Direct implantation of scimitar vein to the left atrium via sternotomy: a reappraisal

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Abstract

OBJECTIVES: There is no consensus about optimal surgical technique for the repair of scimitar vein, an anomalous right pulmonary venous connection to the inferior vena cava. Our current experience with a direct anastomosis of the scimitar vein to the left atrium via sternotomy may be shared with other centres, but has not been widely published.

METHODS: Six consecutive patients (age 6 months to 17 years, mean 5 years) operated on in 2009–12 were retrospectively reviewed. Through median sternotomy and with cardiopulmonary bypass, the mobilized anomalous right pulmonary vein was brought through a large pericardial opening posterior to the right phrenic nerve and anastomosed onto the right side of the posterior left atrium with access via an existing or a surgically created atrial septal defect.

RESULTS: Five patients had primary venous repair and one had a previous failed repair using an intra-atrial baffle. The median cardiopulmonary bypass and cross-clamp times were 88 and 38 min, respectively. The median ventilator time was 1 day and the median stay at the intensive care unit 3.5 days. There were no deaths within a median follow-up of 28 months (range 8–41 months), nor reoperations or instances of pulmonary venous obstruction.

CONCLUSIONS: Anatomic repair of the scimitar vein based on reimplantation onto the left atrium via sternotomy is conceptually appealing. The surgery results in a safe and reliable repair in patients with a wide age spectrum. Durability needs on-going assessment in longer-term follow-up.

Keywords: Pulmonary arteries/veins • Surgery/techniques • Congenital heart disease • Lung/congenital lesions

INTRODUCTION

Scimitar syndrome is a rare congenital anomaly with a reported incidence of 2 in 100,000 live births. The main component of this syndrome usually consists of all pulmonary veins from the right lung forming a common curved channel in the shape of a scimitar sword that enters the inferior vena cava (IVC) at or just below the diaphragm, close to the orifice of the right hepatic vein [1]. Additional features may include hypoplasia of the right lung and dextroposition of the heart. Anomalous systemic arterial supply from the abdominal aorta to the lower segments of the right lung may also be seen. Approximately 70% of patients have an associated atrial septal defect [2]. Presentation is often with features of pulmonary hypertension, recurrent respiratory tract infection or congestive heart failure. Heart failure is more common in infancy and if present, often indicates an associated intracardiac shunt [3].

Surgical correction is usually indicated for patients’ symptoms, scimitar vein stenosis, pulmonary hypertension, the presence of significant left-to-right shunting, as well as right-heart dilatation. Anomalous systemic arterial supply to part of the right lung may often be occluded before surgery. Surgical repair can be challenging and complex; two main current surgical approaches are the intra-atrial baffle repair via a sternotomy utilizing deep hypothermic circulatory arrest in most cases, and direct reimplantation of the scimitar vein to the smallish and posteriorly situated left atrium via a right thoracotomy without cardiopulmonary bypass [4].

Our aim is to describe our current approach based on anatomic consideration and the ease of surgical access via sternotomy on normothermic cardiopulmonary bypass in almost all cases, securing a direct anastomosis of the scimitar vein to the left atrium. This alternative approach is not unique to our centre, but we believe this surgical technique deserves a reappraisal as an alternative option in view of the limitations of the two options mentioned earlier. It can be applied to the majority of patients including small infants and those who had a previous, but failed, intra-atrial baffle repair.
PATIENTS AND METHODS

This is a retrospective review of six consecutive patients operated on for anomalous pulmonary venous connections from the entire right lung to IVC (scimitar syndrome) between 2009 and 2012 by case note review. The institutional research board permission was sought to waive consent.

The preoperative characteristics of patients, as well as the length of their follow-up after the operation, are summarized in Table 1.

None of the patients was ventilated preoperatively; 5 of them were on anti-failure medications. The diagnosis was made in all patients using conventional transthoracic echocardiography. There were no genetic syndromes in any patients.

OPERATIVE TECHNIQUE

All patients received anaesthesia with endotracheal intubation and monitoring lines as needed for open heart surgery. The approach was through median sternotomy and cardiopulmonary bypass was established between ascending aortic cannulation and venous cannulations at the SVC and the low IVC. The IVC cannula was placed after its dissection at diaphragm to free the length of the IVC with its connection to the scimitar vein. In situations where the anomalous pulmonary vein drains to the IVC below the diaphragm, cannulation was done as it is done in usual circumstances and then the isolated anomalous vein was looped for control.

The left pleural cavity was opened widely for the leftward rotation of the cardiac mass. The right pleural cavity was opened electively and hilar dissection carried out to delineate the anatomy. Caution must be exercised as the phrenic nerve is very near the IVC and the hilum. The scimitar vein itself was isolated with a
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RESULTS

The median weight of patients was 9.1 kg (range 6-40 kg, 4 patients <10 kg). Five of six patients underwent primary repair, and one patient had a previous failed intra-atrial baffle repair. Repair was performed under mild hypothermia/normothermia in five patients and in one patient (Patient 2) with dextroposition and the presence of anomalous systemic venous drainage required deep hypothermic circulatory arrest to facilitate surgical access. The median cardiopulmonary bypass time was 88 min and median cross-clamp time was 38 min. All patients had intraoperative transoesophageal echocardiography to confirm a satisfactory pulmonary venous pathway at the end of repair.

There were no operative deaths and primary chest closure was achieved in all patients. The median ventilator time was one day and the median stay at the intensive care unit was 3.5 days (range 2-7 days). The early postoperative echocardiogram did not show any evidence of obstructed venous return from the right lung. Patients who had pulmonary hypertension preoperatively demonstrated reduction in estimated right-heart pressure postoperatively (Table 1). There were no reported phrenic nerve palsy, neurological complications, pleural effusions or postoperative arrhythmias.

The follow-up echocardiograms demonstrated good unobstructed venous return from the right lung with no residual shunts across the atrial septum. There were no late deaths with the follow-up being 100% complete at 8 months, median 28 months and range 8-41 months.

DISCUSSION

Most reports about the operative approach to correct scimitar syndrome are compiled retrospectively and span over a long time. A report by the European Congenital Heart Surgeons Association (ECHSHA) described 68 patients in 19 institutions who underwent surgery for scimitar syndrome over a 10-year period. It did not find any significant differences in terms of outcomes in patients treated mostly with the two main approaches of surgical management: the intra-atrial baffle repair via a sternotomy utilizing deep hypothermic circulatory arrest in most cases and direct reimplantation of the scimitar vein to the left atrium via a right thoracotomy without cardiopulmonary bypass [4, 5]. Fifteen patients in their direct implantation group were operated on through a sternotomy; however, the exact technique was not clearly stated. Similarly, Boston Children’s Hospital recently published their single-centre experience spanning 47 years and covering 80 patients diagnosed with the scimitar syndrome [6]. Thirty-six patients were operated on in their cohort, mostly by the baffle approach, but a few with direct reimplantation. The authors did not report any outcome differences between these groups of patients, nor was it evident whether the direct implantation was achieved through a sternotomy or a thoracotomy.

The surgical approach in many centres may be similar to what we describe in our article advocating an anatomic technique to relocate the scimitar vein, and this deserves a timely reappraisal. Our approach through sternotomy ensures that the scimitar vein is implanted in the most optimal location into the left atrium and helps in correction of all other associated anomalies, if present, and closure of atrial septal defect.

Despite the good mobilization of the ‘anchored’ scimitar vein in the right thoracotomy approach [5], the main disadvantage is the control of the relative remoteness of the left-sided and posteriorly situated smallish left atrium at the time of anastomosis. Direct implantation of scimitar vein by thoracotomy approach, as reported by Yilmaz et al. [7] and Brown et al. [5], does not necessarily involve cardiopulmonary bypass, but the associated intracardiac...
anomalies are not attended to. The risk of vein stenosis tends to remain despite direct implantation; the limited, and at times risky, surgical access could play a role. The suboptimal left atrial position for ‘side biting’ vascular clamping may undermine the quality of the anastomosis.

The traditional intra-atrial baffle repair with a patch obviously has its drawbacks, and deep hypothermic circulatory arrest is often needed during baffle correction. The sharp turn of the anomalous vein into the baffle predisposes to obstruction and thrombus, which remains a significant clinical problem especially in low-weight patients. The challenge may be further enhanced by the smallness of the scimitar vein with venous flow, and indeed this group of younger patients (<3 months) often encounters residual scimitar vein stenosis [4]. Modifications of baffle repairs include IVC transection and implantation as reported by Calhoun and Mee [8] but circulatory arrest is still needed during baffle correction.

In the contemporary ECHSA multicentre study, younger age at repair was a primary risk factor implicated in residual stenosis after the direct implantation technique [4]. However, it must be noted that in the ECHSA population, the weight of patients in the direct implantation group was significantly lower than in the baffle group, most likely reflecting the technical difficulty in executing a baffle repair in a small baby.

Overall, our patient population, albeit a small one, seems to compare favourably in terms of early results with the reported outcomes of these two mainstream approaches. Avoiding a baffle (or any foreign material) by direct implantation will reduce the incidence of scimitar vein stenosis or thrombosis. Since the large end-to-side anastomosis is carried out under direct vision across the atrial septum, and using a well-mobilized vein, the need for circulatory arrest is usually eliminated. The technique is suitable and reproducible for small infants and adult patients alike.

**CONCLUSION**

Anatomic reimplantation of scimitar vein into the left atrium via sternotomy under normothermic cardiopulmonary bypass offers very good early results across age groups. Although the numbers are small, this technique is conceptually appealing and our results are promising for treatment of this heterogeneous group of patients with anomalous pulmonary venous return, including reoperation and low weight at surgery. Durability needs on-going assessment in longer-term follow-up.

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**REFERENCES**


