Mitral valve replacement using a handmade construct in an infant

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Abstract

We recently reported the feasibility of a handmade left atrioventricular valve using decellularized extracellular matrix for 3 months in a failed repair of AV canal defect. We present the use of an extracellular matrix mitral valve for 9 months in an infant with an un-repairable congenitally malformed valve. The valve functioned perfectly without anticoagulation until the patient expired from non-cardiac complications of their fibrillin gene defect. This report adds further experience using valves made from extracellular matrix in the systemic ventricle both in terms of application and in terms of short-term durability of the construct.

Keywords: Extracellular matrix • Infant • Valve • Mitral • Replacement

INTRODUCTION

Options for mitral valve replacement remain disappointing in the paediatric population with very little promise for commercial development. Using a technique described by Dr Cox and associates [1], we describe successful replacement of the mitral valve in a 4-month-old infant who survived for 9 months.

CASE REPORT

The patient presented with heart failure which was believed to be secondary to severe right and left AV valve insufficiency, but without an underlying diagnosis at the time. There was massive right and left atrial dilation with atrial arrhythmias, and she required intubation shortly after presentation. Despite optimal medical management, the child’s clinical condition deteriorated and it was decided to attempt repair of both valves surgically as a life-saving effort.

At operation several important findings were noted; there was severe bullous emphysema throughout both lungs and the cardiac tissue quality was quite delicate, and friable. The tricuspid valve was dilated with poor apposition. The mitral valve leaflets were dysplastic, the anterior leaflet being curled under, severely thickened and shortened with an annular size of 12 mm. The tricuspid valve was repaired with a DeVega annuloplasty stitch with good result; however, despite using a Kaye annuloplasty and anterior leaflet extension with extracellular matrix (ECM) patch, the mitral valve was irreparable. The valve was, therefore, resected including the subvalvular apparatus, and a new ECM mitral valve placed using our previously described technique [2]. Briefly, a 4-ply ECM patch (CorMatrix) was used to fashion the valve by folding over the top edge of the sheet to make a 4-mm cuff. The sheet was then wrapped around a 12-mm dilator (see Fig. 1). The folded-over annulus of the neovalve was tacked with a running stitch to create an 8 ply sewing cuff, and the free edges of the wrapped material were cut to size over a dilator that was upsized by 2 mm and sewn together in a running fashion to form a tube. The distal end of the tube now was trimmed to create interpapillary scallops leaving a ratio of annular diameter to length of 1:1.2 (Fig. 1 inset). The distal arms of the neovalve were then attached to both papillary muscles using pledgetted suture (Fig. 2). Following this, a running 5-0 Prolene suture was used to suture the neovalve to the annulus. Saline float revealed valve competence. Intraoperative echocardiogram revealed no valve insufficiency, no inflow stenosis and no evidence of left ventricular outflow tract obstruction. Given the poor ventricular function preoperatively and suboptimal haemodynamics at the end of the case, the child left the operating room on ECMO.

Echocardiogram performed 24 h later demonstrated neo mitral valve dehiscence at the distal antero-medial papillary muscle attachment. The child was taken back to the operating room urgently. While there was a portion of the papillary muscle attached to the ventricle, the area was quite friable and part of the papillary muscle had torn off. If the antero-septal papillary muscle is friable or there is a single papillary muscle, the valve can be attached transmurally to the left ventricle (which was required in this case). A transmural pledgetted mattress suture was used to anchor the valve (Fig. 2 inset). Intraoperative echocardiogram revealed no mitral insufficiency and no inflow stenosis. Patient was decannulated from extracorporeal membrane oxygenation 4 days later. Regular echocardiograms were done weekly and demonstrated flawless function of the valve. Despite excellent function of the new valve, however, the patient unfortunately expired 8.5 months later secondary to pulmonary complications (pneumothorax with intrapulmonary haemorrhage) likely related to the fibrillin gene defect which was confirmed by genetic testing. Autopsy was unfortunately declined by the family, therefore, tissue was not obtained for histological assessment.
We demonstrate that a custom-made valve constructed as described was able to provide our patient with a competent, non-stenotic, systemic atrio-ventricular valve for a period of almost 9 months, limited by non-cardiac issues. Others have now demonstrated follow-up of a year with similar technique [3]. Alternatives to ECM valve replacement include mechanical valve placement, modified Melody valve placement or pulmonary autograft in Dacron tube (Ross–Kabbani operation). Our findings confirm that the ECM valve can be used as a temporary support, and is a valuable tool for interim support in small infants with irreparable valves and in patients with contra-indications to anti-coagulation or those with systemic infection.

Conflict of interest: none declared.

REFERENCES

