World Journal for Pediatric and Congenital Heart Surgery
1(3) 389-392

DOI: 10.1177/2150135110379620
http://pch.sagepub.com

Emergency Transmediastinal Pneumonectomy for Scimitar Syndrome

Panagiotis G. Sfyridis, MD[1], John K. Papagiannis, MD[2], Irene D. Lytrivi, MD[2], George V. Kirvassilis, MD[3], and George E. Sarris, MD[1]

Abstract

Repair of scimitar syndrome presenting in infancy involves either tunneling or reimplantation of the anomalous vein to the left atrium and may be fraught with serious complications such as thrombosis and secondary pulmonary infarction necessitating pneumonectomy. The authors present the case of a severely symptomatic infant with scimitar syndrome, managed initially with closure of an atrial septal defect in the hope of avoiding a repair with considerable risk of scimitar vein thrombosis and pulmonary infarction. Despite initial clinical improvement, subsequent rapid development of spontaneous massive emphysematous degeneration of the right lung necessitated emergency pneumonectomy, which was accomplished via the median sternotomy approach.

Keywords

congenital heart disease, emphysema/bullae, cardiac anatomy/pathologic anatomy

Submitted April 24, 2010; Accepted June 18, 2010.

Clinical Summary

A 3-month-old female developed tachypnea and tachycardia at 1 month of age and was noted to have a systolic murmur. Chest radiograph showed a hyperlucent right lung with reduced volume. Echocardiogram showed left-to-right shunt through a large atrial septal defect (ASD) as well as partial anomalous venous drainage into the inferior vena cava (scimitar vein). There was evidence of severe pulmonary hypertension and significant dilation of the right ventricle and right atrium (RA). Computerized tomography (CT) scan of the chest revealed a hypoplastic malformed right lung (Figure 1A).

Because of increasingly severe tachypnea and episodes of pronounced bronchospasm and respiratory distress, she was transferred to our facility and required intubation and mechanical ventilation. Cardiac catheterization confirmed systemic-level pulmonary artery pressures, pulmonary/systemic flow ratio (Qp/Qs) of 3.6:1, and angiographically typical scimitar syndrome with a dysmorphic right lung and hypoplasia of the right pulmonary arterial vasculature without systemic arterial supply (Figure 2A). A large secundum ASD was present. The scimitar vein drained into the inferior vena cava (Figure 2B). Another right pulmonary vein drained into the left atrium. With the patient still requiring mechanical ventilation, surgical intervention was performed.

Our initial plan was to attempt reimplantation of the scimitar vein without cardiopulmonary bypass (CPB), followed by closure of the ASD during a brief period of CPB. A right thoracotomy incision revealed a bilobed large and diffusely emphysematous right lung. No gross bullae were present. An incomplete fissure separated a hypoplastic lower lobe from the upper lobe. The aberrant scimitar vein, 3 mm in diameter, was located in this fissure and received tributaries from the lower lobe and the lower part of the upper lobe. There was 1 other (3 mm) pulmonary vein from the upper lobe that drained normally into the left atrium (LA). No systemic arterial supply to the right lung was found.

There was no evidence of pulmonary sequestration.

Even minimal mobilization of the scimitar vein (with the goal of possible direct reimplantation to the LA) resulted in obliteration of its lumen and engorgement of the right lung. Intracardiac redirecting of the small scimitar venous drainage

[1] Department of Pediatric and Congenital Cardiac Surgery, Mitera Children's Hospital, Athens, Greece
[2] Department of Pediatric Cardiology, Mitera Children's Hospital, Athens, Greece
[3] Department of Pediatric Cardiac Anesthesia, Mitera Children's Hospital, Athens, Greece

Corresponding Author:
Panagiotis G. Sfyridis, MD, Mitera Children's Hospital, Department of Pediatric and Congenital Cardiac Surgery, Maroussi, Athens, Greece 151 23
Email: pgsfyridis@yahoo.gr

Abbreviations and Acronyms

ASD	atrial septal defect
CT	computed tomography
CPB	cardiopulmonary bypass
DHCA	deep hypothermic circulatory arrest
LA	left atrium
PA	pulmonary artery
RA	right atrium
SV	scimitar vein

to the LA would require a more complex repair, possibly employing deep hypothermic circulatory arrest (DHCA).The long-term patency of a surgically created intracardiac tunnel was uncertain, and there was an additional right pulmonary vein that connected normally to the LA. Accordingly, the decision was made to repair only the secundum ASD. Because displacement of the right lung to gain access to the aorta and RA proved impossible without hemodynamic compromise, the thoracotomy was closed and a median sternotomy incision was made. Under routine hypothermic CPB and cardioplegic arrest, a large secundum ASD was closed with a patch of autologous pericardium. After weaning from CPB, direct oxymetry documented residual Qp/Qs due to the patent scimitar vein of 1.2:1. Pulmonary artery (PA) pressure decreased to normal (PA systolic pressure = 30 mm Hg). The patient was transferred to the intensive care unit and was hemodynamically stable.

Postoperatively, the patient suffered from increasingly frequent episodes of severe bronchospasm, precluding successful weaning from mechanical ventilation. Two weeks after surgery, sudden onset of rapidly increasing signs of respiratory failure and of cardiac tamponade was observed because of acute massive emphysematous degeneration of the right lung with air trapping and secondary cardiac compression. This condition persisted despite high-frequency jet ventilation. The degree of air trapping and secondary cardiac compression was demonstrated by CT scan (Figure 1B).

Emergency right pneumonectomy was performed through the previous sternotomy incision. At operation, the right lung was massively dilated because of the interim development of multiple giant bullae. The lung extended posterior and inferior to the heart, which was severely displaced. We proceeded with intrapericardial division of the right pulmonary artery. The scimitar vein and the additional right pulmonary vein were each doubly ligated and divided. The right mainstem bronchus was stapled, divided, and covered with a pericardial flap. Pathologic examination of the resected specimen revealed extensive bullous degeneration and hemorrhage of the resected lung without evidence of sequestration.

The patient had a subsequent smooth postoperative course. She remained free of bronchospasm and was discharged home on the 20th postoperative day. At 1-year follow-up, she is asymptomatic and has enjoyed normal development.

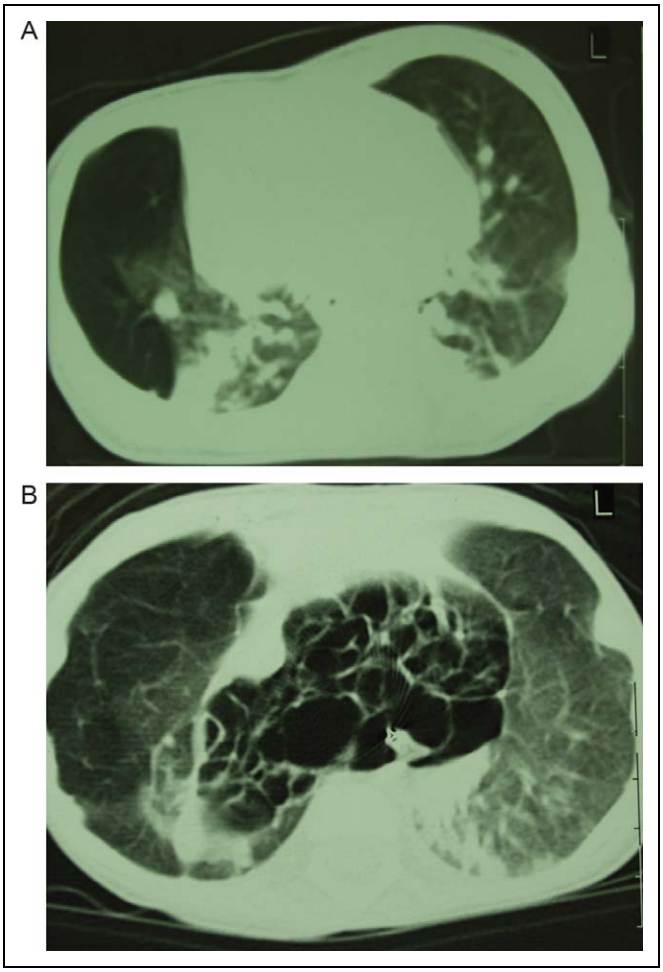

Figure 1. Computerized tomography (CT) scans of chest: (A) Preoperative study showing a hypoplastic malformed right lung. (B) Study done 2 weeks after atrial septal defect (ASD) closure reveals massive emphysematous degeneration of the right lung, air trapping, and secondary cardiac compression.

Discussion

Scimitar syndrome is a rare congenital anomaly consisting of hypoplasia of the right lung, anomalous right pulmonary venous return to the inferior vena cava, and, frequently, systemic collateral supply to the right inferior lobe. There is a bimodal presentation with either an infantile variant or a pediatric/adult form. The former is characterized by higher incidence and severity of associated defects, pulmonary hypertension, heart failure, and significant risk of mortality, whereas the latter is less severe and patients may be asymptomatic on diagnosis.[1]

Surgical treatment of scimitar syndrome is recommended in the symptomatic patient or in the absence of symptoms when left-to-right shunt is greater than 1.5:1, to avoid the sequelae of chronic right heart volume overload, and when other associated cardiac anomalies require repair.[2-8] Infants typically have features of congestive heart failure from significant left-to-right shunting from the anomalous pulmonary

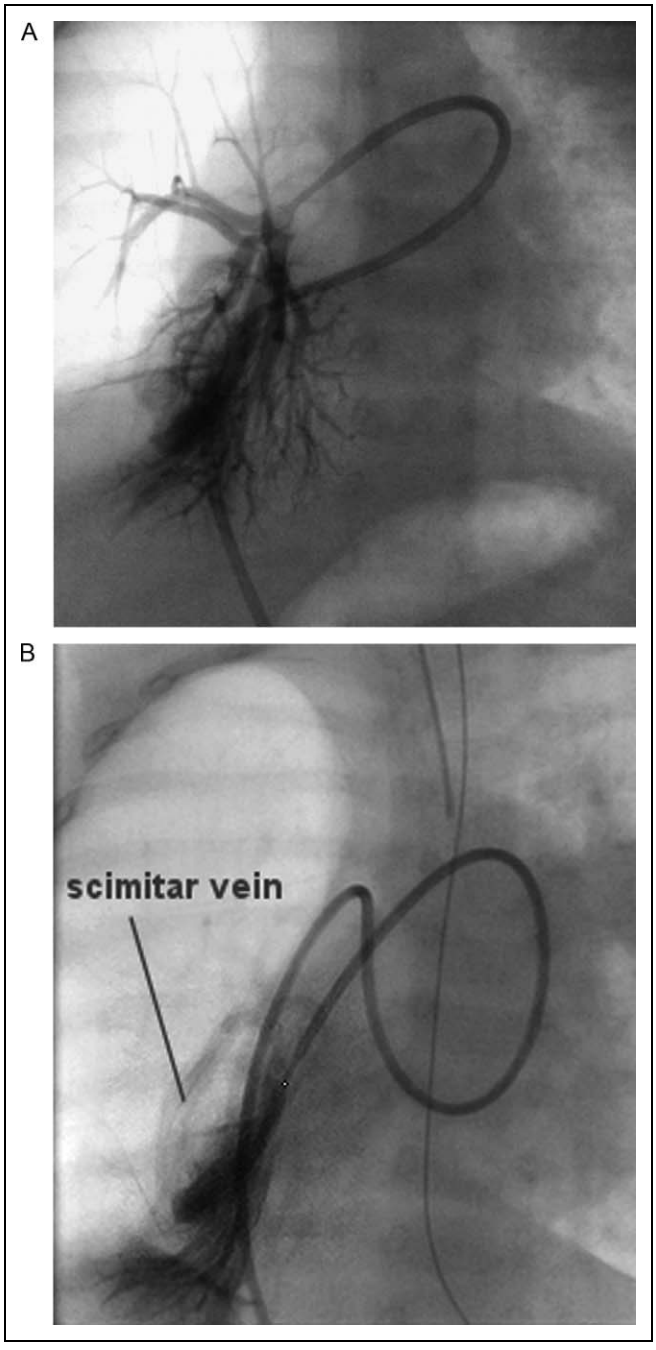

Figure 2. Cardiac catheterization: (A) Hypoplastic pulmonary vasculature of the right lung. Note sparse pulmonary arterial supply to upper, middle, and most of the lower lobe. (B) Scimitar vein draining into the inferior vena cava.

venous drainage and/or from additional cardiac defects, such as an ASD.[2] Pulmonary hypertension is nearly always present in infants and is attributed to relative scimitar vein stenosis and to hypoplasia of the right lung, redirecting most of the pulmonary flow to the left lung, in conjuction with the overcirculation caused by ASD.

Surgical management of scimitar syndrome may involve resection of the right lung drained by the scimitar vein (SV) or by a corrective approach rerouting flow from the SV to the left atrium.[1] There are several surgical corrective options.[5,7] Most commonly, repair through median sternotomy under CPB involves tunneling the orifice of the scimitar vein to the LA via the ASD, if present, or, in its absence, via a defect created in the atrial septum. Creation of this tunnel is often accomplished during a period of DHCA, although the procedure can be performed with continuous CPB. The surgically created tunnels are prone to thrombosis, which may lead to lung infarction and ultimately to the need for pneumonectomy. An alternative approach has been to reimplant the SV to the LA via right thoracotomy without CPB or with CPB if an ASD also requires closure. This approach has been used mostly in older children and adults.[2] A posteriorly located scimitar vein in an infant may be impossible to reroute to the left atrium without kinking and thrombosis.[3,5,7]

In our case, we repaired only the ASD at the time of initial surgery, as we judged the hemodynamic significance of the shunt related to the scimitar vein to be too small to risk creation of an intracardiac tunnel with an uncertain long-term fate, especially in view of the right lung parenchymal and vascular hypoplasia and the presence of 1 other right pulmonary vein draining into the left atrium. Indeed, after ASD closure, the measured Qp/Qs was only 1.2:1.

In retrospect, an initial midline approach to address only the ASD would have been less traumatic and would likely be our preferred approach in a future similar situation. However, this may not have prevented the subsequent deterioration of the right lung and the need for pneumonectomy. With regard to our decision to perform pneumonectomy via the median sternotomy approach, rather than via traditional transthoracic or extrapleural approach, several advantages seem evident in such a dire situation. First, the ill neonate or infant frequently cannot tolerate a lateral thoracotomy postion, especially when the lung is hyperinflated. In addition, ventilation by the anesthesiologist is frequently much easier in the supine position than in the lateral thoracotomy position. If necessary, lifesaving CPB can be instituted promptly. Other advantages include facilitation of a secure proximal bronchial closure in this situation and better access to hilar vessels in the setting of anomalous venous and systemic arterial supply, as seen in scimitar syndrome.

We could not explain either the persistent bilateral bronchospasm postoperatively or the delayed but massive and rapidly progressive emphysematous degeneration of the right lung that necessitated pneumonectomy. The diffuse and massive nature of emphysematous degeneration precluded any conservative resection. We speculate that humoral factors originating from the anomalous right lung may play a role in the bilateral bronchospasm that frequently afflicts patients with severe scimitar syndrome, and this dysmorphic lung (but not the contralateral normal lung) may be inherently prone to emphysematous degeneration.[4,6] This hypothesis is supported by the disappearance of left lung bronchospasm following pneumonectomy.

This case demonstrates some of the difficulties that may be encountered in the management of this uncommon and life-threatening infantile form of scimitar syndrome. Resection of

the dysmorphic lung may be lifesaving for some severely affected infants.

Declaration of Conflicting Interests

The author(s) declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The author(s) received no financial support for the research and/or authorship of this article.

References

1. Brown JW, Gudjonsson U. Scimitar syndrome. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann*. 2006;9:56-62.
2. Brown JW, Ruzmetov M, Minnich DJ, et al. Surgical management of scimitar syndrome: an alternative approach. *J Thorac Cardiovasc Surg*. 2003;125:238-245.
3. Huddleston CB, Exil V, Canter CE, Mendeloff EN. Scimitar syndrome presenting in infancy. *Ann Thorac Surg*. 1999;67:154-159; discussion 160.
4. Kamiyama M, Kamata S, Usui N. Scimitar syndrome treated with pneumonectomy: a case associated with bronchospastic attack. *Pediatr Surg Int*. 2004;20:65-66.
5. Lam TT, Reemtsen BL, Starnes VA, Wells WJ. A novel approach to the surgical correction of scimitar syndrome. *J Thorac Cardiovasc Surg*. 2007;133:573-574.
6. Mason DP, Mihaljevic T, Mazzone PJ, Murthy SC, Rice TW. Extrapleural pneumonectomy for scimitar syndrome. *J Thorac Cardiovasc Surg*. 2006;132:704-705.
7. Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome: twenty years' experience and results of repair. *J Thorac Cardiovasc Surg*. 1996;112:1161-1168; discussion 1168-1169.
8. Vida VL, Speggiorin S, Padalino MA, et al. The scimitar syndrome: an Italian multicenter study. *Ann Thorac Surg*. 2009; 88:440-444.