SCIENTIFIC LETTERS

Mitral valve replacement in infants less than 6 months-old

Prótesis mitral en pacientes de menos de 6 meses

Dear Editor:

Congenital mitral insufficiency (MI) is rare in children. It is usually associated with other heart malformations, and the isolated form occurs much less frequently. Mitral valve replacement (MVR) is an uncommon procedure in the paediatric age group, especially in infants. The most frequent indications for MVR are rheumatic disease, endocarditis, Shone syndrome or a failed atrioventricular canal repair. Despite surgical advances and the use of different materials, MVR is a complex surgery associated with a high rate of complications, and is the procedure for mitral repair that carries the highest mortality (5–52%) and the worst prognosis.

We present the cases of two patients that received care in our hospital and required MVR before age 12 months and reintervention shortly after, which by chance occurred while receiving maintenance treatment with heparin.

A pregnant woman was referred to our hospital for foetal cardiomegaly at 36 weeks’ gestation. Assessment revealed global cardiomegaly, severe tricuspid insufficiency and moderate mitral insufficiency with normal ventricular function and a closed foramen ovale (FO). Premature closure of the FO was diagnosed that required urgent surgery. The neonatal echocardiogram confirmed these findings, with the most salient feature corresponding to a highly dysplastic mitral valve with pulmonary pressures exceeding systemic pressures. The patient did not respond well to treatment and was transferred to the surgical department at age 8 days. Conservative measures were attempted unsuccessfully, so at age 40 days the dysplastic (myxomatous) mitral valve was resected and a 16 mm mechanical prosthetic mitral valve placed in the supra-annular position (CarboMedics, Sorin Group®). The patient had a cerebral infarction at the level of the middle cerebral artery as a complication of surgery. The patient was kept under anticoagulant therapy with heparin, administered by the subcutaneous route given his age. At age 3 months he developed a severe pulmonary oedema secondary to the immobilization of the leaflets by pannus formation, requiring a prosthesis exchange. At age 11 months he had a new episode and underwent surgery again for resection of the pannus and thrombi, and the prosthesis was cleaned and rotated after failure of treatment with fibrinolytic agents. Treatment with acenocoumarol and acetylsalicylic acid (ASA) was initiated after the surgery. The patient has remained asymptomatic since, and is currently 3 years old.

Infant aged 5 months with no relevant history referred by his paediatrician for investigation of a heart murmur and growth failure. The echocardiogram showed severe MI with a retracted posterior leaflet, thickening of the free edge, and significant dysplasia of the subvalvular apparatus with dysfunction of the left ventricle. It also revealed retrograde flow in the left coronary artery, suggestive of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA syndrome). This suspicion led to transfer of the patient to the reference hospital, where the diagnosis was confirmed and coronary translocation from the pulmonary artery to the aorta performed along with a mitral valvuloplasty. At 24 days he showed clinical worsening, and underwent surgery again for placement of a mechanical mitral prosthesis (CarboMedics 18, Sorin Group®). Treatment with heparin was initiated. Six months later, he had a sudden worsening of symptoms with signs of pulmonary oedema, and investigation confirmed the immobilization of the valve leaflets. A new intervention was performed, with resection of the pannus and cleaning and rotation of the prosthesis. The patient was subsequently put on acenocoumarol and ASA. The patient, currently aged 3 years, remains asymptomatic.

The surgical management of MI poses a challenge to the paediatric surgeon due to its considerable complexity. Valve repair is the first-line treatment, as it has the best outcomes, although it is not always feasible. Biological prostheses have the advantage of not requiring anticoagulant therapy, but they degenerate faster. When it comes to mechanical prostheses, the literature has identified the size of the ring and the position of the valve (supra-annular or annular) as factors that influence survival and the need for early reintervention (stenosis, endocarditis and thrombosis). It is imperative that patients are treated with vitamin K antagonist anticoagulants after the surgery.

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In our patients, treatment with heparin was chosen because the adherence to oral administration is worse in children younger than 12 months. Pannus formation (a fibrous tissue overgrowth) was the reason for early reintervention in both patients. This complication is uncommon in the paediatric literature. Although it does not seem to be related to anticoagulant therapy, in both of our patients it developed while they were undergoing treatment with heparin, and did not recur when the patients were treated with a combination of a coumarin drug and ASA.

In short, MI in the paediatric age group continues to be a challenge. Mechanical prostheses have the disadvantage of requiring chronic anticoagulant therapy, which is difficult to adhere to, especially in infants.

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References


M. Colom Seguí, a, b, S. Escrivá Bori, c M.A. de la Fuente Sánchez a, b, F. García Algas b

a Servicio de Pediatría, Hospital Universitari Son Espases, Palma de Mallorca, Spain

b Unidad de Cardiología, Servicio de Pediatría, Hospital Universitari Son Espases, Palma de Mallorca, Spain

c Corresponding author.
E-mail address: marinacolomsegu@gmail.com (M. Colom Seguí).

Typical haemolytic uraemic syndrome: Report of the first case due to Escherichia coli O26:H11 in Spain

Síndrome hemolítico urémico típico: comunicación del primer caso producido por Escherichia coli O26:H11 en España

Dear Editor:

Haemolytic uraemic syndrome (HUS), while rare, is one of the main causes of acute renal failure in the paediatric age group. It is characterised by the development of haemolytic anaemia, thrombocytopenia and acute renal failure. Most cases present with diarrhoea (so-called typical HUS), with Shiga toxin (Stx)-producing Escherichia coli (E. coli), especially serotype O157:H7, being the most frequent aetiological agent. We are presenting a case caused by the O26:H11 strain of enterohaemorrhagic E. coli.

The patient was a girl aged 2 years presenting with fever, vomiting and watery stools with fresh blood of three days’ duration, with asthenia and oliguria in the previous 48 h. The relevant findings of urgent laboratory testing were: haemoglobin, 9 g/dL; platelets, 67,000/μL; creatinine, 3.9 mg/dL; urea, 162 mg/dL; uric acid, 15.9 mg/dL; lactate dehydrogenase (LDH), 5397 IU/L; and 4–5 schistocytes per high-power field in the peripheral blood smear. The patient was admitted to the paediatric intensive care unit for suspected HUS, and her persistent anuria required initiation of continuous venovenous haemodialfiltration, which was followed by peritoneal dialysis for a total of 21 days of blood purification. The complement levels were decreased at diagnosis (C3, 62.1 mg/dL; C4, 10.5 mg/dL) and had normalised three days later, including properdin (28.6 mg/dL) and CH100 (1.125 U/mL) values. The detection of anti-ADAMTS13 antibodies was positive (43.9 AU/mL), although ADAMTS13 activity was normal (64.5%). The investigation was completed with a stool culture that was negative for enterohaemorrhagic E. coli. Due to this result, the sample was analysed by PCR and found to be positive for verotoxigenic E. coli, and genetic characterisation and serotyping identified the isolate as strain O26:H11.

E. coli strain O26:H11 has been an emergent pathogen in Europe since it was first isolated in Germany in 1990. It has been increasingly isolated in cases of HUS to eventually become the second most frequent cause (15–19%) following serotype O157:H7, even surpassing this serotype in some countries between 2008 and 2012. It can also produce two types of Stx, of which Stx2a is strongly associated with the development of HUS. Furthermore, there is evidence that children are more frequently infected by stx2a-harbouning strains, and thus are more likely to develop HUS.

The detection of enterohaemorrhagic E. coli O26:H11 by PCR in our patient constitutes the first time that this strain is found in a child with HUS in Spain. This serotype is associated with infection in younger children, and the form of HUS that it causes may have a similar severity and clinical course to the form produced by O157:H7. The patient was kept in isolation during the early weeks of hospitalisation, and is currently awaiting negative stool culture results to resume her schooling.