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.

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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.

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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