

Direct Reimplantation Technique via a Right Minithoracotomy for Scimitar Syndrome: A Case Report

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ABSTRACT

Scimitar syndrome (SS) is a rare congenital cardiac anomaly defined by an anomalous right pulmonary vein draining of the right lung into the inferior vena cava. We describe a direct reimplantation technique and atrial septal defect closure using cardiopulmonary bypass via a right minithoracotomy on a 24 year-old female SS patient who had an accompanying sinus venous atrial septal defect.

INTRODUCTION

Scimitar syndrome (SS) is a rare congenital anomaly that includes total or partial anomalous venous drainage of the right lung into the inferior vena cava (IVC). Surgical approaches to SS vary according to the anatomical and pathological features of individual cases.

Recently, Brown et al [2003] described an off-pump reimplantation technique via a right thoracotomy for surgical management of SS with an intact atrial septum. We used the same direct reimplantation technique and atrial septal defect (ASD) closure using cardiopulmonary bypass (CPB) via a right minithoracotomy on an SS patient with sinus venous ASD and grade 3 tricuspid valve regurgitation (TR).

CASE REPORT

The 24-year-old female patient was admitted to our outpatient clinic with a diagnosis of SS. The patient's primary complaints were tachypnea and dyspnea. Transesophageal echocardiography (TEE) demonstrated grade 3 TR, anomalous drainage of the right inferior pulmonary vein into the IVC, and sinus venous ASD. Multislice computed tomography (MCT) revealed an anomalous right pulmonary vein

draining into the IVC (Figure 1). Drainage of the left lung and the right upper lobe were normal.

Operative Technique

A double lumen endotracheal tube was inserted, and the left oblique decubitus position was determined. After obtaining a baseline clotting time, anticoagulation was achieved with intravenous heparin (300 IU/kg). The right internal jugular vein was percutaneously cannulated (24 DLP EOPA Arterial Cannulae; Medtronic, Minneapolis, MN, USA). A right anterior minithoracotomy (7 cm) was made through the fifth intercostal space. The right lung was freed from all adhesions, and the inferior pulmonary ligament was divided. The scimitar vein was freed from surrounding tissue to provide greater mobility (Figure 2). The right femoral artery and vein were percutaneously



Figure 1. Preoperative multislice computed tomography revealing the anomalous drainage of the scimitar vein (white open arrow) through the subphrenic inferior vena cava (black arrow) into the right atrium. The white arrow indicates the descending aorta.

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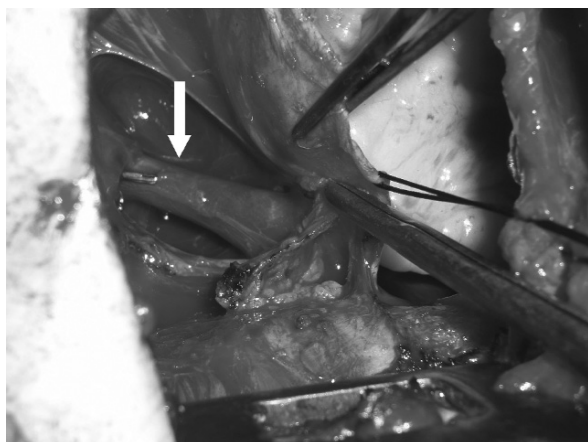


Figure 2. Intraoperative caudal view of the scimitar vein (white arrow).

cannulated (21 F and 29 F Bio-Medicus One-Piece Femoral Arterial Cannulae; Medtronic).

CPB was initiated at a flow rate of 2.4 L/kg per minute. Mild systemic hypothermia (32°C rectal) was maintained. After placing the aortic root catheter, the aorta was occluded with a Cosgrove clamp, and cold blood cardioplegia was induced. Following cardiac arrest, caval snares were snugged down, and the right atrium was opened. The sinus venous defect was closed, and the orifice of the superior vena cava was enlarged with a pericardial patch. The tricuspid valve was repaired with De Vega annuloplasty. The pericardium was opened 3 cm below the phrenic nerve, and a longitudinal incision was made into the left atrium. Holding stitches were taken from the superior and inferior corners of the scimitar vein opening into the IVC. A fine bulldog clamp was used to occlude the scimitar vein and separate it from the IVC. Flow rate and reservoir level were balanced to prevent bleeding from the IVC and entrapment of the venous system. The IVC was closed without clamps (Figure 3). The scimitar vein, after mild spatulation of the open end, was anastomosed to

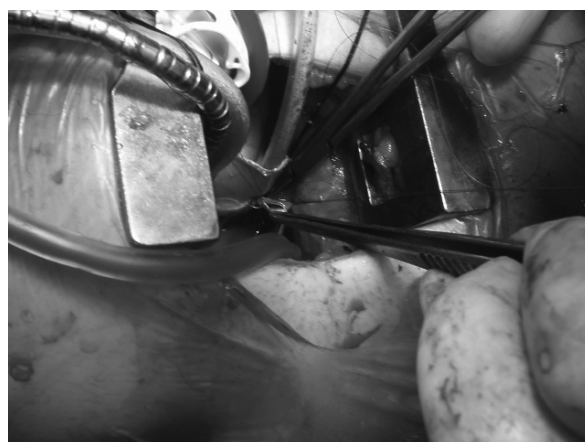


Figure 3. The inferior vena cava was closed without a clamp.

the left atrium in an end-to-side fashion with a continuous, 5-0 polypropylene suture (Figure 4). During the procedure, CO₂ was inflated (4 L/min) into the surgical region and de-airing was achieved. The cross clamp was removed, and the right atriotomy was closed. The patient was weaned off CPB without any inotropic support.

The patient made an uneventful recovery and was discharged on the third postoperative day. Two months later she remained free of symptoms, with increased exercise tolerance and good quality of life. A postoperative MCT showed that the scimitar vein was draining into the left atrium without any stenosis or kinking (Figure 5). Postoperative TEE showed mild TR. There was no residual ASD or superior vena cava stenosis.

CONCLUSION

Correction of SS entails suppressing shunt volume to prevent pulmonary hypertension [Dupuis 1992]. Honey and colleagues [1977] suggested that operative management of SS



Figure 4. A, Intraoperative cranial view of end-to-side anastomosis between the scimitar vein and the left atrium. B, Intraoperative view of the end-to-side anastomosis between the scimitar vein and the left atrium at the end of the operation.

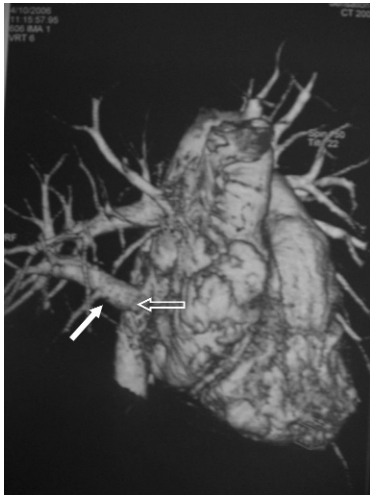


Figure 5. Postoperative multislice computed tomography revealing anastomosis between the scimitar vein (white arrow) and the left atrium (white open arrow).

should differ based on the presence or absence of an associated ASD. Various surgical techniques have been advocated to repair SS, including (1) direct implantation of the anomalous vein to the left atrium [Tornvall 1961], (2) baffling the anomalous venous channel through the right atrium and into the left atrium via an ASD [Puig-Massana 1972], and (3) division and reimplantation of the anomalous vein into the right atrium and baffling blood flow into the left atrium [Torres 1993]. Techniques that have been used in adult SS patients include (1) using the anterior wall of the right atrium to form a tunnel that diverts the anomalous pulmonary vein to the left atrium, (2) making an extracardiac conduit with a polytetrafluoroethylene graft, and (3) intrapericardial-extracardiac baffling to the right superior pulmonary vein [Walles 2002]. There is 1 case report in which a 14-mm Dacron graft was interposed between the orifice of the anomalous pulmonary vein and an enlarged ASD [Tsuchida 1987]. Brown

performed direct anastomoses of the scimitar vein to the left atrium via right thoracotomies in patients without associated intracardiac defects [Brown 2003]. Brown and colleagues did not use CPB and achieved excellent results. This encouraged us to use a similar technique via a right minithoracotomy.

This is the first report describing minimally invasive repair of SS. The aims of corrective surgery for SS in adults are to abolish the left-to-right shunt, preserve drainage of the right lung through the scimitar vein, and avoid thrombosis at the anastomosis. We have shown that the direct reimplantation technique and correction of combined pathologies can be accomplished safely via a right minithoracotomy in SS patients.

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