Case report

Primary repair of transposition of the great arteries with interrupted aortic arch

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Received 3 November 1997; received in revised form 6 January 1998; accepted 14 January 1998

Abstract

Primary repair of transposition of the great arteries with interrupted aortic arch was performed in a 2 day old neonate weighing 2.1 kg. The repair consisted of a direct anastomosis between the ascending and the descending aorta and an arterial switch repair without Lecompte procedure because of the side by side position of the great arteries and an anomaly of the coronary arteries. A total of 28 months after the repair, the patient remains in excellent clinical condition. © 1998 Elsevier Science B.V. All rights reserved.

Keywords: Congenital heart disease; Transposition of the great arteries; Interrupted aortic arch; Primary repair; Cardiac surgery in neonates; Circulatory arrest

1. Introduction

Interruption of the aortic arch (IAA) represents a critical congenital heart disease which necessitates surgical intervention during the first days of life. IAA is usually associated with ventricular septal defect (VSD) or other congenital heart lesions. The association with transposition of the great arteries (TGA) is rare and occurs in ~4% of patients with IAA [1]. The surgical treatment of this complex heart malformation was reported in few papers. In some reports, reconstruction of the aortic arch and pulmonary artery banding was suggested [2–5]. However, only a small number of neonates underwent the primary repair of IAA with TGA [6–10].

2. Case report

A 1 day old cyanotic male neonate weighing 2.1 kg was sent to our institution on a prostaglandin infusion. Echocardiographic examination demonstrated TGA with side by side position of the great arteries, widely patent ductus arteriosus and type B IAA. Balloon atrioseptostomy and angiocardiography were performed. Coronarography showed anomalous origin of both coronary arteries (CA), right coronary artery (RCA) arising from the sinus 1 (left) and left coronary artery (LCA) from the sinus 2 (right).

Surgery was performed the next day after admission when the patient was 2 days old. The morphology and the principles of repair are shown in Fig. 1. The thorax was opened via the midline sternotomy approach. Both the ascendent aorta and the pulmonary trunk, were cannulated with straight metallic cannulae. Venae cavae were cannulated with Pacifico cannulae. Extracorporeal circulation was instituted and pulmonary arteries were occluded with snare. During cooling, dissection of the
aortic arch with its branches was completed. The descendent aorta was dissected well below the take-off of the left subclavian artery. At 16°C, infusion of prostaglandins was stopped and the arterial cannula was withdrawn from the pulmonary trunk. The left subclavian artery was doubly ligated and transected. Snares on both venae cavae were tight and the ascending aorta was cross-clamped. The St Thomas cardioplegic solution was administrated to the aortic root, and cardioplegia was repeated at 30 min intervals. The right atrium was opened and foramen ovale closed with a direct Prolene suture. The circulatory arrest was than instituted, the innominate and the left carotid arteries were snared, and the aortic cannula withdrawn. After transection of the duct, a longitudinal aortotomy was performed on the left side of the ascending aorta. The end of the descending aorta was directly anastomosed with the ascending aorta using a continuous fine polypropylene stitch (7/0 Prolene) and the anastomosis was treated with a biological glue (Tisseal). The extracorporeal circulation was reinstituted.

The repair continued with an arterial switch operation of the transposed great arteries. The ascending aorta was transected 7 mm above CA and the pulmonary trunk was transected below the bifurcation. Both CA were excised with buttons of the aortic wall and reimplanted to the former pulmonary trunk using a 7/0 Prolene. LCA had to be implanted more superiorly using a ‘trap door’ technique. The pulmonary bifurcation was left behind neoaorta which was reconstructed anastomosing the former proximal pulmonary artery with the distal part of the ascending aorta. The miss-match between the two ends of neoaorta was corrected with a small triangular pericardial patch used for enlargement of the distal part of the ascending aorta. The neopulmonary trunk was reconstructed using a pan-taloons-like pericardial patch and anastomosis with the pulmonary bifurcation to the left from neoaorta. The by-pass, cross-clamping and circulatory arrest times were 229, 122, and 40 min, respectively.

The postoperative course was complicated by bleeding, thus necessitating surgical revision, low cardiac output, atrial ectopic tachyarrhythmia and long-lasting respiratory infection which precluded extubation before the postoperative day 27. However, the patient recovered, and on the postoperative day 50, he was discharged from hospital in good condition. Today, the child is 28 months after the repair. Echocardiography shows excellent result of repair with a large aortic anastomosis. The systolic function of both ventricles is normal and there is no aortic or pulmonary regurgitation. Maximum pressure gradient of 40 mmHg is recorded across the neopulmonary anastomosis with a continuous Doppler.

3. Discussion

Less than ten successful primary repairs of IAA with TGA in neonates have been described [6–10]. The majority of survivors had type A IAA [5–9]. The type B was more commonly associated with double-outlet right ventricle [7,9]. All reported patients with the complex of IAA and TGA also had VSD [6–8]. Combination of IAA and TGA without VSD is extremely rare, and we have not found it in the literature.

Heart morphology in our patient looked more like double-outlet right ventricle but aorta took off exclusively from the right ventricle, pulmonary artery from the left ventricle and no VSD was present. Side-by-side position of the ascendent aorta and the pulmonary trunk, extreme miss-match of the great arteries, and, ‘double-looping’ of CA required a modified correction without Lecompte procedure as potential risks of bleeding, left or right pulmonary artery stenosis and CA compression were critical.

Leaving pulmonary bifurcation posterior to the neoaorta should be considered in arterial switch operation of d-TGA with side-by-side position of the great arteries, and in the unusual cases of TGA with the pulmonary trunk situated anterior to the aorta, as in mesocardia. Not to do Lecompte procedure must be considered especially in concomitant presence of certain types of coronary morphology with an increased risk of kinking or compression from an anterior aspect by the neopulmonary artery and its branches (RCA from the sinus 1 and LCA from the sinus 2, single CA from the sinus 1, intramural LCA, and both CA arising anteriorly between aorta and the pulmonary artery very close to a commissure and to each other).

An innovative surgical method avoiding problems related to the miss-match of the great arteries in this
complex heart lesion has been recently reported by Liddicoat and associates from San Francisco [10]. Their method could have been considered in our patient too. It would be, however, technically more difficult and would certainly necessitate longer circulatory arrest time, especially when LCA had to be implanted in a high position.

Acknowledgements

Supported by grant No. 2905-3, Grant Agency, Ministry of Health, Czech Republic (IGA MZ CR).

References