Introduction: Transient elastography is a rapid, noninvasive and reproducible approach to assess liver fibrosis by measuring liver stiffness (LS). However, because the liver is enveloped by a capsule, any variation in parenchymal fluid content could theoretically affect LS. LS has been correlated to central venous pressure (CVP) in an animal model. We aimed to determine the correlation between LS and CVP in children and adults with congenital heart disease.

Methods: In this ongoing prospective study, all patients referred for right heart catheterization were included. Measurements of mean right atrial pressure were obtained under general anesthesia (FiO2=21%) using an Optitorque 5 French catheter. The patients underwent 10 LS measurements (median value taken as representative) by transient elastography (Fibroscan, Echosens, France) within the 24 hours before catheterization. The results of LS are expressed in kilopascals (kPa).

Results: Eleven children (mean age=8±6 yo, 54% male) and 6 adults (mean age=34±17 yo, 66% male) have been included so far. Catheterization indications were pulmonary angioplasty (n=5), Melody valve implantation (n=1), fenestration occlusion after a Fontan procedure (n=1) and pre operative assessment of a complex congenital heart defect (n=10). Mean right atrial pressure was 9.1±3.9 mmHg and mean LS was 8.4±4.2 kPa. Correlation between LS and mean right atrial pressure was excellent for these first 17 patients (r=0.87).

Conclusion: Liver stiffness is a new, rapid and reliable method to evaluate CVP in patients with congenital heart disease. This non invasive parameter could potentially be useful for patients in whom CVP play a key role, especially in patients with a Fontan circulation.

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Vitamin K antagonists self management with COAGUCHEK XS® in children: an observational study

Adeline Basquin (1), Valérie Desilles (1), Emmanuelle Comets (2), Jean Marc Schleich (1)

Centre d’investigation clinique (CIC), Rennes, France – (2) Inserm 0203 – CHU Rennes – Hôpital Pontchaillou, Département de cardiologie et maladies vasculaires, Rennes Cedex 09, France

Introduction: The use of COAGUCHEK XS® (Roche Diagnostics), an INR home measuring device, was authorised by the French Health High Authority on June 2008 for children. Nevertheless, there are few data on safety and effectiveness of patients self management particularly in children.

Methods: Since 2008, 23 children (mean age: 9 y; 16 males, 7 females) and their families were trained to INR self testing and self management of vitamin K antagonists (VKA) (acenocoumarol: n=10, fluindione: n=9, warfarin: n=4). INR controls (self testing or lab) were made at expected dates established by the french law. Additional measurements were authorised in case of need by the family or by the health team if needed.

Objectives: To evaluate safety and efficacy in families performing self management in our centre. Time in therapeutic range (TTR), severe haemorrhage (needing hospitalization and/or blood transfusion) and thrombosis recurrence were observed.

Results: Mean follow-up time was 529 days [1.5 y; range 0.3-2.5y]. Mean TTR was 68.5% and was 90.14% when tolerance range was applied. There was no severe haemorrhage or thrombosis recurrence during follow-up and INR >5 were 0.5% (8/1604 measurements). Lastly, 3.5% additional measurements were performed by the families.

Conclusion: VKA self management using COAGUCHEK XS® was safe and effective in our centre. Clinical studies with larger sample and cost-effectiveness considerations are required.

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Experience with levosimendan as an alternative to catecholamines in children

Fahad Alsiohim (1), Roland Henaine (1), Olivier Bastien (2), Magali Veyrier (1), Corinne Ducrueux (1), Olivier Metton (1), Jean Ninet (1), Sylvie Di Filippo (1)

(1) Hôpital cardiovasculaire Louis Pradel, cardiologie pédiatrique et congénitale, Lyon, France – (2) Hôpital cardiovasculaire Louis Pradel, réanimation cardiothoracique chirurgicale, Lyon, France

The objective of this study was to report experience with levosimendan in hemodynamically compromised children.

Material and methods: all patients aged < 18years with severe myocardial dysfunction and dependent on inotrope support, who received levosimendan infusion, were reviewed for clinical, biological and echocardiographic (LV diameters, shortening fraction, sub-aortic Velocity Time Integral) data. Levosimendan was administered as a continuous intravenous infusion over 24 hours, starting 0.1 micrograms per kg per min and increased up to 0.2micrograms per kg per min. Blood pressure and heart rate were continu-ously monitored, ECG and Echocardiography recorded before, 3 and 5 days after starting levosimendan.

Results: From 2007 to 2011, 8 patients aged 3.5 months to 13years (mean 41months) received 10 courses of levosimendan. All were on milrinone sup-port (mean dose 0.63 mcg/kg/mn) and 3 on associated dobutamine. All achieved the 24-hour course of levosimendan without any complication. Catecholamines were successfully discontinued in 5 cases (3 weaned off at 24th hour and 2 at 5th day). Average milrinone dose dropped to 0.3 mcg/kg/mn at day-5, p=0.028). mean shortening fraction increased from 10.8±1.8 before to 12.7± 2.3 at day-3 (p= 0.67), mean Velocity Time Integral from 6±0.6 to 8.7±1.2 cm at day-3 (p= 0.04).

Five patients (62.5%) were successfully discharged from ICU: 2 are doing well at 1 and 4.5 years follow-up, 1 died from metabolic disease and 2 were readmitted at 1.5 – and 2-month interval and received a second course of levosimendan. The latter 2 patients did not improve and needed mechanical ventilator assistance (1 died and 1 underwent successful heart transplantation). For the remaining 3 patients, first course levosimendan failed and heart transplantation was performed in an emergency setting (pretransplant mechanical ventilator support in 2).

Conclusion: This small size sample study shows encouraging results of levosimendan in children with uncontrolled myocardial dysfunction. Further studies are needed to assess monitoring, timing and indications for repeated dose infusion during follow-up.

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Congenital heart disease detected in adulthood

Aldjia Kachenoura (1), Sihem Touati (1), Mohand Said Issad (1), Zakia Bennou (1), Imène Hachmaoui (2)

(1) CHU Béni Messous, cardiologie, Alger, Algérie – (2) Hôpital de Tamanrasset, médecine interne, Tamanrasset, Algérie

Background: Congenital heart defects are often diagnosed in infancy, or even before birth. But some anomalies are harder to detect than others and may not be diagnosed until much later in childhood or even adulthood. In developing countries many congenital heart diseases are detected in adult- hood.

Objective: The aim of this study was to investigate the epidemiology, clinical aspects, diagnosis and treatment of congenital heart disease screened at adulthood.

Methods: This is a retrospective cohort by medical records analysis enrolled between January 2010 and March 2012 in one center. We investig-ated patients with congenital heart diseases aged over 16 years. All patients
underwent a clinical examination, an echocardiogram, an oesophageal echocardiogram or other radiologic (CT or MRI), to specify the congenital heart disease. All patients had an adequate interventional cardiology, surgical care or medical intervention.

**Results:** We included 44 patients, 23 cases of atrial septal defects (ASD) with mean age 38 years (16 women and 07 men), 8 ASD closed surgically and 15 by percutaneous routes.

- 4 patent ductus arteriosus: mean age 32 years (04 women), closed by percutaneous.
- 4 cases coarctation of the aorta, mean age 32 years (4 women), dilated with percutaneous procedure. 9 pulmonary stenosis, mean age: 30 years, all patients underwent balloon dilation. We investigate also one bicuspid aortic with aortic stenosis and 2 Epstein anomaly.

The outcome was favorable for all patients diagnosed during this period.

**Conclusion:** Congenital heart defects discovered in adulthood are not uncommon and should be supported either by medical, surgical, or interventional.

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**Pulmonary arterial hypertension complicating arterial switch surgery for simple transposition of the great arteries—prevalence and outcomes**

Mariléne Lévy (1), Rachel Cordina (2), David Celermajer (2), Pascal Vouhé (1), Damien Bonnet (1)

(1) Hôpital Necker, cardiologie congénitale et pédiatrique, Paris, France – (2) Congenital Cardiology, Sydney, Australia

Pulmonary arterial hypertension after successful arterial switch operation (ASO) for simple transposition of the great arteries is a rare but important cause of late morbidity and mortality. However this complication is poorly characterised in the literature. In Here, we report the prevalence of this condition and its outcomes from the largest series of ASO procedures yet reported.

**Methods:** Between January 1985 and June 2011, 1137 arterial switch procedures were performed for simple TGA at the Hôpital Necker Enfants Malades in Paris. All patients underwent ASO within the early neonatal period. In all cases, pulmonary pressure was evaluated at routine echocardiography and pulmonary hypertension was confirmed by right heart catheterisation.

**Results:** Of 1137 consecutive ASO cases, 8 patients (0.7%, 95% confidence interval 0.29-1.36%) developed severe PAH in the peri-operative period that persisted beyond 1 month post-operatively requiring PAH targeted therapy. Three of those resolved with treatment after 3, 4 and 5 months respectively.

The prevalence of PAH in a general French paediatric population was estimated to be 3.7 per million. These data suggest that long-term survivors of the ASO have a relative risk for the development of PAH around 2000-fold (point estimated to 3.7 per million. These data suggest that long-term survivors of the ASO have a relative risk for the development of PAH around 2000-fold (point estimate 1757-fold, 95% confidence interval 785-3932) compared to children without congenital heart disease.

**Conclusion:** Pulmonary hypertension is a severe complication of TGA which must be rapidly detected in order to initiate targeted therapy. Reversibility is indeed not excluded since last cases of our series have normalized their pulmonary pressure.

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**Transcatheter closure of ASD in infancy with the Amplatzer septal occluder**

Morgan Recher, Francois Godart, Charles Francart

CHU de Lille, hôpital cardiologique, Cardiologie infantile et congénitale, Lille Cedex, France

Many devices are available for atrial septal defect (ASD) usually in adults and children. We report here one center experience in transcatheter closure of ASD in infancy, using the Amplatzer septal occluder. From January 2003 to March 2010, 10 males and 5 female, mean age 10.9±6.4 months, with weight 6.7±2.5 kg underwent transcatheter closure of atrial septal defect. The patients had significant left-to-right shunting (n=12), right-to-left shunting with hypoxemia (n=3). For the left-to-right shunting, the ASD was closed for pulmonary hypertension (n=10), for recurrent chest infection and bronchiolitis (n=5). The ASD was associated with other congenital heart defect (n=5), prematurity and bronchopulmonary dysplasia (n=6), foetopathy (n=1) and encephalopathy (n=1).

The ASD was evaluated by transthoracic echocardiography (TEE) before the procedure. Percutaneous closure was realized with TEE for all patients. They received intravenous heparin. For 4 patients implantation was performed after a balloon test occlusion (mean 11±3.4 mm diameter). The ASO device size was 10.4±3.9 mm. The fluoroscopy time was 7.9±3.9 minutes and the time of the procedure 55.6±21.8 minutes. No per procedural complications were observed for all but for two (one anomalous device deployment and one insufficient rim). These two patients were subsequently surgically repaired.

Patients left the hospital with a low dose of aspirin for 6 months. During follow-up, four patients had a minimal residual shunt on TEE performed up to one year after the implantation. No later cardiac complication was observed.

To conclude, transcatheter closure of atrial septal defect in infant with the ASO is feasible and a possible alternative to surgical closure. In addition, ASD occlusion improves the respiratory status and reduces the level of pulmonary hypertension. Additional long-term results and a large study are both necessary to establish the future of this population.

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**Outcome of patients with “infantile partial atrioventricular septal defects” necessitating surgical repair within the first year of life**

Florent Paoli (1), Virginie Fouliloux (1), Phyll Sokpisai (1), Loïc Macé (1), Bernard Kreitmann (1), Alain Fraisse (2)

(1) Hôpital de la Timone enfants AP-HM, département de cardiopédiatrie, Marseille, France – (2) CHU la Timone, Marseille, France

**Background:** Precise features of infants with partial and transitional atrio-ventricular septal (PAVC) defect requiring surgical repair remain undetermined, as well as their outcome. We retrospectively analyzed this patient population.

**Patients and Method:** Cases with single ventricle palliation were excluded. Since January 1985, 2000, 11 infants (< 1 year of age) underwent surgical repair of PAVC in our institution, at a median age and weight of 7 (2 to 11) months and 6.2 (3.4 to 7.8) Kg, respectively. All patients had refractory congestive heart failure before surgery. Six patients were prenatally diagnosed and 2 had Down syndrome. Three patients had a small left atrioventricular valve (LAVV) with a Z-score diameter < 2. One infant had and associated aortic coarctation necessitating concomitant repair. Other associated cardiac anomalies included persistant left superior vena cava and interrupted inferior vena cava.

**Results:** All the patients survived after surgical repair, with a median stay of 2 (1 to 21) days in the intensive care unit. One patient with a parachute LAVV was reoperated 1 month after surgery for LAVV insufficiency. After a median follow-up of 4.56 (2.6 to 12.2) years, all the patients are in NYHA class I, with no medication. Echocardiography shows moderate LAVV insufficiency in 2 cases.

**Conclusion:** Surgical repair of PAVC in the first year of life is feasible with excellent mid-term outcome, even in cases with a small LAVV. Patients with “parachute like” LAVV may experience significant regurgitation necessitating reoperation.

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**Syncope unit in pediatric population: a single center experience**

Zakaria Jalal (1), Xavier Iriart (2), Maxime De Guillebon (2), Cecile Escobedo (2), Pierre Bordchar (3), Jean-Benoit Thambo (1)

(1) CHU Bordeaux, cardiopathie congénitale – Dr. Thambo, Pessac, France – (2) CHU Bordeaux, cardiologie congénitale et pédiatrique, Pessac, France – (3) CHU Bordeaux, rythmologie et stimulation cardiaque, Pessac, France