tion surface was found ($r = 0.7$). Attachment of the SAM to the mitral valve was present in 14 patients and confirmed by surgical view in 7 patients.

**Conclusion:** 3DE introduces new pre-operative description of the SAM. Such views could optimize the surgical indication and procedure.

![3D echo of sub-aortic membrane viewed from below](image)

### 309

**Decreased myocardial capillary density in failing functionally univentricular heart (FUVH): a morphometric study**

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**Background:** End-stage ventricular dysfunction is the most common cause of mortality in adults with FUVH. The physiopathology of this condition remains unclear.

End-stage dilated as well as hypertrophic cardiomyopathies experience reduced capillary density. In FUVH physiology, volume overload leads to ventricular dilatation, followed by myocardial hypertrophy. We therefore hypothesize that FUVH also demonstrate a reduced capillary density.

**Methods:** Average capillary density was measured in 13 post-mortem human adult hearts: 5 failing FUVH and 8 controls. Hearts were preserved in 10% formalin. Free wall left ventricular longitudinal and transversal tissue specimens were harvested below the level of mitral valve papillary muscles. A morphometric study was realized after immunohistochemical staining of factor VIII. Capillary surface area was quantified by optical density on 50 different samples per patient. The capillary density was calculated: (capillary surface area / total ventricular surface area) $\times 100\%$ using Image J software.

**Results:** Among the 13 hearts, 6 allowed a correct interpretation of the capillary density: 3 FUVH (2 tricuspid atresia, 1 double-inlet left ventricle, all with end-stage ventricular dysfunction) and 3 controls (congenital heart disease with normal left ventricular function: 2 atrial septal defects with Eisenmenger syndrome, 1 tetralogy of Fallot with pulmonary atresia). There was no significant difference in arterial oxygen saturation between the 2 groups ($71 \pm 1\%$ in FUVH vs $85 \pm 0.8\%$ in controls). Myocardial capillary density was significantly decreased in FUVH compared to controls ($3.58 \pm 1.23\%$ vs $7.69 \pm 3.29\%$, $p<0.0001$).

**Conclusion:** Myocardiac capillary density is decreased in end-stage failing functionally univentricular hearts. Yet, further studies with a larger amount of patients are needed to assert the responsibility of the decreased capillary density for the ventricular dysfunction.

### 310

**Evaluation of performance and quality of prenatal diagnosis of congenital heart disease in Indre-et-Loire over a period of 15 years**

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**Objective:** To assess the evolution of prenatal diagnosis (PD) of congenital heart diseases (CHD) for the population of Indre-et-Loire, a French department.

**Method:** Retrospective analysis of the quantitative and qualitative datas of PD between 2000 and 2005 (period 3) and comparison to studies conducted from 1991 to 1994 (period 1) and from 1995 to 1999 (period 2). The CHD considered as detectable are the major CHD with neonatal symptoms.

**Results:** Period 3: 65.7% of detectable CHD were screened. Using the criteria of PD of period 1, the percentage of CHD detected for periods 1, 2 and 3 is respectively 43.2%, 66.7% and 79.3%. For transposition of the great arteries: 25%, 62.3% and 60%. For hypoplastic left ventricle: 80%, 87.5% and 94.1%. Tetralogy of Fallot: 10%, 42.8% and 75%. Pulmonary atresia with ventricular septal defect: not studied, 50% and 100%. Severe coartation of the aorta: not studied, 25%, 40%. Atrio-ventricular septal defect: 77%, 77% and 80%. Diagnoses made by a Pediatric-cardiologist are correct and complete in 80% of cases.

**Discussion:** Performance of PD of CHD is comparable to those reported in the literature. The improvement of PD over time is due to technological progress, greater experience of ultrasonographers and to the systematic visualization of outflow tract in addition to the four chamber view.

**Conclusion:** Despite the improvement of PD, some life threatening CHD are not screened before birth and require a specific training program for the ultrasonographers.

### 311

**Home INR monitoring of oral anticoagulation therapy in children using a point-of-care INR monitor after an education program**

Fanny Bajolle (1), Dominique Lasne (2), Caroline Elie (3), Damien Bonnet (4)


**Objective:** To assess the evolution of prenatal diagnosis (PD) of congenital heart diseases (CHD) for the population of Indre-et-Loire, a French department.

**Method:** Retrospective analysis of the quantitative and qualitative datas of PD between 2000 and 2005 (period 3) and comparison to studies conducted from 1991 to 1994 (period 1) and from 1995 to 1999 (period 2). The CHD considered as detectable are the major CHD with neonatal symptoms.

**Results:** Period 3: 65.7% of detectable CHD were screened. Using the criteria of PD of period 1, the percentage of CHD detected for periods 1, 2 and 3 is respectively 43.2%, 66.7% and 79.3%. For transposition of the great arteries: 25%, 62.3% and 60%. For hypoplastic left ventricle: 80%, 87.5% and 94.1%. Tetralogy of Fallot: 10%, 42.8% and 75%. Pulmonary atresia with ventricular septal defect: not studied, 50% and 100%. Severe coartation of the aorta: not studied, 25%, 40%. Atrio-ventricular septal defect: 77%, 77% and 80%. Diagnoses made by a Pediatric-cardiologist are correct and complete in 80% of cases.

**Discussion:** Performance of PD of CHD is comparable to those reported in the literature. The improvement of PD over time is due to technological progress, greater experience of ultrasonographers and to the systematic visualization of outflow tract in addition to the four chamber view.

**Conclusion:** Despite the improvement of PD, some life threatening CHD are not screened before birth and require a specific training program for the ultrasonographers.

**Introduction:** The literature demonstrated that children receiving anticoagulation therapy require more frequent monitoring than their adult counterparts due, in part, to the complexity of their underlying medical conditions. Since June 2008, point-of-care International Normalized Ratio (POC INR) monitor are refund by Social Security in France for paediatric patients. The aim of this study is to evaluate the outcomes of home INR monitoring in children through a standardized education and training program.

**Materials and methods:** The POC INR monitor is refund for under eighteen patients with a cardiologic or non cardiologic indications. Participating parents had to complete a one-day intensive education and training program.
After demonstrating theoretical and practical competences, parents commenced home monitoring. We analysed the proportion of INR within the target range, the incidence of anticoagulant related adverse events and the quality of life of children and parents.

**Results:** We included 48 children (between 2 months and 17 years old). Eighteen patients had a mitral valve replacement and 6 an aortic valve replacement. Fifteen patients had a total cavopulmonary derivation, 7 a dilated cardiomyopathy, 1 coronary aneurysms after Kawasaki disease and 1 an antiphospholipid syndrome. Twenty-eight patients received anticoagulation therapy for the first time and 20 had been treated for a long time and switched from venipuncture to point-of-care monitor. The results of the theoretical test of parents were excellent (average 17.3/20). Only 3 families had practical difficulties at home. 65% of INRs were within the target range. There were no haemorrhagic and thrombosis complications. The quality of life was improved.

**Conclusion:** This education and training program was perfectly safe and improved the quality of life of patients and parents. The use of similar education program may serve to improve the outcomes of home monitoring for anticoagulation therapy in children.

### 312 Infective endocarditis in adults with congenital heart disease

**Sylvie Di Filippo (1), Roland Henaine (1), Marielle Gouton (1), Jean Ninet (2), François Sassolas (1), Corinne Ducreux (1), André Bozio (1)**

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The aim of this retrospective study was to describe features of infective endocarditis (IE) in adults with congenital heart disease (CHD).

**Methods:** The records of all episodes of IE diagnosed from 1974, in patients with CHD and more than 18 years of age at diagnosis, were retrospectively reviewed.

**Results:** Forty-four episodes of IE occurred, 36 after 1990 (81.8%), 28 males (63.6%). Age at diagnosis was 30.3±9 years (median 28 years). Ten were recurrent episodes (22.7%). CHD was previously repaired in 15 cases (34%), palliated in 7 (16%) and non-operated in 22 (50%). Dental causes were predominant (34%), followed by cutaneous causes (25%); others were postoperative (4.5%), miscellaneous (7%) or unknown causes (29.5%). A microbial agent as identified in 95.4% of the cases: oral streptococcus and staphylococcus aureus were the leading causative agents (respectively 41% and 36%). Left heart locations were predominant (75%). Severe clinical cardiac complication occurred in 10 cases (23%), an echocardiographic complication in 18 (40%). Twenty-four patients experienced embolic events (54.5%); early surgical treatment was required in 25% of the cases. Three patients died due to IE (6.8%). Antibiotic prophylaxis had been neglected despite known risk in 41% of the cases.

**Conclusion:** IE is an ongoing life-threatening complication in adults with CHD, with a significant morbidity, a high rate of prophylaxis negligence and of recurrence.

### 313 French pulmonary arterial hypertension registry in children: 2-year follow-up data

**Alain Fraisse (1), Claire Dauphin (2), Sylvie Di Filippo (3), Jean Marc Schleich (4), Michel Voisin (5), Pascal Maragnes (6), François Godart (7), Xavier Jais (8), Didier Tardy (9), Pierre Clerson (10), Philippe Acar (11), Damien Bonnet (12)**


**Introduction:** Pulmonary arterial hypertension (PAH) is a devastating disease with poor survival but there are limited data describing its impact in the paediatric population.

**Objective:** The objectives of this national prospective registry were to collect clinical and epidemiological data and to investigate the outcome of children with PAH.

**Methods:** All consecutive patients <18 years with PAH seen in 16 referral PAH centres in France were included and evaluated after 1 and 2 years of follow-up for WHO functional class (FC), 6-minute walk distance (6MWD) and quality of life (QoL: CHQ-PF50 questionnaire). Persistent pulmonary hypertension of the newborn and PAH due to congenital heart disease (CHD) were excluded.

**Results:** Fifty patients were included between May 2005 and June 2006. Mean age at diagnosis was 8.7±4.4 years and male/female ratio was 1/1. The prevalence of pediatric PAH was 4.2 cases/million. A history of prematurity, cancer and major surgery was noticed in respectively 19%, 6% and 4% of patients. At inclusion, 28% of patients were in WHO FC III or IV. Aetiology of PAH was idopathic in 60%; familial in 10%, associated with but not due to CHD in 24%, related to connective tissue disease in 4% and to portal hypertension in 2%.

Nine patients (18%) died during the 2-year follow up. Survival estimates at 1 and 2 years were 86% and 82%. Seventy three percents of patients improved or did not change WHO FC (n = 44). Patients remained stable regarding 6MWD (n = 25), hemodynamics (n = 11), and QoL (n = 19). During the 2-year follow-up, combination of PAH-specific therapies was increasingly prescribed (44% patients vs. 22% at inclusion).

**Conclusions:** This first national paediatric registry showed (1) the presence of multiple PAH aetiologies in children, that could be different from those seen in adults, (2) the association with other co-morbidities, and (3) the stabilization or even improvement in patient’s condition with the therapeutic management recommended in the current era.

### 314 Outcomes and prognosis in patient with TGFBR2 or FBN1 gene mutation

**David Attias (1), Chantal Stheune (1), Caroline Roy (2), L. Faivre (3), Gwenaelle Collod-Beroud (4), Delphine Detaint (1), M.A. Dherue (5), H. Plauchu (6), Mireille Claustres (4), S. Lyonnet (7), A. Vahanian (8), Catherine Boileau (1), Guillaume Jondeau (1)**


**Background:** TGFBR2 mutations were recently recognized among patients with a Marfan-like phenotype and have been associated with poor cardiovascular prognosis (early deaths, aortic dissections or early surgery for thoracic ascending aorta aneurysm).

**Methods:** Clinical features and outcomes were analysed in a group of 71 patients (including 22 children) with a TGFBR2 mutation. Patients were compared with age- and gender-matched patients harbouring the FBN1 mutation (FBN1 group n=243).

**Results:** The proportion of patients with aortic dilatation was similar in the TGFBR2 group and in the FBN1 group in both adults (74% vs. 76%) and children (36% vs. 86%) and aortic dilatation was very variable in both TGFBR2 and FBN1 groups.

The incidence and average age of thoracic aortic surgery for aortic dissection or aortic dilatation were similar in the 2 groups (31% vs. 27% and 35±16 vs. 39±13 years, NS).

Both the frequency and the age of occurrence of aortic dissection were similar in the TGFBR2 and the FBN1 groups for the ascending aorta [14 % vs. 10 %, p=0.26; 38±12 vs. 39±9 years, p=0.82] and for the descending aorta (3/7 vs. 8/243, p=0.72; 34±6 vs. 44±10 years, p=0.16).