Introduction: Syncopesthes are frequent in the pediatric population. The majority is benign but, for a minority of children, a cardiac disease is the underlying cause and has to be recognized as it can be fatal. Syncope units developed in adult population have demonstrated major improvement in diagnostic process, hospitalisation reduction time, with favourable long-term outcome. We report our experience of syncope management in children and adolescents through a dedicated syncope unit.

Methods: In this ongoing prospective study, we enrolled 31 consecutive patients (13±3 yo, 65% male) between Jan 2011 and Mar 2012, referred for loss of consciousness (LOC) in a dedicated pediatric syncope unit involving a pediatric cardiologist, a nurse, a physiotherapist and a psychologist. All patients underwent initial evaluation including medical history assessment, physical examination, 12-lead ECG and echocardiography to exclude non-cardiogenic syncope. If initial assessment was abnormal, they underwent targeted tests that differed according to suspected aetiology. Patients with neurocardiogenic syncope underwent specific physiotherapy training and a consultation with a psychologist.

Results: The most common cause of LOC was neurocardiogenic syncope – 22 patients (71%), psychogenic LOC – 7 patients (23%). One patient (3%) had a long QT syndrome and received beta blocker therapy. One patient had typical epileptic seizure and was transferred to neurologic department. Mean hospitalization duration was 9±6 days. Head-up tilt testing was positive in 62% neurocardiogenic syncope. Echocardiograms and exercise tests were not contributive for diagnosis. After a mean follow up of 9±4 months, including physiotherapist and/or psychologist specific care, syncope recurrence occurred in 4 patients (12%).

Conclusion: Syncope unit in pediatric population with dedicated team improves diagnostic process, reduces hospitalisation and decreases syncope recurrence when adapted follow up is proposed.

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ExtraCorporal Membrane Oxygenator support in the perioperative course of pediatric heart surgery

Zied Matoussi (1), Roland Henaine (1), Olivier Bastien (2), Magali Vey (3), Corinne Dene (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 aim of this study was to assess the results of ECMO as a perioperative support in children with congenital heart disease (CHD) during and after open-heart surgery.

Material and methods: All patients aged < 18 years at surgery who needed ECMO support from 2004 to 2011, were included in the study. Clinical and biological data, demographics and outcomes were retrospectively reviewed.

Results: Twenty-seven patients (19 males) (i.e. 0.5% of total pediatric cardiac surgical procedures performed per year), aged 3days to 18years (mean 16.6years) were placed on ECMO, per-operatively in 10 (37%, for failure to wean off bypass) or during the early postoperative course in 17 (8 >24 hour, 9 >24th hour) for cardiac arrest (33%) or low cardiac output (30%). Surgical repair included: severe form of tetralogy of Fallot (4), complex arterial switch operation (7), complex left heart obstruction (5), Rastelli (4), ALCAPA (1), cavopulmonary anastomosis (3) and miscellaneous (3). Twelve cases were in-hospital preoperatively, of whom 7 were dependent on mechanical ventilatory support.

Five patients died while on ECMO because of multi organ failure (4) or pulmonary hypertension (1). Main complications during support included hemorrhages (15 cases), renal failure requiring peritoneal dialysis (14), hemothysis (13), canulas thrombosis (6), and strokes (4). Only 3 cases were free from complication. Duration of ECMO was 5±± 6days (1 to 16, median 5), of CICU stay 26±16days (10 to 69, median 22). Survival to ECMO was 81.5% (22 patients) and overall survival was 95% (16). Significant predictive factors for mortality were: preoperative clinical status (in-hospital: 25% in alive patients versus 73% in deceased cases, p = 0.011), lactates level at onset of ECMO (mean 6 versus 10, p = 0.004) and duration of aortic clamp (mean 70 versus 110mm, p = 0.05).

Conclusion: This study shows that post-cardiotomy ECMO in children is a valuable therapeutic option as a bridge to recovery, despite high frequency of complications on support.

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Myocardial bridge

Naima Baaddy (1), Salma Fadili (2)

(1) Faculté de médecine HassanII Casablanca, cardiologie, Casablanca, Maroc – (2) Faculté médecine HassanII, Casablanca, Maroc

The myocardial bridge often congenital, is an aberrant relationship between the trunks and epicardial coronary infarction. Although often asymptomatic, this detection coronary angiography (effect of “milking” the compression stroke) was associated with ischemia, heart rhythm disorders and sudden death.

Ten clinical cases have prompted a review of literature on the morphological and functional alterations that characterize patients with myocardial bridge and the current diagnostic and therapeutic method.

The investigative tools currently available in the catheterization laboratoires have clarified how the symptoms and signs of ischemia may occur in these patients, especially when the only angiographic findings appear to be a compression stroke or effect “milking” of a coronary. The quantitative coro coronary angiography and IVUS have clearly demonstrated that stisolic compression phase observed in the angiogram is coupled with a persistent diastolic diameter reduction. The intra coronary Doppler revealed an increase in the speed of retrograde flow and reduced coronary reserve. The diagnosis can be established by a significant reduction in the percentage of luminal diameter with loss of surface, increased coronary and characteristic flow such appearance in IVUS lunate aspect and “finger tip” of the diastolic flow. Early in the intra-coronary Doppler. Medical treatment, interventional catheterization and surgery not only allow a significant reduction or normalization of these alterations, but also a significant improvement in signs of ischemia and angina episodes. The low rate of clinical manifestations and the variability of the morphological, functional and clinical preventing the establishment of solid recommendations for diagnosis and treatment, in light of published cases in the literature that concern only a limited number.

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Ebstein’s disease in adulthood.

Leila Bazdah, Hedi Baccar, Slim Siddhom, Wejdene Ouechtati, Imene Fradi, Hbib Ben Ahmed, Sami Marouane, Meriem Drissa

Hôpital Charles Nicolle, cardiologie, Tunis, Tunisie

Introduction: Ebstein’s disease is a rare congenital malformation with many clinical manifestations depending on the age at diagnosis and the severity of the anatomical pattern. In this study we try to describe the clinical manifestations, the echocardiographic patterns and the outcome of this malformation in adulthood.

Methods and results: Retrospective study about 8 cases of Ebstein’s anomaly in adult patients (5 women and 3 men); the mean age is 34 years old. The clinical manifestations were absent in 6% of cases but the majority of patients have exertional dyspnea or palpitations in 56% of cases. Arrhythmias and congestive heart failure were observed respectively in 6% and 13% of cases. Transthoracic echocardiography demonstrated the presence of Ebstein anomaly in all cases, showing apical displacement of the septal leaflet of the tricuspid valve. Five patients had the A grade of the Carpenter’s classification and the others had the grade B. After an outcome of 11 years, one patient died by refractory heart failure.

Conclusion: Clinical manifestations and outcome of Ebstein’s anomaly depend on the degree of tricuspid valve malformation and many cases are discovered in adults. The prognosis is worsening by arrhythmia and heart failure. Surgical intervention with tricuspid valve repair should be proposed for patients with severe heart failure and intractable arrhythmia. Conservative surgery and anti arrhythmia therapy had enhanced the prognosis of this congenital malformation.