enotheropathy, one a dysfunction of the systemic ventricle and edema, and the last one severe venous insufficiency of the legs. A Melody valve was successfully inserted in all patients; three in the pathway, one after the iliac bifurcation. There was no significant modification of the pressures after valvulation. No acute complication was recorded. There was an improvement of the doppler through the proclive and breathing. However, these were not associated with significant clinical improvement. No thrombosis of the valve accured during the short follow-up (1 to 6 months).

Conclusions: The valvulation of the Fontan circuit is easily performed. We noticed an improvement of the hemodynamic and flow. However, no clinical improvement was recorded as a result. A longer follow-up is needed to appreciate the risks as well as the interest of this procedure.

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Percutaneous insertion of a Melody valve in tricuspid position: technical aspects

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Background: percutaneous transcatheter heart valve replacement of aortic or pulmonary valve is established. Transcatheter atrioventricular valve replacement is described. We report our experience focusing on the technical aspects.

Methods: we retrospectively review the files of patients who received a transcatheter valve in tricuspid position between 2008 and 2012.

Results: Four patients were found. 3 had a heterograft (conduit of 14-mm, Sorin 33 et Edwards Permount 33) and one had a connection between the RA and the RV infundibulum without a valve. Two patients had tricuspid regurgitation as a primary lesion, one had stenotic valve and the last one a mixed lesion. All successfully received a Melody valve from a femoral access. In patients with stenotic lesion, a prolationation using a high pressure balloon was performed before valve implant. In patients with regurgitation, the landing zone was calibrated using a low pressure balloon. These patients were presented to create a landing zone of adequate diameter. Melody valves were inserted using a 22-mm balloon catheter in 3 and a 24-mm in one. All but one were post-dilated. There was no significant regurgitation. The mean gradient across the tricuspid valve felt from 12 to 4.6-mmHg. One patient needed an epicardic pacemaker because of AV block following balloon dilatation. One patient required inotropic support and ventilation following the procedure but recovered after few days.

Conclusion: Transcatheter tricuspid valve insertion is feasible in patients with surgical hetero or homografts after a careful selection. The mechanism of dysfunction must be known. In case of stenosis or mixed lesions, the only question is to know if the stenosis could be relief. In case of regurgitation, it is very important to know the features of surgical substrats and to calibrate the tricuspid orifice. Finally, patients with inappropriate landing zone should be presented prior to valve insertion.

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Potts’ shunt in children with idiopathic pulmonary arterial hypertension: long-term results

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Background: Idiopathic pulmonary arterial hypertension (IPAH) remains a progressive fatal disease. Palliative Potts’shunt has been proposed in children displaying supra-systemic IPAH.

Methods: A retrospective multicenter study to evaluate Potts’shunt in pediatric IPAH.

Results: Between 2003 and 2010, 8 children with supra-systemic IPAH and in WHO functional class IV despite medical PAH therapy underwent Potts’shunt. Age at IPAH diagnosis ranged from 4 to 180 months (median age: 64 months). Surgical procedure was performed in a mean delay of 41.9±54.3 months (from 4 to 167 months, median delay: 20 months) after IPAH diagnosis. Mean size of the Potts’shunt was 9.25±3.30 mm. Two patients, whose medical PAH therapy had been interrupted just after surgery, died at post-operative day 11 and 13 from acute pulmonary hypertensive crisis. After a mean follow-up of 63.7±16.1 months, the 6 children who were discharged from hospital were alive. Functional status improved markedly in the 6 survivors with a WHO functional class I (n=4) or II (n=2) at last follow-up, consistent with significant improvement of 6 minute walk distance [302±95 (51±20% of theoretical values) vs 456±91 meters (68±10% of theoretical values), p=0.038] and decrease of brain natriuretic peptide (BNP) levels [608±109 vs 76±45 pg/ml, p=0.035]. No Potts’shunt was found restrictive at last echocardiography.

Conclusion: Palliative Potts’shunt constitutes a new alternative to lung transplantation in severely ill children with supra-systemic IPAH, carrying a prolonged survival and persistent improvement in functional capacities.

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Conotruncal defects: is the ventricular septal defect always the same?

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Conotruncal defects (CTD) are a group of cardiac malformations heterogeneous from an anatomical standpoint but with a common embryologic origin: an abnormal rotation of the outflow tract. The outlet septum is therefore malaligned or absent, resulting in a ventricular septal defect (VSD).

Aim of the study: To analyze the anatomy of the VSD in hearts with CTD.

Material and methods: We reviewed 200 heart specimens with CTD from the anatomic collection of the French Center of Reference for Complex Congenital Heart Defects: 70 Tetralogy of Fallot (TOF), 53 TOF with pulmonary atresia (TOF-PA), 54 common arterial trunk (CAT), and 23 interrupted aortic arch type B (IAA-B). Special attention was paid to the rims of the VSD viewed from the right ventricular side, the relationships between tricuspid and aortic valves, and the anatomy of the outlet septum.

Results: The VSD was located between the 2 limbs of the septal band (conoventricular) in all hearts. There was a fibrous continuity between tricuspid and aortic valves in 0% of IAA-B, 66% of TOF, 37% of TOF-PA, 1% of CAT (p<0.005). When present, this continuity always involved the anterior tricuspid leaflet. The outlet septum was demonstrable in 81% of IAA-B, 96% of TOF, 39% of TOF-PA, 0% of CAT (p<0.0001).

Conclusion: All CTD share the same VSD, located between the two limbs of the septal band. However, there are some differences regarding the inferior rim of the VSD. The continuity of the aortic valve with the anterior, and not the septal, tricuspid leaflet indicates that this continuity may be a consequence of the malposition of the ventriculo-infundibular fold, along with its outlet septal component, rather than a perimembranous extension of the VSD. Finally, these differences suggest an anatomic continuum from IAA-B to CAT rather than distinct physiological phenotypes, related to various degrees of abnormal rotation of the outflow tract, excessive in IAA-B, incomplete in TOF, TOF-PA and CAT.

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Congenital left coronary ostial atresia or stenosis – a series of four neonatal fatal cases

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Introduction: Congenital left coronary ostial atresia or severe stenosis is an extremely rare coronary abnormality. The clinical picture is either cardiac failure in the small infant or chest pain in the older child or adult patient. This report describes four neonatal cases of this rare abnormality.

Methods: We retrospectively accessed all cases of left coronary ostial stenosis or atresia seen in our center during an 11-year period (2000–2011). Two older patients with this diagnosis seen at 18 months and 4 years were excluded from the study.

Results: Four neonates with the diagnosis of left coronary ostial atresia or stenosis were identified during the study period. One newborn died within minutes, the other within hours after birth because of cardiac failure refractory to all treatment strategies. In both cases left coronary stenosis (one case with a “pinpoint” orifice and the other with a “slitlike” orifice) was diagnosed at autopsy. The third neonate was in cardiac failure due to a severe aortic stenosis. Left coronary ostial atresia was diagnosed during emergency catheter procedure. The infant subsequently died after the aortic dilatation. The fourth infant had a cardiac arrest at the third day of life after normal delivery and neonatal adaptation. She was diagnosed with left coronary ostium atresia by coronary angiography performed because of persistent biventricular dysfunction. She died during the attempt of revascularization surgery at 2 weeks of life.

Conclusion: Congenital left coronary ostium atresia or stenosis is very rare. Coronary angiography is the diagnostic method of choice especially in the small child or infant. Revascularization surgery seems indicated in symptomatic children based on case reports and small series. The clinical picture described here for the first time in the neonate is dramatic and quickly fatal with scarce surgical options. Systematic examination of the coronaries should be part of any neonatal autopsy.

The transcatheter closure of Atrial Septal Defect (ASD) in patients older than 60 years: Retrospective study of 41 cases

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Introduction: Transcatheter closure of ASD is now proposed as first line treatment for the elderly patients.

Purpose: The aim of our study is to evaluating the transcatheter closure of ASD in patients older than 60 years.

Methods: The records of 41 patients (37 women, 4 men) who are over 60 years (69.8±6 years, range 61-82 years), referred between April 1998 and December 2010 for transcatheter closure of secundum ASD, were retrospectively reviewed.

Results: ASD was discovered during the assessment of patients with dyspnea (17) or arrhythmia (17). Average age at time of diagnosis was 62.4±14.89 years (range 14-82 years). 37.8% of cases had a delayed diagnosis.

At time of the procedure, 19 patients (46.3%) had supraventricular arrhythmia and 82.9% of patients were dyspneic. Shunt ratio was 2.50±0.66, defect size 19.67±6.76 mm, stretched diameter 24.36 mm±5.87, device diameter of 24.49 mm±6.58. One patient was recused, because of increased left atrial pressure at occlusion test.

Fluoroscopy time was 7.01±4.41 min. Success rate is 97.5%. Complete closure rate at one day and one month was 84.6% and 94.9% respectively. Systolic pulmonary pressure decreased from 49.37 to 34.58 mmHg at one month.

Hospital stay was 3.58 days ± 2.71. Periprocedural complication rate was 12.2%. 2 complications were major: one death by esophageal hematoma, and one oropharyngeal bleeding, and 3 minors: 3 femoral arteriovenous fistulas.

At late follow-up (34±44 months, range 1 month to 12 years), 34 patients were in stage one, 4 recurred supraventricular arrhythmia, 1 needed pace maker for sinus node dysfunction.

Conclusion: Transcatheter closure of ASD in elderly patients is effective. It does not change rhythmic status but allows symptomatic improvement.

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Circulating endothelial cell levels decrease after vasodilator therapy and are a biomarker of deterioration in pediatric pulmonary hypertension

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Background: Pulmonary vasodilators in general and prostacyclin therapy in particular have markedly improved the outcome of patients with pulmonary arterial hypertension (PAH). Endothelial dysfunction is a key feature of PAH and we previously described that circulating endothelial cells (CECs) could be used as a biomarker of endothelial dysfunction in PAH. We now hypothesized that PAH-specific vasodilator therapy might decrease CEC numbers.

Methods: We quantified CECs in peripheral blood from children with idiopathic PAH (PAH, n = 30) or PAH secondary to congenital heart disease (PAH-CHD, n = 30), before and after treatment and during follow up. CECs were enumerated by immunomagnetic separation with mAb CD41+46-coated beads.

Results: CEC counts were significantly decreased in children after treatment with oral endothelin antagonists and/or PDE5 inhibitors. In 10 children with refractory PAH despite combination oral therapies, SC treprostinil was added and we found a further significant decrease in CEC count during the first month of treatment in every patient. We quantified CEC during 6 to 36 months follow-up after initiation of SC treprostinil and found that CEC count is modified according to clinical status.

Conclusions: CEC counts fall with vasodilator therapy in PAH and could also be used as biomarker of deterioration in refractory pediatric pulmonary hypertension treated with SC treprostinil.

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Use of covered stents for closure of fenestration in extracardiac cavopulmonary connection: technical aspects and mid-term results

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Objective: Closure of fenestration in a cavopulmonary connection (TCPC) is performed with devices used to close intracardiac or aorta-pulmonary connections. This study presents our data regarding the use of covered stents in patients with fenestrated TCPC.

Methods: We retrospectively reviewed the data of all the patients receiving a covered stent to close a fenestration of TCPC between 2005 and 2012.

Results: 38 patients were included. Mean age and weight were respectively 7.6 years and 20 kg. Femoral access was mainly used (32) but also jugular access (5) and trans-hepatic access (1). A Cheatham Platinum Stent was used (34) but we also used Atrium V12 Stents (4). The balloon size was chosen according to the diameter of the conduit. We used BIB balloon (14) or simple balloon (Tyskhal or Balty) (16) (balloon not reported (4)). Mean procedural and fluoroscopy times were respectively 42.5 ± 21 and 7.5±6.6 minutes. Mean central venous pressure rose from 10 to 12mmHg. Mean oxygen saturation increased from 90% to 96%. There was one anaphylactic shock at the end of the procedure and one air embolisation without hemodynamic compromise. Full occlusion was confirmed on angiogram in 36 patients, 2 cases required 2 stents to achieve full occlusion. There was one minimal residual shunt and one failure of the procedure with significant residual shunt in a patient with a conduit made of a Gore-Tex patch between the atrium and the pulmonary arteries. There were no embolic event, acute venous thrombosis or arrhythmias. Patients were treated with anti vitamin K for 6 months then aspirin. Mean follow-up is 49 months without thrombo-embolic complication or desaturation.

Conclusion: Covered stent is an option to close fenestration in extracardiac TCPC. It is safe, easily achieveable with low fluoroscopy time, very low risk of thrombo-embolic events or failure. The good results are sustainable. The main advantage of the technique is to avoid prosthetic device in the atrium.