Antiphospholipid syndrome (APS) is one of several prothrombotic states in which thrombi occur within both the venous and arterial beds. A minority of patients with APS present with an acute and devastating syndrome characterized by multiple simultaneous vascular occlusions throughout the body. “Catastrophic APS” is defined by the clinical involvement of at least three different organ systems over a period of days or weeks with histological evidence of multiple occlusions of large or small vessels. We report the case of a 16-year-old girl referred to our paediatric cardiology unit for NHYA IV and a blowing systolic murmur at the apex radiating to the left axilla. Transthoracic echocardiography (TTE) revealed mitral valve leaflet thickening with vegetations (13 mm) on the edges of both leaflets and moderate mitral regurgitation (MR) (image 1). The diagnosis of Libman-Sacks or non-bacterial thrombotic endocarditis secondary to antiphospholipid syndrome was suggested by repeated negative blood cultures along with persistently elevated anticardiolipin antibody titers. The condition deteriorated to acute thrombotic microangiopathy affecting multiple organs with arterial hypertension and thrombocytopenia. Anticoagulation with warfarin was performed and aspirin, corticosteroids and ACE inhibitors were given. TTE follow-up after 1.5 years revealed no recurrence of MR with normal mitral valve leaflets.

Conclusion: In catastrophic APS an aggressive therapeutic approach is warranted. Valve lesions may become more severe during long term follow-up.

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Exercise testing coupled with Doppler echocardiography: a simple, safe and effective method to assess the severity of coarctation of the aorta.

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Peak systolic Doppler velocity at the aortic isthmus alone is not a good predictor of severity in patients with native or residual coarctation (CoA). A major argument in favour of a hemodynamically significant CoA is the presence of a diastolic gradient (DG) with characteristic “sawtooth” appearance of the Doppler pattern.

Aim of the study: To evaluate, in patients with some degree of aortic isthmus narrowing but without significant DG at rest, if exercise could unmask DG and thus hemodynamically significant CoA.

Material and methods: Fourteen patients aged from 12 to 56 years underwent treadmill exercise testing coupled with Doppler echocardiography. Thirteen had previous CoA repair 8 to 43 years before, one had aortic kinking with mild isthmic narrowing on CT-scan. MRI or CT-scan were performed in 11 patients, showing significant residual stenosis (narrowing >30%) in 8/11. Doppler measurements were performed during exercise testing and 5 minutes after at the suprasternal notch, using a CW-Doppler 2 MHz pedoflux probe. The peak systolic and diastolic gradients through aortic isthmus were measured at rest and at the end of the exercise. A peak DG > 17 mmHg was considered significant.

Results: Mean peak systolic gradient increased from 29 to 65 mmHg with exercise. Significant DG appeared in 4 of 8 patients without any DG at rest. Among the 6 patients with non significant DG at rest, DG remained non significant in 2 and became significant in 4. For the 11 patients with MRI or CT-scan aortic arch evaluation, the specificity of significant DG at exercise was 100%, the sensitivity 75%, predictive positive value 100%, negative predictive value 60%.

Conclusion: Apparition of a significant isthmic Doppler DG at exercise can reliably predict the hemodynamic significance of restenosis after CoA repair, as well as native narrowing of the aorta. This simple, safe and effective non-invasive method may help to identify patients requiring surgery or percutaneous stenting.

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Levosimendan in dilated cardiomyopathy and refractory cardiogenic shock in children.

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Introduction: Levosimendan (Lev) is a new calcium sensitizer and K-ATP channel opener. The documentation regarding this drug is one of the largest ever on the safety and efficacy of a new pharmacological agent in acute heart failure syndromes in adult population. Its use in paediatric is limited to successful weaning from biventricular mechanical support in case reports or small trials conducted in the immediate postoperative period. We report our experience of using (Lev) during refractory cardiogenic shock (RCS) in infants.

Patients and Methods: Four infants aged 2-24 months and suffering from hypokinetically dilated cardiomyopathy were included in this study. All presented with uncontrolled RCS (LVEF <20%) despite conventional inotropes treatment. Lev was intended as a last resort before ECMO. The effectiveness of treatment was monitored by measuring the echocardiographic left ventricular ejection fraction (LVEF) and plasma BNP assay before and 8 days after administration. A total of 15 injections of Lev were realized and studied.

Results: Mean LVEF before and the day after administration of Lev significantly increase from 19.75 % +/- 1.7 to 33 +/- 2.95 (p<0.01) and mean Lev BNP level decrease from 2267 pg/ml +/- 518 to 1673 +/- 372 (p<0.08). These children could benefit from additional treatments of Lev in order to wean amines and defer the circulatory support. In one of these, a total of 6 cycles was necessary without the use of circulatory support for an improvement. In the 4 patients studied, the outcome was favorable. Figure 1 illustrates the individual changes in LVEF and BNP after each Lev injection.

Conclusion: During the refractory cardiogenic shock of the child with hypokinetically dilated cardiomyopathy, levosimendan may improve myocardial function allowing weaning of conventional inotropes and circulatory support. Re-injection may also be necessary. Randomized studies with larger number of patients are needed to confirm these very encouraging results.

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Transcatheter closure of large patent ductus arteriosus using the Amplatzer duct occluder in infants under 2 years of age

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Introduction and objectives: Percutaneous closure of patent ductus arteriosus (PDA) is a well-established technique. We evaluated the usefulness of the Amplatzer duct occluder (ADO) for the percutaneous closure of large patent ductus arteriosus in 16 children less than 2 years of age.