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Case Report

Repair of Scimitar Syndrome with Intact Atrial Septum in a Two-Year-Old Child Via Vertical Right Axillary Thoracotomy

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Abstract

Scimitar syndrome is a form of partial anomalous pulmonary venous connections that involves the right-sided pulmonary veins. It includes a rare constellation of multiple cardiopulmonary congenital thoracic anomalies. Several techniques have been described for repair of such anomaly; however, the most common approach has been median sternotomy. In the current report, we describe our technique of minimally invasive repair via vertical right axillary thoracotomy.

Keywords: Scimitar Syndrome; Vertical Right Axillary Thoracotomy; Minimally Invasive; Partial Anomalous Pulmonary Venous Connections

Introduction

Scimitar syndrome or congenital pulmonary venolobar syndrome is a well-known, albeit rare form of partial anomalous pulmonary venous connection (PAPVC) that was first described in 1836 by George Cooper during an autopsy. It occurs in 3% - 6% of patients with PAPVC [1]. The main characteristic features include PAPVC of the right lung to the inferior vena cava (IVC) or its junction with the right atrium [2,3]. These veins unite usually in one large vein (scimitar vein) that resembles the curved Turkish sword, hence the name, before joining the IVC. Other associated anomalies include hypoplasia of the right lung and right pulmonary artery, dextroposition of the heart, and multiple aortopulmonary collaterals that usually arise from the abdominal aorta and traverse the diaphragm before entering the right lung. This may be associated with an atrial septal defect (usually of the inferior sinus venosus type) or can occur with intact atrial septum. A left-to-right shunt results in enlargement of the right cardiac structures and development of symptoms that necessitate repair. We present a child who was diagnosed with Scimitar syndrome and underwent repair using a two-patch technique through a minimally invasive vertical right axillary thoracotomy (VRAT) approach.

Case

A 2-year-old, 12 kg girl presented to our institution with cardiomegaly (Figure 1) and a murmur. Transthoracic echocardiogram showed enlarged right-sided cardiac chambers and PAPVC of the right lung. Computed tomography scan showed typical features of Scimitar syndrome (Figure 2). Due to the enlarged right cardiac chambers, decision was made to proceed with surgical repair.

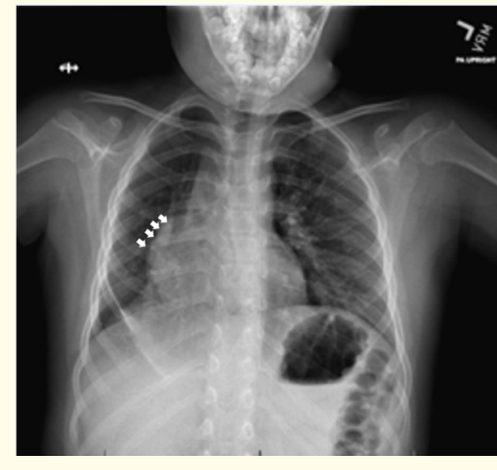

Figure 1: Preoperative Chest X-ray showing cardiomegaly, the radiographic appearance of the scimitar vein (arrow heads), the hypoplasia of the right lung, and the dextroposition of the heart.

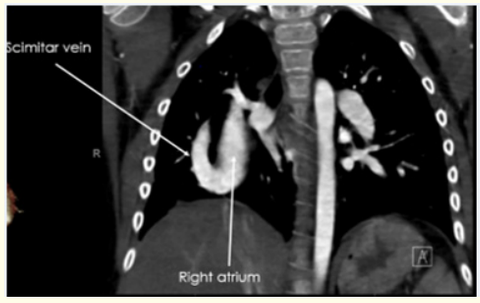

Figure 2: Preoperative computer tomography scan showing the typical scimitar vein and its connection to the inferior vena caval/right atrial junction.

After induction of general endotracheal anesthesia, the patient was positioned in the modified left lateral decubitus with the right arm abducted above the right shoulder. A 6-cm vertical skin incision was made in the right mid-axillary line, extending from the 3rd-to-6th ribs. Generous subcutaneous skin flaps were created with electrocautery and the fibers of the serratus anterior muscle were splitted to expose the underlying intercostal spaces. We entered the right chest through the right fifth intercostal space. The right lung was retracted, and the scimitar vein was visualized at the lung hilum. The scimitar vein was dissected and isolated with a vessel loop (Figure 3). The inferior pulmonary ligament was divided carefully and the multiple systemic aortopulmonary collaterals were visualized and controlled. All anomalous pulmonary blood supply was doubly ligated and divided. The pericardium was then opened anterior to the right phrenic nerve.

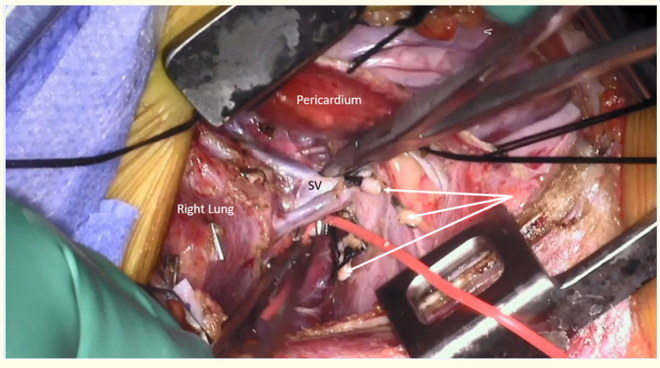

Figure 3: Intraoperative photo showing the view form right thoracotomy with the right lung retracted and the scimitar vein (SV) is dissected and isolated. The multiple systemic aortopulmonary collaterals are ligated and divided (multiple white arrows). SV: scimitar vein.

Heparin was then administered systemically, and normothermic cardiopulmonary bypass (CPB) was initiated via central aortic and bicaval cannulation. The aorta was cross clamped and cardiac arrest was achieved via antegrade cardioplegia.

An oblique right atriotomy was then performed parallel to the right atrioventricular grove. The entrance of the scimitar vein was visualized in the low right atrium, close to the IVC entrance (Figure 4). The septum primum was resected to create an adequately sized atrial septal defect. An appropriately sized bovine pericardial patch was then used to create an intra-atrial baffle (Figure 5 A), thus diverting all the right-sided pulmonary venous return into the left atrium through the surgically created septal defect. The baffle was started inside the IVC orifice around the entry site of the scimitar vein and continued up to the superior edge of the atrial septal defect. A second patch of bovine pericardium was used to augment the IVC/right atrial junction and portion of the right atrial free wall (Figure 5B). This second patch is valuable to minimize the risk of IVC obstruction by the pulmonary venous baffle especially when the pulmonary venous baffle is low in the IVC/RA junction. The heart was de-aired, and the aortic cross clamp (AXC) was removed and the remainder of the right atriotomy was closed in a routine fashion. The patient was subsequently weaned off CPB and once transesophageal echocardiogram confirmed satisfactory repair, the patient was decannulated and Heparin was reversed, followed by placement of a single chest drain and partial closure of the pericardium. The incision was then closed in a routine fashion.

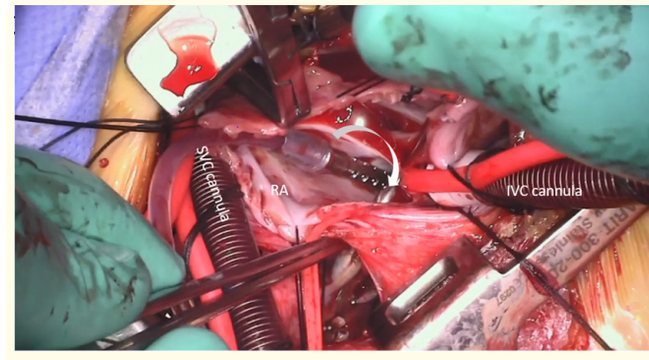

Figure 4: Intraoperative photo showing through the opened right atrium and on cardiopulmonary bypass, the entrance of the scimitar vein into the right atrium. A dilator (curved arrow) is seen passing through the opening of the vein in a retrograde fashion. SVC: superior vena cava; IVC: inferior vena cava; RA: right atrium.

Figure 5a & 5b: Intraoperative photos showing the two-patch technique with (A) the created intra-atrial baffle (asterisk) for directing the scimitar vein to the left atrium, and (B) the second patch (two asterisks) to augment the inferior vena caval/right atrial junction. SVC: superior vena cava; IVC: inferior vena cava.

The aortic cross clamp and cardiopulmonary bypass times were 64, and 91 minutes, respectively. She was extubated in the operating room, and chest tube was removed 48 hour later. She was discharged on Aspirin, 6 days later and pre-discharge transthoracic echocardiogram showed widely patent intra-atrial baffle with no residual shunts and no obstruction at either the inferior vena caval or pulmonary venous pathways. She returned to unrestricted activity 7 days later. At one-year follow-up, she continued to do well with no symptoms.

Discussion

Scimitar syndrome is a rare combination of multiple cardiopulmonary abnormalities that is centered around PAPVC of the right lung and can occur with an intact atrial septum as we described in the current case. This may cause a delay in its diagnosis unless a thorough evaluation of the pulmonary venous anatomy is done.

The most common approach for repair has been median sternotomy in the majority of centers, however, we demonstrated the feasibility of the more cosmetic VRAT approach in these cases. We believe this provides easier exposure to the scimitar vein and facilitates the repair, in addition to the previously demonstrated advantages of VRAT such as the cosmetic superiority and the short hospital stay [4].

We have previously reported our outcomes for repair of a wide variety of heart defects in infants and children using the VRAT approach. Others [5] have demonstrated the superiority of this approach as well which expands beyond the cosmetic aspect to the shorter length of hospital stay and quicker return to full activity [6]. More data will be needed prior to considering this approach as a new standard in such cases.

Conclusion

Repair of scimitar syndrome via vertical right axillary thoracotomy is feasible with excellent surgical results. This approach provides excellent visualization of the anomalous pulmonary veins, satisfactory cosmetic aspect allows rapid return to full activity. This report demonstrates the expanding indications for VRAT in the pediatric population which should be strongly considered in the current era.

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