

See Article page 208.



## Commentary: Novel repair technique for scimitar syndrome

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Geggel and colleagues<sup>1</sup> describe a multipatch repair technique for scimitar syndrome. This retrospective cohort study delineates excellent short- and midterm results. There were no mortalities and no incidence of pulmonary vein obstruction following their multipatch repair technique. The authors postulate that their multipatch repair enlarges the left atrium; avoids acute angles within the baffle pathway; and allows for durable, unobstructed pulmonary venous return to the left atrium.<sup>1</sup>

Scimitar syndrome is a rare subtype of partial anomalous pulmonary venous connection in which there is partial or total anomalous pulmonary venous connection of the right lung to the inferior vena cava (IVC). The characteristic chest radiograph appearance of a scimitar-like descending vein draining to the IVC gives the condition its name.<sup>2</sup> The right lung is occasionally hypoplastic, aortopulmonary collateral arteries to the right lung are common, and a large percentage of patients have an associated atrial septal defect. Presentation is bimodal: infants often present with failure to thrive, cyanosis, respiratory distress, and congestive heart failure, whereas adults may present with recurrent respiratory infections and occasional exertional dyspnea. In pediatric patients, repair is undertaken in a symptomatic child with substantial left-to-right shunt (Qp:Qs, >1.5:1), and right-sided chamber enlargement or pulmonary hypertension are relative indications for repair. Historically, there have been 2 common types of repair of scimitar syndrome: either disconnection from the IVC and reimplantation into the left atrium or intracardiac baffling.

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### CENTRAL MESSAGE

Novel repair technique for scimitar syndrome may minimize the development of pulmonary venous obstruction.

Unfortunately, the results of both repair techniques have been disappointing: Pulmonary vein obstruction occurs in as many as 50% of patients.<sup>3-5</sup>

In the series by Geggel and colleagues,<sup>1</sup> 22 patients underwent scimitar vein repair of which the novel, multipatch repair was used in half of patients (n = 11). The multipatch repair involves complete resection of the atrial septum (including the superior muscular limbus), shifting of the posterior left atrial wall rightward, maintenance of the scimitar vein connection to the IVC, and patch augmentation of the systemic venous return to the right atrium. The placement of these patches effectively increases the pulmonary and systemic venous pathway cross-sectional areas anteriorly while leaving the native connection of the scimitar vein to the IVC untouched. Remarkably, patients who underwent multipatch repair did not develop pulmonary vein obstruction during the follow-up period, whereas 5 out of 11 patients with the conventional repairs developed pulmonary vein obstruction. There was 1 instance of temporary postoperative arrhythmia in the multipatch group.

Geggel and colleagues<sup>1</sup> present a novel repair technique to management of scimitar syndrome. The described multipatch repair was efficacious and safe in the short- and medium term and it had significantly lower rates of pulmonary vein obstruction compared with reimplantation and intracardiac baffle techniques.<sup>1</sup> The results delineated by Geggel and colleagues<sup>1</sup> are very encouraging. We look forward to the authors providing future long-term follow-up regarding the development of pulmonary vein obstruction related to the multiple patches and the development of arrhythmias.

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