velocities at the tricuspid and mitral sides were lower in AoC patients than in healthy group. The E/Ea ratio was significantly higher in patients than in control (6.2±2.1 vs 3.8±1, p<0.000). The LV (left ventricular) ejection fraction was not statistically different between AoC group and controls.

**Conclusion:** In late follow-up despite a satisfactory results after surgery repair of aortic coarctation, left and right ventricular systolic and diastolic performance reveals tendency of decrease.

### 343

**Cardiac mechanics in severely obese children: a 2D speckle strain imaging study**

Eva Zangl (1), Fabien Labombarda [Orateur] (1), Arnaud Pellenzier (1), Dominique Bougle (2), Jean-Christophe Paon (2), Pascale Maragnes (1), Paul Milliez (1), Eric Saloux (1)

**Background:** The prevalence of obesity in children is increasing worldwide. We used 2D speckle strain imaging to investigate whether severely overweight children without hypertension, dyslipidemia, diabetes or sleep apnea, show early cardiac abnormalities. We also investigated the relation between these myocardial features and severity of obesity, fat mass percentage, inflammation and insulin resistance index.

**Methods:** 2D echocardiography, tissue Doppler imaging (TDI) and 2D speckle strain imaging were prospectively performed in obese children and compared to age and sex-matched healthy control subjects. Standard echocardiographic indices of global systolic and diastolic function, early peak diastolic mitral velocity (Ea), longitudinal strain (LS), radial strain (RS) and circumferential strain (CS) were investigated. Z-score body mass index (BMI), body composition, high-sensitive C reactive protein (hs-CRP) and indices of insulin resistance (HOMA-IR) were assessed in the obese children.

**Results:** 32 consecutive obese patients (age: 12.8 [8-17] years; z-score BMI: 5.8 [2.05-8.6]; 15 males; HOMA-IR: 2.1 [0.6-5.7]) were compared to 32 nonobese patients. There was no difference between two groups for left ventricular ejection fraction and conventional diastolic mitral Doppler parameters. Obese subjects showed significantly larger left ventricular wall dimensions (End diastolic diameter: 45±5 vs 43±4 mm; p<0.05; left ventricular mass: 116±31 vs 79±18 g; p<0.005) and signs of early diastolic filling abnormalities on TDI (Ea: 18.1 vs 16.9 cm/s; p=0.02). LS and CS were significantly lower in obese group (LS: –18±2% vs –20±2%; p<0.05; CS: –18±3% vs –20±1±2 %; p<0.05) while RS did not differ. LS and CS were correlated with BMI (Respectively: r=0.5; p<0.05 and r=0.3; p<0.05). There was no correlation between strain parameters and body composition. There was no correlation between strain parameters and hs-CRP. LS was correlated with HOMA-IR (r: 0.45; p<0.05).

**Conclusion:** Obesity in children is associated with significant impairment of longitudinal and circumferential myocardial strain.

### 344

**Molecular patterning of the cardiac outflow tract and coronary arteries of the mouse heart**

Pauline Parisot [Orateur], Magali Théveniau-Ruissiy, Robert Kelly

**Background:** Conotruncal malformations are frequent and often associated with coronary artery anomalies. Coordinated development of the second heart field and cardiac neural crest cells is required for orchestral outflow tract morphogenesis. Defects in one or other cell type result in a spectrum of conotruncal defects observed in human pathology and mouse models. Tbx1, encoding a T-box transcription factor, is the major candidate gene for DiGeorge syndrome and is required for conotruncal development. Tbx1 –/– mouse embryos have a common arterial trunk and proximal coronary artery patterning defects. In the absence of Tbx1 there is a severe reduction in a subpopulation of second heart field cells contributing to subpulmonary myocardium. Sema3C, encoding a neurovascular guidance molecule, is expressed in a Tbx1-dependent domain in subpulmonary myocardium. Sema3C –/– embryos display common arterial trunk with interrupted aortic arch but coronary artery patterning appears normal. Here we present a comparative analysis of the evolution of common trunk in Tbx1 and Sema3C mouse models. These models of truncus display major anatomical differences visible at early stages in the width and the position of myocardial subdomains of the outflow tract and at fetal stages in truncal valve, coronary and great artery patterning. The Tbx1 null phenotype is similar to pulmonary atresia whereas in Sema3C –/- hearts a hypoplastic aorta emerges from the common trunk leading to an interrupted aortic arch. We also investigated potential genetic interaction between Tbx1 and Sema3C, and identified a cryptic early role of Sema3C in pharyngeal arch artery formation revealed in Tbx1+/- embryos suggesting a potential modifier role of Sema3C in DiGeorge syndrome. In addition, we will present data from a microarray analysis of the migestation mouse outflow tract that has identified additional genes expressed in subpulmonary myocardium, potentially contributing to conotruncal and coronary artery development.

### 345

**Safe treatment of infantile hemangiomas with propranolol despite baseline bradycardia**

Bértille Bonniaud (1), Stéphanie Perez-Martin (2), Caroline Bonnet [Orateur] (3), Sylvie Falcon-Eicher (3), Frédéric Huet (2), Pierre Vabres (1)

**Background:** The safety profile of propranolol in infantile hemangiomia (IH) is generally good despite a risk of severe bradycardia. In infants, sinus bradycardia can be a sign of increased vagal tonus, involved in the etiopathology of the sudden infant death syndrome (SIDS), which occurs in the same age group as IH. In addition, infants with IH may combine several other risk factors for syncope or sudden death. Hence, we aimed to determine the frequency of sinus bradycardia in infants before propranolol initiation and cardiac outcome during treatment in children with baseline bradycardia.

**Methods:** Retrospective observational study of infants treated with propranolol for IH from June 2008 to September 2010 at our institution. Prior to propranolol initiation, a detailed baseline cardiological assessment was systematically performed including cardiac ultrasonography and 24-hour electrocardiographic heart rate monitoring. Further at-home long term heart rate monitoring was performed in the infants who had baseline episodes of bradycardia.

**Results:** Significant episodes of sinus bradycardia were found in 3 patients (12.5%), on baseline monitoring prior to propranolol initiation. Cardiorespiratory monitoring at home was thus implemented in these infants and maintained throughout treatment (duration 4 to 8 months). Tolerance was excellent in all three: no further episodes of sinus bradycardia or other adverse events occurred.

**Discussion:** Although asymptomatic, baseline sinus bradycardia may not be uncommon in infants with hemangiomas. It does not seem to preclude the use of propranolol, as no severe sinus bradycardia was noted on propranolol in our series.