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Scimitar Syndrome: Variations In Anatomy And Challenges In Surgical Repairs

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Abstract
Scimitar syndrome is an entity defined by a partial anomalous pulmonary venous return wherein an anomalous right pulmonary vein drains either the entire right lung or a lobe of it, above or below the junction of inferior vena cava and right atrium. The syndrome presents different challenges for the surgical team depending on the anatomical variations. We discuss two cases of the Scimitar syndrome with different sites of drainage of the anomalous pulmonary vein requiring modifications of the technique for correction with or without the use of circulatory arrest.

Keywords: *Scimitar, surgical technique, congenital*

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I. Introduction

Diagnosis of Scimitar syndrome and its associated anomalies itself is difficult; firstly, because of the rarity of occurrence and secondly, of wide anatomical variations in the pulmonary venous drainage. Cardiac CT scan with 3-D reconstruction is an excellent adjunctive diagnostic tool in these cases. These variations often present as surgical challenges which must be dealt with different surgical approaches in each patient including intracardiac rerouting, direct implantation, and creation of in situ pericardial baffle. We, here, discuss two such patients in whom different surgical approaches were used for directing the Scimitar vein to the left atrium.

Surgical Technique

A 2-years young boy presented with complaints of recurrent respiratory tract infections and easy fatigability. Detailed history revealed that he was the first child born of a non-consanguineous marriage with a birth weight of 2.4 kilograms with an unremarkable antenatal period. The child had failure to thrive, evidence of delayed milestones, and fast breathing. Past and family histories were non-significant. On clinical examination, his pulse rate was 104 beats per minute, with a respiratory rate of 24 breaths/ minute and a BMI of 12.46 kg/m2. Cardiac examination revealed an apical impulse in the right sixth intercostal space in the mid-clavicular line which was tapping in nature. There was a grade II/III right parasternal heave, the S1 was normal with a wide fixed split S2 and a loud P2. A grade III/VI mid systolic pulmonary flow murmur was appreciated in the third right intercostal space. 2D transthoracic echocardiography was performed and it disclosed an abnormal right pulmonary vein draining into inferior vena cava (IVC), enlarged right atrium and ventricle, normal systemic venous drainage, intact interatrial and interventricular septum with evidence of severe pulmonary artery hypertension (PAH). The child underwent CT angiography (CTA) for a further detailed description of the anatomy. It confirmed normal atrial, bronchial & abdominal situs, mesocardiac, atrioventricular and ventriculoarterial concordance. The right upper, middle, and lower lobe veins formed a common channel which drained into the suprahepatic IVC at the level of the diaphragm, left pulmonary veins drained into the left atrium, right atrium, and ventricle were dilated and an ostium secundum atrial septal defect was detected (figure 1A). Hence a diagnosis of Scimitar Syndrome was established.

Another 1.5 years young boy was diagnosed with Scimitar syndrome with severe PAH and right ventricular dysfunction on the pre-operative echocardiography and CTA. The right-sided pulmonary veins united to form a common channel that drained in the IVC at the level of IVC- right atrium junction (figure 1B). The child had an aplasia of the upper lobe of the right lung, with cystic changes in the right lower lobe which also received collateral from the descending thoracic aorta which was revealed on CTA.

Both the patients underwent surgical repair with standard median sternotomy. Vertical pericardiotomy was performed and after systemic heparinisation, aortic cannulation was performed. A straight venous cannula was inserted into the right atrium (RA) which was then advanced into the superior vena cava. IVC was cannulated using an angled cannula. In the first child, however, a special precaution was followed before IVC cannulation. The IVC was dissected lower down and the angled cannulation was done at a lower level and more medially. This was not the case with the second child where the Scimitar channel was draining near IVC- right atrium junction. Diastolic cardioplegic arrest following aortic cross-clamping was achieved using St. Thomas cardioplegia solution which was repeated every 20 minutes.

In the first case, the core temperature was gradually reduced to 20° Celsius, circulatory arrest was attained and the right atrium was opened and the IVC cannula was removed. The right atriotomy was extended across the RA- IVC junction. The IVC cannula was removed and the anatomy was delineated. The separate opening of the scimitar channel was identified in the IVC, and to confirm the direction and position of the anomalous right pulmonary venous channel, the right pleura was opened and the scimitar channel was delineated. The opening of this anomalous channel was narrow and therefore it was enlarged by cut-back technique and suturing it with the right atrial wall to form the posterior suture line. Baffling of the right anomalous PV was done using an adequately sized homograft pericardium across the OS-ASD into the left atrium. The right atrium and IVC were then closed and the patient was gradually rewarmed. After gradual weaning off from cardiopulmonary bypass, the post-repair TEE evaluation was performed which revealed hepatic veins draining into IVC which further was seen draining into the RA. The pericardial baffle was seen continuing with the interatrial septum, the pulmonary venous baffle was confirmed draining into the left atrium with no turbulence or gradient across the IVC- RA junction or within the pulmonary venous baffle (figure 2). The child had an uneventful postoperative course with an ICU stay of 3 days and a hospital stay of 7 days.

During the surgical repair for the second patient, a similar cardiopulmonary bypass protocol was employed; the drainage of the right pulmonary vein just below the RA-IVC junction was confirmed. The diaphragmatic part of the IVC was mobilized and IVC cannulation was done similarly. However, in this case, the site of the anomalous pulmonary venous channel drainage was slightly higher than was seen in the previous patient and this allowed us to operate without employing the circulatory arrest. A patent foramen ovale was present which was enlarged and final re-routing of the Scimitar channel was done using an autologous pericardial patch across the foramen ovale into the left atrium. The collateral to the right lung lower lobe was clipped. The patient had severe RV dysfunction which was managed effectively with inotropic support. The child was later discharged with an uneventful postoperative course of 8 days of hospital stay.

II. Discussion

Various techniques of rerouting the anomalous vein have been described and each carries its own merits and demerits. Direct implantation carries the challenge of mobilization of the Scimitar vein, which can further lead to acute angulation and kinking. Brown et al (1) successfully described the advantage of the technique of direct implantation in 9 patients without the use of cardiopulmonary bypass. Although the technique of intracardiac pericardial baffle carries the long-term risk of stenosis, it has the advantage of growth potential. Pericardial baffle size is of utmost importance in this technique to avoid IVC obstruction; it should not be redundant, should not be too small (may create a gradient across the pulmonary venous pathway). Another potential demerit of pericardial baffle creation is the need for deep hypothermic circulatory arrest (DHCA) (2). However, in our second patient, the repair was performed under moderate hypothermia without circulatory arrest. It can be concluded that it is the drainage site of the Scimitar vein which dictates the need for DHCA.

The use of an extracardiac conduit has the potential advantage of performing repairs in anatomically challenging situations without requiring the mobilization of the Scimitar vein; however, it requires lifelong anticoagulation and has no growth potential. It may further be associated with conduit thrombosis. Kowatari et al (3) described another technique using walls of the right atrium and inferior vena cava as a flap to prevent kinking and stenosis of the scimitar vein-left atrium anastomosis.

III. Conclusion

No technique is perfect for the surgical correction of Scimitar syndrome. The procedure needs to be tailored according to the age of presentation and the anatomical variations of the syndrome. The prognosis of the disease depends on the age of presentation and outcomes after surgical repair are suboptimal for infants, which is a risk factor for stenosis after repair (4). Both our patients were beyond infancy and hence, the decision to create in situ pericardial baffle was considered appropriate**.**

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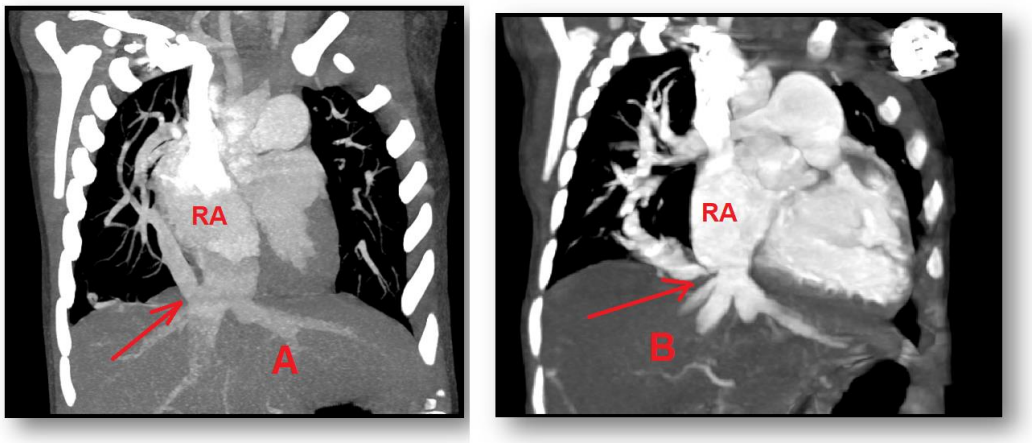

Figure 1: CT Angiography Images Clearly Delineating The Scimitar Vein- Right Upper, Middle, And Lower Lobe Veins Forming A Common Channel (Red Arrow) Draining Into The Suprahepatic IVC At The Level Of The Diaphragm (A); Right-Sided Pulmonary Veins Uniting To Form A Common Channel (Red Arrow) Draining In The IVC At The Level Of IVC- Right Atrium (RA) Junction (B).

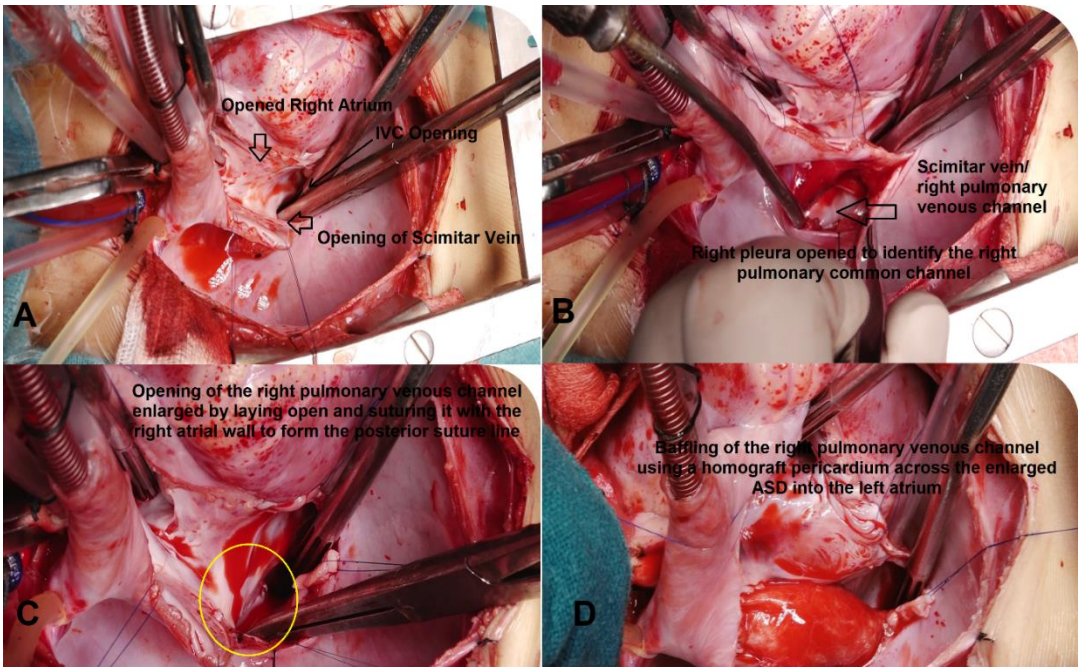

Figure 2: Rerouting Of Scimitar Channel Using A Pericardial Patch Baffle (Patient 1) Using Deep Hypothermic Circulatory Arrest.