Topic 10 – Pediatric and congenital heart disease

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Right atrial area and right ventricular outflow tract akinetic region length predict sustained tachyarrhythmia in repaired tetralogy of Fallot

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Objectives: We investigated the significance of right atrial (RA) and right ventricular (RV) anatomy and function for arrhythmia prediction.

Background: Repaired tetalogy of Fallot (rTOF) patients are at risk of atrial or ventricular tachyarrhythmia and sudden cardiac death.

Methods: One-hundred-and-fifty-four rTOF adults who underwent cardiovascular magnetic resonance (CMR) were studied with the pre-specified end-point of new-onset atrial or ventricular arrhythmia (sustained ventricular tachycardia/ventricular fibrillation) during a longitudinal follow-up.

Results: Median age was 31 (IQR:22-40), median follow-up was 5.6 (IQR:4.6-7.0) years.

Atrial tachyarrhythmia (n=11) was predicted by maximal right atrial area indexed to body surface area (RAA) on cine-CMR (Hazard ratio; HR1.17, 95%CI 1.07-1.28 per cm²/m²; P=0.0005, survival ROC curve analysis, area under curve; AUC 0.74[0.66-0.81], cut-off value 16 cm²/m²). Atrial arrhythmia-free survival was reduced in patients with RAA ≥16 cm²/m² (Logrank; P=0.001).

RV restrictive physiology on echocardiography (n=38) related to higher RAAi (P=0.02) but did not predict atrial tachyarrhythmia (P=0.057). RV restrictive physiology patients had similar RV dilatation and exercise impairment to remaining patients representing a different phenotype from previous reports.

Ventricular arrhythmia (n=9) was predicted by CMR RV outflow tract (RVOT) akinetic area length (HR1.05, 95%CI 1.01-1.09 per mm; P=0.003, survival ROC analysis, AUC 0.70[0.83-0.91]; cut-off value 30 mm) and decreased RV ejection fraction (HR0.93 95%CI 0.87-0.99 per %; P=0.03, respectively). Ventricular arrhythmia-free survival was reduced in patients with RVOT akinetic region length >30 mm (Logrank; P<0.02).

Conclusions: RAA predicts atrial arrhythmia and RVOT akinetic region length predicts ventricular arrhythmia in late follow-up of rTOF. These are simple, feasible measurements for serial surveillance and risk stratification of rTOF patients.

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Cardiac Magnetic Resonance in Children with Acute Myocarditis

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Background: Diagnosis of acute myocarditis (AM) is challenging because its clinical presentation may overlap with that of common infectious diseases or be diagnosed as idiopathic dilated cardiomyopathy (DCM). While Cardiac Magnetic Resonance (CMR) imaging has emerged as an important non-invasive tool in the diagnostic procedure of AM in adults, data on CMR in children remain scarce.

Aim: To describe feasibility of CMR and its contribution for the diagnosis and follow-up of AM or for the etiology left ventricular (LV) dysfunction of unknown origin in children.

Methods: Over a period of 3 years, 43 children underwent CMR for clinical suspicion of AM with or without LV dysfunction of unknown origin. CMR sequences included unenhanced cine-steady state free precession (SSFP), black-blood-prepared T1-weighted images and T2-weighted images and T1-weighted images (EGE) and 3D late gadolinium-enhanced after injection of gadolinium chelate (LGE). The diagnosis of myocarditis was based on the recently consensus criteria. CMR was repeated during follow-up in children with confirmed diagnosis of AM.

Results: AM was diagnosed by CMR in 30/43 children: 22/30 had LV dysfunction, 8/30 had normal LV function but elevated blood levels of troponin I. T2 hyper-signal was present in 21 cases, EGE and/or LGE were present in 29/43 cases. Two children died during hospitalization. All survivors with LV dysfunction had normal echocardiography after a median follow-up of 10 months. 24/30 patients had control CMR that revealed in 4 cases the persistence of inflammation in T2-weighted images and in 6 cases persisting LGE. No children with AM without LV dysfunction developed dilated cardiomyopathy. The remaining 13/43 children without AM on CMR were diagnosed with DCM: 2/13 normalized after 4 and 30 months of follow-up respectively, and 11/13 are still followed for dilated cardiomyopathy.

Conclusion: CMR in children with clinical suspicion of AM or with LV dysfunction of unknown origin is feasible and useful in the diagnostic work-up. It may help to adapt medical targeted therapy and to be more precise in prognosis assessment in infants recently diagnosed with DCM of unknown origin.

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Multiparametric assessment of the Right Ventricle by echography in patients with repaired Tetralogy of Fallot undergoing pulmonary valve replacement: a comparative study with MRI

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Purpose: Evaluation of the right ventricle (RV) using echography is challenging in patients with repaired Tetralogy of Fallot (rTOF). The objective of this study was to evaluate the feasibility and the accuracy of a multiparametric echographic approach including 2D strain and 3D for RV volumes and function assessment, in comparison with MRI.

Methods and results: we performed an echographic study including 2D TAPSE, S’TDI, Tei indice, Fractional area change (FAC), 2D strain and 3D, and an MRI in 26 consecutive patients with rTOF before PVR and one year after surgery. TAPSE, S’TDI and 2D strain parameters were poorly correlated with MRI regarding RV function assessment. FAC was well correlated with REVF before and after PVR (r=0.70, p<0.01 and r=0.68, p<0.01, respectively). Despite RV volumes underestimation, 3D analysis was well correlated with MRI values in both pre and post-operative assessment (r=0.88, p<0.01 and r=0.91, p<0.01 respectively for REVDF; r=0.92, p<0.01 and r=0.95, p<0.01 respectively for RVESV).

Conclusion: Global approach of RV function using 2D (FAC) or 3D (FAC) parameters seems reliable in patients with rTOF. The commonly used TAPSE and S’TDI focused on segmental analysis of RV inflow are less sensitive probably because RV inflow is less affected by RV remodeling related to initial surgical repair.

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Right ventricular function with standard and speckle-tracking echocardiography and clinical tolerance in adults with D-transposition of the great arteries post atrial switch

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**Objectives:** 1/To determine the association between systolic and diastolic function of the systemic right ventricle (RV) evaluated by echocardiography and exercise capacity, and BNP. 2/ To determine the prognostic value of deformation parameters of the sRV in D-transposition of the great arteries and prior atrial switch

**Method:** 20 patients with D-TGV and prior atrial switch (mean age 29±8 years, 6 women) were prospectively evaluated. The systolic and diastolic function of systemic RV were studied using standard ultrasound parameters, and speckle tracking to measure global longitudinal strain (GS), global systolic strain rate (GSRs), global early diastolic strain rate (GSRd), systolic twist and diastolic untwist of sRV. Echographic data were compared with maximum oxygen uptake and BNP performed in the same day. Relationship with clinical events was studied subsequently. A comparison with 20 controls matched for age and sex is being

**Results:** GS, GSRs, GSRd, systolic twist and diastolic untwist of systemic RV were –11.1±2.9%, –1.2±2.4 sec–1, 0.9±1.2 sec–1, 2.4±2.3 ° and –3.5±4.9 ° respectively. No correlation was found between systemic RV diastolic parameters (E, E / A, E / Ea, isovolumic relaxation time, GSRd, and diastolic untwist) and maximum oxygen uptake or BNP. A significant correlation was found only between GS and maximum oxygen uptake (p=0.001). In univariate analysis, parameters of systolic function (S wave peak at tricuspid annulus, GS, GSRs, systolic torsion) were significantly associated to heart failure. In multivariate analysis only peak S wave at the tricuspid annulus (p=0.01) and GS (p <0.0001) were significantly associated with cardiovascular events.

**Conclusion:** The GS is strongly correlated with exercise capacity and cardiovascular events in the TGA palliated by atrial switch. A study on a larger sample will confirm these results.

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**Comparison of 2D and 3D transthoracic echocardiography for measurement of aortic annulus diameter in a paediatric population**

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Precise evaluation of the aortic root geometry is necessary in congenital aortic valve lesions in children, to optimize surgical or percutaneous procedures. The aim of the study was to compare two-dimensional (2D-TTE) and three-dimensional transthoracic echocardiography (3D-TTE) for analysis of aortic annulus and assess feasibility of 3D imaging.

**Methods:** Thirty consecutive children, without heart disease, aged 11±3.6 years old (min 4; max 18; 66.7% boys), were prospectively included in this study. Transthoracic real time 2D and 3D echocardiography (ie 33, Philips, Andover MA, US) was performed using matrix probe (x 3-1, x 7-2 and x 5-1). Multiplanar reconstruction was used to measure in diastole two orthogonal aortic annulus diameters, compared to the measurement in a para-sternal long axis view in 2D-TTE.

**Results:** 3D aortic annulus diameters measurements were obtained in 28 (93.3%) children. Mean annulus diameter in 2D TTE (1.93±0.2 cm) was not significantly different of the mean minimal diameter (1.95 cm: 0.3, p=0.43) but was smaller than the mean maximal diameter (2.03 cm :±0.3, p=0.002) in 3D-TTE. 2D annulus diameter was well correlated to minimal 3D diameter (r=0.89, p=0.0001) and maximal diameter (r=0.88, p=0.0001). A significant difference in the mean minimal and maximal 3D aortic annulus diameters was observed (p<0.0001). 3D horizontal aortic annulus diameter was larger in 15 patients (54%) whereas vertical diameter was larger in 7 patients (25%). Orthogonal diameters were equal in 6 patients (21%). The index of eccentricity was 4±3%.

**Conclusion:** This preliminary study demonstrated the feasibility of 3D-TTE for the assessment of aortic annulus diameter in a standard children population. Because of the asymmetry of the aortic annulus, such 3D measurements could have important issues before aortic valve dilatation or surgical replacement in children.

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**Effects of advanced therapies on echocardiographic and Doppler measures, compared with clinical evaluation and BNP in patients with Eisenmenger syndrome**

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Management options for patients with Eisenmenger syndrome (ES) were limited to palliative measures or transplantation before the advent of advanced therapies (AT). Little data is available on the impact of disease targeting therapies on BNP and echocardiographic variables in patients with ES. We wanted to investigate the effects of AT on echocardiographic measures and BNP in patients with ES.

**Methods and results:** We prospectively included 63 patients with ES who were started on AT (46% were treated with endothelin receptor antagonists, 44.4% with PDE5 inhibitors and 9.6% with an association. Clinical, 6 min walk test and BNP data were collected at baseline (before introduction of AT) and after a mean period of 2 years of sustained therapy.

Mean age was 42±13 years, most of patients were WHO III (97%) whereas a vast majority had post-tricuspid defects. At baseline, echocardiographic findings were consistent with abnormalities of the RV function and adaptation: overall dilated RV (mean inlet 44.9±6.2 mm), reduced TAPSE (16.4±3.5 mm). AT was responsible for an improvement in WHO functional class (p=0.001) and walk distance (±35.1 m; p=0.01), however, it was not associated with any change in BNP (p=0.85). RV systolic function and adaptation was improved (decreased systolic/diastolic duration ratio and total isovolumic time, p<0.001 and p=0.03; decreased RV dP/dT, p=0.05; increased TAPSE and tricuspid Sm, p=0.01 and p=0.03) however, no significant right ventricular structural remodeling was observed. Agreement between the echocardiographic score (previously described as associated with outcomes in ES: reduction in TAPSE, RA area, RA/LA ratio or S/D ratio) and clinical response (decrease in WHO and improved 6MWT) was moderate but higher than BNP.

**Conclusion:** Our results support the notion that therapy is associated with improved symptoms, RV systolic function and adaptation but not with reduction in BNP concentrations.

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**Intracardiac echocardiographic guidance is efficient for transcatheater closure of atrial septal defect in an unselected patient population**

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**Background:** Intracardiac echocardiography (ICE) is an alternative to transesophageal echocardiography (TEE) for transcatheater closure of atrial septal defect (ASD). However, studies on ICE guidance generally include a majority of patent foramen ovale (PFOs) and only few ASDs without rim deficiency. Our aim was to assess transcatheter closure of ASDs under ICE guidance in an unselected patient population.

**Method:** From January 2006 to January 2012 in our institution, all the ASDs in the adult population were closed percutaneously under ICE guidance. During this period, 93 patients (34 males, 59 females, mean age 46.9 years) had transcatheter ASD closure with Amplatzer devices under local anesthesia and ICE guidance. All patients had routine TEE before catheterization.

**Results:** Fifteen patients (13.9%) had deficient rim(s) other than the antero-superior. The median ASD diameter by TEE and device size was 20 and 26 mm, respectively. Ninety cases (96.7%) were successfully closed. Three cases failed because of insufficient rims and/or defect size superior to 40 mm. Minor and transient complications occurred in 10 patients (9.6%). Three patients experienced a major complication with favorable outcome: one arterial femoral wall tear treated by embolization, one blood transfusion for a groin hematoma and one retroperitoneal hematoma. The only risk factor for failure to close the ASD was deficient rims (p=0.05), whereas the size of the...