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Case Report

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Double Aortic Arch And Ventricular Septal Defect With Pulmonary Atresia: A Rare Combination In A Congenital Heart Surgery

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Abstract Rare combinations of congenital heart pathologies can make the management of the surgical repair difficult and may complicate the procedure. We would introduce a newborn diagnosed with ventricular septal defect, pulmonary atresia and double aortic arch and the successful palliative surgery with shunt procedure of the patient. Atypical ductus, double aortic arch and the different configuration of its branches can complicate the usual shunt procedure. Variations in vascular structures may be the reason for complications in congenital heart surgery. Before planning the surgical procedure, vascular anatomy should be presented in detail even for palliative operations. Blalock-Taussig shunt can be performed safely in the case of a double aortic arch.

Keywords Double aortic arch · ventricular septal defect · pulmonary atresia

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INTRODUCTION

Double aortic arch is a variation that occurs due to the persistence of both aortic arches in uterine life [1]. Septal defects, tetralogy of Fallot, transposition of the great arteries, heterotaxy/asplenia syndrome, dextrocardia, right ventricular-dominant atrioventricular canal defect, and double-outlet right ventricle association can be seen in the same individual [1]. We introduce a case report of a pediatric patient diagnosed with double aortic arch, ventricular septal defect, and pulmonary atresia.

CASE REPORT

A 12-day-old and 3 kg in weight term newborn was examined by echocardiography due to the cyanosis with oxygen saturation 80% in room air. On computed tomography angiography, the patient was diagnosed with pulmonary atresia (PA), ventricular septal defect (VSD), and double aortic arch (Figure 1A, 1B, 1C, 1D). The right arch was larger. Its course was through the right side of the vertebral column. On paralel, the left arch was smaller, and the arch branches and ductus arteriosus originated from the left arch. We did not find any sign of compression on the trachea. Because of the PA and severe cyanosis, we decided to perform a shunt operation. Written informed consent for publication was obtained from the patient's relatives. Permission strong[was granted by the patient's relatives to publish the case report.

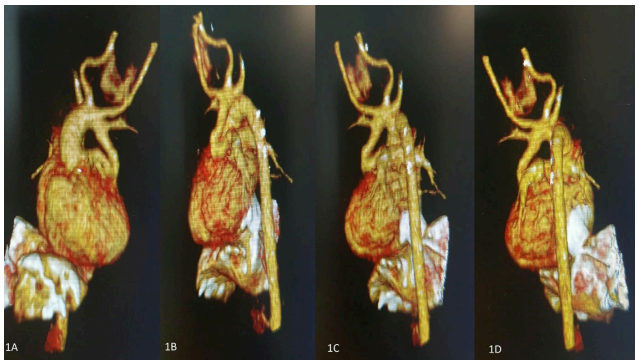

Figure 1. a, b, c, d. Preoperative views of the double aortic arch on computed tomography.

Median sternotomy was performed for better exploration. After complete dissection, the presence of a double arch and ductus arteriosus was confirmed (Figure 2A). For identifying both subclavian arteries, we introduced right and left radial artery catheters previously and a temporary occlusion test was performed for each branch intraoperatively. The ductus arteriosus was connecting the pulmonary confluence to the distal part of the descending aorta. The ductus arteriosus had a tortuous and dilated form and was severely narrowing at the connection of the pulmonary confluence (Figure 2B, 2C, 2D). After a heparin dose of 1 mg/kg was administered, a polytetrafluoroethylene (PTFE) graft of 4 mm was interposed between the right pulmonary artery and the proximal part of the right subclavian artery with an 8/0 polypropylene suture (Figure 3A). When adequate blood supply was obtained through the modified Blalock-Taussig shunt, division of the ductal tissue was performed. The narrowing on the left pulmonary artery was enlarged with a PTFE patch (Figure 3B). Dissection around the aorta was performed and adequate mobility was provided to prevent any compression on the trachea caused by the previous vascular ring effect. Postoperative 2nd day patient tolerated the extubation and acetylsalicylic acid was began 6 mg/kg once a day. On the 7th day after the operation, the patient was discharged without any problem in use only acetylsalicylic acid.

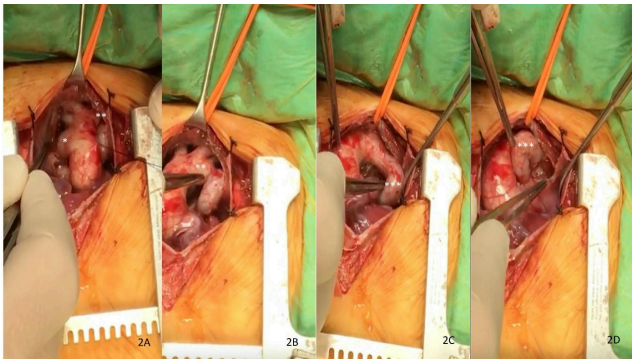

Figure 2. a. Left and right (marked with) aortic arches with branches originating from the left aortic arch. **b.** Left aortic arch (marked with). **c.** Elongated and dilated PDA (marked with).

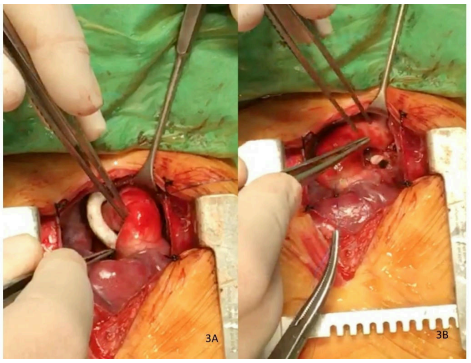

Figure 3. a. Right modified Blalock–Taussig shunt. **b.** Enlargement of the left pulmonary artery using a PTFE graft.

DISCUSSION

The double aortic arch may include archs with the same diameter, or one of them may be hypoplastic. The right arch is frequently dominant [2]. Branches arising from arch (ie truncus arteriosus, carotid arteries) can be arranged in various manners. The double aortic arch produces a vascular ring around the eusophagus and trachea. For isolated double arch,

infants present with respiratory stridor or dysphagia in the early years of their life [2]. The double aortic arch is associated with the tetralogy of Fallot [3], atrioventricular canal defect [1], coarctation of the aorta [1], VSD and pulmonary stenosis [4], or VSD-PA association [5]. Our aim was to determine the appropriate subclavian artery for the shunt procedure with an appropriate angle and to provide adequate mobility for preventing the compression on the trachea. The arrangement of branches on the double aortic arch should be checked with computed tomography in case of complex intracardiac disease.

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