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 (GSRe), 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, GSRe, 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, GSRs, 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). Multplanar reconstruction was used to measure in diastole two orthogonal aortic annulus diameters, compared to the measurement in a parasternal 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=0.009) 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 walked distance (+3.1 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<0.003; 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 transcatheter 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 transcatheter 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 size 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
ASD was a risk factor for major complications (p=0.03). The correlation coefficient between the size of the ASD with TEE and ICE was 0.887.

Conclusion: In an unselected patient population, ICE provides a safe and efficient guidance for device closure of ASD, even for large defects with deficient rims.

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Long-term outcome of 117 patients with univentricular heart and common atrioventricular valve

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Introduction: Few studies investigated the long-term outcome of patients with univentricular heart and common atrioventricular valve.

Method: We retrospectively analysed the medical files of all patients univentricular heart with common atrioventricular valve in the setting of heterotaxy or with unbalanced atrioventricular septal defect preventing biventricular repair.

Results: 117 patients were identified during the study period. 89/117 had a postnatal diagnosis. 28/117 patients never underwent surgery, 25/117 underwent one palliation surgery (Blalock-Taussig-shunt (BTS)/pulmonary banding), and finally, 61/117 patients entered a sequential cavopulmonary connection program: 37/61 had partial cavopulmonary connection at the time of data analysis while 24/61 had total cavopulmonary connection (TCPC). The average age at TCPC was 7.6 years +/-4 years [1.7-16 years]. Three patients were eventually transplanted.

The overall mortality was 59% (69/117): 65% and 30% in heterotaxy and in patients with unbalanced atrioventricular septal defect respectively. Mortality was 85% (24/28) in the subgroup of patients who never underwent surgery, 93% in the subgroup of patients who had a BTS, and 89% after pulmonary banding. In the subgroup planned to have TCPC, 49% died after partial cavopulmonary connection and survival rate was 71.6% [50.7-100] in patients who had TCPC.

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Toward better ventricular pacing in patients with a systemic right ventricle

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Introduction: Patients with transposition of the great arteries (TGA) and atrial redirection, have an important risk of heart failure caused by dysfunction of the systemic RV. Conventional non-systemic ventricular pacing (non-systVP) may even further increase this risk. We investigated whether these patients may benefit from biventricular pacing (BiVP) and/or single-site systemic ventricular pacing (systVP).

Methods: During clinically indicated catheterization in 9 patients with TGA and status post Senning procedure, endocardial ventricular stimulation (overdrive DDD, 80-90 bpm) was applied with temporary pacing leads at the non-systemic ventricle and the systemic ventricle. Acute changes in dp/dtmax and systolic pressure of the systemic ventricle, as induced by non-systVP, systVP and BiVP compared to reference, were assessed with a pressure wire (RADI Medical Systems®) within the systemic ventricle. Reference was AAI pacing with similar heart rate (n=7; filled squares), or non-systVP at a lower heart rate than during stimulation at experimental sites (85 vs. 90 bpm; n=2).

Results (Figure): Systemic dp/dtmax and systolic ventricular pressure were significantly higher during systVP (+15.6% and +5.1%) and BiVP (+14.3% and +4.9%) when compared with non-systVP. In 6 out of 7 patients, dp/dtmax was even higher during BiVP and systVP than during AAI pacing.

Conclusions: Patients with systemic RV, such as patients with TGA and atrial redirection, may benefit from biventricular or systemic ventricular pacing, especially when ventricular pacing is indicated.