Case report

One-stage neonatal corrective repair for d-transposition of the great arteries and complete atrio-ventricular canal

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Abstract

Association of d-transposition of the great arteries and complete atrio-ventricular canal constitutes an uncommon and complex cardiac anomaly usually associated with poor prognosis. We report our experience on one-stage neonatal repair for d-transposition of the great arteries and complete atrio-ventricular canal. Between August 1997 and 2005, four patients (two males and two females) underwent anatomical correction for d-transposition of the great arteries and complete atrio-ventricular canal using an arterial switch procedure and two-patch repair. Mean age and weight at operation were 20 days (range from 3 to 28 days) and 3.2 kg (range from 2.7 to 3.5 kg), respectively. None of the patients received preoperative palliative procedure. Associated lesions were left outflow tract obstruction in three patients and multiple muscular ventricular septal defects in two patients. All four patients survived the operation. There was one in-hospitalised death due to fungal sepsis. One patient required late re-operation for left ventricular outflow tract obstruction and left atrio-ventricular valve regurgitation. For a mean follow-up of 67 months (range from 51 to 90 months) all patients are asymptomatic and with no residual defects. Corrective repair of d-transposition of the great arteries and complete atrio-ventricular canal can be successfully achieved in this very challenging population during the neonatal period.

Keywords: Transposition of the great arteries; Atrio-ventricular canal defect; Arterial switch

1. Introduction

d-Transposition of the great arteries (d-TGA) is the most frequent cause of cyanosis in the neonate. Complete atrio-ventricular canal (CAVC) is a common form of congenital heart disease that is frequently associated with other congenital cardiac defects. For both isolated anomalies, excellent surgical results have been reported. However, surgical approach to d-TGA and CAVC has been classically associated with high morbidity and mortality. Described surgical approaches to deal with this uncommon scenario palliative as well as staged anatomical repair combining an arterial switch operation with a CAVC repair [1,2]. Furthermore, left ventricular outflow tract obstruction (LVOTO) can complicate both d-TGA and CAVC.

In this report we describe our results with four consecutive cases of d-TGA with CAVC repaired in infancy in a single stage procedure.

2. Patients

Between August 1997 and 2005, four patients (two males and two females) underwent corrective repair of d-TGA and CAVC at Children’s Hospital Boston. Median age at operation was 14 days (range 3—28 days). Patient data are summarized in Table 1. Complete anatomical repair was achieved in all patients. Through median sternotomy the pericardium was exposed, excised and treated with glutaraldehyde 0.6% during 15—30 min for patch material. Subsequently, the ascending aorta and main pulmonary artery and its branches were dissected free. Cannulation was achieved with a single arterial and two venous cannulas. The patients were operated on cardiopulmonary bypass and mild hypothermia. Through right atriotomy, the interatrial septum, the pulmonary veins, the cava veins, the common atrio-ventricular valve, and the ventricular septal defect (VSD) were inspected. At this point, wide resection of LVOTO was performed when necessary via incision of the pulmonary artery and through the pulmonic valve under ventricular fibrillation (Fig. 1). In one case a subpulmonary aneurism was excised while in other two resection on fibrous tissue was achieved. In all our patients, accessory tissue from the AV valve attached to the septum was present. This tissue was not part of the subvalvular apparatus so it could be resected. LV size was normal in all
patients. The aorta was cross-clamped and the heart arrested with cold blood cardioplegia. The aorta was transected and the coronary buttons were excised and mobilized to reach the neoaorta. Counterincisions were made in the neoaorta and the buttons were sewn in place. Final inspection and resection of any additional tissue was done once the arterial switch was deemed possible and the heart was arrested with cardioplegia and the aorta transected (Fig. 1). The Lecompte maneuver was achieved and pulmonary artery branches were brought anterior to the aorta and anastomosed end-to-end circumferentially. The VSD was then closed. In order to avoid conduction tissue damage the coronary sinus was left draining to left atrium. The cleft was closed in all patients. Finally the ASD was closed. CAVC was approached with a two-patch technique in all patients. Autologous fixed pericardium or Dacron as patch material were used depending on surgeon’s preferences. The atriotomy was closed and the aortic cross-clamp was removed after complete deairing of the heart. During the rewarming period, the pulmonary defect was repaired with autologous fixed pericardium sewn in place around both coronary buttons, and a pulmonary end-to-end anastomosis was created. Finally, the patients were weaned from cardiopulmonary bypass in the standard fashion. Patient 1 also underwent aortic arch reconstruction and coarctation resection.

Three of the four patients who underwent complete repair are long-term survivors. One patient (d-TGA, CAVC, LVOTO, arch hypoplasia and coarctation) died 2 weeks after surgery of Candida sepsis. During the first 2 weeks this patient had a gradual recovery with echocardiograms showing good ventricular function, no residual LVOTO, and mild to moderate left atrio-ventricular valve regurgitation (LAVVR). After 2 weeks she developed sepsis, and multiple cultures confirmed Candida sepsis. Despite aggressive treatment with amphotericin, the infant died on the 16th postoperative day. The other three patients are alive and well at a median follow-up of 58 months (range from 51 to 90 months). No patients has greater than mild LAVVR, and all have unobstructed LVOTO. One patient was re-operated on at 7 months of age for a subaortic membrane and worsening LAVVR. He underwent resection of the subaortic stenosis and LAVV plasty. At latest follow-up, all patients are asymptomatic, biventricular function is normal with no significant LAVR or residual LVOTO by echocardiography. None of the patients have developed aortic valve insufficiency although one has mild aortic root dilation.

3. Comment

d-TGA with CAVC is a very rare combination of defects, usually associated with a poor prognosis. CAVC repair is more challenging during the neonatal period because AV valve leaflets are thin and delicate. d-TGA associated with a large
VSD can result in adequate systemic oxygen saturation, and patients become cyanotic when severe LVOTO is present. Some surgeons have advocated palliative surgery and/or to postpone anatomic repair [1,2]. Due to the frequent association with LVOTO, pulmonary artery banding is rarely required. Furthermore, banding is a risk factor for aortic regurgitation after the arterial switch operation, and AV valve regurgitation in CAVC [3]. Systemic-to-pulmonary artery shunting is a common palliative approach for severe cyanosis, but when CAVC is present, attendant ventricular volume overload can exacerbate AV valve regurgitation. Moreover, the current approach for both isolated lesions is to perform anatomic repair early in life.

The surgical management of complex forms of d-TGA with LVOTO continues to present a challenge to the cardiac surgeon due to poor results with current approaches and recurrence of LVOTO. Subvalvular stenosis can frequently be excised successfully through the pulmonic valve. After transection of the pulmonary artery the subpulmonary tissue can be resected through careful retraction of the pulmonic leaflets. Three patients had pulmonary subvalvar stenosis, one of those having bicuspid pulmonic valve and supravalvar stenosis. In these patients wide subpulmonary resection was performed. Aortic arch hypoplasia and coarctation is an uncommon associated defect in simple transposition, and is more common with complex forms of d-TGA including double outlet right ventricle (DORV) and the Taussig—Bing complex. In our experience, the presence of aortic arch obstruction is an independent risk factor for death and complications in infants undergoing corrective surgery for d-TGA [4]. The one patient who died in this series also had aortic arch obstruction and coarctation along with d-TGA and CAVC.

LAVVR has been reported as the first cause for late re-operation occurring in up to 30% of patients undergoing repair of endocardial cushion defects [5]. Cleft closure has been related to improved outcomes [6]. In our series all patients had LAVV cleft and mild LAVV regurgitation preoperatively. The LAVV cleft was completely closed without creating stenosis in all patients. At discharge all patients had unobstructed RVOT as well as LVOT. One patient with supravalvar, valvar, and subvalvar pulmonary stenosis had residual LVOTO after repair and subsequently LAVVR worsened during follow-up. He underwent resection of subaortic membrane and mitral valvuloplasty 6 months after first operation. No significant residual LVOTO or LAVVR has been observed. High incidence of residual VSD after correction of CAVC associated to DORV or Tetralogy of Fallot has been reported [7,8]. The importance of size and shape of the VSD patch has been also addressed [6]. In our series, we used a Dacron or autologous pericardium ‘crescent-shaped’ patch to close the VSD. A small, muscular and restrictive (gradient = 70 mmHg) residual VSD was achieved in one patient.

In conclusion, we achieved success performing one-stage neonatal surgical corrective repair of d-TGA and CAVC, with or without LVOTO, in three out of four patients. Associated anomalies including LVOTO and aortic arch obstruction can also be successfully managed surgically in this very challenging population, but aortic arch obstruction remains a risk factor.

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References