51° CONGRESSO NAZIONALE SICP

Società Italiana di Cardiologia Pediatrica
e delle Cardiopatie Congenite

Congiunto con SOCIETÀ ITALIANA DI CHIRURGIA CARDIACA
DOMINIO DELLE CARDIOPATIE CONGENITE

Verona, 27-29 ottobre 2022
Polo Universitario Santa Marta | Silos di Ponente

ABSTRACT BOOK

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DOMINIO DELLE CARDIOPATIE CONGENITE

Venerdì 28 ottobre 2022 - 08.00•09.00
COMUNICAZIONI ORALI
IMAGING

C1

ROLE OF 4D FLOW MAGNETIC RESONANCE FOR RISK STRATIFICATION IN PATIENTS AFTER FONTAN PALLIATION

L. Ait Ali [1], E. Listo [2], E. Valenti [1], S. Salvadori [1], A. Clemente [3], G. Trocchio [2], N. Stagnaro [2], C. Marrone [3], F. Raimondi [4], P. Vitali [3], N. Martini [3], P. Festa [3]

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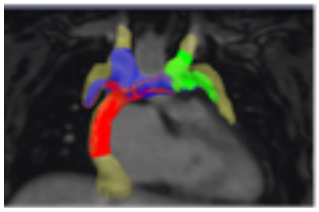
[2] *Ospedale Pediatrico Istituto Giannina Gaslini di Genova, Genova, Italy*
[3] *Fondazione G. Monasterio Regione Toscana (FTGM), Massa-Pisa, Italy*
[4] *Ospedale pediatrico Meyer, Firenze, Italy*

Background: Fontan circulation is one of the more complex hemodynamic patterns in congenital heart disease. Patients after Fontan palliation need extensive hemodynamic evaluation during follow up. 4D Flow Cardiac Magnetic Resonance Imaging (MRI) is emerging as an extremely useful non invasive tool for functional, anatomical and hemodynamic evaluation in patients with congenital heart disease and in particular in this subset of patients.
We sought to evaluate the flow distribution in the pulmonary arteries (PAs) and the contribution of the superior vena cava (SVC) and inferior vena cava (IVC) to pulmonary circulation in patients with functional single-ventricle after Fontan palliation.
Moreover, we aimed to investigate the capability of advanced 4Dflow MRI parameters to predict adverse cardiac outcome.

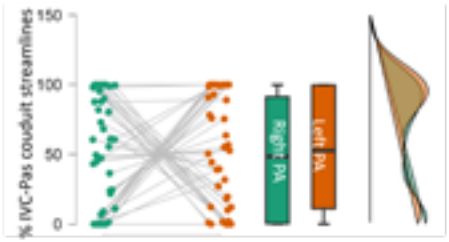
Methods: All consecutive patients after Fontan palliation who underwent cardiac 4Dflow MRI between February 2018 and July 2021 were included in the study. Patients with atrio-pulmonary connection were excluded. Flow was retrospectively quantified in all arterial and venous vessels using a dedicated software (Arterys 4D-flow AI suite). 3D segmentation was manually performed using ITKsnap software. Streamlines quantification and visualization was done using ParaView software (Fig 1A).
Surgical history and clinical data were collected from hospital records. Adverse clinical events were also reported: heart failure, protein-losing enteropathy, New York Heart Association Functional class III or IV.

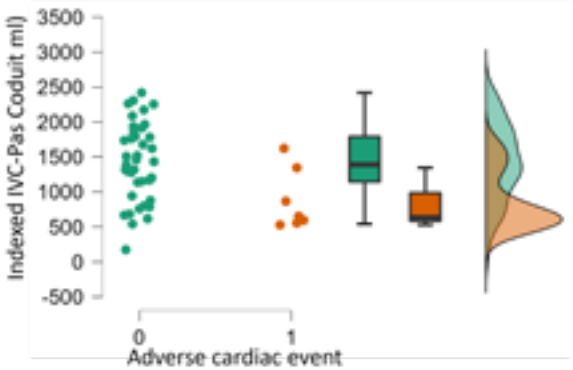
Results: the final study population included 55 patients (37 males, mean age 22 ±10 years. Median age at Fontan was 7,6 (3,6-8) years, in 84 % of patients had an extra-cardiac IVC-Pas conduit . Scan time of 4D flow-MRI was 7.6±1.8 min . Flow distribution was slightly higher in RPA than in LPA (56± 14%). IVC flow accounted for 59%, IQ range: 49;66 % of total systemic venous flow. Streamlines analysis showed that the flow distribution of IVC was slightly predominant for LPA (53± 40%, Fig 1B).
Eight patients experienced an adverse outcome at the time of the study. A logistic regression analysis corrected by age and gender identified as associated with adverse cardiac outcome the following parameters: lower proximal indexed flow, lower mean velocity and higher mean flow jet angle of the IVC-Pas conduit (respectively OR:0.99,p: 0.01, OR:0.87,p: 0.02, OR:1.1,p: 0.04) and higher collaterals flow (OR:1.001,p: 0.004).

Conclusion: 4D flow MRI allows in a single acquisition a comprehensive analysis of Fontan circulation. In our cohort lower flow and velocity and higher jet angle of the IVC-Pas conduit were associated with adverse outcome. Further studies on larger cohorts are needed to understand the usefulness of advanced 4d flow MRI parameters to predict clinical outcome in patients after Fontan palliation.



Stream lines distriubtion





C2

SCREENING OF CORONARY ARTERY ORIGIN BY ECHOCARDIOGRAPHY: DEFINITION OF NORMAL AND DIAGNOSIS OF HIGH TAKE-OFF BY STANDARD ECHOCARDIOGRAPHIC VIEWS IN A HEALTHY PAEDIATRIC POPULATION

P. Marchese [1], C. Viacava [2], N. Assanta [2], G. Corana [2], A. Pizzuto [2], S. Garibaldi [2], E. Franchi [2], G. Santoro [2], M. Cantinotti [2]
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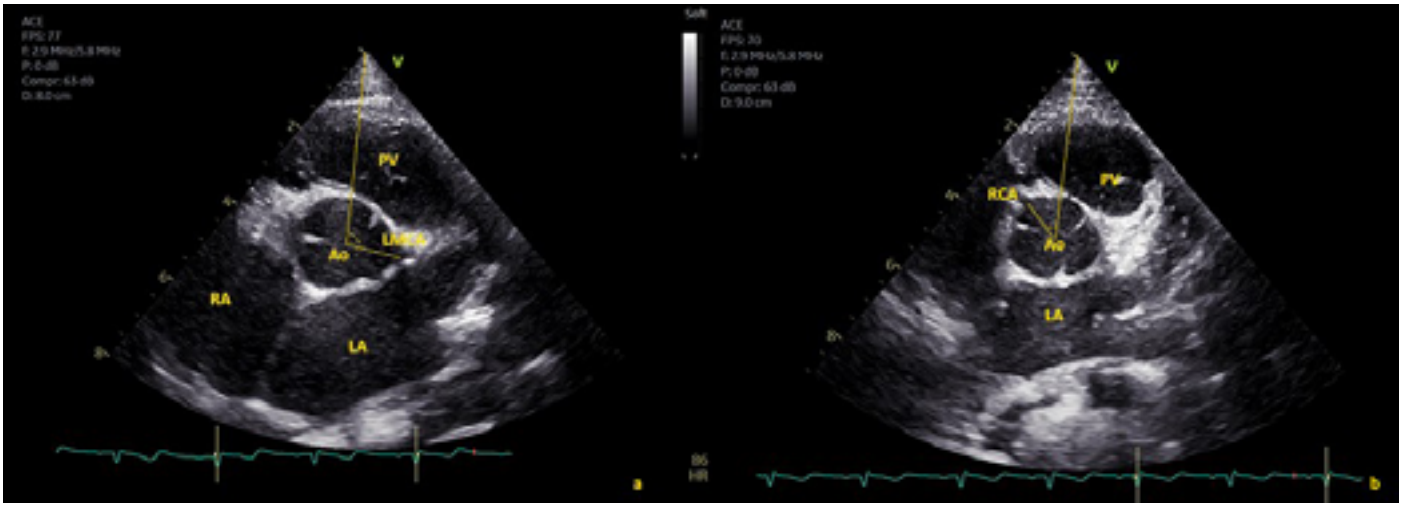
Background: Screening of anomalous coronary artery origin by transthoracic echocardiography is of increasing interest in healthy children undergoing preparticipation screening for sports activities but criteria to define a normal coronary artery origin are poorly defined in young children. This study aims to define the normal origin and angle of emergence of coronary arteries by echocardiography in a large population of healthy children with a special focus on high take-off.

Methods: The distances of the left main and right coronary artery (LMCA and RCA) origins from the aortic annulus were measured in the parasternal long-axis view (LAX). The angle of coronary artery emergence was measured in the parasternal short-axis view (SAX).

Results: 700 healthy subjects (mean age 9.53±5.95 years, range 1 day-17.98 years) were prospectively enrolled. The distance of the RCA and LMCA from the aortic annulus correlated with body surface area, and nomograms (Z-scores) were generated. The RCA origin was below the sino-tubular junction (STJ) in 605 (86.43%), at the STJ in 66 (9.43%), and above the STJ in 29 patients (4.14%). The LMCA origin was below the STJ in 671 (95.86%), at the STJ in 12 (1.71%), and above the STJ in 17 patients (2.43%). For the RCA, an emergence angle <18.5° in SAX could predict a high take-off with a sensitivity of 98.3% and a specificity of 93.1% (AUC 0.998). For the LMCA an emergence angle >119.5° in SAX could predict a high take-off with a sensitivity of 70.6% and specificity of 82.4% (AUC 0.799).

Conclusion: This study establishes nomograms for LMCA and RCA origin in standard transthoracic echocardiographic projections in healthy children. The angle of origin in the short-axis view could predict the height of take-off of coronary arteries.



C3

INTRAVENTRICULAR VORTICES AS A RESERVOIR OF KINETIC ENERGY: CONGENITAL HEART DISEASE'S FLUID DYNAMICS EXPLAINED THROUGH HIGH FRAME RATE BLOOD SPECKLE TRACKING (BST) ECHOCARDIOGRAPHY

P. Marchese [1], N. Assanta [2], E. Franchi [2], C. Viacava [2], G. Corana [2], A. Pizzuto [2], S. Garibaldi [2], G. Santoro [2], M. Cantinotti [2]
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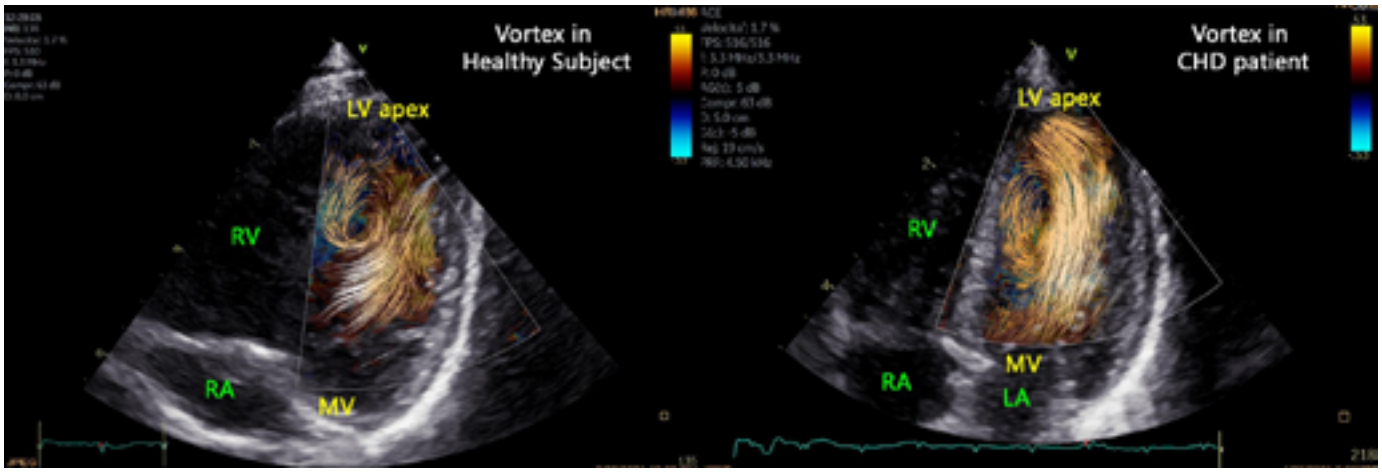
Background: BST (Blood Speckle Tracking) is a newly echocardiographic technique at a high frame rate that allows for observing intraventricular and intravascular fluid dynamics. Ventricular vortices in children have been poorly studied so far, especially in congenital cardiac defects (CHD). This study aims to evaluate the differences in intraventricular vortices in healthy and those with specific CHD.

Methods: Characteristics of LV vortices were analyses based on 4-chamber BST both in healthy and children with CHDs; for each patient, along with the 3 different diastolic phases, the diastolic vortex with the biggest area was used for the final analysis. Multiple linear regression was used to identify factors that independently influence vortex characteristics. all patients had a normal systolic and diastolic function (Ejection Fraction >55%). All measurements were indexed by BSA to overcome differences among patients.

Results: A total of 330 healthy subjects (mean age 6.47±4.12 years, range 0.01-17 years) and 121 children with CHD (mean age 3.49±3.77 years, range 0.01-14.32 years) were prospectively enrolled at a single Center. Feasibility was high, demonstrating a value of 95.6% and 94.3% for healthy controls and CHD patients, respectively. When compared with healthy children, vortices of CHD patients were higher (13.53±8.18 mm/m2 vs 22.55±15.47 mm/m2, p<0.0001), wider (9.97±5.29 mm/m2 vs 17.14±10.6 mm/m2, p<0.0001), with a bigger area (0.8±0.54 cm2/m2 vs 1.47±1.23 cm2/m2, p<0.0001) and occupy a larger surface area in the LV (p<0.0001); no differences in vortex position were founded. When the analysis was conducted on specific CHD, no differences in vortex size were found versus healthy children.

Conclusions: BST echocardiography LV vortex analysis is highly feasible in healthy children and in those with CHD. Left ventricular vortices were bigger in children with CHD compared to healthy counterparts. Since vortices are reservoirs of kinetic energy, our data seems to indicate that children with CHD may have either I) a requirement to produce higher kinetic energy during the diastolic phase to ensure a sufficient cardiac output during the systolic phase, II) a reduced ventricular efficiency/distensibility, and less ability to "compact" the vortex. Future studies are required to confirm these data and to evaluate differences among various CHDs.



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Venerdì 28 ottobre 2022 - 08.00•09.00
COMUNICAZIONI ORALI
IMAGING

C4

HEMODYNAMIC FORCE PATTERN IN A SYSTEMIC RIGHT VENTRICLE: A 4D FLOW MRI STUDY

G. Pasqualin [1], A. Riva [2], F. Sturla [2], F. Bevilacqua [1], M. Lo Rito [1], A. Saracino [1], A. Giamberti [1], A. Redaelli [2], M. Chessa [1], E. Votta [2], M. Lombardi [1]

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Objective: Failure of the systemic right ventricle (SRV) is based on morphological differences between right and left ventricles (RVs and LVs). RV adaptation to systemic afterload includes myocardial hypertrophy and increased circumferential (GCS) over longitudinal global myocardial strain (GLS), with an unknown impact on intracavitary blood flow distribution. The in vivo visualization of intracardiac blood dynamics is nowadays possible through the application of time-resolved three-dimensional phase contrast magnetic resonance (4D Flow MRI). The hemodynamic force (HF) corresponds to the load exerted by the intracavitary blood on the myocardial wall due to pressure gradients. This study aimed to explore the HF pattern in a SRV and to compare it with control LVs and RVs.

Methods: Patients with diagnosis of D-loop transposition of great arteries after atrial switch operation (D-TGA/ASO) and L-loop transposition of great arteries (L-TGA) and a group of age and sex-matched healthy subjects were prospectively enrolled. Cardiovascular magnetic resonance data were acquired on a 1.5-T MAGNETOM Aera (Siemens Healthcare, Erlangen, Germany). Ventricular volumes were calculated on cine steady state free precession sequences (SSFP) and were indexed for the body surface area. Feature tracking analysis was performed on cine-SSFP sequences in long axis for GLS and short axis for GCS. 4D Flow MRI was acquired using a prototype sequence and HFs during the cardiac cycle were computed for SRVs and for the control LVs and RVs. HFs were decomposed in inferior-anterior (HFIA) , septal-lateral (HFSL) and basal-apical (HFBA) components. The ratio between transverse (HFIA and HFSL) and longitudinal (HFBA) HFs (RRMS) was calculated as in Figure 1 for systole and diastole.

Results: We enrolled 12 adults with SRV (6 D-TGA/ASO and 6 L-TGA) and 12 healthy subjects (male to female ratio equal to 5:7 for both groups). Mean age was 41±12 for SRV patients and 42±13 for the control group (p=0.89). SRVs reported comparable end-diastolic volumes (83±18 ml/m2), ejection fraction (60±8%) and GLS (-20.2±3.8%) to control RVs (75±13 ml/m2, p=0.18; 64±5%, p=0.25; -23±5.6%, p=0.21). Differently, SRV mass (55±24 g/m2) and GCS (-18.9±7.8%) were greater than RVs (20±3 g/m2, p<0.001 and -12.4±4, p=0.016) and comparable to control LVs (57±11 g/m2,p=0.7 and -20.2±3.8%, p=0.3).
The SRV systolic RRMS (0.98±0.31) revealed similar values to LVs (0.94±0.27,p=0.78) but lower than RVs (1.32±0.45,p=0.04) (Figure 1). This reflected a significantly increased HFBA with respect to RVs (0.338±0.150 vs. 0.162±0.097,p=0.0025) and similar to LVs (0.462±0.186,p=0.087). Concomitantly, a moderate correlation was demonstrated between SRV systolic HFBA magnitude and GCS (r2=0.47, p=0.013). During diastole, SRVs showed lower HFBA (0.173±0.086) than LVs (0.304±0.104,p=0.0028), revealing a diastolic RRMS (0.74±0.14) comparable to RVs (0.73±0.17,p=0.95) and significantly different from LVs (0.50 ± 0.19,p=0.003) (Figure 1).

Conclusions: Intracavitary HFs in SRVs reveal a partial shift from a RV towards LV pattern. SRV RRMS is similar to LVs during systole, possibly as a result of increased GCS. Inversely, the SRV filling appears to be closely related to the ventricular morphology as suggested by a SRV RRMS comparable to RVs during diastole.

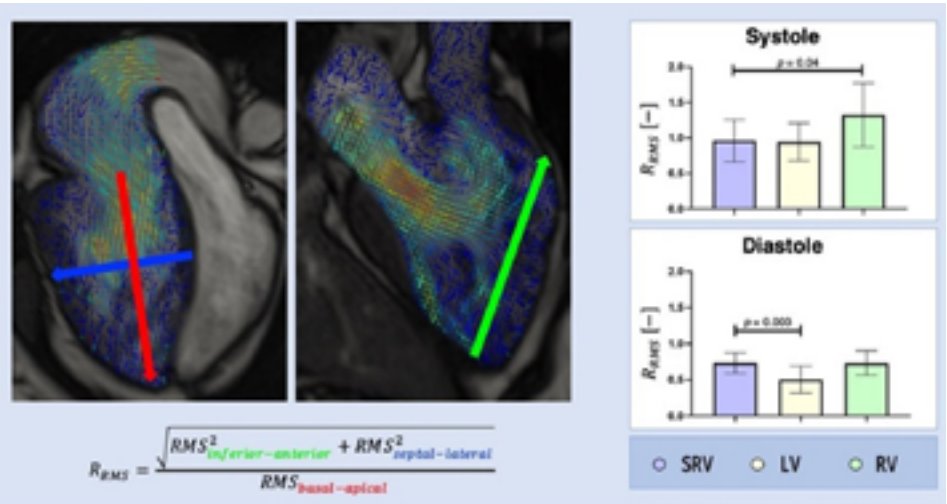

Figure 1. Calculation of the root mean square ratio (R_{RMS}) of HF_{in}, HF_{sl} and HF_{ba} in a SRV and results compared to control LVs and RVs.

C5

ECHOCARDIOGRAPHIC SCORE TO PREDICT NEONATAL SURGERY FOR AORTIC COARCTATION IN NEWBORNS WITH PRENATAL SUSPICION AND PATENT DUCTUS ARTERIOSUS

Y. Bartolacelli [1], D. Palleri [1], A. Balducci [1], L. Ragni [1], T. Hasan [1], G. Egidy Assenza [1], M.E. Mariucci [1], M. Fabi [2], E. Angeli [3], G.D. Gargiulo [3], A. Donti [1]

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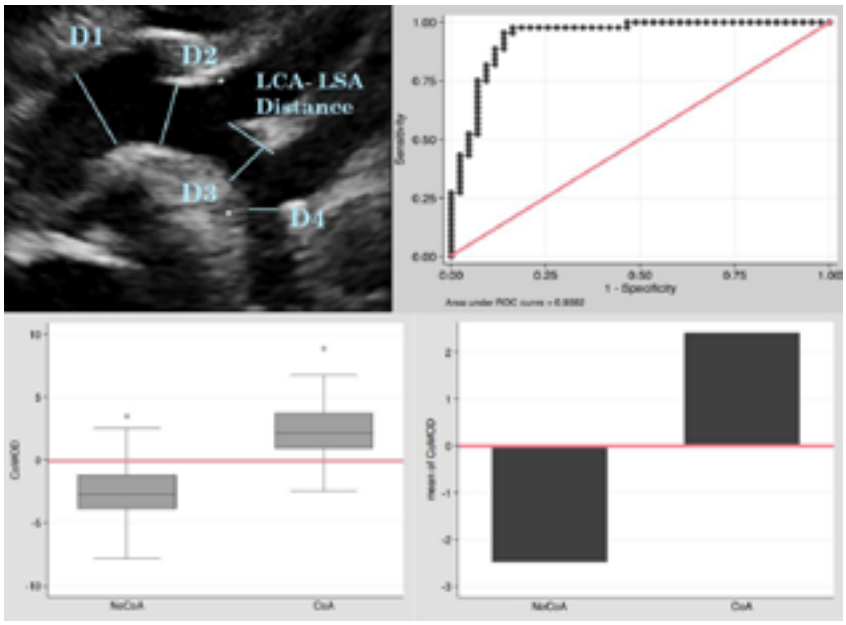
Critical coarctation of the Aorta (CoA) is a ductal-dependent congenital heart disease and it can represent a neonatal emergency requiring surgery in the first weeks of life. Over the time a variety of prenatal echocardiographic scoring systems have been suggested to predict the possible onset of neonatal CoA, but it still remains a challenging diagnosis in prenatal age with a significant number of false-negative and false-positive cases.
The evaluation of upcoming CoA in newborns with prenatal suspicion entails a close echocardiographic monitoring until Arterial Duct closure in a department with paediatric cardiological and surgical expertise. The high rate of false-positive prenatal diagnosis produces unnecessary parental stress and healthcare costs.
The aim of this study was to elaborate an echocardiographic prediction model to be employed at birth, when PDA is still present, in patients suspected of CoA during foetal life, in order to foretell CoA requiring neonatal surgical intervention.

Methods: This retrospective monocentric study included consecutive full-term and late preterm neonates with a prenatal suspicion of CoA born from 01.01.2007 to 31.12.2020. Patients were divided in two groups according to the need of aortic surgery in the first month of life (CoA - NoCoA). All patients underwent a comprehensive transthoracic echocardiographic exam in presence of PDA. Multivariable logistic regression was used to create a coarctation probability model (CoMOD) including isthmal (D4), transverse arch (D3) diameters, distance between left common carotid artery (LCA) and left subclavian artery (LSA), presence/absence of ventricular septal defect (VSD) and bicuspid aortic valve (BAV).

Results: We enrolled 87 neonates (49 male, 56%) with a median gestational age of 272±11.4 days and birth weight 3136±524 grams. Forty-four patients (51%) developed CoA requiring surgical repair. The presence of VSD and BAV was statistically more frequent in CoA group, while Bovine Arch was more frequently observed in NoCoA group. The dimensions D4, D3 and relative z-scores resulted significantly reduced in CoA group and LCA- LSA distance noticeably longer. Our index CoMOD showed an AUC=0.9382, high sensitivity (91%) and specificity (86%) in risk stratification and prediction of CoA in neonates with prenatal suspicion. We classified neonates with CoMOD>0 to be at high risk for surgical correction of CoA in the first month of life, with good PPV (86.9%) and NPV (90.9%).



Conclusions: CoMOD>0 is highly suggestive for the need of early CoA corrective surgery in newborns with prenatal suspicion.

C6

IDENTIFYING PATIENTS AT HIGHER RISK OF SURGICAL COMPLICATIONS IN EBSTEIN'S ANOMALY: ROLE OF THE FUNCTIONAL TRICUSPID ORIFICE ROTATION ANGLE

B. Cavazza [1], G. Pasqualin [2], F. Bevilacqua [4], F. Caldaroni [2], A. Saracino [2], M. Chessa [4], A. Giamberti [2], M. Lombardi [3]
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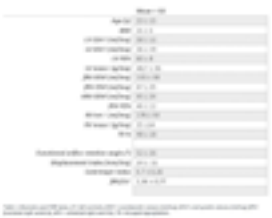
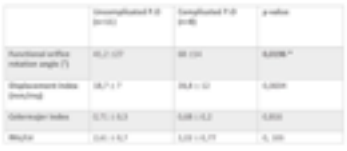
Cardiovascular magnetic resonance (CMR) imaging is the referral method for chamber volume quantification in Ebstein's anomaly (EA) and provides an unrestrictive view of tricuspid valve (TV). In order to improve the therapeutic management of EA, many severity indexes had been proposed over time. Carpentier classification subdivides EA into four groups from type A in which the volume of the true right ventricle is adequate, to type D where there is an almost complete atrialization of the ventricle. Although the progression in surgical TV repair, the indication to intervene is still debated and imaging markers may help in the pre-operative evaluation. The aim of this study was to assess the impact of CMR-derived geometrical severity indexes on the occurrence of post-operative (PO) complications in EA patients undergoing surgery.

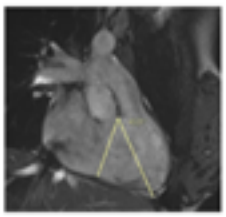
Methods: A retrospective review of EA patients who had surgical repair at Policlinico San Donato from 2015 to 2021 was performed. The pre-operative CMR examination was analyzed.
Ventricular volumes and severity indexes were obtained by steady state free precession (SSFP) sequences. The displacement index was calculated in 4-chamber view as the diastolic distance of the septal TV leaflet from the anterior leaflet of mitral valve indexed for body surface area. The Celermajer index was calculated from a 4-chamber view in end diastole as the sum of the right atrium and atrialized ventricle area over the sum of the functional RV (fRV), the left atrium and left ventricle area. The functional TV orifice rotation angle was measured in 3-chamber RV view between the plane passing through the anatomical TV anulus and the plane passing through the functional TV orifice as in Figure 1 at end-systole. A complicated PO course was defined by the development of moderate or severe TV regurgitation or the onset of sustained bradi- or tachyarryhtmias before the discharge.

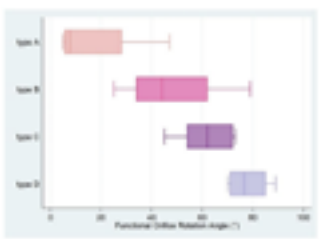
Results: The study included 19 patients (63% males) undergone TV surgical intervention. 15 patients (79%) had Cone Technique , 1 patient (5%) TV repair according to Carpentier, 3 (16%) had TV replacement. Five subjects (26%) received a superior cavo-pulmonary anastomosis.
Biometric and cardiac MRI data are summarized in Table 1. 8 patients (42%) had a complicated PO period: 4 patients developed moderate or severe TV regurgitation (2 of them required re-intervention in the same hospitalization) and 4 patients had sustained arrythmias: bradicardia occurred in 2 (one a complete atrioventricular block requiring pacemaker implantation, one prolonged junctional rhythm) and tachycardia in other 2 patients (one atrial fibrillation and one junctional tachycardia).
The mean value of functional orifice TV rotation angle was significantly higher in the group of patients with complicated PO course 68?15) as compared with the group of patients with regular course (48?27,p=0,0196). No significant difference was shown for the displacement index, Celermajer index and fRV/LV volume between the two groups (table 2)
The functional TV orifice rotation angle showed an increase with the severity of EA according to Carpentier's classification (Figure 2).

Conclusions: Functional TV orifice rotation angle may help to improve the stratification of anatomical severity and to assess the risk of PO complications in EA.









C7

DOUBLE-OUTLET LEFT VENTRICLE: SINGLE-CENTER EXPERIENCE AND LITERATURE REVIEW

M. Lioncino [1], F. Badolato [2], E. Monda [1], S. Maiolo [3], A. De Zorzi [4], G. Calcagni [3], M. Chinali [5], A. Del Pasqua [6], C. Esposito [6], M. Iacomino [6], P. Guccione [6], A. Secinaro [8], L. Galletti [6], A. Carotti [6], M.G. Russo [7], G. Limongelli [1], F. Drago [3], G. Rinelli [3]

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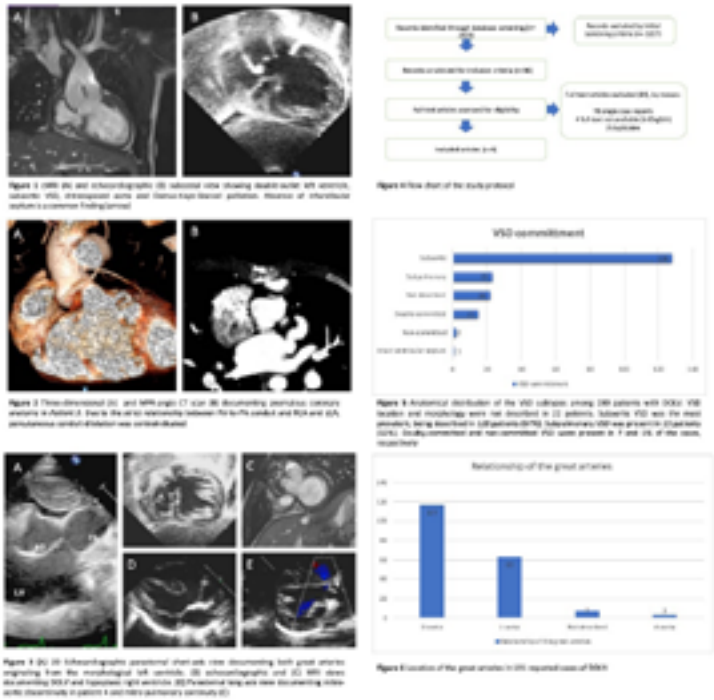
Double-outlet left ventricle (DOLV) is an abnormal ventriculo-arterial connection characterized by origin of both great arteries, or more than 50% of each arterial root, from the morphological left ventricle. The aim of our paper is to describe the anatomic, echocardiographic, and multi-modality imaging characteristics of DOLV and associated malformations, and to assess its surgical outcomes. **Methods:** From 2011 to 2022, we retrospectively reviewed case records, intra-operatory reports and follow-up data of patients diagnosed with DOLV at a paediatric tertiary center. A systematic search was developed in MEDLINE, EMBASE and Web of Science databases, to identify original reports between January 1, 1975 and May 30, 2022, assessing the morphology and surgical outcomes of DOLV. Retrospective cohort studies, cross-sectional and case series were included in the analysis. Single case reports and reviews were excluded. **Results:** At our center, four cases of DOLV were identified. Patient 1 was diagnosed with (S,D,D) DOLV and hypoplastic right ventricle. The aorta overrode a large, doubly-committed VSD with absence of infundibular septum. A tenuous mitro-aortic discontinuity and a well-developed subpulmonary conus were present. Associated abnormalities included crossed pulmonary arteries and two adjacent, side-by-side coronary ostia, located in the anterior facing sinus, which gave origin to the left anterior descending (LAD) and the right coronary artery (RCA). Left circumflex artery (LCx) had a retro-aortic course and originated from the RCA. After pulmonary artery banding, Damus-Kaye-Stansel and Glenn intervention were proposed as first-stage of univentricular palliation. Patient 2 and 3 were diagnosed with (S,D,D) DOLV, subaortic VSD and pulmonary stenosis. Patient 2 underwent Rastelli operation and no anatomic detail was available. Patient 3 showed absence of the infundibular septum and mitro-pulmonary continuity, whereas subaortic conus was well developed. Anomalous origin of the LCx, originating from the posterior facing sinus with retro-aortic course was present. Rastelli procedure was performed to reconstruct right ventricular outflow tract. LCA and RCA were respectively caudal to subvalvular and supravalvular segments of the RV-to-PA conduit. After a 6-years follow-up, severe stenosis of the RV-to-PA conduit was present, nevertheless percutaneous conduit dilatation was contraindicated, due to coronary abnormality, and an aortic homograft was implanted. Patient 4 was diagnosed with (S,D,L) DOLV with subaortic VSD and mitro-pulmonary fibrous continuity. A large subaortic conus was present. Reparation à l'etage ventriculaire was performed to reconstruct RVOT. Follow-up MRI at 8 years showed severe pulmonary artery regurgitation with mild RV dilatation (indexed volume 99mL/m2) and normal RV ejection fraction (54 %) Systematic review:Through our systematic research strategy we scrutinized 96 records for inclusion criteria (Figure 4). After systematic evaluation, a total of 9 reports fulfilled eligibility criteria and were included in our study. Morphological findings and surgical outcomes are summarized in Table 1. Among 191 cases of DOLV included, the most common subtypes of VSD were subaortic (128/191, 67%), subpulmonary (23/191, 12%) or doubly committed (15/191, 7%) (Figure 5). d-transposition of the aorta was present in 117/191 (61%) cases, whereas l-transposition was reported in 63/191 (32%) (Figure 6)



C8

AUGMENTED REALITY IN TEACHING CONGENITAL HEART DEFECT

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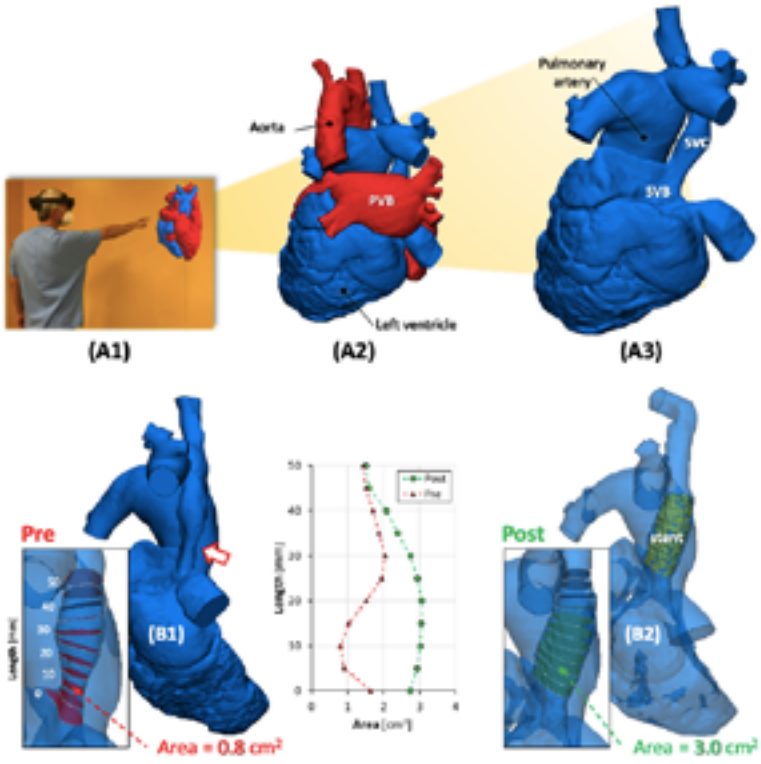
Introduction: Congenital heart disease (CHD) is often comprised of complex three-dimensional (3D) anatomy that must be well understood to assess the pathophysiological consequences and guide therapy. Thus, detailed cardiac imaging for early detection and planning of interventional and/or surgical treatment is paramount. Advanced technologies have revolutionized diagnostic and therapeutic practice in CHD, thus playing an increasing role in its management. Traditional reliance on standard imaging modalities including echocardiography, cardiac computed tomography (CT) and magnetic resonance imaging (MRI) has been augmented by the use of recent technologies such as 3D printing, virtual reality, augmented reality, computational modelling, and artificial intelligence because of insufficient information available with these standard imaging techniques. We hereby investigated the use of augmented reality in teaching CHD.

Study **Aims:** The aim of our study was to investigate whether the use of augmented reality would result in better understanding of the anatomy and transcatheter treatment of a specific CHD named sinus venosus atrial septal defect (SV-ASD).

Methods: 60 medical students were randomly subdivided into three groups (A, B, C) and received the same lecture. The iconographic material was available in different modalities: group A received 2D and 3D images; group B were allowed to access holographic imaging projected on a standard screen; group C were able to interact with the hologram wearing Microsoft HoloLens devices. At the end the of the lecture each group was administered a test. The first part test was composed of six questions testing the comprehension of the pathology in general terms (epidemiology, physiopathology, diagnosis and treatment), the second part of the test was composed of 4 questions and evaluated the capability of the students to understand the relationship among
right pulmonary veins (RVP), superior vena cava (SVC) and sinus venosus ASD and the possibility to close the defect with a covered stent.

Results: The results (figure 1) showed that students from group B and group C performed overall better on the test and demonstrated a better understanding of the second part of the test.

Conclusion: To the best of our knowledge this is the first attempt to evaluate the usefulness of holographic imaging in teaching CHD to medical students. The students who were allowed to visualize holograms either on the screen or wearing HoloLens, understood better the spatial relationships among the sinus venous ASD, SVC, RPV and transcatheter closure technique.



Venerdì 28 ottobre 2022 - 08.00•09.00
COMUNICAZIONI ORALI
CARDIOMIOPATIE

C9

PROGNOSTIC FACTORS IN HYPERTROPHIC CARDIOMYOPATHY IN CHILDREN: AN MRI BASED STUDY

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Background: Clinical and prognostic role of cardiac magnetic resonance (CMR) in adult population with hypertrophic cardiomyopathy (HCM) have been largely assessed.
We sought to investigate the role of CMR for predicting cardiovascular events in children with HCM.

Methods: CMR was performed in 116 patients with HCM (37 sarcomeric mutations, 31 other mutations, mean age 10.4 ± 4.3 yrs). CMR protocol included cine imaging for evaluation of morphology and function and late gadolinium enhancement (LGE). Hard cardiac events (sustained VT, resuscitated cardiac arrest, sudden cardiac death, end-stage heart failure, heart transplant and appropriate ICD intervention) were recorded through a median follow-up of 4 (1-7) years.

Results: During follow-up 21 heart cardiac events occurred. At maximal-rank statistic the optimal cut-point for LGE extent for predicting events was ?2%. Syncope, non-sustained ventricular tachycardia (NSVT) and LGE extent ?2% were independent predictors of events. At Harrel's C statistic combination of LGE extent ?2% and syncope was the strongest model for predicting events. HR of patients with LGE extent ?2% and no history of syncope was 3.6 (1.1-12.2) that increased to 37.6 (5.4-161) in those with LGE extent ?2% and syncope. The median time dependent AUC of LGE extent (0.88, 95% CI 0.86-0.89) was significantly higher than that of syncope (0.63, 95% CI 0.61-0.66, p<0.0001) and NSVT (0.52, 95% CI 0.50-0.53, p<0.0001).

Conclusions: In children with HCM, LGE and syncope were independent predictors of hard cardiac events at follow-up.

Venerdì 28 ottobre 2022 - 08.00•09.00
COMUNICAZIONI ORALI
CARDIOMIOPATIE

C10

PULMONARY ARTERY BANDING FOR REGENERATION OF DILATED CARDIOMYOPATHY IN YOUNG CHILDREN: A SINGLE CENTRE REPORT

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Background: left ventricular dilated cardiomyopathy (LV-DCM) is characterized by a dilation and dysfunction of the left ventricular chamber. The incidence of LV-DCM has been reported 0,57-1,13/100.000 and remains a leading cause of cardiac death in children. Approximately 40% of children die or need cardiac transplantation in the first 5 years after diagnosis. The reported combined medical and surgical approach represents a novel therapeutic strategy with the aim to restore ventricular-ventricular interaction (VVI) and induce myocyte recovery.

Patients and **Methods:** 41 patients below the age of 2 years with an end-stage DCM and Ross-functional-status III-IV were referred to our centre for heart transplantation and received, as additional measure, a surgical PAB. With the hypothesis, that a reversible PAB, along with a child-specific anticongestive therapy, can avoid ore defer a cardiac transplantation, the efficiency of this therapy was retrospectively analysed with the use of clinical, laboratory-chemical and imaging data.

Results: all patients survived surgical intervention to discharge. Follow-up after 74±29 months (n=30), showed an improvement of clinical functional status and a significant improvement, up to normalization, of the analysed data (expressed in mean ± standard deviation): left ventricular ejection fraction (LV-EF) increased from 18±8% to 57±7%, left ventricular end diastolic diameter (LVEDD) decreased from 46±7 mm to 39±5 mm (z-score from
+6,9±1,3 to +0,9±0,8), BNP decreased from 3089±2642 ng/l to 64±82 ng/l. 23 patients have undergone catheter-based de-banding after a median of 10,5 months. 8 patients underwent HTX. 3 patients died, 2 after complete debanding, one 12 years later.
After 1 year left ventricular function normalized in 81% of patients, who were free from transplantation. Freedom from death and transplantation was 81% 1 year after presentation and 76% at 2 and 5 years.

Conclusion: VVI plays a key role in the human heart. The PAB in LV-DCM restores the LV-synchrony by shifting the ventricular septum leftward and thereby reducing left ventricular end-diastolic volume and pressure. Restored VVI and LV-geometry, the PAB-induced pressure overload of the right ventricle and the specific anticongestive therapy, stimulate all together the endogenous regenerative ability of the human heart, leading to recovery of LV-function. Yet further questions concerning cellular mechanism of action, optimal timepoint for intervention and identification of subgroups with risk for treatment failure still need to be examined.

C11

CARDIAC OUTCOMES AFTER IMMUNIZATION WITH MRNA-BASED COVID-19 VACCINES IN ADOLESCENTS

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Background and Objectives: Vaccination against COVID-19 has showed an undeniable public health benefit, but it also could result in potential adverse events. Acute pericarditis/myocarditis is a rare complication of the mRNA-based vaccines and although mostly self-limiting, long-term sequelae remain unclear.

Methods: From September 2021 to February 2022 we enrolled patients admitted at Bambino Gesù Pediatric Hospital in Rome with acute pericarditis/myocarditis with onset after mRNA based COVID-19 vaccines.

Results: We observed 13 patients, eleven male and 2 female adolescent (median age 15 ± 1,6 years) affected by acute pericarditis/myocarditis with onset 1 to 48 days after COVID-19 mRNA vaccination (11 patients after Comirnaty and 2 after Spikevax). All patients presented with chest pain. Diagnosis was made based on clinical presentation, increased levels of troponin-T (mean peak, 641 ± 471 pg/mL) and NT-proBNP and pathognomonic electrocardiographic (ECG) and echocardiographic abnormalities. Seven (54%) of them presented with myopericarditis, three (23%) showed myocarditis and three (23%) had sign of pericarditis. Cardiovascular magnetic resonance (CRM) was preformed in 5 patients among those who had myocarditis/myopericarditis and findings were consistent with myocarditis in all patients, including early gadolinium enhancement and late gadolinium enhancement (LGE).
Alternative virological causes were excluded.
All patients were treated with ibuprofen and were discharged in stable condition with resolved symptoms after few days (mean length of hospital stay, 9 days). Two patient repeated CRM after three months that showed persistent, although decreased, LGE (Figure 1). At this time point the other cardiological findings (ECG, echocardiography, cardiac enzyme, ECG holter and stress testing) were normal.

Conclusions: In our case series all patients showed clinical recovery. However, the evidence of persistent CRM lesions highlights the need for a close and standardized follow-up for all patients who present sign of myocarditis/myopericarditis. Further studies in this population are needed to investigate the personal susceptibility to develop complications after COVID-19 mRNA vaccination.

C12

GENETIC CAUSES OF CARDIOMYOPATHIES IN CHILDREN

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Background: Paediatric cardiomyopathies, including Hypertrophic Cardiomyopathy (HCM), Dilated Cardiomyopathy (DCM), Restrictive Cardiomyopathy (RCM), Left Ventricular Non Compaction (LVNC) and Arrhythmogenic Cardiomyopathy (AC) represent an important cause of childhood heart failure. Despite the increased diagnostic yield of Next Generation Sequencing (NGS), the genetic background of these conditions is only partially resolved, because of their rarity and heterogeneity. However, the assessment of aetiology is of paramount importance in order to define prognosis and, increasingly, therapeutic choices. Moreover, the recognition of genetic causes is the basis for cascade screening in the family.

Aims: This study aimed to identify the genetic causes of cardiomyopathies in children evaluated and followed at the Paediatric Cardiology Unit
Methods
A validated NGS target panel was performed in a cohort of 76 paediatric patients (36 HCM, 24 CMD, 3 RCM, 2 LVNC, 10 AC and one with sudden death) after genetic counselling offered to parents. Furthermore, 9 patients with negative panel (4 HCM, 3 DCM, 2 RCM) for Whole Exome Sequencing (WES). We also studied 34 probands' relatives.

Results: By target panel analysis we identified a pathogenic or likely pathogenic variant in 32/76 patients (42%). Most genotyped patients had a sarcomeric aetiology and MYH7 (11/32, ~34%), MYBPC3 (7/32, ~21%), TNNT2 (3/32, ~9%) resulted the most mutated genes. 11/32 (34%) patients had a complex genotype. Moreover, we identified five syndromic cases: three patients with Noonan Syndrome (one with SOS1 variant and two with PTPN11 variants), one with Danon Disease (with LAMP2 variant) and a child with Alström Syndrome (due to ALMS1 variants). In another patient we found the rare coexistence of two independent autosomal dominant monogenic diseases (Osteogenesis Imperfecta and HCM) due respectively to a COLIA1 variant and a MYH7 variant. By WES analysis we identified a de novo variant in FLNC gene in a child with RCM, a Noonan Syndrome due to biallelic LZTR1 variants, two PLEC1 variants in a child with DCM and Epidermolysis bullosa, and a NKX2-5 variant in a case with RCM.

Conclusions: A definitive molecular genetic diagnosis could be obtained in a substantial proportion of children, including selected individuals with a previously unknown cause of their cardiomyopathy. The genetic test results had direct implications for clinical management, to suggest therapeutic interventions and sometimes to predict prognosis. Moreover, genetic testing performed in family members resulted very important for clinical follow-up and to establish the recurrence risk for subsequent pregnancies.

C13

THE ROLE OF ENDOMYOCARDIAL BIOPSY IN CHILDREN WITH LEFT VENTRICULAR DYSFUNCTION

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Introduction: Endomyocardial biopsy (EMB) is a well-known diagnostic tool for the investigation and treatment of myocardial diseases and so far, remains the gold standard for the diagnosis of myocarditis. Due to its invasivity with a complication rate ranging from 1% to 15%, its role in the diagnostic work-up of pediatric heart failure is not well established.

Aim of this study is to define the clinical role and the safety of EMB in children presenting with severe left ventricular dysfunction.

Methods: We performed a retrospective review of the data of children who underwent endomyocardial biopsy in our Center from 2011 to 2021. Patients with heart transplantation were excluded from this analysis.
Results: During the study period 29 children underwent 30 EMBs for systolic LV dysfunction. Mean age was 7.3 year, 5 children (17%) were < 1 years old and 2 of them had a body weight < 8 Kg. Clinical presentation at admission was: heart failure in 24 pts (83%), AV block in 1, pericarditis in 2, abdominal pain in 1, one child presented with Covid infection and an isolated right ventricular dysfunction was present in 1. Seventeen patients (57%) had a diagnosis of myocarditis, 9 patients received a diagnosis of primary dilated cardiomyopathy, 1 child had Carnitine deficiency, 1 a laminopathy and the patient whose presentation was an isolated right ventricular dysfunction had a Uhl disease (post-mortem finding). Viral PCR on myocardial specimen was positive in 12/17 patients (70%) and viral myocarditis was confirmed. 10 patients had PVB19 viral genome copies (83%), 1 patient had HHV6 viral genome copies and 1 patient had influenza A viral genome copies. EMB results led to a change in medical therapy in 12 patients (40%). Treatment changes were: antiviral therapy in 5 patients with viral myocarditis, immunoglobulin therapy in 6 patients, carnitine supplementation in 1 patient and immunosuppressive therapy in 1 patient with virus negative myocarditis. Regarding complications, cardiac perforation occurred in 3 patients (10%). Weighting 20 Kg, 10,5 Kg and 6 Kg. All these patients survived except one.

Conclusions: In the diagnostic work up of children with systolic heart failure, EMB-derived information are still essential prerequisites to establish an accurate diagnosis of myocarditis and a successful management of these patients. In our experience EMB results changed the clinical management of children with cardiomyopathy in 40% of the cases with a 10% of major adverse advents.

C14

NATURAL HISTORY OF HYPERTROPHIC CARDIOMYOPATHY IN NOONAN SYNDROME WITH MULTIPLE LENTIGINES

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Importance: The natural history and outcomes of hypertrophic cardiomyopathy (HCM) in individuals with Noonan syndrome with multiple lentigines (NSML) have not been well defined.

Objective: To examine clinical features, genotypic results, and outcomes of consecutive patients with NSML and HCM.
Design, Setting, and Participants: A retrospective, longitudinal multicenter cohort of consecutive children and adults with a genetic diagnosis of NSML and HCM between 2002 and 2019 was assembled. Clinical data were collected, including demographics, family history, symptoms, medical therapy, resting electrocardiography (ECG), transthoracic echocardiography, and genetic testing. Data were collected from baseline evaluation to last clinical review.

Main Outcomes and Measures: We defined a priori three different patterns of left ventricular remodeling during follow-up: 1. an increase ?15% of the maximal left ventricular wall thickness (MLVWT), both in mm and z-score (progression); 2. a reduction ?15% of the MLVWT, both in mm and z-score (absolute regression); 3. a reduction ?15% of the MLVWT z-score with a stable MLVWT in mm (relative regression). The primary study endpoint was a composite of HCM-related outcomes including cardiovascular death, heart transplantation, and surgical myectomy.

Results: The cohort comprised 43 patients with NSML and HCM, with a median age at diagnosis of 3.5 (interquartile range [IQR], 0.2-12.2) years. Freedom from composite HCM-related outcomes was 90.6% (95% confidence interval [CI] 81.8-99.4%) 1 year after presentation and 79.6% (95% CI 64.7-94.5%) at 5 years. On multivariate analysis, only MLVWT z-score > 13.4 at diagnosis predicted HCM-related outcomes (hazard ratio, 13.4 [95% CI 1.38-130.19], p-value 0.025). During a median follow-up of 3.7 years (IQR 2.2-7.9), relative regression was the most common type of left ventricular remodeling (n = 12, 36%), followed by progression (n = 9, 27%), and absolute regression (n = 7, 21%).

Conclusions and Relevance: These findings provide insights into the natural history of left ventricular hypertrophy, and can help inform clinicians regarding risk stratification and clinical outcomes in patients with NSML and HCM.

C15

CARDIAC INVOLVEMENT IN CLASSICAL ORGANIC ACIDURIAS: CLINICAL PROFILE AND OUTCOME IN A LARGE PAEDIATRIC COHORT

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Introduction: Cardiac involvement (CI) have been reported in several classical Organic acidurias (OAs) and can contribute to disability and cause premature death. The disorder most strongly associated with cardiac sequelae is Propionic Acidemia (PA). Different types of cardiomyopathies (CMPs) have been described. Despite the improvement in the therapeutic treatment of these patients the overall outcome remains unsatisfying with long-term complications. Liver transplantation (LT) is recognized as a treatment option for selected patients.

Patients and **Methods:** We retrospectively analyzed clinical, biochemical, and molecular records of 258 patients (pts) diagnosed with inborn errors of metabolism (IEM), referred to the Unit of Metabolic Diseases at Meyer Children Hospital from 2000 to August 2021. We have selected individuals with classical OAs for a total of 60 pts.

Results: Of the 60 pts with OAs 27 were males (45%) and 33 females (55%); specifically, 25 individuals received diagnosis of MMA, 20 pts received diagnosis of PA, 9 pts received diagnosis of Isovaleric Acidemia (IVA) and 6 pts received diagnosis of Maple syrup urine disease (MSUD). Median age at diagnosis of the cohort was 364,32 days [IQ 0;15 years].
CI was present in 23 patients (17 pts with PA and 6 pts with MMA) with a mean age of onset of 7.6 years (range 0,1-17.5).
CMP was diagnosed in 18 pts; DCM and Long Qtc were the most recurrent phenotype. DCM was observed in 16 pts (26%). Isolated Long QTc was documented in 4 pts.
Cardioactive treatment was prescribed in 16 (80%) of the 20 patients with CMP.
In 7 patients, despite medical treatment, we observed a progressive reduction of the ejection fraction (EF). In these patients we have proposed LT as a therapeutic option.
Over a median observation period of 5 years, covering 11/60 patients presented a major cardiovascular event or equivalent (MACEs) and death from all causes. 10 of these patients had also CI. Specifically, 5/20 experienced a HF-related event, and 1 HF-death. The rate of occurrence of MACEs was 55% in PA with CMP, 35%in MMA with CMP and 23 % in MMA without CMP. Annual rate of MACEs was 10,1 % for PA with CM, 7,2 % for MMA with CM and 3,9% for MMA without CM. 5/60 patients underwent LT, 3 of these presented also severe heart failure; other 4 are currenty on the waiting transplantation list.

Conclusions: Phenotypic presentation of OAs may be extremely variable and complex.
DCM is also a common finding in some groups of OAs and is associated with poor prognosis.
There are no specific predictors of myocardial damage, therefore affected pts should be enrolled in cardiologic follow-up protocols, despite the absence of cardiovascular symptoms.
Further prospective studies are needed to understand molecular targets involved in cardiac dysfunction in order to start early specific treatment, implement effective treatment strategies and also to evaluate LT as a therapeutic option in the management of CMP.

C16

INCIDENCE OF ACUTE MYOCARDITIS IN THE PEDIATRIC POPULATION IN THE COVID-19 ERA: EXPERIENCE IN A SECOND-LEVEL CENTER

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Aim of The Study: SARS-CoV-2 infection can be associated with myocarditis during acute SARS-CoV-2 infection or as a consequence of COVID-related multisystem inflammatory syndrome of the child (MIS-C). SARS-CoV-2 vaccination has also been related to some cases of myocarditis, especially in adolescent and young adult males. In May 2022, in Italy 1,705,255 pediatric patients aged 5-19 years (20.6% of the pediatric population of the same age) recovered from COVID-19, while children and adolescents vaccinated for SARS-CoV-2 reached the number of 5,131,955 (coverage of 83.7% between 12-19 years). The aim of this study is to evaluate the incidence of acute myocarditis in the COVID-19 era.

Methods: Using our institutional clinical database, we retrospectively analyzed admissions to the Department of Pediatrics at our secondary-level Hospital, between December 2020 (time of introduction of SARS-CoV-2 vaccine in subjects ?16-years-aged) and May 2022, to investigate the incidence of myocarditis and the possible association of the disease with recent infection or vaccination.

Results: Our series refers to 10 cases of myocarditis (10/776 = 1.2% of pediatric hospitalizations), of which 5 cases (50%) of cardiac involvement associated to MIS-C and 5 cases (50%) of primary acute myocarditis. None of the patients admitted for acute SARS-CoV-2 infection showed signs of myocarditis (median age 2 years [range 1 month-16 years]). We observed 6 hospitalizations for MIS-C (6/776 = 0.8% of hospitalizations; median age 8.5 years [5-10], male 50%) of which 5 cases had cardiac involvement (5/6 = 83% of MIS-C; median age 9 years [5-10]). All cases of MIS-C were treated with intravenous immunoglobulin, methylprednisone and low-dose aspirin. Echocardiography showed moderate systo-diastolic dysfunction in two patients (2/5 = 40% of MIS-C with cardiac involvement) with LVEF 42% and 46% respectively, which resolved shortly after starting therapy. Of the 5 admissions for primary myocarditis (5/776 = 0.6% of hospitalizations; median age 15 years [12-17], male sex 100%), 2 cases were observed between 1 and 28 days after the second dose or booster dose of SARS-CoV-2 vaccine, 1 case was observed during convalescence from COVID-19 (first week after negative swab), the remaining 2 cases were not related to recent SARS-CoV-2 infection or vaccination. In all cases of primary myocarditis, normal cardiac function was seen within 3 months from the onset of symptoms, with persistence of late-gadolinium enhancement (at cardiac MRI performed at 4-6 months of follow-up) in only 1 case unrelated to recent SARS-CoV-2 infection or vaccination.

Conclusions: The occurrence of myocarditis after SARS-CoV-2 vaccination was confirmed a rare event (0.2% of pediatric hospitalizations) that mainly involves adolescent male subjects, its incidence was not greater than cases of myocarditis non-correlated to vaccination, and in our series it did not leave sequelae. Overall, the pediatric incidence of myocarditis related to SARS-CoV-2 infection was higher than the incidence of myocarditis after SARS-CoV-2 vaccination (0.6% vs 0.2%). In light of recent data and the current incomplete knowing of pathophysiology of both MIS-C and the possible effects of SARS-CoV-2 vaccination, follow-up of these patients should be continued to evaluate any unexpected long-term effects.

C17

THE INDISPENSABLE HEPATIC ASSESSMENT OF FONTAN PATIENTS: OUR EXPERIENCE

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Background: Fontan-associated liver disease (FALD) is an emerging condition in patients who have undergone surgical correction of univentricular congenital heart disease. Little is known about the epidemiology of FALD, including risk factors for end-organ failure or hepatocellular carcinoma nor are present consensus or guidelines on surveillance. Furthermore, there is a need to understand the role of heart versus combined heart-liver transplantation in this population.

Aim: The purpose of our study is to describe the incidence of FALD in patients undergoing Fontan surgery attending our institution

Methods: From 2017 to 2022, we analyzed 198 adult patients with Fontan circulation. All patients underwent a detailed hepatic examination including laboratory analysis, liver ultrasound and abdominal MRI with hepatospecific contrast agent. Hemodynamic assessment was performed using echocardiography, cardiopulmonary exercise capacity testing and cardiac catheterization.

Results: The mean (±SD) age of the population was 27 ± 4.5 years, with mean duration of the Fontan circulation of 22 ± 2.5 years. All patients underwent cardiac ultrasound annually. Liver parenchyma was unstructured in all patients. 120 patients (61%) were free of nodular lesions hepatics. 78 patients (39%) had hepatic lesions: 48 patients (24%) had nodules < 1 cm and 20 patients (15%) had nodules > 1 cm. Six patients (3%) presented with hepatocarcinoma (HCC). Five out of 6 patients with HCC (83%) presented with elevated alpha-fetoprotein values. In 8 patients (10%) who presented hepatic nodules, the lesion was evident only on abdominal MRI.

Conclusions: We found no hemodynamic risk factors predisposing to the onset of FALD other than the duration of Fontan, which may be considered the major risk factor for the development of FALD. The findings support the use of well-defined surveillance protocols to identify potential precipitants of FALD before liver disease becomes irreversible. The approach should be multifactorial, and the use of hepatospecific abdominal MRI should be favored. In our experience especially in patients who are obese, have poor ultrasound windows, and have a Fontan duration of more than 15 years.

C18

SERUM PROTEOME ANALYSIS IN CHILDREN AND YOUNG ADULT WITH PULMONARY HYPERTENSION: AN OBSERVATIONAL COHORT STUDY

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Background: Pulmonary arterial hypertension (PAH) in children is a complex and heterogenous condition, mostly complicating congenital heart disease (CHD) and carrying an increased risk of mortality. Clinical management is difficult due to the scarcity of data to have an accurate risk assessment. We aim to identify protein proteomic features that correlate with events (life threatening arrhythmias, HF admissions) and might impact on clinical course in children.

Methods: In this observational cohort study, we enrolled children with idiopathic and CHD-PAH in referred to our Institute. Blood samples were collected during routine clinical visits, together with clinical status, biochemical and no-invasive tests. Outcomes were considered deaths or transplant, rehospitalization for heart failure and arrhythmias during one year of follow up.
We performed a proteomic analysis (SWATH) on serum aiming at the identification of a panel of prognostic proteins, evaluating their differential regulation in patients grouped by diagnosis. A number of potentially prognostic proteins were then confirmed by statistical analysis: ANOVA, ROC and Kaplan-Meier curves were used.

Results: We enrolled 34 children (mean age: years 13.8±10.7, median: 10) with idiopathic and CHD-PAH. 347 potentially quantifiable proteins were identify and trough SWATH analysis we selected in these patients a panel of nine highly expressed proteins. On the potentially prognostic proteins, a further statistic validation was performed using ANOVA test and subsequently by ROC and Kaplan-Meier curve. Proteins validated in all analysis were haptgloblin (HPT), thyroxine-binding globulin (TGHB) and transgelin-2 (TAGL2). HPT significantly correlates with an increased risk of admissions for HF. TGHB and TAGL2 significantly correlate with an increased risk of life threatening arrhythmias and particularly with a high risk of arrhythmias in CHD group.

Conclusion: HPT, TGHB and TAGL2 identified in patients with PAH correlates with a high risk of arrhythmia and HF hospitalization and might have a use in clinical management and the evaluation of new therapies in children.

Venerdì 28 ottobre 2022 - 09.00•10.00
COMUNICAZIONI ORALI
FOLLOW UP

C19

MID AND LONG TERM ATRIO-VENTRICULAR MECHANICS IN CHILDREN AFTER RECOVERY FROM ASYMPTOMATIC OR MILDLY SYMPTOMATIC SARS-COV-2 INFECTION

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Background: Clinical manifestations of children's coronavirus disease-2019 (COVID-19) were initially considered less severe compared with adult patients. However, there is now increasing evidence of a "long-tail" of COVID-19 related symptoms lasting for several months after recovery from the acute infection. Long COVID-19-related symptoms and mechanisms are poorly characterized and understood, with several phenotypes reported, often driven by long-term tissue damage (such as lung, heart and brain) and pathological inflammation due to viral persistence and/or immune deregulation.

Purpose: The objective of this study was to evaluate atrio-ventricular mechanics, by means of two-dimensional speckle-tracking echocardiography, in previously healthy children recovered from asymptomatic or mildly symptomatic severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) infection in a long-term follow-up.

Methods: We analysed a cohort of 157 paediatric patients, mean age 7 ± 4 years, who had a confirmed diagnosis of SARS-CoV-2 infection and were asymptomatic or mildly symptomatic for COVID-19. Patients underwent standard transthoracic echocardiogram and speckle tracking echocardiographic study 148 ± 68 days after diagnosis. One hundred seven age, sex, and body surface area comparable healthy subjects were used as control group.

Results: Left ventricular ejection fraction was within normal limits in postCOVID-19 cases and CTRL with no significant differences between the two groups (postCOVID-19: 65.6 ± 4% vs CTRL: 65.0 ± 5%, $p = 0.182$).
Left ventricular (LV) global longitudinal strain (postCOVID-19: -20.5 ± 2.9 %; CTRL: -21.8 ± 1.7 %; $p < 0.001$) was significantly reduced in cases compared with CTRLs. An amount of 11 (7%) postCOVID-19 cases showed impaired GLS values < -17% and 95 subjects (60%) presented with a strain lower than -16% in more than 2 segments. These subjects did not show any difference regarding symptoms or serological findings. Moreover, GLS was significantly reduced in children with disease's onset during the second wave of COVID-19 pandemic, compared with those during the first wave (second wave: -20.2 ± 2.6%; first wave: -21.2 ± 3.4 %; $p = 0.048$). Finally, peak left atrial systolic strain was within the normal range in the postCOVID-19 group with no significant differences compared to CTRL (postCOVID-19: 49.1 ± 12%; CTRL: 49.5 ± 18%).

Conclusions: SARS-CoV-2 infection may affect left ventricular deformation in children despite an asymptomatic or only mildly symptomatic acute illness. Our data show an amount of 60% of children, recovering from asymptomatic or mildly symptomatic COVID-19, with still mild subclinical systolic cardiac impairment in the mid- and long-term follow-up after the infection. This subtle impairment was seen to be worse in children recovering from the second wave of COVID-19 compared to the first one. A follow-up is needed to verify the reversibility of these alterations and their impact on long-term outcomes.

C20

NON-INVASIVE MYOCARDIAL WORK INDICES IN PATIENTS WITH FONTAN CIRCULATION

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Background: Although the Fontan operation has contributed greatly to the improvement of survival in patients with a single ventricle, cardiac function and exercise capacity may deteriorate over time. However, there have been no quantitative analyses of the effects of Fontan circulation on myocardial work.
Recently, a novel non-invasive method for calculating MW has been introduced on the basis of speckle tracking analysis with the estimation of LV pressure from brachial artery cuff pressure.

Purpose: Aim of this study was to evaluate the diagnostic performance of the non-invasive myocardial work indices in predicting subclinical myocardial work impairment in Fontan patients.

Methods: A total of 69 patients were included and compared with healthy age- and sex- matched controls (CTRL).
Ventricular systolic function and global longitudinal strain (GLS) were assessed. Cardiopulmonary exercise test was performed. Global myocardial work index (MWI) was calculated as the area of the LV pressure strain loops. From MWI, global Constructive Work (MCW), Wasted Work (MWW) and Work Efficiency (MWE) were estimated.

Results: The two groups were comparable for blood pressure, weight end height. Mean age of Fontan patients was 21.0±9.2 years. MWI (1162 ± 364 mmHg% vs 1777 ± 240 mmHg%, $p < 0.001$), MCW (1554 ± 450 mmHg% vs 2102 ± 221 mmHg%, $p = 0.001$) and MWE (90 ± 6% vs 96 ± 2% $p = 0.001$) were significantly reduced in Fontan patients compared with healthy CTRL. Moreover, GLS (-13,9± 3,1 % vs -21,2 ± 1.5%, $p < 0.001$) and the ejection fraction (EF) (58,9 ± 4.5% vs 63.3 ± 3.9%, $p < 0.002$) were significantly lower in Fontan patients. Fontan patients with normal EF showed, however, significantly reduced values of MWI compared with CTRL ($p<0.05$).
Fontan patients with functional right ventricle showed significantly reduced MWE compared with patients with functional left ventricle ($p=0.030$).
In univariate analysis, peak VO2 was significantly associated with age, SatO2, MWI. In multivariate regression, lower peak VO2 was associated with older age ($p = 0.003$) and lower MWI ($p = 0.026$).

Conclusions: Fontan physiology is associated with disadvantageous ventricular work. In Fontan patients estimation of MWI may be a more sensitive indicator of myocardial work impairment compared with EF, and is able to predict exercise capacity.

C21

DIAGNOSIS, MANAGEMENT AND OUTCOMES OF RIGHT AORTIC ARCH: FROM FETAL LIFE TO LONG-TERM FOLLOW-UP AT A TERTIARY CENTER

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Background and **Aim:** (1) to report our experience with right aortic arch (RAA) and its variants, association with complete vascular ring (CVR), congenital heart disease (CHD), extracardiac malformations and (2) to describe the long-term follow-up in terms of symptoms and surgery for CVR.

Methods: We collected clinical and instrumental data from 502 patients with RAA referred at our tertiary center.

Results:157 patients (31.3%) had fetal diagnosis (medium follow-up 7.6 years). 330 (65%) had CHD and 89 (17%) genetic anomalies, with DiGeorge syndrome the most frequent (9.5%). Genetic disorders were significantly more frequent in those with CHD (23% versus 7.5%, $p < 0.001$). 58 (11%) had extracardiac anomalies. Among the RAA not forming CVR, left anterior ductus arteriosus with mirror-image (MI) branching pattern of epiaortic vessels (RAA/LADA/MI) was present in 29, 100% associated with CHD (conotruncal in 72%). Right ductus arteriosus (RAA/RDA/MI) was found in 8, 4 with complex CHD. CVR was present in 246 (49%): the most frequent were RAA, left posterior ductus arteriosus with aberrant left subclavian artery (RAA/LPDA/ALSA – 163 cases, 66%) and double aortic arch (DAA – 66 cases, 27%), Left posterior ductus arteriosus (LPDA) with MI branching (RAA/LPDA/MI – 12 cases, 5%) and aberrant left innominate artery (RAA/LPDA/ALIA – 5 cases, 2%) were rare. CVR were most frequently associated with minor CHD (26% versus 17% major CHD). Seventy-one (29%) pts had respiratory symptoms (29 in the first 2 months of life, 22 within the first year of life, 20 afterwards). 80 pts performed lung-function test, 28 (35%) were positive. At CT mean tracheal compression was 46% (57% for DAA, 43% for RAA/LPDA/MI. 109 (45%) were operated (median age 31.2 months). Three among 16 pts were re-operated for persistent symptomatic compression, the majority being DAA.

Conclusions: RAA is commonly associated with CVR, asymptomatic in 2/3 of cases but requiring surgery in 50%. Symptoms resolves almost completely and reoperations are reserved to the DAA. RAA is present along with major CHD in half cases and these patients are at higher risk of genetic syndromes, however not frequent.

C22

3D-ECHOCARDIOGRAPHY FOR EVALUATION OF RIGHT VENTRICLE SYSTOLIC FUNCTION IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT: A COMPARISON STUDY WITH CARDIAC MAGNETIC RESONANCE

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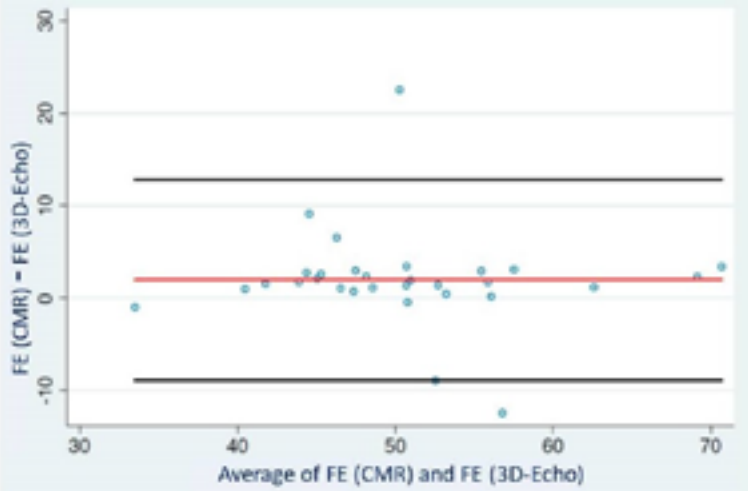
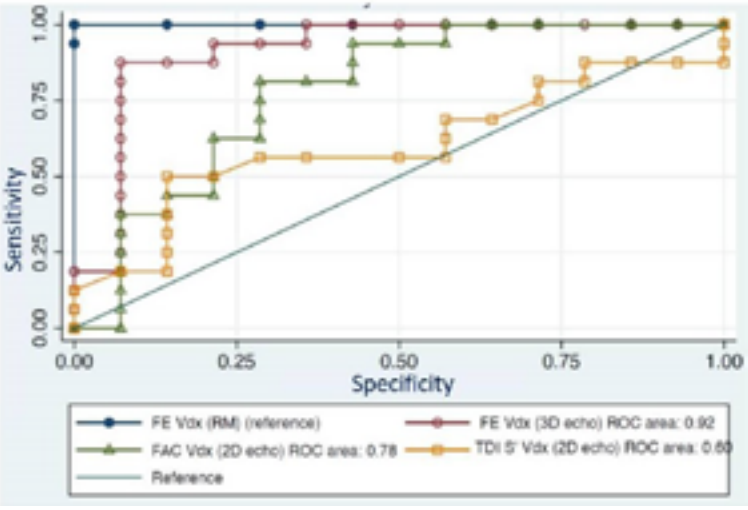
Background: Patients with repaired Tetralogy of Fallot (ToF) need periodic instrumental assessments, due to complications they can meet during lifetime. Right ventricle (RV) dilatation and dysfunction are some examples that explain why those patients regularly undergo Cardiac Magnetic Resonance (CMR), currently the gold standard method to detect these complications.
3D-echocardiography is an emerging tool to study RV, but there is little data supporting its use in congenital heart diseases. This study aimed at evaluating its accuracy in assessing RV systolic function in ToF patients, in comparison to CMR.

Materials and **Methods:** 34 patients were prospectively enrolled after CMR performance. They all underwent standard 2D-echocardiography and Multi-Beat-ECG-triggered 3D Full-Volume acquisition.
Post-hoc analysis of 3D images was performed through a vendor-independent software in 30 patients, resulting in a feasibility of 88%. RV Ejection Fraction (RV-EF) was defined both in 3D-echocardiography and CMR.

Results: A Bland-Altman analysis was performed, showing that 3D-echocardiography overestimates the RV-EF of 1,97% on average (CI 95%: 0.01-3.95, SD: 5.43), compared to CMR.
Secondly, accuracy of different echocardiographic parameters (both 2D and 3D) in detecting a RV systolic impairment was analyzed: ROC curves showed that RV-EF in 3D echocardiography is the measurement that best correlates to CMR in detecting RV dysfunction (AUC 0.92, CI 95%: 0.79-1.00), followed by Fractional Area Change (FAC) (AUC 0.78, CI 95%: 0.60-0.96).
Tricuspid Annular Plane Systolic Excursion (TAPSE) and tricuspid valve annular motion velocities in systole (S'-TDI), instead, showed very poor correlation.

Conclusions: 3D-echocardiography showed good agreement with CMR in defining RV systolic function in ToF patients, even more than the bidimensional measurements cardiologists are used to. This could lead to a wider use of this tool in daily clinical practice, allowing a precise assessment of RV also when CMR is not promptly available.





C23

LATE PULMONARY COMPLICATIONS AFTER ARTERIAL SWITCH OPERATION: A CARDIOVASCULAR MAGNETIC RESONANCE STUDY

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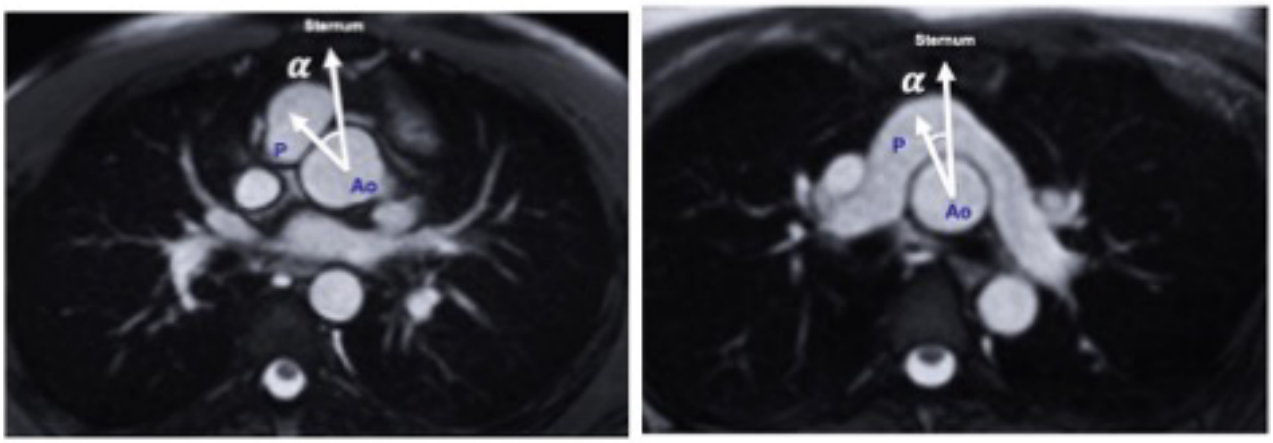
Background: Arterial switch operation (ASO) is the standard of care for D-transposition of the great arteries. Despite excellent early outcomes, supravalvular pulmonary stenosis (PS) and pulmonary branches stenosis (PAS) are common long-term complications that may require reintervention. Factors triggering these complications remain unclear. The study aimed to evaluate, through Cardiovascular Magnetic Resonance (CMR), anatomical features associated with late pulmonary complications after ASO.

Material and methods: We retrospectively analyzed CMR examinations of patients (n=62) who underwent ASO without reintervention for pulmonary complications. On a CMR axial view, the position of the neo-pulmonary artery in relation to the neo-aorta was expressed in terms of the angle between the line connecting the centroid of the two vessels and the line connecting the aorta and the sternum. This angle (?) was measured proximally, as the basal-angle-sternum (BAS) and distally, as the distal-angle-sternum (DAS) (Figure 1).

Results: The mean age of the study population was 16 ± 7 years. PS was reported in 50% (n=31) of patients while the 40% (n=25) developed PAS. In patients with PS, BAS was significantly smaller (p<0.001) than in patients without PS (10.2° ± 7.6° vs. 22.0° ± 9.6°). Conversely, in patients with PAS, DAS was significantly higher (p<0.001) than in patients without PAS (23.1° ± 10.1° vs. 9.0° ± 7.8°). Receiver operating characteristic (ROC) curve analysis reported a BAS cut-off of 15.8° (AUC=0.83, p <0.001) for PS and a DAS cut-off of 14.4° (AUC=0.86, p<0.001) for PAS.

Conclusions: Pulmonary artery position may represent an anatomical risk factor for late pulmonary complications after ASO. These observations may contribute to optimize the surgical strategy of ASO through better manipulation of the neo-pulmonary root and to perform a closer follow-up of patients with anatomical features of increased risk.



α : Basal Angle – Sternum (BAS)

α : Distal Angle – Sternum (DAS)

C24

THE EFFECTS OF EXERCISE TRAINING ON CARDIOPULMONARY EXERCISE TESTING AND CARDIAC BIOMARKERS IN ADULT PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME AND FONTAN CIRCULATION

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Background: Several studies have shown that adult patients with Hypoplastic Left Heart Syndrome (HLHS) and Fontan circulation have a reduced exercise tolerance that affects daily life. Recent studies have investigated the effects of aerobic exercise training in patients with univentricular heart; however, this research topic is still poorly studied. The aim of this study was to evaluate the effects of an aerobic exercise training program on cardiopulmonary exercise testing parameters and cardiac biomarkers in patients with HLHS.

Methods: We enrolled 12 patients with a mean age of 24 ± 2.5 years (range 22-27 years), 50% male, with HLHS at Bambino Gesù Children's Hospital IRCCS. All patients underwent a cardiopulmonary test and blood sampling before (T0) and after (T1) a 4-week aerobic exercise program. Cardiac biomarkers hs-cTnT, NT-proBNP, ST2, GDF-15 were studied.

Results: Data analysis demonstrated an increase in cardiorespiratory performance after 4 weeks of aerobic exercise training activity. In particular, the data showed a significant improvement in test duration ($p < 0.05$), heart rate at rest ($p < 0.05$), heart rate recovery 1 min ($p < 0.05$), VO2 max ($p < 0.01$) and oxygen uptake efficiency slope ($p < 0.05$). At the same time, the data showed a significant reduction in NT-proBNP and ST2 values ($p < 0.01$ and $p < 0.05$, respectively) and a significant increase in GDF-15 ($p < 0.01$). No significant changes were found between the hs-cTnT values.

Conclusions: Our study demonstrated the 4-week efficacy of an aerobic training program in improving cardiorespiratory performance and cardiac biomarker values in adult patients with HLHS and Fontan circulation. To the best of our knowledge this is the first study that has demonstrated the prescription of exercise training in patients with HLHS and the improvement of clinical, functional and biochemical parameters.

C25

HEART RATE AND LEFT VENTRICULAR EJECTION FRACTION IN PEDIATRIC DILATED CARDIOMYOPATHY: HOW MUCH DO WE HAVE TO LOWER IT BY?

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Introduction: An elevated heart rate is associated with an increased risk of death and cardiac transplant in children with dilated cardiomyopathy (DCM). Whether heart rate is a biologic marker to address therapy, is poorly investigated in pediatric age.

Aim: To investigate the relationship between heart rate reduction (HRR) and left ventricular ejection fraction (LVEF) in children affected by DCM, treated with carvedilol.

Methods: This is a multi center retrospective analysis conducted on all children with DCM (aged < 18 years) between 2013 and 2020, with LVEF < 40% and treated with carvedilol. Echocardiographic data on left ventricular function and dimension were collected. The relationship between HRR and left ventricular remodelling (LVEF, left ventricular end-diastolic (LVEDd) and end-systolic diameters (LVESd) diameters) was assessed before and after HRR with carvedilol, using regression analysis.

Results: 100 patients were enrolled (M: 51; age 7 ± 8 y). The average LVEF before treatment was 30.2 ± 10%. All patients were treated with anti HF therapy. Carvedilol was up titrated to the maximal tolerated dose. There was a positive relationship between HRR and increase in LVEF (R2 =0.06, p=0.014). A HRR of > 20% was related to an improvement in LVEF > 13%: mean LVEF at maximum therapeutic dose was 43.7 ± 9.6% (p < 0.0001). The magnitude of LVEF improvement was enhanced by a major reduction in HR. At long term, 3 years follow up, HRR allowed a positive remodelling of LV, with a significant diameters reduction (R2=0.1, p=0.003 and R2 =0.07, p=0.008, for LVESd and LVEDd respectively) and LVEF recovery up to 15% (p<0.0001). No deaths or heart transplant were recorded during follow-up.

Conclusions: This study demonstrates that HRR is safe and improvement in LVEF is related to the degree of HRR.

C26

RESULTS OF PEDIATRIC HEART TRANSPLANTATION PERFORMED IN THE LATE 80S

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Purpose: Analyze results of pediatric heart transplantation (HTx) performed in the late 80s.

Methods: Retrospective single center study enrolling 27 paediatric HTx patients who underwent surgery between 1986 and 1990 at Bambino Gesù Children's Hospital.

Results: Age at HTx ranged from 1 month to 14 years (median 4.6). Indications to HTx were: dilated cardiomyopathy (n = 17), congenital heart disease (n = 5), restrictive cardiomyopathy (n = 3), cardiac tumor (n = 1) and toxic cardiomyopathy (n = 1).
Global survival was 33% (9/27) at 32.3 years of median follow up.
Causes of early death (n = 6) were: acute right ventricle failure due to pulmonary hypertension (n = 2), multi organ failure (n = 2), massive pulmonary thromboembolism (n = 1), intractable bleeding in a Fontan failure patient (n = 1). Twelve late deaths occurred between 1 month and 15 years from HTx at a median time of 7.6 years from HTx. Causes of late death were: acute rejection (n = 4, 2 of which for non-compliance), chronic rejection (n = 4), haemodialysis complications (n = 1), sepsis (n = 1 in a Re-HTx), bleeding from tracheostomy (n = 1), post transplant lymphoproliferative disorder (PTLD, n = 1).
Major complications were: 3 are free from major complications (1 successful pregnancy), 2 underwent re-HTx (1 currently listed for kidney transplant in haemodyalisis), 4 cardiac allograft vasculopathy requiring coronary stenting, 4 PTLD off-therapy.
The immunosuppression therapy is personalized for different toxicity profile: 3 patients are in triple therapy (CNI + mmf/mTOR + steroids), 2 patients are in double therapy (CNI/mTOR + mmf) and 2 are in monotherapy (CNI).

Conclusion: Over 30 years follow up pioneering our experience suggest that pediatric HTx is an acceptable option for end stage cardiac patients.
HTx may prolongs the lifespan and quality of life, changing the underlying heart disease in potentially favorable clinical condition.

C27

TACHIARITMIE NEL FOLLOW-UP A LUNGO TERMINE DI PAZIENTI SOTTOPOSTI AD INTERVENTO DI FONTAN CON CONDOTTO EXTRACARDIACO: INCIDENZA E RUOLO DELL'ANATOMIA VENTRICOLARE NATIVA

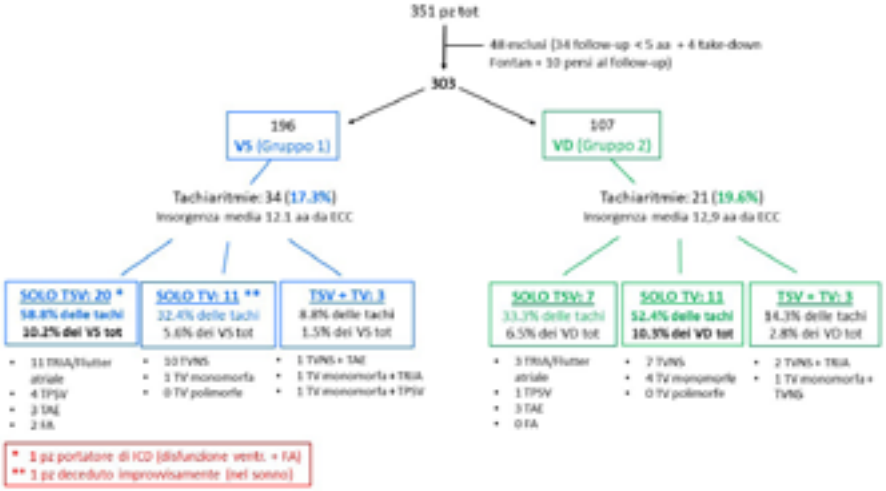
C. Di Mambro, M.L. Yammine, P.P. Tamborrino, M.S. Silvetti, A. De Zorzi, S. Giannico, R. Adorisio, A. Santilli, U. Giordano, A. Carotti, S. Albanese, L. Galletti, F. Drago
DMCCP dell'Ospedale Pediatrico "Bambino Gesù", Roma, Italy

Introduzione e Scopo dello studio: Le aritmie rappresentano una comune e spesso severa comorbidità a lungo termine nelle cardiopatie complesse sottoposte a palliazione chirurgica di tipo univentricolare secondo Fontan. Dal 1971, numerose varianti chirurgiche hanno portato ad oggi a preferire l'utilizzo del condotto extra-cardiaco (ECC). Molti studi hanno riportato la minore aritmogenicità della palliazione extracardiaca (particolarmente per l'incidenza delle aritmie ipercinetiche), anche se il follow-up (FU) dei pazienti negli studi pubblicati in letteratura risulta significativamente più breve rispetto a quello pubblicato per le precedenti metodiche (Fontan classica, tunnel intra-atriale).
In questo studio monocentrico, abbiamo analizzato, in una coorte di pazienti sottoposti a intervento di Fontan ECC, le possibili complicanze tachiaritmiche, la loro epoca di insorgenza e l'eventuale correlazione con la tipologia di ventricolo nativo (sinistro o destro), in un FU a lungo termine non < 5 anni.

Materiali e Metodi: Per l'analisi dell'incidenza di tachiaritmie sopraventricolari e ventricolari, abbiamo retrospettivamente valutato i dati clinici e strumentali di una popolazione di 351 pazienti consecutivamente sottoposti a Fontan ECC, seguiti presso la nostra Istituzione, in FU medio 16 anni (5-31 anni; IQR 11-20 anni). Dei 351 pazienti, 48 (13.7%) sono stati esclusi dallo studio: 34 (9.7%) per un FU < 5 anni; 4 (1.1%) perché sottoposti a take-down della Fontan; 10 (2.8%) perché persi al FU.
La popolazione finale dello studio è stata quindi di 303 pazienti (56% maschi), con età media all'ultimo controllo di 21 anni (8-49 anni; IQR 16-28 anni), sottoposti a Fontan ECC ad un'età media di 3 anni (IQR 3-5 anni). Centonovantasei pazienti (64.7%) presentavano un'anatomia di tipo univentricolare sinistra (Gruppo 1 - VS) mentre 107 (35.3%) di tipo univentricolare destra (Gruppo 2 - VD).

Risultati: Nel corso del FU, 34 pazienti del Gruppo 1 (17.3%) hanno manifestato almeno un episodio tachiaritmico, con insorgenza media dall'intervento di 12.1 anni, mentre 21 soggetti del Gruppo 2 (19.6%) hanno presentato almeno un evento tachiaritmico, ad una media di 12.9 anni dall'intervento. I pazienti con un'anatomia di tipo univentricolare sinistra hanno manifestato in percentuale più tachiaritmie sopraventricolari rispetto ai pazienti con un'anatomia di tipo univentricolare destra (58.8% vs 33.3%), mentre questi ultimi hanno manifestato più tachiaritmie ventricolari isolate (52.4% vs 32.4%). Per quanto attiene, invece, alla contemporanea presenza di aritmie sopraventricolari e ventricolari in uno stesso paziente, l'incidenza nei due Gruppi si è rivelata sostanzialmente simile (8.8% vs 14.3%). - Vedi Figura.

Conclusioni: Questo studio, focalizzato sull'incidenza a lungo termine delle complicanze tachiaritmiche in un'ampia coorte di pazienti sottoposti a palliazione chirurgica secondo Fontan ECC e basato su un periodo di FU tra i più lunghi mai pubblicato, mostra come tale procedura appaia più "aritmogena" di quanto fin qui evidenziato dalla letteratura specializzata. Inoltre, i nostri dati suggeriscono che, seppur non vi sia differenza nell'incidenza generale di aritmie ipercinetiche nei due Gruppi descritti, i soggetti con anatomia ventricolare nativa di tipo sinistro sembrano sviluppare maggiormente tachicardie sopraventricolari, mentre quelli con ventricolo nativo di tipo destro appaiono essere più colpiti da aritmie ventricolari complesse forse frutto dell'impegno sistemico di un ventricolo "non preparato ad affrontarlo"



C28

DILATED CARDIOMYOPATHY WITH SEVERE LEFT VENTRICULAR DYSFUNCTION IN PROPIONIC ACIDEMIA: HOW TO TREAT PATIENT SOLELY WITH LIVER TRANSPLANTATION - A CASE SERIES

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Introduction: Dilated cardiomyopathy (DCM) with reduced left ventricular ejection fraction (LVEF) is a cause of morbidity and mortality in patients affected by propionic academia (PA), a form of organic acidemia caused by an inherited deficiency of the enzyme Propionyl CoA carboxylase. The management of patient presenting with severe LV dysfunction and without metabolic failure is controversial. In some cases, a combined liver-heart transplant has been proposed. We adopted a different strategy, combining recovery of LV using anti heart failure (HF) medical therapy with liver transplantation.

Aims: We propose a different strategy to manage DCM with severe LV dysfunction: maximized anti-HF medical therapy combined to liver transplantation.

Methods: in the group of patients followed at Bambino Gesù Children's Hospital in Rome with diagnosis of PA, we selected those presenting with DCM with severe LV dysfunction (LVEF <35%), and without frequent metabolic decompensation. Firstly, patient underwent a complete cardiac evaluation comprehensive of echocardiogram, cardiac magnetic resonance [MRI], and cardiac biomarkers dosage. Hence, they started anti-HF therapy with beta-blockers up-titration to the maximal tolerated dosage, ACE inhibitor at therapeutic dose, and anti-aldosteronic therapy. On top of beta-blocker, ivabradine was added in those with heart rate > 70 bpm. Sacubitril-valsartan replaced ACE inhibitors in those with persistent severe LV dysfunction. After a period of observation, when LVEF improved to >40%, they have been placed on the liver transplant list. During transplant, they were supported by ECMO and a 24 hours infusion of levosimendan was administered from the time of hospital admission. After 24 hours from transplant, anti-HF therapy was gradually reintroduced.

Results: among all the 17 patients affected by PA actually alive, 6 presented DCM with severe LV dysfunction (LVEF < 35%) as the major manifestation of metabolic disease. Mean LVEF was 30% (from 13 to 35 %). They all started the anti-HF therapy. 5 have been treated also with ivabradine, 1 switched to sacubitril-valsartan. 5 underwent liver transplantation. Mean age at liver transplantation was 17.8 ± 2.2 years. Mean LVEF before liver transplant was 43 % (± 6). All patients were supported by ECMO during surgery, and levosimendan was started at hospital admission and lasted up to 24 hours. All patients survived surgery. One patient died 3 months after liver transplant for pulmonary infection. At last follow up mean LVEF was 48.3% ± 8.8. In particular the first two patient transplanted recovered respectively from 35% to 54% and 48%, after 18 months of follow up.

Conclusions: patient affected by PA and DCM with severe LV dysfunction without metabolic decompensation, may benefit from a composed strategy with anti-HF therapy and liver transplantation. This strategy, may improve LVEF, hence long term outcome of these patients. Further evidence are needed.

C29

ABLAZIONE TRANSCATETERE DELLA TACHICARDIA DA RIENTRO NODALE IN BAMBINI < 26 KG: EFFICACIA E SICUREZZA DELLA STRATEGIA CRIO-ABLATIVA CON MAPPAGGIO DI VOLTAGGIO DEL TRIANGOLO DI KOCH

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L'ablazione transcatetere con radiofrequenza della tachicardia da rientro del nodo atrioventricolare (TRNAV) nei bambini piccoli < 25 kg è ad alto rischio di blocco AV iatrogeno fino all'8,5% dei pazienti. La letteratura pediatrica più recente ha riportato che la crioablazione della via lenta con la strategia del mappaggio di gradiente di voltaggio del triangolo di Koch (TK) per la ricerca del low voltage bridge (LVB), associato alla via lenta di conduzione nodale atrio-ventricolare, è molto efficace e sicura nel trattamento dell'AVNRT nei bambini. Per questo motivo, lo scopo del nostro studio è stato quello di valutare l'efficacia e la sicurezza, sia nella fase acuta che nel follow-up a medio termine, di tale strategia crioablativa in bambini <26 kg.

Metodi e Risultati: Quattordici bambini con AVNRT [64% maschi, età mediana 6,5 anni (IQR 6-8 anni), peso mediano 25,5 kg (IQR 24-26 kg), superficie corporea mediana (metodo Du Bois) 0,95 m^2 (IQR 0,91-0,96 m^2)] sono stati arruolati in questo studio. Tredici pazienti hanno presentato una TRNAV tipica, mentre un paziente sia tipica che atipica. Un jump AV era presente nel 50% dei casi. Un singolo LVB è stato rilevato in 7 pazienti (50%) a livello di M1-M2, in altri 4 (28,5%) a livello di P2, in 1 (7%) a livello di A2 e 2 (14,5%) a livello di M della base del TK.
Un'area mediana dei LVB di 0,22 cm^2 (IQR 0,2-0,3 cm^2) è stata misurata in un'area media del TK di 1,05 ± 0,36 cm^2.
Il successo procedurale acuto è stato del 100%, ottenuto con un numero medio di crioablazione di 5,5 per paziente. In un solo caso persistenza di Jump AV senza eco nodale. Non ci sono state complicazioni peri-procedurali
Tutte le procedure sono state eseguite con esposizione fluoroscopica prossima allo zero (tempo mediano 0,15 minuti, IQR 0-0,7 minuti), in 6 pazienti la fluoroscopia è stata di 0 minuti.
Non ci sono state complicanze o recidive durante il follow-up (mediana: 6 mesi, IQR 6-12 mesi).

Conclusioni: La crioablazione della TRNAV utilizzando la strategia LVB è una procedura molto efficace e sicura anche in pazienti pediatrici molto piccoli.

C30

SHORT-TERM ELECTROCARDIOGRAPHIC ATRIAL REMODELLING AFTER ASD CLOSURE WITH GCO DEVICE IN A PEDIATRIC POPULATION

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Background and Aim: The GORE® CARDIOFORM septal occluder (GCO) is an atrial septal defect/patent foramen ovale (ASD/PFO) closure device with theoretical advantages over other commercialized devices thanks to its softness and anatomical compliance.
Our aim was to evaluate the short and medium-term electrocardiographic changes after percutaneous ASD closure with GCO in a pediatric population.

Methods: We enrolled 39 patients with isolated ASD submitted to trans-catheter closure with GCO from January 2020 to June 2021. EKG was performed before (T0), at 24 hours (T1) and 6 months (T2) after ASD transcatheter closure. P wave dispersion was calculated as the difference between maximum and minimum P- wave duration, PR interval as the interval between beginning of the P wave and beginning of the QRS complex and QT dispersion as the difference between maximun and minimun of QTc intervals. At 6-months from device implantation, the patients were submitted to ambulatory EKG Holter recording.

Results: Patients' age and BSA were 8.2±4.2 years (IQR 4.2-8.3, median 7.0) and 1.0±0.3 m2 (IQR 0.7-1.7, median 0.9), respectively. The strectched ASD diameter was 16.3±4.5 mm (median 16), resulting in QP/QS of 1.7±0.6 (median 1.5). At the baseline mean P wave dispersion was 40±15 msec and decreased to 30±13 msec ($p<0.002$) at 24h, without any further change at 6 months (30±13 msec, $p<0.002$). PR conduction significantly improved at 24 h from device implantation (from 175.0±20.8 to 144.0±22.7 msec, $p=0.018$) and did not significantly change at 6 months (164.0±19.5 msec, p=NS). QTc dispersion decreased at 24 hours (31.7±.20.3, $p<0.02$) and at 6 months (28.0±18.1, $p<0.002$) from device implantation. After device deployment, 2 pts (5%) developed transient, self-limited junctional rhythm and one of them needed a short course of anti-arrhythmic therapy for supra-ventricular tachycardia. No tachy/brady-arrhythmias were recorded at the 6-months follow-up EKG Holter monitoring.

Conclusions: Percutaneous ASD closure with the GCO device results in significant, sudden improvement of intra-atrial, atrio-ventricular and intraventricular electrical homogeneity. This benefit persists unaltered over a medium term follow-up. It might be due to a favourable volumetric remodelling that was not hindered by mechanical impact of the occluding prosthesis and could explain the low rate of arrhythmias found at the mid-term EKG evaluation

	At 0 time	After 24 h	6 months later
Δpm (s)	0,040	0,030	0,030
[p value]		[0,002]	[0,002]
ΔQTc m (s)	41	31,7	28
[p value]		[<0,002]	[0,002]
PRm (s)	0,175	0,144	0,164
		[0,018]	[NS]

C31

MONOAMINE OXIDASE INHIBITION REDUCES CARDIAC DYSFUNCTION IN DYSTROPHIC CARDIOMYOPATHY

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Introduction: Dystrophic cardiomyopathy culminates in heart failure and arrhythmias and is a major burden for Duchenne muscular dystrophy (DMD) patients. Mechanism-driven therapies designed to contrast the development of muscle and cardiac dysfunction are still missing. Antioxidant treatments counteract myocyte injury in mdx mice, a genetic model of DMD, supporting the possible role of enhanced reactive oxygen species (ROS) in the pathophysiology of muscular dystrophies. Previous evidence shows that monoamine oxidases (MAO) represent an important source of ROS, thus contributing to cardiomyocyte damage and dysfunction in different models of heart disease.

Objectives: we tested whether MAO-induced ROS formation contributes to the progression of pathology in dystrophic hearts. Materials and **Methods:** wild type (WT) and mdx mice at 3 and 12 months of age were employed. Cardiac function was determined by echocardiography measuring: fractional shortening, ejection fraction and left ventricle strain. Cardiac structure and fibrosis amount were assessed through histology (H&E, Masson's Trichrome). To test whether reduction in ROS burden could ameliorate cardiac structure and function in mdx mice, MAO-B inhibitor safinamide was administered to WT and mdx mice respectively at 3 and 12 months of age. **Results:** We found that fractional shortening (FS) and ejection fraction (EF) were reduced by 1.2- and 2-fold at 3 and 12 months of age, respectively, in mdx mice vs their WT counterpart. In addition, left ventricle (LV) strain was impaired in mdx mice already at 3 months of age. This functional impairment was accompanied by a 5-fold increase in fibrosis in mdx hearts, evident already at 3 months. Safinamide administration for 30 days, led to a significant improvement of FS, EF and LV strain in mdx mice. Half of the mice in the safinamide cohort showed reduced levels of myocardial fibrosis.

Conclusions: Taken together, these results suggest that pharmacological MAO-B inhibition improves cardiac function in a genetic model of DMD and may represent a clinically relevant target for the treatment of dystrophic cardiomyopathies.

C32

L'EXTRASISTOLIA VENTRICOLARE IN ETÀ PEDIATRICA NEL CUORE SANO: PROGNOSI E IMPLICAZIONI CLINICHE. STUDIO RETROSPETTIVO MONOCENTRICO

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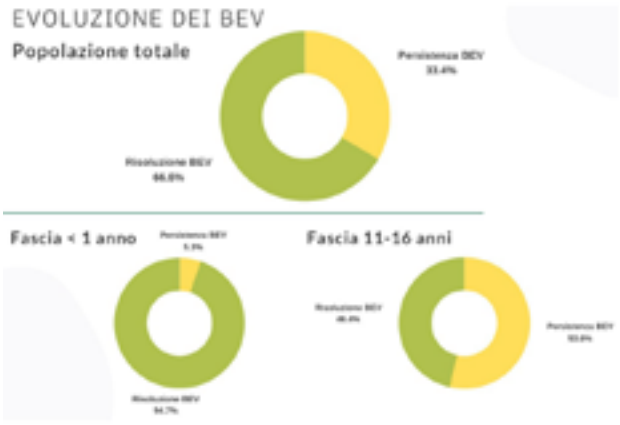
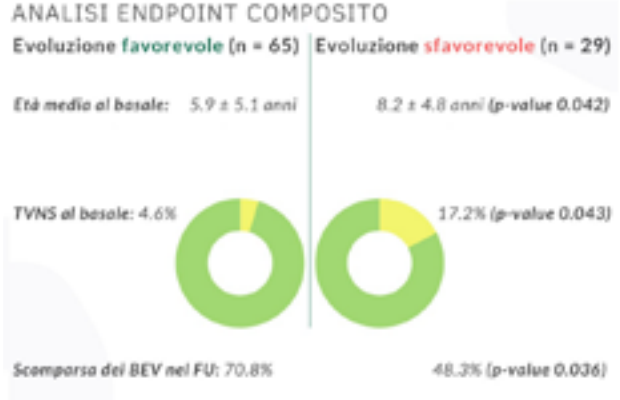
Scopi della Ricerca: I battiti ectopici ventricolari (BEV) focali costituiscono la forma di aritmia ventricolare di più frequente riscontro nella popolazione generale, con un'elevata prevalenza anche nei soggetti sani. Si tratta di un reperto perlopiù benigno, tuttavia i BEV possono costituire l'epifenomeno di una cardiomiopatia misconosciuta, condizionare lo sviluppo di sintomi o di disfunzione sistolica ventricolare sinistra. Lo scopo di questo lavoro è indagare alcuni aspetti ancora controversi in merito a tale entità: evoluzione e prognosi nel lungo periodo nei soggetti in cui sia esclusa una cardiopatia, il riconoscimento di elementi predittivi di un'evoluzione sfavorevole, relazione tra carico aritmico e sviluppo di disfunzione ventricolare, indicazione all'esecuzione di Risonanza Magnetica (RMN) cardiaca o all'avvio di terapia.

Metodi impiegati: Valutazione retrospettiva di ECG Holter 24 ore ed altri esami strumentali (Ecocardiogramma, RMN, prova da sforzo) in 99 soggetti di età 0-16 anni (media alla valutazione basale: 6.6 +/- 5.1 anni), con durata del follow-up > 4 anni (7.2 +/- 2.6 anni) e con >500 BEV ad ECG Holter basale. Criteri d'esclusione: cardiopatia congenita a medio o elevato grado di complessità, cardiomiopatie, canalopatie, pregressa miocardite, disturbi di conduzione o tachicardia ventricolare (TV) sostenuta all'esordio. Si è definito un endpoint composito per definire l'evoluzione sfavorevole dei BEV nel follow-up: documentazione di elevato carico aritmico (> 10% BEV / 24 h), disfunzione ventricolare o TV sostenuta.

Risultati e Conclusioni: Nel 66.6% dei soggetti si è avuta una risoluzione dei BEV, in media dopo 2.6 +/- 2.1 anni: in particolare, la risoluzione dei BEV esorditi nel primo anno di vita è stata pari al 94.7%. Il 31.3% dei soggetti ha presentato > 10% BEV / 24 h ad almeno una delle valutazioni. Il 4% dei soggetti ha presentato TV sostenuta o TV non sostenuta con ciclo RR rapido (> 180 bpm): di questi, in due individui si sono in seguito rilevate forme lievi di cardiopatia. Non si sono rilevate TV polimorfe. Una iniziale disfunzione ventricolare si è riscontrata nel 2% dei soggetti, senza franca correlazione col carico aritmico. La terapia è stata avviata nel 12% degli individui, con un'efficacia nella prevenzione di aritmie complesse o sintomi poco inferiore al 50%. I soggetti con evoluzione sfavorevole avevano mediamente un'età maggiore all'esordio (8.2 vs 5.9 anni), manifestando più frequentemente TV non sostenute alla valutazione basale (17.2 vs 4.6%), mentre non c'è correlazione con la presenza di sintomi o la morfologia dei BEV.
Si conferma la benignità dei BEV in assenza di cardiopatia strutturale. In presenza di TV non sostenuta con ciclo RR rapido o polimorfismo, riteniamo comunque ragionevole approfondimento con RMN; viceversa, in presenza unicamente di elevato carico aritmico, non c'è indicazione ad esecuzione di RMN o ad avvio di terapia. Lo sviluppo di disfunzione ventricolare secondaria ai BEV in ambito pediatrico è molto infrequente e non sembra esserci associazione con carico aritmico. Pur avendo selezionato soggetti a cuore apparentemente sano, in rari casi si sono in seguito slatentizzati quadri più francamente patologici: è utile quindi un costante follow-up, soprattutto in presenza di alterazioni strutturali anche minori.





C34

ATRIAL FLOW REGULATOR DEVICE IN CHILDREN AND IN PATIENTS WITH CONGENITAL HEART DISEASE: AN INTERNATIONAL REGISTRY.

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Background

Atrial Flow Regulator (AFR) (Occlutech, Istanbul, Turkey) is a double disc device made of self-expanding Nitinol wire mesh. The device is structured around a central lumen, intended to maintain a patent communication. Once deployed percutaneously, the central portion of the device stents the atrial septum leaving a preselected calibrated atrial communication.
Its use has been approved in adult population with heart failure and described for pulmonary hypertension.
Only case reports have been published about its use in pediatric population and in congenital heart disease.

Aim

Aim of our study is to describe the use of AFR in children with congenital heart disease and pulmonary hypertension in a multicenter retrospective registry.

Materials and methods

Nine centers worldwide participated in the registry. Inclusions criteria were: patients at any age with congenital heart disease or patients aged < 18 years with pulmonary hypertension or cardiomyopathy needing AFR implantation.

Results

From 2019 to 2022, 29 patients underwent AFR implantation. Median age of the population was 59 months (IQR 34-104 months) and median weight was 17,5 Kg (IQR 10,5-54 Kg). Twenty-four patients had congenital heart disease, 2 patients had a cardiomyopathy and 3 pulmonary hypertension. Indications for AFR implantation were: left heart failure (12pts), pulmonary hypertension (8 pts), severe desaturation in fenestrated Fontan (5 pts) and Fontan failure (4 pts).
Almost all the procedures (93%) were performed under general anesthesia. Venous access was femoral in 24 pts (83%), jugular in 2 pts (7%), transhepatic in 2 pts (7%) and subclavian in 1 pt (3%). Maximum venous sheath was 16 Fr. AFR implantation required balloon pre-dilatation in 20 pts (69%). Implantation success rate was 100%. Two immediate AFR occlusion were registered: 1 in a patient weighing 13 Kg with a Fontan failure in which AFR 4 mm was used to create a fenestration and 1 in a patient weighing 19,5 Kg with a fenestrated Fontan with desaturation in which AFR 4 mm was used. In one case the clotted AFR was immediately removed and substituted with a 6 mm AFR and in the other the clotted AFR was removed and substituted with a 6 mm AFR after 1 month.
At a median follow up of 1 year no major complications occurred. 28/29 patients were alive and the only death was not related to the procedure. One 4 mm AFR clotted 14 months after implantation and was treated with balloon angioplasty.
Thirteen patients improved their NYHA class, 11 patients didn't change their NYHA class and only 1 patient with idiopathic PH worsened his NYHA class.

Conclusions

AFR implantation in children with congenital heart disease and pulmonary hypertension is promising and at short follow up of 12 months seems to have beneficial effects. The AFR has the potential to provide benefits in terms of symptoms and survival to a variety of patients with limited treatment options and indeterminate prognosis.

C35

BEGRAFT AORTIC STENTS: A EUROPEAN MULTI-CENTRE EXPERIENCE REPORTING ACUTE SAFETY AND EFFICACY OUTCOMES FOR THE TREATMENT OF VESSEL STENOSIS IN CONGENITAL HEART DISEASES

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Background and Aim: Stent implantation for the treatment of vessel stenosis in congenital heart diseases has become the preferred method of treatment. Availability of covered stents may decrease complications and may have an important role in the management of patients with complex anatomy. Aim of this study is to evaluate the feasibility and safety of the premounted cobalt chromium stent graft covered ePTFE Aortic BeGraft in a broad spectrum of vascular lesions.

Methods: This is a multicenter retrospective results analysis of 67 implanted Be Graft Aortic stents between 2016 and 2022 in four different European centers.

Results: 63 patients aged 12 years (range 4-59 years), with body weight of 59.5 Kg (range 14-103 Kg) underwent Be Graft stent implantation. Fifty-four patients had aortic coarctation, 3 patients had conduits stenosis in a total cavopulmonary connection (TCPC), 6 patients had stenosis of pulmonary conduit and one patient was treated for a wide portocaval fistula. All the stents were implanted successfully. Median stent diameter was 16 mm (range 12-24 mm) and median length was 39 mm (range 19-49 mm). Four patients received 2 stents. Mean final long sheath size was 12 +/- 3 Fr. Aortic gradient decreased from a mean of 40 +/- 15 mmHg to 12 +/- 6 mmHg. Three major complications occurred. During a conduit stenting procedure, the stent balloon ruptured with some difficulties in retrieval, 1 patient experienced a cerebral embolization without neurological consequences and 1 patient had a femoral artery occlusion requiring vascular surgery for reperfusion. Mean follow up was 11 months. No late complications occurred. One patient had a stent narrowing in aortic position during follow up that needed redilatation.

Conclusions: Be- graft stent can be used safely and effectively in a wide spectrum of congenital heart diseases. Whether these good results will be stable in the long term follow up still needs to be investigated given its recent introduction into clinical practice

C36

IMPATTO DEL DISPOSITIVO GORE CADIOFORM ASD OCCLUDER SULLA FUNZIONE ATRIALE E VENTRICOLARE. STUDIO SU UNA POPOLAZIONE PEDIATRICA

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Introduzione: L'approccio percutaneo costituisce il trattamento di prima linea per la chiusura di difetto interatriale ostium secundum (DIA), in accordo con le attuali linee guida. Il dispositivo GORE® Cardioform ASD Occluder (GCA) ha ricevuto recentemente il marchio CE. Il design del device è potenzialmente innovativo in confronto con altri device autocentranti, in quanto unisce l'elevata morbidezza e compliance anatomica con la possibilità di chiudere difetti ampi fino a 30 mm.
Lo scopo di questo studio è di confrontare i cambiamenti delle proprietà meccaniche delle camere cardiache dopo la chiusura di DIA mediante device GCA.

Metodi: Questo è uno studio prospettico a braccio singolo. Sono stati arruolati pazienti consecutivi di età < 18 anni sottoposti a chiusura percutanea di DIA isolato mediante un singolo dispositivo GCA. L'analisi ecografica è stata eseguita il giorno prima, 24 ore e 6 mesi dopo la chiusura del DIA. E' stato calcolato lo strain longitudinale atriale e ventricolare al fine di studiare il rimodellamento meccanico dopo la chiusura del DIA con il dispositivo GCA. L'analisi dello strain longitudinale è stata eseguita offline (workstation Echopac v.12) mediante metodica speckle-tracking.

Risultati: Sono stati arruolati 70 pazienti tra gennaio 2020 e febbraio 2021. L'età media dei pazienti era 7.9±3.9 anni, il diametro medio del DIA era 17.1±4.5 mm. 38 pazienti (54%) presentavano un DIA complesso per presenza di margini deficitari, difetto multifenestrato e/o fossa ovale aneurismatica. 19 pazienti presentavano indicazione a chiusura chirurgica del DIA. In 15 pazienti è stato posizionato un GCA di taglia 27 mm, in 31 pazienti un GCA di 32 mm, in 15 pazienti un GCA di 37 mm, in 10 pazienti un GCA di 44 mm. È stato riportato un unico caso di embolizzazione precoce del dispositivo. Dei restanti pazienti, nessuno presentava shunt atriali residui a 6 mesi. L'analisi speckle-tracking non ha mostrato nessun cambiamento significativo della funzione longitudinale globale ventricolare dopo la chiusura del DIA. Una riduzione transitoria dello strain longitudinale a 24 ore è stata osservata nei segmenti ventricolari settali basali, in entrambi gli atri e nel ventricolo destro. Alla valutazione a 6 mesi è stato evidenziato un completo recupero di tali modifiche, verosimilmente secondarie ai noti cambiamenti emodinamici immediati dovuti al cambio di distribuzione dei volumi ematici dopo la chiusura del DIA. 6 mesi dopo la procedura solo l'atrio sinistro mostrava una riduzione dello strain longitudinale nei segmenti settali dovuta alla presenza del device in situ, ma con una tendenza al recupero della funzione atriale globale. L'analisi univariata non ha rilevato nessuna correlazione tra i valori di strain nelle camere cardiache e le dimensioni del dispositivo usato, sia assolute che corrette per il peso corporeo.

Conclusione: Questo studio suggerisce che il dispositivo GCA non impatta sulla funzione longitudinale globale e regionale di entrambi i ventricoli. Anche la meccanica atriale risulta preservata, eccetto che per i segmenti settali coperti dal device. Sulla base dei dati attualmente disponibili in Letteratura, questo è il primo device che non altera la meccanica ventricolare, indipendentemente dalle dimensioni del dispositivo usato.

Fig1: Valori di strain longitudinale (%) globale delle camere cardiache e regionale dei segmenti ventricolari basali settali.

	T 0 (basale)	T 24h	T 6 mesi
Ventricolo Sn	-23.2 ± 2.8	-23.0 ± 2.8	-23.5 ± 2.7
Setto basale ant	-20.8 ± 4.6	-19.5 ± 4.4 *	-20.1 ± 4.3
Setto basale post	-19.8 ± 3.3	-18.7 ± 3.6*	-19.2 ± 3.4
Ventricolo Dx	-27.6 ± 5.4	-23.6 ± 5.0*	-27.3 ± 4.6
Atrio Sn	41.4 ± 15.3	29.2 ± 11.4*	39.0 ± 12.9*
Atrio Dx	53.2 ±27.6	44.0 ± 23.8	52.8 ± 22.2

* p<0,05 rispetto al T0.

C37

CHIUSURA PERCUTANEA DEL DOTTO NEL PREMATURO: RISULTATI E FOLLOW-UP

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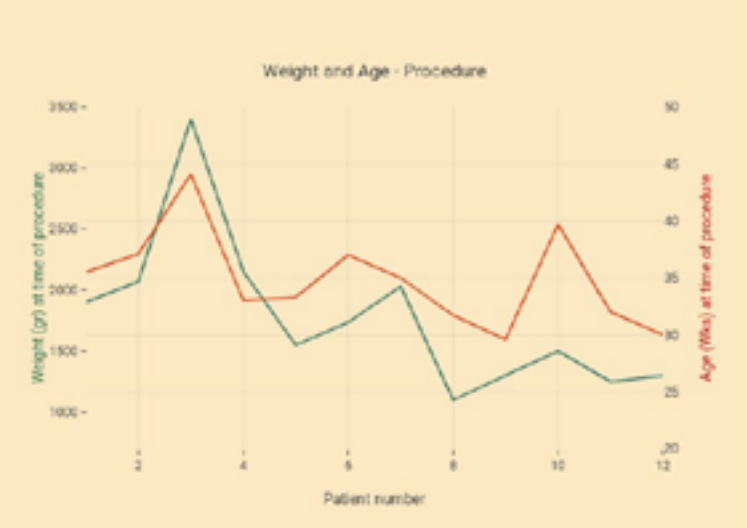
Scopi della Ricerca: In molti paesi europei e negli USA la chiusura percutanea del dotto arterioso (PDA) nel prematuro ha sostituito la chiusura chirurgica nella maggior parte dei casi. In Italia la metodica fatica a diffondersi in modo capillare sul territorio. Nel seguente lavoro si presenta l'esperienza del nostro Centro nella chiusura percutanea del PDA nel prematuro soffermandosi sul follow-up.

Metodi impiegati: La casistica coinvolge 12 neonati prematuri; le procedure sono state svolte tra luglio 2019 e aprile 2022. La tecnica è stata perfezionata con l'esperienza maturata. Viene incannulata la vena femorale con introduttore 3.3F/4F con supporto ECO; si avanza guida coronarica 0.014 (Asahi Sion Blu ES) in atrio destro su catetere JR o MP Mongoose? 3.3/4F, e si sonda poi la tricuspide avanzando la stessa fino all'aorta discendente attraverso il dotto. Si avanza quindi il catetere e si sostituisce la guida coronarica con guida angiografica 0.025 J e quindi il catetere con il sistema di delivery. I device utilizzati per la chiusura sono stati l'Amplatzer Piccolo Occluder (n=10) e l'Amplatzer Duct Occluder II AS (n=2). Il rilascio viene effettuato con guida fluoroscopica ed ecocardiografica.

Risultati e Conclusioni: I pazienti avevano un'età media alla nascita di 27+6 settimane (+/- 3 settimane) e un peso medio alla nascita di 1080.2 +/- 599 g. Sesso maschile nel 58% dei pazienti (n=7). L'età media alla procedura è di 34+5 settimane (+/- 4 settimane), con un peso medio di 1773.8 +/- 622.3g. La procedura ha avuto una durata media di 102 +/-39.1 minuti, con un prodotto dose area (DAP) medio di 0.2 +/- 0.15 Gycm2. In tutti i casi è stata usata una trascurabile dose di mezzo di contrasto (1cc/kg) in singola iniezione a mano, pre impianto (proiezione LL). In un paziente si è riscontrata un'insufficienza tricuspidale di grado lieve post-procedura, regredita completamente nel follow-up; in uno dei primi casi è stato opportuno ristudiare il paziente in giornata 0, per una sospetta protrusione del device verso l'arteria polmonare non confermata dall'angiografia. In un caso si è verificata una protrusione progressiva del disco polmonare, dopo 48 ore dalla procedura, con comparsa di stenosi dell'arteria polmonare sinistra. La bambina al tempo della procedura pesava 1500g, era affetta da ampio dotto (versante polmonare 3.7mm, versante aortico 5.1mm, lunghezza 7mm) ed era stata posizionata una protesi Piccolo 5-4. La bambina è stata ristudiata e sottoposta ad angioplastica dopo 6 mesi dall'impianto; non è stato necessario impianto di stent e la crescita dell'arteria polmonare sinistra è stata adeguata. La sopravvivenza dei soggetti a 30 giorni è del 100%. Con l'aumento dell'esperienza è stato possibile eseguire la procedura su pazienti sempre più piccoli. Si sottolinea l'importanza di utilizzare protesi lunghe 2mm in pazienti di piccolo peso (<2kg), riducendo quindi il rischio di protrusione nei grossi vasi. Riteniamo che sia importante inoltre non sovradimensionare la protesi anche come diametro specialmente nei prematuri di peso poco superiore al kg per rischio di compressione dell'arteria polmonare sinistra.



C38

MICROVESCICOLE CELLULARI E DANNO RENALE NEI PAZIENTI CON FISOPATOLOGIA DA FONTAN

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Scopi della Ricerca: La Circolazione di Fontan si associa ad un inevitabile deterioramento multiorgano progressivo, i cui meccanismi sono in gran parte ancora sconosciuti. Questo deterioramento presenta un'evoluzione insidiosa e si manifesta in modo evidente solo in fase avanzata. In particolare, il danno renale è ancora poco esplorato ed è divenuto oggetto di studi solo di recente.
Le Microvescicole sono, di fatto, frammenti cellulari. Sono costituiti da un involucro di membrana, che racchiude citoplasma, proteine e frammenti di RNA, e vengono rilasciate o come prodotti di scarto o come trasportatori di messaggi. La loro analisi fornisce importanti informazioni sullo stato del tessuto d' origine.
Nel nostro studio abbiamo analizzato le Microvescicole renali di origine urinaria, sia alla ricerca di marcatori di danno renale che per comprenderne meglio la natura nella Circolazione di Fontan.

Metodi impiegati: Abbiamo arruolato 32 pazienti già sottoposti a Fontan, tutti con Condotto Extracardiaco. Sono stati esclusi i pazienti con preesistenti nefropatie o quelli poi trapiantati per failure. Dieci pazienti sani di età paragonabile sono stati arruolati come gruppo di controllo.
Le Microvescicole renali sono state isolate dalle urine dei pazienti ed analizzate per confermarne l'origine renale e per esaminarne le caratteristiche, soprattutto marcatori di infiammazione e di staminalità. E' stato poi dosato anche il fattore Klotho: una proteina di recente scoperta, prodotta quasi esclusivamente dal rene e che, seppure con meccanismi ancora non chiari, presenta una funzione anti-invecchiamento tissutale.
E' stato inizialmente effettuato un confronto tra pazienti e controlli e, successivamente, un' analisi dei dati alla luce di un ampio set di variabili preoperatorie, intraoperatorie, postoperatorie e di follow-up.

Risultati e Conclusioni: Infiammazione e staminalità renali sono risultate correlate tra loro inversamente.
A livello renale, i pazienti Fontan presentavano un evidente stato infiammatorio ed una ridotta capacità rigenerativa tissutale rispetto ai controlli sani. Anche se in modo eterogeneo, verosimilmente per la sua genesi multifattoriale, era evidente un'associazione tra l' entità del danno renale ed alcune delle varibili considerate: l'età alla Fontan, la pressione media nel condotto, le perdite dai drenaggi nel postoperatorio, la saturazione, la stiffness epatica ed il tempo trascorso dalla Fontan.
Klotho merita un'osservazioni a parte: nei controlli sani, come atteso, presentava livelli molto elevati; nei pazienti Fontan, al contrario, risultava ridotto nel preoperatorio e quasi completamente indosabile già in prima giornata.

In Conclusione:
- Le alterazioni dei markers infiammatori e di staminalità delle Microvescicole renali testimoniano un danno tissutale significativo ed in costante progressione.
- Il danno renale, in parte già legato alla cardiopatia in sé, verrebbe poi aggravato dalla Fontan.
- la Fontan sembra "spegnere" quasi totalmente la produzione di Klotho.
- le Microvescicole renali di origine urinaria, oltre che di facile reperimento, sembrano molto promettenti nell'indagine del danno renale da Fontan, anche in fase precoce.
- I markers infiammatori e di staminalità sembrano chiarire alcuni dei meccanismi responsabili del danno sia renale che di altri tessuti.
- La quasi assenza di Klotho, considerata la sua funzione anti-invecchiamento tissutale, potrebbe giustificare molti aspetti delle alterazioni multiorgano oggi ormai note nei pazienti Fontan.

C39

PREDICTIVE ECHOCARDIOGRAPHIC FEATURES OF PERSISTENT DUCT-DEPENDENCY AFTER TRANS-CATHETER TREATMENT OF PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM OR CRITICAL PULMONARY STENOSIS

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Objectives: This study aimed to identify clinical, hemodynamic or echocardiographic predictive features of persistent duct-dependency of pulmonary circulation (PDDPC) after effective percutaneous relief of pulmonary atresia with the intact ventricular septum (PA-IVS) or critical pulmonary stenosis (CPS).

Methods: From 2010 to 2021, 55 neonates with PA-IVS or CPS underwent percutaneous right ventricle (RV) decompression at our Institution. After successfully relief of critical obstruction, 27 patients (Group I) showed PDDPC, whereas RV was able to support the pulmonary circulation in the remaining 28 patients (Group II). Clinical, hemodynamic, and echocardiographic features of these two groups were compared.

Results: No significant difference in clinical and hemodynamic data was found between the groups, although the group I had a lower oxygen saturation at hospital admission. However, tricuspid valve (TV) diameter <8.8 mm, TV z-score <-2.12, tricuspid/mitral valve annular ratio <0.78, pulmonary valve diameter <6.7 mm, pulmonary valve z-score <-1.17, end-diastolic RV area <1.35 cm2, end-systolic right atrium area >2.45 cm2, percentage amount of interatrial right-to-left shunt >69.5%, moderate/severe tricuspid regurgitation, RV systolic pressure >42.5 mmHg, tricuspid E/E' ratio >6.6 showed each significant predictive value of PDDPC. These parameters were used to build a composite echocardiographic score (PDDPC-score), assigning one point each above the respective cut-off value. A score ?4.00 showed high sensitivity (100%) and specificity (86%) in predicting PDDPC.

Conclusion: Clinical and hemodynamic features fail to predict the short-term fate of the pulmonary circulation after successful treatment of PA-IVS/CPS. However, a simple, composite echocardiographic score is useful to predict PDDPC and could be crucial in the management of this frail subset of patients.

C40

THE IMPACT OF DOMINANT VENTRICLE MORPHOLOGY AND ACCESSORY VENTRICLE SIZE ON CLINICAL OUTCOMES AND FUNCTIONAL STATUS IN PATIENTS WITH FUNCTIONAL UNIVENTRICULAR HEARTS

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Objectives: the physio-pathological role of the morphological ventricular dominance (left, FSLV; right, FSRV), and the hemodynamic contribute of an accessory ventricular chamber (AVC), in patients with functional single ventricle (FSV) after Fontan operation are still uncertain due to conflicting data. We analyzed a cohort of Fontan patients to assess and correlate such anatomical features to late clinical outcomes.

Methods: We enrolled all patients after a Fontan procedure who underwent a cardiac magnetic resonance (CMR) and a cardiopulmonary exercise test (CPET) in the previous 3 years. Clinical, CMR and CPET data from the last follow-up visit were retrieved to analyze whether the size of any AVC and the morphological ventricular dominance (FSLV vs FSRV) were correlated with clinical outcomes (NYHA, need for reinterventions or cardiac transplantation, mortality, arrhythmias, liver disease and protein-losing enteropathy) and functional parameters (including FSV ejection fraction and presence of late gadolinium enhancement, LGE, on CMR and peak metabolic equivalents, pMETs, and peak oxygen consumption, pVO2, at CPET). All statistical tests were two tailed and significance was set at 0.05.

Results: we enrolled 50 patients: 29 had FSLV (58%), FSRV in 21 (42%). Median age at evaluation was 19.5 years [IQR 15-26]; median follow-up was 16 years [4-42]. NYHA class III or IV was present in 6%, while 4 (8 %) underwent a re-do Fontan, 2 (4%) entered transplantation waiting list and one of these received a transplant. 2 patients (4%) died at follow-up. Statistical analysis showed that the accessory chamber was larger (>20cc/m2) in FSLV than in FSRV ($p = 0.01$). In the post-operative period, FSRV was associated with higher incidence of low-cardiac output syndrome ($p = 0.043$). In the long-term, there was no statistically significant difference in major clinical outcomes or NYHA class between the two groups. FSLV was associated with a better cardiac function (median FSV ejection fraction 56% vs 52%; $p = 0.041$), less extent of LGE [$p = 0.022$], better functional capacity expressed by METs (14.1 vs 12.3; $p= 0.01$) and pVO2 (1.625
ml/min vs 1.233 ml/min; $p = 0.033$). An AVC was detected in all: its size was <5ml/m2 in 31%, 5-20ml/m2 in 47%, and >20ml/m2 in 22%. A larger AVC was associated with higher need for postoperative ECMO support ($p = 0.007$), while size of AVC was not associated to any statistically difference in clinical outcomes, cardiac function and functional capacity.

Conclusions: In Fontan circulation, a FSLV is correlated to better clinical and functional outcomes when compared to FSRV. On the other side, an AVC appears to be not significantly related to any clinical disadvantage. However, the immediate postoperative course may be influenced negatively by the presence of a larger AVC.

C41

SPECKLE-TRACKING ANALYSIS OF FETAL LEFT VENTRICULAR FUNCTION IN SHONE'S COMPLEX

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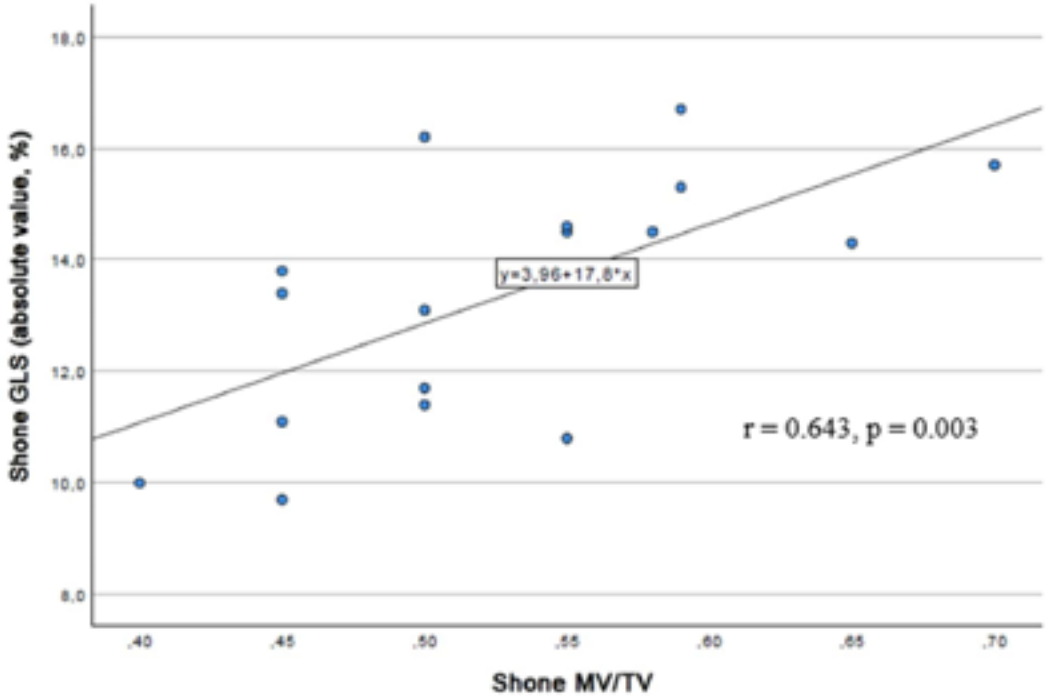
Background and Aim: Shone's Complex is a congenital heart disease characterized by sequential obstructions of left ventricular (LV) inflow and outflow. It can require multiple interventions with a poor long-term prognosis. Prenatal diagnosis is challenging and diagnostic suspicion is raised for the presence of borderline left ventricle with small aortic annulus, similarly to aortic coarctation (CoA). Recent studies have demonstrated LV contractility depression in fetuses with CoA, but assessment of LV function in those with Shone Syndrome has not yet been conducted.
The aim of this study is to assess LV function in fetuses with postnatally confirmed diagnosis of Shone Syndrome using 2D speckle-tracking echocardiography (2D-STE) comparing with fetuses with postnatally confirmed simple aortic coarctation (S-CoA) and those with false-positive aortic coarctation (FP-CoA).

Methods: From January 2010 to December 2019 we retrospectively analyzed the four-chamber view (4CV) of 73 consecutive fetuses referred for suspected CoA and 30 control cases. 2D-STE was used to calculate LV global longitudinal strain (GLS), global circumferential strain (GCS), strain rate (SR) and ejection fraction (EF). Those data were correlated to mitral valve diameter/ tricuspid valve diameter ratio (MV/TV).

Results: Postnatally Shone's Complex was diagnosed in 15 fetuses (20.5%), CoA in 24 fetuses (33%) and FP-CoA in 34 fetuses (46.5%). In fetuses with Shone's Complex GLS (mean -13.2%, SD 2.2, $p < 0.001$), SR (mean -1.1 s?1, SD 0.15, $p < 0.001$) and EF (mean 47.1%, SD 5.1, $p = 0.03$) were significantly reduced compared to fetuses with S-CoA. FP-CoA showed normal LV function parameters but GLS that was significantly reduced compared to controls ($p = 0.01$). Univariate linear regression showed positive correlation between MV/TV and GLS ($p = 0.02$, R2 0.3) in Shone's Complex fetuses.

Conclusions: Fetuses with post-natal diagnosis of Shone's Complex showed markedly depressed LV function compared to S-CoA fetuses. We speculated that this could be related to the same genetic messages causing the anatomical abnormalities. To our knowledge this is the first study describing significant LV dysfunction in fetuses with post-natal diagnosis of Shone's Complex. These data may improve the parental counselling, being the post-operative outcome in patients with Shone's Complex significantly more severe than in those with S-CoA.



C42

ACCURACY OF ECHOCARDIOGRAPHY IN PRENATAL DIAGNOSIS IN DOUBLE OUTLET RIGHT VENTRICLE

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Objective: Double outlet right ventricle (DORV) is a complex conotruncal cardiac malformation represents 3% of congenital heart disease during fetal life, less than 1% of all congenital heart disease. Prenatal diagnosis has high accuracy assessed by fetal echocardiography. Conotruncal malformations are difficult to dignose in fetal life. Recently literature has proved that proper fetal diagnosis contents parental counseling in order to clearly define prognosis and postnatal surgical approach.

The aim of this study was to determine whether improvements in fetal echocardiography can result in increased accuracy of diagnosis of DORV.

Design: Medical records of patients with fetal echocardiography diagnosis of DORV at our institution from 2009 to 2022 were analysed. Pre and post natal diagnosis were matched. We assessed the agreement between fetal echocardiography and postnatal evaluation in determining VSD location, great artery orientation and size, and ventricular size. Neonatal outcome was determined.

Results: The median gestational age at first fetal study at our institution was 21.3 weeks of gestation (range: 16-38 weeks); in our maternal population the incidence of gestational diabetes was 14.3%. There were 0 in utero deaths and 2 termination of pregnancy (4.8%). Among the 41 fetuses with prenatal diagnosis of DORV: 22% had ventricular hypoplasia: in the most of cases with hypoplastic left ventricle; 32% had extracardiac abnormalities and 17% had chromosomal anomalies or a define syndrome (genetic test was performed only in 57%). Heterotaxy syndrome was seen in 5 % of patients. Postnatal diagnosis was confirmed in 90% of cases. The overall accuracy of fetal diagnosis was 83%; 34 of the 41 patients with postnatal evaluation available were correctly diagnosed with DORV prenatally. Among live births, survival was 62% in patients with extracardiac or chromosomal anomalies, compared with 77% in infants without additional anomalies. Four of the 7 patients who died had single ventricle lesions and extracardiac malformations or chromosome abnormalities.

Conclusion: DORV is a rare and often complex cardiac anomaly that can be diagnosed prenatally with high precision. Advances in the last decade in fetal echocardiography and genetic diagnosis may improve the management of future DORV with the goal of providing more accurate and tailored counseling allowing better management for families.

C43

FETAL DIAGNOSIS OF RIGHT AORTIC ARCH AND ABERRANT LEFT SUBCLAVIAN ARTERY: A METHOD FOR POSTNATAL OUTCOME PREDICTION

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Aims: to investigate isolated right aortic arch,left ductus arteriosus and aberrant left subclavian artery (RAA-ALSA)in a quantitative mode: risk stratifying fetuses prenatally and following outcome postnatally.

Methods: the fetal echocardiograms of 41 fetuses with isolated RAA-ALSA carried out between 19 and 37 weeks , period January 2016 and May 2022 , were retrospectively reviewed . Two mesaurementes were studied: the AO-DA angle (angle between the lines bisecting the blood flow of the DA and AO), and the AO-DA distance (distance between the inner boundaries of the AO and the DA)Measurement were taken at the trachea level, all obtained at end sistole. These mesaurements were obtained in the 2° and 3° trimester in 20 fetuses which consist of our study group, in 15 fetuses data were incomplete , 3 are still in follow up ,3 in utero, 1 died in utero due to multiple extracardiac malformations. .

Results: in 20 fetuses diagnosed with RAA-ALSA mesaurements were obtained in the 2° and 3° trimester ,all showed a significant increse in mean AO-DA distance (3,56+/-1.07 mm) respect to normal left arches (1.6+/-0.33 mm) as reported in literature . Comparison of values between a mean AO-DA distance of 3.58 (+/-0.64mm) in 2° trimester with 4 (+/-0.90mm) in the 3° trimester stratified group 1 (10 fetuses) in whom there is a minimal increase of the distance while proceding from 2° to 3° trimester , indicating that the trachea and esofagus are encircled in a ring which is progressively smaller . In contrast, in group 2 (10 fetuses), mean AO-DA distance of 3.34 (+/- 0.44mm) in 2° trimester increased to 8.45 ((+/-1.0mm) in the 3° trimester stratifyig fetuses with a significant less restriction of the ring while the pregnancy progresses to the 3° trimester . The mean difference of the growth 0.39 ((+/-1.05mm) in group 1 compared to 5.1 (+/-0.78mm) in group 2 is statistically significant ($p<0.001$). The AO-DA angle, described in normal fetuses in literature, ranges around 21-22° , in these fetuses with RAA-ALSA due to the parallel alignement of the aortic and ductal arches is significantly lower: it mesured in 2° and 3 trimester,respectively, 2.8°(0.67) to 3.1°(0.68) group 1 and 3.5°(1.4) to 3,4°(0.9) group 2, indicating not significant difference (p 0.56).Postnatal transthoracic echocardiogram confirmed the diagnosis in all newborn .Follow-ups were conducted to obtains the outcomes after delivery The overage follow-up time was 12 months (range 10 days - 36 months).The clinical outcome was symptomatic in 11 (mean age 2 months: range 13 days-6 m): 7 with dysphagya at the ages of 2-11 months , 4 with inspiratory stridor.Postnatal CT scan were performed in 7 and MRI in 4 prior to surgical division of the vascular ring. Patients that required surgical repair of the vascular ring had fetal AO-DA distance that hardly increased or even decreased, as in group 1, meaning a restriction of the circle around esophagus and trachea as the pregnancy progresses. .Asintomatic patients had a much better increase of AO-DA distance ,as in group 2.

Conclusions: Fetal AO-DA distance may help stratifying fetuses with RAA-ASLA . This study supports the hypothesis that fetal AO-DA distance minimal increase or even decrease , during pregnacy progression , may imply compressive vascular ring and assumes clinical value in predicting postnatal risk for intervention. Moreover migth be helpful in counselling,adding more precise anatomical details. Neverthess,the effectiveness of this parameter needs further investigation in larger studies.

C44

ANTIARRHYTHMIC THERAPY IN FETUSES WITH DIAGNOSIS OF TACHYARRHYTHMIA: A TERTIARY REFERRAL CENTER EXPERIENCE

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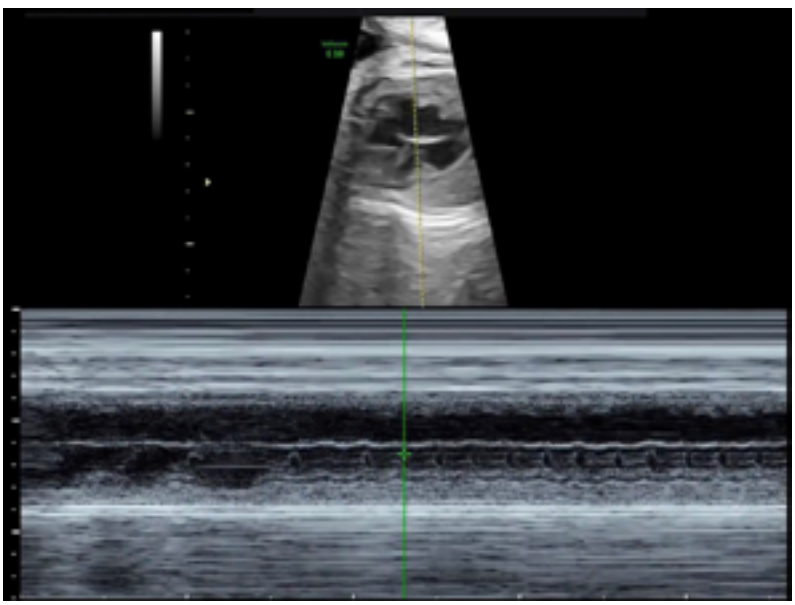
Introduction: Fetal tachyarrhythmias represent a serious condition, potentially leading to heart failure, hydrops fetalis and fetal demise. The aim of this study was to evaluate the impact of early maternal administration of antiarrhythmic therapy on the fetal outcome at our tertiary referral center.

Method: We analyzed data from 114 fetuses with diagnosis of fetal arrhythmia referred to our Paediatric Cardiology Unit between January 2014 and December 2021. We selected fetuses with diagnosis of tachyarrhythmia (n = 30, 26%), including 15 fetuses with atrioventricular reentrant tachycardia, 7 fetuses with ectopic atrial tachycardia, 5 fetuses with atrial flutter, 2 fetuses with ventricular tachycardia and 1 fetus with permanent junctional reciprocating tachycardia. Fetal echocardiography included a complete evaluation of cardiac function to identify signs of hemodynamic impairment. Detailed analysis of fetal heart rhythm was performed using M-Mode, pulsed wave Doppler and tissue Doppler imaging. Eight out of 30 fetuses (27%) presented with signs of heart failure due to tachycardia. Six fetuses (20%) were hydropic because of nearly incessant tachycardia. Oral transplacental antiarrhythmic therapy using flecainide, digoxin or sotalol in single or combination therapy was started in 25 cases as soon after diagnosis (mean gestational age 27 ± 5 weeks), according to the type of arrhythmia.

Results: We observed a good rhythm control in 24/25 cases (96%) with only one non-responder (intrauterine death). Rhythm was completely reverted to sinus in 17 fetuses (68%); in other 7/25 cases (28%), we obtained a partial rhythm control with reduction of recurrences and/or reduction of length of arrhythmia episodes and/or reduction of heart rate during tachycardia. Five out of the 30 patients were not treated: one was lost to follow-up, while in the other 4 cases the mothers were directed straight to caesarean section. Twelve out of 14 fetuses with heart failure or hydrops were treated. Among them, 10/12 patients (83%) achieved complete resolution of the hemodynamic impairment thanks to complete or partial rhythm control. One fetus gained hemodynamic improvement without complete resolution. Therapy failed in one case (intrauterine death). No significant adverse effect requiring drug discontinuation occurred in all mothers. Twenty-five newborns with fetal diagnosis of tachycardia (25/30) were transferred from the birth centers to our Unit in the first days of life. We observed complete resolution of the tachyarrhythmia in 11/25 newborns (44%), with no spontaneous or induced (during transoesophageal electrophysiologic study) arrhythmia episodes during hospitalization. Antiarrhythmic therapy was restarted in the other 14 newborns.



Conclusion: In our population, early oral maternal antiarrhythmic therapy resulted to be safe and effective in fetuses with tachyarrhythmias to obtain a good rhythm control and resolution of fetal heart failure and hydrops fetalis.

C45

SHORT-TERM OUTCOME AND MORBIDITY OF TRANSPOSITION OF GREAT ARTERIES: PRENATAL COUNSELING IMPLICATIONS

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Objective: From to the advent of the Arterial Switch Operation (ASO), the outcome of patients suffering from Transposition of Great Arteries (TGA) simple-type has radically changed, resulting in a substantial change in the prenatal counseling of this congenital heart disease (CHD). We analyzed the experience of our group regarding the accuracy in the diagnosis of simple and complex TGA in prenatal life. The goal of our study is to understand how to improve fetal counseling by examining the accuracy of our prenatal diagnoses and the short-term outcome of patients.

Methods: From October 2017 to May 2022, 65 consecutive fetuses evaluated in our referral center with a prenatal diagnosis of simple and complex TGA types were identified. Prenatal diagnostic accuracy and short-term survival were analyzed for the different types of TGA. Patients with a physiopathology of TGA were included in the group of simple TGA, even with the presence of restricted Ventricular Septal Defect (VSD).

Results: The TGA type was correctly ascertained prenatally in 92.3%. Most fetuses were diagnosed with simple TGA (78.5%). There were 5 discrepancies: two fetuses with prenatal diagnosis of simple TGA had postnatally TGA + VSD in one case and TGA + Coarctation of the Aorta (CoAo) in the other one. Besides, 1 fetus with prenatal diagnosis of TGA + VSD presented postnatally multiple VSDs, CoAo and pulmonary stenosis (PS), 2 fetuses with suspected differential diagnosis between Double Outlet Right Ventricle (DORV) + TGA and TGA + VSD had a postnatal diagnosis of TGA + VSD + CoAo (n=1) and TGA + VSD + PS (n=1). Mean age at Arterial Switch Operation (ASO) was 8.7 ± 4.6 days. Fifteen patients (23%) have been submitted at least one reintervention (surgery or angioplasty). Among them, 73% were reoperated within the third month of life. All four patients (100%) who underwent more than 1 reoperation were affected by complex TGA. The mortality rate was 1.5% (1 pt.) and the cause of death was due to a metabolic disease being defined and not related to CHD or surgery complications.

Conclusions: Simple TGA has a better outcome than the complex forms. We found a discrepancy rate of 7.7% with potential influence on the prognosis of morbidity between the prenatal diagnosis of the TGA type and the definitive diagnosis. Although the diagnostic accuracy by means of fetal echocardiography is extremely high for patients with TGA, the counseling for patients with suspected complex TGA still remains a challenge . Indeed, in this type of TGA, the details that influence the surgical approach and outcome are numerous and tricky to identify during the fetal life.

C46

OUTCOMES AFTER PRENATAL DIAGNOSIS OF FUNCTIONALLY UNIVENTRICULAR HEART

V. Gesuete [1], A. Balducci [1], V.F. Mangerini [2], E. Montaguti [3], C. Ciuca [1], A.G. Quarti [2], F.D. Petridis [2], T. Hasan [1], G. Egidy Assenza [1], A. Perolo [3], P. Pilu [3], L. Ragni [1], G.D. Gargiulo [2], A. Donti [1]

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Background: Despite improved surgical techniques, preoperative and postoperative care, patients with functionally univentricular hearts (UVH) continue to have high morbidity and mortality and a decreased lifespan. The purpose of our study was to determine outcomes after prenatal diagnosis of functionally UVH.

Materials and Methods: We identified all patients with a prenatal diagnosis of functionally UVH presenting between 2000 and December 2021 at our institution. Maternal data, fetal characteristics and data from the postnatal clinical course were collected. Continuos variables are reported as mean ± standard deviation (SD) or as median (range). Categorical data are expressed as frequencies (n) and percentages.

Results: There were 139 patients with a prenatal diagnosis of functionally UVH. The mean gestational age at diagnosis was 20 weeks (range 14-35 weeks) and the mean maternal age was 33,7 years ± 5,6 years (range 20 - 47). 35 (25%) underwent termination of pregnancy. 14 (10%) were lost at follow-up. 90 born alive. Of these, 49/90 (54%) were diagnosed with dominant right ventricle and 41/90 (46%) were diagnosed with dominant left ventricle. The mean gestational age at delivery was 38 weeks (32 – 40 weeks), the mean birth weight was 2900 g ± 600 g (1800 - 4200 g). Preterm delivery before 37 + 0 weeks was reported in 11 (12%) cases, 3 deliveries occurred < 34 + 0 weeks. There were 16 tricuspid valve atresia (TA) (18%), 12 double inlet left ventricle (DILV) (13%), 10 unbalanced atrioventricular septal defect (AVSD) (11%), 6 pulmonary valve atresia and intact ventricular septum (PA:IVS) (6%), 25 hypoplastic left heart syndrome (HLHS) (28%) and 21 double outlet right ventricle (DORV) (24%). In 8 (9%) chromosomal anomalies were identified prenatally, including one case of Turner syndrome, one case of Trisomy 18, and 3 cases of Trisomy 21. In one patient, parents decided for compassionate care.The ages at staged surgery were: initial palliation 27 days ± 0,1 (range 0 – 302 days), cavopulmonary shunt 5,6 months ± 0,89 (range 3,7 – 30 months) and Fontan 6,5 years ± 1,9 (range 1,6 – 11,2 years). One patient underwent heart transplantation at the age of 3 months. Of the 89 neonates with intention-to-treat, 88 (99%) survived 30 days postnatally. The median follow-up time was 9,3 years ± 6,3 and the 1, 5 and 10-year survival rates after initial palliation were 87%, 71% and 50%.

Conclusions: Prenatal diagnosis is the most common and desired path of entry into the system of care for patients with UVH. This is a retrospective longitudinal study that characterizes the outcomes of fetuses with UVH. These results should be explained to parents to ensure informed decisions and counselling.

CB1

NORMAL LUNG ULTRASOUND FINDINGS IN HEALTHY CHILDREN AND IN THOSE WHO HAD COVID-19: DOES COVID-19 LEAVE PULMONARY DAMAGE IN CHILDREN?

P. Marchese [1], N. Assanta [2], A. Pizzuto [2], E. Franchi [2], G. Corana [2], C. Viacava [2], S. Garibaldi [2], G. Santoro [2], L. Gargani [2], M. Cantinotti [2]
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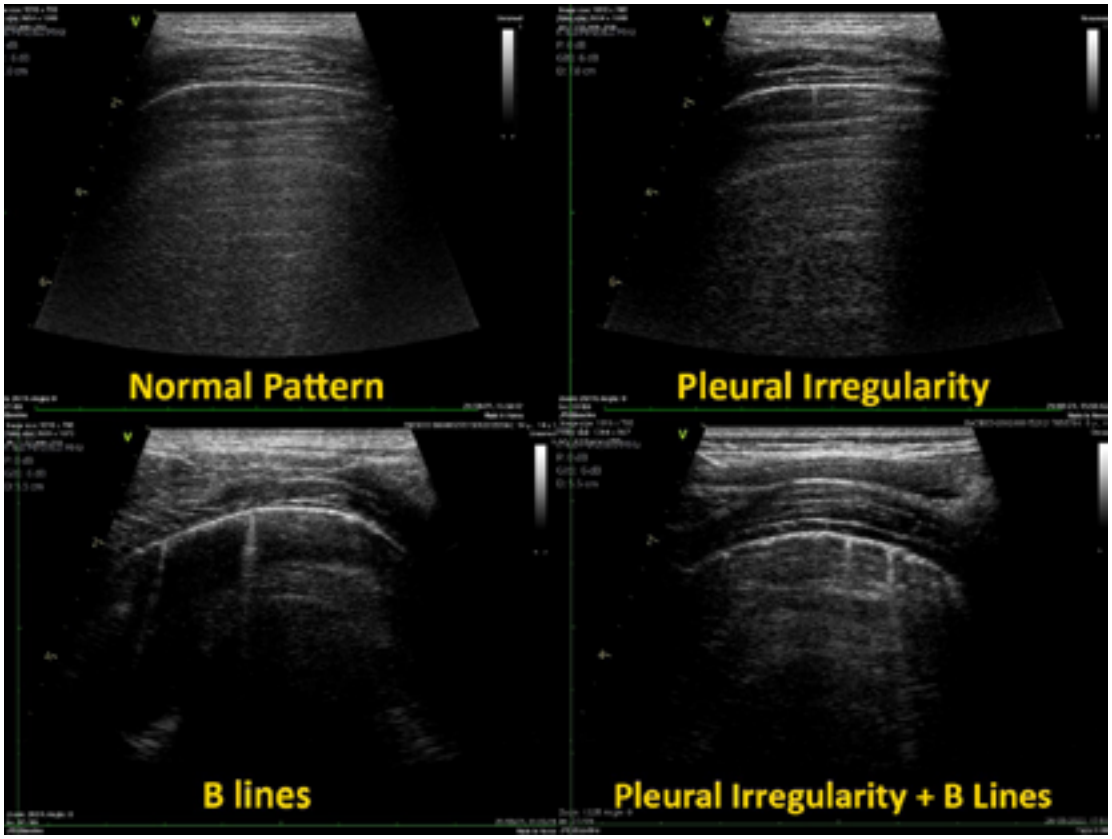
Background: Lung ultrasound (LUS) is gaining consensus as a non-invasive diagnostic imaging method for the evaluation of pulmonary disease in children. This study aims to clarify the importance of artefacts (e.g., B-lines, pleural irregularity) that can be found in children and to compare patients who did not experience Covid-19 and those with previous COVID-19.

Methods: LUS was performed according to standardized protocols. Different patterns of normality were defined: Pattern-1: no plural irregularity, no B-lines; Pattern-2 only mild basal posterior pleural irregularity, no B-lines; Pattern-3 mild posterior basal/para-spine/apical pleural irregularity, no B lines; Pattern-4: like 3 plus rare B-lines; Pattern-5: mild diffuse short subpleural vertical artefacts, rare B-lines; Pattern-6: mild diffuse short subpleural vertical artefacts, limited B-lines; Pattern-7: like pattern 6 plus minimal subpleural atelectasis. Coalescent B-lines, consolidations or effusion were considered pathological.

Results: 459 healthy children were prospectively recruited (mean age 10.564±3.839 years). Children were divided into two groups: Group-1 (n=336), those who had not COVID-19 infection, and Group-2 (n=123) those who experienced COVID-19 infection. Children with previous Covid-19 had a higher value of LUS-score than those who had not (p 0.0002). Children with asymptomatic COVID-19 had a similar LUS-score to those who did not have infections (p>0.05), while those who had symptoms showed a higher LUS-score than those who had no symptoms (p=0.0228).

Conclusions: We report the pattern of normality for LUS examination in children. We also showed that otherwise healthy, who recovered from Covid-19, even mildly symptomatic, had more "physiological" artefacts at LUS examinations.



CB2

LEFT VENTRICLE VORTEX PROGRESSION DURING THE DIASTOLIC PHASES IN HEALTHY CHILDREN: A DEEPER INSIGHT INTO PEDIATRIC DIASTOLOGY BY NEW HIGH FRAME RATE BLOOD SPECKLE TRACKING ECHOCARDIOGRAPHY

P. Marchese [1], N. Assanta [2], E. Franchi [2], G. Corana [2], C. Viacava [2], A. Pizzuto [2], S. Garibaldi [2], G. Santoro [2], M. Cantinotti [2]
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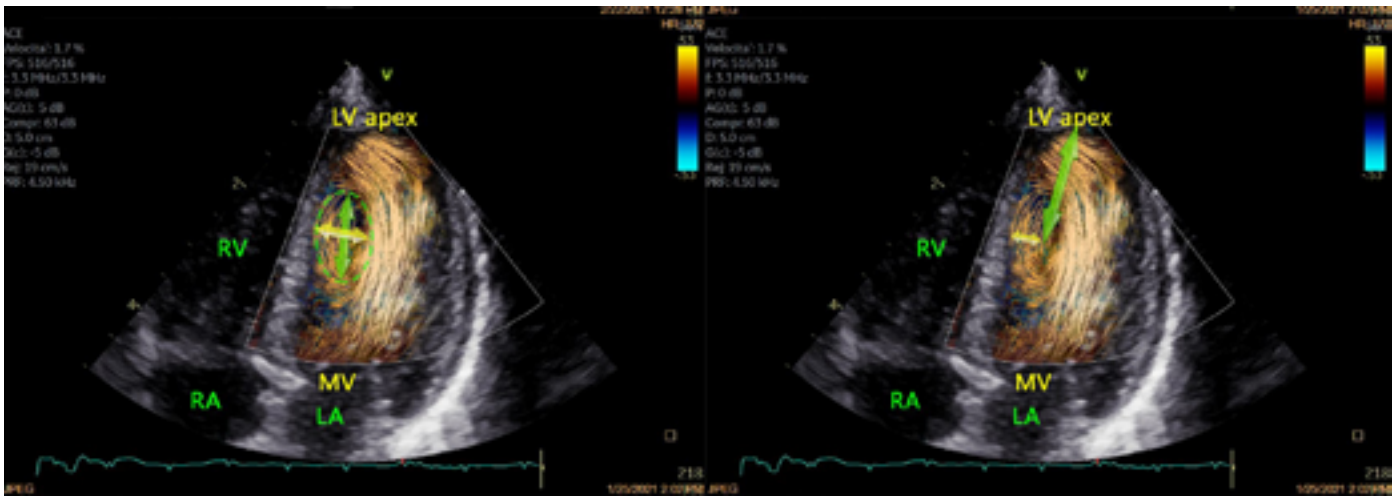
Background: High-frame rate blood speckle tracking (BST) echocardiography is a new technique for the vectorial visualization of intracardiac and intravascular blood flow. Intraventricular vortical flow is well known but poorly studied in the paediatric population. The purpose of this study is to evaluate the characteristics of left ventricular (LV) vortices in healthy children during the diastole.

Methods: Characteristics of LV vortices were analysed based on 4-chamber BST images from 50 healthy children (median age 4.96 years, range 0.06-13.39 years); for each patient, three vortices were analysed along with the entire diastolic phase (early, mid, and late diastole), defining them according to the ECG (after the T wave, during the P wave and during the QRS complex). Multiple linear regression was used to identify factors that independently influence vortex characteristics. Vortices characteristics were all indexed by BSA to overcome age differences.

Results: 50 subjects with a vortex for each of the diastolic phases (early, mid and late diastole) were prospectively enrolled, Vortices in the late diastole were significantly bigger than those in the other diastolic phases. Mean late diastolic vortex height, width and area were 16.19±8.98 mm/m2, 11.48±5.24 mm/m2 and 1.01±0.59 cm2/m2, respectively and they were all bigger than in other diastolic phases (p all <0.05): No differences in vortex height position within the LV were highlighted during the various diastolic phases, but the vortex tended to occur significantly closer to the interventricular septum during the early diastole (0.29±0.11 mm/m2, p=0.01) and then migrated toward the centre of the ventricle during the mid and late diastole, without differences among them (0.34 ±0.1 mm/m2 and 0.35 ±0.08 mm/m2, p=0.6)

Conclusions: We provided a complete analysis of vortex progression over the different diastolic phases in healthy children. Since vortices are known as reservoirs of kinetic energy, the increase in the size of the vortices seems to reflect a constant increase of potential energy through all the diastole. At the end of diastole, the storage of kinetic energy is completed, and the heart is ready for the systole. These data may serve as a baseline for the study of diastolic mechanics in children.



CB3

LEFT VENTRICULAR VORTEX FORMATION TIME, A NEW DIASTOLIC INDEX: COMPARISON AMONG HEALTHY NON-SPORTY CHILDREN AND JUNIOR ATHLETES

P. Marchese [1], N. Assanta [2], C. Viacava [2], E. Franchi [2], G. Corana [2], S. Garibaldi [2], A. Pizzuto [2], G. Santoro [2], M. Cantinotti [2]
[1] *Institute of Life Sciences, Scuola Superiore Sant'Anna, Pisa, Italy*
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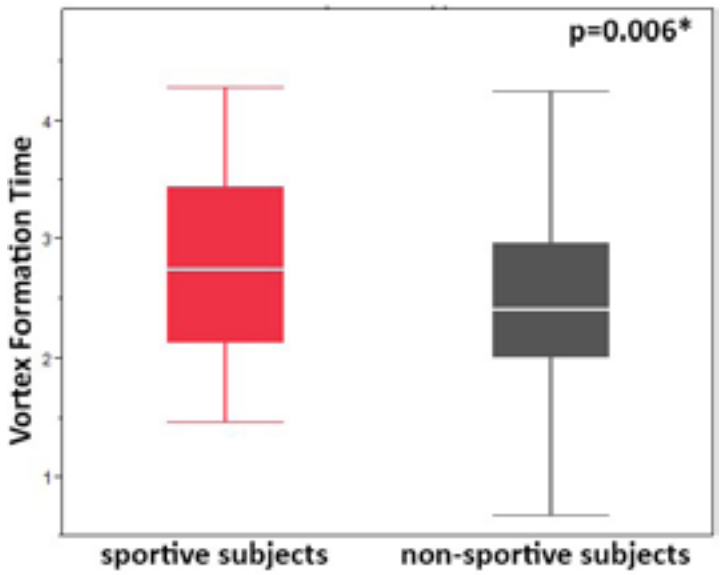
Background: Vortex formation time (VFT) is a continuous measure of the left ventricular (LV) filling that integrates all phases of diastole. Although VFT has been analysed in heart failure, there are currently very few studies in paediatric patients. The aim of this study is to compare VFT in healthy non-sportive children and in young athletes.

Methods: Healthy Caucasian children (<18 years old) were prospectively recruited from May 2021 to February 2022 in the outpatient Pediatric Cardiology Department at the Fondazione CNR Regione Toscana G. Monasterio of Massa. All measurements were gathered from the Apical 4 chamber view. VFT was obtained using the validated formula: 4×(1??)/?×?3×LVEF, where ? is the fraction of total transmitral diastolic stroke volume contributed by atrial contraction (assessed by time velocity integral of the mitral E- and A-waves) and ? is the biplane end-diastolic volume (EDV)1/3 divided by mitral annular diameter during early diastole. Diastolic function was measured by the ratio of mitral peak velocity of the early filling (E) to early diastolic mitral annular velocity (e?) (E/e? ratio) and the ratio of E to mitral peak velocity of the late filling (A) (E/A ratio).

Results: 48 young athletes (15 ±1.9 years, range 12-17.8 years) and 48 healthy age-/BSA-matched sedentary controls (13.4 ±3.1, range 14-17 years) were prospectively enrolled. Junior athletes had lower heart rate (70.15±10.5 bpm Vs 80.5±15.8 bpm, p<0.0001). Although no differences were found in both diastolic and systolic diameters, junior athletes reported higher systolic and diastolic volumes compared to non-sportive adolescents (p=0.0002 and p<0.0001) resulting in a higher stroke volume (72.7±17.9 Vs 57.5±17.24, p<0.0001 while no differences were noted in LV ejection fraction (p=0.09). While no clear differences in all conventional parameters of diastolic function (e.g. E, A, E/A, E/e', p…) were found between the two groups, VFT values were significantly higher in athletes than in non-sporty controls (2.72 ±0.76 Vs 2.31 ±0.54, p=0.006). A modest correlation between VFT and E/A (r=0.43, p=0.0015), E/e' (r=0.24, p= 0.0025) and GLS (r=0.046, p=0.048) was noted.

Conclusions: VFT is a novel measure of left ventricular filling efficiency and was increased in young athletes compared to age-/BSA-matched non-sporty healthy children. This seems to indicate that diastole is more efficient in children and adolescents with a trained heart. This data may serve as a baseline for future studies on diastolic efficiency in junior athletes with suspicion of a cardiac defect and in children with congenital and acquired heart disease.



CB4

HIGHER LEFT VENTRICULAR EFFICIENCY IN JUNIOR ATHLETES THAN NON-SPORTY-ADOLESCENTES REVEALED BY NEWLY ECHOCARDIOGRAPHIC MYOCARDIAL WORK INDEX: A NOVEL METHOD FOR ASSESSMENT OF MYOCARDIAL PERFORMANCE

P. Marchese [1], N. Assanta [2], S. Garibaldi [2], C. Viacava [2], G. Corana [2], E. Franchi [2], A. Pizzuto [2], G. Santoro [2], M. Cantinotti [2]
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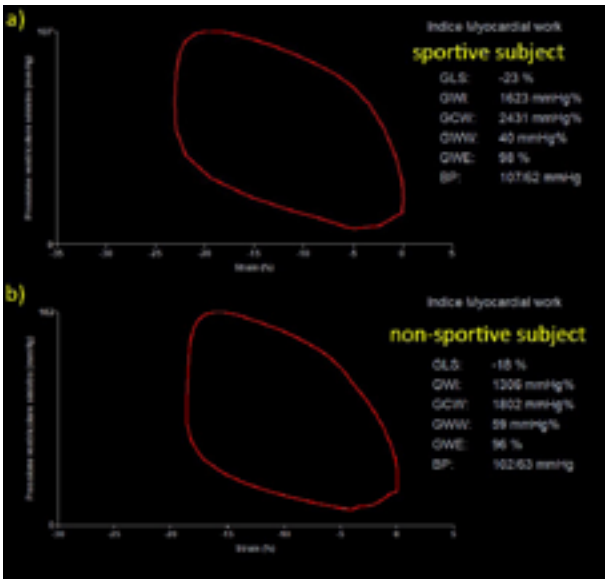
Background: Left ventricular hypertrophy (LVH) in the sportive patient is still a matter of controversy since it is difficult to understand the border between physiological response to exercise and pathological changes. Myocardial Work (MW) is a recently emerged non-invasive method for evaluating myocardial performance by coupling longitudinal strain analysis with noninvasive blood pressure measurements. This study aims to assess the role of LVH in junior athletes.

Methods: Echocardiographic information from healthy paediatric patients was collected from August 2020 to April 2022. Ventricular thickness information was collected from short- and long-axis parasternal views. The ejection fraction was calculated using the biplane Simpson method. The Speckle Tracking Echocardiography (STE) analysis was performed offline directly on the ultrasound machine, based on apical 4,3 and 2 chamber views. Myocardial work indexes derived by the integration of LV STE with systemic blood pressure measured noninvasively

Results: 53 junior athletes (15 ±1.9 years, range 12-17.8 years) were selected and compared with 53 healthy non-sporty age-/BSA-matched adolescents (13.5 ±2.9, range 14-17 years). Athletes had increased interventricular septum (9.8 ±1.6 mm Vs 8.1 ±1.7 mm, p<0.0001) and posterior wall (8.6 ±1.4 mm Vs 7.4 ±1.5 mm, p<0.0001) thicknesses compared to non-sportive subjects, and increase in ventricular mass (160.4 ±41.2 gr Vs 122 ±44.8 gr, p<0.0001). Although no differences were found in both diastolic and systolic diameters, junior athletes reported higher systolic and diastolic volumes (p=0.0002 and p<0.0001), and higher stroke volume (72.7±17.9 Vs 57.5±17.24, p<0.0001)compared to non-sportive adolescents. While no differences were noted among athletes and sporty in LV ejection fraction (p=0.54) junior athletes, had significantly higher values of global longitudinal strain (19.8±1.9% Vs 19.1±1.5%, p=0.04) Global Work Index (1721.9±271.9 Vs 1581.2±231.1, p=0.007) and for Global Constructive Work (2222.8±312 Vs 2093.6±226.6, p=0.02) than non-sporty subjects.

Conclusions: Our indicate that junior athletes have an increased systolic heart performance than non-sport adolescents. While conventional markers to evaluate systolic function (e.g. LV ejection fraction) were unable to unmask these differences, they become evident with novel indices including by speckle tracking echocardiography (e.eg. LV strain and myocardial work). This study seems to demonstrate how the process of LVH in response to training during adolescence does not impair heart function but makes it more efficient.



CB5

FAST-TRACK VIRTUAL REALITY TO FACILITATE 3D RECONSTRUCTION IN CONGENITAL HEART DISEASE

F. Bertelli [1], F. Raimondi [2], C. Godard [3], E. Bergonzoni [1], C. Cattapan [1], E. Gastino [1], F. Galliotto [1], N. Boddaert [2], M. El Beheiry [3], J.B. Masson [3], A. Guariento [1], V. Vida [1]

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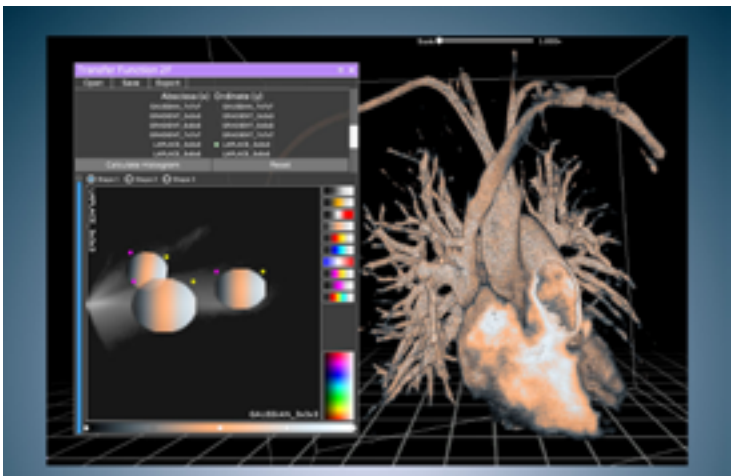
Objectives: A limitation of the clinical use of 3D reconstruction and virtual reality (VR) systems is the relatively high cost and experience required to use hardware and software to effectively explore medical images. We have tried to simplify the process and validate a new tool developed for this purpose with a novel software.

Methods: Five patients with right partial anomalous pulmonary venous return with adequate preoperative imaging acquired by magnetic resonance were enrolled. Five volunteers with no previous experience in the field of 3D reconstruction were instructed to use the software after a short video tutorial. Users were then asked to create a 3D model of each patient's heart with our software and their results were compared quantitatively and qualitatively with a benchmark reconstruction performed by an experienced user.

Results: All our participants recreated 3D models in a relatively short time, maintaining a good overall quality (average quality score? 3 on a scale of 1-5). The overall trend of all the parameters analyzed showed a statistical improvement between Case 1 and Case 5, as users become more and more experienced.

Conclusion: Out software is a very simple software that allows accurate 3D reconstruction in a relatively short time ("fast-track" VR). In this study, we demonstrated the potential use of this tool by inexperienced users, with a significant improvement in quality and time after a few cases were performed. Further studies are needed to confirm the potential application of this technology on a larger scale.



	Case 1	Case 2	Case 3	Case 4	Case 5
Quality Score	3.0±0.2	3.6±0.9	3.2±1.1	3.2±0.9	4.0±0.7
Adjusted Score	2.5±0.2	2.6±0.2	2.9±0.4	3.4±0.3	3.7±0.2

CB6

LATE GADOLINIUM ENHANCEMENT IN GIOVANI ADULTI CON TETRALOGIA DI FALLOT CORRETTA CHIRURGICAMENTE

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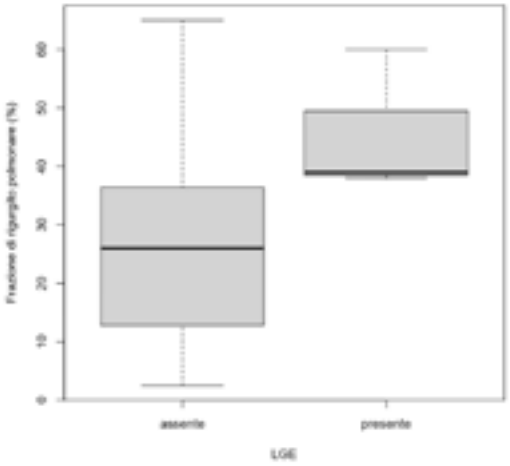
Scopi della ricerca: Nei pazienti operati di Tetralogia di Fallot l'enhancement tardivo di gadolinio (late gadolinium enhancement, LGE) in risonanza magnetica cardiaca (CMR) si localizza a livello del ventricolo destro a causa di processi di cicatrizzazione chirurgica e rimodellamento miocardico. È stata dimostrata una correlazione tra la presenza di LGE e importanti parametri di outcome. Scopo del presente lavoro è verificare la presenza di LGE in una coorte di pazienti con Tetralogia di Fallot sottoposti a correzione chirurgica.

Metodi impiegati: Si tratta di uno studio osservazionale longitudinale prospettico monocentrico. Sono stati reclutati consecutivamente 35 pazienti con tetralogia di Fallot sottoposta a correzione chirurgica, in follow-up presso il nostro Centro al momento dell'esecuzione di una CMR. La presenza di LGE è riportata qualitativamente. Per l'analisi statistica è stato utilizzato il software statistico R (versione 4.2.0). Poiché non è stata riscontrata una distribuzione normale, le variabili numeriche continue sono riportate come mediana e range interquantile (IQR) e sono stati utilizzati test non parametrici per il confronto tra gruppi. Per il confronto tra variabili categoriche è stato usato il test del chi-quadrato.

Risultati: L'età mediana al momento della CMR è stata 18 anni (IQR 15-25.5 anni) con lieve prevalenza del sesso maschile (54,2%). I pazienti sono stati sottoposti ad intervento di correzione ad una età mediana di 15 mesi (8-24 mesi), in circa la metà dei casi (51,4%) preceduto da intervento di palliazione con shunt sistemico-polmonare. Il tipo di correzione più frequente è stato il patch trans- anulare in 21 casi (63.6%), seguito dal patch infundibolare in 8 (24,2%). I pazienti sottoposti a correzione con patch transanulare hanno presentato valori di frazione di rigurgito polmonare significativamente più elevati rispetto agli altri tipi di correzione (36.8%, IQR 27-52.7% vs 12.8, IQR 5.7-17.1%, p<0.001), e conseguentemente di volume telediastolico indicizzato (118 ml/m2, IQR 94- 132 ml/m2 vs 84 ml/m2, IQR 79-89 ml/m2, p< 0.01) e telesistolico indicizzato (60 ml/m2, IQR 51-72.5 ml/m2 vs 45 ml/m2, IQR 36-53 ml/m2, p=0.01). In 12 pazienti (34.3%) vi era stenosi di un ramo dell'arteria polmonare, in due casi associato anche ad ostruzione significativa all'efflusso ventricolare destro (RVOTO). È stata documentata presenza di LGE in 3 pazienti (8,6%). La presenza di LGE si associava a valori di frazione di rigurgito significativamente più elevata (39%, IQR 38.5-49.5% vs 26%, IQR 12.8-36%, p=0.04). Non è stata riscontrata differenza significativa in termini di volumi e di frazione di eiezione del ventricolo destro tra i pazienti con e senza LGE. La presenza di ostruzione destra (RVOTO e/o stenosi dei rami polmonari) non era associata alla presenza di LGE. La presenza di LGE non ha mostrato inoltre una correlazione significativa con l'età all'intervento di correzione, né con il tipo di intervento. Non è stato possibile eseguire un'analisi di outcome in quanto non si sono verificati al follow-up eventi significativi (aritmie ventricolari sostenute, scompenso cardiaco, morte).

Conclusioni: Nella nostra popolazione di studio, la presenza di LGE non era associata a forme di significativa ostruzione ventricolare destra, ma si correlava significativamente con la frazione di rigurgito polmonare.



CB7

IMPACT OF COMPRESSED SENSING (CS) ACCELERATION OF TWO-DIMENSIONAL (2D) FLOW SEQUENCES IN CLINICAL PAEDIATRIC CMR

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Background: Two-dimensional (2D) through-plane phase-contrast (PC) cine flow imaging assesses shunts and valve regurgitations in paediatric CMR, and is considered the reference standard for Clinical quantification Of blood Flow (COF). However, the longer breath-hold (BH) for a flow cine can reduce patient compliance with possibly large respiratory maneuvers altering flow. Compressed sensing (CS) flow has not been widely evaluated in pediatric clinical CMR, although CS flow reduces scan time with persistent accuracy (1-3). Kocaoglu et al applied CS to ascending and descending aorta and SVC with good results. We used main pulmonary artery (MPA) and sinotubular junction (STJ) planes as usual for clinical CMR.

Purpose: We hypothesise that reduced BH time by modest application of CS to 2D cine through-plane flows (Short BH quantification of Flow) (SBOF) retains accuracy while enabling faster and potentially more reliable paediatric flows. We therefore investigate the variance between conventional COF and new SBOF cine flows in paediatric CMR.

Methods: Paediatric patients were enrolled (1.5T, Avanto FIT/Aera, Siemens). Aortic (AO) and MPA COFs were planned from cines of the left and right ventricular outflow tracts. For AO flow the plane was placed at the STJ, and MPA flow was acquired above the pulmonary valve. The same planes were used for COF and SBOF flows at nominally similar parameters except the moderate application of CS exploiting redundancy across the cine frames, SBOF being segmented CS cines not real-time. CVI42 (5.10; Circle CVI) was used for flow analysis (single observer, 3 years' experience). Paired t-tests found the overall differences, and variability was defined at ±2SD for LVSV, RVSV, LVCO, RVCO, and Qp/Qs.

Results and Discussion: 20 patients (mean age 13.7, range 10-17y) were enrolled (12 CHDs, 7 cardiomyopathies or other diseases). The BH times were COF mean 11.7s (range 8.4-20.9s) vs SBOF mean 6.5s (min 3.6-9.1s). For STJ flows the differences and variabilities between the COF and SBOF flows were SV 6.95±13.6(ml/beat), CO 0.16±1.35(l/min) and for MPA flows SV 2.95±12.3(ml/beat), CO 0.27±0.96(l/min) and for Qp/Qs were 0.04±0.19 by ml/beat and 0.02±0.23 by l/min. The mean differences were nonsignificant, and variability between SBOF and COF was similar to intrasession repeatability of COF in a separate paediatric population at our centre (unpublished), that might arise from physiological flow changes, possibly in terms of pre and post load and heart rate. With shorter BHs also assisting patient compliance of the SBOF, physiological flow effects might be reduced, although given the variability in COV this was unconfirmed.

Conclusion: Moderate CS applied to clinical segmented-cine paediatric phase-contrast flows in the STJ and MPA planes did not degrade flow repeatability or cause bias. SBOF assists clinical flows by shorter BHs and also may aid compliance and reduce physiological variations

CB8

AN INSIGHT LOOK INTO ARTERIAL TORTUOSITY SYNDROME

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Background and aim: We report our experience with Arterial Tortuosity Syndrome (ATS), a rare (<1:1 000 0000 live births), autosomal recessive, connective tissue disorder characterized by:

- elongation and widespread tortuosity of large and medium-sized arteries;
- propensity towards aneurysm formation, vascular dissection, focal stenosis of the pulmonary arteries.

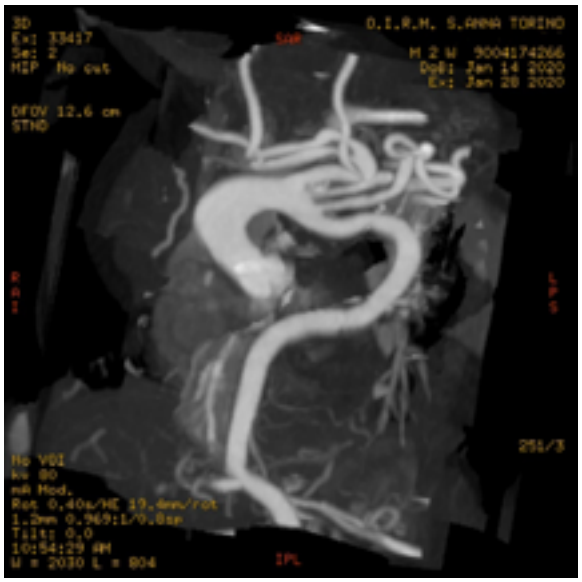
Molecular genetic testing establishes the diagnosis with the finding of biallelic pathogenic variants in SLC2A10, causing a loss of function of GLUT10. The clinical manifestations and prognosis vary depending on the arteries affected. Prenatal diagnosis may be suspected by fetal imaging and confirmed by prenatal molecular diagnosis.

Case presentation: A 36-year-old not primiparous mother was referred to fetal cardiology at 33 weeks of gestation. Fetal echocardiogram showed an elongated and tortuous right pulmonary artery, as well as an elongated aspect of the ascending aorta and aortic arch. Pregnancy and delivery were uneventful. The neonate presented large for gestational age, with a voluminous bilateral inguinal hernia and a hiatal hernia. Genetic testings found homozygous variant c.685>T in SLC2A10 and confirmed the heterozygosity in both non-consanguineous parents.

Methods: An angio-CT (Figure 1) study was performed, with the following result. Extremely tortuous aspect of the arteries of the circle of Willis and its branches, tortuosity and kinking of carotids in the intracranial tract and basilar and vertebral arteries, with absence of images referable to aneurysmal dilatation. Thoracic and abdominal aorta, supraortic trunks, internal jugular veins, branches of abdominal aortic, and arterial iliac-femoral axis characterized by a tortuous course but free from signs of stenosis or dilatation. Left pulmonary artery elongated with superior take off. The lobal and interlobar pulmonary of both lungs small in caliber and tortuous. Left aortic arch placed in the first thoracic tract with passage into the right hemithorax at the level of the left atrium with an elongated and tortuous course. Elongated transverse arch with normal origin of the supra-arctic trunks that present tortuous along their entire course.

Results: Patient asymptomatic at 2 years of life. Hemodynamic stability. Persistent foramen ovale (2 mm), with left-to-right shunt of small entity. Enlargement of main pulmonary and of both its branches (10 mm at the origin). Slight enlargement of the ascending aorta (21,5 mm). Follow-up every six months.

Conclusions: ATS is a connective tissue disorder involving great to medium-sized arteries. Morbility and morbidity vary according to the vessels involved. Angio-CT is essential to establish the prognosis.



CB9

NON-INVASIVE EVALUATION OF COMPLEX CONGENITAL HEART DISEASE IN A PRETERM NEW BORN USING 4D-FLOW TECHNIQUE WITHOUT ANAESTHESIA AND CONTRAST MEDIUM

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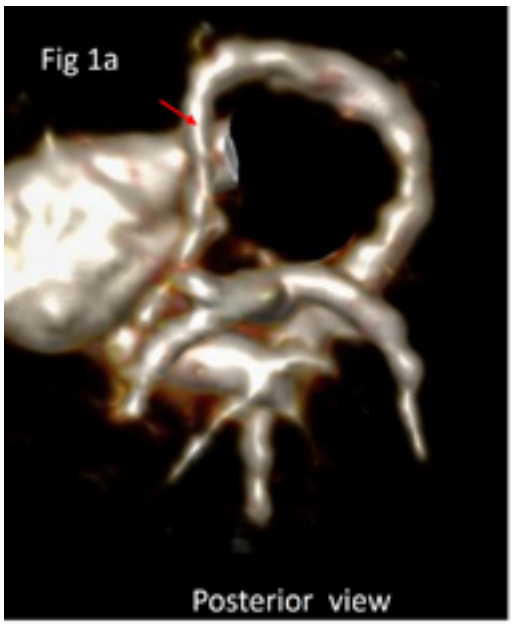
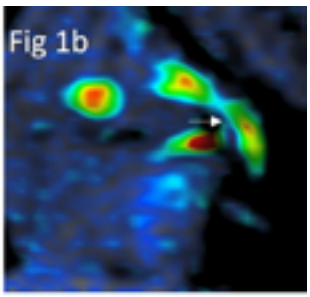
Purpose: Cardiac Magnetic Resonance (CMR) is a validated method in the evaluation of complex congenital heart disease (CCHD). The use of CMR in newborns is limited by risks associated with general anaesthesia. 4D-flow CMR sequence is increasingly used in CHD and aortic disease. In this study we sought to evaluate the feasibility of the 4D-flow for evaluation of in a preterm newborn with CCHD using the feed-and-sleep technique (avoiding general anaesthesia), without medium contrast.

Method: A foetus with CCHD was born at 35 weeks in our centre for premature rupture of membranes by vaginal delivery. Weight at birth: 3000 g, Apgar 8/9, peripheral O2 sat: 90 %. The prenatal diagnosis was confirmed by echocardiography: heterotaxy syndrome, common atrio-ventricular valve, severe pulmonary stenosis; left anomalous pulmonary venous return and a small aorto-pulmonary collateral were suspected as well. The neonate presented a good adaptation to the extrauterine life. At 8 days of age he was scheduled to a CMR study in order to evaluate the pulmonary venous return and quantify the QP/QS. The neonate was fed after the application of CMR electrodes and 1 mg of melatonin was orally administered. Afterwards he was wrapped in a blanket and placed in a MR-compatible bed with compatible MRI ear muffs Around 30 min after feeding the CMR examination starts. The examination was comprehensive of a 4D-flow sequence in axial plan performed with a temporal resolution of <40ms and a reconstructed isotropic spatial resolution of 1.2 x 1.2 x 1.2 mm. A PCMRA was subsequently reconstructed to evaluate venous and arterial vessels. Systemic and pulmonary flows were calculated in post processing using a dedicated software (Arterys 4D-flow AI suite). Time acquisition of 4D flow was < 3min.

Results: the suspected diagnosis was confirmed: in particular partial anomalous left pulmonary venous return into left anonymous vein through a vertical vein (red arrow fig 1a) malposition of the great arteries, pulmonary stenosis and right aortic arch (fig 1b) and a small aorto-pulmonary collateral (white arrow Fig1b) . The shunt was calculated as QP/QS 0.9. The newborn was discharged at 19 days age. At follow-up the infant growths well, O2 Sat progressively decreased. At 6 months he was admitted for elective surgical palliation (Glenn anastomosis and partial pulmonary venous return repair). Anatomic findings of CMR were confirmed by pre-operative CT scan (fig !C) and the surgical report.

Conclusion: CMR evaluation using the feed-and-sleep technique in this preterm neonate with CCHD allowed a non-invasive diagnosis without using ionizing radiation, contrast medium avoiding also general anaesthesia. The 4 D flow sequence is a valid technique for the evaluation of vessels flow and anatomy. Further larger studies are needed to validate the technique in a large population of congenital heart disease.





CB10

IS INITIAL CORONARY DILATATION A RISK FACTOR FOR PERSISTENT VASCULAR RISK IN LONG-TERM FOLLOW-UP OF OLD KAWASAKI DISEASE PATIENTS?

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Background: There is increasing evidence that patients with Kawasaki disease (KD) are at increased risk of accelerated atherosclerosis, specially involving the coronaries, this risk is believed to be mediated by persistent inflammation.
However, there is non-conclusive data if this persistent inflammation can involve other vessels other than the coronaries, thereby increasing their long-term vascular risk. The aim of this study was to assess vascular dysfunction in longterm follow-up of healed KD patients.

Patients and Methods: Forty Kawasaki disease patients were recruited from Cairo University Children Hospital, exclusion criteria was patients with persistent coronary aneurysms and an interval of less than five years after the acute stage of KD and any additional cardiovascular or systemic disorder. Patients were divided into two groups, those who developed coronary aneurysms that regressed during follow up (Group C-GC) and patients who never developed coronary aneurysms (Group B-GB). In addition, twenty controls were enrolled in the study (Group A-GA). Patients were assessed by Brachial Flow-Mediated dilation (BFMD), Carotid intima media thickness (CIMT), and Aortic biomechanics by aortic strain (AAS).

Results: CIMT was higher in GC compared to GA and B (0.6 vs.0.38 and 0.39, P<0.001) respectively. Aortic strain was impaired in GC in contrast to GA and B (12 vs. 18.4 and 18.9, P<0.001) respectively. BFMD was reduced in GC compared to GA and B (5.7 vs. 9.7 and 9.3, P<0.001) respectively.

Conclusion: This study shows strong evidence of persistent vascular dysfunction in old KD patients even without persistent coronary aneurysms. This warrants longterm follow up of these **Patients and** tailored thromboprophylaxis to prevent vascular events.

CB11

ISOLATED LEFT VENTRICULAR NON COMPACTION: A SCARY DIAGNOSIS LOOKING FOR DIAGNOSTIC CRITERIA. CLINICAL FEATURES AND FOLLOW-UP OF 22 PEDIATRIC PATIENTS

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By definition, isolated LVNC (ILVNC) occurs in the absence of other cardiac and non-cardiac congenital abnormalities. The left ventricular apex and the free wall are the most frequently affected segments, even though both ventricles can be influenced in some cases (22–38%). It has been suggested that LVNC may be due to the premature interruption of the final stage of myocardial compaction during embryonic life. Clinical presentation of LVNC is highly variable. It can occur at any age, ranging from asymptomatic to end-stage heart failure, and it can be associated with arrhythmias, syncope, sudden cardiac death or thromboembolic events (or combinations thereof).

We analyze clinical features and imaging aspects of 22 patients. We included patients with isolated LVNC diagnosed in pediatric age (0-18 years) followed up at the Pediatric Cardiology Unit of the Children's Hospital of Parma. We reviewed medical records from December 2005 to May 2021.

Five out of the 22 patients had symptoms at diagnosis, two more during follow-up; symptoms are often exertion related. Only the patient who had antenatal diagnosis had heart failure during the first year of life and needed therapy. Myocardial function improved significantly during follow-up. In the majority of patients, non-compaction was more prominent in the myocardial segment of the LV apex and lateral wall.

LVNC is a cardiomyopathy that can have different clinical manifestations. Echocardiographic and MR diagnostic criteria have been proposed, but we still lack universally accepted diagnostic criteria and, moreover, we lack a clear correlation between imaging findings and clinical manifestations. Further studies are needed to establish clear diagnostic criteria and a proper follow-up.

CB12

ECHOCARDIOGRAPHIC AND CMR FEATURES OF PATIENTS WITH REPAIRED TETRALOGY OF FALLOT IN THE LONG-TERM FOLLOW-UP: A SINGLE-CENTRE EXPERIENCE

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Background: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart disease and the population of ToF repair survivors is growing rapidly. Children and, more frequently young adults, with repaired ToF develop late complications. Sudden cardiac death and life-threatening ventricular arrhythmia remain a concern in patients with repaired tetralogy of Fallot.
The aim of this study was to describe and analyze long-term follow-up of patients with repaired ToF.

Methods: This is a retrospective multi-centre cohort study. Consecutive 87 patients with repaired ToF who did not undergo pulmonary valve replacement were included. Mean age of all patients was 19±8. The electrocardiographic (ECG), cardiopulmonary exercise testing (CPET), echocardiographic and cardiac magnetic resonance (CMR) data were reviewed retrospectively. Patients were screened according the age at surgery (<12 months and >12 months) and the time since the repair (< 25 years and ? 25 years).
All patients were screened during a median follow-up of 2.9 years and significant sustained and non-sustained ventricular tachyarrhythmias at ECG Holter were recorded.

Results: CPET values were not significantly different in patients with more than 25 years since the corrective surgery. RVESV at CMR, pulmonary regurgitation at CMR, RA and LA areas and RV basal- and mid- diameters at echocardiography were significantly increased in patients with more than 25 years since the corrective surgery
Patients with older age at surgery (>12 months) have significantly reduced LVEF at CMR, V02max at CPET, larger echocardiographic RA and LA areas and larger RV basal diameter.
During a median follow-up of 2.9 years, 12 patients had documented ventricular tachycardia at ECG Holter. On univariate analysis, RVEF and LVEF at CMR (p=0.049 and 0.005, respectively), right atrial area (P=0.02), right ventricular outflow tract gradient (P=0.017), were related to ventricular arrhythmias.
On a multivariable analysis, the echocardiographic right ventricular outflow tract gradient was found to be significantly related to ventricular arrhythmias (P=0.001).

Conclusion: Long-term survival and clinical condition after surgical correction of ToF in infancy is generally good and the late functional status in ToF - operated patients could be excellent up to 25 years after the repair. However, patients with older age at surgery have relative reduced functional status and larger right ventricular dimensions compared with those undergoing early repair. The echocardiographic right ventricular outflow tract gradient is an independent predictor of ventricular arrhythmias in patients with repaired TOF.

CB13

CARDIAC ATROPHY AND ACE INHIBITORS IN DUCHENNE CARDIOMYOPATHY: 10 YEARS IS THE RIGHT MOMENT TO START THERAPY?

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Background: Dilated cardiomyopathy (DCM) is a leading cause of morbidity and mortality in patients affected by Duchenne muscular dystrophy (DMD). DCM is commonly characterized by a hypokinetic and dilated phenotype, typically involving left ventricle (LV). It is a progressive disease: prevalence and severity increase with age. Nowadays, angiotensin converting enzyme inhibitors (ACEi) are indicated in all patients older than 10 years in order to prevent DCM onset and progression.

Aim: The aim of this study is to analyse efficacy and timeliness of ACEi in Duchenne patients.

Methods: We conducted a retrospective analysis on all patients affected by Duchenne muscular dystrophy followed in our Institution from 2010 (?), and treated with ACEi. Therapy was documented. We recorded echocardiographic data about LV dimension and function, at first cardiovascular assessment, at one year from ACEi start, and at last follow up. Relative wall thickness (RWT) was calculated. We considered a RWT< 0.32 as a marker of cardiac atrophy. We analysed RWT trend as a marker of LV remodelling during time, in relation with anti-remodelling therapy.

Results: We enrolled 45 patients. Median age at first CV assessment was 10.3 y (±2.9). All patients were treated with ACEi: 48% was treated with Enalapril, 26% with Ramipril, 17% with perindopril, and the remaining with Fosinopril or Lisinopril. Median age at ACEi start was 11.2 (± 2.9).
72% of patients presented a reduced RWT (< 0.32) at first CV evaluation (average value 0.29 ± 0.05). After 1 year of treatment the percentage of patient with cardiac atrophy was unchanged (p = 0.52): 6 patients progressed and 6 regressed. At last follow up (35.5 months, ±7.7) the prevalence of cardiac atrophy was slightly increased (78% - p = 0.15): 9 patients (28%) developed cardiac atrophy during follow-up, despite ACEi therapy, while 6 (19%) regressed. Notably, neither progression nor regression were statistically significant (p= 0.15, p = 0.4 respectively).

Discussion: these data attest the progression of DCM in DMD patients is slow, and ACEi seem to be useful do delate its course: 60% of patients remained stable at 1 year, 50% at last follow up. However, the most important data is the high prevalence of cardiac atrophy at ACEi start: three-quarters of patients presented a reduced LV mass.

Conclusion: ACEi seem to be effective in delaying DCM progression. However, our data evidence cardiac atrophy is already present at ACEi start. Therefore, it should be considered to earlier start therapy with ACEi in order to prevent loss of cardiac muscle. Further studies will be needed.

CB14

HEPATOCELLULAR CARCINOMA AFTER THE FONTAN PROCEDURE: SINGLE CENTER EXPERIENCE

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Background: Hepatocellular carcinoma (HCC) is a recognized long-term complication following the Fontan operation, but outcomes and risk factors for mortality have not been characterized in this population. Furthermore, HCC is frequently difficult to diagnose and treat because of cardiac complications, coagulopathy, and congenital abnormalities.

Aim: The purpose of this study was to: (1) describe the outcomes in patients with HCC diagnosis after Fontan palliation; (2) identify risk factors for mortality in patients with HCC undergoing the Fontan operation; (3) perform a screening algorithm that enables early HCC diagnosis.

Methods: We retrospectively identified all patient with Fontan palliation and a diagnosis of HCC from January 2017 to December 2021 afferent to our adult congenital heart disease center

Results: Among a total of 198 patient who had undergone the Fontan operation, there were 7 (3,5%) HCC cases (42,9% males). The median age at the time of HCC diagnosis was 30 years (range 22-40) and the median duration from Fontan operation to HCC diagnosis 26 years (range 20-35). The median age at Fontan operation was 3,6 years. The most common type of Fontan connection was extracardiac conduit (5 patients, 71%), one patient (14%) had atriopulmonary Fontan and one (14%) had atriopumonary Fontan subsequently converted in extracardiac conduit. All patients had a unique morphologically left ventricle with preserved contractile function (mean EF 57,8%). The median value of alphafetoprotein at the time of HCC diagnosis was 584 ng/ml (range 2,3 - 2931). Two patients (29%) had normal values of alphafetoprotein. In six patients (86%) abdominal ultrasound was negative or showed only micronodular liver lesions and the liver injury was suspected on follow-up cardiac MRI. The mean tumor size was 20 ± 5 mm and nobody had metastasis at the time of HCC diagnosis. Liver cirrhosis was present in all patients. Among the 7 patients with HCC, nobody had symptoms at the time of diagnosis, including abdominal pain, jaundice, ascites, dyspnea or fever. Patients received multiple forms of therapies. Administered treatments included: transarterial chemoembolization (n=4, 57%), surgical resection (n=2, 29%), radiofrequency ablation (n=1, 14%), listing for liver transplant (n=2, 29%), medical therapy with sorafenib (n=1, 14%). Interestingly, two patients (29%) presented HCC two years after heart transplantation performed for Fontan failure (reasons for transplant were ventricular dysfunction in one case and protein losing enteropathy in the other). Of the 7 patients with HCC, two (29%) died during a median follow-up of 30 months. The cause of death was metastatic disease with multiorgan failure in both cases.

Conclusions: This study demonstrates high mortality following a diagnosis of HCC in adult patients with Fontan (29%). Notably, initial liver imaging can be negative for chronic liver disease and alpha-fetoprotein can be normal. In keeping with previous experience, our study suggests that liver imaging or alphafetoprotein alone, may be insufficiently sensitive to screen patient at risk for HCC. A multi-system approach is needed and the use of hepatospecific abdominal MRI should be favoured.

CB15

USE OF PROPRANOLOL IN TREATING HEMANGIOMAS; CARDIAC SIDE EFFECTS: (OUR 11 YEARS EXPERIENCE)

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Infantile hemangiomas (IHs) are the most common benign tumors of infancy occurring in 5% to 10% of infants; exhibiting a characteristic sequence of growth and spontaneous involution. Most infantile haemangiomas do not require therapy Treatment is indicated in case of large hemangiomas that have tendency to complicate. In 2008, the effectiveness of the beta-blocker propranolol for infantile haemangiomas was discovered; by now there is extensive worldwide experience.

Aim: The purpose of this study is to investigate the safety of propranolol, when used in pediatric age to treat infantile hemangioma (IH), especially related to its cardiac side effects as being considered a heart drug, as the main reason for parents and even physicians to resist starting the treatment.

Method: This is a retrospective study with a sample of 476 patients diagnosed with infantile hemangioma and treated with systemic propranolol during the time interval from January 2011 to December 2021, in our center as a single center in Albania. We observed clinical recorded propranolol adverse events experienced in hospital or outpatient based on parental reporting and measured the impact of propranolol on blood pressure (BP) and heart rate (HR) by comparing the means before starting treatment, 2 hours and 2 weeks after starting treatment.

Results: This study showed that symptomatic adverse events caused from propranolol were mild and severe adverse events were very rare. The most common complaints were agitation, reduced feeding, paleness, and sweating. But only in 28 (5.9%) cases these symptoms were severe enough to review the treatment where 1.8% had more severe respiratory symptoms, 2.7% experienced hypoglycemia and only 1.2% heart related symptoms, symptomatic hypotension, or bradycardia. Mean blood pressure reduction with treatment was significant, especially after achieving the maintenance dose 2 mg/kg/bw. BP under the 5-th percentile was registered in 2.9% of cases but only in 4 patients it was symptomatic. While heart rate reduction was noticed from the first dose, but under the 5-the percentile it was registered only in in 2% of cases, and only 2 of them experienced symptoms.

Conclusion: Propranolol has been successful in treating problematic infantile hemangiomas, with minimal, not significant cardiac side effects. where most of them are easily overcame by temporarily interrupting treatment. Given the favorable risk-benefit profile, propranolol should be the first-line therapy for infantile haemangiomas that require treatment.

CB16

THE EFFECTS OF PHYSICAL ACTIVITY ON HEART FAILURE BIOMARKERS IN PEDIATRIC PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME AND FONTAN CIRCULATION

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Background: Hypoplastic left heart syndrome (HLHS) is one of the most complex cardiac defects seen in the newborn and remains probably the most challenging to manage of all congenital heart defects. Several studies have investigated the possible effects of physical activity in patients with HLHS, however this topic is still not fully understood. The aim of this study was to evaluate the effects of physical activity on heart failure biomarkers in patients with HLHS and Fontan circulation.

Methods: We enrolled 28 (18 male) patients with HLHS and Fontan circulation and a mean age of 10.4 ± 2.7 years. Each patient underwent a cardiological examination, echocardiogram and blood tests. Physical activity was assessed by submitting the International Physical Activity Questionnaire (IPAQ) to the children together with their parents. Based on the score obtained, the patients were divided into two groups. In group I (12), children who carried out regular physical activity (1380 ± 260 METs/min/week). In group II (16), on the other hand, the children who, according to the questionnaire scores, led a sedentary life (568 ± 68 METs/min/week) were considered. Therefore, we measured heart failure biomarkers hs-cTnT, NT-proBNP and ST2 in all patients.

Results: Statistical analysis showed a significant difference in physical activity (METs/week/min) between the two groups ($p<0.001$) while there was no significant difference in right ventricular function between the two groups: RVFAC 36.29 ± 4.8% vs 35.14 ± 5.3% ($p>0.05$) in group I and group II, respectively. Regarding cardiac biomarkers, the data analysis did not show a significant difference in hs-cTnT between group I and group II (3.2 ± 0.8 vs 4.1 ± 0.4 ng/L, $p>0.05$), while we found a significant difference in NT-proBNP and ST2 levels between group I and group II 65.98 ± 20.03 vs 102.95 ± 23.65 ng/L ($p <0.01$), 21.0 ± 8.6 vs 24.6 ± 7.5 ng/mL ($p<0.05$) respectively.

Conclusions: Our study showed, for the first time, a reduction in NT-proBNP and ST2 levels in the group of children who engaged in regular physical activity compared to the group of sedentary children.

CB17

COEXISTING GENETIC FINDINGS IN A COHORT OF PATIENTS WITH LABORATORY CONFIRMATION OF 22Q11.2 DELETION SYNDROME

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Background: The 22q11.2 deletion syndrome (22q11.2DS) is the most frequent chromosomal microdeletion syndrome with an incidence of 1:3000-1:6000 live births.
Patients with 22q11.2DS display a highly heterogeneous phenotype characterized by variable involvement of multiple organs and systems.
Up to 70% of patients show congenital heart defects (CHDs), mostly conotruncal anomalies (CTDs). Sometimes patients may present with atypical or complex clinical features that deserve further investigation in order to identify secondary diagnosis.
The identification of potential additional genetic mutations responsible for overlapping phenotype or atypical clinical features may lead to improved clinical outcome and management of these patients.

Methods: This is a retrospective, observational study. We collected data on patients diagnosed with 22q11.2 DS followed-up in the Pediatric Department from January 2015 to September 2021.
We excluded patients without a genetic confirmation despite clinical phenotype suggestive of the syndrome.
We included all 22q11.2DS patients with a laboratory confirmation through multiplex ligation-dependent probe amplification (MLPA), fluorescent in situ hybridization (FISH), comparative genomic hybridization array (CGH-array) or single nucleotide polymorphism (SNP-array).
Cardiac phenotype was systematically collected and the presence of a dual diagnosis was retrospectively identified.

Result: Of a total of 110 patients referred to our Department, we excluded 32 patients for missing laboratory confirmation; the final cohort consisted of 78 22q11.2DS patients. MLPA was performed in 3 patients(3.8%), FISH in 40 patients(51.2%), CGH-array in 31 patients(39.7%) and SNP-array in 4 patients(5.1%).
We identified a coexisting variant in 8 patients analized with CGH-array(25.8%) and in 1 patient with SNP-array(25.0%). We identified additional microdeletions and microduplications involving chromosomes 5,16,2, X and 7.
CTDs were present in 1/9 patient (11.1%), aortic arch anomalies in 3/9 patient(33.3%), septal defects in 1/9 patients(11.1%) and 1/9 (11.1%) had no CHD. In 2/9(22.2%) an echocardiographic assessment was missing for poor compliance.

Conclusions: Incidence of dual diagnosis and additional CNVs with a possible functional significance in patients with 22q11.2DS is remarkable.
The presence of coexisting mutations may be responsible for an overlapping phenotype and/or clinical atypical features that impact on the multidisciplinary approach for these patients.
The use of accurate diagnostic tools (i.e. CGH-array and SNP-array) allows the diagnosis of both 22q11.2DS and additional mutations in order to provide a more personalized clinical management and accurate genetic counseling.

CB18

THE BURDEN OF SUSPECTED CARDIAC DISEASE ON AN ACADEMIC PEDIATRIC EMERGENCY DEPARTMENT

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Background: The admission to the Paediatric Emergency Department (PED) for symptoms potentially related to cardiac etiologies can be challenging due the variability of severity and to possible different underlying conditions.

Objective: The aim of our study was to describe the rate of admissions to a tertiary care PED due to symptoms possibly associated with cardiac conditions.

Methods: We retrospectively analyzed the database of our PED from 01th January 2019 to 31th December 2021 including a potentially cardiological disease as diagnosis. We categorized these diagnosis as follows: arrhythmias, chest pain, syncope/lipothymia, heart failure (including cyanosis, edema, pleural effusion), cardiac arrest. We reported the prevalence of each category as percentage .

Results: In the period indicated, in our PED 861 children were admitted with symptoms possibly related to cardiac conditions, accounting for 2% of total visits (44438 patients admitted). Almost half of the reasons for PED visits was syncope/lipothymia (413 admissions, 48%), while chest pain accounted for 263 visits (30,5%). Almost all of them were not related to an heart disease. Arrhythmias accounted for 94 visits (11%) and 23% of them were supraventricular tachycardias. Only 15 patients were diagnosed with heart failure (1,7%) and all of them had an associated congenital heart disease (CHD). A miscellaneous of conditions such as edema, pleural effusion and cyanosis were present in 71 patients (8%), but not related to heart failure. Lastly, cardiac arrest was observed in 5 cases, three of which with an underlying CHD. Overall, only 44 patients had a CHD (0.1% of the total admissions). Of note, during the SARS-CoV-2 pandemic in 2020 there was a 42% fall in PED accesses for potentially cardiac symptoms.

Conclusions: Our study showed that children admitted to a tertiary care PED with symptoms possibly related to heart disease mostly presented non cardiac conditions, particularly syncope/lipothymia and thoracic pain not requiring cardiological referral. Although heart diseases are relatively rare in the pediatric population referring to PED, patients must be carefully evaluated because of the risk of potentially severe evolution, especially in children with CHD.

CB19

STUDIO RETROSPETTIVO MONOCENTRICO SULLE MIOCARDITI IN ETA' PEDIATRICA: DALLA DIAGNOSTICA AL FOLLOW UP

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Background: La miocardite in ambito pediatrico appare una sfida impegnativa per il clinico, dalla scelta degli strumenti diagnostici iniziali, alla terapia, fino al tipo ed alla durata del follow up.

Scopo dello Studio: Lo scopo di questo studio retrospettivo, condotto su pazienti in età pediatrica affetti da miocardite, è stato quello di analizzare la presentazione clinica, la diagnostica iniziale, il follow up e la prognosi. Lo studio è stato condotto presso il centro di Cardiologico pediatrico - nel periodo compreso dal 2011 al 2021.

Materiali e Metodi: Il gruppo esaminato comprendeva 17 pazienti di età inferiore ai 5 anni (età media di 22 +/-14 mesi), affetti da miocardite fulminante (8/16) o da miocardite acuta (9/17), con quadro clinico di scompenso cardiaco ed ecocardiografico di severa compromissione della funzione sistolica. Sono stati valutati: clinica, comorbilità, esami ematici e anatomopatologici (biomarcatori, ricerca virus, analisi bioptica), imaging, terapia e follow up. I parametri ecocardiografici sono stati valutati all'ingresso e al follow up (1-3-6-12 mesi). In tutti i pazienti è stata monitorata ecocardiograficamente la presenza di insufficienza mitralica (all'esordio e durante tutto il periodo di monitoraggio).

Risultati: Tutti i pazienti analizzati hanno presentato miocardite ad eziologia virale (prevalentemente da PVB19, ma anche HHV2, EBV e Covid19) e un quadro di scompenso cardiaco acuto che ha necessitato di supporto inotropo e/o ventilazione meccanica invasiva. La diagnosi è stata confermata con la biopsia endomiocardica e/o risonanza magnetica cardiaca. La maggior parte dei pazienti è stata trattata con terapia immunosoppressiva ed in alcuni casi infusione endovenosa di immunoglobuline umane. E' stato analizzato un periodo di follow up della durata di 3-12 mesi, al termine del quale è stato rilevato un miglioramento significativo dei parametri ecocardiografici analizzati ($p<0.001$) (FE e grado di insufficienza mitralica), una riduzione significativa del diametro telediastolico (DTD), del diametro antero-posteriore (DAP) dell'atrio sinistro, dell'anulus mitralico e del rapporto atrio sn/radice aortica. E' stato inoltre evidenziato che, in media, i pazienti con miglioramento della funzione ventricolare sinistra erano gli stessi in cui la patologia aveva esordito con un quadro di miocardite fulminante ed FE severamente depressa, laddove coloro che avevano esordito con un quadro di miocardite acuta, presentavano a 6 mesi disfunzione sistolica di grado variabile. La severità e il grado di insufficienza mitralica all'ingresso non si sono dimostrate predittive del rischio a lungo termine.

Risvolti Clinici: Sebbene la miocardite tenda a presentarsi maggiormente nei primi anni di vita, le evidenze ottenute in queste fasce di età risultano ancora poche e contraddittorie.
Non di rado, nella pratica clinica si tende a prendere decisioni che non sempre risultano basate su evidenze forti o protocolli universalmente condivisi, in particolar modo dopo la pandemia da Covid19 che ha visto emergere una nuova entità clinica nota come MIS-C (Sindrome multi infiammatoria sistemica del bambino).

Conclusioni: La nostra esperienza ha confermato che i pazienti con miocardite virale e severa compromissione emodinamica hanno beneficiato di un trattamento precoce ed aggressivo che ha consentito un outcome positivo nell'88% dei casi. Un quadro iniziale di miocardite fulminante, sebbene più severo, porta ad un massiva attivazione della risposta immunitaria che agisce direttamente sulla replicazione virale, laddove un quadro di miocardite acuta può decorrere in maniera più subdola ed insidiosa, esitando in cardiomiopatia dilatativa. Nonostante la presentazione clinica severa, una terapia mirata ha mostrato di poter previene il rischio di disfunzione sistolica a lungo termine e il rimodellamento inverso del cuore.

CB20

THE EFFECTS OF PHYSICAL INACTIVITY IN ADULT PATIENTS WITH CONGENITAL HEART DISEASE DURING THE COVID-19 PANDEMIC

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Background: The ongoing SARS-CoV-2 pandemic has infected more than 500 million people worldwide. Several measures have been taken to reduce the spread of the virus and the saturation of intensive care units: among them, lockdown has been declared in Italy 9th March 2020. As a result, gyms, public parks, sports fields, outdoor play areas, and schools, as well as multiple commercial activities, have been closed. The consequences of physical inactivity can be dramatic in ACHD patients, in which the benefit of regular exercise is well known.

Aim: In this study we investigated the effects of reduced exercise training during the COVID-19 pandemic on exercise capacity of adult patients with congenital heart disease (ACHD).

Methods: We selected, from our database, patients aged ? 18 years who performed an exercise or a cardiopulmonary exercise test before the lockdown (from October 2019 to February 2020) and one year after the lockdown. Patients were divided in two homogeneous groups according to age and sex and physical activity (sedentary versus regular physical activity). Exclusion criteria were: presence of significant clinical and/or therapeutic changes between the two tests; significant illness occurred between the two tests, including Sars-CoV 2 infection; interruption of one of the tests for reasons other than muscle exhaustion. We analyzed the following parameters before and after the lock-down: a) physical activity in minutes per week; b) BMI; c) duration of exercise test, d) maximal oxygen consumption (VO2 max). The parameters were analysed with paired Student t-test.

Results: Thirty-eight patients (60.5% males) met the inclusion criteria. Before the lock-down, 20 patients (group A) aged 25 ± 6 years old had a sedentary lifestyle, and 18 pts (Group B) aged 26 ± 6 years old had a lifestyle characterized by regular physical activity (RPA). During the Sars-CoV2 pandemic, 1 patient in Group A started physical activity at home (pilates 120 min weekly) and 4 pts of group B (10.5%) stopped their training, In Group A: BMI, duration of the exercise test and VO2 max did not show any difference. In group B: a) physical activity before and after the lock-down reduced with statistical significance from 115 ± 46 min/week to 91 ± 64 min/week (- 21%, p = 0.03); b) BMI did not change; c) duration of exercise test was fairly stable; d) VO2 max at cardiopulmonary exercise test showed a significant reduction before and after the lock down (p = 0.04). Results are displayed in Figure 1.

Conclusion: Sars-CoV2 pandemic dramatically changed habits of ACHD patients, significantly reducing their possibility to exercise. Our data analyzed in this extraordinary situation demonstrated once again that the physical inactivity in ACHD worsen functional capacity as highlighted by VO2 max. For this reason, regular exercise should be always encouraged to preserve functional capacity and avoid morbidity.

Figure 1

	Group A (n = 20)			Group B (n = 18)		
	Before LD Mean (±SD)	After LD Mean (±SD)	p-value	Before LD Mean (±SD)	After LD Mean (±SD)	p-value
RPA min/wk	-	-	-	115 (46)	91 (64)	0.03*
BMI	22 (3)	23 (4)	0.37	22.2 (2.7)	22.8 (2.6)	0.98
Duration (s)	496 (140)	556 (100)	0.85	580 (280)	599 (59)	0.35
VO_2 max ($mlO_2*kg^{-1}*min^{-1}$)	24.3 (4.0)	23.9 (4.9)	0.77	31.0 (5.0)	27.0 (3.8)	0.04*

CB21

EXERCISE CAPACITY AND PREDICTORS OF MORBIDITY AND MORTALITY IN PATIENTS WITH FONTAN CIRCULATION

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Background: Discordant results regarding the correlation between poor outcomes in patients with congenital heart diseases (CHD) and Fontan patients at cardiopulmonary exercise test (CPET) were shown in previous literature.

Aim: The aim of our study is to identify the instrumental predictors of mortality and need for transplantation in Fontan population.

Material and Methods: We included in our study all patients who had undergone Fontan procedure in our Institution and performed CPET until 2018. CPET and echocardiographic data were collected retrospectively from medical records and outpatients' visits. To perform statistical analysis, we compared CPET results of Fontan patients transplanted/listed to heart transplantation or death with other Fontan patients.

Results: Eighty-nine Fontan patients were included (76 extracardiac, 3 atrio-pulmonary, 10 lateral tunnels). The mean age at Fontan surgery was 10 years old (SD 6.8 years). The mean age at CPET was 18 years old (SD 7 years).
During follow-up 3 patients died, 1 patient undergone heart transplantation (HTx) and 3 are on active transplantation waiting list. At univariate analysis VO2 at anaerobic threshold, chronotropic index, VO2 peak were related to poor prognosis (p=0.015, p=0.007, p=0.001 respectively). Reduced EF was related to impaired VO2 peak results (p=0.039) while atrioventricular valve regurgitation was related to reduction of O2 peak saturation (p= 0.047).
At multivariate analysis evaluating VO2 at anaerobic threshold, VO2 peak saturation, VO2/ heart rate, chronotropic index, and FEV 1, only chronotropic index was an independent predictor of poor prognosis or advanced NYHA class (p=0.037; p=0.015).

Conclusions: In our experience even if most parameters of CPET are associated with increased risk of death or need for heart transplantation at multivariate analysis only chronotropic index appears to be an independent predictor of poor outcome and it is related to impaired quality of life.
A higher sample of patients would be necessary in order to draw conclusive results regarding the usefulness of CPET in the prognostic evaluation of Fontan patients.

CB22

LIVER STIFFNESS EVALUATION USING ACOUSTIC RADIATION FORCE IMPULSE ELASTOGRAPHY IN PEDIATRIC AND ADULT PATIENTS WITH CONGENITAL HEART DISEASE

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Background & Aims: Hepatic complications are common in patients with congenital heart disease and, therefore, an adequate hepatic surveillance is fundamental. Liver biopsy represents the gold standard for staging hepatic fibrosis but it's not suitable for routine setting. Acoustic radiation force impulse (ARFI) elastography allows to assess hepatic stiffness in a non-invasive and reproducible way. The usefulness of ARFI imaging has been described in adult Fontan patients but only few studies have been reported in the pediatric Fontan population and no one in CHD others than Fontan.
The aims of this study were to assess liver stiffness, using ElastPQ acoustic radiation force impulse elastography, in pediatric and adult patients with CHD, to compare liver stiffness values with healthy controls and to analyze possible associations between ARFI values and clinical, biochemical, cardiac and hepatic parameters.

Materials and Methods: Pediatric and adult patients that underwent heart surgery for CHD were prospectively enrolled between October 2018 and October 2020. Controls subjects matched for age and sex to the case group were also included. The latest laboratory tests and echocardiogram available were collected. ARFI measurement were performed by a single expert radiologist using the Philips Healtcare® ultrasound with ElastPQ software.

Results: Fifty subjects were enrolled for the study: 20 Fontan patients (13 males, median age 8.4 years), 13 non-Fontan (9 males, median age 4.8 years) and 17 controls (6 males, median age 10 years). The median values of ARFI elastography were significantly higher in patients with CHDs compared to control subjects ($p<0.01$). Patients with morphological right ventricle overload showed significantly higher results ($p=0.02$). The cut-off of 5.7 kPa at elastography was used to discriminate between normal liver and liver with signs of congestion or fibrosis. All controls subjects showed ARFI values <5.7 kPa whereas only 25% of Fontan Patients and 46% of non-Fontan were below that threshold. Liver stiffness values were positively correlated with time from surgery and age at liver evaluation ($p<0.01$). The number of platelets and white blood cells were inversely related to liver stiffness measurements ($p=0.04$ and $p=0.05$ respectively). The AST to platelet ratio index positively correlated with ARFI elastography results ($p<0.03$). No significant correlations between ARFI results and other biochemical or cardiac parameters were found.

Conclusions: Our data showed that the median values of liver stiffness measured with ElastPQ pSWE were significantly higher in patients with CHDs and, in particular, in those with morphological right ventricle overload. Liver stiffness values were also correlated with time from surgery and age at liver evaluation. The number of platelets and white blood cells were inversely related to liver stiffness measurements supporting the need for a screening of portal hypertension in these patients. The AST to platelet ratio index was also correlated to ARFI elastography results suggesting that liver stiffness may reflect the evolution of liver fibrosis.
In conclusion, our study demonstrated, for the first time in literature, that ARFI elastography (pSWE) with ElastPQ software can be a useful tool to assess liver stiffness in patients with Fontan circulation and other congenital heart disease.

CB23

NUOVI ANTICOAGULANTI ORALI NEI PAZIENTI ADULTI CON CARDIOPATIA CONGENITA: STATO DELL'ARTE ED ESPERIENZA DEL NOSTRO CENTRO

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Introduzione: Sempre più cardiopatici congeniti adulti (GUCH) necessitano un trattamento anticoagulante. Le principali indicazioni al trattamento sono le aritmie e la profilassi tromboembolica primaria e secondaria. Le attuali opzioni disponibili sono gli inibitori dei fattori vitamina K dipendenti (VKA) e i nuovi anticoagulanti orali (NOAC) . L'aderenza al trattamento con VKA non è sempre ottimale considerate le loro peculiarità e la necessità di monitoraggio continuo. I NOAC sono stati proposti come valida alternativa tuttavia dalla recente letteratura emergono dati discordanti circa l'efficacia e la sicurezza. Lo scopo del nostro studio è stato quello di verificare l'efficacia e la sicurezza dei NOAC nei pazienti GUCH che necessitavano trattamento anticoagulante.

Materiali e Metodi: Sono stati inclusi nello studio tutti i pazienti GUCH che hanno iniziato un NOAC dal gennaio 2019. Sono stati esclusi pazienti che hanno iniziato il NOAC precedentemente o che erano già in trattamento con VKA. Il periodo di valutazione della sicurezza è stato fissato entro 30 giorni dall'inizio dei NOAC e a 1 anno. Il follow-up ha avuto luogo a 30 ± 7 giorni, 1 anno ±28 giorni. I principali endpoint considerati sono stati: eventi tromboembolici e sanguinamento maggiore. Come endpoint secondari sono stati considerati : sanguinamenti minori , mortalità, ospedalizzazione.

Risultati: Dal gennaio 2019 a gennaio 2020 sono stati reclutati 11 pazienti (età mediana 47 anni [IQR 35-61], 72% maschio) Tutti i pazienti hanno avviato NOAC a dosaggio pieno (apixaban 53%; rivaroxaban 18%; dabigatran 27%) per aritmie atriali e sono stati sottoposti a cardioversione elettrica dopo un mese. Le aritmie atriali erano la fibrillazione atriale (63%) e tachicardia atriale (9%) e flutter (27 %) .Il 93% aveva un punteggio CHA2Ds2-VASc di 0 e il 9 % (1 paziente) di 2. il 100% dei pazienti aveva un HASBLED, < 2.Complessivamente, il 18 % aveva circolazione di Fontan (connessione atriopolmonare con condotto intracardiaco), 9 % ipertensione polmonare e 36% aveva una storia di insufficienza cardiaca. Due pazienti avevano avuto un evento tromboembolico (TIA). A 30 giorni dall' inizio dei NOAC, nessun paziente ha sviluppato eventi tromboembolici o sanguinamenti maggiori. Il 27 % dei pazienti ha sviluppato un sanguinamento minore, epistassi (n = 2) e sanguinamento gengivale (n = 1), entro i primi 30 giorni. Tutti i pazienti con eventi avversi minori (n = 3) avevano una età < 65 anni con cardiopatia di grado moderato o complesso di cui una in circolazione Fontan. Ad un anno non si sono evidenziati sanguinamenti maggiori e/o eventi tromboembolici, 2 pazienti sono stati ricoverati per scompenso cardiaco (1 con circolazione di Fontan e 1 con Malattia di Ebstein).

Conclusioni: L'analisi della recente letteratura mostra risultati discordanti sull'efficacia e la sicurezza dei NOAC nei GUCH. Il nostro studio, seppur con numeri limitati e dati parziali mostra tassi di complicanze in linea con quanto fin ora riportato confermando un buon profilo di efficacia e sicurezza dei NOAC. Tuttavia alla luce dei dati fin qui disponibili sono necessari ulteriori studi prospettici e idealmente randomizzati in questa categoria di pazienti per validare l'uso dei NOAC.

Tabella 3 – Endpoint

Tipo di Cardiopatia	Sanguinamenti maggiori a 30 g	Sanguinamenti minori a 30 g	Sanguinamenti maggiori a 1 anno	Sanguinamenti minori a 1 anno	Eventi tromboembolici a 30 giorni	Eventi tromboembolici a 1 anno
Circolazione di Fontan	0	1	0	1	0	0
Ipertensione polmonare – Eisenmerger	0	1	0	1	0	0
Tetralogia di Fallot	0		0	1	0	0
Malattia Di Ebstein	0		0	0	0	0
TGA -S/P switch atriale sec. Mustard	0	1	0	1	0	0
DIA -S/p chiusura chirurgica	0		0	0	0	0

CB24

USO DI IVABRADINA IN TETRALOGIA DI FALLOT E TACHICARDIA GIUNZIONALE ECTOPICA POSTOPERATORIA

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Introduzione: L'ivabradina è un nuovo farmaco inibitore selettivo della corrente If del sodio; esso agisce principalmente a livello del nodo del seno provocando un effetto bradicardizzante senza effetti sull'inotropismo e sulla conduzione atrioventricolare. Il canale If, infatti, localizzato nelle cellule pacemaker, si attiva in iperpolarizzazione e consente al potenziale di membrana di diventare via via più positivo fino al raggiungimento del potenziale soglia per l'innesco del potenziale d'azione. Studi precedenti hanno in realtà dimostrato l'utilità di ivabradina non solo per rallentare la frequenza sinusale ma anche in caso di tachicardia atriale e tachiacrdia giunzionale ectopica.

Caso clinico: Il nostro paziente, affetto da Tetralogia di Fallot, viene sottoposto all'età di 5 mesi ad intervento chirurgico di correzione con patch transanulare e chiusura DIA tipo seno coronarico. Durante la degenza in terapia intensiva postoperatoria presenta numerosi episodi di tachicardia sopraventricolare, cardiovertiti con adenosina. Viene intrapresa terapia con amiodarone ev, con discreta risposta; all'ECG si osservano frequenti extrasistoli sopraventricolari isolate e ripetitive (coppie e triplette). Terminata l'infusione ev si intraprende terapia per os con amiodarone e si associa betabloccante, non ottenendo però un buon controllo del ritmo; inoltre la presenza di episodi numerosi e sostenuti di aritmia ad elevata frequenza si ripercuote sui parametri emodinamici del piccolo. Per la persistenza degli episodi aritmici si intraprende terapia con ivabradina alla dose di 0,02 mg/kg per os 2 volte al giorno. Si osserva ritmo sinusale stabile per cui successivamente si sospende terapia con amiodarone e si continua quella con ivabradina. Non si osservano aritmie né durante la degenza né nel successivo follow up dopo la dimissione, effettuato con Holter ECG seriati. Dopo circa 3 mesi la terapia viene sospesa, in assenza di aritmie ipercinetiche sopraventricolari e ventricolari.

Conclusioni: L'ivabradina è un farmaco inibitore selettivo del canale del sodio a livello delle cellule pacemaker; nato come farmaco bradicardizzante per la sua azione sul nodo del seno, è stata dimostrata la sua utilità anche nel trattamento di aritmie sopraventricolari da esaltato automatismo e da rientro atriale. È ipotizzabile quindi la presenza dei canali If anche in foci ectopici, che ne consentirebbe l'utilizzo anche come vero e proprio farmaco antiaritmico, sfruttando la sua peculiare azione non interferente con l'inotropismo e la conduzione atrio ed intraventricolare.

CB25

EFFECTIVENESS OF ANTIARRHYTHMIC THERAPY IN FETUSES WITH DIAGNOSIS OF LONG VA TACHYCARDIA

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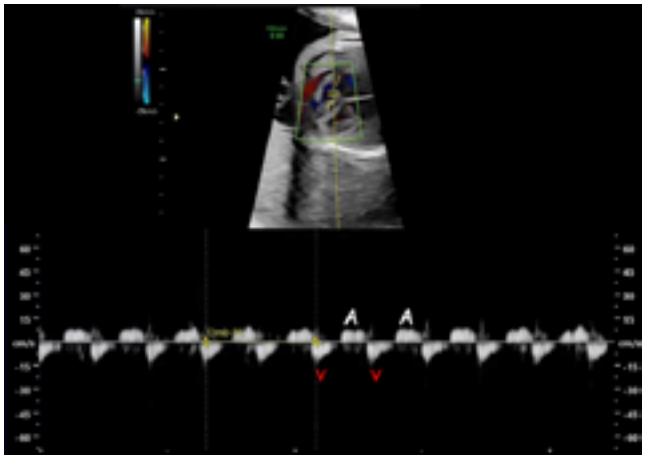
Introduction: Long ventriculo-atrial (VA) tachycardias, as ectopic atrial tachycardia (EAT) and permanent junctional reciprocating tachycardia (PJRT) are rare in fetal life. They are more refractory to treatment with worse prognosis compared with more common types of supraventricular tachycardia (SVT), as atrioventricular reentrant tachycardia or atrial flutter. Antiarrhythmic drugs as flecainide or digoxin are usually administered in case of SVT but, due to the lower incidence of long VA tachycardia, limited data are available about the optimal therapy in this specific setting during intrauterine life. The aim of this study was to evaluate the effectiveness of early maternal administration of antiarrhythmic therapy in fetuses with diagnosis of long VA tachycardia referred at our tertiary referral center.

Methods: Among 114 fetuses with diagnosis of fetal arrhythmia referred to the Paediatric Cardiology Unit of our Institute between January 2014 and December 2021, we selected 8 fetuses (7%) with long VA tachycardia, including 7 fetuses with diagnosis of EAT and 1 fetus with diagnosis of PJRT. Fetal echocardiography was performed to evaluate the fetal heart rhythm using either M-Mode, pulsed wave Doppler and tissue Doppler imaging. EAT was diagnosed in the presence of "warming-up" and "cooling-down" phenomena with variable AV relationship, instead PJRT was suspected in case of long VA interval with fixed 1:1 AV relationship and sudden onset and offset. Three fetuses (37%) presented with signs of heart failure (n = 2) or hydrops fetalis (n = 1) due to tachycardia. Mean heart rate during EAT was 224 ± 27 bpm. PJRT had a mean heart rate of 250 bpm. Oral transplacental antiarrhythmic therapy using flecainide or digoxin was started as soon after diagnosis (mean gestational age 27.2 ± 5.3 weeks).

Results: Flecainide was the most used drug in this study, obtaining in our patients with EAT 100% of complete rhythm control (5/5), including those with hemodynamic impairment. Fetal heart failure and hydrops fetalis were completely solved thanks to antiarrhythmic therapy in all 3 fetuses. Digoxin failed in the other two patients with EAT, therefore it was successfully replaced by flecainide and sotalol. Digoxin was only effective in onepatient with PJRT. No adverse effect requiring drug discontinuation occurred in all mothers. Outcome was good for all newborns after birth.

Conclusion: In our cohort of patients, oral maternal antiarrhythmic therapy resulted to be safe and effective in fetuses with diagnosis of long VA tachycardia. Our preliminary experience suggests that flecainide can be successfully used to treat EAT, including fetuses with signs of hemodynamic impairment, improving outcome.



CB26

ALVT A CASE REPORT

C. Morelli [1], F. Fratta [1], F. Del Gaizo [1], L. Di Pietto [1], A. Giudicepietro [1], M. Giordano [1], L. Spinelli Barrile [2], A. Ammendola [2], F. De Fazio [1], M.G Russo [1], G. Oppido [3]

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Introduction: The aorto left ventricular tunnel (ALVT) It is defined as a rare congenital heart disease (CHD) in which where is a paravalvular communication between the aorta and the left ventricle. The incidence is unknown, estimates ranging from 0.01% to 0,5% of (CHD). The prenatal diagnosis is possible but is challenging and should be considered in any fetus with aortic regurgitation. We describe a case of a 33-year-old pregnant woman sent to our reference unit to perform a fetal echocardiography for suspected aortic stenosis at 24 weeks of gestation.

A pregnant woman of 33 year's old was invited to perform a fetal echocardiographic examination in our Pediatric Cardiology Unit of the Monaldi's Hospital, University 'L. Vanvitelli' of Naples, for suspected aortic stenosis at ultrasound screening scan performed at 21 weeks of gestation. Extracardiac fetal anatomy appeared normal. The fetal echocardiographic was performed at 22 weeks of gestation. The four-chamber scan appeared abnormal. The left ventricle was dilated and hypokinetic with signs of fibroelastosis. There was mild mitral regurgitation. In left ventricular outflow tract (LVOT) were seen subaortic and supravalvular defects. (fig 1). The aortic valve appeared dysplasic, Doppler color flow demostrating regurgitation around LVOT and consequent rapid run-off of blood. In right ventricular outflow tract (RVOT) the pulmonary artery appeared normal. At Doppler color flow in three vessels view (3V) the ascending aorta appeared dilated. In three vessels and trachea view (3VT) and in sagitall view of aortic arch was found regurgitation. With these elements Aorto-Left ventricular tunnel was suspected. Although no associated genetic syndromes are known, invasive technique by amniocentesis was offered. The couple agreed to perform it and the result was normal (46XY). Follow-up was performed every 4 weeks and there were no signs of heart failure but at 37 weeks were also clearly visualized apical trabeculae in the myocardium of the left ventricle. At 39 weeks of gestation birth a male infant of 3112 g by Cesarean section. The infant was immediately transferred to Monaldi's Hospital in Pediatric Cardiology Unit and the prenatal diagnosis of ALVT was confirmed with a bicuspid and dysplastic aortic valve without signs of stenosis. The left ventricular appeared spongy with presence of middle apical trabeculae and systolic function was slight reduction.tunnel entry orifice misured 4 mm, the tunnel exit orifice misured 12 mm . Cardiac surgery was performed on day 3 of life. In median sternotomy under cardiopulmonarybypass, the ALVT was closed through its longitudinal incision, direct closure of direct communication on the ventricular side and by patching of heterologous pericardium on the aortic side, DIA closure os with patch, ductus arterial closure, Bicuspid and dysplastic aortic valve.The post-operative course was uneventful and the infant was discharged at one month and one week. Duration of ICU and hospital stay were 6 and 36 days, respectively.

CB27

SELECTIVE LEFT VENTRICLE PACING IN PEDIATRIC PATIENTS WITH CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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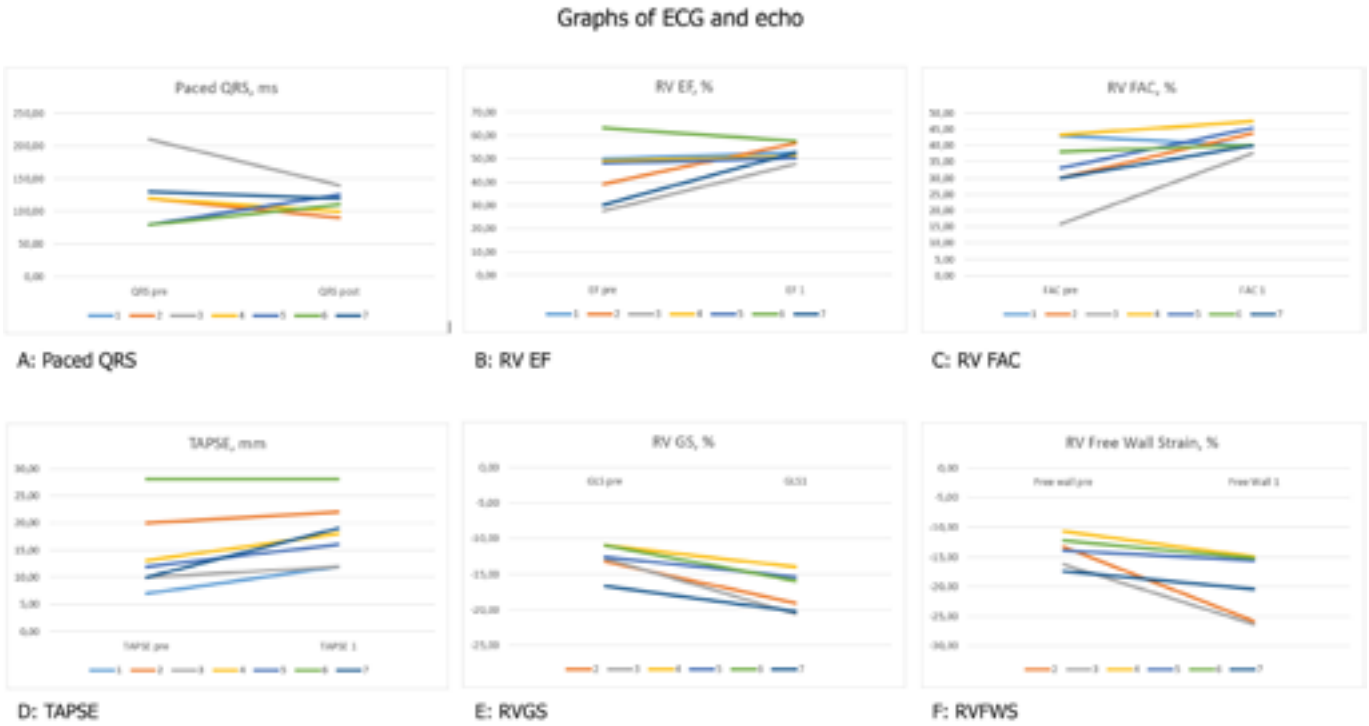
Background: Congenitally corrected transposition of the great arteries (CCTGA) has a systemic right ventricle (RV). Atrioventricular block (AVB) and systolic RV dysfunction frequently occur. Permanent pacing leads positioned in the subpulmonary left ventricle (LV) may worsen RV dysfunction. Selective pacing has been shown to preserve systolic function in children.

Objective: Aim of the study was to seek out if three-dimensional-electroanatomic mapping systems (3D-EAM) guided selective LV septal pacing preserves RV systolic function in CCTGA patients with AVB.

Methods: Retrospective analysis of CCTGA patients who underwent EAM-guided selective LV septal pacing. A 3D-pacing map guided ventricular lead implantation in septal sites with narrower paced QRS. ECG and echocardiograms were obtained following the pacemaker implantation up to 1 year to evaluate the RV function: ejection fraction (EF), fractional area change (FAC), tricuspid annulus plane systolic excursion (TAPSE), tricuspid lateral annular systolic velocity (S'-TDI), RV global strain (RVGS), RV free wall longitudinal strain (RVFWLS). Data are reported as median (25th-75th centiles).

Results: 7 CCTGA patients aged 15 (9-18) years, with complete/advanced AVB, underwent LV selective pacing (5 DDD, 2 VVIR). No procedural complications occurred. Ventricular pacing was >90%. At 12-month post-implantation there was a significant increase of FAC, TAPSE, RVGS, RVFWLS. The improvement of EF and S'-TDI was not significant. QRS duration at baseline and post-selective pacing showed no significant changes: 120 (80-120) ms vs. 120 (100-125) ms. Figure 1.

Conclusion: 3D-EAM-guided selective LV septal pacing preserved RV systolic function in paediatric patients with CCTGA and AV block after short-term follow-up.



CB28

TRANSVENOUS LEAD ADVANCEMENT IN PAEDIATRIC PACING TO OVERCOME GROWTH STRETCHING

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Background/Introduction: One of the main complications of transvenous leads implanted in paediatric patients is the stretching of the lead caused by the somatic growth. It may cause pacing and sensing defects and lead dislodgement or even fracture. Absorbable lead ligature and atrial loop may reduce this risk. However, the loop may induce traction or may unroll too early and therefore impair lead function. Lead extraction and replacement is another solution, although it has some procedural risks in young patients. Lead advancement through pushing it from the pocket may solve growth-induced traction and spare the electrode throughout childhood until post-puberty.

Purpose of the study is the retrospective analysis of the outcome of the transvenous lead advancement in children with a pacemaker (PM) in a single tertiary paediatric center.

Methods: consecutive patients with a VVIR PM implanted for isolated congenital complete atrioventricular block (no structural heart disease) in alternative right ventricular (RV) pacing sites, with lead stretching underwent a trial of lead advancement during general anaesthesia, cefuroxime antibiotic profilaxis, from 2014 to 2021. After venous angiography showed venous patency, the PM pocket was opened, the lead was released from subcutaneous adherences and with a stylet was gently advanced to create a semi-loop in the atrium without dislodging the tip. Lead data (threshold, sensing, impedance) were compared before and after the procedure. Data are expressed as median (25th-75th centiles)

Results: 8 patients underwent PM implantation at 6.8 (5.8-8.0) years of age, 20 (18-21) kg, 116 (106-120) cm, with the lead positioned at parahisian(3)/mid-septum (5 pts) sites. During a follow-up of 3 (1-5) years, advancement procedures were 1.5 (1.0-4.3) per patient. Between procedures, delta age was 16 (12-20) months, height 8 (7-11) cm and weight 5 (3-7) kg. All leads were successfully advanced without any procedural complications. Procedure time (skin to skin) was 96 (73-108) minutes, fluoroscopy was 0.5 (0.2-1.4) mGy, 16 (10-46) microGy/m2. Electrical lead parameters did not showed significant differences between consecutive control times.

Conclusion: the advancement of transvenous leads in children is a safe and effective procedure, with low fluoroscopy exposure, without significant acute and chronic complications. This procedure seems to preserve transvenous leads until growth has completed.

CB29

ABLAZIONE TRANSCATETERE CON RADIOFREQUENZA E CRIOENERGIA DELLE VIE ACCESSORIE LATERALI DESTRE IN ETÀ PEDIATRICA: EFFICACIA E SICUREZZA IN ACUTO E NEL FOLLOW UP A MEDIO TERMINE

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Introduzione: In letteratura l'ablazione transcatetere con radiofrequenza (RFTA) delle vie accessorie (VA) laterali destre in età pediatrica mostra un'efficacia in acuto del 92-95%, con successo a distanza del 72%. La crioablazione evidenzia un ottimo successo in acuto (97%), con un tasso di recidiva del 20%. Lo scopo di questo studio è valutare il tasso di successo in acuto e a lungo termine, nonché i fattori di rischio per recidive.

Metodi: Sono stati analizzati retrospettivamente i dati clinici e procedurali di una coorte di 53 pazienti in età pediatrica con VA laterale destra, sottoposti a RFTA o crioenergia da gennaio 2010 ad aprile 2022. La VA è stata identificata in 3 gruppi: laterale destra (LDx), anterolaterale destra (ALDx) e posterolaterale destra (PLDx).

Risultati: Su 53 pazienti, che presentavano almeno una VA (57% maschi; età media 12.2 anni, peso 47.5 kg), un caso era associato con anomalia di Ebstein. Venticinque pazienti (47.2%) appartenevano al Gruppo LDx, 14 (26.4%) al Gruppo ALDx e i rimanenti (14, 26.4%) al Gruppo PLDx. In 4 casi vi erano VA multiple (7.5%). La VA era manifesta nel 72% ed occulta nel 28% dei casi.
La RFTA è stata eseguita in 36 casi (68%) con successo in acuto del 86%, mentre la crioablazione in 17 pazienti (32%) con successo in acuto del 82%. La tabella 1 mostra il tasso di successo in acuto in FR e crioablazione rispettivamente nei gruppi LDx, ALDx e PLDx. In 8 su 53 pazienti abbiamo registrato un insuccesso nella prima procedura ablativa. Solo 4 pazienti su 53 (7.5%) hanno mostrato recidive nelle prime 24 ore. Una ricorrenza a distanza si è presentata in 8 pazienti (22%; 4 pazienti gruppo LDx, 1 gruppo ALDx e 3 gruppo PLDx), sottoposti a RF come prima ablazione. L'approccio mediante accesso venoso giugulare destro è stato utilizzato in 11 su 53 casi (21%), con VA ALDx, documentando un tasso di successo completo (100%) indipendentemente dal tipo di energia erogata. Abbiamo registrato una sola complicanza: fistola artero-venosa nella sede della puntura femorale. Tempo mediano di fluoroscopia nella prima procedura 0.4 minuti in RF vs 0.6 minuti in crioablazione. Un totale di 16 pazienti è stato sottoposto a nuova procedura ablativa, con un'efficacia in 10 mediante RF (83%) ed in 4 con crioablazione (100%). Anche dopo procedure multiple, abbiamo evidenziato un tasso di insuccesso a distanza (follow-up 28 mesi) del 22% (5 pazienti VA LDx e 3 VA PLDx) nelle procedure in RF e del 12% (2 pazienti VA PLDx) in crioenergia.

Conclusioni: L'ablazione della VA destra ha mostrato una maggior efficacia in acuto nei casi con localizzazione laterale e antero-laterale, con ottimi risultati anche in crioenergia. Indipendentemente dal tipo di energia erogata, il principale insuccesso sia in acuto sia a distanza è legato ad una VA PLDx. Il tasso di successo a distanza rimane limitato, paragonabile ai dati in letteratura, registrando migliori risultati con l'uso della crioablazione. L'ablazione delle VA laterale destre si conferma una procedura sicura nella popolazione pediatrica indipendentemente dal tipo di energia utilizzata ed accesso vascolare.

Tabella 1: Tasso di successo in acuto in FR e crioablazione in base alla localizzazione della via accessoria.

	RF		Crioablazione	
Localizzazione VA	Procedura, n (%)	Successo in acuto, n (%)	Procedura, n (%)	Successo in acuto, n (%)
LDx	18 (50)	17 (94.5)	7 (41.2)	6 (85.7)
ALDx	6 (16.7)	6 (100)	8 (47)	7 (87.5)
PLDx	12 (33.3)	8 (66.7)	2 (11.8)	1 (50)
Totale	36 (100)	31 (86)	17 (100)	14 (82)

CB30

ICD IN PEDIATRIC ARRHYTHMOGENIC CARDIOMYOPATHY: INDICATIONS AND OUTCOME: A SINGLE CENTER EXPERIENCE

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Arrhythmogenic cardiomyopathy (ACM) is a very rare condition among pediatric patients. Once diagnosed, the risk stratification is mandatory in order to prevent sudden cardiac death which can sometimes be the first manifestation of the disease as well. Moreover, it must be periodically reassessed due to the progression of the disease.

Metodi impiegati: In this single center retrospective observational study, all pediatric patients (<18 y.o.) diagnosed with ACM since 2009 in Our Institution were collected. Only the patients with a confirmed diagnosis according to the "Padua Criteria" were enrolled. ICD implantation was decided according to the current recommendations/clinical studies, and outcome was recorded during follow-up.

Risultati e Conclusioni: Twenty-two patients had a "definite" (21) or "borderline (1) ACM diagnosis (13 females) according to the "2020 Padua Criteria". Out of them, 16 patients (72.7%) received an ICD: 3 (18.8%) a transvenous device and 13 (81.2%) a subcutaneous one (S-ICD). In 4 (25%) patients, ICD was implanted for secondary prevention (2 hemodynamically unstable sustained VT and 2 hemodynamically stable sustained VT). In the other 12 (75%) patients, it was implanted for primary prevention: in 2 with class I indication (severe right ventricular dysfunction), in 8 with class IIa indication (syncope, non-sustained VT, moderate ventricular dysfunction) and in 2 with class IIb due to several "minor" risk factors.
During the follow-up (5.59 ± 3.4 years), appropriate DC-shocks were delivered only in 3 (18.8%) patients for sustained VTs, without defibrillation failures. Inappropriate shocks occurred in 2 cases: one for T-oversensing and one for high-rate sinus tachycardia exceeding the VT limit.

Conclusions: ICD is indicated in the majority of pediatric patients due to the high risk of SCD. Despite that, appropriate ICD therapies occurred in only a minority of patients. The rate of inappropriate shocks is even more rare. This reassuring follow-up may be due to the efficacy of medical antiarrhythmic and anti-remodeling therapy, promptly started once the diagnosed is made. Of course, other risk factors and protective ones remain still unknown and a specific pediatric risk stratification model is advised in the future.

CB31

CLINICAL PROFILE OF CARDIAC ARREST SURVIVORS IN A PEDIATRIC POPULATION: EXPERIENCE IN A TERTIARY REFERRAL CENTER

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Background: Sudden cardiac death (SCD) in pediatric patients is rare: a recent European study targeting a population under 18 yrs reports an estimated incidence of 1:100.000 person/year. Notably, death was unexpected in more than 70% of victims and was frequently triggered by effort. Although a cardiac cause is suspected in the large majority of cases, autopsy results are negative in about 40%. As many uncertainties remain, important lessons can be learned from patients surviving potentially lethal arrhythmias.

Aim: In order to assess the clinical and genetic profile of a series of pediatric cardiac arrest survivors.

Patients and Methods: From September 1987 to May 2022 we studied a population of 12 patients, 7 females, age ranging from birth to 18yrs, seen at our cardiac outpatient clinic following resuscitated sudden cardiac arrest.

Results: In four patients (zero to four yrs) a torsade de pointes was the recorded arrhythmia; in all genetic study we found mutations associated with LQTS: respectively LQT1 in a four yrs female pt, LQT3 in a female newborn, a Calmoduline mutation (CALM1) in a female infant, and a Jervell Lange Nielsen syndrome in a four yrs male child. In four children, 2 males and two females, age 8-14 yrs, all with events during effort, a diagnosis of polymorphic ventricular tachycardia was confirmed by genetic studies reporting RYR2 mutations; a male patient aged 6 had a short-coupled torsade de pointes degenerating to ventricular fibrillation. He was resuscitated by an ICD previously implanted for syncope and familial history of sudden death; a male patient with hypertrophic cardiomyopathy aged 14 was resuscitated for ventricular fibrillation and later required heart transplantation: genetic test showed two mutations in MYH7 and SCN5A. A 14-years old girl affected by tuberose sclerosis, carrying an ICD, presented with ventricular fibrillation, despite the absence of cardiac rhabdomyomas on the echocardiogram. ICD interrogation showed that the episode begun with high frequency atrial fibrillation, later degenerating into VF. One female patient aging 13 had asystole during effort. Following a diagnosis of hypoplastic coronary ostium of LAD, she underwent cardiac transplantation.

Conclusions: The substrate and triggers of cardiac arrest in children are heterogeneous and often involve rare genetic conditions. Appropriate work-up may reveal the etiology of the underlying disease, with crucial potential for cascade screening in families. These results on cardiac arrest survivors might inform operational algorithms such as molecular autopsies in victims, in order to minimize current gaps in knowledge on pediatric sudden death.

CB32

ROLE OF FETAL ECHOCARDIOGRAPHY IN PRENATAL SURGICAL PREDICTION IN DOUBLE OUTLET RIGHT VENTRICLE

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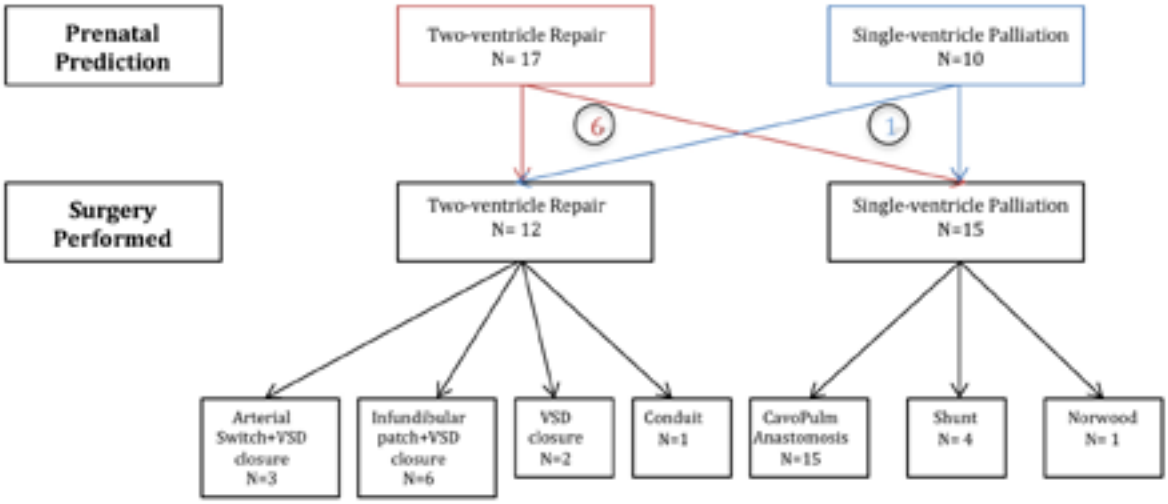
Objective: Double outlet right ventricle (DORV) is a complex conotruncal cardiac malformation which affect less than 1% of all congenital heart disease. Prenatal diagnosis has high accuracy assessed by fetal echocardiography. Recently literature has proved that proper fetal diagnosis contents parental counseling in order to clearly define prognosis and postnatal surgical approach, regarding uni or biventricular repair.

The aim of our study was to define if accuracy in prenatal echocardiography diagnosis can improve parental counseling predicting the type of surgical correction required.

Design: Medical records of patients with fetal echocardiography diagnosis of DORV at our institution from 2009 to 2022 were analysed. Pre and post natal diagnosis were matched. The surgical prognosis, distinguishing between two-ventricle repair and single-ventricle palliation, was compared with the postnatal approach.

Results: Our population consisted of 41 fetus with none in utero deaths, 2 terminations of pregnancy and 39 live births. We compared postnatal echocardiograms or autopsy results of all these patients. The accuracy of fetal echocardiography in making a correct diagnosis of DORV was 90%.
In our study population 27 (66%) patients had surgery; of those who were not included: 4 had a different postnatal diagnosis; 6 died before undergoing corrective surgery; 3 was lost to follow-up and 1 was recently born. Among patients who underwent surgery, 44% had two-ventricle repair and 56% had single-ventricle palliation. Accurate prenatal prediction of surgical approach was made in 74% of this population.

Conclusion: DORV is a rare and often complex cardiac anomaly that can be diagnosed prenatally with high precision. By implementing diagnostic accuracy of fetal echocardiography, important anatomical details can be determined which allows accurate prenatal counseling regarding surgical prognosis.



CB33

PREGNANCY AND DELIVERY OF ADULT CONGENITAL HEART DISEASE (ACHD) WOMEN: A SINGLE CENTER EXPERIENCE

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Background: The number of adult congenital heart disease (ACHD) patients is constantly growing up, as the result of the increased survival of patients with complex CHD. In this population, the number of women having a pregnancy is also growing up, requiring specific counselling, follow-up and care before and during pregnancy until childbirth.

Aim: To report data about childbirths of ACHD women during last years at a single regional reference center where these patients can have follow-up during pregnancy until delivery.

Methods: We reviewed data about childbirths of ACHD women from January 2018 to April 2022 at our center. Data collected for patients: demographic data, modified WHO and NYHA class, diagnosis, cardiac surgery or interventional procedure, use of cardiovascular drugs during pregnancy, type of delivery and indication, peripartum complications. Data collected for neonates: gestational age (GA), Apgar score, birth weight, diagnosis of CHD.

Results: During the reference period, 23 ACHD women gave birth to their babies at our center, after a follow-up during pregnancy. The median age was 34.1 years (range 26-46). Patients belonged to I to IV modified WHO risk classes and to I to III NYHA classes. Diagnoses were: Fontan circulation, pulmonary hypertension in repaired atrioventricular septal defect (AVSD), bicuspid aortic valve with dilation of the ascending aorta, aortic valve stenosis, repaired tetralogy of Fallot, pulmonary valve atresia after surgical valvotomy, surgically repaired coarctation of the aorta (CoA), transposition of the great arteries after arterial switch operation, repaired total anomalous pulmonary venous return, repaired septal defects, repaired aortic and pulmonary stenosis, mitral valve prolapse. 15 women underwent at least one cardiac surgery procedure, 1 a cardiac interventional procedure, 7 no procedure. Use of cardiovascular drugs during pregnancy was recorded in 5 patients. All women had singleton pregnancies. A vaginal delivery was observed in 5 women (21.7%), spontaneous in 4 and operative in 1; a cesarean section was performed in 18 women (78.3%). Indications for cesarean section were obstetrical in 44.4% and cardiologic in 55.6%. Patients did not present peripartum complications due to CHD. Mean GA at delivery was 38 weeks and 2 days (range 33+6/40+1, 3 preterm births). Apgar score was 7-10 at 1 minute in 22 neonates and at 5 minutes in all. Birth weight was adequate for GA in all except 2 cases: 1 small for GA (SGA) and with intrauterine growth restriction (IUGR), 1 large for GA (LGA). Among neonates, there were 3 cases of CHD (prenatally diagnosed): critical pulmonary stenosis, CoA, ventricular septal defect.

Conclusions: In our population, we did not record any significant maternal peripartum complication due to CHD. Cesarean section had a high frequency, but in quite half of cases the indication was obstetrical. Among neonates we reported preterm birth in few cases and one SGA and IUGR newborn. The recurrence rate of CHD is higher than usually reported. The small number of cases could limit the relevance of data.

CB34

CONJOINED TWINS WITH HEARTS OR HEART MALFORMATION? ROLE OF MULTI-MODALITY IMAGING AND REVISION OF THE LITERATURE

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A 26 years old G000 female requested a second opinion consultation at 13 6/7 weeks' gestation for suspected conjoined twins at first level obstetric scan. Evaluation included detailed trans-abdominal and trans-vaginal ultrasound performed by maternal-fetal medicine specialist, trans-abdominal and trans-vaginal fetal echocardiogram performed by pediatric cardiologist with expertise in fetal cardiology and multidisciplinary consultations. Ultrasound findings included evidence of monocorial/monoamnyotic twin pregnancy, confirmation of thoracopagus conjoined female fetuses, evidence of shared heart and liver and of facial anomalies in one twin. Fetal echocardiography revealed the presence of complex congenital heart disease. Parents refused invasive genetic test and they chose for termination of pregnancy. Postmortem micro-CT was performed in order to correctly define heart defect and associated anomalies.

Aims: This paper aims to systematically review the literature regarding conjoined twins (CT) and the use of multimodality imaging in the diagnose and estimation of prognosis.

Methods: A systematic literature review was conducted to identify publications concerning CT, prenatal diagnosis of CT and in utero multimodality imaging.

Results and Conclusion: CT are rare anomaly. Prevalence has been estimated to be 1 in 200,000 births. They are divided in symmetric (more frequent) and asymmetric (parasitic) and are classified according to side of fusion and shared organs, with thoracopagus and omophalopagus type the most frequent (70%). Etiology is uncertain. Female predominance has been observed with a ratio of 3:1. "Fusion" and "fission" theories are postulated to explain the mechanism but an universal consensus lacks. Other defects (neural tube defects, congenital heart disease, genitourinary anomalies musculoskeletal defects, gastrointestinal malformations) are frequently associated. Survival of CT is precarious and depends mainly on the type of CT, the shared organs and the possibility of surgical treatment. Most of them die in utero or during the very early perinatal period. Prenatal diagnosis is possible, even if challenging. Prenatal multimodality imaging is essential to reach all prognostic information. In utero multidisciplinary management is mandatory to reach precise diagnosis of type of CT and of shared organs and to assess the postnatal feasibility of separation. Counseling should start as soon as the diagnosis of conjoined twins is made.

CB35

SUCCESSFUL IMPLANTATION OF A MICRA LEADLESS PACEMAKER IN A YOUNG PATIENT WITH SURGICALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES

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Introduction: Late onset of complete heart block is a potential complication after heart surgery. Leadless pacemakers have emerged in recent years as an alternative to the conventional ones. The Micra® transcatheter pacing system TPS (Medtronic, Minneapolis, MN, USA) is the only leadless pacemaker currently approved by the FDA for use in the USA.
We present the case of successful implantation of Micra ® in a CHD pediatric patient.

Case report: We present the case of a 16year-old female with D-TGA with VSD who had underwent neonatal arterial switch operation (ASO). During follow-up, no significant residual defects were noted. The girl grew up well, participating in sport. At the age of 16, she started complaining of episodes of dizziness without loss of consciousness. The basal ECG showed sinus rhythm. At echocardiography no significant residual defects were noted. The Holter ECG registration was performed, and paroxysms of advanced heart block with pauses up to 15 seconds were revealed. To exclude anatomical substrates, a cardiac MRI was performed and confirmed the lack of significant abnormalities. The implantation of a permanent pacemaker was then planned. A leadless pacemaker was selected for its small size, for its minimal cosmetic concerns and because it could allow the patient to practice physical activity. The procedure was performed under general anesthesia, with two femoral accesses. The device was positioned in the middle part of the ventricular septum. There were no complications, and both echocardiogram and chest X-ray confirmed the corrected position. The patient was discharged in 3rd day after implantation. She restarted sport activity without psychological conditioning. At midterm control, the device was working properly with good parameters and low percentage of stimulation.

Discussion: The implantation of traditional pacemaker is a complex issue in CHD because of the anatomy which often requires epicardial devices; moreover it is complex in pediatric patients who are a special population for their growth and their needs. Literature about leadless implantation in children is scarce, but all the studies seem to confirm that this approach significantly reduces complications. Our case demonstrates for the first time in the world the feasibility and efficacy of this procedure in a girl with D-TGA. The limitation of this procedure is the potential necessity to remove the device in the future (the Micra ®AV has a projected longevity of 12 or more years). Other open issues are the possibility of vascular complications secondary to the large delivery system and the absence of atrial pacing.

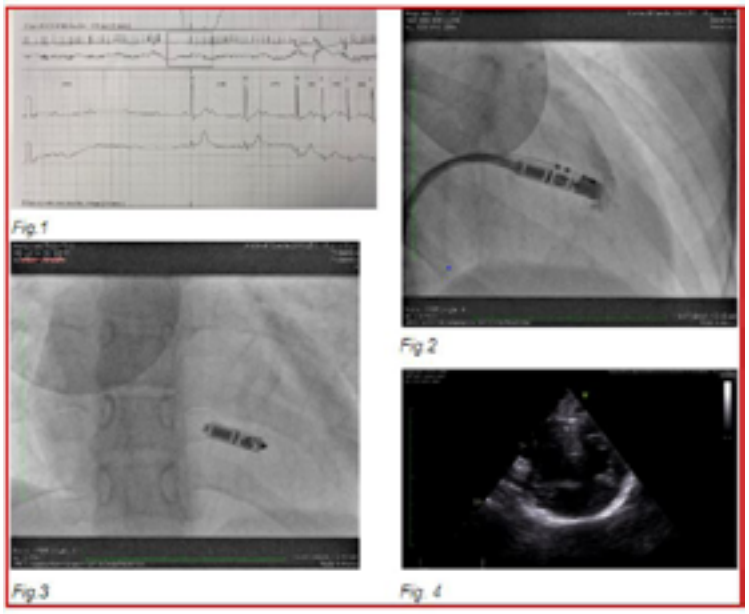

Fig 1: ECG showing a 15'' pause
Fig. 2 (2A): device with delivery system
Fig. 3(2A): device in correct position after removing delivery system
Fig 4: 2D echo short axis view with device in the septum

Conclusions: Leadless pacemaker implantation was feasible and effective in a young girl with transposition of the great arteries after arterial switch operation. This may be used as a reference in similar cases, when a pacemaker implant is required at a young age, if anatomy allows it and if no high percentage of pacing is necessary.
Long term follow-up studies and a larger number of patients are required.

Declaration of competing interest
There are no conflicts of interest.

CB36

SUPRAVENTRICULAR AUTOMATIC TACHYCARDIAS IN PAEDIATRIC AGE: A RETROSPECTIVE STUDY OF 23 PATIENTS IN A TERTIARY MEDICAL CENTRE.

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Background: Supraventricular automatic tachycardias (SAT) represent about 15% of supraventricular tachycardias in paediatric age. To date studies concerning SAT consist of small series of patients; compared to re-entry tachycardias, resistance to drug therapy and a tendency to become incessant should be noted. In this study our intent was to describe a series of SAT in a mid-term follow-up.

Material and Methods: From April 1995 to September 2021 23 patients (14 females and 9 males) from birth to 18 years were observed in a tertiary medical centre because of SAT, which include focal atrial tachycardias (FAT), multifocal atrial tachycardias (MAT), junctional ectopic tachycardias (JET) and atrial fibrillations (AFib); we considered them together, because of common electrophysiological mechanisms; electrocardiographic aspects, clinical symptoms, response to acute and long term treatment were considered.

Results: The diagnostic criteria for FAT were met in 16 patients, MAT in 4, JET in 1 and AFib in 2.
FAT occurred earlier (median 19 days old) than the other forms (8 years for MAT and 6 years for Afib); JET started during foetal period and continued after childbirth. Clinical presentation was paucisymptomatic in most cases (74%) and the median maximum heart rate at the onset was 215 bpm. A stable sinus rhythm was reached in a minority of cases with only one anti arrhythmic drug: 57% of patients required multiple antiarrhythmic agents with a median time to cardioversion of 10 days. The most successful strategy was the association of a beta-blocker with flecainide (6/16 FAT, 1/4 MAT, 1/2 AFib). In 2 FAT we must use 4 antiarrhythmic drugs to sinus conversion. JET was cardioverted by ivabradine. At a median 49 follow-up months 21 pts (91%) are in sinus rhythm and 15 (65%) still in maintenance therapy. In 2 pts with FAT evolved in chronic atrial fibrillation.

Conclusions: Our experience confirmed that in SAT time to conversion to sinus rhythm is usually longer than in re-entry tachycardias; moreover in a mid-term follow-up the use of a combination of anti-arrhythmic drugs is often needed; finally in rare cases it may happen an evolution in atrial fibrillation: in these patients it seems necessary to investigate about genetic aspects, identifying which genes may be involved in promoting the onset of tachycardia and influencing the prognosis.

CB37

IS LATE COMPLEX SURGERY IN SYSTEMIC RIGHT VENTRICLE AFFORDABLE?

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Introduction: Some congenital heart diseases (CHD) are characterized by a systemic right ventricle (SRV). The most common clinical scenarios are: complete transposition of the great arteries (TGA) previously treated with atrial switch repair, congenitally corrected TGA, anatomical/functional single right ventricle with Fontan palliation. The management of the dysfunction remains an ongoing challenge and the heart transplantation represents the only option in end-stage heart failure. However, patients with SRV frequently undergo aortic dilation which may need additional surgical procedures several decades after first correction. Surgeons are concerned in regard to use cardioplegic arrest on these ventricles as it may contribute to function decline. The usual solution adopted is waiting for heart transplant.

Case report: Herein we describe two similar cases of patients with SRV who have undergone reoperation for aortic root dilatation. No similar cases have been described in the literature.
The first patient was a 36-year-old man who had undergone a Fontan palliation for unbalanced complete atrioventricular canal defect with right dominance, transposition of the great arteries, right aortic arch and right isomerism. Follow-up cardiac MRI revealed a diameter of 52 mm, 58 mm, 67 mm at the aortic root, sino-tubular junction and ascending aorta respectively. Furthermore, the echocardiogram showed a severe aortic regurgitation, a mild regurgitation of the common atrio-ventricular valve and an EF of 55%. The patient was in NYHA class II. An aortic root replacement with a mechanical Bentall operation was performed.
The second case was a 36-year-old man with an aortic root dilatation (47 mm) and a severe aortic regurgitation after a Senning operation that was successfully managed by a mechanical Bentall procedure. The preoperative echocardiogram showed a volume overload of the SRV with a mild reduction of the systolic function (shortening fraction 40%), a severe tricuspid valve regurgitation (TR) secondary to annular dilatation (42 x 52 mm) and tethering of the leaflets. In both cases a Del Nido cardioplegic solution was preferred.
The postoperative course of both patients was uneventful, they were discharged home in good conditions. At the follow-up the patients were in NYHA functional class I. They did not show worsening of ventricular function and the second patient demonstrated an improvement in the degree of TR (moderate).

Discussion and Conclusion: There is an increasing population of ACHD patients with a post-surgical SRV. Their underlying pathology is often associated with late aortic aneurysm formation and possibly aortic regurgitation. The surgical management of these patients can be very complex and challenging because it exposes the SRV to a further ischemic arrest period. Despite waiting for heart transplantation is the most adopted approach, in our opinion, additional surgical correction carries an acceptable risk and provides good early outcome.

CB38

HOME MONITORING IN PAEDIATRIC HEART FAILURE (HOPE-HF): SINGLE-ARM PROSPECTIVE STUDY

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Introduction: Telemedicine have improved prognosis in adult patients with chronic HF. During pandemic COVID 19, the rate of cardiovascular death increased. Currently, no studies have been conducted on children.

Hypothesis: To affirm the feasibility of a new model of tele monitoring in a paediatric population with high-risk HF and in waiting list for heart transplant (HTx).

Methods: Chronic paediatric HF pts with advanced NYHA class and/or waiting list for HTx received a system of continuous tele-monitoring of vital parameters (heart rate, arrhythmias, systemic blood pressure, oxygen saturation, breathe frequency) through a patch applied on the chest. Data were sent to "virtual clinic" and daily analysed by a specialized nurse, who notified to clinicians allowing them to detect critical signals of a deteriorated status early. Primary endpoint was the number of day (percentage) of adherence to telemonitoring at 3 months, fixed to a minimum of 18 hours per day of patch application. Secondary endpoints were hours (median per day) of patch application at 3 months.

Results: 10 pts enrolled (9,8 ± 5,8 years; 6 M; CHD with single ventricle: 5) and 7 completed the study; 1 withdrew, and 2 ended early the study for hospitalization. Adherence to telemonitoring at 3 months ranged between 3.13% and 34.5%. Dermatitis patch-related was the most encountered complication with a median time ranged between 5h15m and 17h32m. The system was adapted with introduction for commercially electrodes. In 2 pts telemedicine allowed to detect serious events requiring urgent admission.

Conclusions: Telemonitoring was effective to detect adverse events in children with severe HF and in waiting list for HTx. Current patch commercialized limited the use in children because of development of dermatitis, differently from adult population. Patch designed for children skin is necessary to improve compliance.

CB39

INCIDENCE AND PREDICTORS OF PERICARDIAL EFFUSION FOLLOWING SURGICAL CLOSURE OF ATRIAL SEPTAL DEFECT IN CHILDREN: A SINGLE CENTER EXPERIENCE

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Objectives: To evaluate the incidence of pericardial effusion (PE) after surgical atrial septal defect (ASD) closure and to investigate the presence of predictive risk factors for its development.

Methods: We collected data of 203 patients followed at Bambino Gesù Children's Hospital of Rome who underwent cardiac surgery for ASD correction between January 2015 and September 2019.

Results: A total of 200/203 patients with different types of ASD were included. Patients were divided in two groups: group 1, 38 (19%) who developed pericardial effusion (PE) and group 2, 162 (81%) without PE. No differences were noted between the two groups with regards to gender or age at the surgery. Fever in the 48 hours after surgery was significantly more frequent in group 1 than in group 2 (23.7% vs 2.5%; $p<0.0001$). ECG at discharge showed significantly ST segment elevation in children who developed PE, 24.3% vs 2.0% in those who did not ($p<0.0001$). Group 1 patients were divided in two subgroups on the basis of the severity of PE: 31 (81.6%) with mild and 7 (18.4%) with moderate/severe PE. Patients with moderate/severe PE had a significative higher BMI value (median 19.1 Kg/m2) (range 15.9-23.4, $p=0.004$)

Conclusions: Presence of fever and ST segment elevation after surgery predict subsequent development of PE suggesting a closer follow-up for this categories of patients. A higher BMI appears to be associated to a higher risk of moderate/severe PE.

CB40

INITIAL EXPERIENCE WITH MICRO MULTIPLANE TRANSOESOPHAGEAL PROBE IN INFANTS DURING CARDIAC SURGERY

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Background and aim: The use of intraoperative transoesophageal echocardiography (TEE) has been shown to be of significant benefit to patients undergoing surgery for congenital heart disease.
We assessed the utility and safety of a micro multiplane transoesophageal probe in small infants undergoing cardiac surgery.

Materials and Methods: 9 infants undergoing cardiac surgery at our Institution were included in the study. Intraoperative echocardiography with Philips s8-3t microTEE probe done using Epiq platform was utilized to study preoperative anatomy and assess postoperative results. The transoesophageal echocardiogram was used to evaluate the appearance of endocardial borders, atrioventricular and semilunar valves, systemic and pulmonary venous inflow as well as atrial and ventricular septum using standard views and positions. This included high oesophageal, mid-oesophageal, and transgastric views. Pulse wave, continuos wave and colour flow Doppler were applied in these projections.

Results: 9 patients aged 4 days- 1,2 years (mean 5.8 months) and weighing 2,5-8.3 kg (mean 5.9 kg) underwent perioperative evaluation using microTEE probe. There were two transposition of the great arteries, one congenitally corrected transposition of the great arteries, two atrioventricular septal defects, two double outlet right ventricle and two ventricular septal defect. The micro transoesophageal echocardiogram probe was passed without difficulty and complications in all patients. Good quality 2D and Colour Doppler images were obtained in all patients.

Conclusions: Given the increase in the number of neonatal surgical procedures for congenital heart disease and guided by the need for a transoesophageal echocardiographic probe small enough to be used in infants weighing > 2,5 kg, a miniaturized multiplane micro-TEE probe was introduced some years ago. The micro-TEE provides high quality, useful diagnostic images reducing the risk for complications such as airway compromise, oesophageal or gastric trauma and hemodynamic alterations. It is widely accepted that intraoperative transoesophageal echocardiogram in paediatric patients offers clear clinical benefits for the identification of residual lesions and reduced re-operation rates.

CB41

ANATOMICAL CORRECTION OF TAUSSIG-BING ANOMALY WITH AND WITHOUT AORTIC ARCH PATHOLOGY, EXPERIENCE AND RESULTS OF THE ARTERIAL SWITCH OPERATION

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Background: Taussig-Bing anomaly (TBA) is a rare congenital cardiac anomaly and frequently associated with aortic arch pathology (more than 50% of the patients) such as interrupted aortic arch (IAA), coarctation and arch hypoplasia. Also, various types of coronary anomalies are frequently seen. Different surgical techniques and strategies (neonatal single stage correction with ASO, staged repair, three patch technique) are described to deal with this complex anomaly. Although, technically demanding in this setting, the arterial switch operation (ASO) associated with arch surgery is the most utilised approach for TBA correction. The aim of the present report is to describe our results of TBA correction with ASO.

Methods: Between December 2007 to November 2021, 10 consecutive patients with TBA underwent surgical correction at our Institution. Eight (80%) underwent single stage neonatal anatomical correction with ASO, one had staged repair with anatomical correction with ASO performed at 4 months of age due to complex single coronary anatomy and the other one had intraventricular repair (Kawashima operation).

Results: There were 5 male and 5 female patients, with a median age of 20 days (range 7 dys to 12 Yrs) and median body weight of 3.5 kg (range 2.8-35kg). Median cardiopulmonary bypass time was 244 min (136-405 min) and the median time of aortic clamping was 141 min (60-182). Aortic arch anomalies that required concomitant surgery was present in the 80% (8/10) of Patients and in 7 of them aortic arch was repaired using cerebro-myocardial perfusion on the beating heart in order to reduce clamping time. Lecompte manoeuvre was performed in 8 patients of 9 who underwent ASO. Concomitant right ventricular outflow tact patch enlargement was necessary in 6 (6/9). In the series only one patient experienced complete heart block that required permanent pace-make implant, while none required veno-arterial ECMO. There were no early deaths, all patients were discharged and entered follow-up which was available for all and complete. There was only one late death (10%) due to pulmonary hypertension in a foreign patent presenting late at 3 months of age who underwent single stage repair with ASO. The patient survived first operation and presented at follow-up after 2 years with severe pulmonary hypertension, severe mitral insufficiency and moderate RVOT stenosis. Median follow-up was 5.8 years (range, 6 months - 14,5 years). Overall survival was 100% at 1 year, 90% at 2, 5, 10, 14 years. All patients were in New York Heart Association class I at last follow-up. Freedom from reintervention was 96% (95% confidence interval) and 65% (95% confidence interval) at 1 and 5 years, respectively, in the hole cohort and after ASO. The most common reintervention was RVOT patch enlargement due to RVOT stenosis which occurred in 3 patients at a median of 12 months after ASO.

Conclusions: Patients with Taussig-Bing anomaly and aortic arch obstruction can undergo neonatal single stage ASO with excellent survival and very low morbidity. In complex coronary anomalies staged repair may represent a safe strategy to manage TBA.

CB42

THE ACHD PATIENT IN CATH LAB. IMPACT AND BENEFITS OF A DEDICATED INTERVENTIONAL TEAMWORK

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Background: The number of cardiac catheterization procedures devoted to adult patients with congenital heart disease (ACHD) is becoming a growing issue in the catheterization laboratories of adult and pediatric cardiac institutions. These complex patients feature a wide array of cardiac conditions and comorbidities and deserve a tailored care, especially if invasive cardiac procedures need to be carried out.

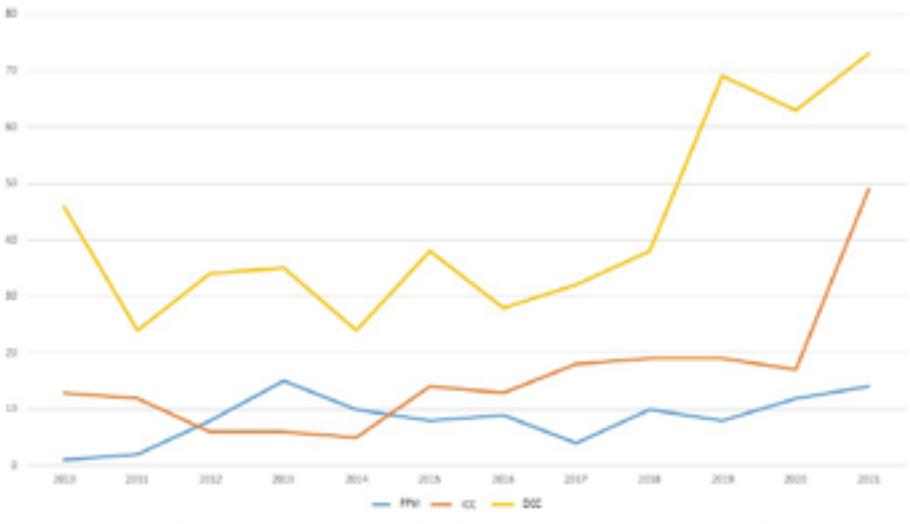
Aim: We sought to evaluate the impact of a dedicated ACHD unit operating since 2017 by a retrospective analysis of the cardiac catheterization data of all ACHD patients treated since 2010 in our institution.

Methods: Moderate or complex ACHD (according to the 2020 ESC Classification) followed at our ACHD unit and who had at least one diagnostic (DCC) or interventional cardiac catheterization (ICC) procedure during the time frame 2010-2021 were selected for the study. This dataset was further divided into three main subsets: 1) diagnostic procedures, 2) interventional procedures, 3) percutaneous valvular replacement.

Results: During the study time, 1443 patients (mean age 27.9 ± 3.5 yrs) fulfilled the study entry criteria with a total of 796 invasive procedures carried out. After 2017 there has been a steady increase in the number of diagnostic and overall interventional procedures, while the pulmonary implantation procedures (PPVI) remained quite stable throughout the entire time frame of the study (Figure 1)
The rise of DCC since 2017 has been mainly due to the start of a dedicated Fontan clinic program which encompasses a scheduled timeline of invasive assessment of these patients, according to the current AHA/ACC and ESC guidelines. The main reason for the number of ICC was also due to an increasing number of Fontan patients needing conduit stenting and/or collateral vessel occlusion. Furthermore, in 2020 there was a sharp pandemic-related reduction of the procedures, largely recovered in 2021.

Conclusions: ACHD patients are a growing population who deserve unique clinical and invasive care. Fontan patients need a specific cardiac catheterization pathway along with non invasive functional testing to avoid premature cardiac failure and overwhelming comorbidities. Our experience, as reported in previous studies, confirms that the institution of a Unit for ACHD allows to provide optimal care for these complex patients.

Figure 1



CB43

IMPELLA DEVICE FOR MECHANICAL SUPPORT OF THE SYSTEMIC CIRCULATION IN A PEDIATRIC CENTER

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Introduction: Management of cardiogenic shock in pediatric population is always a challenge. At present, the most common used system for mechanical support in children is the extracorporeal membrane oxygenation (ECMO), which usually require surgical implantation. Impella devices are now approved for short-term support (4–6 days depending on the device) for treatment of refractory or ongoing cardiogenic shock after AMI or following open-heart surgery. Case series have been published about Impella use in pediatric cardiogenic shock both for circulatory support and for cardiac unloading during ECMO assistance. Aim of this study is to describe our experience about the use of Impella devices in a population of adolescents and young adults mainly for cardiac unloading in association to ECMO assistance.

Materials and Methods: This is a retrospective review of Impella device percutaneously implanted at our center from 2019 to 2022.

Results: In the last 3 years a total of 7 percutaneous Impella implants were performed. Median age and weight were 17 years (14-22 years) and 50 Kg (range 40-70 Kg). In 5 pts Impella was implanted in association to ECMO assistance in order to achieve a complete cardiac unloading, in 2 pts Impella was used alone for cardiac support. Indications to assistance were: myocarditis in 4 patients, heart failure in dilated cardiomyopathy in 2 and cardiogenic shock in congenital heart disease in 1. The last patient had a history of congenitally corrected transposition of great arteries and is one of the few cases described of Impella support in the right systemic ventricle.
In 6 patients arterial femoral access was used (5 pts a 14 Fr introducer, 1 pt a 12 Fr introducer). In one patient the Impella device was implanted through a carotid access, obtained by surgical cutdown. In 6 patients a CP Impella (3.5) was used, only in one patient Impella 2.5 was implanted. During percutaneous insertion, no acute complications were encountered. Mean duration of support was 8 days (range 1 to 17 days). Indications for Impella explant were: LV function recovery in 4, LVAD implant in 2 and death in one. Impella removal was performed by manual compression of the arterial femoral access in all except one in which Impella was surgically removed with surgical reconstruction of the femoral artery.
During Impella support only one major complication was registered: one patient presented bleeding from the femoral access, treated by a surgical revision of the access site.

Conclusions: Percutaneous implantation of the Impella system is feasible for a long period in a pediatric/young adult population. The association of Impella with ECMO for cardiac unloading allows the use of the smallest models of Impella, reducing vascular access complications. Short-term findings demonstrated an acceptable safety profile of the device for temporary circulatory support in adolescent patients presenting with refractory cardiogenic shock.

CB44

DOES BIVENTRICULAR REPAIR INFLUENCE THE AORTIC HEIGHT INDEX IN PATIENTS WITH TETRALOGY OF FALLOT?

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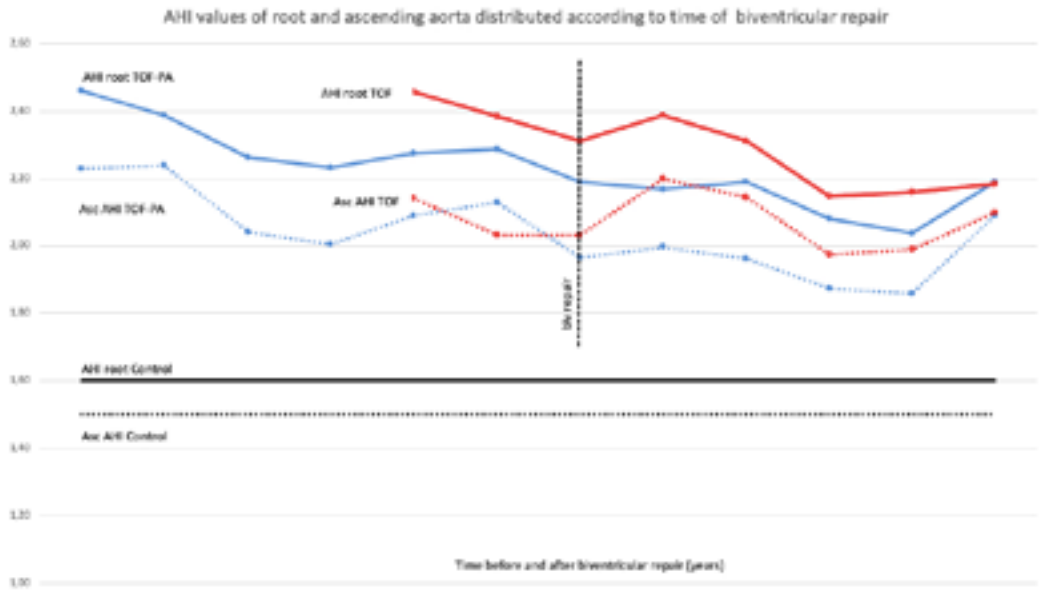
Background and aim: Patients with Tetralogy of Fallot (TOF) are assumed to have an enlarged aorta due to increased blood flow in the systemic outflow and to intrinsic wall tissue abnormalities. Whether biventricular repair affects the aortic size has been poorly investigated and little is known about the expected aortic height index (AHI) in TOF patients. We report the AHI and the impact of biventricular repair on a cohort of TOF patients.

Patients and Methods: We retrospectively reviewed preoperative and postoperative echocardiography (one cardiologist, two blind measures) of 88 patients, with varying degrees of TOF, who underwent biventricular repair between January 2014 and December 2019. Patients with incomplete data were excluded. 53 patients (35/18 M/F; TOF-PA 26 pts; TOF 27 pts) were enrolled and the aortic size (root and ascending aorta) and biometric parameters were collected. We evaluated pre and post-operative AHI and the effect of biventricular repair on it. A normal AHI value was calculated on a control group.

Results: AHI data of the root and ascending aorta were distributed based on the time of operation and compared between TOF-PA and TOF patients. At biventricular correction the mean age was different (5,3 ± 4,2 and 0,6 ± 0,3 yrs in TOF-PA and TOF patients respectively; $p<0.01$) however the AHI values did not differ significantly (Root: 2,2 ± 0,3 and 2,3 ± 0,3; ascending aorta: 2,0 ± 0,3 and 2,0 ± 0,2 in TOF-PA and TOF respectively). In the control group the mean AHI for root was 1,6 ± 0,1 and for ascending aorta was 1,5 ± 0,1. The figure evidences the AHI changes of the root and ascending aorta of TOF-PA and TOF patients before and after the operation. Despite a downward trend of AHI values, they did not differ significantly over the observed period. However, there was a significant difference with the control group ($p<0.01$).

Conclusions: Biventricular repair does not appear to affect aortic size in TOF patients. Despite the observed downward trend of AHI after surgery, the AHI in TOF-PA shows that reduction can occur regardless of surgical repair. In TOF patients with antegrade flow this possible natural trend may be masked by the younger age to biventricular repair.



CB45

TRANSCATHETER CLOSURE OF "SURGICAL" OSTIUM SECUNDUM ATRIAL SEPTAL DEFECTS WITH GORE® CARDIOFORM ASD OCCLUDER

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Objective: To evaluate the GORE® Cardioform ASD Occluder (GCO) (WL Gore & Associates, Flagstaff, AZ, USA) device for closure of ostium secundum atrial septal defects (ASD) with predicted indication for surgical correction.

Background: Closure of large ASD in small children by transcatheter approach is still challenging. This study evaluated the results of GCO in this subset of patients in a tertiary referral center.

Methods: Between January 2020 and March 2022, 97 children underwent transcatheter ASD closure at our Institution. Of them, 38 had a large defect (diameter/weight >1.2 or diameter/BSA >20 mm/m2), predicted suitable for surgery and underwent closure with GCO. Procedure results and mid-term outcome are reported.

Results: Patients' age and weight were 5.5+1.5 years and 19.7+4.7 kg, respectively. Absolute and relative ASD size was 21.5+3.6 mm, 1.1+0.2 mm/kg and 27.7+4.6 mm/m2 respectively, resulting in QP/QS of 2.0+0.8. Three patients were sent to surgery after balloon sizing. Four of the remaining 35 patients who underwent device deployment, needed rescue or elective surgery due to device embolization (n=1), device instability (n=2) or new-onset tricuspid valve regurgitation (n=1). Procedure feasibility was 88.6%. Major complications were recorded in 2 patients (5.7%). Minor complications were recorded in 5 patients (14.3%). Complete closure at discharge was 90.3% (28/31 pts) rising to 100% at the last follow-up evaluation. Wireframe fractures rate at the 6 months examination was 52%, without clinical and instrumental consequences.

Conclusions: Percutaneous treatment with GCO device is effective and safe in a high percentage of ASD children with predicted indication for surgical correction.

CB46

BALLOON ANGIOPLASTY FOR NATIVE AORTIC COARCTATION IN NEONATES AND INFANTS WITH HIGH SURGICAL RISK

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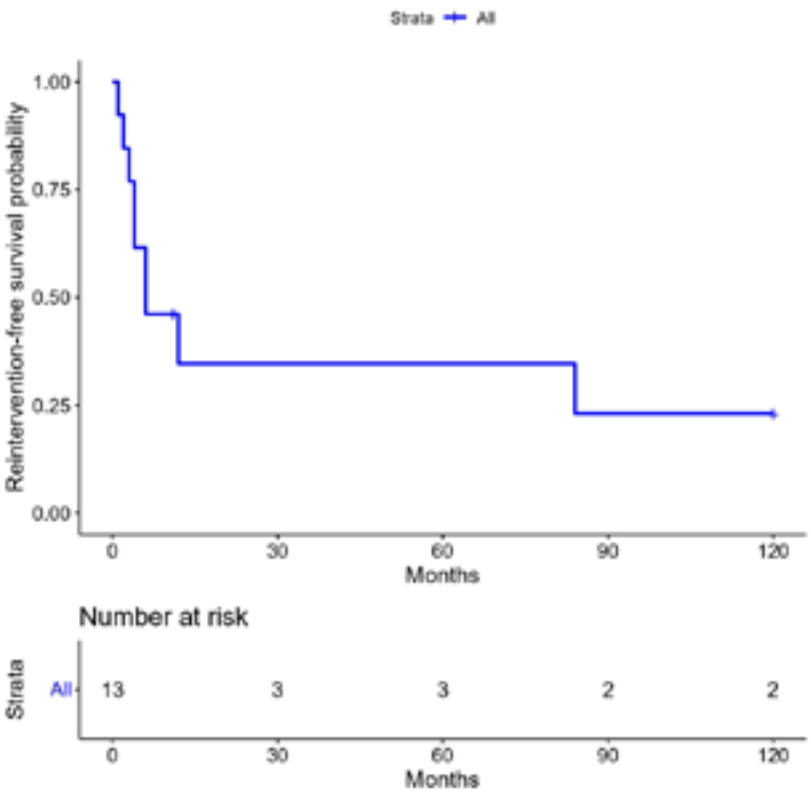
Background: Surgical repair for native aortic coarctation (CoA) is the current standard of treatment in the neonatal period and infancy. In selected cases balloon angioplasty may be performed as a palliative strategy to stabilize patients with high surgical risk. The aim of the study was to explore the efficacy and safety of balloon angioplasty in neonates and infants with native CoA and high surgical risk at clinical presentation.

Methods: This is an observational retrospective study. All patients of age <12 months who underwent percutaneous balloon angioplasty for native CoA between January 2010 and May 2022 were identified form the institutional database and included in the study. Medical records were reviewed regarding patient profiles, concomitant congenital heart defects, surgical and percutaneous procedures. R statistical software 4.2.0 was used for analysis. Since numeric variables were not normally distributed, non-parametric tests were performed.

Results: Thirteen patients were included in the study. The median procedural age was 1 month (IQR 0.5 – 4 months) and the median weight was 3.2 kg (IQR 2.8 – 5.5 kg). High surgical risk at presentation was determined by acute decompensated heart failure (53.8%), low-body weight <2.5 kg (31%), concomitant major extracardiac anomalies (eg. oesophageal atresia, 15.4%) or a combination of them. Mini-Tyshak balloon (NuMED, Inc., Hopkinton, NY, USA) was the only type of balloon catheter employed. A single balloon diameter was used in 69.3% of patients, 2 different balloon diameters in 30.7%. Balloon angioplasty produced a large decrease in the invasive peak systolic pressure gradient (median ± IQR, 50±15 mmHg vs 5 ±3.5 mmHg). The most common vascular access was the right axillary artery (53.8%). Femoral artery access was used was used in 38.4% of cases. In one case a right carotid artery access with surgical cut-down was used. No major and minor procedural complication were reported. A second catheter intervention for recurrent CoA was required in 3 cases (23.1%) at a median age of 12 months (IQR 8-48 months). Surgical reintervention was performed in 6 of 13 (46.1%) at a media age of 3.5 months (IQR 2.25-5-5 months) after initial balloon angioplasty. The hazard of reintervention was highest early. The median freedom from any reintervention was 79.6% (95% CI, 57.1% -100%) at 3 months, 34.6% (95% CI, 15.3% - 78.2%) at 1 year, 23.1% (7.3% - 72.3%) at 5 and 10 years. Freedom from surgery was 51.9% (95% CI, 30.3-89.1%) at 5 years and remained unchanged at 10 years. In the low-body weight group, we observed a trend of higher frequency of recurrent coarctation (100% vs 55.6%, p=ns) and earlier presentation (5 months vs 11 months, p=ns). One patient developed a small aortic aneurysm at balloon dilatation site and recurrent CoA and therefore he needed surgery.



Discussion and Conclusion: Balloon angioplasty for native CoA was effective and safe in a cohort of neonate and infants with immediate high surgical risk. Recurrent coarctation was higher in patients with low-body weight. Repeat angioplasty is effective in recurrent CoA and surgery may be avoided in a non-negligible proportion of patients.

CB47

ECMELLA TREATMENT IN PATIENTS WITH CARDIOGENIC SHOCK AND ACUTE HEART FAILURE: IS IT FEASIBLE IN PEDIATRIC AGE?

A. Ionata [1], R. Adorisio [2], E. Bellettini [2], G. Ingrasciotta [2], E. Mencarelli [2], S. Alfieri [2], D. De Angelis [2], G. Grutter [2], M. Rebonato [2], R. Formigari [2], G. Butera [2], L. Galletti [2], A. Amodeo [2]

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Background: MCS is always advocated to improve hemodynamic status and favour ventricular recovery in children with acute myocarditis. Recently, the combination of short-term mechanical circulatory support by left-sided Impella (LV-Impella) with extracorporeal life support (ECMO), called ECM-ELLA, in patients with cardiogenic shock due to different aetiology has emerged as a promising strategy in adult population.

Methods: We present a case series of 4 pts (all female; mean age: 16 years ± 1.8) presented with acute heart failure and cardiogenic shock, treated with ECMELLA strategy.

Results: 4 patients were admitted to Bambino Gesù Paediatric hospital with severe acute heart failure and reduced ejection fraction (mean LVEF: 18.5% ± 2.4). The recognized aetiology were: 3 acute myocarditis, 1 due to a metabolic disorder after non cardiac surgery. All patients underwent to ECMELLA strategy in acute phase, with a mean time of ECMO placement from admission of 26 hours (from 2 to 72 hours), and a mean time of LV-Impella placement of 53 hours (from 6 to 120 hours). This MCS strategy was used for 12 ± 8.1 days on average. ECMO was discontinued after 7 ± 4,4 days, while LV-Impella was removed after 11 ± 8.3 days. Complications encountered were: retroperitoneal haematoma during the LV-Impella implant procedure; one bleeding from the cannula site after ECMO placement. No sequelae were reported. After 1 month, 3 patients fully recovered (mean LVEF: 56.6 ± 10.4%; mean days at recovery: 25 ± 4,7) and 1 was successfully bridged to durable LVAD implant (after 20 days from admission, 11 days after ECMO removal and soon after LV-Impella removal).

Conclusions: ECMELLA strategy is effective in children to bridge to myocardial recovery since it provides proper circulatory support while reducing wall stress and, therefore, adverse cardiac remodelling.
Its prompt use should be considered also in paediatric patients with resistant cardiogenic shock, in order to set the proper strategy according to the aetiology of heart failure.

CB48

CHIUSURA PERCUTANEA DI DIFETTI INTERASTRALI MULTIFENESTRATI IN ETÀ PEDIATRICA

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Introduzione: I difetti interatriali (DIA) sono una delle più comuni patologie riscontrabili all'interno delle cardiopatie congenite. La chiusura percutanea di tali difetti risulta oggigiorno la tecnica di scelta, in quanto ha dimostrato di avere buoni risultati in termini di efficacia ed un numero di complicanze inferiori se rapportata alla chiusura chirurgica.

Scopo dello Studio: Valutare l'efficacia, in termini di shunt residui, e la sicurezza, in termini di complicanze procedurali e post procedurali, della chiusura di DIA multifenestrati, con singolo device e con doppio device.

Metodi: Nel centro di cardiologia pediatrica di Padova dal 1/01/2020 al 31/05/2022 sono state eseguite 110 procedure di chiusura di difetti interatriali per via percutanea. La valutazione pre-operatoria dei pazienti è stata basata su ecocardiografia trans-toracica e trans-esofagea 2D e 3D. In fase intra-operatoria è stata eseguita valutazione basata sia su ecocardiografia trans-esofagea 2D e 3D. La valutazione post-operatoria, basata su ecocardiografia trans-toracica 2D, ha preso in considerazione la presenza di shunt residui valutati a 24 ore e dopo 6 mesi dal posizionamento dei device. I device usati, singoli o multipli, scelti in base alle caratteristiche anatomiche e del paziente sono stati: GORE® Cardioform ASD Occluder, GORE® Cardioform Septal Occluder e Amplatzer septal occluder.

Risultati: La procedura è stata efficace in tutti i 110 pazienti arruolati. È stato utilizzato un device AGA in 17 casi, Occlutech in 24 casi, GSO in 33 e GCA in 36 pazienti. Il setto risultata multifenestrato in 16 casi (14,5%). Per la chiusura di tali difetti si è utilizzato un doppio device in 4 casi (GCA+GSO nel totale dei casi). Nei restanti 12 pazienti si è ottenuta una occlusione con singolo device (GCA 5 casi 41,7%; nei restanti 7 casi GSO 58,3%) mediante sovradimensionamento rispetto al sizing del difetto principale.
A 24 ore la prevalenza di shunt residuo era 18,75% nei multifenestrati vs 97% nei difetti singoli, a sei mesi non vi erano shunt residui in entrambi i gruppi.

Conclusioni: In età pediatrica i DIA multifenestrati sono relativamente frequenti. Se trattati in eta pre-adolescenziale, le dimensioni assolute del setto totale permettono la chiusura completa dei difetti con un singolo dispositivo in circa il 75% dei casi. Quando ciò non sia fattibile, l'utilizzo di un secondo dispositivo di piccole dimensioni è comunque una opzione sicura ed efficace a correggere tale difetto.

EP01

SCIMITARRA A CLESSIDRA

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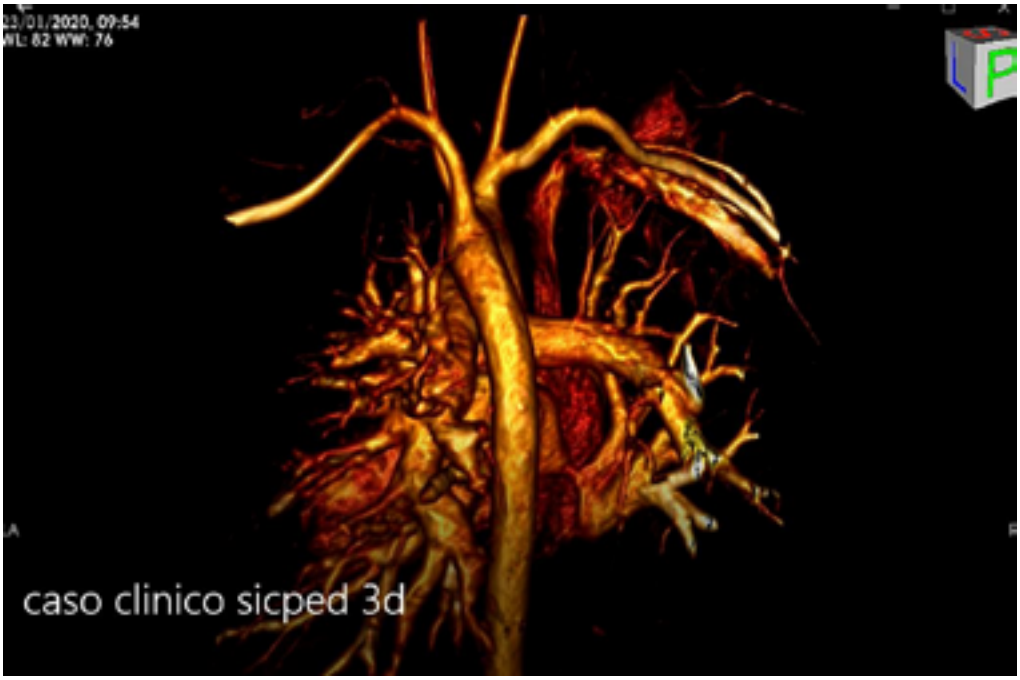
La sindrome della scimitarra rappresenta una complessa sindrome congenita nella quale è presente, fra l'altro, un ritorno venoso anomalo delle vene polmonari di destra in un collettore venoso. L'entità dello shunt sinistro-destro secondario influenza la precocità della presentazione clinica e il timing diagnostico- correttivo.
Di seguito viene riportato il caso di un giovane, ora di 21 anni, seguito da diversi anni presso l'ambulatorio delle cardiopatie congenite per follow up di pregressa correzione cardiochirurgica (2003) di DIA tipo ritorno venoso anomalo parziale in sindrome della scimitarra. L'intervento cardiochirurgico di cui è disponibile l'atto operatorio, è stato eseguito effettuando un'anastomosi longitudinale fra il collettore venoso e l'atrio sinistro con abbocco diretto delle due strutture anatomiche. E'stato quindi ricostruito il setto interatriale con l'utilizzo di patch in pericardio autologo.Il decorso post operatorio è stato regolare .Durante gli anni successivi all'intervento il bambino ha eseguito controlli cardiologici regolari che hanno rilevato ottima crescita , stato di benessere generale , assenza di aritmie e una buona capacità funzionale .Nel corso degli anni non è stata evidenziata suscettibilità ad infezioni respiratorie. Negli ultimi anni il ragazzo ha effettuato diversi test al cicloergometro che non hanno rilevato aritmie da sforzo né anomalie ecg anche ad alto carico di lavoro; nella norma la saturazione a riposo. Nel 2020 data la scadente finestra ecocardiografica è stato richiesto controllo con cardio RMN (2021).
Alla cardio RMN si è documentato un ventricolo destro moderatamente dilatato con funzione sistolica globale conservata con evidenza di shunt sinistro-destro significativo (con Qp:Qs stimato pari a 1.9). Dalla ricostruzione bi e tridimensionale si è apprezzata la sostanziale ricanalizzazione del dotto collettore venoso a livello della giunzione atrio-cavale inferiore in sede sovra-diaframmatica; si è evidenziata la stenosi della pregressa anastomosi vascolare tra il dotto collettore e l'atrio sinistro, anastomosi caratterizzata da una morfologia a clessidra per focale marcata riduzione del calibro (calibro minimo di circa 6-7 mm); il setto interatriale appare integro.
L'angio Tc eseguita successivamente ha confermato i dati anatomici rilevati alla cardio RMN, in particolare la pervietà con morfologia a clessidra della connessione chirurgica tra il dotto collettore e l'atrio sinistro; la presenza di questa pur ridotta connessione anatomica, potrebbe consentire un re-intervento con approccio ibrido di correzione della cardiopatia .



EP02

CARDIOVASCULAR RISK FACTORS AND GENETIC PREDISPOSITION: AN UNUSUAL CASE OF AORTIC REGURGITATION AND LVOT OBSTRUCTION

A. Balducci [1], M. Molisana [1], D. Palleri [1], C. Ciuca [1], Y. Bartolacelli [1], L. Careddu [2], E. Angeli [2], A. Donti [1], G.D. Gargiulo [2]

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A 38-year-old obese man presented to our outpatients' clinic for progressive dyspnea and chest pain during exertion. The patient had a history of percutaneous patent foramen ovale closure, uncontrolled hypertension and congenital blindness secondary to bilateral cataract. At the clinical evaluation a new ejective systolic murmur, best heard on the aortic area, was revealed. He presented neither crackles at lung auscultation, nor hepatomegaly or extremity edema. His blood pressure was 170/75 mmHg, the other vital signs were normal.

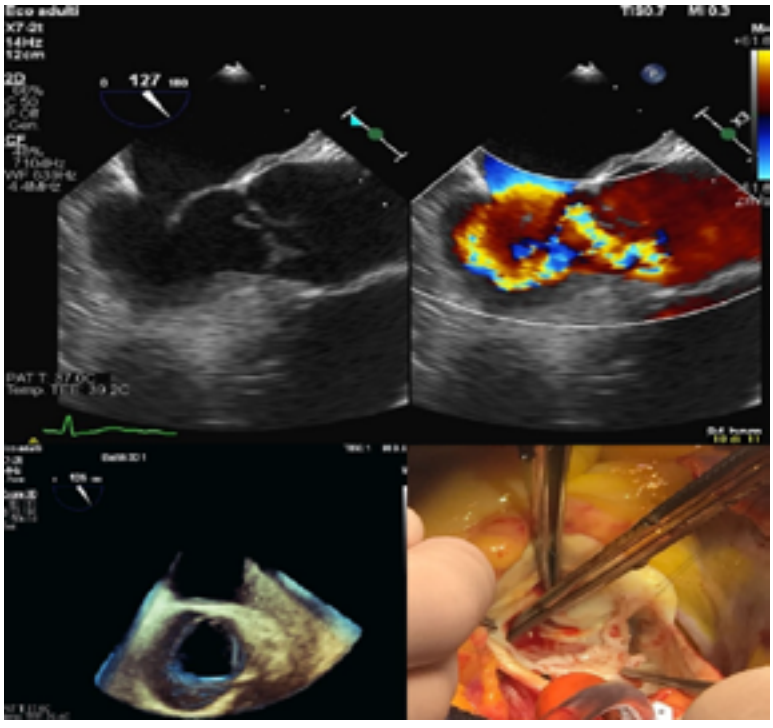
Electrocardiogram showed a first-degree atrio-ventricular block with signs of left ventricular hypertrophy. Transthoracic echocardiography showed: a slightly reduced left ventricular ejection fraction, a severe left ventricle dilatation and a mild left atrial dilatation, a severe aortic regurgitation and a mobile ill-defined subaortic structure prolapsing into the left ventricular outflow tract (LVOT) causing a severe obstruction. To better characterize this finding, a transesophageal echocardiography was performed. An abnormal movement of the aortic right coronary cusp, losing the coaptation point and everting totally in the LVOT was revealed and confirmed on 3D reconstruction. This finding was interpreted as right coronary cusp flail, which was consistent with the severe aortic regurgitation but not with the LVOT gradient.

Patient symptoms together with the echocardiographic findings made the surgical solution recommended. Intraoperative inspection of the aortic valve showed a tricuspid aortic valve with fibromuscular metaplasia of the cusps' structure and the delamination of the right coronary cusp, splitting in two layers: one layer adherent to the valve annulus and the other protruding into the LVOT. The native aortic valve was then removed and a mechanical prostheses was implanted. The patient had a regular peri-operative course without complications. In humans and animals models, valve cellular and extra cellular integrity is actively maintained in a homeostatic balance by paracrine interactions between valve interstitial cells (VIC) and valve endothelial cells (VEC), which inhibit endothelial-to-mesenchymal transition. The exposure to different types of stress entails a valve change in dimension, shape and stiffness secondary to the interaction between cells and extracellular matrix. Several studies demonstrated the role of TGF? overexpression in the development of lens opacity in patients with congenital cataract. In VIC and VEC environment, TGF? can activate mesenchymal signaling programs and in canine valve cultures the combination of TGF?1 overexposure and cyclic strain induced myofibroblastic differentiation in VIC.

Uncontrolled hypertension is a cause of excessive shear stress and together with an anomalous autocrine and paracrine signalling pathway, involving TGF? cascade, may result in valve metaplasia and disruption.

These mechanisms could explain the peculiar finding of a cusp delamination causing severe valve regurgitation and the presence of valve muscular metaplasia projecting through the LVOT.

Valve tissue remodeling is a complex process, where genes and environment act together to produce the valve phenotype. The genetic predisposition in an adult with a congenital eye disease in association with the mechanical stress of arterial hypertension led to a peculiar fibromuscular degeneration of the aortic valve needing surgical replacement. Echocardiography played an important role for working diagnosis but surgical evaluation was essential to characterize this unusual condition.



EP03

FONTAN-ASSOCIATED LIVER AND RENAL DISEASE: A PROSPECTIVE SINGLE CENTER STUDY TO EARLY DETECT LONG TERM COMPLICATIONS

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Background: Fontan operation represents the surgical palliative option for congenital heart disease with single ventricle physiology. With the improvement of surgical techniques, we are facing a growing population with multiorgan complications: in particular, hepatic and renal dysfunction remain mostly unknown.

Materials and Methods: Patients that underwent Fontan palliation in our Centre between 1985 and 2016 were included in this prospective study. We excluded patients with major congenital renal anomalies, those that underwent cardiac transplantation and redo-Fontan patients. All the subjects underwent clinical evaluation, laboratory exams with complete renal and hepatic function, transient hepatic elastography and complete cardiac evaluation. We aimed at 1) evaluate the cardiovascular, renal and hepatic dysfunction and 2) show the relationship between renal and hepatic results and cardiovascular status and the effect of time on multiorgan involvement.

Results: We enrolled 45 patients, 60% male. Medium time from Fontan completion was 13 years 6 months (167 months), SD ± 6.8 years (82 months).
Data from echocardiographic evaluation shows that patients with functionally single right ventricle have worse diastolic function ($p<0.05$); AV valve insufficiency is associated with low VO2 max at cardiopulmonary testing ($p<0.05$). 23% patients have a mildly reduced glomerular filtration rate and 3% moderately reduced. Renal evaluation shows glomerular dysfunction (associated with systolic dysfunction $p<0.05$) and chronic tubular alterations (associated with diastolic dysfunction $p<0.05$). Renal dysfunction doesn't appear to be worse in patients with a past history of dialysis. Hepatic tests show that GGT alterations are more frequent than AST elevation (66% vs 9%). Hepatic stiffness higher than reference value is present in 32% of the population, with a lower VO2 max at cardiopulmonary testing ($p<0.05$).

Conclusions: Fontan patients show hepatic and renal dysfunctions, that worsen with time and appear to be related with functional ad cardiological tests. This suggests that Fontan patients need a multidisciplinary follow-up.

EP04

PARVOVIRUS B19 MYOCARDITIS IN A COVID19 MIS-C SYNDROME: CAUSE OR CAUSALITY?

E. Bellettini, E. Mencarelli, G. Ingrasciotta, M. Rebonato, P. Francalanci, L. Galletti, A. Amodeo, R. Adorisio
IRCCS Istituto Ospedale Pediatrico Bambino Gesù, Roma, Italy

Background: Myocarditis has been recognized as one of major complication during COVID 19 infections and vaccination. Few data are currently available on viral detection during myocardial damage in pediatric population. We report a case of myocarditis PVB 19 related during MIS-C syndrome.

Case presentation: A previously healthy 13 years old boy presented with abdominal pain, vomit and fever at the Emergency Department in Bambino Gesù Hospital on January 2021. Previous infection by SARS-CoV2 was diagnosed two months before, without symptoms. He was admitted with the suspicion of appendicitis and antibiotic therapy was started. Abdominal ultrasound showed minimal fluid distension in absence of significant parietal thickening. Nasopharyngeal swabs was positive for a new SARS CoV-2 infection.
Clinical scenario evolved rapidly with frequent diarrhoeic discharges, pallor, hypotension and galloping rhythms. Laboratory tests showed a progressive rise in inflammation indices (C reactive protein, procalcitonin, ferritin and leucocytosis with lymphopenia) and cardiac enzymes: NTproBNP 9.000 ng/ml, troponin 1.500 ng/ml (NV < 14 ng/ml). At the echocardiogram a severe left ventricular dysfunction without dilation and with mild pericardial effusion were immediatly detected. Due to rapid progression to cardiogenic shock, an haemodynamic support was offered with orotracheal intubation and inotropes.
Myocardial biopsy was performed disclosing an inflammatory lymphomonocyte interstitial infiltrate (CD3++, CD4+, CD8++, TIA-1+, Granzyme B+, CD20+/- and MUM1-) with myocellular cytotoxic aggression and necrosis. Parvorus B19 genome was found on myocardial muscle while COVD 19 was not detectable. The patient was treated coupling: 1. high-dose iv immunoglobulins (2 g/kg), 2. CytoSorb ® for 3 consecutive days. 24 hours after Immunoglobulin infusion LVEF promptly recovered from 25 to 50%. After 48 hours Anakinra was added. Low molecular weight heparin at therapeutic dose and high dose iv diuretics were added. General clinical condition and vital parameters gradually normalised, permitting inotropes weaning and extubation on the seventh day. Helmet Cpap support was used until clinical improvement. Sars-CoV-2 swabs were negative after 7 days.
The patients was discharged after 20 days in NYHA class I, with normal cardiac function and normalization of cardiac biomarkers. No major arrhythmias were detected during hospitalization.
Cardiac MRI performed after 3 months showed no myocardial tissue alterations and normal biventricular function. Aldactone was administered for 6 months. After 18 months the patients is asymptomatic, cardiac echocardiogram, EKG and functional tests are normal.

Conclusion: in this case Multisystem Inflammatory Syndrome due to SARS-CoV2 infection was associated to viral myocarditis by Parvovirus B19. The huge inflammatory stimulation probably activated cardiac damage by a latent, recent Parvovirus B19 infection, leading to severe cardiac dysfunction and cardiogenic shock. The therapeutic strategy with the promptly use of immunoglobulin and CytoSorb permitted to modulate immune response with the complete recovery of cardiac muscle.

EP05

RELAPSE OF MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN DURING THE CORTICOSTEROID TAPERING, DESCRIPTION OF TWO CASES

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The MIS-C is a syndrome that affects young individuals arising with fever and multisystem organ involvement (cardiac, hematologic, gastrointestinal, renal, respiratory, dermatologic or neurological). It is caused by a massive inflammatory response to the SARS-CoV-2 infection. It shares many similarities with Kawasaki syndrome, therefore many cases of relapse after MIS-C were not expected. We present two cases of children who experienced relapse of MIS-C during the corticosteroid tapering.

A 11-years old boy suffered from MIS-C with gastrointestinal and cardiac involvement. In particular, the echocardiography showed pericardial effusion in subacute phase and dilation of both coronaries, with no signs of ventricular dysfunction. The patient was treated with intravenous immunoglobulin and methylprednisolone (3 mg/kg/die) two weeks after the start of symptoms.
About eight weeks after the diagnosis, when the patient was tapering the prednisone, he presented thoracic pain and fever. The blood exams showed alteration of C-reactive protein, ferritin, increase of D-dimer and NTproBNP. The cardiological evaluation revealed clear clinical and instrumental signs of relapsing pericarditis (typical pain, EKG, evident pathological signs in the echocardiography). It was thereby decided to prescribe Anakinra, a biological drug functioning as IL-1 receptor antagonist, with an immediate clinical improvement. The echocardiographic control exposed the progressive resolution of the pericarditis, and a minor myocardial and coronary involvement (slight septal dyskinesia, altered left ventricular function and Global Longitudinal Strain, left coronary ectasia).

A 12-years girl was diagnosed with MIS-C at another hospital and treated with intravenous immunoglobulin and prednisone (1 mg/kg), two weeks after the beginning of the symptoms. Arthritis, gastroenteric involvement and atrioventricular block of first grade (PR 280 ms) occur at the onset.
About three weeks after the first treatments, once the deltacortene was suspended, she developed fever, asthenia, diffused arthralgia. The blood exams showed a high CRP and ferritin, an increase in hepatocellular enzymes and bilirubin. The patient was treated with corticosteroid. A month later, during the tapering down of corticosteroid, she re-presented fever, arthralgia and vomit. The EKG showed an atrioventricular block (PR 250ms). Tapering down the corticosteroids, the clinical condition worsened and the blood labs came back with high inflammatory markers and an alteration of the liver profile. The patient was transferred to our Institute. The experts agreed on defining the clinical picture as a relapse of MIS-C. The echocardiography detected a slightly septal dyskinesia and a posterior pericardial effusion. The tapering down program of corticosteroid was pursued at a slower pace. During the follow up, the AV block was detected intermittently and the cardiac magnetic resonance resulted normal.

By describing these two cases, we desire to point out the possibility of relapses. During the relapses, the cardiological manifestations were more aggressive, with ventricular dysfunction not present during the first episode. One patient even required a more aggressive therapy. We suggest to monitor carefully the cardiological involvement if patients with MIS-C present suspicious symptoms during the corticosteroid tapering, even without cardiovascular signs.

EP06

LINCOLN-DANIELSON HEART: FIRST 3D RECONSTRUCTED CASE AND A REVIEW OF LITERATURE ABOUT ONE OF THE RAREST HEARTS IN PEDIATRIC CARDIOLOGY

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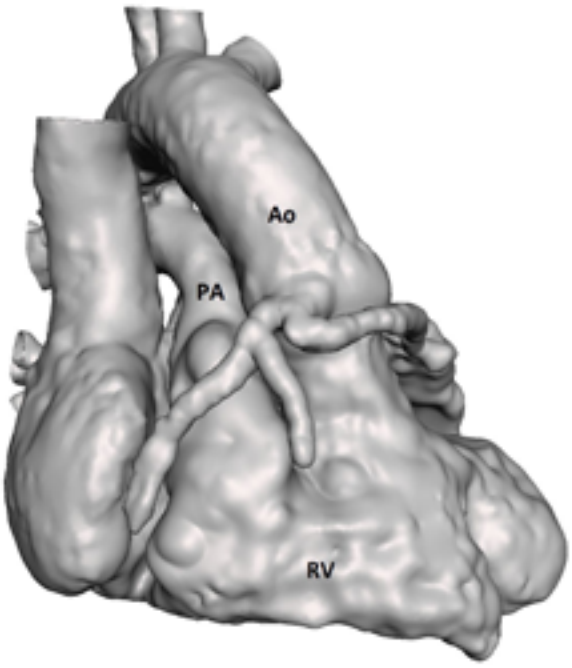
Background: The so-called Lincoln-Danielson heart is an extremely rare congenital heart disease characterized by the presence of a double outlet right ventricle with L-malposition of the great arteries. Due to its rarity, its main features and possible surgical treatments are often overlooked by textbook and relevant literature.

Methods: In our clinical practice we recently encountered an example of this congenital defect and we made a 3D model that clearly illustrates its main anatomical features. We then searched the scientific literature and reviewed all previously published cases to clarify its peculiarities and to help surgeons decide the best therapeutic approach bases on past experiences.

Results: A 3D model was created from a CT scan acquisition and 3D printed using a commercially available printer. We also rendered a short video navigation of this model and explained the internal and external morphology. We then identified a total of 50 other published cases from 1892 to 2013 and we described the differences and the similarities between them.

Conclusions: Double outlet right ventricle with L-malposition of the great arteries is a unique heart defect with an excellent prognosis in most cases, is associated with left juxtaposition of the atrial appendages and can coexist with any type of ventricular septal defect. The main predictor of surgical treatment is the type of VSD, the ventricular dimensions and the presence of pulmonary stenosis.



EP07

IPERFERRITINEMIA E VERSAMENTO PERICARDICO: UN CONNUBIO IMPERFETTO

G. Calderoni [1], G. Favia Guarnieri [1], D. Capodiferro [1], G. Ladisa [1], G. Scalzo [2], A. Filannino [1], M. Travascia [1], N. Laforgia [1]
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Outborn, nato a termine (39w) da parto eutocico con peso alla nascita di 3000 g (AGA), da genitori non consanguinei. Riferita gravidanza normodecorsa con esecuzione di test del DNA fetale risultato nella norma. In anamnesi ostetrica remota, precedente MEF di un feto di sesso femminile a 35 settimane, sine causa apparente. Giunge alla nostra osservazione in 20° giornata di vita per riscontro di scarso accrescimento ponderale, iperferritinemia, ipertransaminasemia con indici di flogosi negativi.
All'esame obiettivo all'ingresso riscontro di lievi dismorfismi facciali, aracnodattilia, capezzoli invertiti, criptorchidismo bilaterale, modesta ipotonia assiale. Fig.1
L'ecografia transfontanellare mostra asimmetria dei ventricoli laterali e dilatazione della cisterna magna con ipoplasia del verme cerebellare, confermati successivamente con RMN. L'ecografia dell'addome mostra fegato di volume aumentato con aspetto diffusamente iperecogeno e reni aumentati di volume con scarsa differenziazione cortico-midollare. L'ecocardiografia evidenzia versamento pericardico circumferenziale, di modesta entità.
Nel sospetto di emocromatosi neonatale, viene effettuata RMN total body che non risulta significativa per accumulo marziale parenchimale epatico e la biopsia delle ghiandole salivari minori che risulta nella norma.
La ricerca delle mutazioni per sindrome di Prader Willi risulta negativa e si avvia esoma clinico, con indicazione alla valutazione del gene PMM2, sul braccio corto del cromosoma 16, per il sospetto di CDG-1 (congenital disorders of glycosylation 1), di cui viene rilevata una eterozigosi composta di due varianti missenso (una di origine paterna e l'altra di origine materna).
In seguito a deterioramento clinico con difficoltà di alimentazione, diarrea cronica e aumento progressivo del versamento pericardico circumferenziale, viene avviata terapia medica con FANS, corticosteroidi e diuretici, senza risultati. In seguito a IVU da E. Coli con ulteriore aumento del versamento pericardico e iniziali segni di tamponamento (Fig. 2), a 98 giorni di vita viene effettuata pericardiocentesi con drenaggio estemporaneo di 75 cc di liquido citrino e posizionamento di drenaggio a permanenza. Il quadro clinico non si modifica e l'exitus sopraggiunge a 100 giorni di vita per coagulazione intravascolare disseminata e multiorgan failure.
I disordini congeniti della glicosilazione sono patologie genetiche a trasmissione autosomica recessiva o recessiva legata al cromosoma x, caratterizzate dalla alterazione degli enzimi coinvolti nella glicosilazione proteica e con coinvolgimento multiorgano. La forma ad esordio neonatale è associata a prognosi peggiore (exitus entro l'anno di vita) e l'interessamento cardiaco (cardiomiopatia ipertrofica o dilatativa, versamento pericardico), sebbene raro, rappresenta un ulteriore fattore prognostico negativo. Ad oggi non esistono trattamenti specifici, ma solo sintomatici.
L'ipotesi di CDG deve essere considerata in quei neonati con scarso accrescimento ponderale, alterazioni neurologiche, note dismorfiche e iperferritinemia.

EP08

IPERFERRITINEMIA E VERSAMENTO PERICARDICO: UN CONNUBIO IMPERFETTO

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Outborn, nato a termine (39w) da parto eutocico con peso alla nascita di 3000 g (AGA), da genitori non consanguinei. Riferita gravidanza normodecorsa con esecuzione di test del DNA fetale risultato nella norma. In anamnesi ostetrica remota, precedente MEF di un feto di sesso femminile a 35 settimane, sine causa apparente. Giunge alla nostra osservazione in 20° giornata di vita per riscontro di scarso accrescimento ponderale, iperferritinemia, ipertransaminasemia con indici di flogosi negativi.
All'esame obiettivo all'ingresso riscontro di lievi dismorfismi facciali, aracnodattilia, capezzoli invertiti, criptorchidismo bilaterale, modesta ipotonia assiale. Fig.1
L'ecografia transfontanellare mostra asimmetria dei ventricoli laterali e dilatazione della cisterna magna con ipoplasia del verme cerebellare, confermati successivamente con RMN. L'ecografia dell'addome mostra fegato di volume aumentato con aspetto diffusamente iperecogeno e reni aumentati di volume con scarsa differenziazione cortico-midollare. L'ecocardiografia evidenzia versamento pericardico circumferenziale, di modesta entità.
Nel sospetto di emocromatosi neonatale, viene effettuata RMN total body che non risulta significativa per accumulo marziale parenchimale epatico e la biopsia delle ghiandole salivari minori che risulta nella norma.
La ricerca delle mutazioni per sindrome di Prader Willi risulta negativa e si avvia esoma clinico, con indicazione alla valutazione del gene PMM2, sul braccio corto del cromosoma 16, per il sospetto di CDG-1 (congenital disorders of glycosylation 1), di cui viene rilevata una eterozigosi composta di due varianti missenso (una di origine paterna e l'altra di origine materna).
In seguito a deterioramento clinico con difficoltà di alimentazione, diarrea cronica e aumento progressivo del versamento pericardico circumferenziale, viene avviata terapia medica con FANS, corticosteroidi e diuretici, senza risultati. In seguito a IVU da E. Coli con ulteriore aumento del versamento pericardico e iniziali segni di tamponamento (Fig. 2), a 98 giorni di vita viene effettuata pericardiocentesi con drenaggio estemporaneo di 75 cc di liquido citrino e posizionamento di drenaggio a permanenza. Il quadro clinico non si modifica e l'exitus sopraggiunge a 100 giorni di vita per coagulazione intravascolare disseminata e multiorgan failure.
I disordini congeniti della glicosilazione sono patologie genetiche a trasmissione autosomica recessiva o recessiva legata al cromosoma x, caratterizzate dalla alterazione degli enzimi coinvolti nella glicosilazione proteica e con coinvolgimento multiorgano. La forma ad esordio neonatale è associata a prognosi peggiore (exitus entro l'anno di vita) e l'interessamento cardiaco (cardiomiopatia ipertrofica o dilatativa, versamento pericardico), sebbene raro, rappresenta un ulteriore fattore prognostico negativo. Ad oggi non esistono trattamenti specifici, ma solo sintomatici.
L'ipotesi di CDG deve essere considerata in quei neonati con scarso accrescimento ponderale, alterazioni neurologiche, note dismorfiche e iperferritinemia.

EP09

LONG-TERM CARDIOVASCULAR OUTCOME OF CHILDREN WITH MIS-C: ITALIAN MULTICENTER EXPERIENCE

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Introduction: MIS-C is a multisystem inflammatory syndrome characterized by severe systemic hyper-inflammation with multi-organ failure and cardiac involvement. The aim of this study is to describe long term cardiovascular outcome in a cohort of pediatric patient with MIS-C, admitted to two Italian Pediatric Referral Centres.

Methods: All paediatric patients affected by MIS-C, admitted to Bambino Gesù Children Hospital and Giannina Gaslini Institute COVID Centers from March 2020 to March 2022 were evaluated. Patients with MIS-C and cardiac involvement were daily revaluated with ECG and echocardiogram during acute event. Troponin were repeated every 6 hours; NT-proBNP once a day. Holter monitoring was performed in patients with cardiac conduction disturbances or arrhythmias. After discharge, patients were regularly evaluated at 1 month, 3 months, 6 months and 1 year with ECG, echocardiogram and cardiac biomarkers. In patients older than 8 years cardiac magnetic resonance (CMR) was performed.

Results: Between March 2020 and March 2022, 45 children (mean age 11 ±4,1 years, male 55%) with MIS-C diagnosis were included, 43 of whom showed cardiac involvement.
Among them, 58% had ventricular dysfunction, 52% pericarditis, 26% coronary involvement, and 13% showed arrhythmias. All patients reached one month of FU, 80% reached 3 months of FU, 70% 6 months and 65% had 1 year of FU.
At the onset, ECG abnormalities were present in 89% of patients, and progressively decreased to 20% at discharge, 3% at 1 month of FU; whilst no ECG anomalies were detected at FU. Arrhythmias were present in 13% of patients during acute events and disappeared at discharge and during FU. Pericarditis were detected in 52% of MISC and resolved in all patients but 4% at discharge, at 1 and 3 months of FU, no more pericarditis were evident after 6 months from diagnosis. Coronaritis were observed in 26% of children during acute phase, only 8% had coronary involvement at discharge and at 1 month of FU. No more coronaritis were detected beyond month 3 of FU. LV dysfunction was present in 58% of patients during acute event, and showed a rapid recovery after therapy start with a full LV function recovery at discharge and thereafter. Mean EF was 48 ± 11% at diagnosis, 58 ± 3 % at discharge, and 63% during FU. At the onset elevated values of NTproBNP and hsTNT were detected (mean value 10521 ± 10377 pg/m and 188 ± 290 pg/ml respectively), and then progressively decreased during hospital stay and normalized at discharge (mean NTproBNP 122 ± 162 pg/ml and hsTNT 4.4 ± 3.2 pg/ml) and during FU. CMR at FU was performed in 22% of patients and did not show presence of myocardial scars. All patients recovered after therapy and no deaths for MIS-C were recorded.

Conclusion: Our experience showed that cardiac involvement in MISC patients is almost the rule. LV dysfunction is the most frequent manifestation, followed by pericarditis. However, patients clinical course was satisfactory and no additional events have been observed during FU. Early treatment is necessary for correct management and early recovery of patients.

EP10

UN RARO CASO DI CARDIOMIOPATIA DILATATIVA IDIOPATICA IN DUE GEMELLI OMOZIGOTI, DALL'ASSISTENZA ECMO VA AL TRAPIANTO CARDIACO: LA GESTIONE INTENSIVISTICA

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Background: La cardiomiopatia dilatativa (DCM) è una condizione caratterizzata da dilatazione e contrazione alterata di uno o entrambi i ventricoli. I pazienti affetti hanno una funzione sistolica gravemente compromessa e possono sviluppare insufficienza cardiaca conclamata (HF). Le manifestazioni di presentazione possono includere aritmie atriali e/o ventricolari e la morte improvvisa può verificarsi in qualsiasi stadio della malattia.
Per la diagnosi è necessaria evidenza di dilatazione e contrazione alterata del ventricolo sinistro o di entrambi i ventricoli (ie. F.E. ventricolare sinistra <40%). La malattia è considerata idiopatica quando le cause primarie e secondarie di malattie cardiache (ie. miocardite e malattia coronarica) vengono escluse dalla valutazione comprensiva di anamnesi ed esame obiettivo, test di laboratorio, angiografia coronarica, ecocardiografia e biopsia endomiocardica quando indicato.
Tra i pazienti con DCM idiopatica, si stima che fino al 50% abbia ereditarietà. Nessun criterio clinico o istologico, a parte l'anamnesi familiare e un attento esame dei parenti, è stato derivato per distinguere la malattia familiare da quella non familiare. La modalità di trasmissione è generalmente autosomica dominante, sebbene siano state descritte anche ereditarietà autosomica recessiva, legata all'X e mitocondriale.
DCM è attualmente responsabile di decessi e ricoveri ogni anno. Inoltre, il DCM idiopatico è l'indicazione primaria per il trapianto cardiaco. Spesso questi pazienti necessitano di supporto dei parametri vitali nella fase acuta di scompenso fino al posizionamento di ECMO VA come bridge to transplant.
Si evince come la gestione intensivistica del perioperatorio, in un contesto multidisciplinare, risulti fondamentale per la stabilizzazione clinica del paziente in un programma di trapianto.

Caso clinico: Due pazienti di 16 anni, gemelli omozigoti con anamnesi patologica remota muta senza alcuna terapia cronica domiciliare e senza una alcuna nota familiarità per cardiomiopatia anche in età giovanile o per morte improvvisa, manifestano, entrambi nell'arco di un mese, uno scompenso cardiaco in cardiomiopatia ipocinetico-dilatativa di nuovo riscontro.
Le principali cause primarie e secondarie di cardiomiopatia dilatativa sono state escluse ai primi accertamenti diagnostici.
I pazienti vengono inizialmente stabilizzati con terapia medica in reparto cardiologico. In seguito al rapido scadimento della performance cardiovascolare si rende necessario il trasferimento in terapia intensiva per impianto di assistenza meccanica con ECMO-VA e successivamente ad atrioseptectomia per venting del ventricolo sinistro.
Vengono entrambi sottoposti, nell'arco di una settimana, a trapianto cardiaco in regime di emergenza nazionale pediatrica. Il decorso post operatorio, seppur gravato da alcune complicanze, gestite senza esiti, è stato poi regolare per entrambi con progressiva ripresa di una adeguata funzione cardiaca e dimissione in reparto di cardiochirurgia.
Di cruciale importanza, durante la degenza in terapia intensiva sono state la gestione dell'assistenza ECMO-VA e la gestione dell'immediato post operatorio in seguito a trapianto cardiaco. Una gestione multidisciplinare tra anestesisti-intensivisti, cardiologi, cardiochirurghi, tecnici di perfusione cardiovascolare, psicologi e personale infermieristico di rianimazione e di sala è stato cruciale per una ottimale gestione di questo raro caso.
Attualmente sono ancora in attesa i risultati dei test genetici per eventuali rare malattie familiari.

Metodo: Analisi della cartella clinica informatizzata e cartacea dei due pazienti, raccolta dei diari clinici medico-infermieristici, consultazione delle valutazioni multidisciplinari, dei file di imaging e dei referti laboratoristici.

EP11

CHIUSURA PERCUTANEA DEL DIFETTO INTERATRIALE: TO SIZE OR NOT TO SIZE?

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Introduzione: la chiusura del difetto interatriale (DIA) è tra le procedure più frequenti nel laboratorio di emodinamica pediatrica ed è una procedura a basso rischio e con un tasso di efficacia molto alto. Recentemente, alcuni Centri, con casistiche prevalentemente focalizzate su pazienti adulti, hanno iniziato a mettere in discussione l'utilità dell'utilizzo del sizing del DIA, proponendo algoritmi di selezione del device sulla base dell'ecocardiografia 2D e 3D sia transtoracica (TTE) che transesofagea (TEE).
Scopo dello studio: valutare il valore incrementale del sizing del DIA per l'algoritmo decisionale del tipo e delle dimensioni del dispositivo da utilizzare in una coorte di pazienti pediatrici.

Metodi: Dal 01/09/2021 al 31/12/2021 sono state eseguite 27 procedure di chiusura percutanea di difetto interatriale. I pazienti sono stati sottoposti ad ecocardiografia transtoracica 2D e 3D pre-operatoria ed ecocardiografia transesofagea 2D e 3D intra-operatoria. Tre operatori (1 ecocardiografista, 1 emodinamista con <5 anni di esperienza e 1 emodinamista con >10 anni di esperienza) sono stati invitati ad esprimere un giudizio sulla misura di device utile alla chiusura del difetto e se reputavano superfluo l'utilizzo del sizing sia sulla base al TTE che sulle immagini del TEE. A prescindere dalle indicazioni a priori degli operatori, il sizing è stato eseguito in tutti i pazienti, e le dimensioni del difetto sono state scelte in funzione del sizing.

Risultati: dei 27 pazienti selezionati, un paziente ha avuto indicazione a chiusura chirurgica dopo il sizing. Il diametro medio dei restanti 26 pazienti 16,8?4,6 mm. Sono stati utilizzati device AGA/Occlutech in 13 pazienti e device Gore in 13 pazienti.
L'ecocardiografista ha espresso la necessità di eseguire il sizing in tutti i pazienti, la sua valutazione è stata identica alla decisione presa dopo il sizing nel 35% dei casi dopo TTE e nel 42% dei casi dopo TEE. Nel 41% e nel 44% dei casi, rispettivamente, il device sarebbe stato sottodimensionato di almeno una misura.
Anche l'emodinamista più giovane non si sarebbe sentito confidente di chiudere il DIA senza sizing in nessun caso, tuttavia la decisione della misura del device risultava coerente a quella identificata dopo il sizing nel 42% dopo TTE e nel 76,5% dopo TEE. L'emodinamista più esperto avrebbe evitato il sizing nel 50% delle procedure, dopo il TEE. L'agreement tra scelta a priori e sizing saliva dal 32% dopo TTE al 77% dopo TEE. Nei 13 pazienti in cui si sarebbe evitato il sizing la misura identificata al TEE è sempre risultata quella appropriata.

Conclusioni: In età pediatrica, la predittività della compliance dei rim al sizing non è scontata. Quando le caratteristiche anatomiche appaiono favorevoli, un operatore esperto potrebbe soprassedere all'esecuzione del sizing, tuttavia il rischio di sottodimensionare o sovradimensionare la protesi di due o più misure è un evento non trascurabile in età pediatrica (tra il 6 e il 20%, a seconda dell'esperienza dell'operatore).

EP12

PERCUTANEOUS APPROACH TO RESIDUAL PULMONARY BIFURCATION STENOSIS IN CONOTRUNCAL DISEASES

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The residual stenosis after right ventricle outflow tract surgery represents a major issue to manage in the pediatric and adult patient with conotruncal defects. Despite a detailed multimodality imaging, the anatomy of distal pulmonary trunk and pulmonary artery bifurcation may be challenging in these patients.
The aim of this study was to analyze retrospectively the outcome of the percutaneous transcatheter treatment in pediatric patients with post-surgical stenosis of pulmonary artery bifurcation.
We enrolled 32 patients with a median age of 5.9 years. Standard high-pressure balloon dilation was attempted in 26 patients, effective in five of them. Pulmonary branch stenting was performed in 10 patients, effective in six. A kissing balloon approach was chosen in 9 patients (6 after angioplasty or stenting failure), this technique was effective in eight cases. Finally, a bifurcation stenting was performed in 11 patients (second step in 9 cases), effective in all the cases. None of the patients approached by kissing balloon needed a bifurcation stenting.
In conclusion, standard balloon angioplasty and standard stenting might be ineffective in post-surgical stenosis involving pulmonary artery bifurcation. In this population, kissing balloon or bifurcation stenting, followed by side branch de-jailing, may be more effective in relieving the gradient.

EP13

STUDIO ELETTROFISIOLOGICO TRANSESOFAGEO PER IL FOLLOW UP DI TACHICARDIE PAROSSISTICHE SOPRAVENTRICOLARI NEONATALI

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Introduzione: La tachicardia parossistica sopraventricolare (TPS) rappresenta la più comune forma di aritmia che insorge in età neonatale. Nella maggior parte dei casi è dovuta alla persistenza di vie accessorie atrioventricolari che residuano dopo la vita fetale causando aritmia da rientro; generalmente tali vie scompaiono nel corso del primo anno di vita del bambino. Lo studio elettrofisiologico transesofageo (SETE) consente di dimostrare l'eventuale persistenza dell'aritmia dopo il primo anno di età e di decidere quindi l'eventuale prosecuzione della terapia antiaritmica.

Scopo dello studio: Valutare l'affidabilità dello studio elettrofisiologico transesofageo nel follow up di questi pazienti.

Materiali e Metodi: Abbiamo osservato retrospettivamente 8 pazienti giunti alla nostra osservazione in età neonatale per tachicardia parossistica sopraventricolare. Tutti i pazienti hanno effettuato ad una prima valutazione ecocardiogramma e Holter ECG e sono stati dimessi in terapia farmacologica efficace per il controllo del ritmo (con uno o due farmaci). Successivamente hanno effettuato un follow up con Holter ECG ogni 3 mesi in terapia farmacologica. Al raggiungimento di un anno di età la terapia antiaritmica è stata sospesa ed è stato eseguito uno studio elettrofisiologico transesofageo comprensivo di stimolazione atriale a frequenze crescenti e di protocollo di stimolazione atriale programmata con singolo, doppio e triplo extrastimolo. In caso di inducibilità di TPS la terapia è stata ripresa e lo studio è stato ripetuto dopo un anno in wash out.

Risultati: Abbiamo osservato 8 pazienti giunti per TPS in età neonatale. Sette di questi avevano un cuore strutturalmente normale; una paziente presentava coartazione aortica corretta con aortoplastica. Dopo un anno di terapia antiaritmica, 4 pazienti (50%), di cui la metà in duplice terapia (flecainide associata a betabloccante e/o ad amiodarone) risultavano negativi allo SETE; questi pazienti, a cui è stata sospesa definitivamente la terapia antiaritmica, non hanno più presentato episodi aritmici nel successivo follow up. I restanti 4 hanno ripreso e proseguito la terapia (duplice nella metà dei casi) per un altro anno e successivamente ripetuto lo studio in wash out: di questi una paziente continuava a presentare aritmie (la paziente con coartazione aortica corretta); gli altri 3 risultavano non inducibili. Nel successivo follow up, di questi 3 pazienti solo una ha mostrato una recidiva di aritmia.

Conclusioni: Lo studio elettrofisiologico transesofageo è una metodica semplice, poco invasiva ed affidabile per identificare i soggetti in cui poter sospendere la terapia per TPS. La persistenza dell'aritmia nel nostro studio è risultata indipendente dal trattamento farmacologico effettuato in mono o duplice terapia. Abbiamo osservato inoltre che in caso di inducibilità di aritmia dopo un anno di terapia, lo studio può essere ripetuto in seguito consentendo di interrompere la terapia in un secondo momento.

EP14

IMPIANTO DI LOOP RECORDER IN SOGGETTO DI ETÀ PEDIATRICA CON SINDROME DI DANON E ARITMIA EXTRASISTOLICA VENTRICOLARE

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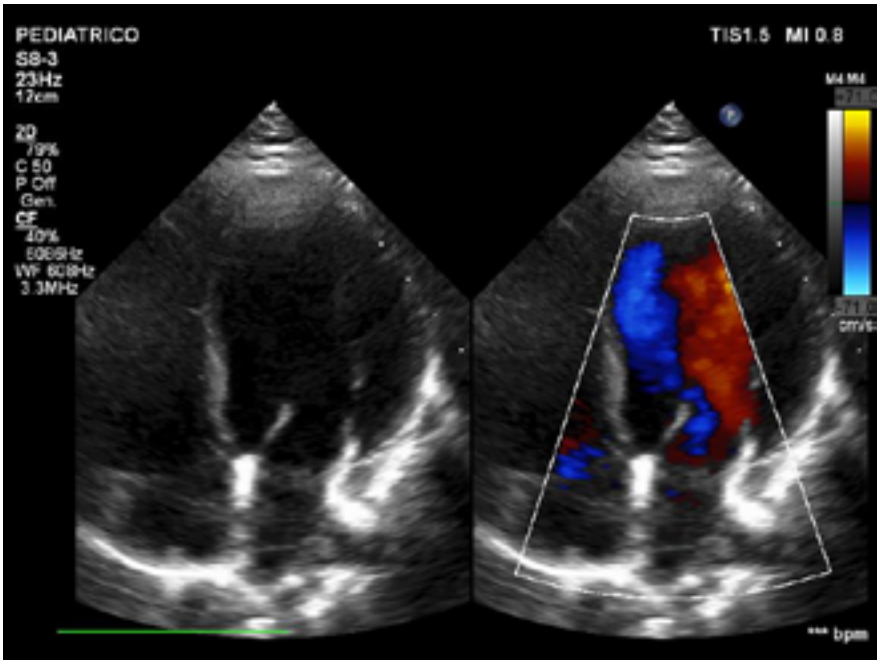
Introduzione: La sindrome di Danon è una rara malattia da accumulo legata al cromosoma X causata da mutazione del gene LAMP2, che si esprime a livello cardiaco con cardiomiopatia ipertrofica e conseguente scompenso cardiaco ed aritmie.

Caso clinico: La piccola C. giunge alla nostra osservazione all'età di 3 anni per aritmia extrasistolica ventricolare. La nonna materna è deceduta improvvisamente all'età di 36 anni, con riscontro all'esame autoptico di cardiomiopatia ipertrofica. La mamma, deceduta a 28 anni per scompenso cardiaco refrattario, era affetta da sindrome di Danon con cardiomiopatia ipertrofica non ostruttiva ad evoluzione dilatativa, episodi di fibrillazione atriale parossistica e tachicardia ventricolare sostenuta, sindrome di Wolff-.Parkinson-White, già sottoposta ad impianto di ICD ed in lista per trapianto cardiaco. La piccola, asintomatica, presenta alla nostra prima valutazione un ecocardiogramma normale; l'Holter ECG delle 24 ore documenta numerose extrasistoli ventricolari isolate, monomorfe, talora ordinate in cadenza bigemina. Viene avviata analisi genetica che accerta la presenza della mutazione germinale nel gene LAMP2 c.928G>A (correlata in letteratura alla malattia di Danon). Nel corso del follow up la paziente, sempre asintomatica per sincope e/o cardiopalmo, presenta comparsa di lieve ipertrofia concentrica del ventricolo sinistro; all'Holter persistenza di rare extrasistoli ventricolari monomorfe, non ripetitive. All'età di 5 anni si esegue impianto di loop recorder iniettabile; la procedura risulta ben tollerata e priva di complicanze e si osserva un ottimo sensing dell'onda R (1,03 mV). Viene inoltre intrapresa terapia con betabloccante. Avviato il monitoraggio a distanza, si osservano nel primo mese dall'impianto alcuni falsi episodi di tachiaritmie dovuti a doppio conteggio per la presenza di onde T di elevato voltaggio; dopo riprogrammazione del dispositivo non si verificano ulteriori falsi allarmi. Nel corso di circa 3 anni il monitoraggio con loop recorder ha permesso di documentare l'assenza di episodi bradi e tachiaritmici sostenuti. Si è inoltre osservata al follow up clinico la riduzione del burden aritmico, con scomparsa pressocchè totale delle extrasistoli ventricolari.

Conclusioni: L'impianto di loop recorder rappresenta una procedura sicura ed affidabile in età pediatrica per il monitoraggio di pazienti a rischio aritmico che non abbiano indicazione evidente all'impianto di defibrillatore. In particolare, la sindrome di Danon si esprime fenotipicamente con la comparsa di cardiomiopatia ipertrofica nella seconda-terza decade di vita; è quindi necessario identificare i soggetti di età pediatrica che possano essere più a rischio di eventi aritmici improvvisi.



EP15

ORIGINE ANOMALA DELL'ARTERIA INTERVENTRICOLARE ANTERIORE DALL'AURICOLA SINISTRA: CASE REPORT

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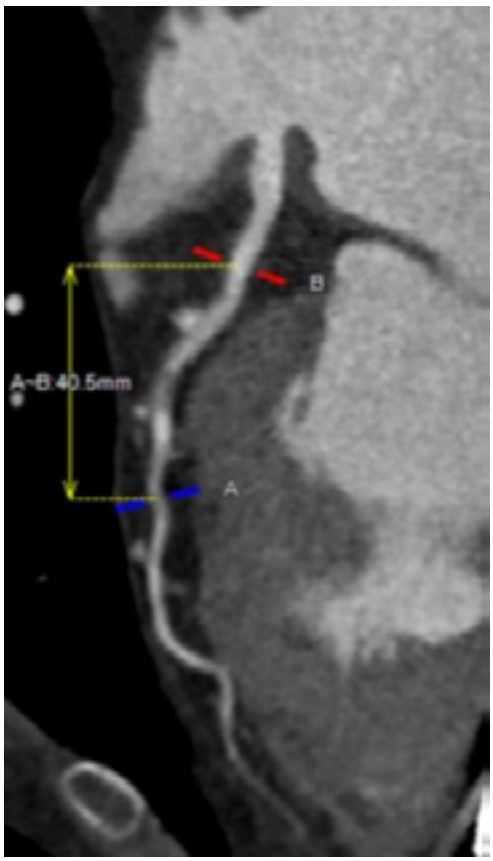
L'incidenza totale delle anomalie coronariche congenite si attesta intorno al 5.6% (1) e il loro impatto clinico varia da casi asintomatici diagnosticati accidentalmente, fino a casi di ischemia miocardica severa e morte cardiaca improvvisa.
Descriviamo un caso di origine anomala dell'arteria interventricolare anteriore (IVA) dall'auricola sinistra, diagnosticata mediante TC cardiaca.
Metodi impiegati: Una donna di 71 anni giunse alla nostra osservazione per scompenso cardiaco congestizio acuto, in corso di un episodio di fibrillazione atriale a rapida risposta ventricolare.
In anamnesi riportava ipertensione arteriosa e blocco atrioventricolare completo, per cui nel 2013 fu sottoposta ad impianto di pacemaker bicamerale.
Un mese prima del ricovero per scompenso cardiaco, il suo pacemaker registrò alcuni episodi di tachicardia ventricolare non sostenuta, per cui aveva programmato ulteriori accertamenti.
Durante il ricovero presso il nostro centro gli esami di laboratorio mostrarono valori aumentati di troponine e di nt-proBNP.
L'ECG di ingresso mostrava ritmo da pacemaker con stimolazione bicamerale.
All'ecocardiogramma il ventricolo sinistro appariva di normali dimensioni con discinesia settale ed ipocinesia della parete anteriore con una funzione sistolica globale moderatamente ridotta (FE 45%). Le camere destre risultavano normali e non vi erano valvulopatie di rilievo. La paziente mostrò beneficio clinico e soggettivo con la terapia diuretica.
Fu quindi sottoposta a studio coronarografico. Le coronarie visualizzate risultarono pervie, tuttavia vi era una origine anomala della circonflessa del seno coronarico destro (Figura 1). Inoltre l'IVA non era visualizzabile e la perfusione della parete anteriore risultava sopperita da modesti vasi provenienti dall'arteria circonflessa.
Per meglio comprendere l'anatomia coronarica, la nostra paziente fu quindi sottoposta a TC cardiaca. Alla TC l'origine dell'IVA risultò chiaramente anomala, a partenza dall'auricola sinistra (Figura 2).
Risultati e Conclusioni: Al meglio delle nostre conoscenze, questo è il primo caso descritto di origine anomala dell'IVA dall'auricola sinistra. Questa estremamente rara anomalia congenita può risultare complessa in termini di gestione clinica, soprattutto in caso di ischemia miocardica acuta. Il quadro clinico di presentazione del nostro caso è stato rappresentato da scompenso cardiaco acuto verosimilmente su base ischemica.
La nostra paziente presentò episodi di fibrillazione atriale parossistica, chiaramente connessa con la formazione di trombi, soprattutto all'interno dell'auricola sinistra, e quindi con una ulteriore riduzione della perfusione dell'IVA anomala.
La paziente fu dimessa in terapia anticoagulante orale e con terapia antiaritmica con Amiodarone per prevenire eventuali recidive di fibrillazione atriale. Venne inoltre posta indicazione a intervento cardiochirurgico di bypass aortocoronarico e di legamento dell'auricola sinistra.



EP16

CARDIAC OUTCOMES IN CHILDREN WITH MIS-C AFTER SIX MONTHS FOLLOW-UP

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Background and Objectives: Cardiac involvement is very common in multisystem inflammatory syndrome in children (MIS-C). Myocardial dysfunction, coronary abnormalities are prominent features. In this study we aim to evaluate the early and 6-month follow-up outcomes of MIS-C.

Methods: This is a longitudinal 6-month cohort study of 20 children admitted and treated for MIS-C from October 2020 to September 2021 at Bambino Gesù Pediatric Hospital in Rome. Patients were followed 4-6 weeks and 4-6 months postadmission

Results: The median age of children was 7 years at admission, 50% were male. Acutely, 9 (45%) patients had left ventricular (LV) systolic dysfunction, 4 of which (20%) required intensive care with vasoactive support, 5 (25%) had coronary dilatation (z score <2.5) and 1 (5%) presented a first and second degree atrioventricular block. 6 patients weren't affected by cardiac abnormalities. All patient received at admission immunomodulatory treatment consisting of methylprednisolone and intravenous immunoglobulin (IgEv). The median length of stay was 13 days (Figure 1). At 4-6 weeks postadmission 1 (5%) patient had persistent dilated coronary artery. None had LV systolic dysfunction or heart rhythm abnormalities. At 4-6 months all patients returned to functional baseline with normal LV systolic function and resolution of coronary abnormalities. No persistent heart rhythm abnormalities were observed. Cardiovascular magnetic resonance performed during recovery in patients with systolic dysfunction didn't revealed any myocardial fibrosis.

Conclusions: A prompt treatment with immunomodulatory drugs, specifically methylprednisolone and intravenous immunoglobulin, led to a rapid recovery and no cardiac sequelae were observed. However, to date the cardiac long-term outcomes in MIS-C patients are unknown, thus a strict follow-up and larger cohorts studies are deeply needed.

EP17

DUCTUS ARTERIOSUS THROMBOSIS EXTENDING INTO THE PULMONARY ARTERY- CASE REPORTS

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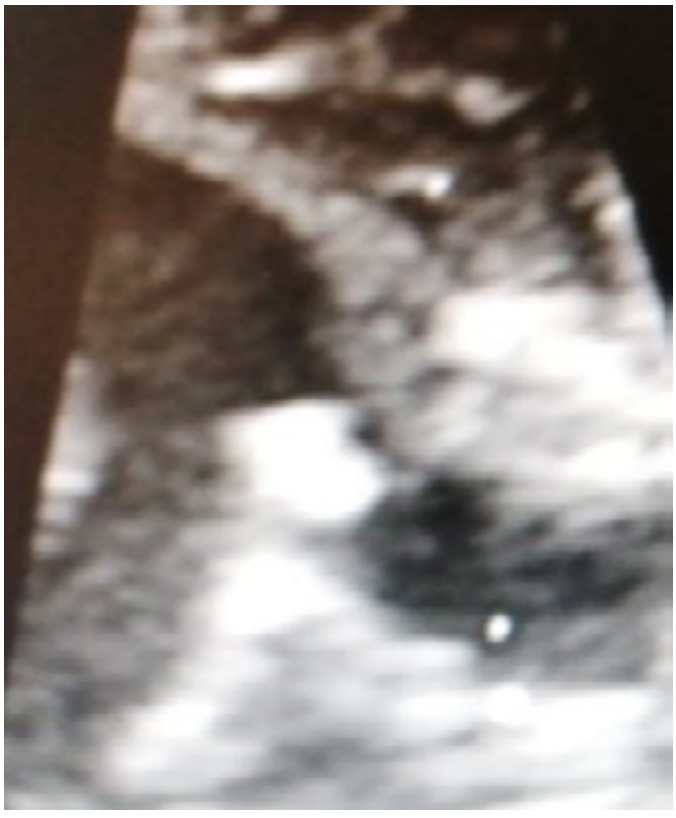
Introduction: Thrombosis of ductus arteriosus and pulmonary arteries is a rare newborn's condition. Few data concerning its pathogenesis exist; however ductal aneurysm is recognized as a potential cause of thrombosis and prenatal closure. Although thrombotic risk factors may play a role, many cases occur in their absence. Currently, no specific guidelines regarding treatment are available. We describe two cases of ductus arteriosus and pulmonary arteries thrombosis diagnosed in our department.

Case 1: male, born at 41 weeks of GA by emergency C-section, because of cardiotocographic alterations, with Apgar score 5 at 1' and 8 at 5' and asphyxia. He underwent therapeutic hypothermia and invasive ventilation for 3 days. Concomitant thrombocytopenia required platelet transfusions. On the first day, echocardiography showed right ventricular hypertrophy and dysfunction and signs of pulmonary hypertension. No flow was detectable in the ductus arteriosus. At second check, a thrombus, of 6 x 9 mm in size, within the ductus was clearly visible, extending into the pulmonary artery, just before the origin of the left branch, without significant Doppler flow changes. CT angiography excluded pulmonary embolism. Since hemodynamic and respiratory values were normal, no urgent invasive intervention was necessary and unfractionated heparin was started. The thrombus decreased in size and, after 10 days, low molecular weight heparin replaced unfractionated heparin. Thrombophilia screening resulted negative. The patient was discharged at 1 month and continued therapeutic low molecular weight heparin administrations for other 2 months, when he switched to aspirin 5 mg/Kg per day. There were no symptoms nor irregular growth. A calcific thrombus attached to the wall of the pulmonary artery persisted.

Case 2: Male, born at 35 weeks of GA with Apgar score 7 at 1' and 8 at 5'. The mother was febrile at delivery. The neonatal course was uncomplicated and antibiotic treatment was administrated. At 5 days of life, physical examination revealed a systolic murmur. Echocardiography showed the presence of ostium secundum atrial defect and thrombotic obliteration of the ductus arteriosus extended into the pulmonary trunk, at the origin of the left pulmonary artery, its size was 3,5x2,9 mm. Right ventricle was mildly enlarged and hypertrophic; systolic function was preserved and no indirect signs of pulmonary artery hypertension were detected. The patient was completely asymptomatic without signs of respiratory or hemodynamic failure. Due to hemodynamic stability and major risk of bleeding of late preterm condition, we started a prophylactic dose of low molecular weight heparin. Thrombophilia screening was negative. The following echocardiographic evaluations documented a mild reduction in thrombotic dimensions and development to calcific thrombus attached to the wall of the pulmonary artery.



Conclusions: Patients affected by ductus arteriosus thrombosis extended to pulmonary arteries may present with symptoms of hemodynamic decompensation, likely due to intrauterine ductus arteriosus closure as possible cause of perinatal asphyxia, or may be totally asymptomatic. Many factors, still unknown, may interact in its pathogenesis. In both cases, the thrombus dimensions did not grow thanks to the use of heparin. As most cases may be asymptomatic, this condition may frequently remain underdiagnosed.

EP18

CARDIOVASCULAR RISK AND TYPE 1 DIBETES IN ADOLESCENT: EARLY ECHOCARDIOGRAPHIC CHANGES AND METABOLIC STATE, STUDY ON A COHORT OF PATIENTS

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Background: Children with type 1 diabetes mellitus are at higher risk to develop early adult-onset cardiovascular disease. Although cardiovascular complications are uncommon in children, some studies have demonstrated subclinical signs of cardiac dysfunction in adolescents with type 1 diabetes mellitus. Diabetic cardiomyopathy is a condition in which heart failure occurred in the absence of coronary artery disease, hypertension, and valvular heart disease; it is due to early complex metabolic disorder in cardiomyocytes that lead to morphological and functional abnormalities of the myocardium. The aim of this study is to determine early echocardiographic changes in adolescents with type 1 diabetes mellitus.

Material and methods: This study includes 30 adolescents affected by type 1 diabetes mellitus without any complications (mean age 14,6 ± 1,83 years) and 30 healthy adolescents (mean age 14 ± 1,52 years). Left ventricular remodeling and diastolic dysfunction have been assessed with conventional echocardiography and with pulsed wave (PW) Doppler echocardiography.

Results: Results of diabetic group and healthy children were compared. The results showed left ventricular diastolic dysfunction as reflected by significantly increased early filling deceleration time in the patients with diabetes (p-value= 0,009). Compared with the control group, patients with diabetes showed a significantly increased in the left ventricular wall thickness (p-value 0,005, p-value 0,006, respectively for the interventricular septum and posterior wall).
There was a correlation between diabetes duration and increased of interventricular septum thickness (p-value 0,012). In the group with diabetes, E/A ratio was significantly lower in female patients compared with male patients. Diabetic patients were divided into two groups according to their HbA1c level (<8% and > 8%). Results showed a significant decreased E/A ratio and increased early filling deceleration times in in patients with HbA1c level > 8% (p-value 0,002, p-value 0,023, respectively).

Conclusion: Our results suggested an early left ventricular remodeling and diastolic dysfunction in adolescent with type 1 diabetes. A chronic worse glycemic control (HbA1c level and diabetes duration) may be the main risk factor for these alterations. A further follow-up and speckle-tracking echocardiography may be useful to assess the relevance of these subclinical signs of cardiac dysfunction.

EP19

RIGHT VENTRICULAR FUNCTION AND OUTCOMES IN LOW BIRTHWEIGHT NEONATES WITH HYPOPLASTIC LEFT HEART SYNDROME

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Introduction: contractile dysfunction and arrhythmias are frequent complications in Systemic Right Ventricle (SRV) and could be explained by profound myofiber disorganization. Moreover, preterm birth affects the cellular structure of RV and carries an increased risk of cardiovascular morbidity, altered systolic and diastolic functions. Although the outcome of cardiac surgery in neonates with low birthweight (LBW) has improved, LBW remains a risk factor for palliation. We sought to estimate whether the LBW affects the SRV function and to describe LBW clinical outcomes after stage I Norwood palliation in neonates with hypoplastic left heart syndrome (HLHS).

Methods: from January 2018 to April 2022 we retrospectively reviewed our experience in 24 consecutive pts with HLHS. Mortality and in-hospital complications (NEC, ECMO, cerebral insult, arrhythmias, and others) were the clinical endpoints. SVR systolic function was analyzed before and after I-stage palliation through the routinely used echocardiographic parameters (RVFAC and TAPSE). We considered 2.5 kg birthweight as a cut-off value for LBW.

Results: mean Gestational Age (GA) was 37 weeks (range 31-39) and mean birthweight was 2.783 ± 0.560 kg. Normal APGAR score was present in all. Seven pts (29%) were LBW, of which two were preterm (31 and 33 weeks GA). Twelve pts (50%) underwent Norwood-Sano Stage I palliation and the rest received Norwood BT-shunt. All 7 LBW (100%) had in-hospital complications compared to 6 normal BW (35%), in detail 3 LBW (43%) compared to 1 normal BW (6%) underwent ECMO (p=0.03). In-hospital mortality was higher in LBW group (3 (43%) vs 2 (12%), p=0.08)). At birth SVR function did not differ among the two groups (RVFAC 42.3 ± 5.0 % in LBW vs 45.2 ± 8.2 %, p=0.44 in normal BW; TAPSE 8.5 ± 0.3 in both, p=0.67) and RVFAC at did not correlate with BW (R2 0.09, p=0.154). The entire population showed a decrease in SVR function 2 weeks after stage I Norwood palliation compared to birth (RVFAC 44.5 ± 7.6, TAPSE 8.5 ± 0.3 at birth vs RVFAC 41.4 ± 5.5, TAPSE 6.3 ± 1.8 2 week after Norwood, p=0.12 and p<0.05 respectively) and a complete recovery at 2 months (RVFAC 45.0 ± 5.3, p=0.82; TAPSE 8 ± 0.6, p=0.07).

Conclusion: LBW affects in-hospital mortality and complications but did not show any effect on SRV both at birth and at 2 months after stage I Norwood palliation. Therefore, our preliminary results did not show an increased impairment in RV function in LBW with HLHS compared to preterm without congenital heart diseases.

EP20

PRENATAL DIAGNOSIS OF LEFT VENTRICOULAR ANEURYSM

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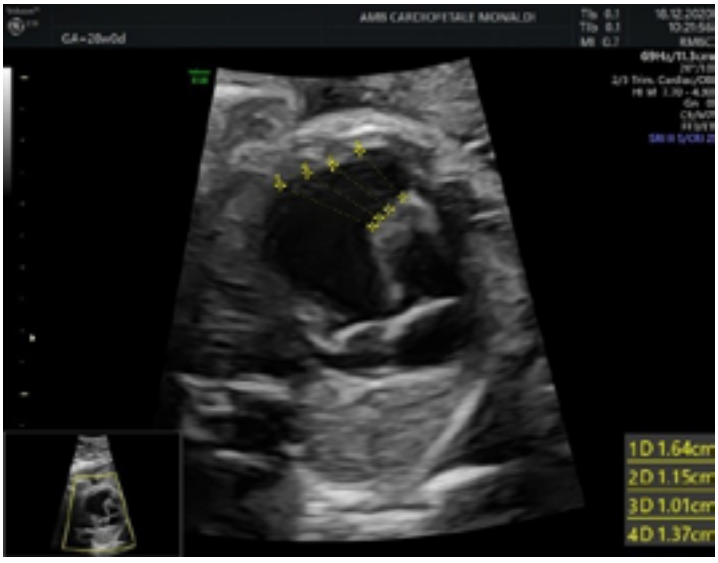
Ventricular outpouching is a rare finding in prenatal sonography and the main differential diagnoses are diverticulum, aneurysm, and pseudoaneurysm in addition to congenital cysts and clefts. The vario modes of fetal presentation of congenital ventricular outpouching include an abnormal four-chamber view on fetal two-dimensional echocardiogram, fetal arrhythmia, fetal hydrops, and pericardial effusion. Left ventricular aneurysm (LVA)/nonapical diverticula are usually isolated defects. Apical diverticula are always associated with midline thoracoabdominal defects (epigastric pulsating diverticulum or large omphalocele) and other structural malformations of the heart. Most patients with LVA/congenital ventricular diverticulum remain clinically asymptomatic but they can potentially give rise to complications such as ventricular tachyarrhythmias, systemic embolism, sudden death, spontaneous rupture, and severe valvular regurgitation. The treatment of asymptomatic LVA and isolated congenital ventricular diverticulum is still undefined.
We describe the case of a patient who came to our observation at 24 w for suspicion of a hypoplastic right ventricle, with evidence on fetal echocardiography of marked dilation of the apex of the left ventricle,
attributable in size (<1 cm) to diverticulum of the ventricular wall. Controls were performed at 28 and 31 W, and fetal echocardiographic measurements showing an increase in the size of the extroflexion and initial signs of regional hypokinesia In the last check at 31 W, a left ventricular apex aneurysm was highlighted (> 1 cm) with marked depression of the ventricular kinetics and pericardial effusion. An emergency caesarean section was therefore performed, with a birth weight of the baby of 1.7 kg.

The neonatal echocardiogram confirmed the diagnosis of left ventricular aneurysm, apical site with depression of the ventricular function (EF 10%), therapy with inotropes was started with no recovery of function with exitus after 17 g of the newborn.



EP21

PRENATAL DIAGNOSIS OF SUSPECTED CHD WITH CCHB AND PMAIVF AT BIRTH

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Congenital complete heart block (CCHB) is a rare cardiac disorder affecting 1 in 15–20,000 patients. 1) Congenital complete heart block is associated with congenital heart disease in ~14–42% of cases . 2) Atrioventricular block is classified as congenital if diagnosed in utero, at birth, or within the first month of life 3).

PMAIVF Congenital /idiopathic pseudoaneurysm of the mitral-aortic intervalvular fibrosa is a rare cardiac anomaly. The typical finding of echocardiography is a pulsatile echo-free sac that expands in systole and collapses in diastole. 4)P-MAIVF is often associated with infection or surgical trauma 5)

We describe a case of a woman at 26 W referred to our center for prenatal diagnosis of large septal atrial defect . At fetal scan we detected a exuberant tissue at the level of the atrial septum in probable contiguity with the mitral valve. The baby was born prematurely due to rhythm abnormalities. Birth weight was 1,800 kg. EKG at the entrance showed a complete heart block with average rate of 68 bpm , at the two-dimensional (2D) transthoracic echocardiograms in 5 chamber and left venticular long axis we detected an echo-free sac in the mitral-aortic intervlvular fibrosa that expanded in systole and collpased in diastole. Other cardiac anomalies were: atrial septal defect and mild aortic regurgitation. Left ventricular function was moderately depressed. After 2 months in NICU the newborn was discharged and now is in close follow-up with 2D Echo and Ekg –Holter to establish the right timing to implant the pmk.

EP22

SUPRAVENTRICULAR TACHYCARDIA IN A CHD PEDIATRIC PATIENT: IS IT TRIGGERED BY SARSCOV2 INFECTION?

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Introduction: The coronavirus disease 2019 is an highly contagious infectious disease and widespread worldwide, which has multiorgan consequences. Mostly studies in adults demonstrate aftermaths of the virus on respiratory system, but there are many hemodynamic complications and electrophysiologic abnomalities. Some reports describe life-threatening arrhythmias as a potential severe complication in COVID-19 adult patients, especially who suffer from pre-existing heart disease (such as inherited arrhythmias syndrome) and other comorbidities (renal failure, hypertension, diabetes, ARDS, cancer). Up to now literature regarding the heart rhythm disorders in pediatric patients affected by COVID-19 infection is quiet lacking. Mostly pediatric studies report the most frequent arrhythmic involvement during multisystem inflammatory syndrome in children (MIS-C), in particular 7-60% of patients have cardiac rhythm disorders. The acute pediatric COVID-19 infection is usually asymptomatic, mild and mostly healthy children don't develop severe cardiac manifestations. They tend to have less life-threatening arrhythmia (PVCs, PACs, incomplete right bundle branch block, first degree AVB, supraventricular tachycardias). Nevertheless there is not actually an exact percentage of heart rhythm disorders COVID-19-related. Rare case reports describe the association and the outcome between CHD and arrhythmias or between healthy children and severe arrhythmias. We are reporting an unusual pediatric arrhythmic case COVID-19- related.

Case presentation: A 16 years-old teeneger with surgically corrected congenital heart disease (atrio-ventricular septal defect-ASVD) and a dual chamber pacemaker for post surgical complete AV block reported palpitations in the two days preceding access to the paediatric emergency department. Remote control of the device at the beginning of symptom showed the onset of atrial flutter from 2 days before and this supraventricular tachyarrhythmia has been never detected at the previous periodic PM controls in telemedicine. At the arrival at emergency department the patient presented stable hemodynamic condition and was completely asymptomatic, the EKG confirmed an atrial flutter 2:1 at heart rate of 120 bpm (Figure 1). The pre-admission nasopharyngeal swab was positive for COVID-19 with all the other blood exams negative, especially inflammatory tests or troponin. An echocardiogram was performed without signs of enlargement of both atria nor valve insufficiency nor ventricular dysfunction. Anticoagulant therapy with subcutaneous heparin was started without considering an antiarrhythmic therapy, but the pacemaker was reprogrammed (DDIR 75-160 bpm) by obtaining a median heart rate of 80 bpm with persistent heart rhythm disorder.
After 15 days of quarantine the nasopharigeal swab was negative. The remote control of the device showed the spontaneous restoration of the sinus rhythm in correspondence with the negativization of the swab. No more supraventricular arrhythmias have been detected in the next short and medium follow up.

Conclusion: No previous literature about atrial flutter in congenital heart disease has been described during a COVID-19 infection before, especially in pediatric population. For the first time ever we recorded an atrial flutter which occurred spontaneously in a CHD pediatric patient during the COVID19 infection. The girl presented stable hemodynamic condition and the self-resolving trend of the tachyarrhythmia in correspondence with the microbiological negativization might suggest the role of this virus as trigger factor of cardiac arrhythmia.

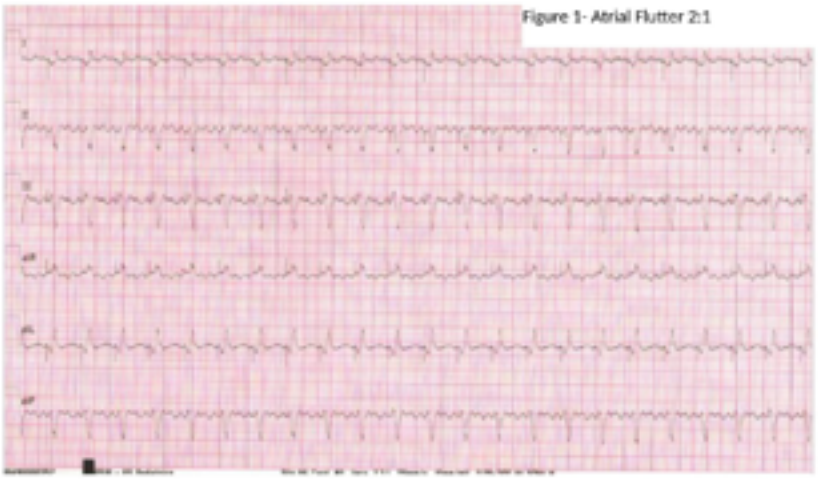
Figure 1- Atrial Flutter 2:1

EP23

UN CASO DI CARDIOPATIA CONGENITA ASSOCIATA A DIABETE NEONATALE CON IPOPLASIA PANCREATICA

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G.A. prematuro di 34+6 settimane, IUGR, PN 1390 g (SGA 1°centile), Apgar 7/8. Nelle prime ore di vita per soffio sistolico 2/6 e sospetta agenesia del dotto di Aranzio, eseguiva valutazione cardiologica che evidenziava una cardiopatia congenita tipo canale atrioventricolare parziale con ampia comunicazione interatriale ostium secundum e piccolo ostium primum, associato a stenosi polmonare in valvola displasica ed aspetto "goose neck" del tratto di efflusso ventricolare sinistro, senza ostruzione significativa. Valutazioni ecocardiografiche successive mostravano un progressivo peggioramento della stenosi polmonare, la cui evoluzione in grado severo a circa un mese imponeva il trasferimento presso un centro di secondo livello. Il paziente è stato sottoposto a due tentativi di dilatazione percutanea dell'anulus valvolare con beneficio iniziale e rapida nuova progressione della stenosi, per cui a 6 mesi eseguiva intervento cardiochirurgico con patch transanulare e correzione del CAV.

Durante la degenza in TIN erano state riscontrate due ulteriori problematiche: iperglicemie che avevano necessitato di insulinoterapia, inizialmente in infusione continua e quindi tramite microinfusore, e segni di colestasi e insufficienza pancreatica esocrina con necessità di terapia enzimatica sostitutiva, confermati dal deficit di elastasi pancreatica e dall'ipoplasia pancreatica con agenesia delle vie biliari alla colangioRM.
L'associazione di una cardiopatia congenita complessa al diabete neonatale con insufficienza pancreatica, ha condotto all'esecuzione di indagini genetiche: il sequenziamento NGS per diabete monogenico è risultato negativo, mentre il CGHarray ha evidenziato una delezione interstiziale in eterozigosi sul cromosoma 8p23 coinvolgente il gene GATA4.

Discussione: La delezione 8p23 è una condizione rara, seppur la sua prevalenza non sia nota. Ad oggi sono riportati 70 casi di delezione interstiziale o terminale sul braccio corto del cromosoma 8. Le manifestazioni sono varie, le più tipiche sono restrizione della crescita fetale, basso peso neonatale, ritardo di crescita, deficit cognitivo, problemi comportamentali (iperattività, impulsività), cardiopatie congenite (CAV, difetti settali, stenosi polmonare, TOF), anomalie craniofacciali, ernia diaframmatica.
Una cardiopatia congenita è presente nel 75% dei pazienti con delezione terminale e nel 94% con delezione interstiziale. La forma più tipica è rappresentata dal CAV, che costituisce la metà dei casi di cardiopatia congenita associata a delezione 8p23. Il caratteristico fenotipo con cardiopatia congenita è dovuto al coinvolgimento del gene GATA 4, localizzato a livello prossimale della banda 8p23.1, che codifica per il fattore di trascrizione GATA4 coinvolto nell'embriogenesi cardiaca.
Mutazioni a carico di GATA4 sono state inoltre descritte come possibile causa di insorgenza di diabete neonatale. È noto, infatti, che GATA4 e GATA6 prendono parte allo sviluppo pancreatico. Tuttavia, un'alterazione a carico di GATA4 rappresenta una causa rara (0,5% dei casi) di diabete neonatale con agenesia pancreatica. In letteratura sono descritti solo 6 casi di mutazione di GATA4 associati a diabete, a differenza dei numerosi (>140) associati ad una cardiopatia congenita, cosa che suggerisce una bassa penetranza del fenotipo pancreatico rispetto al fenotipo cardiaco.

Conclusioni: Con il caso riportato vogliamo sottolineare l'importanza di eseguire valutazioni genetiche in cardiopatie complesse con interessamento di altri organi. È importante, infatti, conoscere una condizione seppur rara, per poter garantire una più appropriata e personalizzata gestione del paziente sia a breve che a lungo termine.

EP24

A CASE OF MASSIVE FETAL CARDIAC RHABDOMYOMA: ULTRASOUND FEATURES AND MANAGEMENT

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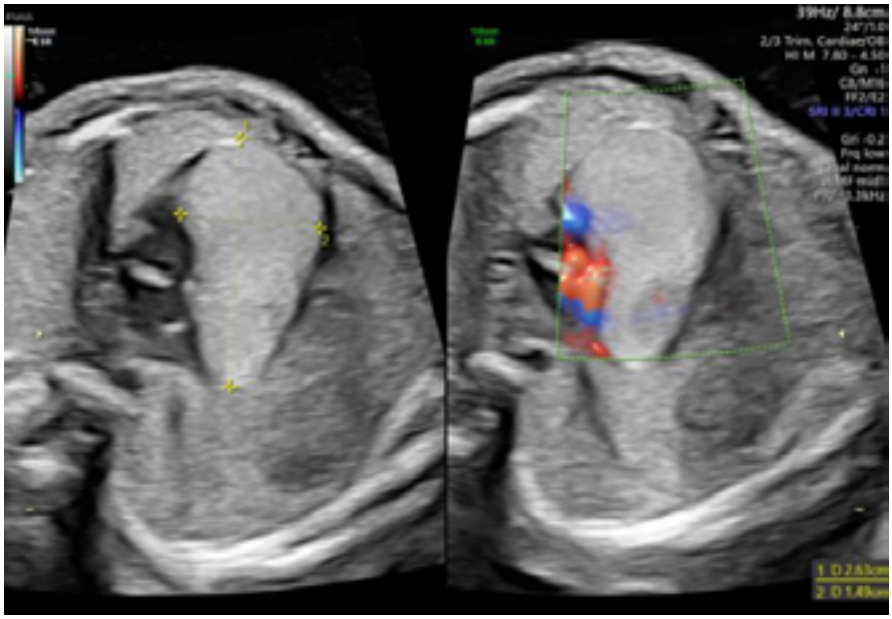
Introduction: Primary heart tumors are an extremely rare finding during prenatal examination. Fetal cardiac tumors are usually benign, with variable clinical manifestations. We report the case of a massive fetal cardiac rhabdomyoma.

Case report: A 31-year-old woman was referred to our clinic at 23 weeks of gestational age because of the finding of a large cardiac mass during the second trimester screening ultrasound, raising the suspicion of cardiac fibroma. Non-invasive prenatal testing and nuchal translucency thickness assessed during the first trimester aneuploidies screening were within normal limits. Echocardiographic examination showed a hyperechogenic homogeneous area of 26x15 mm at the atrio-ventricular junction (Figure 1). It was extending throughout the whole cardiac diameter and moved synchronically with cardiac chambers. Pulsed wave and Color Doppler assessment of atrioventricular and semilunar valves showed normal waves form. A modest pericardial effusion was shown but cardiac frequency was normal. A fetal echoencephalography was performed, showing no visible pathological lesions. Fetal magnetic resonance confirmed the presence of an expansive cardiac neoformation of 29x13x12 mm. The woman underwent an amniocentesis, resulting in a normal female karyotype 46 XX and normal array CHG. However, molecular analysis of genes TSC1 and TSC2 revealed an heterozygotic missense variant of gene TSC2 associated with Tuberous Sclerosis (TS). The same mutation was later found in the asymptomatic mother as well. So the mass was most likely referrable to a single large rhabdomyoma of progressively increasing dimensions. At 37 weeks of gestational age, the mass had reached the remarkable dimension of 48x42 mm, raising the fear of fetal heart failure and malignant arrythmias.
At 39 weeks and 5 days of gestational age, the patient underwent a cesarean section for failed induction and delivered a newborn of 2980 grams, with normal umbilical cord gas analysis and Apgar score. The baby was promptly transferred to Neonatal Intensive Care Unit for cardiac monitoring.
At electrocardiogram (ECG), a ventricular pre-excitation was found, with a marked and diffused ST down-sloping. Echocardiogram confirmed the presence of an intrapericardial wide mass of 42x28 mm strictly connected to the right ventricle, the right atrium and the posterior wall of the left ventricle without obstruction. Postnatal therapies with Everolimus and Flecainide were started.

Conclusions: Among cardiac tumors, rhabdomyomas are the most common. Even though symptoms and prognosis are determined by their size, location and rhythm or flow impairment, they are mostly asymptomatic and are often diagnosed as incidental findings during second or third trimester of pregnancy. Rhabdomyomas might increase in size in utero, with possible arrythmias or hemodynamic impairment in case of large tumors. The association with TS is known so the detection of a rhabdomyoma should be considered a warning sign for TS. Rhabdomyomas are quite often multiple and located along the intraventricular sept. In this case, it was a single large tumor, developing externally along the ventricular wall, making differential diagnosis and management more challenging. New treatments with mTOR inhibitors such as Sirolimus or Everolimus have been proposed to stimulate involution of rhabdomyomas and reduce the necessity of intervention.



EP25

FETAL AORTIC VALVULOPLASTY: EXPERIENCE OF TWO CASES

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Background: Fetal aortic stenosis may progress to hypoplastic left heart syndrome with a poor prognosis. We report two cases of fetal aortic stenosis treated with fetal aortic valvuloplasty using ballon valvuloplasty.

Case presentation: After fetal anesthesia by intrafunicular injection fentanyl and contextual curarization, an ultrasound-guided balloon aortic valvuloplasty was performed in two fetuses with severe aortic stenosis and left ventricular dysfunction at 28 weeks of gestational age between July 2021 and March 2022.
Under continuos echocardiography guidance, a 18-gauge cannula with a stylet needle was introduced into the maternal abdomen and directed from the fetal thorax to left ventricular apex, aiming toward the aortic valve. After advancing the cannula just beneath the aortic valve, the stylet needle was removed and a 0.014 in guide wire was introduced across the valve. A 3 x 12 mm (15 atm, case 1) and a 3,5 x 10 mm (case 2) ballon was used and the balloon was inflated five times (15 atm, case 1) and four times (14 atm, case 2). In the first fetus, mild pericardial effusion developed and resolved within 48 h. In the second case, severe bradycardia occurred so fetal intracardiac administration of adrenaline was performed with resumption of an adequate heart rate. In addition, a mild pericardial effusion developed and resolved in a few days. No maternal complications occurred in either case.
The first fetus was delivered by elective Caesarian section with a birth weight of 3,5 kg at 38 weeks of gestation. Postnatal echocardiography showed bicuspid aortic valve with severe stenosis and moderate left ventricular disfunction. The infant was started on inotrope therapy to improve cardiac function and underwent two aortic balloon dilatations that resulted in residual moderate aortic regurgitation and stenosis. At the time of this writing, he was 10 months and doing well in follow up.
The second infant was delivered by elective Caesarian section with a birth weight of 3,2 kg at 38 weeks + 5 days of gestation. Postnatal echocardiography showed a diminutive left ventricle, severe aortic stenosis and parachute mitral valve with severe mitral stenosis. He underwent aortic balloon dilatation. At the time of this writing, he was 5 days of life and required prostaglandin and high-dose inotrope therapy.

Conclusions: Fetal aortic valvuloplasty seems to be a feasible procedure. It has been reported that it has the potential to prevent progression to hypoplastic left heart syndrome in selected foetuses with severe aortic stenosis. We have reported our initial experience.

EP26

INITIAL EXPERIENCE WITH FETAL CARDIOVASCULAR MAGNETIC RESONANCE

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Background: Fetal cardiovascular magnetic resonance (CMR) imaging may provide a valuable adjunct to fetal echocardiography in the diagnosis of congenital heart disease (CHD). We assessed the utility and safety of fetal CMR.

Materials and Methods: Four fetuses (gestational age 29-31 weeks) were examined using 1.5 T CMR scanners between March and April 2022 at our Istitution. All fetuses had a suspicion for congenital heart disease at fetal echocardiography. All partecipants provided written informed consent. The CMR protocol consisted of anatomical balanced steady state free procession images in the sagittal, transversal and coronal planes. Cine image acquisitions were acquired in 4-chamber, 2-chamber and short axis views using Doppler Ultrasound (DUS) gating. Fetal CMR images were evaluated by 4 observers in consensus, all together with extensive experience in fetal cardiology, echocardiography and CMR.

Results: Scan duration was approximately 20 to 40 minutes without maternal and fetus complications. CHDs were confirmed in all fetuses: one transposition of great vessels, one interruption of aortic arch, one aortic stenosis and one aortic coarctation. In fetus with transposition of the great vessels CMR demonstrated interatrial communication via the restrictive foramen ovale. In fetus with aortic stenosis, fetal CMR demonstrated that the mitral valve was severely underdeveloped and left ventricular contractility reduced making biventricular correction more difficult.

Conclusions: Fetal CMR is a useful and safety complement to fetal echocardiography for evaluating cardiac function and both and vascular anatomy in a wide spectrum of CHDs. CMR added clinically useful information that affected planning of delivery, postnatal care and/or parental counselling.

EP27

DIAGNOSTIC REFERENCE LEVELS FOR PEDIATRIC HEART CATHETERIZATIONS: SETUP OF A NATIONAL PLATFORM

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Obiettivi: Gli stati europei sono tenuti a stabilire, usare e rivedere regolarmente i livelli di riferimento diagnostici (DRL) per le procedure in cui si utilizzino i raggi X, ma fino ad ora non ci sono stati DRL italiani per cardiologia interventistica pediatrica (IC). Valutare i DRL per i bambini è una sfida a causa dell'enorme variazione del peso del paziente.

Metodi: Questo studio prospettico osservazionale della durata di circa 36 mesi ha l'obiettivo di raccogliere i dati di circa 5000 cateterismi diagnostici e/o interventistici con l'obiettivo di trovare i DRL nazionali da un'indagine multicentrica, studiare i parametri di complessità relativi all'esposizione, sviluppare una metodologia affidabile per definire il DRL attraverso le curve dell'indice di dose rispetto al peso del paziente. Ulteriori vantaggi saranno: mettere a punto una struttura utile per l'indagine sui DRL, per creare una collaborazione tra i centri per le revisioni future dei DRL; dimostrare la fattibilità di un registro nazionale delle dosi.

Progetto: Un team dedicato alla dose (fisici medici, cardiologi, tecnologi delle unità di ricerca) sarà incaricato di svolgere le seguenti attività preliminari: elaborazione di una specifica tecnica dettagliata per il servizio di piattaforma web; preparazione di regole standardizzate per la raccolta dei dati (categorizzazione di procedure, nomenclatura, tipo di informazioni dosimetriche e cliniche, formati, ecc.). I dati raccolti includeranno: cateterismi diagnostici e diversi gruppi di interventi, ad esempio chiusura del setto interatriale (ASD), valvuloplastica aortica (AV), trattamento della coartazione dell'aorta (COA), dotto arterioso (PDA), valvuloplastica polmonare (PV). Saranno raccolti i seguenti indici di dose: kerma totale d'aria (Ka, r), dose area product (DAP), tempo totale di fluoroscopia (TF); numero di cine-acquisizioni (N). Le informazioni sulla dose saranno fornite dal DICOM Radiation Dose Structured Reports (RDSR) prodotto dalle unità angiografiche.

Conclusioni: Tale studio multicentrico prospettivo osservazionale consentirà di stabilire dei DRL nazionali età, peso e procedura correlate, in modo da ottimizzare e standardizzare le dosi da utilizzare per ogni singolo cateterismo diagnostico e/o interventistico in età pediatrica nel panorama italiano.

EP28

LA GRAVIDANZA NELLE DONNE CON CARDIOPATIA CONGENITA: IL CASO DELL'OSTRUZIONE ALL'EFFLUSSO SISTEMICO E IL CASO DELL'OSTRUZIONE ALL'EFFLUSSO POLMONARE

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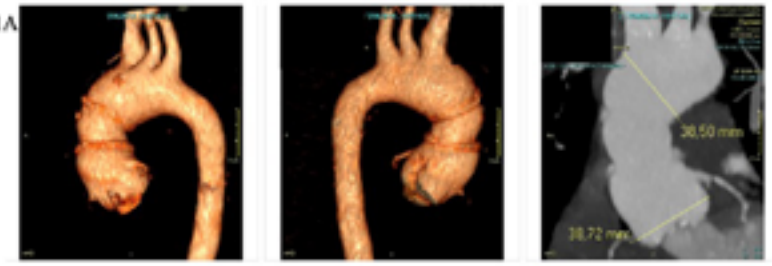
Introduzione: I cambiamenti fisiologici a carico del sistema cardiovascolare che si verificano durante la gravidanza possono risultare drammatici per le donne con cardiopatia congenita, specialmente se gli efflussi risultano coinvolti. La stenosi aortica severa asintomatica e la stenosi polmonare moderato-severa sono classificate dalla Word Health Organization come a rischio III/IV. Presentiamo la gestione clinica di due gravidanze, insorte spontaneamente e non programmate in donne affette da severa ostruzione all'efflusso sistemico in un caso e polmonare nell'altro.

Casi clinici: Il primo caso è una donna di 35 anni affetta da stenosi valvolare aortica su valvola bicuspide e dilatazione dell'aorta ascendente, sottoposta ad intervento di plastica valvolare aortica e sostituzione dell'aorta ascendente con protesi vascolare retta in Dacron (22 anni). Il quadro clinico-strumentale pregravidico (figura 1a) evidenziava una stenosi aortica asintomatica di grado severo (Gradiente pressorio massimo/medio: 46/32 mmHg) e dilatazione dell'aorta ascendente prossimale (43 mm). Durante la gravidanza, si documentava un progressivo incremento del gradiente trans-valvolare aortico (gradiente pressorio massimo/medio al terzo trimestre: 155/78 mmHg) e peggioramento della dilatazione dell'aorta ascendente (50 mm). Il neonato (2031g, 91° percentile4) è stato assistito con ventilazione a pressione positiva tramite Neopuff nel primo minuto di vita (Apgar score al 1° minuto: 4) con successivo adeguato adattamento neonatale (Apgar score: 8 al 5° minuto, 9 al 10° minuto). A due settimane dal parto si confermavano (figura 1b) immodificate le dimensioni dell'aorta ascendente e la severità della stenosi aortica (gradiente massimo/medio: 110/55 mmHg) per cui è stata eseguita valvuloplastica percutanea con miglioramento del quadro.
Il secondo caso è una donna di 30 anni affetta da Sindrome di Shone con coartazione aortica, bicuspidia congenita e stenosi sottovalvolare aortica. Nell'arco della sua storia chirurgica ha eseguito: intervento di coartectomia mediante flap della succlavia (5 mesi), resezione di membrana subaortica (18 mesi), intervento di Ross Konno (7 anni), sostituzione del condotto ventricolo destro-arteria polmonare (21 anni) e posizionamento di valvola protesica biologica per via percutanea in posizione polmonare (22 anni). Il quadro clinico-strumentale pregravidico evidenziava una stenosi severa e sintomatica della valvola protesica (gradiente transvalvolare polmonare massimo/medio: 74/35 mmHg). Durante la gravidanza, non vi sono state significative variazioni dei reperti ecocardiografici (gradiente medio-ventricolare: 72 mmHg, pressioni del ventricolo destro: 100 mmHg). La giovane ha riferito sporadici episodi di cardiopalmo in assenza di correlato significativo agli Holter ECG. La neonata (1617 g, 52° percentile) ha presentato buon adattamento neonatale (Apgar score al 1° e al 5° minuto: 9). I controlli successivi al parto non hanno mostrato sostanziali modifiche del quadro clinico-strumentale. In entrambi i casi, il parto è avvenuto tramite taglio cesareo elettivo a 32 settimane post-amenorrea, in respiro spontaneo e in anestesia loco-regionale, previo incannulamento delle arterie femorali nell'eventualità di dover ricorrere alla circolazione extra-corporea. Non si sono verificate complicanze maggiori intra- né post-procedurali. Il decorso clinico dei due neonati è stato gravato da problematiche di lieve entità e in ogni caso non dissimili da quelle di neonati della stessa età gestazionale.

1A



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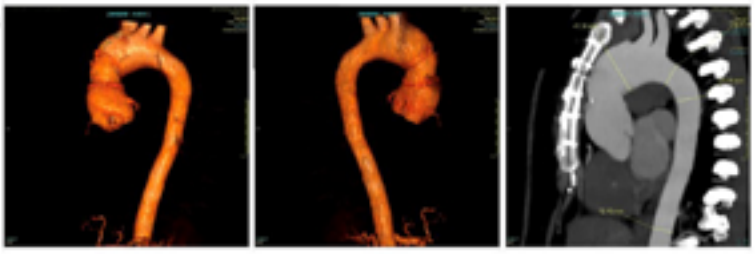

Conclusioni: La condivisione della gestione clinica delle due gravidanze qui descritte fornisce un'importante esperienza clinica.

EP30

GIANT CELLULAR MYOCARDITIS: WHICH STRATEGY SHOULD BE USED TO TREAT?

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Introduction: Giant cell myocarditis (GCM) is a rare disease histologically characterized by the presence of giant multinucleated cells along with extensive inflammatory infiltrates with large areas of necrosis. The pathophysiology of GCM is not well understood beyond its recognition as an autoimmune disorder attributable to T-lymphocyte–mediated myocardial inflammation. The prognosis is poor, and death often ensues unless a heart transplant is performed. The use of immunosuppressive therapy, together with guideline directed medical management of heart failure and arrhythmias, has significantly altered the prognosis. We describe a case of fulminant GCM with severe cardiogenic shock treated by intense immunosuppressive therapy combined with mechanical circulatory assist (MCS) devices, followed by left ventricular assist device (LVAD) implantation.

Case report: A 17-year-old girl with no significant medical history went to the emergency room of a local hospital for fatigue and worsening dyspnea in the previous week. No fever or infectious symptoms were reported. Clinical and chest X-ray revealed acute pulmonary oedema. On echocardiogram, the left ventricle (LV) was dilated with severely reduced ejection fraction (EF 30%) with normal right ventricular function. The patient was initially treated with iv diuretics and transferred to the intensive care unit of our hospital. On admission, she presented with cold extremities, pallor, hypotension (BP 75/55mmHg), lactic acidosis (10 mmol/L), elevation of cardiac biomarkers (hsTnT 1291 pg/mL; NT-proBNP 29.000 pg/mL). Electrolytes, complete blood count and liver function were normal. The echocardiogram revealed dilation and worsening of LVEF (10%) and severe mitral regurgitation. The ECG showed sinus tachycardia (HR 110 bpm) with diffuse ST segment depression. Treatment with iv diuretics and inotropes (adrenaline) was started and mechanical circulatory support (MCS) devices with arteriovenous extracorporeal membrane oxygenation (ECMO) in addition to femorally LV-aortic microaxial pump (Impella) were placed, improving clinical status, and achieving hemodynamic stabilization. Cardiac catheterization with right ventricular endomyocardial biopsy (EMB) was performed. Histological examination revealed an intense inflammatory infiltrate of lymphocytes and plasma cells, with some eosinophils and abundant multinucleated giant cells, as well as low-load Epstein Barr virus (200 copies/ml), Human Herpes virus-6 (<500 copies/ml) and Parvovirus B-19 virus (<100 copies/ml). On the following day, based on this finding, combined immunosuppression was started with iv immunoglobulin (IVIG 2g) for the first two days in addition to iv methylprednisolone 1g for the first three days, followed by a prednisone taper, with a starting dose of 50 mg/day. Because of worsening LV dilatation and development of non-sustained ventricular arrhythmias, antithymocyte globulin (1 mg/kg) iv daily for 3 days was added and tacrolimus and mycophenolate mofetil were started. 20 days after admission, a second EMB was performed: reduction in inflammatory infiltration, without fibrosis. As the persistence of severe cardiac dysfunction depending from Impella, despite guideline-directed medical therapy, MCS support was converted to durable LVAD Heart Mate III as a bridge to recovery or alternatively, as bridge to transplantation.

Conclusion: Our case shows that combination of immunosuppressive therapy along with MCS was helpful to achieve hemodynamic stabilization and preserve organ function in rapid onset GCM with cardiogenic shock refractory to medical therapy.

SESSIONE E-POSTER

EP31

PULSE-OXIMETRY ROLE IN EARLY DETECTION OF CRITICAL CONGENITAL HEART DISEASE

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Background: Congenital heart disease is the most common birth defect and represents nearly 24-40% of all deaths caused by congenital anomalies. Critical congenital heart disease (CCHD), which encompasses the more severe forms, is present in 2.5 to 3 in 1000 live births. In general, prenatal ultrasound or physical examination alone can easily miss newborns with CCHD. Failure to diagnose CCHD early in life can result in high morbidity and mortality rates, because symptoms frequently present after closing of the pulmonary ductus arteriosus, after nursery discharge. Pulse-oximetry screening is a low-cost, painless, noninvasive test that increases the ability to identify newborns with CCHD before they clinically decompensate.
The AHA (American Heart Association) has endorsed newborn pulse-oximetry screening to capture these infants prior to hospital discharge. Despite the urgency of diagnosis, CCHD is missed 1 in 3 newborns. Pulse-oximetry screening is a low cost, noninvasive and painless bedside diagnostic test that can be completed by a technician just in 45 seconds. Many studies sow that Pulse-oximetry screening for CCHD has a less than 1% chance of giving false positive results.

Objective: To develop strategies for the implementation of safe, effective, and ef?cient screening.
The aim of our study is to increase the pre-detection rate of newborns with CCHD before the symptoms appear.

Methods: We prospectively enlisted well newborn term and near term infants from January 2017 to December 2020 in a non-public Maternity structure. All pregnancies was followed according approved protocols with fetal morphologic ultrasound and with cardiac fetal consultation in all cases suspected for CCHD by cardiologist .
Near 24 hours of life trained nursing staff performed pulse- oximetry on the right hand and either foot, using an EDEN-H100B pulse oximeter. Those with saturations > 95% with 3% difference between hand and foot measurements passed. Infants with saturations< 86% failed. Infants with saturations between 86-94% or ?3% difference in saturations were tested again up to 2 additional screens in room air with standard sea level protocol. Providers were notified and echocardiograms ordered for all infants deemed to have failed.

Results: A total of 3205 infants completed the protocol. 0.7% of CCHD screening were made before 24 hours of life, 96.6% after the first 24 hours and 2.7% of newborns have been discharged without CCHD screening. We found 2 case with CCHD (ductus dependent) during the CCHD screening at 24 h of life. Diagnosis was confirmed with heart ultrasound. Prostaglandin was started immediately till surgical intervention had place.
We had only 1 case with CCHD (ductus dependent) the CCHD screening could not discover. None of the infants fall the CCHD screening.

Conclusions: Screening for low blood oxygen saturation through the use of pulse-oximetry monitoring to detect CCHD in well-infant and intermediate care nurseries is a low cost, non invasive and painless bedside diagnostic test, especially in countries with limited resources.
Most of CCHD are ductus dependent and 30% could not discover by CCHD screening. A second screening should be done , maybe before discharge.
The sensitivity of our pulse oximeter should be revised.

EP32

EVALUATION OF LEFT VENTRICULAR MECHANICS IN HEALTHY PEDIATRIC PATIENTS AFTER COVID19 MRNA VACCINATION

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Background: The Covid-19 pandemic has caused substantial morbidity and mortality over the past two years. The development of a safe and effective vaccine has been the key to restoring normalcy by early 2021. The debate on possible adverse effects on myocardial function in children is unfortunately still open. Few cases of myocarditis and hyper-inflammatory syndrome after COVID-19 mRNA vaccine administration were described in the past year. However, whether COVID-19 mRNA vaccine can induce subtle reduction of myocardial function in children is still unknown.

Purpose: The objective of this study was to evaluate ventricular mechanics, by means of two-dimensional speckle-tracking echocardiography, in healthy children in a short-term follow-up following COVID-19 mRNA vaccine administration.

Methods: We analysed a cohort of 19 paediatric patients who underwent COVID-19 mRNA vaccination with Cominarty BioNTech Pfizer vaccine in the previous six months. Patients underwent standard transthoracic echocardiogram and speckle tracking echocardiographic study at our Institution. Nineteen age, sex, and body surface area comparable healthy subjects not subjected to vaccination against Covid 19 were used as control group.

Results: No statistical differences were observed in terms of left ventricular end-diastolic (LVEDD) and end-systolic (LVESD) diameters in the two groups (p=0.554 and p=408), even when normalized for the respective Z-Scores. Moreover, the analysis revealed that the mean ejection fraction (EF) of vaccinated patients was similar both in vaccinated children and in controls (mean EF in vaccinated = 60.7% ± 5, controls = 61.2% ± 7 (p = 0.134)).
Interestingly, also global longitudinal strain (GLS) was similar and not statistical different in the group of vaccinated patients and in CTRLs (-19.5% ± 10 and -20.5% ± 5, respectively (p = 0.121)).

Conclusions: In conclusion, there were no significant differences in left ventricular strain and EF between the group of vaccinated patients and the control group.
It would be interesting to expand the sample size to provide more reliable data.

EP33

ELECTROCARDIOGRAPHIC AND ECHOCARDIOGRAPHIC FOLLOW-UP IN CHILDREN WITH MILD-MODERATE SARS-COV2 INFECTION

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Background: Cardiac alterations associated with COVID-19 have been described even in pediatric population, for example: arrythmias, cardiac dysfunction, coronary injuries. Given the persistence, in some patients, of symptoms after the disease, we focused on evaluation of possible late cardiovascular involvement in children with past paucisymptomatic SARS-CoV2 infection.

Methods: A single centre retrospective study was performed and included 120 children with previous mild COVID-19 disease, who were non-hospitalized. Demographics, symptomatology, ECGs, echocardiographic data were collected. We included patients infected from the first wave on and excluded patients with established cardiac involvement in MISC.

Results: The median age of the group was 8 years (IQR 4-11 years), with a 57,5% of male and 42,5% of female. The mean of the period between infection and the cardiologic follow-up was 5,05 months, the median time 5 months (STD 2,98; 1-2 quartile 3-7 months; IQR 4 months), with a mode of 7 months. (Table I) We divided our cohort into 3 groups: asymptomatic group (20%), paucisymptomatic group (58%) and the moderate group (20%). 8 on 120 patients (6%) complained palpitations as post-covid sign, nevertheless only two patients had ECG signs of tachycardia. Sinusal arrythmias were seen in 35 (29%) children (of which only two with tachyarrythmias), focal block of right bundle branch in 9 (7%) patients. The heart rate of almost all the patients was within normal limits for age (median 90 bpm), only one patient had supraventricular tachyarrythmia, ectopic supraventricular heartbeats, ventricular repolarization with tendency to ST segment depression. PR interval: although in almost all the patients it was within normal range for age, we noted a tendency to shortening. Only in two patients (1,6%) there were an effective shortening of the PR interval below the minimum limit for age, but in 25 patients (21%) we noted an approximation of the PR interval to the minimum limit for age. However, the correlation index of PR interval behaviour and severity of symptoms indicated a weak correlation (0,016). (Table II-III) About echocardiographic findings, all the patients had normal value of EF%, FS%, TAPSE; we noted PAPS>20 mmHg in 14 patients (11,6%) and a PAPS>24 mmHg in 2 patients (1,6%). The median PAPS in our cohort was 15 mmHg (IQR 10-17) (Table IV). We found mitral regurgitation in 4 patients (3,33%), mild mitral insufficiency (1 patient) and thinning of foramen ovale (1 patient). No coronary abnormalities were detected.

Conclusion: Cardiac evaluation of paediatric patients with previous covid-19 disease is mandatory to assess late cardiac symptoms that have been well described in patients with more severe COVID-19 forms or affected by MIS-C. We included in our series only paucisymptomatic patients and we had only slight but significant cardiac involvement. Despite these results, we recommend a cardiac follow-up also in paucisymptomatic children to improve certainty of complete recovery from covid-19 because of cardiac viral tropism and parental concern about a possible late cardiac involvement.

Age (years)	8 (4-11)*
Months from infection	5 (3-7)*
Male	57,5%**
Female	42,5%**

*median (interquartile range)
**%

Table 1

Mean	119,8
median	120,0
Mode	120,0
Range of variation	95,0
Interquartile range	20,0
Variance	338,6
Standard Deviation	18,3
Asymmetry	0,1
kurtosis	0,0

Table 2

Age	6-11 month	1-2 years	3-4 years	5-7 years	8-11 years	12-17 years
Mean	100,3	104	111,16	116,09	125,3	128,5
STD	11,7	8,3	16,54	14,6	19,2	17,2
1 quartile	89,75	96,5	100,75	106,5	120	120
Median	104	110	114	120	126	124
2 Quartile	104,5	110	120	128,5	135	140
IQR	14,75	13,5	19,25	22	15	20

Table III

EP34

A 4-MONTHS OLD INFANT WITH TRICKY ECHOCARDIOGRAPHIC IMAGES: HOW CLINICAL FEATURES MUST ALWAYS GUIDE ULTRASOUND EXAMINATION

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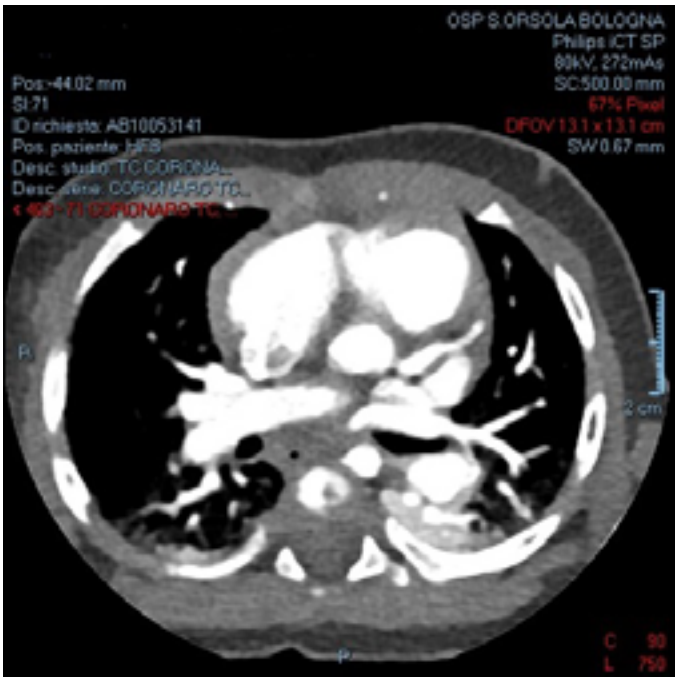
We report a case of a 4-months old female infant, previously healthy, who developed hyperpyrexia with maculopapular rash on her trunk, treated with amoxicillin and clavulanic acid without benefit. After 4 days, because of a non-resolution of the clinical features, she was admitted to our hospital in good clinical condition, with still high-grade fever, no more rash and 2/6 Levine systolic murmur; rest of physical examination was normal. Laboratory findings showed a remarkably high level of C-reactive protein (14.28 mg/dl), serum acute phase reactants including erythrocyte sedimentation rate (85 mm/h) and platelets count (895.000/mmc) with a normal total blood count, no bacterial growth detected in any of her culture and negative viral serologies. She was referred to paediatric cardiologist for prolonged fever. Electrocardiogram showed sinus rhythm, QRS axis directed to the right and no sign of myocardial ischemia. At transthoracic echocardiogram a 9-millimeter atrial septum defect was detected with left to right shunt and moderate right heart dilatation. Normal left ventricle contraction and dimensions. Challenging assessment of anatomy of coronary arteries was highlighted by a suspicious coronary fistula between left anterior coronary artery (LAD) and left pulmonary artery with a remarkable dilatation of left main coronary artery (2.74 mm z score +4.15), LAD (3.95 mm z score +7.46) and also both proximal (2.7 mm z score +4.68) and mid (1.9 mm z score 2.86) right coronary artery. (IMG1)

Clinical, laboratory and echocardiographic findings could not certainly exclude an incomplete Kawasaki Disease (KD). Considering that, intravenous immunoglobulin (2 g/kg) and acetylsalicylic acid treatment (5 mg/kg) were prescribed.

In order to rule out if ultrasound findings could support diagnosis of incomplete KD or refer to a coronary fistula a coronary computed tomography has been performed (IMG 2) and it has rejected the hypothesis of a coronary fistula because of the absence of abnormal connections between the distinct anatomical sites.

Taking that into account, a strict follow-up has been scheduled and 3 months later coronary dilatation was partially improved, both right and left coronary artery (LMA 2.4 mm z score 2.56, LAD 2.8 mm z score 5.88, proximal RCA 2.2 mm z score 2.6). Seven months later coronary dilatation has not been detected anymore and it could have been possible to stop acetylsalicylic acid treatment. We now will check our patient into 2 years.

In conclusion we want to remark how challenging was our imaging diagnosis and how many misleading factors we came across. First of all, we know that KD is really uncommon under one age of year, as rare are also coronary fistulas. Furthermore, incidental finding of atrial septal defect with a dilation of right ventricle with a pulmonary hyperflow could on one hand have masked the absence of connection between LAD and left pulmonary artery and on the other hand have mimicked a coronary fistula draining in left pulmonary artery. With all these tricky factors, we decided to treat our patient as a KD and it has revealed to be the right strategy, as the great outcome our patient achieved.



EP35

NUOVI FARMACI PER IL TRATTAMENTO DELLA INSUFFICIENZA CARDIACA CON RIDOTTA FRAZIONE DI EIEZIONE DEL VENTRICOLO SINISTRO E TACHIARITMIE NELL'ADULTO CON CARDIOPATIA CONGENITA O GENETICAMENTE DETERMINATA

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La fisiopatologia dell'insufficienza cardiaca (IC) nel cardiopatico congenito adulto è variegata, spesso differente dall'IC dell'adulto senza cardiopatia congenita. E' incerta la risposta alle terapie anti-scompenso standard. In questo contesto assume rilievo il possibile ruolo di farmaci non tradizionali o di più recente introduzione. Abbiamo usato due nuovi farmaci consigliati per il trattamento della IC con ridotta frazione di eiezione (rFE), sacubritil/valsartan e SGLT2, per trattare due adulti con cardiopatia congenita o geneticamente determinata con IC, rFE e tachicardia ventricolare (TV), resistenti alle terapie anti-scompenso standard. Il primo paziente è portatore di "Trasposizione delle grandi arterie" sottoposta a switch arterioso, plastica dei rami polmonari, patch di allargamento del tronco dell'arteria polmonare e occlusione trans-catetere del difetto interatriale. All'età di 21 anni si ricovera per scompenso cardiaco e progressivo calo ponderale. Ha una rFE del ventricolo sinistro (VS) (38%), aree di ipoperfusione da stress e a riposo in assenza di kinking delle coronarie reimpiantate e di lesioni coronariche epicardiche alla coronarografia selettiva. Refrattario alla terapia anti-scompenso con ACEi, antialdosteronici e lasix ev, si somministra sacubritil/valsartan (valsartan, bloccante del recettore dell'angiotensina, e sacubritil, inibitore della neprilisina, enzima che degrada il peptide natriuretico promuovendo vasodilatazione e natriuresi). Per le TV viene somministrato bisoprololo, subito sospeso perché scarsamente tollerato per astenia e nausea. Si tratta con levotiroxina l'ipotiroidismo subclinico da tiroidite cronica autoimmune. Il paziente presenta relativa stabilità clinica per diciotto mesi. Si ricovera un mese dopo infezione da SARS CoV2 per recidiva di dolore toracico e lieve dispnea a riposo, presenta stabile sat.O2 95-96%, ricorrenti salve di TV a 180-190/min. Con ivabradina, antiaritmico inibitore della corrente If nel nodo senoatriale, si ottiene un ottimale controllo della frequenza cardiaca e delle tachiaritmie. La TAC torace mostra fibrosi ed enfisema para-cicatriziale, esiti di pleurite. Per dispnea per sforzi lievi e dispnea parossistica notturna, si ottimizza la terapia associando dapagliflozin, inibitore del co-trasportatore renale di sodio-glucosio con azione diuretica e natriuretica. Dopo l'introduzione di dapagliflozin si riduce la dose di diuretico. Il follow-up a breve termine documenta stabile compenso II classe NYHA, buona tollerabilità del farmaco, stabili valori pressori senza necessità di variare la posologia di sacubritil/valsartan. Il secondo paziente è affetto da distrofia muscolare di Becker con "Cardiomiopatia dilatativa e TV", in terapia con bisoprololo, diuretici e ACEi, a basso dosaggio per astenia e ipotensione. A 24 anni presenta un importante deterioramento clinico, dispnea per sforzi minimi e inappetenza, frequenti salve di TV polimorfa, importante rFE (35%). Si somministra sacubritil/valsartan, tollerato alla dose massima di 49mg/93mg per lipotimia, con evidente miglioramento clinico, brevi e occasionali crisi di cardiopalmo, miglioramento dell'appetito e della FEVS (44%). Nel nostro lavoro presentiamo due casi di cardiopatia congenita con insufficienza cardiaca e rFE: 1)cuore biventricolare con VS sistemico e aritmie ventricolari maligne, già sottoposto a molteplici procedure cardiochirurgiche, 2)cardiomiopatia dilatativa e tachiaritmie ventricolari in distrofia muscolare di Becker. Nuovi farmaci, entrati nell'armamentario delle terapie anti-scompenso, sono ancora di utilizzo non codificato nel cardiopatico congenito ma appaiono ben tollerati ed efficaci nella gestione di cardiopatie con differente e variegata fisiopatologia.

EP36

SYSTEMIC SIROLIMUS: A NEW ERA FOR PULMONARY VEINS STENOSIS TREATMENT?

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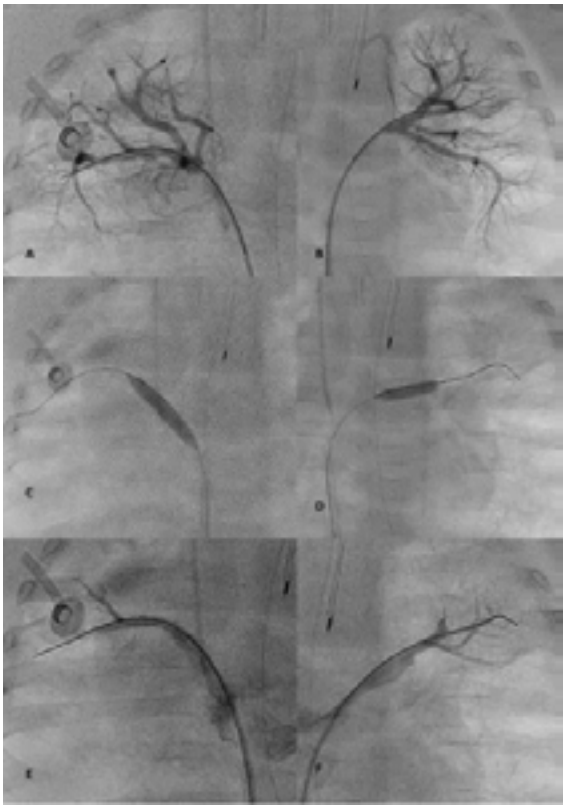
Introduction: Pulmonary veins stenosis, though rare, represents a great challenge of paediatric cardiology, due to its malignant natural history despite surgical or percutaneous treatment. Even when prompt and optimal intervention is performed, early and symptomatic recurrence is observed among most cases, with progressive and potentially fatal decline of clinical status. However, researchers are testing new pharmacological therapies, systemically or through drug-eluting stents administrated, in order to improve overall survival. We describe a case report of an infant in which percutaneous intervention followed by systemic sirolimus medication ameliorated short-term prognosis.

Case description: A male baby was born at 30+5 weeks of gestational age of twin pregnancy by urgent caesarean section, due to IUGR condition. Jaundice and preterm anaemia characterized neonatal period and X-ray showed progressive bronchodysplasia. The baby needed non-invasive ventilatory support, with low levels of oxygen supplementation, for his first 3 months of life. At 2 months old, he also reported profound cardiorespiratory depression requiring cardiopulmonary resuscitation. After this episode, he remained hemodynamically stable for the following period. Screening echocardiography documented spontaneous arteriosus ductus closure, ostium secundum atrial defect with left-to-right shunting and, by the 4th month of life, gradual onset of indirect signs of pulmonary artery hypertension with right chambers' dilatation and progressive development of three pulmonary veins stenosis. Sildenafil treatment was set up, then the infant was transferred to our centre at 6 months of age, with satisfying levels of oxygen saturation on high flow ventilatory support, despite some episodes of dyspnoea. Our assessment confirmed the diagnosis of severe pulmonary veins stenosis with pulmonary artery hypertension (mean pressure of 50 mmHg detected through pulmonary regurgitation), and the following day the baby underwent cardiac catheterization under general anaesthesia and intubation. Post-capillary pulmonary artery hypertension, severe stenosis of left superior, right superior and right inferior pulmonary veins were documented and treated through multiple angioplasties, with both conventional and sirolimus drug-coated balloons. Severe bradycardia and desaturation temporally complicated the procedure, but subsequent respiratory and hemodynamic stability was obtain. Post-procedural echocardiography showed successful reduction in pulmonary veins gradient and a mean pulmonary artery pressure of 23 mmHg. Sildenafil treatment was withdrawn, whereas systemic sirolimus administration at the doses of 0,05 mg/kg was started. Blood count, kidney and liver function, thyroid and lipid profile, lymphocytes subpopulation and serum sirolimus levels were regularly in range, thus sirolimus doses were titrated to 0,07 mg/kg. The following course was uncomplicated and subsequent echocardiography confirmed the persistence of optimal post-procedural result. No more episodes of cyanosis or hemodynamic failure occurred. The patient was discharged with loop diuretic and sirolimus medications. CT scan angiography was scheduled at follow-up.



Conclusions: Despite the natural predisposition to severe recurrence of pulmonary veins stenosis, aggressive combination of interventional treatment and target therapies against neo-intimal and myofibroblast proliferation, like mTOR inhibtors, should be considered, aiming to slow the progression of the disease and to improve short-term prognosis. However, major evidence regarding the real efficacy on long-term outcomes is still lacking, and larger studies are further required.

EP37

A SUCCESSFUL TREATMENT OF A SEVERE PULMONARY ARTERIAL HYPERTENSION IN A 2-MONTH-OLD FEMALE WITH INCONTINENTIA PIGMENTI. A CASE REPORT AND REVIEW OF LITERATURE

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Incontinentia pigmenti (IP) is a rare X-linked dominant neuroectodermal dysplasia, usually lethal in males before birth. Females survive because of X-inactivation mosaicism. IP is caused by a loss-of-function mutations in the IKBKG gene, formerly known as NEMO, located on the Xq28 locus. The NEMO gene is a kinase subunit that activates the NF-?B pathway, which helps to prevent TNF-?-induced cell apoptosis by regulating the expression of genes related to the immune system, inflammation, cell adhesion and apoptosis.
IP occurs in approximately 1:40-50,000 births. Its global incidence is 27.6 new cases per year worldwide: 65-75% are sporadic mutations, while 25-35% are familial.
The clinical features of IP are variable, however in the classic presentation, cutaneous lesions are the first manifestation. They appear in infants at birth or in the first few months of life, and evolve through 4 peculiar, overlapping stages: vesiculobullous, verrucous, hyperpigmented, and atrophic/hypopigmented lesions that correlate with typical histopathologic changes. These cutaneous lesions in all stages tend to affect Blaschko lines on the trunk and extremities.
IP involves primarily skin but can also affect skin appendages (alopecia and nail dystrophy), teeth (hypodontia, microdontia, delayed dentition, conical teeth) and the oral cavity (micrognathia, prognathia, high palate), central nervous system (seizures, mental retardation, microcephaly), eyes (retinal detachment, pigment retinopathy, retrolental dysplasia, strabismus, cataract), and blood cells (eosinophilia). More rarely, cardiovascular abnormalities (such as ASD, tricuspid insufficiency, abnormal shunt of the right PV into SVC or left ventricular endomyocardial fibrosis causing severe mitral regurgitation) and/or pulmonary hypertension have been reported in infants with IP.
Only six cases of PAH without associated congenital heart diseases have been described in IP patients until now. Four of them died before one year of age.
Herein we report the third case of successful treatment of severe PAH in a 2-month-old female infant with IP.
In our case, we had a sudden worsening of PAH that required a severe combined therapy with oral vasodilators (Sildenafil and Bosentan) and intravenous Iloprost plus an anti-inflammatory treatment with intravenous bolus of mPDN and subcutaneous weekly Etanercept injections. The therapy we started is similar to that used by Atallah et al. Unfortunately, we had to stopped Entanercept and Iloprost after few administrations because of an important pulmonary bleeding that required reintubation, evacuative thoracentesis and broad-spectrum anti-infective therapy.
After 40 days of PCICU hospitalization our patient progressively improved and was transferred to Cardiological ward, where she was discharged with double oral anti-hypertensive pulmonary therapy (Bosentan and Sildenafil).
In the follow-up, the evolution was favorable with progressive improvement of the PAP confirmed by RHC after 4 and 12 months [Table1]. PAH drugs were tapered down, and finally all PAH treatments were stopped after 24 months. Subsequent controls did not show signs of PAH. The girl is now 3 years old and she is still undergoing a multidisciplinary follow-up for neurological retardation in the absence of seizures and ophthalmological examinations for ocular anomalies.
This case suggests that aggressive therapy combining vasodilators and anti-inflammatory treatment for PAH might change the course of this devastating complication in IP.

	Basal right heart catheterization (RHC)	RHC after 4 months of anti-hypertensive therapy	RHC after 12 months of anti-hypertensive therapy
RV [mmHg]	50/6	34/11	26/7
Estimated PAP [mmHg]	51	32	24
mPAP [mmHg]	41	22	19
PCW [mmHg]	2	7	10
dAo [mmHg]	51/28/39	62/33/48	80/30*
CI [l/m/mq]	4,09	5,8	4,13
PVRi [WU/mq]	9,54	4,46	2,18
Rp/Rs	1,05	0,19	-

Table 1. Hemodinamic findings before aggressive combination therapy (anti-hypertensive therapy with bosentan, sildenafil, intravenous iloprost and anti-inflammatory treatment with intravenous bolus of methylprednisolone and subcutaneous weekly Etanercept injections) compared to datas after 4 months and 1 year of double anti-hypertensive pulmonary therapy (bosentan and sildenafil) in our case.
RV, right ventricle; estimated PAP, pulmonary arterial systolic pressure; mPAP, mean pulmonary pressure; PCW, pulmonary capillary wedge, dAo, descending aorta; CI, cardiac index; PVRi, pulmonary vascular resistance indexed; Rp/Rs: pulmonary to systemic resistance ratio.
* non-invasive measurement

EP38

A CASE OF A 9-YEAR-OLD GIRL AFFECTED BY GLYCOGEN STORAGE DISEASE TYPE III: DIET SEEM TO PLAY A PROTECTIVE ROLE IN SEVERE CARDIOMYOPATHY'S PROGRESSION

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GSD III is a rare autosomal-recessive metabolic disorder characterized by mutations in the AGL gene that encodes the glycogen debranching enzyme and this leads to an accumulation of abnormal glycogen called "limit dextrin" (LD) in affected tissues. Four clinical types of GSD III are described. The most common form is subtype IIIa that shows hepatic, skeletal muscle and cardiac involvement. Cardiac storage of LD is described in 58% of GSD IIIa affected patients and it can evolve to a severe cardiomyopathy in 15% of cases.

Herein we present a challenging case of a GSD III-affected 9-year-old girl that received a molecular diagnosis of GSD III (c.3304G>A/C.2681+1G>A mutation) at 2 years of age.

At 3 years of age, the echocardiogram showed an initial myocardial thickness (IVSd 10 mm, z score +3,47 and LVPWd 12 mm, z score +4,52) with a mild obstruction gradient on LVOT.

At 8 years of age, she was hospitalized for lower limb pain, fatigue and poor exercise tolerance. Glycemic levels were normal with a dietary regimen based on frequent high-carbohydrate meals associated with cornstarch. The echocardiogram revealed a worsening of cardiomyopathy with a marked increase in the thickness of IVSd (30 mm, z score +14.94) and LVPWd (20 mm, z score +6.78), a mild obstruction gradient (basal: 30 mmHg, during Valsalva maneuver: 40 mmHg) on the LVOT and a severe diastolic dysfuction (TDI lateral E/E': 29). ECG showed marked biventricular hypertrophy with a significant ST-tract depression in all leads, especially from V4 to V6 (max 26 mm) and prolonged QTc (480 ms). Holter-ECG resulted negative for arrythmia. Cardiac enzymes were elevated: NTpro-BNP value was 5273 ng/L (n.v.<450) and CK-MB value was 51,6 ?g/L (n.v <3.1).

The case was discussed for cardiac transplantation but after a careful review of literature, we decided to attempt a modification of the current dietary regimen in favor of a high-fat, high-protein and low-carbohydrate diet. In the literature we found six pediatric cases and two adult cases of GSD III and severe myocardiopathy that after a minimum of 12 months of ketogenic diet showed a significant improvement of IVSd and LVPWd thickness, cardiac enzymes and reversal of cardiac symptoms, avoiding cardiac transplantation.

After 9 months of follow-up, we can report preliminary results of the ketogenic diet that seems to play a protective role in severe cardiomyopathy's progression also in our patient.

The last echocardiographic examination revealed an initial improvement of the cardiomyopathy with a reduction in the thickness of the interventricular septum (IVSd 24 mm, z score +11.68) and the left ventricle posterior wall (LVPWd 19 mm, z score +6.38), a mild obstruction gradient (30 mmHg) on the LVOT and a moderate diastolic dysfuction (TDI lateral E/E': 19). ECG showed marked biventricular hypertrophy with a significant ST-tract depression in all leads, especially V5 and V6 (max 20 mm) and prolonged QTc (500 ms).

Holter-ECG resulted negative for arrythmia. Cardiac enzymes were still elevated but showed a significative reduction: NTpro-BNP value was 2708 ng/L (n.v.<450) and CK-MB value was 29,5 ?g/L (n.v <3.1).

EP39

EFFETTO DELLA TERAPIA MEDICA SULLA DILATAZIONE AORTICA NEI PAZIENTI PEDIATRICI CON VALVOLA AORTICA BICUSPIDE NORMOFUNZIONANTE

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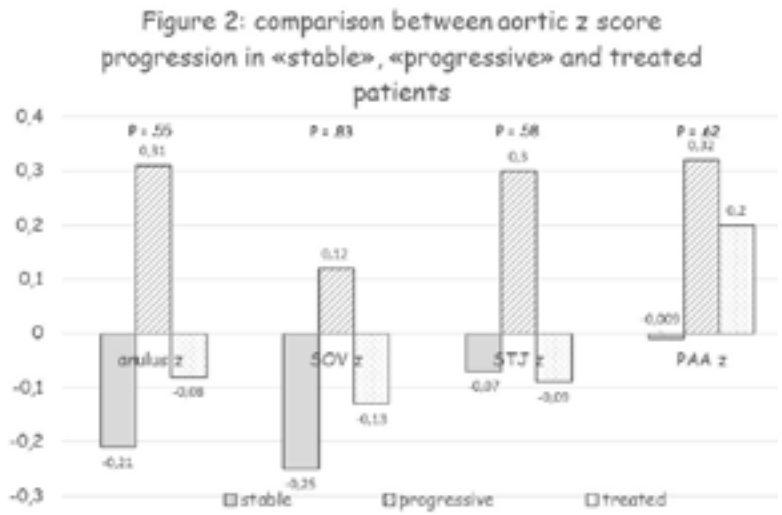
Scopi: Attualmente non ci sono evidenze circa l'efficacia o meno della terapia medica nel contrastare la dilatazione aortica nei pazienti pediatrici con valvola aortica bicuspide (BAV) normofunzionante. La finalità dello studio è stata quella di descrivere l'andamento dei diametri della radice aortica nei pazienti pediatrici con BAV normofunzionante non trattati e nei pazienti con documentata progressiva dilatazione della radice aortica trattati con terapia medica.

Methodi impiegati: È stata condotta un'analisi retrospettiva su 191 pazienti pediatrici con BAV normofunzionante seguiti dal 2005 al 2021 con ecocardiogrammi transtoracici seriati.

Risultati: Nel 46.3% dei pazienti è stata osservata la presenza di una dilatazione della radice aortica, localizzata prevalentemente a livello dell'aorta ascendente prossimale e giudicata di grado lieve. Ad un follow-up medio di 3.7 ± 2.7 anni fra i 175 pazienti non trattati il 52.6% ha presentato una progressione della dilatazione della radice aortica o una dilatazione di nuova insorgenza (gruppo "progressive"), mentre il 47.4% ha presentato diametri aortici normali e stabili (gruppo"stable"). L'8% dei pazienti non trattati con una dilatazione aortica giudicata di grado lieve alla prima valutazione ecocardiografica hanno presentato al follow-up una normalizzazione dei diametri aortici. I pazienti del gruppo "progressive" presentavano più frequentemente una BAV con rafe (73.9% vs 57.8%, p = .037) ed un'insufficienza valvolare aortica di grado lieve (76% vs 45.8%, p = .00007). Trenta pazienti nel gruppo "progressive" sono stati trattati con terapia medica (prevalentemente losartan), gruppo "treated". Ad un follow-up medio di 3.3 ± 2.3 anni non si sono osservate differenze significative nella progressione dello z score aortico fra i pazienti "stable", "progressive" e "treated".

Conclusioni: Attualmente non ci sono specifiche raccomandazioni in merito alla terapia medica dei pazienti pediatrici con BAV e dilatazione aortica. In una coorte di pazienti con BAV normofunzionante si è osservato che nei soggetti con progressiva dilatazione aortica è frequente la presenza del rafe e di una lieve insufficienza valvolare aortica. La terapia medica non ha modificato la dilatazione aortica nei soggetti con dilatazione lieve e progressiva, pertanto la pratica diffusa di prescrivere una bassa dose di losartan nei pazienti con BAV normofunzionante e lieve dilatazione aortica non appare giustificata. Saranno necessari trial randomizzati e controllati per validare queste osservazioni.

Figura: confronto fra la progressione dello z score della radice aortica nei pazienti "stable", "progressive" e "treated": nonostante un trend verso la stabilizzazione dello z score nei pazienti trattati non si sono osservate differenze significative fra i gruppi.



EP40

ESPERIENZA MONOCENTRICA NELLA CHIUSURA PERCUTANEA DEL FORAME OVALE PERVIO IN ETÀ PEDIATRICA

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Background: La chiusura percutanea del Forame Ovale Pervio (PFO) è una strategia terapeutica per la prevenzione dell'ictus criptogenetico ben validata nella popolazione adulta; il suo ruolo nell'età pediatrica è invece meno consolidato.

Scopo della ricerca: Lo scopo di questa ricerca è di analizzare in modo retrospettivo la casistica di pazienti pediatrici sottoposti a trattamento percutaneo di PFO nel nostro centro.

Metodi impegnati: Dal 2006 al 2021, sono state eseguite 21 procedure in pazienti pediatrici (età media: 14 +3 anni; sesso femminile: 58%, peso: 59 + 20 kg).L'indicazione alla chiusura percutanea è stata posta in 18 casi (85,7%) a seguito di un evento ischemico cerebrale, in 2 casi (9,5%) per presenza di emicrania invalidante ed in un caso (4,8%) per ridurre il rischio di embolia paradossa associata ad un intervento di chirurgia addominale. La procedura percutanea è stata eseguita in tutti i pazienti in anestesia generale e con monitoraggio ecocardiografico transesofageo. In 4 pazienti (19%) il PFO era associato ad aneurisma del setto interatriale (che in 2 casi presentava fenestrazioni accessorie su cui si realizzava lo shunt); in un paziente, l'anatomia del setto interatriale era caratterizzata , invece, dalla presenza di un lungo tunnel. La chiusura percutanea del PFO è stata eseguita in 20 casi (95%) utilizzando un singolo device. In 5 pazienti, l'anatomia del setto ha richiesto l'utilizzo di un dispositivo per difetto interatriale. In un singolo caso con ampio aneurisma multifenestrato è stato necessario impiantare 2 dispositivi (di cui un device specificatamente per PFO ed un device per difetto interatriale). Nessuna complicanza peri e post procedurale è stata riportata. In tutti i pazienti, è stato instaurato un trattamento antiaggregante per 6 mesi dopo la procedura.

Conclusioni: La chiusura percutanea del PFO in età pediatrica in centri ad alto volume è una procedura sicura ed efficace. In pazienti con pregresso evento ischemico cerebrale, il trattamento percutaneo può esser utilizzato in prevenzione secondaria per ridurre il rischio di ricorrenza.

EP41

NUOVE PROSPETTIVE FARMACOLOGICHE NEL TRATTAMENTO DELLO SCOMPENSO CARDIACO NELLE CARDIOPATIE CONGENITE CON VENTRICOLO DESTRO SISTEMICO

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Nuove opzioni farmacologiche sono attualmente disponibili nel trattamento dello scompenso cardiaco ma i dati principali sono limitati al paziente con cardiopatia acquisita e disfunzione ventricolare sinistra. A causa della notevole diversità fisiopatologica, queste novità terapeutiche non possono essere automaticamente traslate nelle cardiopatie congenite con ventricolo destro sistemico e fisiologia biventricolare. Questo sottogruppo specifico di pazienti è rappresentato essenzialmente dalla trasposizione delle grandi arterie corretta con intervento di switch atriale (TGA S/P swAtr) e la trasposizione congenitamente corretta delle grandi arterie (ccTGA) Nonostante la sopravvivenza a medio-termine sia ormai buona, la disfunzione ventricolare destra è presente invariabilmente a lungo termine. L'insufficienza tricuspidale, le anomalie di conduzione e le tachiaritmie spesso complicano la storia clinica di questi pazienti.
In questo lavoro presentiamo la nostra esperienza nell'utilizzo del sacubitril/valsartan nel trattamento della disfunzione del ventricolo destro sistemico esemplificandolo attraverso un caso clinico. Il nostro paziente è portatore di trasposizione delle grandi arterie a setto interventricolare intatto, corretta con intervento di Mustard. All'età di 25 anni, circa 2 settimane dopo risoluzione dell'infezione da SARS CoV2, si ricovera con quadro di edema polmonare acuto complicato da flutter atriale con conduzione AV 2:1 e frequenza ventricolare di 150b/min. Il VD appare molto dilatato e presenta compromissione degli indici di funzione sistolica longitudinale e globale. È ottenuta progressiva stabilizzazione del compenso emodinamico con infusione endovenosa di diuretici dell'ansa e antagonisti del recettore dell'aldosterone e ciclo di levosimendan ed è ristabilito il ritmo sinusale con amiodarone. In considerazione della persistenza in classe funzionale NYHA III nonostante terapia medica ottimizzata scegliamo di utilizzare il sacubitril/valsartan, titolato sino al massimo dosaggio. La terapia è risultata ben tollerata e dopo due mesi di terapia il paziente è stabilmente in ritmo sinusale in classe NYHA II, con riduzione significativa dei valori di NT-proBNP (da 4142 pg/ml a 342 pg/ml).
È incerta la risposta del ventricolo destro sistemico alle terapie standard per l'insufficienza cardiaca, fatta eccezione per i diuretici che hanno un ruolo importante per il controllo dei sintomi da ritenzione idrica. Ad esempio, nonostante i beta-bloccanti possano dare benefici prognostici a dosi target elevate, spesso determinano l'insorgenza di bradicardia clinicamente sintomatica in pazienti predisposti ad anomalie della conduzione atrio-ventricolare (ccTGA) o a disfunzione del nodo del seno (TGA S/P swAtr). Il sacubitril/valsartan è un'associazione di un sartanico con un inibitore della neprilisina, enzima che degrada il peptide natriuretico, favorendo vasodilatazione e natriuresi. E' stato dimostrato che il suo utilizzo porta a un significativo miglioramento della sopravvivenza e della qualità di vita dei pazienti con scompenso cardiaco da disfunzione ventricolare sinistra, tanto da essere inserito in classe I di raccomandazione nelle ultime linee guida ESC 2021. Attualmente non può essere suggerita alcuna raccomandazione nelle cardiopatie con ventricolo destro sistemico e fisiologia biventricolare in quanto le osservazioni sono di natura retrospettiva o aneddotica, ad eccezione di un singolo studio longitudinale di 18 pazienti (Zandstra TE, et al. Heart 2021;0:1–6). Nella nostra limitata esperienza il farmaco risulta efficace e ben tollerato. È auspicabile l'avvio di trial randomizzati multicentrici in questa popolazione di pazienti.

EP42

FIRST IMPLANTATION OF EDWARDS SAPIEN 3 VALVE IN BIOINTEGRAL BIOPULMONIC VALVE

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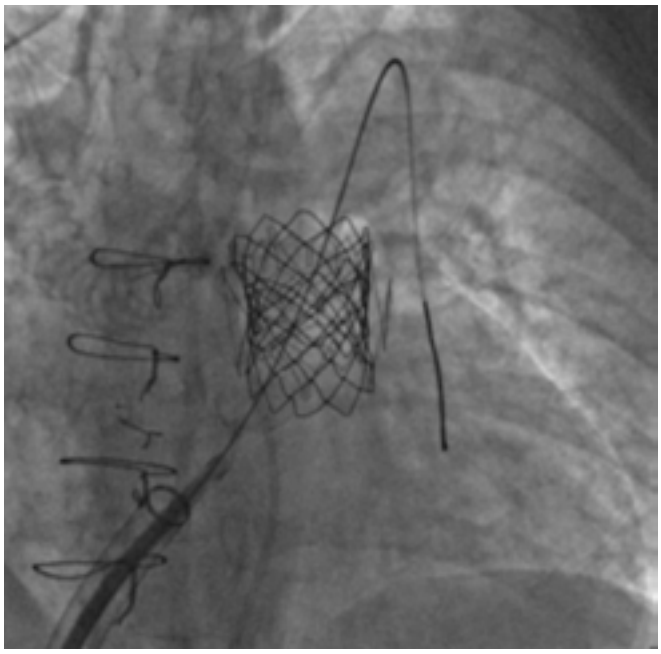
Case presentation: A 42-year-old female with tetralogy of Fallot underwent total correction at the age of one with a transannular patch of the pulmonary valve. Twenty-eight years later, she had an injectable bioprosthesis (BioIntegral Biopulmonic n° 27) surgically inserted (off-pump) in the pulmonary position due to pulmonary valve insufficiency. Subsequent well-being with regular cardiological follow-up and one pregnancy delivered at term without complications. Thirteen years later she was referred to our outpatient clinic for exertional dyspnea.

Diagnostic workup: At physical examination, the patient had a 4/6 systolic ejection murmur. Transthoracic echocardiography revealed severe transprostethic stenosis (mean gradient 50 mmHg – maximum gradient 78 mmHg). Based on the clinical and instrumental assessment, the patient was deemed suitable for valve-in-valve transcatheter pulmonary valve replacement (TPVR). Cardiac CT confirmed the presence of pulmonary stenosis and guided sizing for transcatheter pulmonary valve replacement.

Therapeutic workup and outcome: A 45 mm covered stent (Numed-CP stent) pre-mounted on a 26 mm balloon-in-balloon catheter was placed in BioIntegral pulmonary valve preparing a landing zone and covering calcification. CP stent was then post-dilatated with a Bard Atlas Gold 26 mm at 10 atm. An Edwards Sapien 3 26 mm pulmonary valve was advanced through a Dry Seal 26F and correctly implanted (inflated nominal+1ml). Pressure gradient and pulmonary angiography showed no regurgitation and no residual stenosis. Coronary angiography was done at baseline, after stenting and also after pulmonary valve implantation to exclude coronary compression. The post-operative period was uncomplicated and the patient experienced complete symptomatic resolution. Follow-up echocardiography at one month showed normal prosthetic function (peak gradient 18 mmHg) with no sign of pulmonary valve insufficiency.

Take-home messages: In this setting TPVR is widely accepted as an alternative to surgery. This is the first report of percutaneous Edwards Sapien 3 valve-in-valve implant in Biointegral Biopulmonic injectable valve. We documented the safety and efficacy of such procedure prior CP stent placement.



EP43

SACUBITRIL/VALSARTAN NOT ONLY IN SYSTEMIC RIGHT VENTRICLE: A PRELIMINARY STUDY

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Introduction: Heart failure is one of the main causes of mortality, morbidity, hospitalization and poor quality of life. Among Adult patients with Congenital Heart Disease (ACHD), the data show a prevalence of approximately 20-50% of heart failure and represent an important cause of death. Sacubitril/Valsartan is an established treatment for heart failure with reduced ejection fraction. However, to date, few studies have been published, with controversial data on the efficacy and safety of this drug in ACHD.

Aim: The purpose of the study is to evaluate the efficacy and tolerability of Sacubitril/Valsartan in ACHD, undergoing complete Fontan palliation and in patients with biventricular pathophysiology but with systemic right ventricle.

Methods: Since January 2022, we started to treat the above mentioned subset of patients with Sacubitril/Valsartan. Inclusion criteria were:
a) severe systemic ventricular systolic dysfunction with ejection fraction ?40%;
b) NYHA class II or III.
We evaluated clinical and laboratory parameters at baseline and at three months after administration of Sacubitril/Valsartan.
The primary endpoint was to evaluate the effectiveness of Sacubitril/Valsartan by improving symptoms and/or the NYHA Class and the quality of life according to APPROACH-IS scale.
The secondary endpoint was to test the decrease of NTproBNP and the functional capacity at the ergometric test.
Adverse drug events were also monitored by monitoring creatinine, the onset of hypotension up to drug intolerance and worsening of symptoms or instrumental tests.

Results: We enrolled 5 patients between January and February 2022:
1) All patients showed a reduction of 47% in the value of NTproBNP
2) All patients reported an improvement in quality of life and NYHA Class
3) Two patients who presented ascites showed complete resolution of the effusion
4) The ergometric test performed after three months revealed a better response to effort with an increase in duration of 15%

Conclusions: Our preliminary data are promising and show the efficacy and safety of Sacubitril/Valsartan in ACHD, as reported in patients with heart failure. Further studies are needed to confirm these data.

EP44

EARLY PROGNOSTIC EVALUATION OF CORONARY ARTERY ANEURYSM AND DILATATION IN COVID-19 AND NON- COVID CHILDREN

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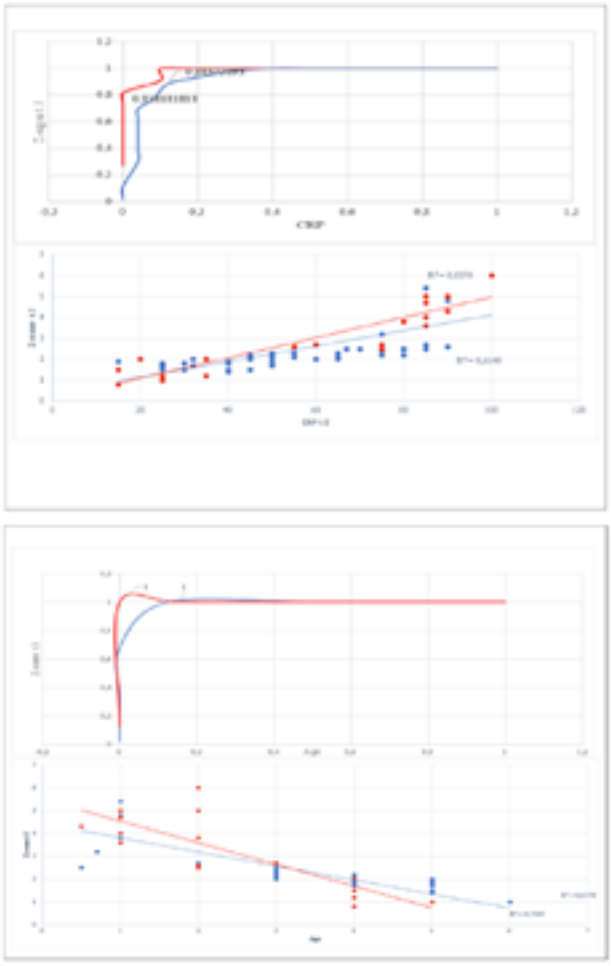
Background: COVID-19 is an infectious disease associated with cardiovascular implications. Although infection with SARS-CoV-2, is usually mild in children, some children later develop a severe inflammatory disease that can have manifestations similar to or Kawasaki disease (KD) with coronary artery dilation or aneurysms have been described. In 2020, the Centers for Disease Control provided a case definition of manifestations for what was ultimately termed multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19 The initial reports of MIS-C described a KD-like illness. There is an increasing appreciation that MIS-C is distinct from KD, but the disorders share some clinical features including coronary artery aneurism (CAA). The treatment of coronary artery ectasia and aneurysms has been adapted from the standard treatment of KD. We aimed to evaluate the risk of coronary artery abnormality development using baseline demographic and laboratory data, including echocardiogram findings in covid children.

Methods: We recruited 89 consecutive children from January 2018 to January 2022 (mean age 2 years) M/F = 1.5 non-covid (68) with KD and covid (21) who had diagnostic criteria for KD or MISC, both received standard IVIg treatment in our pediatric cardiology unit. The following variables were assessed in the risk model: demographic factors, including age, sex, race, ethnicity, and clinical factors; and baseline laboratory studies including white blood cell count (WBC), hemoglobin, hematocrit, platelets, C-reactive protein (CRP). The baseline maximum z score of the proximal right coronary artery and the proximal left anterior descending artery was measured, and coronary artery abnormality was defined as a z score of the coronary artery internal diameter of 2.5 or more. We selected those that correlated with the onset and three-month z scores. We used ROC curves to evaluate the predictive value of these parameters and their specific weight on CAA risk at three months.

Results: We report that age, z-score, WBC, and CRP at baseline are correlated with the risk of persistence of CAA at three months. We evaluated whether there was an increased risk of CAA at three months in both covid and non-covid populations with the same risk factors. We then made a comparison of these cut-offs in the two populations to verify if there was a loss of specificity in the covid population compared to the non-covid one. The cut-offs were evaluated for age and CRP which exceeded the risk at three months for CAA increase. ROC curve relates the sensitivity and specificity of a test to the cut-off value. From the evaluation of these risk curves, it emerges that both populations have the same risk of CAA (Figure).



ROC curves and Pearson correlation index in the two Covid (red) and non-covid (blue) populations.

Conclusion: Patients with covid have mean values ??of CRP, and WBC slightly higher but for them, they have the same risk of persistence of CAA compared to the non-covid population. The onset of the disease at about one year of life appears to be related to a higher risk index for AAC. Multi-center collaborations are key to understanding the natural history, refining diagnostic criteria, developing risk stratification algorithms, and determining the best management.

EP45

TRANSCATHETER CLOSURE OF OSTIUM SECUNDUM ATRIAL SEPTAL DEFECTS WITH GORE® CARDIOFORM ASD OCCLUDER IN PEDIATRIC AGE: SINGLE CENTER EXPERIENCE

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Background. This perspective, observational study evaluated safety and efficacy of the GORE® Cardioform ASD Occluder (WL Gore & Associates, Flagstaff, AZ), a compliant and potentially innovative prosthesis recently approved for closure of ostium secundum atrial septal defects (ASD).

Methods. Between January 2020 and March 2022, 97 children underwent transcatheter ASD closure at our Institution; between these patients 63 with hemodynamically significant ASD were submitted to trans-catheter closure with GORE® Cardioform ASD Occluder. Primary endpoints were procedural success and safety. Secondary endpoints were closure rate and clinical safety at 1-month follow-up.

Results. Patients' age and weight were 7.0±3.5 years (range 3-17, median 6.0) and 27.0±13.8 kg (range 13-67, median 21.5), respectively. ASD diameter was 18.4±4.5 mm (median 18.5), resulting in QP/QS of 1.8±0.7 (median 1.5). Thirty-five patients (36%) were considered "surgical" candidates due to challenging septum morphology, ASD rim deficiency or ASD diameter/patient weight ratio >1.2. Device placement was successfully achieved in all but four patients (94,6%), who needed rescue or elective surgery due to device embolization (n=1), device instability (n=2) or new-onset tricuspid valve regurgitation (n=1). No cross-over with different devices was recorded. Median procedure and fluoroscopy times were 58 and 10 min, respectively. Major adverse events were recorded in 6.3% (4 pts). Complete closure rate was 95.2% at discharge, rising to 98.4% (62/63 pts) at 1-month evaluation, without cardiac or extra-cardiac adverse events. "Challenging" procedures were more time-consuming but as effective and safe as the "simple" ones.

Conclusions. The GORE® Cardioform ASD Occluder device was highly effective and versatile in closure of ASDs with different anatomy and size, even in challenging settings.

EP46

EBSTEIN'S ANOMALY: A SINGLE CENTER SURVEY OF A RARE CONGENITAL HEART DISEASE

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Ebstein's anomaly represents about 1% of all congenital heart disease and is characterized by apical dislocation of the septal and posterior tricuspid valve flaps, with lower dislocation of the functional annulus, dilation of the "atrialized" portion of the right ventricle, redundancy, fenestration, and limitation of the anterior cusp and dilation of the right atrioventricular junction. The malformation spectrum in Ebstein's anomaly can vary from minimal dislocations of the septal and inferior cusps up to an imperforate membrane or a muscular plane between the internal part and the trabecular areas of the right ventricle. Often other heart diseases are associated with Ebstein's anomaly, which can exert a further deleterious effect on cardiopulmonary physiology. Due to the unlimited anatomical variety with which this anomaly can occur, the clinical presentation is extremely variable and can range from a severely symptomatic infant to a casual diagnosis in an adult.
This retrospective study was performed to examine the clinical and surgical outcome of patients with Ebstein's anomaly followed in our Pediatric Cardiology Unit between 1975 and 2021. The mean follow-up period was approximately 21 years and 9 months (ranging from 18 months to 47 years). At least one associated congenital heart disease was described in 77% of patients. Nine patients had electrocardiographic changes, among them, 22% with ventricular pre-excitation, underwent surgical ablation. 38% of the patients underwent cardiac surgery.
Age at diagnosis ? 12 months, hepatomegaly, the need for mechanical ventilation and medical therapy, and the coexistence of other cardiac anomalies are factors associated with a worse outcome.

EP47

GLUCOCORTICOIDS AS SINGLE FIRST-LINE THERAPY IN MIS-C: A SINGLE-CENTRE EXPERIENCE

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Introduction: Multisystem inflammatory syndrome in children (MIS-C) is a rare but life-threating hyperinflammatory disorder, that usually develops 2-6 weeks following SARS-CoV-2 infection. It was firstly described in literature in April 2020 and many aspects of it have to be defined yet. There are currently no universally accepted guidelines for the treatment of MIS-C, although authoritative panels and various scientific societies have published clinical guides recommending the use of intravenous immunoglobulin (IVIg) with steroids as a first line of therapy.
However, there is a lack of comparative efficacy research and randomized controlled trials to compare different treatment strategies in the literature.

Objective: The aim of our observational, monocentric study is to describe clinical, laboratory and instrumental features of MIS-C patients hospitalized in our Centre, with a special attention to cardiac involvement, therapeutic management and short to mid-term outcome.

Materials and Methods: From April 2020 and June 2021, 31 patients with a diagnosis of MIS-C, according to WHO criteria, were hospitalized in our Centre, Regina Margherita Children Hospital, in Turin. We analyzed electronic medical records to collect data. Two patients were not included for insufficient data.
Results. 29 patients received a MIS-C diagnosis, fulfilling WHO criteria, 55% of them were male and medium age was 8.6 years (range 3-14 years). Fever was the most frequent clinical manifestation at admission to the Emergency Unit (87%), followed by muco-cutaneous alterations (60.5%) ad gastrointestinal disorders (51.7%). All patients had high levels of anti-SARS-Co-V-2 IgG antibodies. Blood tests showed a marked increase of inflammatory index (C-reactive protein - medium value 183 mg/L, procalcitonin - m.v. 19 ng/ml, serum ferritin - m.v. 917 ng/ml). Cardiac function was characterized by high NT-proBNP levels (m.v. 7.125 ng/L) and increased troponin levels in 79% of patients. 44.8% of patients showed ventricular disfunction with a variable reduction of the ejection fraction. 10.3% showed a coronary involvement (6.89% transitory ectasias, 3.5% persistent small aneurysms). Intensive care unit hospitalization was needed in two patients, only one of them underwent inotropic therapy. Mechanical ventilation or ECMO were not needed. No patient died.
All the patients received an intravenous high-dose steroid therapy as first line treatment, in 27.6% of cases a step-up to anakinra was done, with good clinical improvement. Only 13.8% of patients, after showing a coronary involvement, received IVIg associated with steroids. Complete restitutio ad integrum was reached in 28 patients.
Comparing the different groups of patients, we found out that the patients with cardiac involvement had a significant increase of the following laboratory values: C-reactive protein (p value 0,049), NT-proBNP (p value 0,023) and Troponin I (p value 0,019). NT-proBNP > 5.000 ng/L was a negative prognostic factor because it was related to myocardial dysfunction at echocardiography.

Conclusions: Our data highlight the effectiveness of intravenous high-dose steroid therapy, eventually associated to a step-up with anakinra. Although this result is promising, further and larger studies are still needed.

EP48

WHEN THE GOING GETS TOUGH THE TOUGH START PLAYING: A CHALLENGING CASE OF FONTAN FENESTRATION

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History: L.G., Double inlet left ventricle with aortic coarctation and aortic stenosis. He underwent multiple palliative interventions that ended with a fenestrated Fontan at the age of 6. In 2015 he underwent fenestration closure with a 4 mm ASD device. In the last years the patient started to suffer from protein losing enteropathy , low cardiac output syndrome (class NHYHA IV), moderate cyanosis (Oxygen saturation at rest 90%) and elevated cavopulmonary pressure (mean 18 mmHG). The patient was not suitable for heart transplant because of hyperimmune state. In 2021 because of moderate Fontan conduit caliber reduction a covered CP stent was positioned. Clinical condition worsened so critical that the patient was not able to be discharged from the hospital. Since the clinical scenario didn't improve despite high doses of diuretics, we decided to re- fenestrate the conduit and implant a Cardiomems system in order to continuosly monitor PA pressure.

Intervention: Patient was intubated and ventilated under general anesthesia. A femoral vein and artery were cannulated and a bolus of 5000 UI of heparin was given. The Fontan conduit and the systemic atrium were delineated by angiography. A Brockenbrough needle was advanced trough a 8 fr long sheath (Medtronic) to the selected perforation site of the conduit. Due to the stiffness of the PTFe membrane the strength of the Brockenbrough needle alone was not enough. So the stiff side of a 0.14 coronary guidewire was inserted into the needle and the other side was connected to a electrosurgical knife. This system finally allowed to cross the Goretex conduit. After creating an AV loop the guidewire was stabilized and the fenestration was progressevely dilated with high pressure balloons starting with a Conquest 7 mm x 20 up to 30 atm and then an Atlas balloon 12 x 20 mm up to 16 atm. A 16 Fr long sheath was then advanced into the systemic atrium and, according to patient's anatomy, a medium size Occlutech AFR device 8 mm was chosen and deployed analogue to a conventional ASD closure. A final angiography revealed an excellent position with right to left shunt. Mean pressure in the cavopolmonary circuit remained stable (about 18 mmHg), mean oxygen saturation decreased to 86%. During the same procedure through a 16 fr long sheath the Cardiomems system was implanted in the lower lobe of the right pulmonary artery. No complications occurred.
During follow up the continuous PA pressure monitoring allowed medical therapy adjustment with a progressive reduction of diuretic dose.

Learning Points of The Procedure: Creation of a Fontan fenestration is safe and effective also in challenging anatomical situations. Cardiomems is a very useful tool in PA monitoring and subsequently medical therapy adjustment.

EP49

MID- TO LONG-TERM OUTCOMES OF AORTIC ARCH REPAIR IN NEONATES AND CHILDREN WITH AORTIC COARCTATION: A SINGLE CENTER EXPERIENCE

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Background: Aortic coarctation is a congenital narrowing of the aorta usually distal to the origin of the left subclavian artery, accounting for 4 to 6 % of all congenital heart defects. Repair of aortic coarctation in neonates and infants is mostly performed surgically and it is associated with a low mortality rate, however, a recurrence of the stenosis (recoarctation) occurs in 10 – 30% of cases.

Aim of The Study: evaluate the mid- to long-term follow-up after aortic coarctation surgical repair focusing on morbidity in terms of recoarctation rate, particularly in relation to different types of surgical approaches and to age at time of intervention.

Methods: we performed a retrospective review of our institute's clinical, echocardiographic and surgical data regarding patients that underwent surgical correction of native aortic coarctation between 1971 and 2018. Patients with left heart syndrome and those with a borderline left ventricle requiring Norwood procedure were excluded.
Recoarctation was defined as a peak systolic gradient of ? 35 mmHg at doppler echocardiography study in addition with the persistency of a diastolic run off. We considered one year after the first surgical intervention as a cut off for an early recoarctation.

Results: 136 patients were included. The median age at the intervention was 39 days (IQR, 13 – 309), 59 patients (43,4%) underwent surgery under the age of one month. 107 patients (78,7%) were treated by extended end-to-end anastomosis (EEEA), 17 (12,5%) by end-to-side anastomosis (ESA), 5 patients (3,7%) with prosthetic (Vaskutek ®) patch aortoplasty, and 1 (0,7%) underwent subclavian flap aortoplasty. The median follow-up was 8,7 years (IQR, 1,8 – 14,9 years). There were two early deaths (1,5%) and no late deaths. Six patients were lost at follow-up. 32 (23,5%) had a recoarctation, of which 20 early one (62,5%), and 12 late one (37,5%). 23 patients (16,9%) underwent a reintervention for the treatment of recurrent coarctation. 20 underwent a balloon angioplasty and 3 a surgical repair. Extended end-to-end anastomosis resulted to be a protective factor for the late recoarctation ($p = 0,034$). Age at surgery ? 30 days was independently associated with a higher rate of early recoarctation ($p = 0,003$).

Conclusions: the results of our study confirm, in agreement with literature, that extended end-to-end anastomosis is an effective approach for aortic coarctation repair and that, compared with other surgical techniques, represents a protective factor for recoarctation in the mid- to long-term follow-up. Patients who are subjected to surgery under one month of age have a higher risk of recoarctation in the first postoperative year.

EP50

UN CASO DI MIS-C CON GRAVE INTERESSAMENTO CARDIACO

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Introduzione: La Multi-system-inflammatory-syndrome in children (MIS-C) è una complicanza che si manifesta da 2 a 6 settimane dopo l'infezione da Sars-Cov2. L'interessamento cardiaco in corso di MISC è frequente e severo; spesso va incontro a rapido miglioramento dopo terapia appropriata. L'evoluzione a lungo termine del danno cardiaco è poco conosciuta.

Report-clinico: A.S 17 anni, con pregressa infezione da Sars-Cov2 30 giorni prima dell'esordio della MIS-C. Accedeva al PS per febbre da 3 giorni (Tmax 39°C), esentema maculopapulare alle estremità, iperemia congiuntivale e fissurazione labiale; leucocitosi con linfopenia e aumento degli indici di flogosi; TC-torace nella norma. Al quarto giorno peggioravano le condizioni generali: persisteva febbre, compariva dolore toracico, si incrementavano gli indici di flogosi e si positivizzavano gli indici di miocitolisi. All'ECG sopraslivellamento dell'ST in sede inferiore; alterazioni omosede della cinetica ventricolare sinistra e versamento pericardico. Diagnosi cardiologica di miopericardite. Per la comparsa di insufficienza respiratoria acuta ipocapnica , la paziente è stata sottoposta a ventilazione meccanica non invasiva. TC-TORACE con e senza mdc: "multiple aree di aumentata densità con aspetto a vetro smerigliato a distribuzione peribroncovascolare e dei lobi inferiori. Versamento pleurico bilaterale". In considerazione dei dati clinici, di laboratorio e della pregressa infezione da Sars-cov2, è stata posta diagnosi di MISC. Veniva avviata terapia con immunoglobuline (IVIG), metilprednisolone, aspirina a dosaggio-antiaggregante, fondaparinux .Il quadro ecocardiografico mostrava ingrandimento delle cavità cardiache destre (segno di McConnel, 60/60, flusso bifido in arteria polmonare come da sovraccarico acuto secondario a polmonite diffusa.) Veniva, somministrato secondo ciclo di IVIG e bolo di metilprednisolone ed avviata terapia antiscompenso. La paziente ha presentato bradicardia asintomatica con FC 30 bpm per cui è stata interrotta terapia ?- bloccante. Graduale miglioramento clinico: defervescenza e negativizzazione degli di flogosi in 8° giornata; normalizzazione delle cavità cardiache destre e della FE in 12° giornata. E' stata dimessa dopo 20 giorni in buone condizioni cliniche generali ed inserita in un programma di follow-up multidisciplinare comprendente esami seriati , fisiokinesiterapia e Rm-cardiaca.

Conclusioni: La MISC può presentarsi, anche in maniera subdola, con interessamento multisistemico sino a grave scompenso-circolatorio ed insufficienza respiratoria. Il ritardo nell'avvio del trattamento immunomodulante, come nel caso in esame, potrebbe avere un impatto negativo sull'evoluzione dei quadri più gravi di MISC. Il clinico deve pertanto, saper riconoscere e trattare precocemente questa patologia. E' utile un monitoraggio cardiaco continuo per identificare tempestivamente aritmie-maligne anche di natura ipocinetica, come suggerito dal riscontro nella nostra paziente di grave bradicardia sinusale post picco infiammatorio. Riteniamo che tali aritmie ipocinetiche siano secondarie all'infiammazione e all'edema del tessuto di conduzione. Nel caso descritto abbiamo osservato un insolito e transitorio coinvolgimento del cuore destro verosimilmente da ascrivere alla grave polmonite interstiziale.

EP51

ROLE OF ?P(V-A)CO2/?C(A-V)O2 RATIO AS A HEMODYNAMIC MARKER IN PEDIATRIC CONGENITAL CARDIAC SURGERY PATIENTS: A SINGLE CENTER PROSPECTIVE STUDY

I. Tomasella [1], M. Taiana [1], B. Martinez Lopez de Arroyab [1], A. Russo [1], A. De Vigili [1], M. Ceola Graziadei [1], E. Bresadola [2], G.B. Luciani [1], L. Gottin [1]

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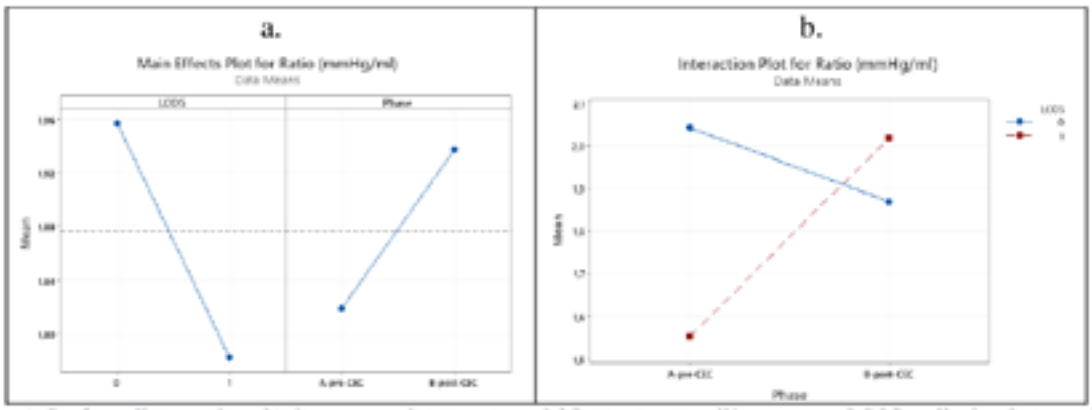
Introduction: Low Cardiac Output Syndrome (LCOS) is one of the most frequent complication in pediatric patients with congenital heart disease undergoing corrective surgery. This condition is associated with a high morbility and mortality rate so early recognition is crucial. Up to date, several perfusion markers such as serum lactate concentration and central venous saturation (ScVO2) have been used to recognize LCOS. Nevertheless, these markers seem to be not completely reliable in order to identify a state of global tissue hypoxia. [1]

Aim: The aim of the study is to establish the ability of the variation of Ratio ?P(v-a)CO2/?C(a-v)O2 ratio to detect LCOS development and the reliability of classical perfusion markers to identify LCOS in a pediatric cardiac surgery population.

Methods: Arterial and central venous blood samples were collected and analyzed at different stages of the surgical procedure and postoperative period. At any stage the parameters analyzed were: PaCO2, PaO2, PvO2, PvCO2, SaO2, ScVO2, lactate, hemoglobin, cerebral and somatic NIRS, inotropic doses, ERO2, ?P(v-a)CO2, creatinine. LCOS was defined as the development of organ dysfunction leading to a higher inotropic and vasoactive requirement more than usual CPB weaning support or ICU pharmacological support. Furthermore, acute kidney injury (AKI) was defined according with the Pediatric modified Acute Kidney Injury Network Criteria. Study population included all cyanotic and not cyanotic pediatric congenital heart disease patients (between 0-14 years) undergoing corrective and palliative cardiac procedures. Fisher's test and the non-parametric Mann-Whitney test was used for statistical analysis.

Main Results: 98 patients undergoing corrective and palliative cardiac surgery from June 2018 to October 2020 have been included in the study. 46 children of 98 (46,9%) developed LCOS. No statistically significant association between the increase of Ratio and the onset of LCOS was found neither in total population nor in the cyanotic heart disease group. According to literature our Ratio cut off was set at 1.4 mmHg/ml. [2][3] However, between pre-CPB phase and post-CPB phase an average increase in the Ratio values by + 31% was observed in patients developing LCOS. Furthermore, the data showed a decrease in the mean value of Ratio in cases in which LCOS did not develop. The statistical significance was 94% with a p-value 0,06.

Conclusion: The study shows that Ratio can not be identified as a reliable marker of LCOS in the pediatric population affected by congenital heart disease and undergoing cardiac surgery. However, an increase in Ratio, although not statistically significant, is present when LCOS occurs. In conclusion, the trend of the Ratio would be useful not as a single marker but as a component of a much more integrated approach to hemodynamic monitoring of LCOS. Moreover, the study confirms that serum lactate, ScVO2, ?P(v-a)CO, ERO2 are strong markers both pre and post-operative of LCOS.



a) Grafico effetto medio e b) diagramma di interazione del Ratio rispetto all'insorgenza LCOS e alle due fasi pre-CEC e post-CEC.

EP52

CARDIOMIOPATIA E CUORE D'ATLETA SEPARATI DA UNA LINEA SOTTILE

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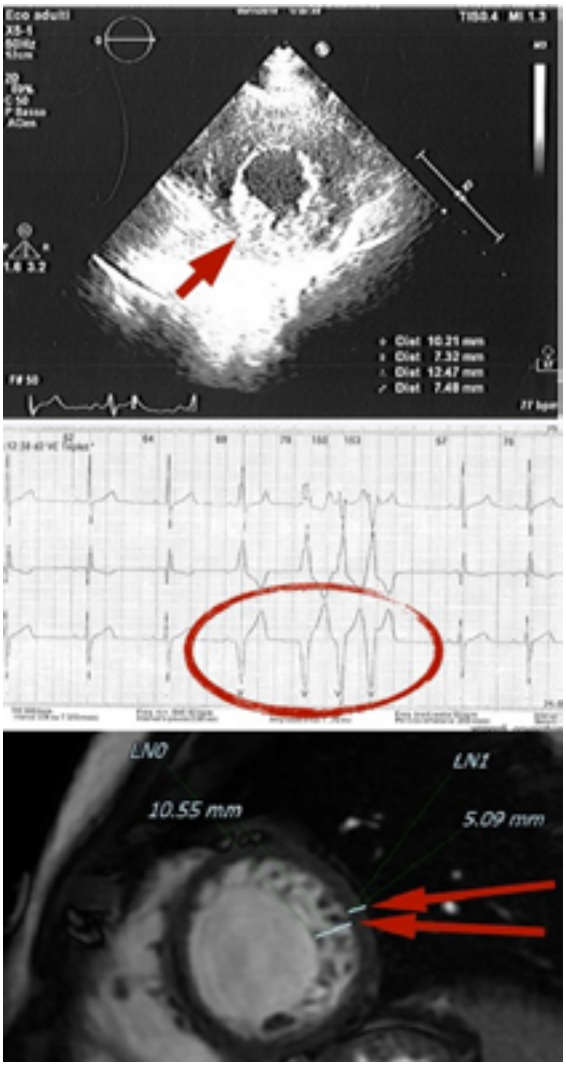
Descrizione di un caso clinico.

Metodi impiegati: Ragazzo di 12 anni, pratica ju-jitsu ad elevato livello agonistico.
Durante un ricovero in chirurgia pediatrica per torsione del testicolo, all'ECG preoperatorio evidenza all'ECG di mancanza delle forze elettriche sinistre (scarsa progressione onda R da V2-V6), emiblocco posteriore sinistro, deviazione assiale destra (QRS 135°), ripolarizzazione nella norma, consigliato ecocardiogramma di completamento. All'ecocardio evidenza di accentuata trabecolatura a livello del ventricolo sinistro (NC/C 1,8-1,9). Consigliato Holter ECG (3 canali) che evidenzia una tripletta di extrasistoli ventricolari notturna; riposizionato un Holter a 12 derivazioni risultato nella norma. Alla luce dei reperti riscontrati si decide di bloccare l'attività agonistica e procedere con gli accertamenti. La RMN cardiaca conferma l'aspetto di ipertrabecolatura del ventricolo sinistro (NC/C 2). Si invia il ragazzo presso il centro di riferimento nazionale per ulteriore valutazione, la quale conferma all'ecocardiogramma la diagnosi di miocardio non compatto (NC/C 2.1); eseguita prova da sforzo risultata nella norma ed Holter ECG con isolate extrasistoli ventricolari. Data la forte motivazione del ragazzo, l'assenza di aritmie significative all'ECG sotto sforzo e agli Holter eseguiti in corso di allenamento, le normali dimensioni e la normale funzione sistolica ventricolare sinistra, viene concessa l'idoneità sportiva agonistica per 6 mesi.
Inizia la pandemia e gli allenamenti vengono drasticamente ridotti. Al controllo cardiologico un anno dopo evidenza all'ECG di presenza delle forze elettriche sinistre, all'ecocardio riduzione della trabecolatura del ventricolo sinistro (NC/C 1.4-1.7), all'Holter ECG assenza di aritmie significative; viene effettuato prelievo genetico per cardiomiopatie e consigliato impianto di loop-recorder che il paziente e i genitori rifiutano per il rischio intrinseco legato ai traumi sul torace che la pratica sportiva del ragazzo comporta. Al controllo sei mesi dopo ulteriore riduzione della trabecolatura del ventricolo sinistro (NC/C 1.1), all'Holter ECG assenza di aritmie significative. Arriva il risultato dell'esame genetico che risulta negativo per varianti patogenetiche correlabili con cardiomiopatie.
Il ragazzo può riprendere l'attività sportiva agonistica e continua a raggiungere risultati eccellenti a livello nazionale.



Risultati e Conclusioni: Questo caso clinico mostra quanto vi sia una sottile linea che separa il cuore d'atleta dal cuore con cardiomiopatia; la presenza dell'isolata tripletta di extrasistoli ventricolari ha acceso un campanello d'allarme al quale hanno fatto seguito una serie di accertamenti volti alla ricerca o all'esclusione di una cardiomiopatia sottostante. Il decondizionamento ha aiutato per dirimere il dubbio nella gestione di questo caso clinico, permettendo al ragazzo di proseguire la sua normale attività sportiva agonistica. Può sembrare un eccesso bloccare l'attività sportiva agonistica di un promettente giovane atleta, in questo caso fortunatamente la pandemia è stata stranamente utile, ma alla luce dei pregressi tristi episodi di morti improvvise in giovani sportivi affetti da cardiomiopatia aritmogena non diagnosticata che hanno dato segno di se solo con extrasistoli ventricolari, forse vale la pena fermarsi un attimo e fare tutti gli accertamenti necessari per far praticare l'attività sportiva nel modo più sicuro possibile.

EP53

TRANSCATHETER SECUNDUM ATRIAL SEPTAL DEFECT CLOSURE USING INTRACARDIAC ECHOCARDIOGRAPHY IN ADULT PATIENT WITH AZYGOS/EMIAZYGOS CONTINUATION OF THE INFERIOR VENA CAVA

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Introduction; Percutaneous closure of an isolated secundum atrial septal defect (ASD) has become the first-line treatment in patients with sufficient rim. In patients with secundum ASD, the procedure is mostly performed through the femoral vein. Standard techniques include the use of balloon-size technique and intracardiac echocardiography (ICE), the latter as an accepted alternative to the transoesophageal echocardiography (TEE). Congenital anomalies of the deep thoracoabdominal venous system are caused by variations in the development during embryogenesis. As described in the literature, knowledge of this anatomic arrangement is important in percutaneous cardiopulmonary procedures.

Case summary: A 53-year-old female patient was admitted to our cardiology department with recent complaint of dyspnea. Physical examination revealed a fixed splitting second heart sound. Chest radiography showed prominent pulmonary vasculature. Transthoracic echocardiography revealed enlarged right heart cavities and TEE confirmed a 20 x 17 mm secundum ASD, with just enough rims for percutaneous interventional closure, except the anterior rim (2 mm).
The patient was admitted to catheter laboratory for percutaneous closure of secundum ASD through the right femoral approach.

Results. The ICE catheter was inserted via 9 F long venous sheath from the left femoral vein and advanced to attempt through the Inferior Vena Cava (IVC) to enter into the right atrium. Venography was performed because of the abnormal course of the catheter and an azygos/hemiazygos continuation of the left iliac vein, up to the Superior Vena Cava (SVC).
For this double IVC with hemiazygos continuation of the left-side IVC and for a very acute angle of the draining azygos/hemiazygos veins into the SVC, it was not possible to advance the ICE into the right atrium through this venous system. The right femoral vein was therefore catheterised twice: proximally with a 7 F sheath, which was used for preoperative right cardiac catheterization and pulmonary pressure measurement and replaced with 12 F long sheath for device implantation; distally with 9 F sheath for the ICE study in the right atrium.
The interatrial septal defect was passed through using a superstiff-0.035' Amplatzer wire and directed toward the left superior pulmonary vein. The dimensions of the ASD were confirmed by the sizing balloon, and compared with the ICE. For the small anterior rim, a 37 mm Gore Cardioform occluder was loaded and advanced through the sheath, and the defect was closed using the routine protocol. The position and stability of the device were evaluated on ICE. No post-procedure residual shunt was detected.
Postoperatively, computed tomography angiography was performed through the lower extremity. The patient was discharged 2 days later with oral aspirin.

Conclusions: The interventional endovascular maneuvers normally coded for vascular access, materials, and technique, must be carefully re-evaluated in case of congenital anomalies involving the systemic venous return to the right atrium. These anomalies are usually asymptomatic and incidentally found, and the major clinical significance of their recognition is to prevent misdiagnosis, which can have implications on invasive procedures and abdominal hemorrhage (a highly feared complication).

EP54

UN CASO DI AORTA QUADRICUSPIDE

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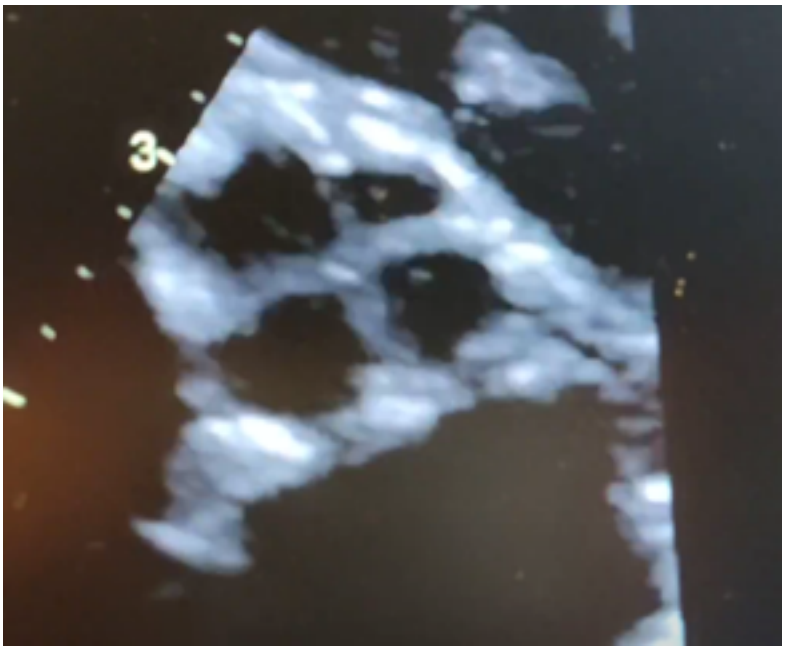
Descriviamo il caso di una neonata nata a termine da parto eseguito con ventosa, gravidanza normodecorsa senza alcuna segnalazione di anomalie. Ci viene richiesta la programmazione di un ecocardiogramma per la presenza di familiarità per cardiomiopatia dilatativa primitiva da parte materna. La neonata ha un esame obiettivo negativo.
All'ecocardiogramma riscontro di valvola aorta quadricuspide con conformazione ad X, cuspidi simmetriche di pressocchè ugual misura ("tipo a" secondo classificazione di Hurwitz e Roberts) , continente, normali flussi transvavolari, normali i diametri aortici. Non altre anomalie congenite associate, non altre anomalie valvolari.Normali dimensioni e funzione biventricolari.
ECG nei limiti di norma per età.
Normale crescita.
E' stato programmato un primo controllo ecocardiografico a breve distanza dal primo che ha mostrato quadro invariato. La piccola cresce bene. Non sono segnalate problematiche cliniche attive di alcun genere.
Abbiamo dunque impostatato follow up ecocardiografico a distanza.

La valvola aortica quadricuspide è una rara anomalia cardiaca congenita spesso diagnosticata accidentalmente. Si stima un'incidenza compresa tra lo 0.008% e lo 0.033% su serie autoptiche e tra lo 0.013% e lo 0.043% su serie ecocardiografiche.
Tale anomalia sembra avere una lieve prevalenza nel sesso maschile.
Secondo la classificazione di Hurwitz e Roberts del 1973 si distinguono sette varianti anatomiche di valvola aortica quadricuspide, vengono indicate con le lettere dalla "a" alla "g" in base alle dimensioni relative delle cuspidi. Si va dalla valvola con quattro cuspidi di uguali dimensioni (tipo a) alla valvola con quattro cuspidi completamente diverse l'una dall'altra (tipo g). Il tipo più comune è il tipo a; di frequente riscontro è anche il tipo b, con tre cuspidi uguali e una piccola cuspide accessoria.
L'aorta quadricuspide è prevalentemente un difetto congenito isolato.
In letteratura la storia naturale di tale anomalia prevede l'evoluzione verso l'insufficienza valvolare, che rimane l'anomalia emodinamica predominante. E' descritta inoltre necessità verso la quinta-sesta decade di vita di intervento cardiochirurgico di sostituzione valvolare legata sempre all'insufficienza. La frequenza della comparsa di disfunzione valvolare non sembra essere correlata con la morfologia della valvola. L'evoluzione verso la stenosi valvolare aortica è più rara.
In letteratura è riportato che l'aorta bicuspide e l'arta quadricuspide presentano prevalenze simili in termini di valvulopatia ed artopatia sebbene per l'aorta bicuspide si presentino ad un'età minore.
La valvola aortica quadricuspide può costituire un substrato per l'endocardite, indipendentemente dalla sua morfologia.
Vista la rarità di tale anomalia è difficile stabilirne l'andamento clinico e quindi la gestione.
Per il caso descritto abbiamo optato attualmente per un controllo ecocardiografico annuale.



EP55

ISOLATED PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT: CARDIAC SURGERY OR WATCH AND WAIT STRATEGY?

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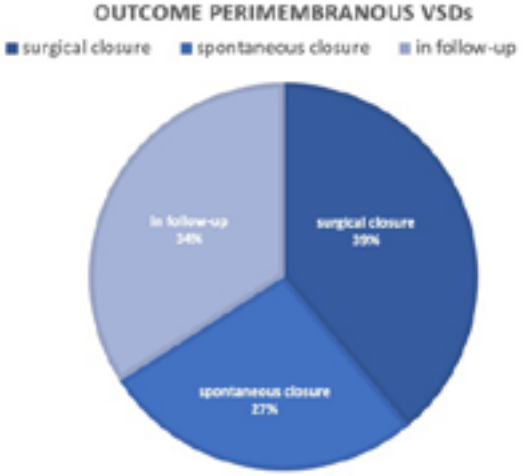
Introduction: Isolated ventricular septal defects (VSDs) are the most common congenital cardiac anomaly in children.
The most frequent type of isolated VSD is muscular VSD whereas perimembranous ones are the least frequent.
The aim of our study is to carry out an in-depth evaluation of the isolated perimembranous VSD and analyze its evolution through time in order to identify the best follow-up strategy and predictable outcomes.

Methods: We searched our database containing 596 patients diagnosed with isolated VSD in the Pediatric Cardiology Clinic in the period between 1995 and 2020. Among these patients we selected 185 of them being diagnosed with isolated perimembranous VSD within the first 60 days of life.

Results: 39% of patients (n=72) underwent surgical closure (average age of procedure 346 days old) with most cases being large defects (n=51). 27% of patients (n=50) experienced spontaneous closure of the defect (average age of closure 838 days old) which occurred more frequently in presence of small defects. Remaining 34% of patients (n=63) were followed up on outpatient basis.
The mean follow-up length for all included patients was 3.9 years.
28% of overall patients (n=52) required anti-decompensation therapy which was used more frequently in patients with medium and large defects.
Of these 52 patients, 54% (n=28) were treated with cardiac surgery whereas the remaining 46% (n=24) were managed conservatively and placed on follow-up according to a watchful waiting strategy. Notably, 18% (n=6) of conservatively managed patients experienced spontaneous closure of VSD.
Among patients treated surgically (n=72), 2.7% of cases (n=2) developed atrioventricular block which required the placement of a permanent pacemaker and 26.4% (n=19) developed a residual shunt.

Conclusions: Perimembranous VSDs have a heterogeneous outcome as shown in our cohort of study.
We identify defect size as a possible outcome predictor since small defects were less associated with requirement for anti-decompensation therapy and were more prone to spontaneous closure (overall spontaneous closure in 50 patients whose 52% had small size defects, 44% had medium size defects and 4% had large size defects). Furthermore, of all patients who underwent surgery, 70.8% had a large defect.
From our registry it is deductible that spontaneous closure of isolated perimembranous VSD tends to occur around the average age of 2.3 years of life (later compared to average age of 0.9 years of life when surgical closure is generally performed).
Moreover, among patients who presented decompensated (n=52), watchful waiting strategy was adopted for 46% cases (n= 24) with a favorable observed outcome. Notably, among these patients managed conservatively, 6 of them experienced spontaneous closure of the defect and the remaining ones, despite the unresolved defect, did not report any complications such as aortic insufficiency or endocarditis in the follow-up period.
Conservative strategy thus seems feasible and safe especially in selected patients who are not in advanced heart failure or who don't have large defects. In these types of patients, cardiac surgery approach can be spared, but it has still to be considered for patients presenting with advanced heart failure or bearing large size defect which are unlikely to experience spontaneous resolution.



CN1

L'INFERMIERE IN CARDIOLOGIA PEDIATRICA: L'ARTE DELLA CURA PEDIATRICA INFERMIERISTICA

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L'arte della cura significa sviluppo della sensibilità della creatività in ambito sanitario della conoscenza di sé, del riconoscimento del altro e della dignità umana nell'ambito della condotta professionale, dell'interazione infermiere- persona e della spiritualità. L'obiettivo come infermiere del reparto de cardiologia pediatrica è quello di verificare se i tirocinanti infermieristici applicano l'arte della cura infermieristica pediatrica, attraverso non solo delle procedure di routine ma anche dell'approccio umano sia verso il paziente pediatrico e i loro familiari.
Oltre a verificare l'applicazione della cura infermieristica pediatrica, l'obiettivo è quello di trasmettere ai tirocinanti l'esperienza acquisita per poter fronteggiare le diverse problematiche che si riscontrano in ogni paziente pediatrico, dando a loro la possibilità di crescere a livello professionale e umano.
Come strumento di valutazione farò un questionario per valutare il grado di conoscenza acquisita e le loro difficoltà.
Questo questionario verrà applicato in forma anonima ad ogni tirocinante.
Alla fine di questa esperienza spero di poter contribuire alla crescita della figura infermieristica pediatrica rendendo più consapevoli le nuove generazioni del ruolo importante che andranno a svolgere.

CN2

ASSISTENZA INFERMIERISTICA AL PAZIENTE PEDIATRICO SOTTOPOSTO A CHIUSURA DILAZIONATA DELLO STERNO: ANALISI RETROSPETTIVA MONOCENTRICA ED ELABORAZIONE DI UNA PROCEDURA

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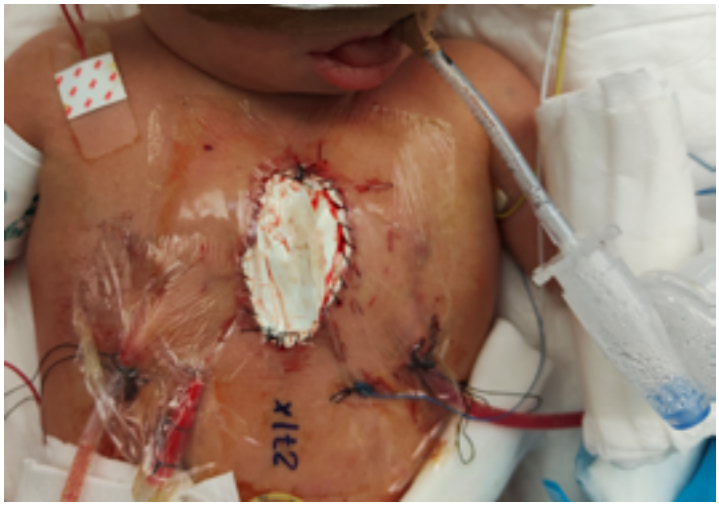
Introduzione: La chiusura dilazionata dello sterno (Delayed Sternal Closure, DSC) rappresenta una delle opzioni terapeutiche per la gestione postoperatoria del paziente pediatrico sottoposto ad intervento cardiochirurgico di correzione di cardiopatia congenita.
Questa tecnica consiste nel mantenere il torace aperto anche nel postoperatorio, coprendo il cuore e i grandi vasi con una membrana in PTFE (politetrafluoroetilene). Questa membrana viene coperta con un film in poliuretano che permette di vedere al di sotto di essa.

Obiettivi:
- Analizzare l'esperienza clinica del centro oggetto dello studio per evidenziare la frequenza di presentazione della DSC;
- Elaborare una procedura di presa in carico di questa tipologia di pazienti

Materiali e Metodi: Nel centro di cardiochirurgia pediatrica dell'azienda ospedale-università di Padova è stato formato un GLAM (gruppo di lavoro aziendale multiprofessionale) che:
- Ha eseguito ricerca bibliografica utilizzando le parole chiave Delayed stenal closure, open chest, thoracic surgery, critical care nursing, congenital heart defects. Ha poi eseguito un'analisi retrospettiva monocentrica osservazionale prendendo in considerazione tutti i pazienti pediatrici operati tra gennaio 2018 - dicembre 2020 che hanno richiesto la tecnica di DSC
- Sulla base dell'entità del problema ha poi elaborato una procedura di accoglimento in Terapia Intensiva nel postoperatorio di questa tipologia di pazienti

Risultati: dei 535 pazienti operati nel periodo di osservazione, 51 (9%) hanno richiesto la tecnica di DSC. La patologia più frequente che ha richiesto questa tecnica è rappresentata dalla trasposizione completa delle Grandi Arterie (D-TGA) (12 pazienti, 23%), seguita dalla Tetralogia di Fallot (ToF) (9 pazienti, 18%). L'età media al momento dell'intervento è stata di 1,69 giorni, con 40 pazienti (78%) che avevano meno di 12 mesi di vita. Il tempo medio di apertura del torace è stato di 3.75 giorni (1.00-28.00). 14 pazienti (27%) hanno richiesto il concomitante utilizzo di ECMO; 3 pazienti (6%) hanno presentato complicanze correlate alla DSC, tra cui le principali sono state il sanguinamento dalla ferita mediastinica e la deiscenza superficiale della stessa. Entro il ricovero si sono verificati 9 decessi (18%).
Dopo questa prima analisi, la procedura è stata elaborata partendo dai documenti già presenti all'interno dell'UO (accoglimento postoperatorio del paziente pediatrico) e tarandola per il paziente con DSC, fornendo alcune specifiche attività e note tecniche su come gestire questi pazienti.

Conclusioni: La DSC è una tecnica sicura ed efficace per la stabilizzazione postoperatoria del paziente pediatrico sottoposto a correzione cardiochirurgica di cardiopatia congenita.
La sua frequenza nel centro analizzato è del 9%, quindi in una percentuale esigua ma molto complessa nella gestione. Poiché il paziente con DSC richiede articolari attenzioni che devono far parte dell'assistenza infermieristica, la proposta di standardizzare l'accoglimento di questa tipologia di paziente permette a tutto il personale, anche il meno esperto, di sapere quali attenzioni prestare in questi particolari casi



CN3

VALUTAZIONE DELL'EFFICACIA DI UN INTERVENTO EDUCATIVO SULLE RISPOSTE EMOTIVE ALL'INDUZIONE DELL'ANESTESIA GENERALE NEI BAMBINI SOTTOPOSTI A PROCEDURE DI EMODINAMICA

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Introduzione: L'ansia preoperatoria è spesso vissuta dai bambini che devono essere sottoposti a procedure di Emodinamica in anestesia generale, ed è associata ad un numero significativo di esiti negativi come un aumentato fabbisogno postoperatorio di farmaci analgesici e sedativi o risvegli dall'anestesia più agitati. Spesso la causa maggiore dell'ansia preoperatoria è una disinformazione da parte dei bambini e dei genitori su come si svolgerà l'induzione dell'anestesia.

Obiettivo: Lo scopo di questo studio è verificare l'efficacia di un intervento educativo rivolto ai bambini e ai genitori che devono essere sottoposti a anestesia generale per migliorare le reazioni emotive associate ad essa.

Metodi: Sono stati arruolati 84 pazienti, affetti da cardiopatia congenita, sottoposti a procedure di Emodinamica elettive in anestesia generale. I pazienti sono stati randomizzati in 2 gruppi: il gruppo intervento che ha ricevuto un intervento educativo da parte del personale infermieristico della sala operatoria e il gruppo controllo che ha ricevuto l'educazione standard. Le reazioni emotive sono state misurate attraverso la Children's Emotional Manifestation Scale (CEMS) e la 4-point Behavior Scale.

Risultati: i pazienti del gruppo intervento hanno mostrato delle reazioni emotive migliori all'induzione dell'anestesia generale rispetto al gruppo controllo. L'intervento educativo ha avuto lo stesso effetto sia che i bambini abbiano già vissuto altre anestesie sia che non ne abbiano mai sperimentato una.

Discussione: l'implementazione di un intervento educativo, effettuato il giorno prima della procedura attraverso la simulazione e la manipolazione dei presidi che verranno utilizzati, migliora le reazioni emotive dei bambini affetti da cardiopatia congenita che devono essere sottoposti a anestesia generale.

Venerdì 28 ottobre 2022
08.30•10.30
COMUNICAZIONI NURSING

CN4

APPROCCI ALL'APPORTO CALORICO NEL PERI-OPERATORIO ED ESITI ASSOCIATI ALLA NUTRIZIONE IN NEONATI CON CARDIOPATIA CONGENITA

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Introduzione e Scopo della ricerca: Le cardiopatie congenite (CHD) sono tra le più diffuse anomalie congenite, con un'incidenza di circa lo 0,8- 1% dei nati vivi. Alcuni neonati necessitano del trattamento con prostaglandine per il mantenimento della pervietà del dotto arterioso, indispensabile per la sopravvivenza, in attesa dell'intervento cardiochirurgico correttivo o palliativo. Tra i fattori determinanti il successo della terapia chirurgica, rientra l'accrescimento ponderale del neonato. Tuttavia, in letteratura non sono riportati approcci condivisi e standardizzati circa l'utilizzo della nutrizione enterale in questi neonati. Scopo della ricerca e? di descrivere, attraverso una revisione narrativa della letteratura, gli approcci più diffusi all'apporto calorico nel peri-operatorio e gli esiti associati alla nutrizione in neonati con cardiopatia congenita in trattamento con prostaglandine.

Metodi impiegati: E' stata condotta una revisione della letteratura utilizzando le banche dati Pubmed e CINAHL. Sono state elaborate stringhe di ricerca ad hoc per le due banche dati, utilizzando parole chiave e termini MeSH. La ricerca, effettuata nel mese di gennaio 2022, ha portato alla selezione di 9 articoli per la revisione.

Risultati e Conclusioni: Sono stati inclusi 7 studi osservazionali, uno studio controllato randomizzato e una revisione narrativa della letteratura. In generale, gli studi si ponevano l'obiettivo di descrivere le pratiche di nutrizione nel peri-operatorio e valutare l'insorgenza e di complicanze potenzialmente associate alla nutrizione. Si riscontra notevole variabilità e scarsa coerenza nelle pratiche nutrizionali, in particolare rispetto all'esistenza di protocolli condivisi e all'adozione della nutrizione precoce. La considerazione della nutrizione enterale quale pratica sicura risulta essere disomogenea tra le varie realtà internazionali. Tuttavia, sembra che i neonati che vengono nutriti precocemente non presentino un maggiore rischio di sviluppare complicanze e abbiano un migliore decorso post-operatorio rispetto a quelli non nutriti (p= 0.002).
Alcuni degli approcci più diffusi potrebbero risultare non efficaci e implicare un maggiore impiego di risorse umane ed economiche, incidendo sugli esiti dei pazienti e sui costi per il Sistema Sanitario. Si rende necessario un consenso da parte dei professionisti sanitari, basato su prove di efficacia per la condivisione di definizioni chiare e protocolli standardizzati.

CN5

SOPHIA OBSERVATION WITHDRAWAL SYMPTOMS - PEDIATRIC DELIRIUM: STUDIO OSSERVAZIONALE NEI PAZIENTI AFFETTI DA CARDIOPATIA CONGENITA POST TIP"

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Introduzione: I bambini sottoposti a intervento cardiochirugico ed esposti per un tempo prolungato a terapia analgosedativa, possono sviluppare crisi d'astinenza, delirio o alterazioni dello stato di coscienza.
L'aumento del tasso di incidenza di tali disturbi nei bambini critici è motivo di preoccupazione, e il mancato riconoscimento o diagnosi comporta conseguenze gravi sul bambino, come l'aumento delle complicanze post-operatorie, l'allungamento del periodo di degenza e l'aumento della morbilità e mortalità. Nasce, per questo, l'esigenza di valutare i segni e i sintomi mediante una scala in grado di misurare contemporaneamente sia il delirio che le crisi d'astinenza.

Materiali: La Sophia Observation Withdrawal Symptoms – Pediatric Delirium (SOS-PD) è uno strumento scientificamente validato, affidabile e clinicamente pratico per un iniziale inquadramento e per le successive misurazioni della severità dei sintomi delle crisi d'astinenza e/o delirio nei bambini critici. È applicabile a tutti i bambini di età compresa tra 0 e 16 anni in terapia con benzodiazepine, oppiacei o sedativi in infusione continua e/o intermittente per un periodo di tempo variabile.

Metodi: È stato condotto uno studio osservazionale presso un reparto di Degenza Pediatrica nei mesi di Luglio-Settembre , durante il quale è stata somministrata la scala SOS-PD a tutti i bambini di età inferiore a 16 anni che hanno ricevuto in Sala Operatoria e in Terapia Intensiva farmaci sedativi e analgesici oppioidi. I bambini osservati sono stati valutati entro 2 ore dal trasferimento in Degenza Pediatrica e almeno due volte nell'arco delle 24 ore, se il punteggio totale ottenuto dall'osservazione di sintomi è uguale o maggiore di 4, il bambino è positivo e deve essere valutato fino a quando non risulta in due rilevazioni consecutive negativo, punteggio minore di 4.

Risultati: A seguito dell'utilizzo della scala SOS-PD è emerso che, crisi d'astinenza e delirio sono manifestazioni frequenti nei pazienti pediatrici ai quali sono stati somministrati farmaci analgesici oppioidi e sedativi. Infatti, su 28 pazienti presi in esame, 7 pazienti sono risultati positivi sia a crisi d'astinenza che delirio, 3 pazienti sono risultati positivi esclusivamente a delirio, 1 paziente è risultato positivo solo a crisi d'astinenza e 17 pazienti sono risultati negativi sia a crisi d'astinenza che delirio.

Conclusioni: Dall'osservazione si rende necessario implementare lo strumento SOS-PD come scala di valutazione delle problematiche post sedazione e analgesia come nel caso riscontrato, per consolidare il campione ed analizzare e validare i dati raccolti; integrando alla scala, anche, un protocollo condiviso dal team multidisciplinare per il trattamento specifico e standardizzato di queste manifestazioni, in modo da garantire la salute del bambino e prevenire l'insorgenza di complicanze. Lo studio osservazionale, pur con i limiti legati al campione, ha messo in luce che, la scala SOS-PD è uno strumento di valutazione affidabile e valido per lo screening precoce di crisi d'astinenza e delirio, anche, nei bambini cardiopatici.

CN6

LA COMPLESSITÀ ASSISTENZIALE IN UN HUB COVID PEDIATRICO: STUDIO OSSERVAZIONALE RETROSPETTIVO

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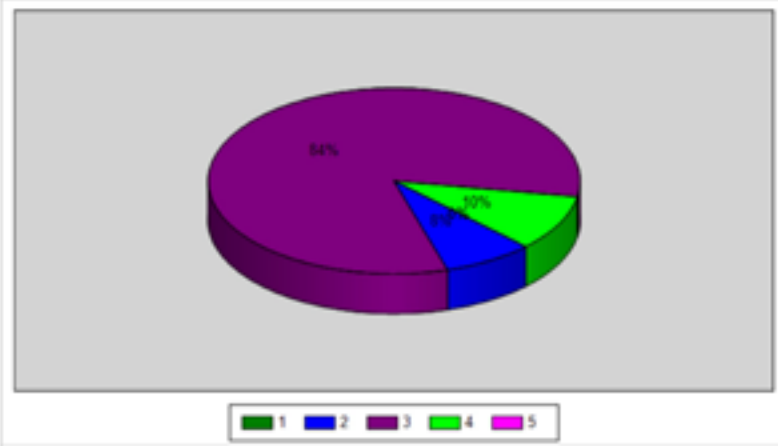
La diffusione del COVID-19 ha portato alla definizione di un nuovo profilo paziente con una propria Complessità Assistenziale (CA), che è utile ed importante da misurare per migliorare l'efficacia e l'efficienza del sistema.
Il presente lavoro si pone l'obiettivo di quantificare la CA dei pazienti pediatrici positivi al SARS-CoV-2 ricoverati presso un Hub COVID Pediatrico.
È stato condotto uno studio di tipo osservazionale retrospettivo. I Bisogni Assistenziali Infermieristici (BAI) dei pazienti oggetto di studio sono stati rilevati attraverso la consultazione delle cartelle cliniche. Successivamente, i dati raccolti sono stati elaborati ed inseriti all'interno del software ICAcode, che ha consentito la determinazione degli Indici di Complessità Assistenziale (ICA).
Il campione è composto di 66 pazienti, di cui 40 di sesso maschile e 26 di sesso femminile, con un'età media di 5,3 anni. Una parte consistente del campione si è attestato nelle Classi di Gravità (CdG) 3 (84%), mentre si è riscontrato un 8% nella CdG 2 e un 10% nella CdG 4. La degenza media nel reparto è stata di 7,5 giorno. Gli interventi più onerosi, in base al fattore tempo, sono stati il monitoraggio dei parametri vitali, dell'alimentazione e dello stato idrico, l'esecuzione di prelievi ematici, la somministrazione di farmaci e l'educazione terapeutica del caregiver, nonché il monitoraggio dell'ambiente e il mantenimento dell'isolamento, ma soprattutto il mantenimento della comunicazione e dell'interazione con il paziente e il caregiver.
Questo lavoro ha evidenziato, seppur con alcuni limiti, la possibilità di generare informazioni e dati utili nel contesto clinico-assistenziale e organizzativo, definendo la CA della popolazione pediatrica affetta da COVID-19 ed evidenziandone i BAI.

Figura 1 - Complessità assistenziale per gli interventi del Dizionario Attività.

Figura 2 - Distribuzione delle classi di gravità dei pazienti da ICAcode.



CN7

DALLA TEORIA ALLA PRATICA. PIANIFICARE L'ASSISTENZA INFERMIERISTICA CON ICNP™ IN UNA DEGENZA PEDIATRICA CARDIOCHIRURGICA

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Introduzione: La Fondazione Monasterio lavora dal 2012 all'informatizzazione della cartella clinica integrata. Al progetto di digitalizzazione lavora un team multidisciplinare composto da coordinatori infermieristici, infermieri, medici, personale sanitario, farmacisti ed informatici. Negli anni il team ha lavorato alla raccolta dati, alla valutazione delle buone pratiche e degli esiti infermieristici, alla comunicazione tra operatori sanitari per assicurare la continuità di cura, alla riduzione degli errori di prescrizione, registrazione di informazioni più precise, chiare, complete ed accessibili. Lo sviluppo si è reso necessario sulla pianificazione infermieristica puntando al linguaggio International Classification for Nursing Practice™, anche in ambito pediatrico. L' ICNP™ è una terminologia standardizzata per documentare la pratica infermieristica, inclusa tra i linguaggi riconosciuti dall'Organizzazione Mondiale della Sanità. E' un prodotto dell'International Council of Nurses (ICN), distribuito da SNOMED International, in seguito all'accordo stipulato nell'agosto 2020. La pianificazione assistenziale avviene attraverso l'enunciazione di diagnosi infermieristiche, interventi e outcome, documentando ciò che si è fatto e l'eventuale risoluzione del problema, permettendoci di seguire con precisione il percorso assistenziale del bambino e la sua famiglia, valutandone l'efficacia.

Materiali e Metodi: Il gruppo di lavoro aziendale ha immaginato l'implementazione del linguaggio ICNP™ all'interno della cartella infermieristica informatizzata, attreverso incontri tra tutti i professionisti coinvolti, confrontandosi sulle singole parti sviluppate/da sviluppare, condividendo i progressi e le nuove criticità..

Risultati: Sono state analizzate le scale di valutazione previste dalla procedura aziendale - "Valutazione Multidimensionale" - compilate dagli infermieri in ingresso, durante la degenza e alla dimissione (Humpty Dumpty; Retos; Glamorgan; Barthel, Braden; Pyms), analizzati i vari item e da questi estratte le diagnosi infermieristiche, gli interventi e gli outcomes di interesse. Sono stati valutati i bundle presenti nelle schede di valutazione dei presidi / device, essenziali al fine del contenimento del rischio infettivo, e anche per questi strutturato il percorso secondo ICNP™, così come per i singoli bisogni assistenziali.
Sono stati scelti degli outcome assistenziali di interesse con una dashboard di monitoraggio.

Conclusioni: Tale percorso è stato seguito al fine di pianificare percorsi assistenziali personalizzati sicuri e di qualità, seguendo il modello assistenziale ICNP™. Il gruppo di lavoro ha raggiunto ad oggi alcuni degli obiettivi programmati, ed è in corso la divulgazione del piano al gruppo degli informatici per lo sviluppo e la strutturazione del processo. Da lavorare ancora sulla condivisione del linguaggio e la formazione per tutto il personale.

CN8

LA PREVENZIONE DEL CAVO ORALE NEL BAMBINO CON CARDIOPATIA CONGENITA: L'INFERMIERE SPECIALISTA CLINICO

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Premessa: Alcuni studi, effettuati in tutto il mondo, provano che i pazienti con cardiopatie congenite sono predisposti ad una scarsa igiene orale, da cui l'insorgenza di gengiviti e carie.
Sovente troviamo questi pazienti in sala operatoria per bonifiche dentarie da eseguire in narcosi per la gravità con cui giungono alla nostra attenzione.

Presso l'ospedale pediatrico Bambino Gesù, nel 2021 sono stati trattati 21 casi di bambini con anomalie cardiache, in sala operatoria, per eseguire cure odontoiatriche in anestesia generale. 16 di questi per eseguire cure di carie ed estrazioni, 8 anche ablazione del tartaro. Numero che è raddoppiato rispetto agli anni precedenti: 9 casi nel 2020, 11 casi nel 2019. Un caso del 2020 è rientrato in sala per la stessa procedura a distanza di soli 8 mesi.
Da questo l'importanza di proporre un lavoro per mettere in atto un programma di prevenzione al fine di migliorare la salute orale, e da questa la salute generale dei bambini affetti da cardiopatie.

Materiali e Metodi: In accordo con il dipartimento di cardiologia, si è proposto di analizzare i pazienti cardiopatici al fine di crearne una valutazione attraverso l'indice di placca e di sanguinamento, tramite sondaggio parodontale e indice DMFT.
L'idea suppone di registrare per ogni paziente una cartella parodontale.
L'igiene professionale includerà l'educazione del bambino e dei suoi genitori ad una corretta esecuzione dell'igiene orale domiciliare nonché ad una corretta alimentazione.
Si ha l'ardire di voler eseguire controlli di rivalutazione a 3, 6, 9 e 12 mesi riconsiderando gli indici della prima osservazione.
Inoltre, in occasione dell'arruolamento, sarà somministrato un questionario ai genitori per indagare il grado di conoscenza circa l'importanza delle cure per la salute orala nei pazienti con cardiopatia congenita.

Risultati Attesi: Questo lavoro, oltre a promuovere la prevenzione primaria e secondaria riguardo la salute orale, avrà certo risvolti sulla gestione delle sedute di sala operatoria presso il nostro ospedale, ottimizzandone le liste.
Nell'attuazione di questo programma diventa essenziale la collaborazione con altre figure professionali: oltre il pediatra, l'odontoiatra e il cardiologo, l'igienista dentale e la nutrizionista, per partire dalle basi del problema quali l'educazione alla salute orale ed una corretta alimentazione non cariogenica da unire a quella che già dovrebbe essere in atto per la patologia e relativa terapia in corso.

L'infermiere di pratica avanzata è un professionista che ha sviluppato le proprie competenze professionali in un determinato contesto clinico o organizzativo, attraverso un percorso formativo universitario o di rilievo professionale tale da consentire un più approfondito approccio metodologico ai problemi della persona o della comunità assistita.
Infermiere specialista è l'esperto clinico nell'accertamento e la pianificazione dell'assistenza nell'area di competenza, ha una conoscenza approfondita dei percorsi diagnostico-terapeutici dei propri assistiti, è in grado di sviluppare i propri interventi secondo le più recenti indicazioni dell'evidence-based nursing.

Per giungere a quello che abbiamo formulato essere il nostro scopo, ovvero il miglioramento della salute orale nei pazienti cardiopatici congeniti, la figura dell'infermiere esperto in cardiologia pediatrica, rappresenterà la chiave di unione in questo importante studio multidisciplinare.

CN9

ALLATTAMENTO MATERNO NEI PAZIENTI AFFETTI DA CARDIOPATIA CONGENITA. STUDIO OSSERVAZIONALE RETROSPETTIVO

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Introduzione: I benefici dell'allattamento materno esclusivo sul corretto sviluppo del bambino e sulla prevenzione di numerose patologie sono ampiamente riconosciuti dall'Organizzazione Mondiale della Sanità (OMS), che considera l'allattamento uno degli obiettivi prioritari di salute pubblica a livello mondiale. Stime ufficiali indicano che, nel mondo, solo il 40% dei bambini di età compresa tra 0 e 6 mesi è allattato esclusivamente al seno, e in appena 23 Stati il tasso di allattamento al seno supera il 60%.

Obiettivo dello Studio: Lo studio è finalizzato ad analizzare l'esperienza di allattamento materno; valutare quanto gli interventi di educazione sanitaria influiscono sulla reale esperienza di allattamento, per apportare eventuali migliorie nelle strategie educativo-assistenziali pre e post partum.

Materiali e Metodi: Il questionario utilizzato per lo studio è stato somministrato, attraverso indagine telefonica, ad un campione di 60 madri di neonati cardiopatici. La raccolta dati è avvenuta dal 15 Gennaio 2021 al 15 Febbraio 2021. L'adesione allo studio è su base volontaria.
RISULTATI. L'analisi dei dati ha permesso di attribuire grande importanza e valore all'assistenza, operata dal Team della struttura ospedaliera, in termini di educazione e supporto all'allattamento. Tuttavia, in determinate situazioni, il quadro clinico del neonato e la relativa patologia cardiaca hanno influito sul non ottenimento di un allattamento esclusivo e prolungato nel tempo. È rilevante evidenziare che, una considerevole percentuale di madri, ha allattato da 6 fino a 12 mesi di età del bambino (come suggerito dalle raccomandazioni OMS e UNICEF).

Conclusioni: Una corretta educazione sanitaria all'allattamento al seno e un'assistenza orientata ai bisogni di madre e di bambino rappresentano strumenti fondamentali per ottenere un allattamento efficace. L'importanza dell'attività assistenziale e di supporto nel post partum si riscontra, in particolar modo, in situazioni di neonati patologici, quando le molteplici criticità neonatali possono costituire un ostacolo per l'allattamento.

CN10

STATO D'ANSIA DEI GENITORI NEL PREOPERATORIO

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Contesto: L'intervento cardiochirurgico in età pediatrica costituisce un evento psicologicamente stressante sia per il paziente sia per i genitori, associandosi a fantasie di perdita che determinano elevati livelli di angoscia in entrambi. L'elaborazione psichica del genitore può essere o meno favorita dallo stato emotivo che sperimenta all'ingresso del figlio in sala operatoria. La letteratura concorda sul fatto che l'ansia preoperatoria del genitore debba essere gestita dal personale sanitario poiché costituisce un fattore di rischio per l'insorgenza di problematiche emotive e comportamentali al risveglio in terapia intensiva che possono avere un impatto sulla gestione clinica del paziente. L'ansia sperimentata al momento della separazione può avere infatti delle conseguenze sulle modalità di interazione con il figlio, sul tipo di accudimento che instaurerà con lui e sulla compliance con il personale medico-infermieristico. Essendo dimostrato che il culmine dell'ansia preoperatoria è raggiunto al momento della separazione, vissuta da entrambi come un abbandono e, pertanto, con emozioni di paura, angoscia, frustrazione e, nel genitore, anche di colpa, nel nostro centro, per agevolare il distacco nel presala operatoria, è presente lo psicologo e, quando possibile, l'anestesista somministra la preanestesia in presenza dei genitori.

Obiettivo: Scopo della ricerca è rilevare se il distacco dal bambino nel presala operatoria determini nel genitore un'ansia clinicamente significativa e se essa correli con variabili relative all'anagrafica del paziente e del genitore.

Metodi: L'indagine, svolta al momento della separazione nel presala operatoria, prevede il colloquio psicologico, l'osservazione clinica, la somministrazione al genitore di un questionario che indaga i dati anagrafici, il livello di ansia auto-percepita e la percezione dello stato di coscienza del bambino a seguito della preanestesia, e la somministrazione dello State-Trate Anxiety Inventory Y-1 per l'autovalutazione dell'ansia di stato.

Riflessioni e Conclusioni: Lo studio ha coinvolto un campione di n°57 genitori (n°29 madri n°28 padri) di bambini con età compresa tra 1 giorno e 13 anni, dei quali n°14 al primo intervento cardiochirurgico e n°15 a intervento successivo.
La raccolta dati si è focalizzata sul momento della separazione dal figlio e ha incluso, oltre all'anagrafica del paziente e alle informazioni relative all'anamnesi, età dei genitori, grado di istruzione, presenza di altri figli, numero di interventi cardiochirurgici effettuati, percezione dello stato di coscienza del figlio, valutazione dell'ansia auto-percepita mediante una Visual Analogic Scale e il questionario self report STAI Y-1.
Dall'analisi statistica è emerso che n° 57 genitori hanno sperimentato un'ansia di stato clinicamente significativa che nelle madri è risultata correlare con l'ansia auto-percepita, l'età del bambino, il numero di interventi cardiochirurgici effettuati e lo stato di coscienza percepito nel figlio, mentre nei padri è risultata correlare soltanto con l'ansia auto-percepita.
I dati raccolti fanno emergere delle riflessioni rispetto allo stato emotivo del genitore e alla necessità di fornire anche nel presala operatoria un adeguato supporto psicologico. Si rivela fondamentale individuare precocemente e tempestivamente possibili fattori di rischio nel sistema familiare e supportare in maniera continuativa le coppie che sperimentano un elevato livello di ansia con l'obiettivo di ridurne l'impatto sul figlio nell'immediato postoperatorio, favorire in loro una risposta adeguata alle necessità di cura e agevolare l'outcome del paziente.

CN11

PAZIENTE AFFETTO DA SINDROME DA ANNEGAMENTO

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Paziente femmina di 2 anni di età, giunta presso l'U.O di terapia intensiva pediatrica post sindrome da annegamento e dopo primo soccorso da parte del 118. Condizioni critiche. Allertato ECMO team che non ha ritenuto necessario posizionare supporto cardio-polmonare.
Paziente in stato severo di ipotermia, ipossia ed acidosi metabolica. Gestita fin da subito con monitoraggio continuo dei parametri vitali ed elettroencefalogramma continuo, ventilazione meccanica e sedazione.
Dopo un lento riscaldamento, iniziatosi lento risveglio e svezzamento dalla ventilazione meccanica efficace. Dopo un ricovero di 4 giorni in terapia intensiva e di 4 giorni nel reparto di degenza pediatrica non si riscontra nessun segno di danno ipossico cerebrale e si riscontra una ripresa totale delle funzioni vitali.
In questo percorso la rete tempo dipendente è stata efficace ed efficiente, dalle prime manovre e dai primi contatti la situazione è apparsa molto critica, ma grazie alla sinergia dei percorsi il risultato è stato molto positivo. Considerando questo risultato e i possibili eventi presso il nostro centro di pazienti pediatrici affetti da sindrome da annegamento, l'obiettivo è la stesura di un percorso diagnostico, terapeutico, assistenziale personalizzato al fine di porre le basi per una gestione efficace e sicura, monitorando il processo dal dispatch telefonico, alla dimissione, includendo gli outcome assistenziali di riferimento durante il ricovero e nel follow up.

CN12

GESTIONE INFERMIERISTICA DELLA DESATURAZIONE E DELL'ALIMENTAZIONE NEI PAZIENTI CON ATRESIA DELLE ARTERIE POLMONARI

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Introduzione: L'atresia delle arterie polmonari è una rara e complessa cardiomiopatia congenita. Una strategia di riabilitazione dei vasi polmonari centrali consiste nella realizzazione di shunt e condotti tra l'aorta o il ventricolo destro e questi vasi centrali al fine di ottenere una dilatazione di questi vasi per favorire la ricostruzione di una vascolarizzazione polmonare.
Tale patologia comporta grave desaturazione per ipoafflusso delle arterie polmonari con cianosi periferica e difficoltà nell'alimentazione dei pazienti pediatrici.

Obiettivo: L'obiettivo di tale ricerca mira a comprendere e gestire le complicanze dovute alla desaturazione, il ricorso ad un'ossigeno-terapia efficace.
Importante per la riabilitazione interventistica è l'aumento di peso dei neonati che spesso non hanno un'alimentazione idonea. Per tale motivo è necessario ricorrere a presidi per l'alimentazione artificiale come SNG o PEG.

Materiali e Metodi: Si effettuerà una revisione sistematica delle ricerche inerenti tale cardiomiopatia ricavate dalle banche dati ricercando le strategie riabilitative e interventistiche, la gestione dei pazienti riguardo la desaturazione e l'alimentazione.

Risultati: Gestire al meglio la desaturazione nei neonati che spesso presentano grave cianosi periferica ricorrendo ad ossigenoterapia; cercare di calmare il neonato comprendendo i motivi del pianto che provoca l'insorgenza di cianosi periferica in quanto durante le crisi insorge desaturazione marcata; cercare di garantire un'alimentazione sufficiente per l'accrescimento in modo da permettere di procedere con la riabilitazione.

Conclusioni: I pazienti affetti da Atresia delle arterie polmonari, presentando grave cianosi periferica e desaturazione marcata, nonostante l'ossigeno-terapia nei momenti di pianto, provocano preoccupazione durante l'assistenza. Importante è riconoscere i motivi di tale desaturazione e attuare tutte le strategie possibili per aumentare la sensazione di comfort dei neonati in modo da far cessare il pianto e di conseguenza migliorare la cianosi e i livelli di SpO2.
Riconoscere l'importanza e comprendere i presidi migliori a cui fare ricorso in caso di difficoltà nell'alimentazione.

CN13

I BENEFICI DERIVANTI DALLA TERAPIA DEL DOLORE DURANTE LA RIMOZIONE DI DRENAGGI TORACICI NEI PAZIENTI PEDIATRICI, SOTTOPOSTI AD INTERVENTO CARDIOCHIRURGICO

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Introduzione: ogni bambino sottoposto ad intervento di cardiochirurgia sarà provvisto di almeno un drenaggio toracico, che potrà essere di locazione pleurica o mediastinica. Già la presenza stessa del presidio provoca dolore, portando ad esempio difficoltà al paziente nell'espandere correttamente la cassa toracica durante la respirazione. Questi poi vengono rimossi nelle prime giornate post operatorie non appena le perdite risultano esigue, sempre previa rx del torace.

Obiettivo: l'obiettivo di tale ricerca è incentrato nella rimozione di tali presidi: spesso la pratica del quotidiano, porta alla rimozione dei drenaggi senza l'utilizzo di terapia sedativa/antidolorifica sufficiente ad annullare completamente la pratica agli occhi del paziente, ritenendo che la somministrazione porterebbe ad una inefficace risposta di espansione polmonare. Scontato dire che tale manovra, anche se rapida, è inevitabilmente dolorosa ed è stato notato come venga vista nel paziente come la pratica di cui aver più paura. Motivo per il quale in questa ricerca ci si va ad incentrare sul beneficio sia fisico che psicologico che potrebbe portare una adeguata terapia pre-rimozione drenaggi toracici.

Materiali e Metodi: si effettuerà una revisione sistematica delle ricerche inerenti all'uso di terapie estemporanee per controllare il dolore ricavate dalle banche dati. Verranno inoltre raccolti dati empirici, in accordo con i cardiochirurghi della nostra unità operativa.

Risultati: non far provare alcun dolore al paziente e ridurre al minimo il trauma psicologico, in particolare per pazienti come questi che sono, per la loro condizione, spesso costretti ad essere sottoposti ad altri interventi cardiochirurgici, permettendo di affrontare il periodo post operatorio più serenamente.

Conclusioni: la rimozione dei drenaggi è sempre un momento abbastanza sentito, sia per la sua importanza clinica che emotiva. E' importante che la gestione e rimozione di tale presidio possa avvenire nella più totale sicurezza e serenità del paziente, il quale in seguito dovrà ancora affrontare tutta la restante riabilitazione prima della dimissione.

CN14

INTEGRAZIONE DELLE CURE PALLIATIVE IN CARDIOLOGIA PEDIATRICA

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Introduzione: Le cure palliative pediatriche vengono definite dall'OMS come l'attiva presa in carico"globale" di un corpo, mente e spirito di un bambino e della sua famiglia. L'integrazione delle cure palliative per un bambino richiede un piano assistenziale personalizzato, sviluppato sulla base di obiettivi che migliorino la qualità della vita del paziente e della sua famiglia , tali obiettivi vanno realizzati in collaborazione con quest'ultimi. Date le crescenti comorbilità, molti pazienti trarrebbero beneficio dal coinvolgimento delle cure palliative lungo la traiettoria della malattia, collegamento attuabile con la terapia intensiva cardiologica.

Obiettivo: L'obiettivo delle cure palliative pediatriche è fornire supporto e ridurre la sofferenza ai bambini con malattie gravi e alle loro famiglie, attuando nel miglior modo possibile il care del bambino, al fine di ridurne il disagio e ottimizzare la compliance che indirettamente migliora tutti i parametri vitali. Questi obiettivi sono raggiunti attraverso una valutazione interdisciplinare esperta e attraverso la gestione dei sintomi fisici e psicologici, fornendo una comunicazione di alta qualità per facilitare il processo decisionale e la pianificazione anticipata delle cure.

Materiali e Metodi: prendendo in considerazione un caso clinico presentatosi presso la nostra unità operativa della cardiologia cardiochirurgia pediatrica, si vuole descrivere come è avvenuto il collegamento con la rete delle cure palliative pediatriche domiciliari e come questa è organizzata, la relazione con la famiglia e l'educazione terapeutica fornita dagli infermieri di reparto.

Risultati: Sicuramente il coinvolgimento della famiglia e l'educazione terapeutica fornita dai professionisti garantisce una migliore qualità nella gestione della malattia del bambino, identificando tutti i loro bisogni.

Conclusioni: Il ponte tra la terapia intensiva cardiologica e le cure palliative non è ancora così immediato. La famiglia partecipa attivamente alla cura ed è chiamata a prendere decisioni spesso difficili. I bisogni della famiglia sono molteplici: educativi-formativi, spirituali, economici e sociali. L'accompagnamento del bambino nella dimensione del domicilio e la prospettiva di un fine vita con un sostegno alla famiglia, aiuta nella gestione del quotidiano.

CN15

IL CASO DI C.: L'EVOLUZIONE DELL'ATTESA

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Riportiamo il caso di un bambino di 15 mesi che, nel corso della sua degenza, nel periodo tra febbraio e maggio 2022, presso la nostra struttura di cardiologia pediatrica, ha sviluppato molteplici problematiche. La diagnosi principale indicava ventricolo destro doppia uscita, trasposizione delle grandi arterie, difetto interatriale, atresia della arteria polmonare e microdelezione del cromosoma 22 con sordità genetica. Nel corso del ricovero il bambino ha subito una prima indagine in emodinamica, con successivo intervento cardiochirurgico di shunt tra arteria anonima e arteria polmonare ed esplorazione del DIV dall'aorta, seguito da un post operatorio complicato da infezione della ferita chirurgica, infezione dell'accesso venoso centrale e numerose crisi per ipovolemia. L'assistenza infermieristica erogata ha coinvolto diversi ambiti, dal supporto emodinamico in emergenza, alle manovre assistenziali di base, con costante supporto alla famiglia, rivelatosi fondamentale viste le restrizioni date dal periodo di emergenza sanitaria.

CN16

THE LESSER THE BETTER: È SEMPRE POSSIBILE FARE BENE CON MENO APPROPRIATEZZA DEI PROTOCOLLI DOSE

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Introduzione: Negli ultimi anni numerosi sforzi sono stati attuati al fine di ridurre al minimo la dose di radiazioni ionizzanti soprattutto nel campo dell'emodinamica pediatrica. Tali sforzi hanno portato al miglioramento sia delle macchine e dei protocolli legati ad esse ma anche ad una migliore consapevolezza e responsabilità del tecnico di radiologia. La maggior parte degli angiografi moderni permette la selezione del pacchetto di esposizione in relazione al peso del paziente, con la possibilità di ridurre il numero p/s che vengono emessi null'unità di tempo.

Scopo dello studio: Scopo dello studio è stato quello di dimostrare l'efficacia dell'utilizzo dei protocolli con esposizione radiologica ridotta nel garantire una adeguata risoluzione dell'immagine per l'operatore.

Materiali e Metodi: Nell'ultimo anno (2020-2021) abbiamo attuato nella nostra emodinamica un protocollo ai pazienti pediatrici affetti da Difetto Interatriale utilizzando il protocollo radiologico più basso possibile (CARD <6Kg) indipendentemente dal peso del paziente coordinandoci con l'operatore, al fine di ridurre al minimo la dose, conservando la qualità dell'immagine ad un livello più che accettabile. Abbiamo quindi confrontato questi dati con i dati relativi al periodo 2018-2019.

Risultati: Abbiamo studiato 206 pazienti trattati tra il 2018 ed il 2021 per chiusura di DIA OS singolo con protesi di Amplatzer e con un peso tra i 20 ed i 35 kg.
Tali pazienti sono stati suddivisi in due gruppi:
gruppo A (2018-2019) dove i parametri di acquisizione venivano selezionati in relazione al peso effettivo del paziente (CARD <20Kg – CARD <40Kg – CARD Adulti), utilizzando una scopia con pulsazioni variabili da 7.5 a 10 p/s;
gruppo B (2020-2021) trattati utilizzando dei protocolli a bassa dose (CARD<6 kg) indipendentemente dal peso dei pazienti.
I due gruppi erano comparabili per età (gr 8.9+/-4,2 versus gr B 8.5+/-5.7; p=0.1) e per peso (gr A 33.5+/-17.4 versus gr B 29+/-15,4; p=0.2). La durata della procedura era 107 +/- 29,14 min nel gr A e 111+/-60 min nel gr B p=0.06).
La dose assorbita nel gr A era significativamente aumentata rispetto al gr B (gruppo A 18.13+/-33 mGy- 491.7+/-1257,7 µGym^2, gruppo B 14+/-24.2 mGy - 214.8 +/- 462.7 µGym^2, p 0.001). Il tempo totale di scopia espresso in minuti era 8.6+/-2.4 min nel gruppo A versus 9,6+/-6,03 min nel gruppo B (p= 0.2).
Infine non vi erano differenze tra i due gruppi in termini di risultati e complianze.

Conclusioni: La strategia di utilizzare parametri di acquisizione predefiniti con basso grado di esposizione (CARD <6Kg), riducendo le pulsazioni a 4p/s, determina una riduzione significativa dell'esposizione radiologica al paziente, mantenendo una buona risoluzione radioscopica. Tale significatività si è mantenuta nonostante l'utilizzo nel 2021 di nuovi dispositivi oltre all' Amplatzer (GORE- Occlutech) che hanno richiesto maggior definizione e l'utilizzo del Biplano.

Venerdì 28 ottobre 2022
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CN17

MONITORARE PER MIGLIORARE

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Presentiamo il database, ideato e introdotto in reparto, finalizzato a monitorare la gestione degli accessi venosi centrali presenti presso la nostra struttura. I dati raccolti e inseriti nel database hanno la finalità di monitorare la corretta gestione e l'eventuale sviluppo di complicanze in relazione a questi presidi.
I criteri per l'inclusione nello studio sono stati i seguenti:
- Ricovero presso uno dei centri di costo presenti in reparto (alta e bassa intensità; pediatria, cardiologia e chirurgia pediatrica; day hospital e day surgery)
- Accesso di tipo venoso centrale (CICC, FICC e PICC)
- Impianto eseguito presso la nostra unità operativa o altre strutture dell'ASST GOM Niguarda, dal 1/03/22 al 30/09/22

I criteri di esclusione sono stati i seguenti:
- Impianto del presidio presso una struttura esterna all'ASST GOM Niguarda

Il campione finora analizzato comprende 17 accessi centrali.
Tra le finalità del database troviamo:
- Identificare se ci sono bambini più a rischio di sviluppare complicanze
- Monitorare l'insorgenza di complicanze a breve e a lungo termine
- Valutare la corretta esecuzione delle medicazioni

Questo lavoro si correla direttamente allo studio effettuato lo scorso anno riguardante la revisione delle indicazioni per la gestione e la medicazione degli accessi centrali. Il database fino a questo momento si sta rivelando un valido strumento, soprattutto, per il monitoraggio sull'esecuzione della medicazione da parte del personale infermieristico; rappresenta inoltre uno stimolo per l'identificazione precoce delle complicanze.

Venerdì 28 ottobre 2022
10.30•12.30
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CN18

L'ASSISTENZA INFERMIERISTICA AL PAZIENTE PEDIATRICO E ADULTO CON CARDIOPATIA CONGENITA TRAPIANTATO DI CUORE

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Introduzione: Il primo trapianto di cuore in un paziente adulto fu eseguito da Christian Barnard presso il Groote Schuur Hospital in Sud Africa il 3 dicembre 1967. Solo tre giorni dopo, il 6 dicembre del 1967 a New York, Adrian Kantrowitz eseguì il primo trapianto di cuore su un neonato di 19 giorni affetto da atresia della tricuspide che visse solo 6 ore.
Secondo i dati attuali riportati dal ISHLT (International Society For Heart and Lung Trasplantation) in tutto il mondo, attualmente, vengono effettuati circa 600 trapianti pediatrici l'anno con una sopravvivenza complessiva del 90% ad 1 anno, del 75% a 5 anni e del 65% a 10 anni. Uno dei maggiori problemi non ancora risolti è rappresentato dalla mortalità dei pazienti pediatrici in lista di attesa per trapianto ortotopico di cuore. Tale condizione è dovuta al fatto che la disponibilità di organi è di gran lunga inferiore al numero di pazienti in lista di attesa.
Le patologie cardiache che, in età pediatrica, richiedono il trapianto di cuore sono diverse a seconda dell'età del paziente. Infatti, mentre per il bambino con età inferiore all'anno vi è una prevalenza di cardiopatie congenite, nelle età successive le principali malattie cardiache responsabili di scompenso cardiaco refrattario alla terapia medica, e che quindi richiedono il trapianto di cuore, sono rappresentate dalle cardiomiopatie dilatative, ipertrofiche e restrittive.
Tutti i pazienti, prima di essere inseriti in lista per un trapianto di cuore, devono essere valutati attentamente al fine di escludere la presenza di patologie in altri apparati, o di un danno d'organo irreversibile, che dopo il trapianto potrebbero compromettere l'outcome del paziente stesso.
Inoltre, un'attenta analisi dello stato psico-sociale del paziente e della famiglia è fondamentale per garantire una perfetta aderenza del ricevente all'iter diagnostico terapeutico dopo il trapianto di cuore.

Obiettivo: L'obiettivo è quello di fornire e garantire, al paziente e alla sua famiglia, un supporto per ciò che concerne l'aspetto clinico-assistenziale, l'aspetto psicologico e quello logistico, oltre che attuare una buona educazione terapeutica, in previsione della dimissione a domicilio. Questi sono obiettivi che si raggiungono attraverso una gestione multidisciplinare, coinvolgendo più figure che collaborano tra di loro.

Risultati: Il coinvolgimento della famiglia e l'educazione terapeutica fornita dai professionisti, garantisce una migliore gestione della malattia del bambino/adulto e una migliore qualità di vita, identificando tutti i propri bisogni.

Conclusioni: Si vogliono evidenziare le attività che l'infermiere deve svolgere, per poter garantire sia un'assistenza infermieristica efficace ed efficiente che il cambiamento di vita che la persona è chiamata ad attuare. Uno degli intenti di questo lavoro è stato di mostrare le responsabilità che l'infermiere ha nei confronti del paziente trapiantato e le attività infermieristiche che è chiamato a svolgere per poter garantire un buon successo dell'intervento. L'elaborato, inoltre, pone l'accento sull'importanza del ruolo infermieristico nell'assistenza del paziente nella fase post-operatoria e sull'autonomia acquisita nello svolgimento delle attività. Si è voluto poi mostrare l'utilizzo del "processo di assistenza infermieristico", un metodo che permette l'individuazione dei problemi del paziente e la pianificazione delle attività che portano alla risoluzione di questi.

CN19

NURSING TRANSCULTURALE: LA NUOVA SFIDA PER LA PROFESSIONE INFERMIERISTICA

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La nuova realtà sempre più multietnica, multinazionale e in continuo movimento in cui viviamo pone nuove sfide all'assistenza infermieristica e a tutte le professioni sanitarie. E' indispensabile quindi acquisire competenze transculturali spendibili nell'esercizio della pratica clinica, organizzativa e formativa all'interno del panorama delle dinamiche di salute in un mondo sempre più interconnesso.
Per acquisire tali competenze è fondamentale conoscere non solo i nuovi scenari del mondo salite ma anche lo studio di alcuni modelli che tengano conto dei vari aspetti che concorrono alla salute degli individui.
Lo scopo del lavoro è la condivisione non solo di alcuni argomenti trattati all'interno del corso effettuato ma anche l'analisi del modello di Purnell e della sua ricaduta sulla cura culturalmente competente nella pratica clinica e organizzativa.

CN20

QUALITY OF LIFE OF FAMILIES WITH CHILDREN WITH CONGENITAL HEART DISEASE": STUDY PROTOCOL

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Congenital heart disease (CHD) represents a heterogeneous group of the most common congenital defects. Advances in medicine have allowed for a notable increase in the survival rates of children born with CHD, even in the most complex cases, which almost mitigate their pathology from being lethal to chronic. The quality of life (QoL) experienced by such children is influenced by that perceived by their parents. However, international literature has rarely considered the entire family nucleus, often focusing its attention on the mother–child relationship. The same literature recommends studies that investigate both parents and the child with CHD over time.
This study aims to analyse the temporal trend of the QoL of children with CHD as well as that of their parents, following the children's hospitalization for an invasive procedure.

Longitudinal study. A sample of families (children with CHD and their parents) were will be enrolled during the patient's discharge from hospitals and examined every three months for one year. The hypothesis adopted by this study is that there is an interdependence between the subjects in the study that is capable of influencing the individual perception of QoL. Over time, this perception of QoL could be vary in relation to the individual perception of the components of the triad of care.

This study want to identify the variables and their temporal trend, which was attributable to the family unit that – together with the physical and clinical ones –could influence the QoL of children with CHD. Studying the QoL of the triad of care with the longitudinal method will allow the identification of the predictors and the interdependence of the QoL of the child and the parents in order to improve the elaboration of guidelines providing better assistance to the patient and the caregivers.

Venerdì 28 ottobre 2022
10.30•12.30
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CN21

UN MODELLO EDUCATIVO DI TRANSIZIONE PER GLI ADOLESCENTI CON CARDIOPATIA CONGENITA: STUDIO TELEMACO. RISULTATI AD INTERIM DI UNO STUDIO RANDOMIZZATO CONTROLLATO E MULTICENTRICO

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Razionale: La transizione dell'adolescente con cardiopatia congenita verso l'età adulta rappresenta un momento di criticità considerando i bisogni fisici e psicologici peculiari dell'età evolutiva. In questo scenario, il modello educativo di "Transition Clinic" (TC) costituisce un elemento cruciale per l'erogazione di interventi educativi standardizzati e di supporto, con approccio multidisciplinare, durante la transizione dell'età pediatrica a quella adulta. Tuttavia, le evidenze a supporto dell'efficacia reale del modello TC nella promozione della percezione dello stato di salute dell'adolescente con cardiopatia congenita sono limitate. Pertanto, tale lavoro ha l'obiettivo di presentare i risultati ad interim di uno studio randomizzato controllato e multicentrico (studio TELEMACO) sull'efficacia del modello TC nella promozione dei livelli di engagement e di soddisfazione nell'adolescente con cardiopatia congenita nella fase di transizione.

Materiali e Metodi: Studio randomizzato controllato (RCT), in singolo cieco, con due braccia di studio: gruppo sperimentale in cui è stato erogato il modello TC versus gruppo di controllo con un approccio gestionale usual care. La raccolta dei dati è avvenuta al baseline, a tre e a sei mesi dall'arruolamento, attraverso l'utilizzo di validi strumenti di misurazione self-report. Gli adolescenti eleggibili all'arruolamento dovevo presentare i seguenti criteri di inclusione: età compresa tra 12-18 anni, diagnosi di cardiopatia congenita moderata e/o complessa, volontà alla partecipazione allo studio.

Risultati preliminari: Sono stati arruolati 70 pazienti (n=35 nel braccio sperimentale; n=35 nel gruppo di controllo). Nello specifico, non emergono differenze significative nel tempo per i livelli di engagement tra i due gruppi, nonostante a livello descrittivo il gruppo sperimentale presenta livelli più alti di engagement. Per quanto riguarda invece la soddisfazione, il gruppo sperimentale presenta valori di soddisfazione significativamente maggiori rispetto al gruppo di controllo a sei mesi (Gruppo TC= 86.91±11.16; Gruppo UC= 79.11±17.64; $p < 0.043$). Infine, sussiste una correlazione positiva tra engagement e soddisfazione ($r=0.286$, $p < 0.027$), tale per cui: maggiori sono i livelli di engagement maggiore è il livello di soddisfazione auto percepito.

Conclusioni: I risultati preliminari dello studio TELEMACO hanno dimostrato un'efficacia significativa del modello TC nella promozione dei livelli di soddisfazione. Il proseguo dello studio TELEMACO nei vari centri di arruolamento consentirà di corroborare l'efficacia del modello TC su tutti gli altri esiti di salute self-report indagati (es. qualità di vita, bisogni di salute) negli adolescenti con cardiopatia congenita nella fase di transizione.

CN22

COMPETENZE COMUNICATIVE E RELAZIONALE: UN ABILITA' ESSENZIALE NELLA PROFESSIONE INFERMIERISTICA

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Le competenze comunicative e relazionali di un infermiere rappresentano un elemento fondamentale per poter garantire un approccio professionale, olistico ed individualizzato. Queste competente come la capacità di comunicare (intelligenza emotiva, risoluzione dei conflitti), lo sviluppo delle proprie skills relazionali, le abilità personali e sociali sono particolarmente importanti.

Obbiettivi: Il seguente lavoro è volto ad evidenziare come le competenze comunicative e relazionali supportate dal confronto con i colleghi possono essere uno stimolo che spinge a migliorarsi utilizzando delle strategie quali:
Incremento della autostima professionale infermieristica. Miglioramento della comunicazione.
Rinforzo della consapevolezza dell'autonomia professionale.

Metodi: Per il raggiungimento degli obbiettivi si è scelto di svolgere una revisione narrativa della letteratura; utilizzando parole chiave come: skills, comunication, nurse, relational, patient, benefits.

Risultati: Acquistare le competenze comunicative e relazionali permettono di costruire un rapporto di fiducia, di migliorare la qualità dell'assistenza e il benessere del professionista.
Sono emerse anche quali strategie si sono rivelate utili al miglioramento di tali competenze attraverso alcune raccomandazioni.

Conclusione: Tutti professionisti sanitari, in particolare gli infermieri possono garantire un percorso lavorativo e assistenziale migliore attraverso le conoscenze e le capacità comunicative e relazionali.

CN23

QUAL È L'ESPERIENZA VISSUTA DEI PAZIENTI ADULTI CON CHD DURANTE LA PANDEMIA DI COVID-19? RISULTATI DI UN'ANALISI FENOMENOLOGICA INTERPRETATIVA

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La pandemia di COVID-19 ha modificato profondamente l'erogazione dell'assistenza sanitaria in tutto il mondo, con un impatto significativo sulle persone affette da malattie croniche, comprese le persone adulte con cardiopatia congenita (ACHD), le cui esigenze sanitarie sono state oscurate dai problemi causati dalla pandemia. Inoltre, durante la pandemia, i pazienti ACHD erano particolarmente a rischio a causa delle loro condizioni cliniche e della loro elevata vulnerabilità, oltre che aver dovuto affrontare importanti cambiamenti nel percorso terapeutico, come il passaggio dai sistemi di erogazione delle cure a programmi di telemonitoraggio realizzati per ridurre il più possibile l'esposizione ospedaliera, con un impatto significativo sulla vita dei pazienti ACHD. Tuttavia, ad oggi, nessuno studio ha esplorato le esperienze vissute di questi pazienti, che possono essere rilevanti per identificare i loro bisogni e possibili interventi mirati. Pertanto, questo studio mirava a descrivere l'esperienza vissuta di pazienti ACHD durante la pandemia di COVID-19.
È stato condotto uno studio qualitativo, con analisi fenomenologica interpretativa. La raccolta dati è stata eseguita con interviste semi-strutturate, su un campione di pazienti ACHD selezionato con metodo propositivo. Le interviste sono state audio registrate (previo consenso dell'assistito) e successivamente trascritte fedelmente per permettere l'analisi dei dati. I dati sono stati analizzati secondo la metodologia IPA, il rigore è assicurato dall'utilizzo delle tecniche di triangolazione, bracketing e member-cecking.
Sono state intervistati otto pazienti ACHD, di cui 6 donne. Il range di età variava da 35 a 56 anni. Dall'analisi qualitativa sono emersi tre temi ed otto sottotemi. Il primo tema "Paure in più", si compone di 3 sottotemi: Le paure individuali; Problematiche relazionali; Problematiche cliniche. La maggior parte dei pazienti intervistati ha manifestato un atteggiamento di paura dovuto al COVID-19 poiché la consapevolezza della "fragilità" data dalla loro patologia sommata ad un'informazione proveniente da fonti diverse, a volte contraddittoria, ha generato incertezza nel paziente e timore di contagiarsi.
Il secondo tema "Le strategie attuate", si compone di 3 sottotemi: La prevenzione del contagio; Il supporto; Le agevolazioni lavorative. Dalle interviste emergono quali sono state le strategie messe in atto dai pazienti ACHD per prevenire il contagio, ingannare il lento scorrere del tempo e riempire i vuoti lasciati dalla mancata opportunità di avere relazioni sociali.
Il tema 3 è stato identificato con "Miglioramento dei servizi" e si compone di 2 sottotemi: Il ruolo dei professionisti sanitari; Cambiamento delle infrastrutture. Dalle interviste è emerso la necessità di allargare la rete di servizi attraverso centri ambulatoriali lontani dalla sede centrale, attraverso il servizio domiciliare o con l'implementazione della telemedicina in risposta alla pandemia.
Il COVID-19 ha apportato una serie di modifiche che hanno intaccato non solo l'individualità ma anche l'organizzazione sanitaria, ciò ha avuto delle ripercussioni su tutti i pazienti ma soprattutto su quelli con patologie croniche come i pazienti ACHD. I risultati di questo studio possono essere utili per gestire future situazioni di disagio da parte dei pazienti ACHD, col fine di realizzare interventi sanitari mirati, atti ad implementare i servizi per questa tipologia di pazienti.

CN24

LA PERCEZIONE DEL FINE VITA DEI PAZIENTI ADULTI CON CHD. RISULTATI DI UN'ANALISI FENOMENOLOGICA INTERPRETATIVA

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I pazienti ACHD possono andare incontro ad insidiose complicanze, influenzando negativamente la sopravvivenza sia in termini di qualità di vita, sia in termini di durata della vita stessa. Inoltre, nel corso della progressione della malattia, i pazienti ACHD manifestano bisogni che diventano sempre più complessi con l'avvicinarsi della fine della vita, con la conseguente necessità di garantire loro cure e assistenza appropriate, personalizzate e centrate non solo sul paziente ma anche sulla sua famiglia. Per ottimizzare l'approccio personalizzato è importante conoscere la percezione che i pazienti ACHD hanno sul fine vita. In letteratura emerge che il tema sul "fine vita" viene trattato e affrontato con molta difficoltà sia dai pazienti che dai professionisti sanitari, con evidenza in merito discordanti. Perciò, l'obiettivo è esplorare la percezione dei pazienti ACHD relativamente alla tematica del fine vita.
È stato condotto uno studio qualitativo fra aprile e luglio 2021, con analisi fenomenologica interpretativa. La raccolta dati è stata eseguita con interviste semi-strutturate a distanza, su piattaforma online, su un campione di pazienti ACHD selezionato con metodo propositivo. Le interviste sono state audio registrate (previo consenso dell'assistito) e successivamente trascritte fedelmente per permettere l'analisi dei dati. I dati sono stati analizzati secondo la metodologia IPA, il rigore è assicurato dall'utilizzo delle tecniche di triangolazione, bracketing e member-cecking.
Sono stati intervistati otto pazienti ACHD, con un'età tra 35 e 53 anni, di cui 5 donne. 5 intervistati sono coniugati, e 7 pazienti sono di religione cattolica. Dall'analisi qualitativa sono emersi tre temi ed undici sottotemi. Primo tema: "La percezione di sè e della malattia". Nascere e crescere con una CHD, specie se complessa, è un'esperienza che segna il paziente nel corpo e nell'anima, un'esperienza accompagnata da sofferenza fisica e mentale. Un'esperienza in cui il funzionamento alterato del corpo non è solo un ingranaggio malfunzionante, ma porta con sé un significato, un sentimento, una nuova percezione di sé e della vita. Il secondo tema: "La comunicazione". La comunicazione circa il fine vita risulta essere complessa e articolata considerando che il tempo della comunicazione tra medico e paziente costituisce esso stesso tempo di cura. Ciò che verrà riferito al paziente cambierà in maniera importante il decorso della sua esistenza, dunque, il momento comunicativo deve essere scelto con minuziosa precisione. Il terzo tema: "Affrontare il fine vita". Il fine vita è un momento difficile e delicato per i pazienti e i famigliari, in primis, ma anche per gli operatori sanitari che assistono il malato. L'obiettivo è quello di accompagnare la persona ad una morte serena nel rispetto della dignità umana rispettando le decisioni e volontà espresse. L'accompagnamento inizia ben prima, attraverso la preparazione del paziente e dei famigliari all'evento. Inoltre, l'accompagnamento necessita di personalizzazione e quindi, del coinvolgimento di un team multidisciplinare.
Il fine vita negli ACHD è un argomento importante che influenza in maniera significativa la loro esistenza. In tal senso, sono necessari maggiori studi per approfondire quanto emerso e per poter aiutare gli ACHD ad affrontare il fine vita nel rispetto dei principi etici.

Venerdì 28 ottobre 2022
10.30•12.30
COMUNICAZIONI NURSING

CN25

I COMPORTAMENTI DI SELF CARE ATTUATI DALLE PERSONE ADULTE AFFETTE CARDIOPATIA CONGENITA: RISULTATI DI UNA REVISIONE SISTEMATICA DELLA LETTERATURA

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Il self-care si riferisce ad un processo decisionale naturale che gli individui con patologie croniche mettono in atto nella scelta dei comportamenti al fine del mantenimento della salute attraverso pratiche di promozione della salute e di gestione della malattia quando essa si verifica.
Numerose evidenze empiriche hanno evidenziato molteplici benefici nelle persone con patologie croniche che attuano comportamenti di self-care. Ciò diventa inevitabilmente necessario per i pazienti con CHD, soprattutto con l'avanzare dell'età con precarietà e criticità della condizione clinica. Tuttavia, ad oggi, pochi ricercatori hanno studiato i comportamenti di self-care attuati da adulti con CHD, e le evidenze disponibili ne fanno emergere una visione eterogenea e frammentata. Pertanto, l'obiettivo di questo studio è sintetizzare le evidenze disponibili relativi ai comportamenti di self-care attuati dai pazienti adulti con CHD.
È stata condotta una revisione sistematica della letteratura, con sintesi narrativa, il cui protocollo è stato registrato su PROSPERO (CRD42020218662). Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement e flowchart hanno guidato la selezione degli articoli da includere. Due ricercatori, in modo indipendente, hanno costruito le stringhe di ricerca ed interrogato le banche dati PubMed, Embase, Cinahl, and Scopus. La qualità degli articoli da includere è stata valutata seguendo le indicazioni del Joanna Briggs Institute da utilizzare nelle revisioni sistematiche.
La ricerca bibliografica nei database ha identificato 310 articoli, di cui 17 articoli sono in linea con i criteri di inclusione ed esclusione precedentemente individuati, ed hanno approfondito i comportamenti di self-care attuati da pazienti adulti con CHD. Di questi, solo 2 articoli sono stati guidati dalla Middle Range Theory of Self-Care of Chronic Illness, gli altri articoli si sono focalizzati su aspetti specifici del self-care, sottolineandone ulteriormente l'eterogeneità degli studi disponibili. Self-care maintenance è stato descritto da 11 articoli: solo il 45% dei pazienti ACHD attua adeguati comportamenti di self-care maintenance. Inoltre, molti pazienti con ACHD fumano sigarette, il 40% è sottopeso o sovrappeso-obeso e solo il 30% dei pazienti ACHD ha implementato un livello ottimale di attività fisica. Un paziente su tre con ACHD non ha ricevuto il vaccino antinfluenzale e un paziente su cinque con ACHD ha riferito una scarsa aderenza al trattamento medico in corso e alle visite di follow-up. Adeguati livelli di self-care monitoring sono adottati dal 27.3% dei pazienti con ACHD, e il 23.3% adotta adeguati livelli di self-care management: entrambi questi costrutti sono stati poco studiati.
Anche se le evidenze sul comportamento di auto-cura dell'ACHD sono scarse, gli studi sottolineano uno scenario allarmante, che rende necessarie ulteriori ricerche e l'implementazione di un supporto sanitario su misura per la peculiarità di questa popolazione. A questo proposito, sono fondamentali il ruolo e le competenze degli infermieri clinici avanzati, che fanno da ponte tra il bisogno sanitario dell'ACHD e il contributo specifico di ciascun operatore sanitario, lavorando in team multidisciplinari.

Venerdì 28 ottobre 2022
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COMUNICAZIONI NURSING

CN26

CENNI TEORICI SULL'ARTE TERAPIA E LA MEDICAL ART THERAPY

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L'arte terapia, considerando il suo significato più ampio, si riferisce alle applicazioni del linguaggio artistico, delle sue tecniche e dei suoi mezzi a scopo riabilitativo, preventivo e terapeutico. Nel 1994 l'Associazione Americana di Arte Terapia ha ufficialmente riconosciuto una branca dell'arte terapia che prevede l'utilizzo dell'espressione artistica con persone affette da malattie organiche che hanno subito un trauma a livello fisico e sono sottoposte a procedure mediche complesse. L'uso della terapia espressiva in campo medico si basa sul potere vitale dell'attività creativa e dell'immaginazione anche in un momento drammatico della vita di una persona che può essere causato dalla presenza di una malattia. L'obiettivo è quello di contribuire al processo di cura, facilitare l'elaborazione e l'integrazione di un'esperienza potenzialmente traumatica promuovendo e sostenendo la dimensione espressivo-comunicativa.

Venerdì 28 ottobre 2022
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CN27

ESPERIENZE CLINICHE DI ARTE TERAPIA IN UN REPARTO DI CARDIOLOGIA E CARDIOCHIRURGIA PEDIATRICA E DELL'ETÀ EVOLUTIVA

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Considerando l'ospedalizzazione come un'esperienza traumatica e tenendo presente le teorie sul trauma, si può affermare che esso provochi una caduta delle capacità di mentalizzazione. Gli effetti del trauma si stabilizzano nella memoria corporea alla quale si accede attraverso canali non verbali, per questo motivo l'espressione non verbale dell'arte può essere utile come strumento terapeutico. In questa sede verranno presentati alcuni interventi terapeutici mediati dall'utilizzo di tecniche espressive in un reparto di cardiologia e cardiochirurgia pediatrica e dell'età evolutiva. Saranno esplorati casi clinici in età pediatrica e in età adulta durante un periodo di ospedalizzazione e in attesa di trapianto cardiaco. L'obiettivo è quello di focalizzarsi sul valore dell'espressione artistica nell'esplorare e comunicare il mondo interno dei vissuti emotivi legati all'ospedalizzazione e alla malattia.

Venerdì 28 ottobre 2022
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CN28

IL TECNICO DI CARDIOLOGIA: RUOLO CRUCIALE NEL FOLLOW-UP DEI CARDIOPATICI CONGENITI ADULTI

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Introduzione: Il Tecnico di fisiopatologia cardiocircolatoria e perfusione cardiovascolare (TFCPC) fonda la propria formazione come responsabile della gestione della circolazione extracoporea durante gli interventi di cardiochirurgia, ma nel nostro Centro è riuscito ad affermarsi anche nell'ambito della cardiologia compreso il follow-up dei pazienti ACHD (Adult Congenital Heart Disease).
L'unità operativa dedicata all'assistenza ACHD nasce per soddisfare le esigenze di questi pazienti e per garantire loro una continuità terapeutica. Per questo, è necessaria un'organizzazione sanitaria con programmi specifici previsti da un team pluri-specialistico, con conoscenze anatomiche e fisiopatologiche delle cardiopatie congenite in storia naturale e/o operate.
L'assistenza di questi pazienti è un processo che dura tutta la vita e richiede strategie di pianificazione anticipate delle cure.

Scopo: Lo scopo di questo studio è descrivere l'importanza del TFCPC durante il follow-up dei pazienti ACHD.

Metodi: Analisi delle competenze del tecnico nella pratica quotidiana di un centro di terzo livello.

Risultati: La nostra unità di ACHD è composta da: 1 Responsabile Cardiologo, 2 Cardiologi esperti, 2 TFCPC (Sonographer/Elettrophysiologist),1 Infermiere, 1 capo tecnico e 1 caposala.
Questo team si basa su un modello Anglosassone che prevede la presenza di tecnici di cardiologia con compito di interfaccia medico-paziente. Gli esami di primo livello (ecocardiogramma color Doppler, ECG dinamico secondo Holter, controlli PM, Test da sforzo/cardiopolmonare) vengono eseguiti ed elaborati per fornire un piano terapeutico con corretta stratificazione del rischio.
L'Ecocardiografia è tra gli esami chiave nella valutazione del follow-up di questi pazienti, che seguono un percorso diagnostico con timing secondo Linee Guida, ma talvolta personalizzati in base alle condizioni del paziente. Tale esame richiede un Sonographer esperto in CHD capace di eseguire le seguenti valutazioni: M-mode, 2D e 3D, doppler pulsato, continuo e color, speckle-tracking; valutazione della presenza di versamenti pericardici/pleurici. Al termine dell'esame, il tecnico fornisce una prima interpretazione dei dati registrati, che vengono successivamente sottoposti all'attenzione del cardiologo.
È stato visto che il test da sforzo o test cardiopolmonare TDS/TCP, insieme alle immagini dell'ecocardiogramma, rimane un'indagine che fornisce informazioni fondamentali per la prognosi ed eventuali strategie terapeutiche. Il TCP richiede un tecnico esperto, capace di interpretare i dati forniti dai test. Le informazioni ottenute dal TCP, tra cui il consumo di ossigeno (VO2), sono importanti indici predittivi in pazienti a rischio di ospedalizzazione o di morte/trapianto. Il tecnico in collaborazione con il medico svolge un ruolo cruciale nell'acquisire, interpretare e refertare il TCP, analizzando i dati ECG e cardiorespiratori.
Un aspetto innovativo è quello di aggiornare costantemente il database dell'Unità Operativa, favorendo l'analisi statistica e permettendo anche elaborazioni di studi retrospettivi e protocolli prospettici.
Il risultato è quello di supportare e sollevare il medico dall'esecuzione pratica degli esami, in modo da poter concentrare, dedicare e designare la strategia/terapia migliore per il paziente.

Conclusioni: Sulla base dei risultati ottenuti in termini di miglioramento per la cura e l'assistenza del cardiopatico congenito adulto, si può concludere che la figura del tecnico di fisiopatologia cardiocircolatoria e perfusione cardiovascolare è diventata fondamentale per il follow-up di questi pazienti fragili e cronici che necessitano di cure costanti e personalizzate.

CN29

PATIENT ADVOCACY: ALLEANZA TRA PAZIENTE, PERSONALE MEDICO E PERSONALE SANITARIO, ISTITUZIONI E AZIENDE

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Durante la Pandemia Covid-19 le associazioni e maggiormente le associazioni di pazienti, hanno dimostrato il loro importante ruolo nel supportare le tantissime persone affette da patologie croniche e rare e le loro famiglie, ad affrontare quel difficile periodo (ALTEMS, 2020).

Ciò ha permesso alle associazioni che si occupano di cardiopatie congenite di avere maggior consapevolezza della propria forza di reazione alle difficoltà e alle persone affette da cardiopatia congenita a prendere in mano la propria vita e cercare attraverso altre persone con le stesse difficoltà a dare delle risposte ai propri bisogni. Attraverso l'associazione Aicca, (associazione di pazienti affetti da cardiopatia congenita), si è riuscito a creare un percorso di empowerment ed engagement che ha permesso di costruire un'organizzazione capace di operare in ambito sanitario a sostegno e tutela dei pazienti.

Tappa fondamentale del percorso per l'associazione è stata l'opportunità di formazione attraverso il Corso di alta formazione "Raise the patient voice" organizzato da Janssen con il supporto scientifico dell'Alta Scuola di Economia e Management dei Sistemi Sanitari (ALTEMS) dell'Università Cattolica del Sacro Cuore.

Il Corso ha messo in luce l'importanza fondamentale delle associazioni di pazienti e il loro ruolo sempre più centrale nello sviluppo della sanità del futuro.

Esse stanno diventando un soggetto che concorre alla costruzione e allo sviluppo di alcune delle più importanti politiche sanitarie, in oncologia come nelle malattie rare, nelle malattie metaboliche come nella reumatologia, nelle cardiopatie congenite.

La collaborazione e l'alleanza tra tutti gli stakeholder, garantisce un'ottimizzazione delle cure e mette il paziente non più solo al centro delle cure ma accanto a coloro che si occupano del suo benessere. Il paziente è il maggior esperto di sé stesso e per tale ragione più di altri deve avere la capacità di poter suggerire per sé le modalità più idonee per rispondere a suoi bisogni. Tale capacità messe insieme tramite le associazioni di pazienti, possono cambiare il modo di vedere la cura e cambiare la vita.

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