entheropathy, one a dyfunction 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 been 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 Perimount 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 mixted lesion. All successfully received a Melody valve from a femoral access. In patients with stenotic lesion, a predilatation 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 mixted 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 peptid (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 anatomic 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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