Cardiovascular Pathology 25 (2016) 165–168

Contents lists available at ScienceDirect

Cardiovascular Pathology

Clinical Case Report

Novel valve replacement with an extracellular matrix scaffold in an infant with single ventricle physiology

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ARTICLE INFO

Article history:
Received 25 July 2015
Received in revised form 10 September 2015
Accepted 12 September 2015

Keywords:
Congenital heart disease
Univertricular heart
Valve replacement
Bioengineered tissue

ABSTRACT

Valve replacement in children with functionally univentricular hearts remains challenging. The absence of small prostheses, the lack of growth, and the need for anticoagulation limit these procedures. We describe a 1-year follow-up of an extracellular matrix scaffold tube used as systemic atrio-ventricular valve in an infant.

1. Introduction

Surgery for systemic atrio-ventricular valve (AVV) regurgitation in children with functionally univentricular heart (fUVH) is challenging [1]. The use of porcine small intestine submucosa extracellular matrix (CorMatrix™ Cardiovascular Atlanta, GA, USA) scaffold has been reported in surgery for congenital heart disease [2,3] as it provides an interim collagen framework that allows host own cells to repopulate and regenerate tissue. Recently, effective use of a handmade CorMatrix™ AVV has been reported, but its durability is not known [4–6]. We report a 1-year follow-up with a CorMatrix tube implanted as systemic AVV in an infant with fUVH.

2. Case report

2.1. Clinical history

A neonate with prenatal diagnosis of fUVH underwent bidimensional echocardiography and cardiac catheterization at birth which demonstrated an unbalanced atrioventricular septal defect with an hypoplastic right ventricle (RV), D-Transposition of the Great Arteries, subpulmonary stenosis, and a severe AVV regurgitation (Fig. 1A–B).

Following pulmonary artery banding and systemic AVV plasty at the age of 86 days, he developed a residual AVV regurgitation (Fig. 1C), causing congestive heart failure and mechanical ventilation dependency. At the age of 128 days, he underwent AVV replacement with a handmade CorMatrix tube (Fig. 2A). After uneventful redo-sternotomy, through a right atriotomy, the native anterior AVV leaflet was resected, together with accessory chordae tendineae and subpulmonary muscle bundles, to avoid SIS ECM valve impingement in native subvalvar apparatus. On the back table, a four-ply CorMatrix patch was open and hydrated and rolled, and its free edge portion was sutured with 5–0 polypropylene, to create a simple cylindrical tube, whose length was calculated leaving a ratio of annular diameter (measured at preoperative 2D echocardiography) to length of 1:1.2 (Fig. 2B). This tube was inserted in the AVV annulus and anchored distally to RV papillary muscles with interrupted pledgetted stitches, proximally to AVV annulus with a 5.0 polypropylene continuous suture. Saline float revealed competence of the new valve. Postoperative Trans-Esophageal Echocardiography (TEE) demonstrated new leaflets-wide opening, with no regurgitation and no ventricular outflow tract obstruction (Fig. 2C).

Postoperative course was characterized by multiple failed extubations, and a tracheostomy was required 3 months later. Prolonged antibiotic intravenous therapy was necessary because of positive blood cultures for multidrug-resistant *Pseudomonas Aeruginosa, Candida Albicans*, and *Enterobacter Cloacae*.

Disclosures: Conflicts of interest: none.

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http://dx.doi.org/10.1016/j.carpath.2015.09.003
1054-8807/

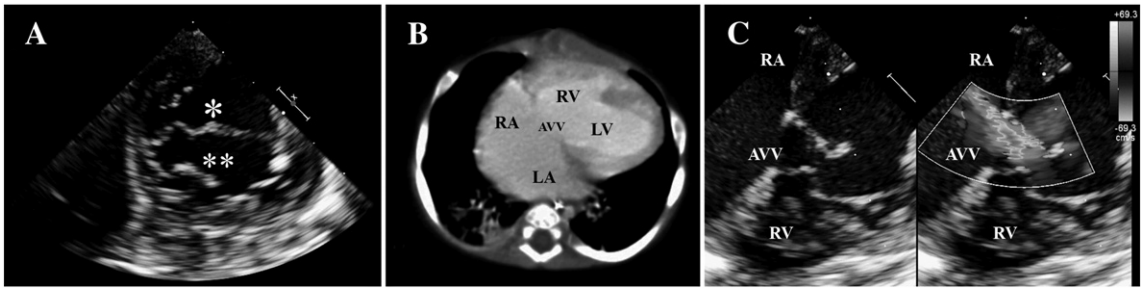

Fig. 1. ○. Bidimensional echocardiography, short axis view. Superior (*) and inferior (**) bridging leaflet of AVV (A);
○. Computed tomography scan, transverse view. The common native AVV of the unbalanced atrio-ventricular septal defect (B);
○. Parasternal 4 chamber view. After palliative plasty, a significant residual AVV regurgitation is detectable at bidimensional and Doppler echocardiography (C). Left atrium; left ventricle.

Three dimensional echocardiography performed 4 months later demonstrated excellent CorMatrix tube valve function (Fig. 2D). He was discharged on home mechanical ventilation and vasodilator and diuretic therapy, 8 months after AVV replacement.

Three months later (at the age of 16 months), during elective clinical and hemodynamic evaluation, an echocardiography exam demonstrated a severe right atrium dilatation, no AVV regurgitation, but a thickened CorMatrix tissue, with AVV stenosis (gradient 18/9 mmHg). Due to infection and progressing hemodynamic impairment, inotropic support and antibiotic therapy was started. He subsequently underwent uneventful surgery consisting in bidirectional cavo-pulmonary shunt and intraoperative dilation with Hegar dilator of the CorMatrix tube that was mildly stenotic, soft, with no calcification. Postoperatively, after successful weaning off inotropic support and inhaled nitric oxide, he was transferred to intermediate care before discharge, but due to sepsis unresponsive to antibiotic therapy, he died on postoperative day 28.

2.2. Autopsy findings and histopathology examination

Autopsy showed an intact CorMatrix valve, with moderate stenosis, a fibrous cloth, and no calcifications and/or vegetations on both sides. Histology of the CorMatrix valve showed the extracellular matrix (ECM) scaffold sleeve encircled by fibroblast hyperplasia on the atrial and ventricular side associated with fibrosis, an intense chronic inflammatory infiltrate, no signs of acute damage, and no calcifications (Fig. 3).

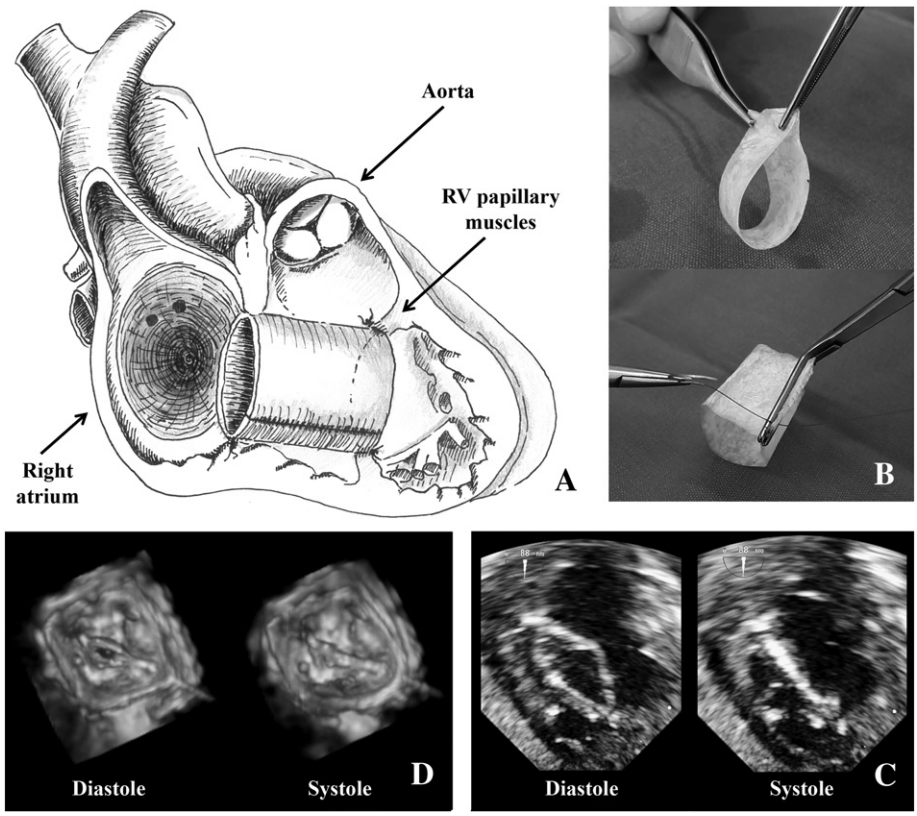

Fig. 2. ○. Diagram showing CorMatrix valve replacement. The tube is inserted in the AVV annulus and anchored distally to the right ventricular papillary muscles. The proximal part of the sleeve is anchored to the AVV annulus (A);
○. Back table construction. CorMatrix scaffold is rolled and sutured to create a cylindrical tube (B);
○. Postoperative *trans*-esophageal bidimensional (C) and 3D transthoracic echocardiography (D). It is showed an excellent tube systolic and diastolic function.