Intracardiac defects and vascular ring from aberrant subclavian artery: single-incision approach.

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Abstract

We present a rare case of atrial septal defect and ventricular septal defect with a vascular ring. The ring was formed by a right-sided aortic arch with an aberrant left subclavian artery that gave rise to a patent ductus arteriosus connecting to the main pulmonary artery. We performed a single-stage repair of the intra-cardiac defects and division of vascular ring with a novel sternotomy approach instead of the traditionally practised dual approach. Our novel approach included implantation of the aberrant left subclavian artery to the left carotid artery after transection. We could perform single-stage division of vascular ring along with the closure of both septal defects.

Case Report:

A 2-year-old girl was referred for management of restrictive perimembranous ventricular septal defect (VSD), moderate sized secundum atrial septal defect (ASD) and right aortic arch with a left aberrant subclavian artery (ASA). The left ASA ran a retrooesophageal and retrotracheal course with indentation of the oesophagus and trachea. Additionally, a large patent ductus arteriosus (PDA) connecting the main pulmonary artery (MPA) to left ASA completed a rare case of vascular ring (Figure 1).

Steps of surgery:

The child underwent surgery through mid-sternotomy employing cardio-pulmonary bypass (CPB) under general anaesthesia. After sternotomy, the thymus was excised completely to expose and loop the innominate vein. That exposed the right carotid artery and left carotid artery on either side of the trachea. This was followed by further diligent dissection into the uppermost aspect of the posterior mediastinum between the trachea, left carotid artery and ascending aorta to trace entire length of the ASA and loop it so as to complete the mobilisation (Figure 2A).

After establishing the cardio-pulmonary bypass (CPB), the ductus was divided and sutured. Under mild hypothermia, the ASA was mobilised proximally as well as distally and was transected near its aortic origin while the subclavian end was brought anteriorly and was anastomosed with the left carotid artery in an end to side fashion using 7/0 prolene continuous sutures (Figure 2C). The removal of clamps revealed good pulsatile flow with satisfactory distention of the ASA.

Thereafter, under cardioplegic arrest, the VSD (autologus pretreated pericardial patch) and ASD (direct closure) were closed from right atrium. Patient was rewarmed and cross clamp was released. All the steps of open-heart surgery were along the standard lines. Child was discharged on the seventh day with good pulsations of the left radial artery. The child remains symptom-free and has well felt pulsations in left radial artery 3 months after surgery.

Discussion:

A right aortic arch with a left aberrant subclavian artery, as seen in this patient, is the second most common form of vascular ring anomaly after the double aortic arch[1]. In our case, the ring was completed by a PDA running between MPA and ASA which is unusual (Figure 2B).

The conventional approach is division of the ligamentum arteriosum or PDA to relieve compression caused by the vascular ring allowing the structures to retract away from the tracheo-oesophageal complex[2]. This can be also achieved with minimally invasive methods but with a higher chance of recurrence of symptoms requiring re-intervention[3]. Translocation of the ASA to the carotid artery via lateral thoracotomy has better outcomes than the above approaches[2,3] but this technique applies to isolated vascular rings without coexisting intracardiac lesions and our child had large septal defects that also required closure in the same sitting. A single approach to treat vascular rings with intra-cardiac defects that has a lower risk of re-intervention remains undescribed.

In our patient, a lateral thoracotomy approach was not considered in view of co-existing intra-cardiac shunts. A dual incision approach comprising of supraclavicular access and a sternotomy had obvious disadvantages of increase in operative time with attendant morbidities and 2 visible scars. Moreover, complete translocation of the ASA without a CPB from sternotomy was risky as the ASA was posterior to the trachea and oesophagus with chances of injuring the oesophagus during dissection and mobilisation. In view of these, it was thought that a midline approach using CPB was ideal because it gave access to the vascular ring, increased safety and also provided the option of closing the intracardiac defects. Besides, it also provided better access to mobilise the ASA after exit from its tracheo-oesophageal relation.

Conclusion: To conclude, our single-stage and single-incision approach has several additional advantages and early result have been encouraging.

1. It enabled total correction of vascular ring and also closure of septal defects in a single stage and single incision.
2. Complete removal of pulsatile blood flow posterior to the trachea and oesophagus without injuring them.
3. Minimal damage to the second and third part of the subclavian artery which is associated with increased risk of vascular compromises[4].
4. It was quicker, cosmetically superior and less morbid as compared to other approaches[5].
5. It eliminated chances of the recurrence of symptoms due to transection and reimplantation of ASA.

Conflicts of interest:

Conflict of interest: None declared

Author contribution statement:

Jineel Raythatha – Conceptualization; Formal analysis; Investigation; Methodology; Project administration; Visualization; Writing – original draft; Writing – review & editing

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Figure legends

Figure 1: Computer Tomography

A: Sagittal section showing TBC

B: Axial view of TBC by the vascular ring

Figure 2: Pre and post operation photographs and illustrations

A: Pre-operative photograph. Aorta and right ventricle (RV) are visualised on right. LCC is retracted to reveal posteriorly located ASA L represents lateral.

B: Illustration of TBC by VR.

C : Post-operative photograph. End-side anastomosis bulb of ASA to LCC seen.

D : Illustration of Interruption of ASA at origin from aorta, distal transection and anastomosis to LCC, and PDA ligation.

Keys for figures: AO = Aorta; ASA = Left aberrant subclavian artery; CCA = Common Carotid Artery; LCC = Left Common Carotid artery; MPA = Main Pulmonary Artery; O = Oesophagus; PDA = Patent Ductus Arteriosus; SCA = Subclavian Artery; TBC= tracheo-bronchial compression; T = Trachea; VR = Vascular Ring.