Norwood modification for hypoplastic left heart and right aortic arch

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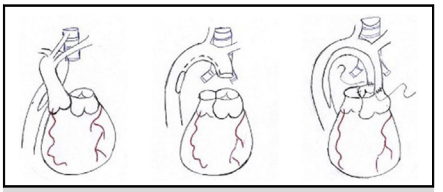


Technique of leftward transposition of ascending aorta for right aortic arch in Norwood.

CENTRAL MESSAGE

Right aortic arch in hypoplastic left heart syndrome poses challenges during the stage 1 Norwood operation requiring a modified approach.

Video clip is available online.

Right aortic arch (RAA) in hypoplastic left heart syndrome (HLHS) or variant is extremely uncommon and poses challenges to aortic reconstruction during the Norwood Stage 1 operation.[1] We describe our surgical technique for stage 1 palliation of a case of hypoplastic left ventricle associated with RAA that continued as descending aorta on the right side of the spine.

CASE REPORT

Consent was obtained for publication of this report from the parents of the patient. A 1-week-old infant, born full-term and weighing 4 kg, was referred to our institution with a diagnosis of unbalanced complete atrioventricular canal defect, hypoplastic left ventricle, RAA which descended on the right side of the spine, right-sided ductus arteriosus, aortic coarctation and a left subclavian artery originating from the main pulmonary artery. There was also a left superior vena cava that drained to the coronary sinus and an interrupted inferior vena cava that continued as the left-sided hemiazygous vein. The diagnosis was confirmed by computed tomography angiogram (Figure 1). In addition, cardiac catheterization showed a Qp/Qs of 3.6 and normal pulmonary veins. During the Norwood operation, the patient was placed on cardiopulmonary bypass via arterial cannulation of the distal main pulmonary artery and venous cannulation of the right atrium (Video 1). The branch pulmonary arteries were snared soon after

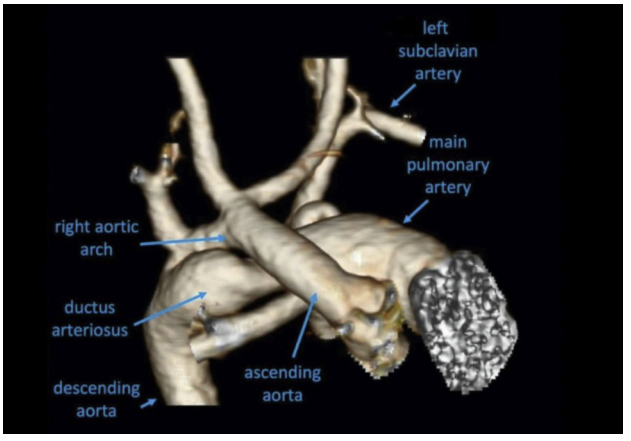
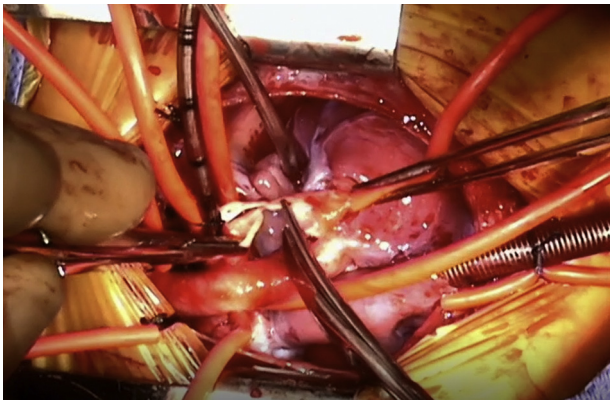


FIGURE 1. Computed tomography angiogram showing complex single ventricle anatomy with right aortic arch and right descending thoracic aorta.



VIDEO 1. Video demonstrating our technique of Norwood modification for hypoplastic left ventricle and right aortic arch. Video available at: https://www.jtcvs.org/article/S2666-2507(22)00239-5/fulltext.

commencement of cardiopulmonary bypass and the left subclavian artery arising from the distal main pulmonary artery was doubly ligated and divided. The arterial cannula was advanced and snared within the ductus arteriosus. The main pulmonary artery was transected just below the bifurcation and the distal end was closed with a fenestrated patch of pulmonary homograft. A 5-mm ringed polytetrafluoroethylene graft (Gore-Tex, W. L. Gore & Associates, Inc) was anastomosed to the right ventricle using periscopic technique. The ascending aorta was then clamped just proximal to the origin of the left common carotid artery and antegrade cardioplegia was administered for myocardial arrest. The ascending aorta was divided at the same level as the division of the main pulmonary artery. The proximal portions of the aorta and main pulmonary artery were sewn together for approximately one-third of the circumference of the aorta. The distal ascending aorta was then transposed leftward to the lateral aspect of the divided main pulmonary artery such that arch reconstruction could be performed in a technically easier fashion on the right lateral aspect of the neoaorta (Figure 2). The ductus arteriosus was then divided under circulatory arrest. The transverse arch incision was continued down the descending aorta well past the isthmus. The right lateral aspect of the arch was then patched with pulmonary allograft using continuous 7.0 PROLENE (Ethicon). The remaining circumference of the proximal aorta and pulmonary artery were encompassed in the patch. The distal anastomosis of the Sano conduit was performed, and the patient was gradually weaned from cardiopulmonary bypass with good hemodynamics. The patient did well in the postoperative period and was discharged home. A pre-Glenn cardiac catheterization demonstrated good hemodynamics with no residual arch obstruction (Figure 3). The patient subsequently underwent a left-sided Glenn operation at 8 months of age and continues to do well.

DISCUSSION

RAA is extremely uncommon in HLHS or variant. The RAA can descend on right or left side of the spine. Norwood modification for RAA with left descending thoracic aorta has been successfully described with the reconstructed aortic arch lying either in front or behind the trachea.[2-5] However, a RAA with right descending thoracic aorta poses more challenges for Norwood type reconstruction. Only one successful case of Norwood modification for RAA with right descending thoracic aorta has been described using a large diameter Kommerell diverticulum.[5] In this case, following excision of the ductus, the large Kommerell diverticulum was mobilized to the right side of the trachea and anastomosed to the neoaortic root.[5] However, the technique can only be used when there is an unusually large diverticulum. Our modification of leftward transposition of the ascending aorta is a more reproducible technique. Leftward

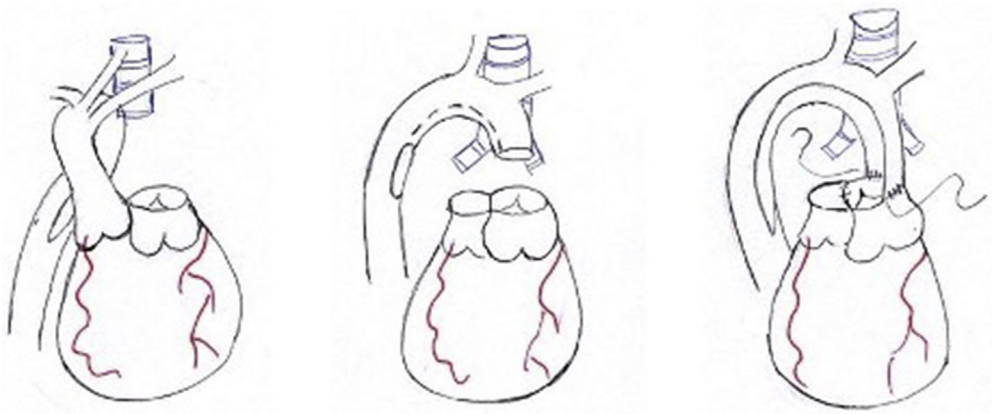


FIGURE 2. Technique of leftward transposition of ascending aorta for right aortic arch in Norwood.

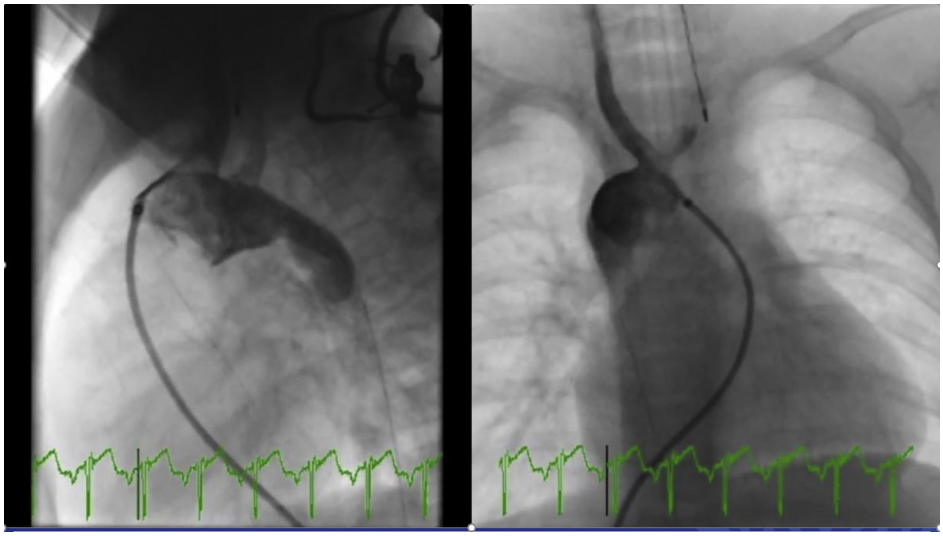


FIGURE 3. Pre-Glenn cardiac catheterization showing unobstructed neoaortic arch.

transposition of the distal ascending aorta onto the leftward aspect of the Damus–Kaye–Stansel anastomosis allows for a technically simpler augmentation of the neoaortic arch. Interestingly, review of literature on HLHS with RAA revealed common association with unusual origin of either the innominate or left subclavian artery.[2-5] In our case, it arose from the distal pulmonary artery. To potentially avoid any steal of pulmonary blood flow, we chose to ligate it. However, one could have reimplanted it either in the neoaorta or left carotid artery,[E1] at the cost of prolonging the operation.

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E-Reference

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