Long-Term Survival of Aortic Atresia Following Biventricular Corrective Surgery

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Abstract. A female born with aortic atresia, large ventricular septal defect, normal mitral valve, and left ventricle is well at 21 years of age following biventricular repair. She had palliative surgery at 15 days and closure of ventricular septal defect with placement of a valved conduit from the left ventricular apex to descending aorta at 15 months. Conduit was replaced at 34 months and at 10 and 21.5 years of age.

Keywords: Aortic atresia — Left ventricular apex to descending aorta conduit — Cardiac MRI

Case Report

This adult female was born in 1980 following a full-term pregnancy and normal vaginal delivery to a gravida 1, para 1 mother. Birth weight was 2.36 kg. At 4 days of age she developed respiratory distress and decreased peripheral perfusion. Infusion of PGE1 resulted in significant improvement. On auscultation, the second heart sound was single and a 3/6 systolic murmur was audible at the left sternal border. The ECG showed sinus rhythm, right axis and severe right ventricular hypertrophy. The chest radiogram revealed a normal heart size and increased pulmonary vascular markings. M-mode echocardiography revealed aortic atresia with a normal mitral valve and left ventricle (LV).

Cardiac catheterization and angiography at 15 days demonstrated a large outlet ventricular septal defect (VSD), a wide ductus arteriosus (DA), and isthmic coarctation of the aorta (CoA). There was left to right shunting across the large VSD and right to left shunting through the DA up the aortic arch and down the descending aorta (DAo). Qp/Qs was 4.5:1 L/min/m², and pulmonary vascular resistance was 1.44 U × M².

At 27 days, the patient underwent bilateral branch pulmonary artery banding (PAB), placement of an 8-mm Goretex™ conduit from the pulmonary trunk to the coarcted juxta-ductal aorta, and ductal ligation. During the course of the first year, the patient exhibited progressive cyanosis and failure to thrive. A retrograde catheterization (no venous access) and angiography (Fig. 1) at 14 months of age revealed a trans conduit pressure gradient of 20 mmHg. Right ventricular (RV) pressure was systemic with pulmonary arterial branches tightly banded. Aortic oxygen saturation was 68% on room air. Corrective surgery was undertaken at 15 months under cardiopulmonary bypass and hypothermia. Bilateral pulmonary branch arterioplasty, division of pulmonary-aortic conduit, VSD closure and trans-diaphragmatic insertion of a composite graft, consisting of a 14-mm right angle metallic stent and a 15-mm Carpentier-Edwards valved conduit between the LV apex and abdominal aorta were performed [1]. The early postoperative period was complicated by a cerebral vascular accident resulting in left upper extremity paresis.

The second year of life was characterized by normal growth parameters. At 27 months of age, a left heart catheterization and angiography revealed no conduit stenosis or regurgitation. Mild Coa was demonstrated with a reverse gradient of 20 mmHg. Over the following 6 months, severe conduit valve regurgitation developed prompting reoperation. At the age of 34 months the malfunctioning conduit was replaced with a 16-mm Carpentier-Edwards conduit and the apical stent was replaced with an 18-mm right angle stent. Other than left upper extremity paresis, growth and development remained normal. A cranial CT scan revealed a right-sided postinfarction encephalomalacia, but no medical treatment was required.

Over the next 6 years the conduit valve gradually became regurgitant and stenotic. A pressure gradient of 70 mmHg was demonstrated at cardiac catheterization at 10 years. The pressure gradient across the CoA remained 20 mmHg. At reoperation at 10 years of age, the calcified porcine valve with minimal pannus formation was removed and replaced with an 18-mm modified Hancock valved conduit that was anastomosed to the previously placed apical stent [3, 7]. Postoperatively, isolated premature ventricular contractions (PVC) and transient nonsustained ventricular tachycardia occurred, but no long-term pharmacological intervention was required.

At cardiac catheterization and angiography at 14 years of age (Fig. 2), a 10 mmHg pressure gradient was demonstrated across the porcine valve and there was a further 10 mmHg gradient between the distal apical conduit and the abdominal aorta, as well as a 5 mmHg reverse pressure gradient across the Coa. Mild conduit valve regurgitation was present. LV contractility was normal.

At 21 years the patient developed recurrent syncope. A 24-h holter monitor demonstrated sinus rhythm, isolated PVCs, and
runs of nonsustained supraventricular tachycardia. A 2-D echocardiogram revealed mild global LV dysfunction. Conduit stenosis and regurgitation were noted. Due to a prominent to-and-fro murmur over the left upper abdomen and lower thorax, a contrast enhanced magnetic resonance imaging study was performed (Fig. 3). Aneurysm formation at the site of conduit anastomosis with the abdominal aorta was suspected and confirmed at angiography. Catheterization revealed a 15 mmHg pressure gradient across the porcine valve. The pressure gradient across the coarctation was 5 mmHg.

At surgery (21.5 years), a thin-walled 2.5 × 3 cm pseudo-aneurysm found at the disrupted suture line between the distal apical conduit and the descending aorta (DAo) was resected. Bacterial cultures were negative. The apical metallic stent and the conduit were replaced with a #22 composite porcine valve conduit. A modified right atrial maze procedure was also carried out [8]. No coarctation repair was attempted. The patient recovered fully and was discharged home 1 week following the surgery with a well functioning conduit, good LV function, and in sinus rhythm. Medications included an anticonvulsant, diuretics, antiplatelets, and afterload reducing agents.

Discussion

Aortic atresia is most often part of the hypoplastic left heart complex. Such patients are candidates for univentricular (RV) repair [6, 9, 11]. This report deals with a rare form of aortic atresia associated with normal mitral inflow, normal sized LV, and a large outlet VSD. A patient with this anatomy should be considered for biventricular surgical repair.

The surgical procedures performed in our patient reflect the trends that prevailed in the 1970s. In neonates the primary objective was to establish a lasting pulmonary artery to aorta flow and to protect the pulmonary vascular bed through branch PAB. Once the patient reached a suitable age, a biventricular repair was accomplished by patch closure of the VSD and establishment of a single outflow from the LV by an apical stented conduit connected to the abdominal aorta.

Carrel first proposed the LV apex-to-DAo bypass in 1910, but it was not until 1955 that Sarnoff et al. applied it to clinical use for severe aortic valve disease [2, 14]. They used the Hufnagel caged ball prosthesis, later perfected by Starr and Edwards [5, 15].

The advent of modern surgical techniques, advances in mechanical and biological aortic valves, has