Porcine Xenograft Aortic Root Replacement in a Three Month Old With Severe Truncal Insufficiency

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Outcomes for truncus arteriosus repair are impacted significantly by the severity of truncal valve dysfunction. When satisfactory repair of the regurgitant truncal valve is unattainable, replacement is required. Given our experience in children with stentless porcine xenografts in the aortic position and the incidence of early valve failure for aortic homografts in infants, we replaced a severely regurgitant truncal valve with a full-root porcine xenograft in a 3-month-old infant. The initial and early result are encouraging, suggesting that the stentless porcine xenograft may be considered an option in cases where primary repair of the truncal (or aortic) valve is not possible.


Although surgical repair of truncus arteriosus is associated with improving outcomes, patients with severe truncal valve insufficiency have significantly increased morbidity and mortality [1]. When primary repair of the truncal valve is not achievable, replacement is required to improve survival and quality of life [2]. The ideal prosthesis for systemic semilunar valve (SSV) replacement in children remains elusive. Earlier, we reported the successful application of a stentless porcine xenograft for aortic valve replacement in a 7-year-old patient [3]. Herein we report the successful replacement of the SSV with a stentless porcine xenograft (Fig 1; Freestyle [Medtronic, Inc., Minneapolis, MN]) in a 3-month-old born with truncus arteriosus and severe truncal valve insufficiency.

A newborn child (3.4 kg; 39 weeks gestation) presented to our center from an outside hospital with truncus arteriosus, severe truncal valve insufficiency, and compromised perfusion. On day of life 6, the child underwent salvage palliation by the following: (1) truncal valve repair employing commissural plication and leaflet resuspension; and (2) bilateral branch pulmonary artery banding. At 4 weeks of life, re-repair of the truncal valve was performed along with bands removal and pulmonary artery isolation to a 6-mm, polytetrafluoroethylene right ventricle to pulmonary artery Sano conduit. The postoperative echocardiogram revealed mild to moderate insufficiency and mild stenosis of the truncal valve. The child responded well initially and was extubated. However, at 3 months of life the child demonstrated failure to thrive and feeding intolerance, necessitating a Nissen fundoplication and feeding gastrostomy. A subsequent echocardiogram demonstrated recurrent, severe truncal valve insufficiency and ventricular dilation (Fig 2) necessitating surgical intervention. A 19-mm full-root, stentless porcine xenograft was used for truncal replacement for systemic semilunar valve (SSV) replacement in children remains elusive. Earlier, we reported the successful application of a stentless porcine xenograft for aortic valve replacement in a 7-year-old patient [3]. Herein we report the successful replacement of the SSV with a stentless porcine xenograft (Fig 1; Freestyle [Medtronic, Inc., Minneapolis, MN]) in a 3-month-old born with truncus arteriosus and severe truncal valve insufficiency.

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valve replacement [3]. The ventricular septal defect (VSD) was closed with a fenestrated polytetrafluoroethylene patch, given the small pulmonary arteries (3 to 4 mm), and a right ventricle to pulmonary artery valved conduit (12-mm Contegra; Medtronic, Inc) was implanted.

The child recovered well and postoperative echocardiograms revealed a competent neo-truncal valve and preserved ventricular function (Fig 3). The child was discharged to a rehabilitation facility for transitioning and then discharged home a total of 3 months after truncal valve replacement. For prophylaxis, 40.5 mg of aspirin and 0.5 mg/kg of clopidogrel were administered daily. At 7-month follow-up, the child presented with poor weight gain, severe Freestyle valve insufficiency, and moderate left ventricle dilatation. The Freestyle valve was inspected intraoperatively for repair or replacement. Interestingly, a 1-mm thick fibrin layer similar to that observed inside stented vessels was found covering the left ventricular outflow tract, the 3 bioprosthetic valve leaflets, and root. The layer seemed to originate from the VSD fenestration but was not causing any significant obstruction other than tethering the leaflet excursion. The fibrin layer was removed and the underlying Freestyle valve appeared normal. Postoperative echocardiogram demonstrated the prosthesis to be without insufficiency or stenosis. The child’s clinical status improved and he was discharged home on 1 mg/kg of enoxaparin twice daily and 40.5 mg of aspirin daily. Follow-up at 11 months after initial implantation showed no insufficiency or stenosis. Ventricular function was normal with left ventricular internal dimension in diastole of 2.89 cm (z score = 1.03).

Comment

Severe truncal valve insufficiency is associated with increased mortality and typically requires repair or replacement [2]. When satisfactory repair is not achievable, traditional options for truncal valve replacement are the same as for other forms of SSV dysfunction. These options include mechanical prostheses, aortic homografts, pulmonary autografts (non-truncus), and xenograft bioprostheses.

All options for SSV replacement in children have inherent limitations. Mechanical valves have documented success in adults and children [4, 5] but use is limited in the pediatric population due to patient-prosthesis mismatch, lack of growth, and the need for life-long anticoagulation [5]. Aortic homografts avoid anticoagulation but are susceptible to calcification and regurgitation, leading to high early and late reoperation rates [6]. In cases of SSV dysfunction where an adequate pulmonary valve is present, the use of pulmonary autografts in the aortic position (Ross procedure) remains controversial due to the creation of 2-valve disease and the incidence of autograft insufficiency [7].

Xenograft valves have traditionally been avoided in pediatric populations because of concerns for accelerated calcification and early valve failure [8]. However, the latest generation of bioprostheses has shown increased durability, excellent hemodynamics, freedom from long-term anticoagulation therapy, and freedom from aortic regurgitation and stenosis in adults [9]. Given the limitations of traditional options for SSV replacement in children, we have explored the application of xenograft...
prostheses to this population. Early results with the Freestyle are encouraging [3].

In this very challenging case, we implanted a stentless porcine xenograft in the SSV position in the youngest patient reported to date. The patient developed a fibrin sheath in the left ventricular outflow tract and Freestyle valve 7 months after initial implantation. The precise mechanism of fibrin deposition is unknown, although it may be related to turbulence from the ventricular septal fenestration, decreased leaflet excursion and leaflet washing in a small patient, or clopidogrel resistance in an infant [10]. A similar observation of fibrin-like deposition has been reported by Ohnaka and colleagues with a bioprosthetic valve 27 months after initial implantation in an adult patient [11].

The woven polyester cuff of the Freestyle is of variable height (Fig 1). Therefore, the prostheses must be rotated to facilitate coronary button implantation. For typical coronary anatomy, this involves a 180° rotation of the prosthesis to match the least-tall cuff with the posterior coronary. In most cases, the patient’s left coronary button is implanted into the porcine root noncoronary sinus, and the patient’s right coronary button is implanted anteriorly in the location of the porcine coronary.

In very small patients, the right pulmonary artery and left coronary artery must be mobilized to allow for unobstructed left coronary button implantation. Left ventricular outflow enlargement may be necessary as 19 mm is the smallest Freestyle currently available.

References

Birt-Hogg-Dubé Syndrome in a Patient Presenting With Familial Spontaneous Pneumothorax

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Birt-Hogg-Dubé (BHD) syndrome is a recently discovered autosomal-dominant disease caused by a mutation in the folliculin gene. We report a patient with familial spontaneous pneumothorax who was found to have BHD syndrome. Patients with a personal and family history of pneumothoraces and computed tomographic (CT) findings of multiple pulmonary cysts should alert the thoracic surgeon to this syndrome; additional evaluation and testing may be warranted.


Birt-Hogg-Dubé (BHD) is a genetic syndrome characterized by skin lesions, renal tumors, and recurrent pneumothoraces. Patients with a family history of pneumothoraces and suggestive CT findings should be referred for genetic testing and offered screening for the detection of renal neoplasms.

A 28-year-old man presented to our emergency department with a 1-day history of progressive dyspnea and a chest roentgenogram that demonstrated bilateral pneumothoraces (Fig 1). The patient was a nonsmoker and had no significant past medical history. However, his family history was notable for recurrent pneumothoraces affecting both his mother and his maternal grandmother. A CT scan of the chest demonstrated several parenchymal cysts, which raised concern for BHD syndrome (Fig 2). The pneumothoraces were evacuated with bilateral pigtail catheters, and the patient was discharged home the following day. Further testing was conducted on an outpatient basis. His α1-antitrypsin serum level was normal, as were anti-RNP, Ro, and La antibody levels. Abnormal sequence analysis of the folliculin (FLCN) gene was consistent with BHD syndrome. Given the association of BHD with renal neoplasms, the patient underwent renal ultrasonography, which was unremarkable.

Comment

BHD is an autosomal-dominant genetic syndrome that was first described in 1977 [1]. The initial report of the