Thirty-one patients were included. The DCPT was performed on average at 9 years old and the hepatological assessment 15 years later. All patients had sinusoidal dilatation and fibrosis and cirrhosis was present in 10 patients (38%). A 25-year-old woman was diagnosed with multifocal HCC. No significant difference was found between the two groups regarding functional ventricle type, time since surgery, age at surgery, early CVP or CVP measured at the time of biopsy.

In conclusion, one-third of patients had histologically proven cirrhosis and two-thirds had extensive fibrosis. There was no evidence of any factor associated with more severe fibrotic changes. Large cohorts are needed for the study of this rare but decisive disease in the prognosis of patients.

Keywords Single ventricle; Fontan; cirrhosis; Heart related liver disease; Hepatocellular carcinoma

Disclosure of interest The authors declare that they have no competing interest.

References

[1] Kiesewetter CH, Sheron N, Vettukattill JJ, Hacking N, Stedman B, Millward-Sadler H, et al. Hepatic changes in the failing Fontan circulation. Heart 2007;93:579—84.

[2] Wu FM, Kogon B, Earing MG, Aboulhosn JA, Broberg CS, John AS, et al. Liver health in adults with Fontan circulation: a multicenter cross-sectional study. J. Thorac. Cardiovasc. Surg 2017;153:656—64.

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P3

Four-Dimensional flow magnetic resonance imaging in cardiovascular diseases: Who can benefit?

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Introduction Four-dimensional flow cardiac magnetic resonance (4D CMR) is an emerging imaging modality for qualitative and quantitative analysis of cardiovascular pathologies. Despite numerous advantages, its distribution and use remain limited suggesting practical or technical difficulties in using this technique routinely. We sought to report our preliminary experience in real life of 4D CMR in unselected children with congenital heart disease (CHD). We focused on its feasibility and ability, compared with adults with CHD as control.

Methods We herein report 50 4D CMR examinations over a one-year study period. This modality has been applied as a complementary imaging when conventional imaging modalities were unsatisfactory. Quality was classified according to qualitative and quantitative criteria by two blinded radiologists.

Results This study included 22 children and 28 adults (mean age 29.5 ± 18.5 years old [0.3; 54], mean weight 61.0 ± 22.7 kg [3.8; 105.0], mean height 159.8 ± 5.5 cm [52; 192]). In infant and children mean age was 10.6 ± 6.1 years old [0.3; 18], mean weight was 36.8 ± 24.5 kg [3.8; 105.0], mean height was 149.4 ± 42.1 cm [52; 192]. Clinical indications were 36% of tetralogy of Fallot, 18% of aorta disease, 27% of complex CHD, 9% of ventricular septal defect and 10% of valvulopathy. The feasibility of this examination was excellent, while 100% exams were performed with no need for general anesthesia whatever the indication or patient (infant, child or adult). Average duration of exams were 465 ± 90s [339—610]. The overall quality of exams was satisfactory; 63% of good quality and 23% of medium quality. The evolution of quality over time has shown a progressive improvement which seems to correspond to a 3-month long learning curve. The only predictive factor identified for quality was the experience ($X^2 = 4.8$; $P = 0.03$ in CHD).

Conclusion Based on our preliminary experience, 4D Flow has become a complementary imaging modality accessible in current practice and open to all patients, infant, child or adult, without restriction of age, weight, size or pathology. The quality of this examination was satisfactory and seems to require an estimated learning curve of 3 months according to our experience.

Disclosure of interest The authors have not supplied their declaration of interest.

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P4

Clinical and genetic data of 20 new patients with SMAD3 mutations type 3 Loeys Dietz syndrome (LDS) and reviews of the literature

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Background Pathogenic variants in SMAD3 (type 3 LDS, Marfan-like connective tissue disorder) cause thoracic aortic aneurysms and dissections, along with aneurysms and rupture of other arteries. Generally, these aggressive vascular damages are associated with multisystemic signs including skeletal abnormalities and premature osteoarthritis. Variable expressivity and incomplete penetrance are commonly associated.

Methods Aortic status, events, and clinical features were abstracted through retrospective review of medical records 20 new patients (28.8 years-old (6—60)) from 8 families from our Reference Centre. After a complete review of the literature, we collected a total of 49 unique variants of different nature (missense, truncating and splicing variants) from 152 individuals of 58 unrelated families. The aim of this study was to look for genotype-phenotype correlations between the mutations of this gene and the severity of the phenotype.

Results Aortic aneurysm and/or dissection are the main vascular findings, affecting respectively 57% and 32% of all type 3 LDS patients described. In our cohort of patients, half presents an aortic dilatation (10/20), 10% an aortic dissection. Three of our patients displays an aortic dilatation during their childhood (at 8 and 7 years old and a surgery for an aortic dilation at 10 years old). Aneurysms and dissections can also be seen in other arteries in 27% (35/138) and particularly intracranial aneurysms found in 20% of patients (21/103), like in two of our patients, one presented an iliac artery dissection. Arterial tortuosity is also frequent, particularly in carotids, representing about a third of the patients (36/117 = 31%), 2 of our 8 patients.

Conclusions SMAD3 pathogenic variants cause thoracic aortic aneurysms and dissections in the majority of individuals with variable age of onset and reduced penetrance. We confirmed that there

is no correlation between the mutation type and the phenotype severity.
Aneurysms-Osteoarthritis syndrome (AOS), genotype-phenotype correlation, heritable thoracic aortic aneurysms and dissections (hTAAD), Loeys-Dietz syndrome, SMAD3, Marfan-like connective tissue disorder, TGFβ pathway.
Disclosure of interest The authors declare that they have no competing interest.

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P5

Surgical closure of the patent ductus arteriosus by anterior mini-thoracotomy in very preterm infants

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Introduction Patent ductus arteriosus (PDA) is an important cause of morbi-mortality in preterm newborns.
Purpose Our study aimed to analyze efficacy and safety of surgical closure of PDA using anterior mini-thoracotomy in very low weight preterm babies.
Materials and methods Monocentric and retrospective study including 21 preterms < 1.3 kgs, who underwent surgical closure of PDA through anterior mini-thoracotomy, between 2010 and 2016.
Results Mean gestational age (GA) at birth was 25.9 ± 1.2 weeks, mean weight at birth was 734 ± 133 gr. Mean age at the time of surgery was 25.4 ± 9.6 days. Mean corrected age and weight at surgery were 29.6 ± 1.6 weeks of GA and 1058 ± 166 gr respectively. 90.5% of neonates had at least one trial of ibuprofen before surgery. 18 patients (85.7%) were ventilated before surgery. Median follow-up was 68.5 days [11 to 273 days] after surgery. No death related to surgery occurred. 3 patients died 49, 65 and 204 days after surgery, due to sepsis, not considered related to surgery. Immediate post-operative echocardiography showed non significant residual shunt in only 1 patient (4.8%), and complete closure in the 20 remaining babies. Median time to extubation was 6 [3—16] days. One patient (4.8%) had a local complication (wound infection) and 5 patients (23.8%) presented transient instability, either hemodynamic ($n = 2$ patients (9.5%)), respiratory ($n = 1$ (4.8%)) or combined ($n = 2$ (9.5%)).
Conclusion Surgical PDA closure using anterior mini-thoracotomy is an effective and safe technique under experienced hands, for PDA closure in very low weight preterm babies. This technique needs to be compared with transcatheter PDA closure currently proposed for those very small babies.
Keywords PDA; Anterior mini-thoracotomy; Very preterm babies; Weight under 1.3kgs
Disclosure of interest The authors declare that they have no competing interest.

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P6

Electrical cardiometry and detection of left ventricular failure in right ventricular heart diseases

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Background Early and easy to do detection of left ventricular (LV) failure is crucial to improve following and outcomes of patients with right ventricular (RV) overload in congenital heart diseases. Electrical cardiometry (Osypka medical) is easy handling, even in medical office or in pre-hospital condition, and can provide cardiac output, and a new contractility index (ICON) supposed to be independent from load conditions. ICON have never been previously challenged to our knowledge.
Objectives We aim to compare ICON with the only contractility parameter independent from load conditions: the elastance slope (Emax).
Methods Using porcine models of Fallot repaired and pulmonary hypertension (PH), we assess LV function using conductance catheter and electrical cardiometry devices over 4 months after surgery. We measured ICON, Emax, Contractile reserve (ΔEmax) and VIC (respiratory variations of ICON) at basal state and after adrenergic stimulation (Dobutamine).
Results Three animals of each group were compared with 6 controls. Non parametric correlation (spearman) hightlights at basal state a non significant and low correlation between ICON and Emax and ΔEmax ($r = 0.5$). However after Dobutamine, correlation is important and strong with $r = 0.98$ between ICON/Emax (0.05) and 0.89 between VIC/Emax. We did not find strong correlation between ΔEmax and VIC or ΔICON.
Conclusion These results obtain on a small in vivo/animal cohort highlight than electrical cardiometry device could be a useful and easy handling (4 skin patchs) tool for LV failure and loss of contractility early screening, specially after adrenergic stimulation and stress conditions. It could provide precious help in patients following.
Disclosure of interest The authors declare that they have no competing interest.

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P7

Are grown-up patients with congenital heart disease and mechanical valve using self-testing INR device? Experience feedback in a French population

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Background The Coaguchek INRange® is a self-testing meter to measure the INR, that is reimbursed in France since August 2017 for patients with mechanical valve.