Surgical downsizing of a prosthetic ring in congenital mitral valve regurgitation

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DESCRIPTION

Isolated congenital mitral valve regurgitation is an extremely rare disease in children in the first year of life.¹ It can progress quickly to severe regurgitation, leading to left ventricular enlargement and remodelling, with decline in ejection fraction.² In such cases, the dilated annulus and leaflet abnormalities are complex and difficult to manage. Because it is usually complex, intervention is ideally postponed allowing time for annular growth and tissue maturity.³

We present a case of a 2-month-old girl with a history of poor feeding, respiratory distress, pallor and poor weight gain. The baby was evaluated by paediatric cardiology due to a heart murmur grade IV/VI on Levine scale. Transthoracic echocardiography showed an arcade mitral valve (Hammock) with major valvar regurgitation, dysplastic leaflets, with incomplete coaptation, an extremely dilated valvar ring (z-score +9.54)⁴ (video 1) and massive dilation of the left atrium. There were no other valve or coronary artery abnormalities. Even though there was some improvement with anticongestive medication, she remained on cardiac failure and was proposed for surgical intervention at 6 months of age. Informed consent was obtained from parents prior to the intervention, concerning the need for an intraoperative modifying device.

Surgical findings: huge circular mitral annulus (26 mm), a large anterior leaflet, thick and yellowish pliable tissue, anomalous subvalvular apparatus with a large anterior papillary muscle attached to a small posterior papillary muscle and short chordae. The surgical intervention consisted of longitudinal division of both papillary muscles, resulting in three separate heads and mitral annuloplasty. Because we were aiming for anteroposterior reduction without too big an annulus size reduction, we chose a rigid Carpentier Edwards Physio Ring. As the smallest available size was too large for this patient, the posterior part of the ring was cut out over a length of 12 mm. The annulus was bent, the two ends were

Video 1 Transthoracic echocardiogram before and after surgery, from an apical four-chamber view with and without colour Doppler.

Video 2 Transoesophageal echocardiogram before and after surgery, with mitral valve with and without colour Doppler.

joined together and covered by its own Dacron tissue. The final diameter was approximately 20 mm maintaining the original shape (figure 1). Transthoracic echocardiogram after surgery showed a vast reduction of mitral regurgitation and better leaflet coaptation (video 2).

Two years after the surgery, the child has reached the 75th centile of weight, progressively with lower symptoms of cardiac insufficiency, maintaining captopril therapy for minor mitral regurgitation

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Figure 1 Carpentier Edwards Physio Ring transformation during surgery.

Patient's perspective

As parents, we were extremely concerned about our baby's failure to thrive during the first months of life. As soon as she initiated medication, she improved but we were hopeful on surgery results. The fear of losing her was always present but steeped after the surgery, as she started to gain weight and feel much better. We are very grateful to all the medical teams that did the best for our little child.

Learning points

- ▶ Anticongestive medication should be optimised but can be insufficient to support adequate thrive until the ideal intervention time.
- ▶ Although there are no consensual surgical treatments for congenital mitral regurgitation, all efforts should be made to preserve the native valve.
- ▶ Modifications of prosthetic material can be safe and life-saving.

with no stenosis (median mitral gradient 4.9 mm Hg) and systolic pulmonary artery pressure of 40 mm Hg.

Mitral regurgitation is extremely rare among congenital heart diseases. For that reason, surgical management may not be consensual or strictly established. Native valve preservation ought to be always aimed for. This case clearly exemplifies how medical treatment was insufficient and surgical treatment was effective. Each rare case must be evaluated by a multidisciplinary experienced team to best portray its treatment.

Contributors RAS and SC have collected all data (images, medical records, etc), took care before and after surgery, during ICU internment. IM made the diagnose and is her main doctor. JN was the main surgeon who operated the baby.

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