discharge. Of the 21, 14 (74%) were arrhythmia free at a mean follow-up of 2.8 years. Only 3 of the patient included in the study had anomalous pulmonary venous return. The European Congenital Heart Surgeons Association study group¹ reviewed 68 patients who underwent surgery for scimitar syndrome, and in their report only 1 patient underwent a Cox maze operation. The report unfortunately does not describe the details of the surgery or the effectiveness of the procedure.

CONCLUSIONS

The Cox maze 4 lesion set can be performed safely and effectively in patients with Scimitar syndrome. The lesion set needs to be modified in the setting of anatomic abnormalities.

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EDITORIAL COMMENTARY

The bad guy, good guy scenario with a scimitar sword: A case to learn from

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Today it would not be acceptable to close an atrial septal defect and leave the anomalous scimitar vein draining to the inferior vena cava (IVC). The case presented by Clarke and colleagues¹ in this issue of the *Journal* highlights how the care of patients with congenital cardiac defects has dramatically improved. Clearly, in the early 1980s when the first surgery was done for this patient, transesophageal echocardiography, magnetic resonance imaging, and computed tomographic scans were not routine in refining the diagnosis. In contrast, the use of one of these modalities is now essential in delineating anatomic pathophysiology.

A surgeon closing an atrial septal defect must go through the basics to check that the diagnosis is accurate. An oversized IVC should raise a flag of an abnormal drainage, and certainly failure of resolution of volume overload should raise another. We assume that this case also demonstrated the other features of scimitar

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Central Message

Repair of scimitar syndrome in adults with arrhythmia requires knowledge of defect morphology and lesion map for the maze procedure. Baffling or reimplantation could be used.

See Article page e105.

syndrome—namely, the arterial supply of the right lung, the dextroposition, the hypoplasia of the right lung, pulmonary hypertension, and other associated lesions such as horseshoe lungs—although the specific findings were not detailed in the report. Most of these features are present in the infantile form of the syndrome and may not be present in the milder adult form.

Surgical techniques have varied through the years, with outcomes varying with the type. The techniques ranged from reimplantation to baffling, with numerous

FIGURE 1. Schematic drawing for the technique of in situ pericardial repair of scimitar syndrome. SVC, Superior vena cava; SV, scimitar vein; LA, left atrium; RA, right atrium; IVC, inferior vena cava.

permutations of how these techniques are performed. Typically, pulmonary veins do not behave well with a circular anastomosis, and they are notorious for neointimal hyperplasia and recurrent obstruction, with this being manifested more in the infantile form. With the larger caliber pulmonary veins in adults, the results may be better. I have preferred baffling with autologous pericardium and augmentation of the IVC if needed. On a few cases we have used “in situ pericardial baffle” to route the scimitar

vein to the left atrium. Figure 1 depicts the technique. Cannulation of the IVC is done low, without dissection of the pericardial reflection posteriorly and without snaring the IVC. The junction of the scimitar vein and the IVC is opened and the posterior angle sutured together, making the angle less acute and moving the opening away from the lumen of the IVC. Then a generous opening in the wall of the left atrium is created behind the atrial septum, with the posterior lip everted to the back intact in situ pericardium. Finally, a patch of autologous pericardium is used to roof the baffle from the vein to the anterior lip of the left atrial opening.

Adults with congenital heart disease, repaired or unrepaired, who have arrhythmias now represent a growing population. These patients clearly need special attention and understanding of the anatomic variations and access. This case demonstrates what is needed to ablate atrial fibrillation during the repair of scimitar vein, with cut and sew, radiofrequency, and cryoablation lesion sets. Managing such arrhythmias requires innovation and knowledge of anatomy. The basic Cox maze principle should still be applied in such cases.

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