from a vascular perspective, a virtual resection was already made. Providing that the surgery could restore the size of the cavity and sufficient rigidity to the ventricular wall to prevent dyskinetic motion, the complete excision of the tumors could be undertaken.

Complete resection of giant fibromas involving the AV groove can be accomplished. Reconstruction maintaining ventricular and valvular dimensions by simple means is easily achieved. The cardiac function at short- to medium-term follow-up is excellent, with resolution of symptoms.

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An approach to interrupted aortic arch associated with transposition of the great arteries

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Interrupted aortic arch (IAA) is an uncommon anomaly that often is associated with other conotruncal anomalies[1,2] and carries significant mortality.[2-5] IAA is present in approximately 1.3% of patients with congenital heart disease. The combination of IAA with transposition of the great arteries (TGA) is rare. Only about 6% of children with IAA have associated TGA.[1] Surgical management of these patients is complex and associated with high mortality.[3] The world experience with surgical repair of IAA in combination with TGA is limited to a few case reports.

Clinical Summary

A neonate (6-day-old girl weighing 4.0 kg) with a diagnosis of situs solitis, atrioventricular concordance, ventriculoatrial discordance, TGA, atrial septal defect (ASD), ventricular septal defect (VSD), and IAA type A underwent a 2-stage surgical repair.

The left subclavian artery was 3.5 mm, and its origin from aortic arch was separated from the descending aorta by approximately 2 cm (Figure 1, *A*). The descending aorta was fed via a duct of 6 mm equivalent in size to the descending aorta. There was a marked disparity in size between the hypoplastic ascending aorta and the dilated main pulmonary artery. The ascending aorta was 6 mm in diameter. The main pulmonary artery was approximately twice the size of the ascending aorta.

Owing to significant distance between the aortic arch and descending aorta, a primary complete repair through a midline sternotomy would have been technically difficult. Thus, repair of the IAA was performed first through a left thoracotomy. A patch of an aortic homograft was used to create an interposition graft from the arch to the descending aorta. The proximal suture line was interrupted with a separate continuous suture at the site of the future arch augmentation, so that the proximal suture line could be cut across during patch augmentation. The patent ductus arteriosus was ligated and pulmonary artery banding was performed (Figure 1, *B*). However, the aortic size was still determined by the origin of the left subclavian artery, which was 3.5 mm in diameter. The next day, the patient underwent the arterial switch, VSD closure with a bovine pericardial patch, ASD closure with direct suture, aortic arch reconstruction with a homograft patch, and pulmonary artery debanding. Ascending aorta and arch reconstruction was achieved with a second patch of the aortic homograft to augment the ascending aorta and entire aortic arch extending into the previously placed homograft conduit (Figure 1, *C*). Aortic crossclamp time was 160 minutes. Cardiopulmonary bypass time was 263 minutes including a 26-minute period of circulatory arrest with continuous cerebral perfusion at 20°C. The patient is doing very well and is asymptomatic at 5 months of follow-up.

Discussion

Although a 1-stage repair for IAA associated with complex conotruncal anomalies is the preferred surgical strategy,[4,5] a 1-stage repair of IAA associated with TGA carries a high mortality.[3] Although TGA with coarctation of the aorta can be repaired through a midline sternotomy as a 1-stage procedure, it is more difficult to deal with IAA in such setting because the distance from the aortic arch to the descending aorta in patients with TGA can be quite significant. This would make repair of the IAA through a

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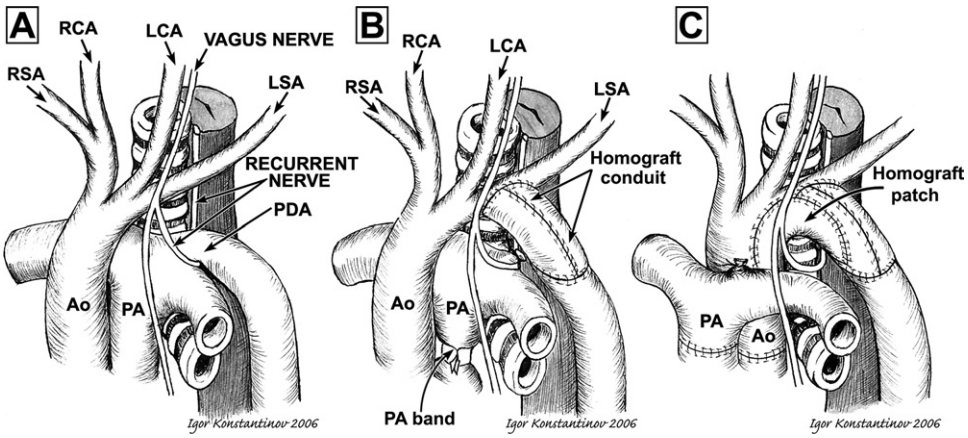

Figure 1. A, IAA distal to left subclavial artery (type A) with TGA. B, Continuity of the aortic arch was established with a homograft conduit, the patent ductus arteriosus was ligated, and the pulmonary artery was banded. C, Reconstruction of the aortic arch with a homograft patch and arterial switch operation was performed via the midline sternotomy. *RSA*, Right subclavian artery; *RCA*, right carotid artery; *LCA*, left carotid artery; *LSA*, left subclavian artery; *PDA*, patent ductus arteriosus; *PA*, pulmonary artery; *Ao*, aorta.

midline sternotomy a challenging and potentially more hazardous procedure. An attempt to repair the IAA through a midline sternotomy would also prolong the duration of cardiopulmonary bypass necessary for the subsequent arterial switch operation and VSD closure.

In summary, a 2-stage repair with generous augmentation of the entire aortic arch and arterial switch operation with VSD closure provided a good result in a patient with the combination of IAA and D-TGA.

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