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Surgical correction for scimitar syndrome by right thoracotomy and direct anastomosis in children

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ABSTRACT

Importance: Scimitar syndrome (SS) is a rare type of congenital heart disease characterized by total or partial anomalous venous drainage of the right lung to the inferior vena cava. However, the surgical repair techniques for SS vary according to patients' anatomical and pathological features.

Objective: This study was performed to analyze the mid-term results of a less invasive surgical correction technique for SS in children.

Methods: Eleven patients with SS who underwent surgical repair from January 2012 to March 2020 were retrospectively analyzed. The anomalous scimitar vein (SV) was directly reimplanted to the left atrium, and the concomitant atrial septal defect was simultaneously repaired with cardiopulmonary bypass.

Results: Three male and eight female patients were included in the study. Their mean age was 3.1 ± 1.3 years, and their mean body weight was 12.8 ± 3.0 kg. Most patients had symptoms, such as upper respiratory tract infection, dyspnea, and recurrent pneumonia, and two patients had pulmonary hypertension. None of the 11 patients who underwent direct SV reimplantation by right thoracotomy developed bleeding, arrhythmia, heart failure, or perioperative death, and no patients required reoperation during a mean follow-up period of 36.6 ± 15.2 months. Postoperative echocardiography revealed no restenosis or obstruction of the anastomosis in any patients.

Interpretation: Surgical repair for SS by right thoracotomy and direct anastomosis of the SV to the posterior wall of the left atrium is safe and effective, with good long-term patency of the reimplanted SV and a low mortality rate.

KEYWORDS
Scimitar syndrome, Congenital heart disease, Surgical repair, Right thoracotomy, Direct anastomosis

INTRODUCTION

Scimitar syndrome (SS) is a rare type of congenital heart disease characterized by total or partial anomalous venous drainage of the right lung to the inferior vena cava (IVC). The first reported case was published in 1836 by Cooper.[1] In 1960, Neill et al[2] described the scimitar sign and provided a detailed description of the syndrome. SS can be associated with several other abnormalities, including a hypoplastic right lung, dextroposition of the heart, and an anomalous systemic arterial supply to the right lung with or without pulmonary sequestration. SS can also be associated with other congenital heart defects, the most common of which is atrial septal defect (ASD). Other

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reported defects include coarctation of the aorta, ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and persistent left superior vena cava.[3] The term "scimitar" used to describe this syndrome is derived from the shadow created by the anomalous pulmonary vein on the chest radiograph. This shadow extends from the lateral superior position of the right lung to a more medial location and increases in caliber as it descends toward the cardiophrenic angle. The appearance of the shadow closely resembles that of a curved Turkish sword or scimitar.[2]

The goal of surgical repair in patients with SS is to reconstruct the right pulmonary venous return to the left atrium (LA). However, the surgical approaches vary according to the anatomic and pathologic features of each case. The most common procedure involves construction of a long baffle in the right atrium (RA) from the ostium of the scimitar vein (SV) to the ASD to reroute the venous blood flow to the LA. This procedure requires cardiopulmonary bypass (CPB) and with or without deep hypothermic circulatory arrest (DHCA). The primary stenosis of the SV or the stenosis at the confluence of the SV to the IVC may not be resolved. We herein describe 11 patients with SS who underwent treatment using a novel approach through a right thoracotomy with or without CPB.

METHODS

Ethical approval

The study was approved by the Ethics Committee of Beijing Children's Hospital (Number: 2020-Z-132) and was conducted in accordance with the Helsinki Declaration. This was a retrospective study and was exempt from informed consent from patients.

Inclusion and exclusion criteria

The inclusion criterion was confirmation of right anomalous pulmonary venous return to the IVC on preoperative chest radiography, echocardiography, and computed tomography angiography (Figures 1 and 2). The exclusion criterion was the presence of a supracardiac or intracardiac partial anomalous pulmonary venous connection on chest radiography, echocardiography, and computed tomography angiography.

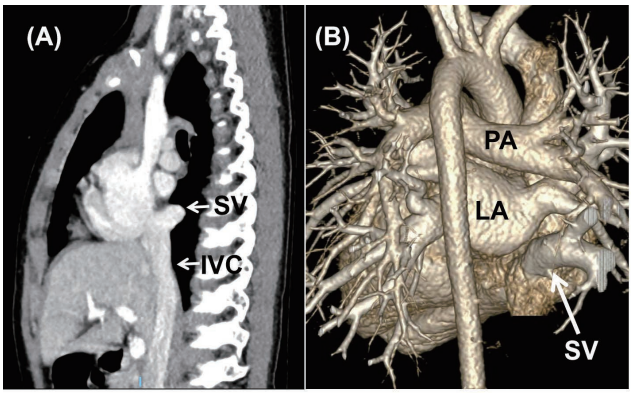

FIGURE 1 (A) Three-dimensional reconstructed image by maximal intensity projection (MIP) demonstrated anomalous pulmonary vein (scimitar vein) returns to the inferior vena cava (arrows). (B) Three-dimensional reconstructed image by volume rendering technique (VRT) showed scimitar vein returned to the inferior vena cava (arrow), other three pulmonary veins drained into the left atrium. SV, scimitar vein; IVC, inferior vena cava; PA, pulmonary artery; LA, left atrium.

Surgical procedures

The surgical procedures were performed through the direct anastomosis of the SV to the posterior aspect of the LA using a right thoracotomy with CPB for simultaneous repair of the ASD. The right thoracotomy was established and the pericardium was opened. Great vessel dissection was then performed in preparation for aortic and caval cannulation (Figure 3). During the dissection procedure, particular attention was given to the right phrenic nerve, which passed along the right side of the superior vena cava and continued inferiorly along the right side of the pericardial sac within the fibrous pericardium, anterior to the root of the right lung. The SV ran anterior to the hilum or coursed along the anterior surface of the right lung; in some cases, it was located within or ran along the posterior surface of the right lung. Limited hilar dissection was performed to free the anomalous pulmonary vein along its course from the lung to the IVC. In patients with right

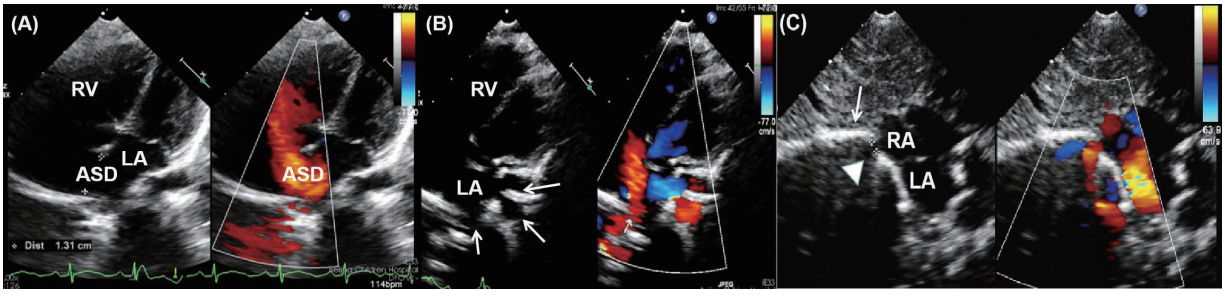

FIGURE 2 (A) Subcostal four chamber view with color Doppler showed the interatrial shunt and significant enlargement of the right atrium and right ventricle. (B) Subcostal four chambers view with color Doppler indicated the three pulmonary veins returned to the left atrium (arrows). (C) Subcostal view depicted the scimitar vein (arrowhead) drained into the inferior vena cava (arrow). ASD, atrial septal defect; LA, left atrium; RV, right ventricle; RA, right atrium.

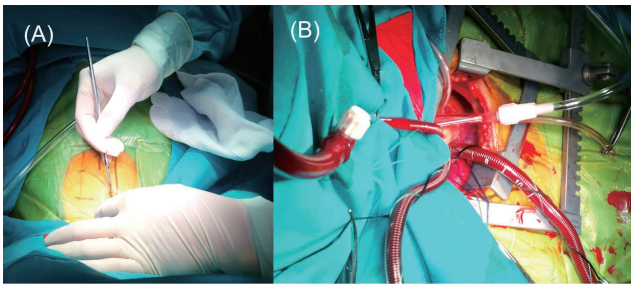

FIGURE 3 (A) Right thoracotomy started from the skin incision which was created obliquely between the anterior and posterior axillary folds with the upper point at the third intercostal space and the lower point at the sixth. (B) Cardiopulmonary bypass was successfully established by cannulation of the aorta, superior vena cava and inferior vena cava.

lower lobe sequestration, the systemic blood supply to the sequestrated lobe was detected and ligated. The aorta was cannulated and secured with a single purse-string suture. Bicaval venous cannulation was performed in a routine fashion with direct placement of the IVC cannula at the level of the hepatic vein entrance, thus establishing CPB. Under moderate hypothermia (defined as 33–36°C), the aorta was cross clamped and cold cardioplegic solution was delivered to the aortic root. The RA was opened through a longitudinal incision approximately 1.5 cm away from the atrioventricular groove, and the ASD was clearly identified. The SV was transected at the level of the diaphragm, or the diaphragm was incised from the IVC hiatus rightward and laterally to meet the point at which the SV entered the IVC. The distal end of the SV was closed with running suture. A small pericardial window was opened on the posteroinferior portion of the pericardial sac, which was posterior to the phrenic nerve; the window was located about 1.5 cm away from the phrenic nerve to avoid injury. The interatrial groove was dissected to expose a generous area of the LA. A small linear atriotomy was made on the LA. After mild trimming of the free end, the SV was opened longitudinally to increase the orifice size because it is frequently stenotic in patients with SS. The SV was then anastomosed to the LA in an end-to-side fashion with continuous 6-0 polypropylene running suture (Prolene; Ethicon Inc., Somerville, NJ, USA). The ASD was repaired by a bovine pericardial patch or polyethylene terephthalate patch (Dacron patch; DuPont, Wilmington, DE, USA) in the RA with 6-0 polypropylene running suture. The incision on the RA wall was closed in two layers using a 6-0 polypropylene running suture. Air was expelled before the suturing was completed. The aortic clamp was removed, and the patient was weaned from CPB. Finally, the right thoracotomy incision was closed.

In all patients, intraoperative transesophageal echocardiography demonstrated patent pulmonary venous return with blood flow of normal velocity through the reimplanted SV and the anastomosis.

RESULTS

We retrospectively analyzed the data of 11 patients who were diagnosed with and treated for SS in our hospital from January 2012 to March 2020. The patients comprised three male and eight female children. Their mean age at diagnosis was 3.1 ± 1.3 years (range, 7 months to 5 years), and their mean body weight was 12.8 ± 3.0 kg (range, 7.9–18.0 kg). The patients' clinical features and diagnostic findings, including their age at presentation, symptoms, associated anomalies, cardiac catheterization data, surgical treatment, and outcomes, are shown in Table 1.

Among the 11 patients with SS who underwent direct SV reimplantation by right thoracotomy, the major symptoms

TABLE 1 Clinical features and diagnostic findings of the 11 patients with scimitar syndrome

Patient No.	Age	Gender	Body weight (kg)	Symptoms	Associated cardiac defects	PAP (mmHg)	Qp/Qs	Flow rate at the anastomosis (cm/s)	Outcome and F/U (month)
1	7 mo	M	8.1	Recurrent pneumonia	ASD	50	1.9	82	Patent/8
2	11 mo	F	7.9	Recurrent pneumonia, heart murmur	ASD	39	2.2	109	Patent/17
3	2 y 6 mo	M	10.8	Recurrent pneumonia	ASD	19	1.5	97	Patent/27
4	3 y 6 mo	F	12.9	URT infection	ASD	26	1.8	116	Patent/29
5	4 y	F	13.7	Respiratory failure	ASD	29	2.1	67	Patent/33
6	4 y	F	13.5	Heart murmur, tachypnea	ASD	31	2.2	85	Patent/39
7	4 y	F	15.5	Recurrent pneumonia, respiratory failure, distress	ASD	28	2.9	122	Patent/41
8	2 y 6 mo	F	11.5	URT infection	ASD	42	2.4	79	Patent/43
9	5 y	F	18.0	Heart murmur	ASD	26	1.7	105	Patent/49
10	3 y 6 mo	M	13.0	Recurrent pneumonia	ASD	37	2.3	119	Patent/55
11	3 y 6 mo	F	16.2	Cough, dyspnea	ASD	33	2.1	126	Patent/62

M, male; F, female; PAP, pulmonary artery pressure; Qp/Qs, pulmonary to systemic flow ratio; F/U, follow-up; URT, upper respiratory tract; ASD, atrial septal defect.

were pneumonia and recurrent upper respiratory tract infection. At the time of preoperative cardiac catheterization, the median systolic pulmonary artery pressure was 32.7 ± 8.3 mmHg (range, 19–50 mmHg), and six patients had mild to moderate pulmonary hypertension (≥30 mmHg). No aortopulmonary collateral arteries were found on cardiac catheterization in any patients. The preoperative median pulmonary blood flow to systemic blood flow ratio was 2.1:1 (range, 1.5:1–2.9:1). An ASD was observed in all 11 patients, and all were repaired simultaneously; the mean CPB time was 87 ± 22 minutes (range, 53–126 minutes), the mean aortic cross-clamping time was 68 ± 21 minutes (range, 39–110 minutes). The patients recovered steadily after surgery without bleeding, arrhythmias, heart failure, perioperative death, or any other complications.

All patients were followed up every 3 to 6 months by echocardiography. During the mean follow-up period of 36.6 ± 15.2 months (range, 8–62 months), echocardiographic examination proved that all reimplanted SVs were patent without evidence of stenosis or obstruction, and the velocity at the venoatrial anastomosis was normal in all 11 patients (Table 1). No consistent pulmonary hypertension, heart failure, reoperation, or late mortality occurred.

DISCUSSION

SS is a rare and complicated condition associated with congenital cardiopulmonary defects. Previous reports have indicated that the estimated prevalence of SS is 1 to 3 per 100 000 births.[4] However, this prevalence is probably underestimated because many patients have no symptoms and therefore remain undiagnosed. SS can be seen in infancy, childhood, or adulthood. Although patients with SS may have no or minimal symptoms, presentation in infancy is usually due to severe heart failure, pneumonia, or upper respiratory tract infection from a significant left-to-right shunt, and onset in infancy has a poorer prognosis than onset in adulthood. This poorer prognosis may be due to the shunt from the anomalous pulmonary venous return, but it is more likely a result of a concomitant congenital heart lesion, such as an ASD, which is the most commonly associated cardiac anomaly in patients with SS and was present in all of our patients.[3] Dupuis et al[4] classified SS into 2 forms: infantile and childhood/adult. Patients with infantile form are younger than 1 year and always present with severe heart failure, pneumonia, or upper respiratory tract infection from a significant left-to-right shunt. Patients with childhood/adult form are older than 1 year and presented with recurrent respiratory infection with relatively good heart function. Therefore, our preference is to repair the ASD or other congenital heart defects simultaneously through the right thoracotomy incision, which resolves the left-to-right shunt problem in a single procedure.

The diagnosis of SS should be considered if an infant presents with failure to thrive, cyanosis, respiratory distress, or congestive heart failure in association with dextroposition of the heart and hypoplasia of the right lung upon birth.[5] The diagnosis may be difficult because of the wide range of symptoms at clinical onset, especially in children with concomitant congenital heart lesions. Therefore, preoperative catheterization should be considered to confirm the diagnosis, identify the specific course of the anomalous pulmonary venous drainage, determine the presence of SV stenosis and pulmonary hypertension, measure the degree of the left-to-right shunt, and detect any associated cardiac abnormalities.[4]

The indications for correction of SS may include congestive heart failure, recurrent pneumonia, a pulmonary-to-systemic blood flow ratio of >1.5, or stenosis of the anomalous pulmonary vein with pulmonary hypertension, because stenosis of the SV may cause pulmonary hypertension in infants.[4,6] Furthermore, it is important to reroute the SV and repair the associated cardiac defects, such as a ventricular septal defect or an ASD. More than half of the patients in our study had an elevated pulmonary artery pressure of ≥30 mmHg at the time of preoperative catheterization. The symptoms of tachypnea, respiratory distress, and even repeated respiratory infection were improved in most of our patients after rerouting the abnormal pulmonary veins and repairing the associated ASD because the left-to-right shunt with pulmonary volume overload was abolished.

Although no cases of pulmonary sequestration were observed among the 11 children in our series, pulmonary sequestration or anomalous systemic arterial supply to the lower part of the right lung can be seen in some patients with SS.[7] In such cases, the sequestered piece of lung is a mass of abnormal lung tissue that does not communicate anatomically with the tracheobronchial tree, and it is often supplied by an anomalous artery, usually from the descending aorta. The venous drainage is mainly via the SV to the IVC. The diagnosis of pulmonary sequestration is confirmed by thoracotomy or preoperative enhanced computed tomography. The indication for resection of the sequestered lobe is chronic recurrent infection caused by the significant left-to-right shunting. Once the sequestered piece of lung has been resected, the SV was no longer to be reimplanted.

Several surgical correction techniques for rerouting the anomalous right pulmonary venous return to the LA have been reported, with most requiring DHCA. The choice of technique remains highly surgeon-dependent.[8,9] The procedure can be performed under direct implantation of the anomalous vein to the LA, which was done in the present study.[4,8] Alternatively, an intracardiac patch can be created to baffle the blood flow from the anomalous vein to the LA through the ASD.[9,10] Moreover, the anomalous

pulmonary vein can be dissected and reimplanted into the RA and an intracardiac baffle can be created to redirect the blood flow to the LA.[6] However, regardless of the repair procedure used, the poor outcomes in many surgically treated patients are attributed to thrombosis of the venoatrial anastomosis. Pulmonary venous stenosis, obstruction, or thrombosis remain the major obstacles to successful repair in patients with SS. The primary challenge of direct reimplantation is how to maintain the correct orientation of the SV so that it does not kink at the hilum when the lung is reinflated because the vein may become more horizontal rather than vertical during this transposition. Distortion of this vessel can obstruct blood flow with consequent thrombosis, which may lead to immediate infarction of the right lung, pulmonary hypertension, and hemoptysis.[11] In such cases, subsequent lobectomy or right pneumonectomy is often necessary as a bail-out procedure to treat these complications.[4] We usually stitch two to three markers using 5-0 polypropylene suture longitudinally along the SV to ensure that the reimplanted SV lies correctly before initiation of the anastomosis. The small window on the pericardial sac and the small incision on the LA wall should be made near the SV to avoid tension at the anastomosis site. A potential surgical complication during baffle repair is obstruction or stenosis of SV at the junction of the anomalous vein to the IVC and thrombosis caused by the primary stenosis of the SV.[12] The reimplantation procedure can avoid this because it provides a short, straight pathway to the LA. The orifice of the vein can also be enlarged by a longitudinal incision during transection of the SV or before the anastomosis procedure. Several stitches can also be placed using 5-0 polypropylene suture near the phrenic nerve in patients with SV stenosis; this may help the surgeon to recognize the nerve and thus avoid injury. During the average follow-up period of 36.6 ± 15.2 months in the present study, all 11 reimplanted SVs remained patent without any stenosis or thrombosis.

Continuous CPB or DHCA can be used for SS repair. All of our 11 patients had a concomitant ASD. No other associated cardiac defects need to be repaired. Compared with the long baffle repair procedure, there is no need to manipulate the IVC by our direct anastomosis to the LA. Therefore, we can avoid DHCA, and its associated risks, such as stroke, respiratory failure, and renal failure.

A median sternotomy is the traditional approach in correcting SS and provides the advantage of excellent exposure.[8-10] However, this conventional approach is associated with morbidity, postoperative discomfort, and a large midline scar, which may evoke psychological distress, especially in young female patients. Therefore, transition from standard median sternotomy to a less invasive incision in the repair of SS is very important. We completed all 11 correction procedures in this study through a right thoracotomy, which not only has superior cosmetic results but is also associated with low morbidity and mortality. No patients in our study developed postoperative hemothorax, needed a reoperation for bleeding, or died during the perioperative period. This technique leaves the sternum intact, which may increase the postoperative recovery rate.[13]

Several reports have described similar cases and treatment techniques but with small numbers of pediatric and adult patients.[5-8] Our report of 11 patients is the largest series among all published pediatric series of direct implantation by right thoracotomy. We recommend this simpler procedure, in which baffles are no longer necessary. But there are still some limitations in our study. It is a retrospective study from a single center, the selection bias and the limited patients' number might reduce the accuracy of the results. A multicenter and prospective study is required to improve the results.

In summary, SS is a very rare congenital heart disease. If patients with SS present during infancy or have other associated congenital heart diseases that make them appropriate surgical candidates, operative repair to correct the abnormal SV return may be recommended. Division of the SV and reimplantation into the LA for correction of SS is safe and effective, with favorable long-term patency of the reimplanted SV and a low mortality rate.

CONFLICT OF INTEREST

All Authors declare that they have no conflict of interest.

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