Surgical Management of the Scimitar Syndrome

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Abstract

The morphology variations of the so-called scimitar vein are many and varied. We present a synthesis of 92 published investigations of the overall scimitar syndrome. We reviewed the clinical presentations, diagnostic modalities, surgical approaches, and outcomes. Diagnostic information was provided by clinical presentations, radiographic findings, transthoracic and transesophageal echocardiography, computed-tomographic angiography, magnetic resonance imaging, angiocardiography, and ventilation/perfusion scans. These investigations served to elucidate the origin, course, and termination of the scimitar vein, the intracardiac anatomy, the presence of associated defects, and the patterns of any accompanying pulmonary lesions. In short, they defined the disease prior to surgical intervention. Of the patients described, up to four-fifths presented during infancy, with cardiac failure, increased pulmonary flow, and pulmonary hypertension. Associated cardiac and extracardiac defects, particularly hypoplasia of the right lung, are present in up to three-quarters of cases. Overall operative mortality has been cited between 4.8% and 5.9%. Mortality was highest in patients with preoperative pulmonary hypertension, and those undergoing surgery in infancy. Despite timely surgical intervention, post-repair obstruction of the scimitar vein, intra-atrial baffle obstruction, or stenosis of the inferior caval vein were reported in up to two-thirds of cases. The venous obstruction could not be related to any particular surgical technique. On long term follow-up, one sixth of patients reported persistent dyspnoea and recurrent respiratory infections. Any infants presenting with heart failure, right-sided heart, and hypoplastic right lung should be evaluated to exclude the syndrome. An increased appreciation of variables will contribute to improved surgical management.

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Running title: Scimitar syndrome

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Mortality was highest in patients with preoperative pulmonary hypertension, and those undergoing surgery in infancy. Despite timely surgical intervention, post-repair obstruction of the scimitar vein, intra-atrial baffle obstruction, or stenosis of the inferior caval vein were reported in up to two-thirds of cases. The venous obstruction could not be related to any particular surgical technique. On long term follow-up, one sixth of patients reported persistent dyspnoea and recurrent respiratory infections. Any infants presenting

with heart failure, right-sided heart, and hypoplastic right lung should be evaluated to exclude the syndrome. An increased appreciation of variables will contribute to improved surgical management.

Key words: Scimitar syndrome, Anomalous pulmonary venous connection, Pulmonary sequestration, Horse-shoe lung, Congestive heart failure, Hypogenetic lung syndrome, Congenital pulmonary venolobar syndrome, Vena cava bronchovascular syndrome, Mirror-image lung syndrome

Introduction

The Scimitar syndrome is a rare congenital malformation characterized by totally or partially anomalous pulmonary venous connection of the right lung to the inferior caval vein or right atrium, variable hypoplasia of the right lung, a right-sided heart with normally related cardiac chambers, and variable systemic arterial supply to the right lung.[1-7] The phenotypic feature is the anomalous pulmonary venous connection, which produces the characteristic shadow on the chest radiograph that resembles the Turkish scimitar.[8-10]

The incidence of the syndrome is relatively low, representing no more than around one-twentieth of all cases of partially anomalous pulmonary venous connection.[7] The estimated prevalence is from 1 to 3 per 100,000 births. It is associated with other congenital cardiac malformations in up to three-quarters of cases.[11-13]The first description of the findings now known as the Scimitar syndrome was made at autopsy by Cooper and Chasinat in 1836. The findings were then recognised in an asymptomatic live patient in 1949 by Dotter.[1,2,14] It was Halsz and associates, however, who first used the term "scimitar", in 1956, when describing the appearance of the abnormal vein. Neil and associates, in 1960, then named the syndrome after the Turkish sword.[15,16] A right lower lobectomy in a patient with the findings now recognised as representing the syndrome had been performed in 1950 by Drake and Lynch,[17] while Kirklin, Ellis and Wood had corrected the overall findings in 1956.[18]

Due to the small number of reported cases, and limited description of clinical details, it is difficult to draw conclusions regarding the indications and outcomes for surgical as opposed to non-surgical management. For the purposes of our review, however, we have evaluated all clinical and necropsy studies describing individuals recognised as having the syndrome and undergoing either conservative or surgical management (Table E1).

We assessed the locations of the anomalous pulmonary venous connection to supradiaphragmatic or infradiaphragmatic inferior caval vein, the course of scimitar vein in relation to the hilum of right lung, the degree of hypoplasia of the right pulmonary veins, the source of anomalous arterial supply to right lung, any associated congenital cardiac lesions, the degree of hypoplasia of either the right or left lung, and the size and pattern of drainage of the left-sided pulmonary veins. This has permitted us to collate the anatomical details, the diagnostic challenges, the associated cardiac anomalies, and the techniques and outcomes of management, including reinterventions.

Methods

We searched the literature to identify the described instances of scimitar syndrome, with detailed anatomical description, associated cardiac and non-cardiac anomalies, any association with so-called heterotaxy syndrome, the surgical and non-surgical therapeutic options, and their outcomes. The search engines employed were PubMed, Google Scholar, Cochrane Database for Systematic Reviews, Cochrane Central Register of Controlled Trials, Ovid Medline, ACP Journal Club, Ovid EMBASE, and Database of Abstracts of Review of Effectiveness in all languages.

Analysis was based initially on the individualized review of 91 investigations.[1-4,6-20,E1-E72] These were then incorporated from the results of the retrospective study emanating from the multi-centric European Registry, which itself had collated the findings from 485 patients from 51 institutions, being carried out under the auspices of the Association for European Pediatric Cardiology and the European Congenital Heart Surgeons Association.[5,E72]

Due to the limited size of the reported series, and the heterogeneity of the clinical state at the time of surgical intervention, along with the difficulties in selection of appropriate cardiac quantifiable end-points, we were

unable to perform a meta-analysis. Our analysis therefore, is based on review of case reports and small case series, along with incorporation of data from the more extensive series reported from the Joint Multicentric Registry.[1-20,E1-E72]

Morphogenesis

The errors accounting for this developmental constellation are not well understood. As the lung bud development progresses, its primary blood supply changes from a plexus derived from the descending aorta to the portion of the ultimate pharyngeal arches that give rise to the pulmonary arteries, a transition that is complete after the 7^{th} week of gestation. With normal development, pulmonary venous connection to the left atrium is completed by the 11^{th} week of gestation. In 1999, Huddleston and Mendeloff suggested an insult could have occurred which would culminate in failure of this "hand-over", resulting in the persistence of systemic arterial supply to the right lung from the abdominal aorta, and underdevelopment of the right pulmonary artery and right lung.[E2] The syndrome represents a more primitive derangement than the usual forms of partially or totally pulmonary venous connections. It has been suggested that it may be related to disturbed laterality signalling.[E50]

Results

Demographics

The age of reported patients ranges from 1 day to 72 years. In the Joint Multicentric Registry, median age at diagnosis was 0.48 years, with an interquartile range from 0.1 to 5.2 years. Among 485 reported cases, with a ratio of 1.4 females to 1 male, almost three-fifths were neonates or infants, around one-quarter were aged between 1-10 years, and one-fifth were older than 10 years.[3-5] There was no identifiable regional or ethnic predominance, with isolated reports of bronchial isomerism and familial occurrence were seen.[11,12,16,E22,E25,E26]

The clinical presentation varies from being asymptomatic to significant heart failure. The age at presentation can accordingly differ significantly from infancy to adulthood.[E27] Some have sub-classified the syndrome into the groups of isolated presentation in infancy, presentation in infancy with congenital heart diseases, and isolated presentation during adulthood (Table E1).[3,11,12] In this regard, half of adults or adolescents reported in the multicentric European study present as isolated cases with a slight degree of right pulmonary hypoplasia, one-third were asymptomatic, while half reported a history of recurrent infections of the respiratory tract.[3-5,9,11,12,E2,E20-E22]

Diagnosis

The diagnosis is straightforward when the scimitar sign is recognised on the chest radiograph in association with hypoplasia of the right lung, a right sided mediastinal shift, a right-sided heart, or an opacified right hemithorax.[20,E27,E49] On occasion, however, a meandering but normally connected pulmonary vein may be confused for a scimitar vein.[E27,E49,E50] The scimitar sign, nonetheless, may be absent due to hypoplasia of the right lung and a right-sided heart, or a prominent thymus, these changes obfuscating the venous shadow.[E27,E49,E51]

Accurate anatomical information prior to surgery is desirable, as the surgical strategy depends on the configuration and site of drainage of the scimitar vein. The restricted imaging windows limit the usage of transthoracic echocardiography in assessing the exact site of drainage in the prenatal period, and in adults.[E29,E51,E52]Transesophageal echocardiography is useful in confirming the diagnosis preoperatively, and in postoperative assessment of the patency of the scimitar vein.[E18,E32,E49,E52-E55,E57]

On duplex sonography, the normal pulmonary venous flow is biphasic or triphasic, with one or two peaks in systole, one peak in diastole, and reverse flow at atrial contraction. The flow in the scimitar vein, however, is monophasic, extending throughout the cardiac cycle with no reverse flow with atrial contraction. This feature is better visualized by transesophageal echocardiography.[E18,E51-E53,E56]Shibuya and associates were

able to characterize the anomalous venous connection in nine-tenth of their cases when using color flow mapping, as opposed to only one-sixth without.[E52]

Contrast computed tomography, and/or 3-D magnetic resonance angiography, now provide comprehensive assessment of the intracardiac anatomy, the diameter, trajectory, and site of drainage of the scimitar vein, and its relationship to the hilum of right lung (Figures 1 & 2). The technique may now safely be used to replace conventional angiography for definitive diagnosis.[E54,E55,E57-E59,E61]

Angiocardiography may still be indicated in doubtful cases, for coil embolization of the aortopulmonary collateral arteries, or for therapeutic intervention should the scimitar vein be stenosed.[3-5,11,12,E20,E21] Ventilation/perfusion scans are recommended as a routine by some investigators for pre- and postoperative evaluation.[E17] Routine bronchoscopy and bronchography are also recommended by others.[E17,E28]

Surgical Anatomy

The Scimitar vein typically drains the entire right lung, but may occasionally drain only the lower or middle lobe.[E28]It can course either anterior or posterior to the hilum of the right lung. It is not always be wide and curved like a scimitar, but rather may be straight, thin or multiple. It connects usually to the inferior caval vein, but cases are described with connection to the superior caval vein or the right atrium.[19,E2,E22,E29] When connecting to the inferior caval vein, the site of connection is usually subdiaphragmatic, but can be supradiaphragmatic or to the inferior cavoatrial junction.[E30] The vein was reported as stenotic in one-fifth of patients, this being a harbinger for a poor prognosis.(Figure 2)[E50]

Left-sided scimitar veins have also been described.[6-10] For example, in 2005, Juraszek and associates described an isolated case in whom all the left pulmonary veins joined together in a confluence below the left atrium, and then drained subdiaphragmatically into the right-sided inferior caval vein.[8] Such left-sided connections are less frequently obstructive, but may cause refractory pulmonary hypertension.[9] Pulmonary arterial hypertension may occur in one-third of all cases, being observed mainly in neonates and infants with congenital heart diseases, but only rarely in isolated cases.[3-5,11,12]

Associated cardiac and extracardiac anomalies were reported in one-third to three-quarters of patients.[E31] Cardiac anomalies included atrial and ventricular septal defects, functionally univentricular hearts, double outlet right ventricle, tetralogy, tricuspid atresia, anomalous origin of the left coronary artery from the pulmonary trunk, double chambered right atrium, left-sided obstructive lesions, and stenosis of the inferior caval and pulmonary veins.[3-5,E32,E33,E40-E72]

Associated extracardiac anomalies included patency of the arterial duct, coarctation of aorta, arch obstruction, abnormal lobation of the right lung, hypoplasia of the right lung, pulmonary sequestration, the scimitar vein draining the entirety of both lungs, interruption of the inferior caval vein, obstruction of the scimitar vein at its caval connection, pulmonary arterial hypoplasia, and aortopulmonary collateral arteries.[E34-E36] Hemivertebras, proximal rib fusion, subnumerary ribs, and micromelia of the arms have all been reported at necropsy.[54-56] Hypoplasia of the right lung to varying degree has been reported in cardiac position.[3-5,E72]

An anomalous arterial supply from the descending thoracic or abdominal aorta typically perfuses the lower or middle lobes, but may infrequently perfuse the whole right lung.[E37-E39] Hyperperfusion of the left lung was reported in three-quarters of cases on pulmonary scintigraphy.[E28] Tracheobronchial abnormalities in the form of sequestration, diverticulums, stenosis or atretic bands have been reported. There are no reported cases with a normal bronchogram.[E28] Najm and associates reported just over two-fifths incidence of pulmonary sequestration in their series of 32 patients.[19] Bronchial sequestration, however, is rarely present.[7,E17] There are, nonetheless, 14 patients reported with horseshoe lungs, the two lungs being fused at the postero-inferior segments posterior to the heart and in front of esophagus.[11,12,19,E41-E43] The right pulmonary artery and right bronchus usually cross the midline to supply part of the left lung.

Surgical Approaches and Management

Management, including indications, timing, and type of intervention remain nebulous.[3-5,11,12,19-20,E2] During

infancy, unless pulmonary hypertension develops, medical treatment is indicated to offset heart failure, and allow growth before surgical repair. Pulmonary hypertension, however, must be addressed before surgery, either by medical intervention or by reducing pulmonary blood flow by coil embolization of aortopulmonary collateral arteries.[3-5,11,12,E72] Some authors recommend early surgical intervention if cardiopulmonary bypass and cardiac arrest are not used in the repair procedure.[E7] In general, surgical correction is recommended in all symptomatic patients, and in asymptomatic patients with pulmonary-to-systemic flow ratios greater than 1.5:1, or pulmonary-to-systemic flow ratio lesser than 1.5:1 in the settings of clinically treated pulmonary hypertension, stenosis of the scimitar vein, or concomitant cardiac lesions.[19,E1,E21]

The indications for surgery in adults is still debated, since many adults with the syndrome lead a normal life without surgical treatment. The general consensus, nonetheless, is that surgery is indicated in symptomatic patients, or when the pulmonary-to-systemic flow ratio is greater than 1.5 in asymptomatic patients. Diuretics, sodium potassium/ATPase inhibitors, β-agonist, ipratropium bromide, antibiotics and cortisones are used on an individualized basis.[3-5,11,12,E72]

Principles of surgical treatment are creation of an unrestricted pathway for the scimitar vein through an existing or newly created interatrial communication, without causing tension, torsion, kinking, flattening, or stenosis of the scimitar vein, Concomitant anomalies are also repaired. In symptomatic patients when the vein cannot be rerouted to the left atrium, lobectomy, pneumonectomy, or heart lung transplantation are considered according to the situation. Multiple individual techniques are reported for surgical correction (Table E2).

Division and reimplantation of the scimitar vein into the right atrium without cardiopulmonary bypass

In 1956, Kirklin, Ellis and Wood reported the first total correction of the lesions making up the syndrome by translocating the scimitar vein to the right atrium without utilizing bypass. By means of a Bailey atrioseptopexy, the flow was then directed to the left atrium through the oval foramen.[18,E3]

Division and translocation of the scimitar vein into the right atrium

Shumacker and Judd described this technique in 1964. They divided the Scimitar vein along with a cuff of the inferior caval vein, transplanting it to the right atrium adjacent to the oval fossa such that flow could be directed to the left atrium. The cuff of caval venous wall provides more robust tissue for anastomosis. The inferior caval vein itself was patched to avoid stenosis. The technique has since been used with satisfactory results (Figure 3).[13,19,E5,E6]

Direct implantation of the scimitar vein to the left atrium via median sternotomy

This involves mobilisation of the vein, bringing it through a large pericardial opening posterior to phrenic nerve, and anastomosing it to the left atrium.[E66]

Division and translocation of the scimitar vein into the left atrium via right thoracotomy without cardiopulmonary bypass

This technique was described for both infants and adults without concomitant cardiac anomalies. Through a right anterolateral thoracotomy, scimitar vein was harvested with a venous cuff and sutured to the left atrium (Figure 4).[E2,E6-E8]

Division and translocation of the scimitar vein into the left atrium using Dacron graft interposition via right thoracotomy.

This technique was described in 1968, interposing a 20 mm Dacron graft.[7,19]

Division and translocation of the scimitar vein into the left atrium using polytetrafluoroethylene graft interposition

Lam and associates anastomosed a polytetrafluoroethylene graft between the scimitar vein and the left atrium (Figure 5).[E9]

Division and translocation of the scimitar vein into the left atrium via pericardial tunnel

This innovative method was described in 2014 by Lugones and Garcia. The scimitar vein was transected at the level of diaphragm and both ends were over sewn. A large incision was made in right lateral wall of the pericardium. Then under cardioplegic arrest, a pericardial anastomosis was created to a large vertical left atriotomy (Figures 6A-6C).[E10]

Partitioning of the inferior caval vein and rechanneling of the scimitar vein into the left atrium

In 2003, Calhoun and Mee used a pericardial patch to partition the inferior caval vein,[E11] creating a posterior compartment receiving the scimitar venous blood which was sutured to the left atrium. The anterior compartment receiving the systemic venous blood remained connected to the right atrium (Figures 7A, 7B).

Intra-atrial baffle

Either polytetrafluroethylene[E12] or the free wall of the right atrium[E13] have been used to baffle the orifice of scimitar vein through an existing or newly-created interatrial communication to the left atrium (Figures 8A, 8B).

Division and translocation of scimitar vein into the left atrium with venous flap.

This technique involves harvesting the Scimitar vein together with a venoatrial flap and anastomosing it directly to the left atrium (Figure 9).[E65]

Division and translocation of the scimitar vein using a ringed polytetrafluoroethylene extracardiac conduit

This technique involves creation of a pericardial window, with a ringed polytetrafluroethylene interposition graft then sewn end-to-end between the divided scimitar vein and the left atrium (Figure 10).[E70]

Atrial septectomy, and intra-atrial baffling

This technique, employing a polytetrafluroethylene patch was used in a patient with an intact atrial septum.[E14]

Scimitar vein cut back technique

The technique of cutting up into the right atrium along the venoatrial junction resulted in taking away the angle from the anastomosis with the left atrium, thus decreasing the risk of stenosis at that site.[19,20]

Pericardial tunnel technique

This technique was used for the vertical scimitar vein, defined as an angle of insertion to the inferior caval vein of less than 45°.[E69] Under cardioplegic arrest, flow was directed to the left atrium inside a pericardial tunnel.[E69]

Repair by a novel multipatch technique

This novel operation enlarges the left atrium, and avoids circuitous pathways or tension on scimitar vein. The operation included resection of the atrial septum, making a V-shaped incision made at the venoatrial junction or into the scimitar vein, and anastomosing it to the left atrium. An autologous pericardial patch was used to separate the atriums, with an additional patch needed to augment the inferior caval vein.[E68]

In situ pericardial roll repair for distant anomalous pulmonary venous return

This innovative technique, using dual pericardial rolls, is suitable for multiple anatomic configurations, and can be used for both infants and adults.[E64]

Modified in situ pericardial rerouting and repositioning of atrial septum

This approach is a modification of the so-called "sutureless" repair. The atrial septum was repositioned to create a wide opening in the lateral aspect of the left atrium, ensuring an adequate pathway.[E67]

Pneumonectomy

Right pneumonectomy has been advised when the scimitar vein courses posterior to lung hilum to emerge from under the diaphragmatic surface of right lung. In this setting, it is difficult to mobilize the vein without kinking or twisting.[13,E2, E15-E17] Other indications for pneumonectomy are post-repair thrombosis or fibrosis of the scimitar vein, or recurrent pulmonary venous stenosis.[13,E15-E17]

Lobectomy/resection of sequestered lung

Several authors have performed lobectomy for pulmonary sequestration with recurrent right-sided pneumonias.[E17,E18]

Lung transplantation

Transplantation has been recommended for persistent postoperative pulmonary hypertension and recurrent pulmonary venous stenosis following surgery.[9,E2]

Orthotopic cardiac transplantation

A hybrid approach, or early cardiac transplantation, was recommended in the setting of a functionally univentricular heart and a severely hypoplastic pulmonary artery.[13,E19]

Correction of associated cardiac anomalies

The associated congenital cardiac anomalies are treated on their own merits.[1-5,E72]

Transcatheter therapy / surgical ligation of major aortopulmonary collateral arteries

Aortopulmonary collateral arteries are managed by either embolization using detachable mini balloon, mini coils, or tissue adhesive or by surgical transthoracic ligation. Balloon angioplasty and balloon expandable stainless steel Palmaz stent have been employed for stenotic anomalous pulmonary veins.[3-5,9,13,19,E2,E20-E24,E60-E72]

Short- and Long-term Outcomes

Early surgical outcomes are good for most individuals undergoing surgical correction. The overall risk of surgical repair is between 4.8% and 5.9%.[5,11,13,19,E2,E5-E7,E10,E17,E22] The causes of death were presentation in infancy with concomitant congenital cardiac and extracardiac anomalies, severe pulmonary arterial hypertension, severe pulmonary hypoplasia with sequestration, respiratory failure and congestive cardiac failure. Patients undergoing resective procedures have a higher risk of death compared with those undergoing corrective surgery (36% vs 3%) (Table E3).[5,11,13,19,E2,E5-E7,E10,E17,E22]

Those patients with severe pulmonary hypoplasia, diffuse right-sided bronchiectasis, post baffle thrombosis / fibrosis of scimitar vein, recurrent pulmonary vein stenosis following dilatation / stenting and hypoplastic left heart syndrome continue to present a significant challenge.[3-5,E6] Despite timely surgical intervention, operative mortality remains high in this subset of patients, and heart / heart-lung transplantation may be the only surgical option.[9,13,E2,E19]

Regardless of the surgical techniques, venous or baffle obstruction are reported in between one-sixth and two-thirds of cases.[3-5,9,E6,E22,E23,E62] The specific technique did not seen to influence the rate of postoperative pulmonary venous obstruction.[3-5,9,11-13,19,E6,E22,E23,E62-E64,E71,E72]

In the joint European Registry, over nine-tenths of patients underwent corrective surgery, with the remaining patients having lung resection. In the series, almost half the patients were medically treated. At a mean follow-up of 7 years, late mortality was 4% in those surgically treated, and 6.5% in medically treated patients. Postoperatively, one-fifth of patients had scimitar venous stenosis, and 2% had total occlusion. This complication was related to neither the baffle nor reimplantation, and was more frequent in neonates

and infants. Two-thirds of these patients underwent reintervention or reoperation at a median of 0.8 years after repair, with an interquartile range from 0.5 to 1.2 years.[5] Persistent dyspnoea and recurrent respiratory infections were documented in 15% of surviving patients 4.5 years after surgery.[3,5]

Baffle repair or reimplantation techniques did not influence mortality or reoperation in the majority of the studies.[3-5,11,12,E6,E7] The prognosis for infants with pulmonary hypertension is generally poor with or without surgery. The reported one-year survivals are 62.5% and 54% with surgical and medical treatment, respectively, suggesting the advantage of surgical intervention for symptomatic neonates.[11,12,E5]

The optimal management of symptomatic adults currently remains unclear. The proponents for surgical intervention, nonetheless, have demonstrated excellent long-term functional results.[5,13,E6,E7,E22]Dupuis and associates, however, obtained a good result in only one-third of their surgically treated patients. Of 85 medically treated patients, 79 remained asymptomatic.[11,12] Because of the frequent association of right lung malformations, many patients have poor lung perfusion despite having an unrestricted pathway.[19]

At a median follow-up of 7.2 years, in a cohort of 81 patients, Chemin and associates demonstrated persistent reduction of lung volume and air flow reflecting pulmonary hypoplasia.[E44]

Discussion

Although patients with scimitar syndrome may be asymptomatic, presentation in infancy, occurring in three-fifths, usually has shunt-induced congestive heart failure and pulmonary hypertension.[3-5,9,11,12,E2,E20-E22,E44] The diagnosis should be suspected when an infant presents with tachypnoea, repeated chest infections, or other signs of heart failure in association with a right-sided heart and hypoplasia of the right lung. Adults, however, may remain asymptomatic.[3-5,9,11,12,E2,E20-E22]

Causative factors for severe symptoms and pulmonary hypertension are manifold. They include shunting from the scimitar vein or aortopulmonary collateral arteries. Associated congenital heart disease is another problem, as is a hypoplastic pulmonary vascular bed and a volume-overloaded left lung. Primary pulmonary hypertension of the newborn can also be a problem, along with pulmonary venous obstruction, or failure of involution of the pulmonary arterioles due to excessive pulmonary flow.[11,12,e13,e45,e46] Pulmonary histopathology has demonstrated an increased muscularity of pulmonary arteries less than 300 μm in diameter, suggesting failure of regression of mural thickness due to excessive pulmonary blood flow from birth.[e46,e47]

The optimal surgical repair involves redirection or re-implantation of the scimitar vein to the left atrium without using synthetic vascular substitutes, without causing distortion, without attenuation of scimitar venous or pulmonary venous flow, and without kinking the contiguous vascular structures. That this ideal has yet to be achieved is evident from the numerous surgical techniques described, along with follow-up studies that report an undesirable incidence of post-operative venous or baffle obstruction. Reintervention, either surgical or by catheter, varies from 11% to 30%.[19,E22,E60,E62-E64,E71]

Surgical strategies vary according to the configuration and drainage site of the anomalous pulmonary vein. Because of this, it is desirable to obtain accurate anatomical information in the preoperative period. The restricted imaging windows limit the usage of echocardiography in assessment of the exact course and location of the entry point of scimitar vein. Contrast-enhanced and multidetector computed tomography, and/or cardiac magnetic resonance imaging provide comprehensive assessment of the anomalous venous connection and flow characteristics. They are now safely used to replace angiography in the definitive diagnosis.

With the evolving knowledge of the complex variable anatomy, surgeons began to adopt individualized surgical techniques to suit the different pathological variants. Long-term outcomes of intracardiac baffling have been disappointing in infants due to pulmonary venous obstruction or inferior caval venous stenosis.[3-5,19,E2,E3,E12,E21,E71,E72] The anatomical factors contributing to such obstruction include variable scimitar venous anatomy, the acute angle taken by the blood as it traverses the baffle to reach the left atrium, shrinkage and thrombosis of the baffle, and stenosis of the orifice of the scimitar vein.[19,E2,E3]

Although deep hypothermic circulatory arrest facilitates visualization of the infradiaphragmatic opening

in small children, it is associated with an increased incidence of neurologic events.[3-5,E72] In an attempt to improve the results of baffling, several teams have proposed ingenious techniques. Puig Massana and Revuelta, for example, used the free wall of the right atrium.[E13] Calhoun and Mee compartmentalized the inferior caval vein into posterior and anterior compartments.[E11] Spray advocated cutting back the venoatrial junction to remove the angle from the anastomosis.[19,20] Geggel and associates resected the atrial septum, employed a V-shaped incision at the scimitar venoatrial junction, and employed multiple patchs.[E68] Chan and colleagues described atrial septectomy and intra-atrial baffling.[E14] The long-term results of all these innovative procedures is unknown.[E14]

Seeking to avoid the use of baffles, and stenosis of the scimitar vein, other ingenious techniques of division and translocation of scimitar vein with or without cardiopulmonary bypass have been proposed. They include division and translocation of scimitar vein and right atrium adjacent to a pre-existing interatrial communication, or creation of a defect if one is lacking.[13,19,E5,E6] Janssen and colleagues divided the scimitar vein, bringing it through pericardial opening posterior to the phrenic nerve before anastomosing it to the left atrium.[E66] Brown and colleagues divided and translocated the scimitar vein into the left atrium via right thoracotomy without using bypass.[E7] This technique is also recommended in cases of posteriorly placed scimitar vein, for which some investigators have suggested pneumonectomy.[E7,E17]While it eliminated the long, complex, and angulated pathway associated with intracardiac baffling, the reported incidence of pulmonary venous obstruction has been high, likely due to pulmonary venous distortion or anastomotic stricture.[3-5,e4,e12,e22,e71,e72] A recent series from Boston described an incidence of pulmonary venous obstruction of 46% with intracardiac baffling, and 68.8% with reimplantation procedures.[E22]

The use of pericardial rolls as developed at the Cleveland Clinic avoids direct circumferential anastomosis of the delicate pulmonary veins, making it ideal for infants and adults with multiple small, separate pulmonary veins, and multiple anatomic configurations. Early outcomes are encouraging, with complete relief of presenting symptoms and unobstructed pulmonary veins. Longer-term follow-up is needed.[E64]

Repair when the pulmonary veins are distant from the left atrium is challenging, and may eventuate in a higher prevalence of pulmonary or inferior caval venous stenosis, or intracardiac baffle obstruction.[3-5,13,E62-E64,E67,E72]

A myriad of surgical techniques have been described for managing distant anomalous pulmonary veins, including mobilization and direct implantation into the left atrium, use of interposition grafts, and creation of an intra-atrial baffle f through a pre-existing or newly constructed atrial septal defect.[3-5,E22,E71,E72] The risk of postoperative obstruction increases with circular direct anastomoses, acute angulation of veins, the requirement for increased mobilization, and handling or clamping at the time of surgery.[3-5,13,E62-E64,E67,E72]

Although the pericardial tunnel, or "sutureless", technique as described by Sun and colleagues addresses several drawbacks that increased risk of pulmonary venous obstruction of other techniques, it is difficult to apply in cases of marked right-sided positioning of the heart, and hypoplasia of the right lung.[E69] Lugones and associates described a modification of their original rerouting technique using the sutureless concept, along with repositioning of the atrial septum.[E10,E67] **Since the vertical form of scimitar vein is shorter than the horizontal form and have sharper angulations to the inferior caval vein and are located distal to the atrial wall, the tunnel technique minimizes anastomotic tension and is the preferable option.[E10,E69] The tunnel technique has shortcomings, nonetheless, including bleeding due to tissue friability, especially in infants, and injury to the phrenic.[E67]For horizontal variety of scimitar vein, a pericardial tunnel might compromise the morphology of the scimitar vein and cause obstruction. Thus, direct implantation is more reasonable for horizontal scimitar veins.[E67]**

Opinion is divided regarding the approach to aortopulmonary collateral arteries.[E21,E22] While some have demonstrated the beneficial effects of coil embolization on reduction of pulmonary hypertension and congestive heart failure, others have an inconsistent experience.[11,12,E2,E4,E20-E46] The majority recommends coil embolization in symptomatic neonates and infants to delay surgery.[E21,E22] Our analysis from published

series demonstrates that seven-tenths of infants had such aortopulmonary collaterals, and half underwent coil occlusion.[3-5,E60,E72] Surgery was delayed until after infancy in half of those undergoing coil occlusion. Such occlusion may postpone, or even eliminate, the need for surgical repair in the absence of concomitant hemodynamically significant cardiac lesions.[3-5,9,11,12,E2]

Prevention and treatment of postoperative pulmonary venous stenosis remains a formidable surgical challenge. It may progress from proximal obstruction to peripheral pulmonary veins, a phenomenon appropriately termed by Caldarone and colleagues as "relentless". Postoperative pulmonary venous obstruction is more common among patients with pre-existing obstruction, and distantly located scimitar vein. The "sutureless" technique has been used in handling these patients with variable success.[E62-E64,E72] Surgical and percutaneous intervention for pulmonary of inferior caval venous obstruction have been reported with poor results.[E34,E61,E72]

Presently, right pulmonary lobectomy, or more rarely a right pneumectomy, has been limited for severe right pulmonary hypoplasia, recurrent upper respiratory tract infections unresponsive to medical treatment, diffuse bronchiectasis, persistent hemoptysis, or intra-atrial baffles thrombosis after corrective surgery.[3-5,13,E15-E18,E72]

Some investigators, in the setting of a functionally univentricular heart and severely hypoplastic pulmonary arteries, and for those with post-repair persistent pulmonary hypertension, have recommended lung transplantation.[9,E19]

There is an incidence of up to 11% paralysis of the phrenic following surgery.[E6,E67] It is generally higher following direct reimplantation compared to use of an intra-atrial baffle.[4,5,E72] The higher incidence in younger patients has been attributed to dissection of scimitar vein and pericardium near the right phrenic nerve.[3-5,11,12,E6,E67,E72]

Study Limitations

Our analysis was limited because of its retrospective nature, the rarity of the syndrome, the variety of surgical techniques used, and the long time period of the study. Radiological imaging was not routinely performed in all patients at follow-up and the incidence of undiagnosed occlusion/stenosis of the rechanneled pulmonary vein is unknown.

Conclusions

On the basis of our review, we conclude that surgical correction is recommended in all symptomatic patients, and in asymptomatic patients with pulmonary over- circulation or with a clinically treated pulmonary hypertension, stenosis of the scimitar vein, and concomitant cardiac lesions. Surgery in adults, although debatable, is recommended in symptomatic patients with pulmonary over-circulation.

Outcomes of intracardiac baffling have been disappointing; especially in infants. Direct reimplantation of the scimitar vein, although successful in small series with short-term follow-up, remains associated with an undesirable incidence of pulmonary venous obstruction. It may, nonetheless, be a desirable option in horizontal scimitar veins. Other ingenious techniques exist for those with features such as a vertical scimitar vein, severe hypoplasia of the right lung, extreme rightward positioning of the heart, and short scimitar veins. Knowledge of all these approaches should contribute to the armamentarium of cardiac surgeon faced with this rare congenital anomaly.

Author's contribution

Author's name	Concept/ design	Data analysis/ interpretation	Drafting article	Critical revisi
Ujjwal Kumar Chowdhury	?	?	?	?
Robert H. Anderson	?	?	?	?
Lakshmi Kumari Sankhyan	?	?	?	?
Niwin George	?	?	?	?

Author's name	Concept/ design	Data analysis/ interpretation	Drafting article	Critical revisi
Niraj Nirmal Pandey	-	?	?	?
Abhinavsingh Chauhan	-	?	?	?
Yatin Arora	-	?	?	?
Shikha Goja	-	?	?	?

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Figure Legends

Figure 1 : Coronal maximum intensity projection image (A) and volume rendered image, anterior view (B) show an anomalous vein (*) draining the venous return of the right lung into the suprahepatic inferior vena cava (IVC). Volume rendered image, posterior view (C) demonstrates an anomalous systemic artery (arrowheads) from abdominal aorta supplying the lower lobe of right lung.

Figure 2 : Oblique coronal maximum intensity projection image (A) and volume rendered images, anterior view (B) and posterior view (C) shows an anomalous vein (*) draining the venous return of the right lung into the suprahepatic inferior vena cava (IVC). Note is made of tight stenosis (white arrow) of the anomalous vein just before draining into the IVC.

Figure 3 : Schematic drawing of Shumacker and Judds modification of repair of scimitar syndrome. After transecting the scimitar vein with a caval cuff, the proximal end of the scimitar vein was transplanted to the posterolateral wall of the right atrium (RA) adjacent to a pre-existing atrial septal defect (ASD). A short

segment of polytetrafluoroethylene patch was used to baffle flow across the scimitar vein and ASD to left atrium (LA).

Figure 4 : Indiana University (Brown's) modification. Through a right thoracotomy, the scimitar vein was transected with a vena caval cuff. After spatulating, the proximal end of the scimitar vein was translocated into the LA using a side biting clamp without CPB.

Figure 5 : Lam's technique of repair of scimitar syndrome for short scimitar vein. Under CPB, after transecting the scimitar vein, a segment of polytetrafluoroethylene graft was used for anastomosis between the scimitar vein and LA.

Figure 6A-6C : Step-by-step demonstration of division and translocation of the scimitar vein into the LA via pericardial tunnel (Lugones and Garcia's modification). 6A: After resecting the scimitar vein, both ends were over sewn. 6B: A large incision was made in the right lateral wall of the pericardium and the scimitar vein was anastomosed to the pericardium. 6C: Under circulatory arrest, a large vertical left atriotomy was performed. The pericardium was sutured to the left atriotomy and free wall of the RA.

Figure 7A,7B : Calhoun and Mee's modification of repair of scimitar syndrome. 7A: Under circulatory arrest after transecting the inferior cavoatrial junction, a pericardial patch was used to partition the IVC into anterior and posterior compartments in patients with short, lung-enveloped scimitar veins. 7B: After incising the inferior aspect of LA, and interatrial septum, the posterior half of the IVC containing the scimitar vein was sutured to the LA and the anterior lip of IVC was sewn to the RA.

Figure 8A,8B : 8A: An artist's rendition of the anomalous scimitar vein (SV) entering the IVC below the diaphragm above the hepatic veins. 8B: The scimitar vein was directed from the orifice in the IVC to the LA through the ASD. The right atriotomy was extended downwards into the inferior cavoatrial junction for adequate exposure and the requirement of patch augmentation, if needed. Under circulatory arrest, a large synthetic or pericardial patch was sutured from the caudal end of the opening of the scimitar vein all around the margin of the ASD to complete the repair.

Figure 9 : Kowatari's technique of repair of scimitar syndrome using the walls of the right atrium and IVC as a flap, and anastomosing directly to the LA under CPB and cold crystalloid cardioplegic protection.

Figure 10 : Gurerra's modification of repair of scimitar syndrome. Under normothermic CPB and cardioplegic myocardial protection, the scimitar vein was disconnected from the IVC. The IVC was repaired by a continuous suture. The scimitar vein was anastomosed end-to-end to the ringed ploytetrafluoroethylene conduit. The reinforced conduit was tunneled behind the IVC and was anastomosed to the postero inferior LA wall.

[Abbreviations: D: Diaphragm; HV: Hepatic veins; P: Pericardium; PA: Pulmonary artery; PTFE: Polytetrafluoroethylene; SVC: Superior caval vein]

Table E1: Summary of the published investigations documenting the diagnosis of anomalous connection of the right superior caval vein to the morphologically left atrium and its management

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
1.	Najm HK et al, 1996[19]	1975-1995	32 Infant (19), Adult (13)	Age at diagnosis: Median 7 months (range 1 day-70 years) Age at presentation: Infant 25 days (1-335 days), adult >1 year 10.5 years (2.5-70 years)	SVIVCRA junction (n=28) SVRA (juxta IVC) (n=2) SVInfradiaphragmatic IVC (n=2) IVC SV (n=3) LA Associated anomalies (20), ASD (21), sequestration (17), hypoplastic right lung (17), dextrocardia (15), PV stenosis (7), LSVC (4), MV prolapse (4), COA (4), PDA (4), horseshoe lung (3), Azygos continuation of IVC (3), small LV (3), VSD (2), RVMB (1), RAA (1), VACTERL (1), Turner (1) Diagnosis: Cath (29), Echo alone (3), Qp:Qs 0.9-5.9:1	Palliative procedure-infant (12), adult (2) No intervention: infant (7), adult (2) Hypothermic CPB (32), circulatory arrest (8), ACCT 62 min CPBT (25-89 min), CPBT 94 min (42-195 min). Intracardiac baffle pericardial patch (14), PTFE patch (3) Perfusion scan (15) reduced flow right lung 24% (15)	Surgery-mean 8.9 years (1.6-17 years) no deaths Medical mean 5.7 years (0.7-18 years) Post repair PV stenosis (8), reoperation for PV stenosis (2)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
2.	Huddleston CB et al, 1999[E2]	1972-1997	12	Age at presentation: mean 6 weeks (2 days-4 months) Female 4	SV infradiaphragmatic IVC (8) IVC infradiaphramatic (4) SV LA Hypoblastic right lung (12), tracheal stenosis (1), shones complex (1) Clinical: Tachypbnoea CHF (12), NEC (1), HMD (1), cyanosis, high PVR, rightleft shunt (1) PAP systolic: 73.9±21.8 mmHg (40-110mmHg) Qp:Qs 3.1±1.5:1	Hypothermic circulatory arrest (12), intra-atrial baffling (2), division+translocation to LA or RA with baffle (5), pneumonectomy (4) for baffle obstruction, coil embolization MAPCAs (2), bilateral lung transplantation (2), medical treatment (2),	Overall mortality (4) Follow-up median 8.8 years, post pneumonecton primary procedure/baffle occlusion – survivors doing well

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
3.	Brink J et al, 2015[E6]	1974-2012	21	Age at operation: Median 5.4 years (2.5 months-16.7 years) Female (10)	SVIVC ASD (11), VSD (1), ASD+VSD (1), VSD, PDA(1), lung sequestration (1), MAPCA (13), hypoplastic right lung (16)	CPB (19), non-CPB (2), sternotomy (18), thoracotomy (3), baffle repair (15), reimplantation to RA (4), to LA (1), pneumonectomy (1), concomitant ICR (15), ACCT 79±37.5 min (29-164 min), CPBT 115.2±51.5 min (52-263 min), circulatory arrest (11), mean arrest time 28.5 min (19-50 min)	Hospital death (1), late death (3), suprasystemic PAP postoperative-died (2), mean follow-up 13.7±10.3 years (1.3-38.5 years), all survivors NYHA I, Nil medications

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
4.	Gao YA et al, 1993[9]	1964-89	13	Age at catheterization: 1 day-6 months (78±74 days), Female (8)	SVIVC Pulmonary hypoplasia (12), associated cardiac malformations (11), MAPCA (9), hypoplasia left heart (7), bronchial sequestration (2), horseshoe lung (4), CHF (11), pulmonary hypertension (12)	Surgery or transcatheter therapy (10), MAPCAs-surgical ligation (3), coil embolization (4), balloon angioplasty (2) stenosis left sided PV (1), stenosis anomalous right PV (1), occlusion MAPCAs significant improvement-CHF, PAH recurred underwent surgery	

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
5.	Juraszek AL et al, 2005[7]	2005	1	22 years female	ASD-OS closure at age 3 years Decrease exercise tolerance, CXR- mild cardiomegaly, cath- left sided pulmonary veins joined in a confluence behind the LA drained, right sided infradiaphragmatic IVC, RSPV-cranial part of RSPVC	Surgery: side to side anastomosis-left pulmonary venous confluence to LA, ligation and division SV-IVC junction	Follow-up: 6 years MRI-unrestricted anastomosis, doing well
6.	Chan DYL et al, 2015[E14]	2015	1	52 years female	Clinical: Increasing dyspnoea, fatigue, CXR- large vessel – right border of diaphragm CT- SV draining right lung infradiaphramatic IVC, TEE-Intact atrial septum, mild PAH, Qp:Qs: 2.1:1	CPB circulatory arrest. Right pleural cavity open-large SV draining to infradiaphramatic IVC Atrial septectomy, intra-atrial baffling, PTFE patch	Follow-up 4 months, asymptomatic, patent IVC-LA baffle, normal RV size

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
7.	Chemin A et al, 2013[E23]	1976-2013	81	Age at diagnosis: <1 year (67), median 1.5 month (IQR: 0.1-6 months)	Antenatal diagnosis (6), CHF (18), respiratory distress ()19), cardiac murmur (8), incidental finding (7), right lung hypoplasia (81), hypoplastic RPA (57), MAPCA (58), ASD (58), vertebral anomalies (10), PH (36), death (26)	Not mentioned	Follow-up-median duration 7.2 years (2.4-13.4), median age at follow-up 9.8 years (4.5-14.4), high respiratory morbidities with 38% and 43% children, pulmonary infections/ wheezing, one-third-rehospitalized, significant lower TLC-infantile variety

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
8.	Wang H et al, 2018[13]	1994-2015	47	Median age at heart failure 37.7 years-isolated adult	Isolated infantile (1), CHD infantile (2), isolated adult (16), ASD, VSD, UVH, HLHS, Shones complex, mitral atresia, pulmonary sequestration, left anomalous pulmonary vein return, RPA hypoplasia, ALCAPA, TOF, TA, DORV SV obstruction at IVC (19, 43.2%)	Intra-atrial baffle (12), reimplantation (5), right pneumonectomy (1). Overall mortality baffle or reimplantation 37.5% (3 of 8) infants, 0% non-infants	Coil embolization, non-operatively managed (28), medical follow-up 2.74 years (0.46-9.78), 3/6 infantile, 4 of CHD infantile-died At 3.55 years median follow-up living 12 (80%), died 3 (20%).

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
9.	Dusenbery SM et al, 2013[E22]	1964-2013	80	Female (51) Age at diagnosis (year) 0.4 (0-66.8) Age at follow-up (year) 10.8 (0.71-5) <1 year = 46, > 1 year=34	APC (56), S obstruction (17), non-cardiac anomalies (15), cardiac anomalies excluding ASD 30 (38%), VSD, UVH, LPV stenosis, TA, COA, heterotaxy, subaortic stenosis, bilateral PA hypopxia, TOF, PDA, IAA, heterotaxy Symptomatic <1 year 27/46, >1 year 11/34, Qp:Qs (n-80) 1.6 (0.9-2.5) MAPCAs-ipsilateral lung (56), coil occlusion (31), surgical ligation (12), no intervention (13) SV (all right lung) IVC (51) Separate IVC pulmonary RA (15) veins LA SV obstruction-preoperative obstruction (17), <1 year (13), >	Not mentioned	Medical follow-up 6.2 years (0.4-2 years)-61/80 survived SV surgery was performed 33/61 patients, postoperative PV obstruction (18/36) who had SV surgery, 13/28 with baffle procedure, 5/8 with reimplantation Postoperative SV stenosis occurs with similar frequency with both baffle and reimplantation

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
10.	Masrani A et al, 2018[E61]	2001-2016	16	Female (13) Age mean 39.5 years (14 days-72 years) Pediatric age 14 days, 7 months, 9 years, - 3 patients	CT (12), MRI (3), CT+MRI (1) SV Infradiaphragmatic IVC (10) SV supra IVC (6) Right lung hypoplasia (9) CHD (4), dextroposition (8) Pulmonary sequestration (4) RV enlargement-most common radiological findings	Not mentioned	Not mentioned
11.	Tjang YS et al, 2008[14]		1	19 year Female	Systolic murmur, no recurrent pneumonia, ECG, RAD, RVH, CXR, destropsotion Qp: Qs 1.8:1, No ASD, MRA-SVC infradiaphragmatic IVC	Hypothermic circulatory arrest, atrial septectomy, baffle scimitar vein	Follow-up: 5 years, NYHA I Echo – no stenosis of intra-atrial tunnel

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
12.	Shibuya K et al, 1996[E51]	1974-1993	27	1 day-15 years Female (18)	SV total (21), partial (6), dextro/mesocardia 70% SVIVC (12), IVC-RA junction (11), low RA (4)	Surgery (18), interventional cath or both No surgery or interventional cath (9)	Died (4) due to severe associated anomalies
13.	Idris MT et al, 1998[E46]		1	27 year/Female	CXR, TE – dextrocardia, dilated right heart, MVA 1.9 cm^2, no ASD Cath Qp:Qs 2.5:1, PAP 30/15 mmHg, SVIVC	Hypoplastic right lung, SV to infradiaphragmatic IVC	Discharged home, patent surgical anastomosis

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
14.	Vida VL et al 2018[5]		485	year (282) 1-10 (113) >10 year (90) Female (305)	51 institutions, dextrocardia (240), mesocardia (83), CHD (299), ASD (243), right pulmonary hypoplasia (346), severe PPH (46), cardiac symptoms (227), respiratory (243) Qp:Qs 2.1:1 SAS to (R) lung (248), coil embolization SAS (177),	Not mentioned	Time to last follow-up-years 7.2 (2.2-14), STP HD 17 (6.1%) STP LD 11 (39.%) CMP mortality 13 (6.3%) Late death 5 (12%) Symptoms at follow-up 172/451 (37%) Post-operative stenosis / occlusion SV 63/254 (25%)- Reoperation 42 (67%), Untreated 21 (33%)
15.	Pfammatter JP et al 1997[E40]	1997	1	40 weeks gestation	Dexocardia, single S2, Grade 2/6 murmur, Cath at 6 weeks- left sided PVLA, right sided SV IVCm, PAP 67/19, PVR 4 woods unit/m^2, ASD, PDA, MAPCAs	Not mentioned	Coil occlusion: MAPCAs done on follow-up (19 months) doing well

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
16.	Schramel FM et al, 1995[E17]	1967-1992	7	4 years, 29 years, 23 years, 27 years, 24 years, 46 years, 5 years	SVIVC- CP angle (5), Hypoplasia-bronchial tree, tracheal bronchus SV – ICV / LA (2)	Reimplantation of the SV into LA, resection of the sequestered lobe (2), reimplantation of SV (1), right pneumonectomy (1), none (3)	Postoperative thrombosis / fibrosis SV needing pneumonectomy (2), remaining patients uneventful recovery
17.	Salazer J 1995[E18]		5	Female (2) Age mean 3.4 years (6 months-11 years)	SV Subdia IVC (3), peak velocity 0.6-1.0 m/sec, ASD (1) SVsupradia IVC (2), MAPCA right lung (2)	Atrial septal patch in patient with SV+ASD, lobectomy (1) without chest inf recurrent	Not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
18.	Morgan JR 1971[E29]			22 years	Respiratory wheez decrease exercise tolerance, CXR destrocardia, decreased pulmonary vascularity, lung scan, decrease perfusion right lung, cath, PAP 35 mmHg, decreased size RPA, SV draining to LA scimitar sign with normal pulmonary venous drainage	Not mentioned	Not mentioned
19.	Murphy, Kerr and Kirklin 1971[E13]	1960-68	6	8, 15, 20, 32, 57 years Female (3)	scimitar syndrome	CPB, hypothermia, intra-atrial tunnel, tissue tunnel, pericardium, Dacron graft-used	Hospital death (12), survived (5) Follow-up – not mentioned
20.	Mardini MK et al, 1982[6]	1982	3	8 months, 3 years, 2.5 months Male (3)	Recurrent RTI, perfusion scan – less perfusion (R) lung compared to left Qp:Qw 1.5:1	Not mentioned	Not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
21.	Mathey J et al, 1968[8]	1968	3	9 years (F), 7 years (F), 15 years (M)	Recurrent RTI, murmur-ESM-PA cath, CXR-scimitar sign cath confirmed the diagnosis	Sternotomy (1), right thoracotomy (2), SVRA (1), division + translocation SVLA (1), division + translocation SVgraft 20 mm – LA (1)	Postoperative- uneventful
22.	Lloyd IE et al, 2014[E35]	2014	2	7 months (F), 2 weeks (M)	Case 1: SS, SVIVC, additional ASD, cleft, ATL, retroesophageal, hypoplastic RPA, LSVCCS, hypoplastic right lung Case 2: SS, SVIVC, associated TGA, VSD, PDA Skeletal-proximal rib fusion 7^{th}, 8^{th}, subnumerary ribs, 10, 11 Hemivertebrae T8-T9 Micromelia-upper extremities	Necropsy study	Not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
23.	Mas C et al 2000[E47]	2000	1	3 years	Asymptomatic, Loud S2 (A2), CXR-mesocardia, hypoplastic right lung Echo, Angio-abnormal venous flow right side IVC-RA junction Right lungSV which formed an arch draining to IVC/RA SV-IVC junction 5mm, SV-10mm at hilum	Coil embolization: IVC-SV junction	Follow-up at 5 months Complete occlusion lower end SV
24.	McBride ME et al, 2009[10]	2009	1	New born female	SS with HLHS (MS, aortic atresia variant) Cath hypoplastic right lung, SS	Successful norwood operation with Sano modification, returned at age of 5 months successful Glenn	At the time of reporting – surviving

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
25.	Lam TT et al 2006[E9]	2006	2	4 years, 14 years	Cath SS, secundum ASD, Qp:Qs 2.6:1	CPB, bicaval cannulation, cannulation, cardioplegic arrest, SV-IVC junction divided 12 mm, 16mm PTFE graft interposed between the divided SV to LA ASD closed with pericardial patch	Echo at 4 months, 6 months – unobstructed laminar flow from SV to LA
26.	Eckhauser AW et al, 2013[E19]	2013	5	1 day male, 5 year female, 1 day female, 1 day female, 1 day male	Case 1: SS, HLHS, DORV, bilateral SVC, severe PA hypoplasia (Z-6.5), MAPCA (large) – right lung Norwood with a 5mm Sano shunt (SV, APC left unaddressed) At 11 months bilateral SCPC attempted-abandoned because hypoplastic RPC	Case 1: SS, HLHS, DORV, bilateral SVC, severe PA hypoplasia (Z-6.5), MAPCA (large) – right lung Norwood with a 5mm Sano shunt (SV, APC left unaddressed) At 11 months bilateral SCPC attempted-abandoned because hypoplastic RPC	Tracheostomy Home- died age 3 years (respiratory infection)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
					Case 2: SS, HLHS, norwood with a 5mm Sano, ligation APC, SV intact, at 10 years of age- right hemi Fontan, left BDG, stenting, proximal RPA, PVR 3.7/ wu/m^2 Age 18 years, extracardiac TCPC	Case 2: SS, HLHS, norwood with a 5mm Sano, ligation APC, SV intact, at 10 years of age- right hemi Fontan, left BDG, stenting, proximal RPA, PVR 3.7/ wu/m^2 Age 18 years, extracardiac TCPC	Doing well saturation >90%

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
					Case 3: SS, HLHS, severe RPA hypoplasia (3.3 mm, Z-2.2), cath obstruction SV (entire right lung) hybrid procedure, bilateral PA band, ductal stent severe CHF Age 5 months-biatrial heart transplantation, atrial septum left intact with SVRA ECMO-tracheostomy	Case 3: SS, HLHS, severe RPA hypoplasia (3.3 mm, Z-2.2), cath obstruction SV (entire right lung) hybrid procedure, bilateral PA band, ductal stent severe CHF Age 5 months-biatrial heart transplantation, atrial septum left intact with SVRA ECMO-tracheostomy	Follow-up 6 years, doing well

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
					Case 4: SS, HLHS, dextrocardia, bilateral SVC, SV RA, severe RPA hypoplasia, right pulmonary sequestration Hybrid procedure-bilateral PA band, inter-atrial stent Age: 5 months-bicaval cardiac transplantation, reimplantation SVLA	Case 4: SS, HLHS, dextrocardia, bilateral SVC, SV RA, severe RPA hypoplasia, right pulmonary sequestration Hybrid procedure-bilateral PA band, inter-atrial stent Age: 5 months-bicaval cardiac transplantation, reimplantation SVLA	Discharge home with tracheostomy

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
					Case 5: SS, HLHS, right lung hypoplasia, LSVCCS, Norwood with a 3.5mm RMBT, SV-cavo atrial junction, mild RPA hypoplasia, moderate sized APC Age 5 months-unsuitable for 2nd stage palliation with an elevated PVR Age 8 months-stenting proximal BT shunt, balloon angioplasty instent SV	Case 5: SS, HLHS, right lung hypoplasia, LSVCCS, Norwood with a 3.5mm RMBT, SV-cavo atrial junction, mild RPA hypoplasia, moderate sized APC Age 5 months-unsuitable for 2nd stage palliation with an elevated PVR Age 8 months-stenting proximal BT shunt, balloon angioplasty instent SV	Survived

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
27.	Farnsworth AE et al, 1974[E34]	1974	2	1 day	SS, right lung markedly hypoplastic, dextroposed heart, MAPCA pierced the diaphragm, PFO (8mm), VSD (18mm), RV wall hypertrophied	SS, right lung markedly hypoplastic, dextroposed heart, MAPCA pierced the diaphragm, PFO (8mm), VSD (18mm), RV wall hypertrophied	Necropsy study
				45 years	SV subdiaphragmatic IVC, Qp:Qs 2.7:1 Right bronchogram-middle lobe from upper lobe RALT-SV to LA anastomosed through RA under CPB	SV subdiaphragmatic IVC, Qp:Qs 2.7:1 Right bronchogram-middle lobe from upper lobe RALT-SV to LA anastomosed through RA under CPB	Survived at the time of reporting

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
28.	Calhoun RF and Mee RBB, 2003[E11]	2003	2	Not mentioned	Infantile SS with short lung-enveloped scimitar veins, portioning of the inferior cavoatrial junction using a pericardial patch with the SV draining into the posterior compartment which was anastomosed to the LA	Infantile SS with short lung-enveloped scimitar veins, portioning of the inferior cavoatrial junction using a pericardial patch with the SV draining into the posterior compartment which was anastomosed to the LA	1 patient recovered fully from CHF, 2nd patient underwent heart lung transplantatio
29.	Brown JW et al, 2003[E7]	1990-2000	9	Age at presentation: 7 months-43 years (Mean, 11.5±17.6 years), Female (8)	Recurrent pneumonia, SS, SV infradiaphragmatic IVC, extra lobar sequestration (3), Qp:Qs 1.6-3.1:1.0, no ASD	SV to LA anastomosis without CPB via right anterolateral thoracotomy	HD, LD-Nil Follow-up mean 55±46 months (a patent anastomosis without any evidence of restenosis)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
30.	Dupuis C et al, 1992[12]	1960-1990	122	Not mentioned	22 university centers, scimitar syndrome, average follow-up 22 years, pneumonia (38), slight DOE (23), retraction of hemothorax (8), hemoptysis (7), dextrocardia (98), hypoplastic lung (102), ASD (16), PDA (3), SVC to CS (1), azygos continuation IVC (1), cortriatriatum (1), vertebral malformation (1), scoliosis (1), horseshoe kidney (1), leftright shunt <50% (100/122 years), PAP-normal (94), slightly elevated (28)	22 university centers, scimitar syndrome, average follow-up 22 years, pneumonia (38), slight DOE (23), retraction of hemothorax (8), hemoptysis (7), dextrocardia (98), hypoplastic lung (102), ASD (16), PDA (3), SVC to CS (1), azygos continuation IVC (1), cortriatriatum (1), vertebral malformation (1), scoliosis (1), horseshoe kidney (1), leftright shunt <50% (100/122 years), PAP-normal (94), slightly elevated (28)	Follow-up 1 year (20), 1-10 years (52), >10 years (50) Surgery (12), deaths (4), without surgery (79), deaths (0)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
31.	Jimenez M et al, 1988[E31]	1988	1	6 months male	Recurrent RTI, CXR- hypoplastic right lung, dextroposition-heart, eventration diaphragm, sequestration right lung lobe, Doppler echo-SV (7mm diameter) diaphragmatic portion IVC, IVC gradient 8mmHg, Cath-IVC-RA gradient 9mmHg	Medical treatment	Follow up 6 months, asymptomatic

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
32.	Kamiyama M et al[E34]		1	4 months, female	Recurrent attacks bronchospasm, strider, CXR- lung hypoplasia, mediastinal shift to the right, scimitar shadow, echo- absent RPA, dilated MPA, LPA, PDA Cath- absent RPC, SV IVC, MAPCA from abdominal aorta, VSD, PDA, suprasystemic PAP, 8 months age	VSD,ICR, PDA ligated 2 months postop- Severe bronchospasm- ventilated for 3 months Right thoracotomy- congested, unilobulated, poorly ventilated lung (R) pneumonectomy Extubated 3 days later	Follow-up (11 years), asymptomatic

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
33.	Huebesch P et al, 1989[E52]	1989	1	66 years, female	SOB, mastectomy for breast carcinoma-16 years ago, fixed spitting S2, sonography-abdominal-distended IVC, atypical vessel entering below the diaphragm Cath-O2 step up at IVC, CT-SV entering subdiaphragmatic IVC	Refused surgical treatment	Not mentioned
34.	Geggel RL et al, 1993[E28]	1993	1	1 year, female	Asymptomatic, CXR (R) lung hypoplasia, mediastinal shift to right, 2 curvilinear densities in the right chest, oxygen step up at IV, PAP-normal, Qp:Qs 1.7:1 IVC-RA junction SV RA SVC-RA junction	No surgery	Not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
35.	Gikonyo DK et al, 1986[E30]	1986	4	1 female	All symptomatic at birth, CHF, cyanosis, respiratory distress syndrome, SV infradiaphragmatic IVC, MAPCA-abdominal aortic RML (case 1) VSD, tubular, hypoplasia aortic arch, PDA, single umbilical artery, TOF, PDA (right-sided), aberrant LSA	Necropsy finding	Not mentioned
36.	Frank JL et al, 1986[E41]	1986	6	1 day female	SS infradiaphragmatic IVC, SS with horseshoe lung, right lung hypoplasia	Died on operation table	
				9 months female	SS infradiaphragmatic IVC, SS with horseshoe lung, right lung hypoplasia	18 month right pneumonectomy	Discharged home, lost to follow-up

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
				10 months female	SS infradiaphragmatic IVC, SS with horseshoe lung, right lung hypoplasia	Right pneumonectomy advised – Refused	Lost to follow-up
				4 months female	SS infradiaphragmatic IVC, SS with horseshoe lung, right lung hypoplasia	Medical follow-up	Not mentioned
				1 day female	SS infradiaphragmatic IVC, SS with horseshoe lung, right lung hypoplasia	Right pneumonectomy	Stormy postoperative-discharged home on 21 month
				1 month female	SS infradiaphragmatic IVC, SS with horseshoe lung, right lung hypoplasia	Medical follow-up	Not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
37.	Co-Vu JG 2010[E61]	2010	1	3 days	Non-dysmorphic, CXR cardiomegaly, TTE/MRI-HLHS with SS, hypoplastic (right) lung, 50% of normal size, small RPA, single APC, right lower lung	Initial Norwood with sanomodification, vascular plug to MAPCA, 6.5 months - cardiac transplantation, ECMO support	Not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
38.	Dupuis C et al, 1993[11]	1970-1987	25	15 female	12 European centers, age at hospital admission because of severe symptoms, M1 wk (8), 1-8 weeks (12), 3,4,6 months (3), 8 months (2), CHF (14), RDS (8), dextroversion, severely hypoplastic right lung (19), SV image (6), vertebral malformation (3), cath – systolic PAP increased in all patients 60-120 mmHg, PFO (19), ASD (6), hypoplastic RPA (21), right lung total drainage (20), partial drainage (5), MAPCA to right lung (23)	Medical treatment alone (10), death (7), survivors (3), good result (2) Surgical treatment (15), death (9), survivors (6), good result (5), respiratory problem (1)	As mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
39.	Vida VL et al, 2013[4]	2013	44	25 female, median 3 months	9 Italian centers **Isolated infantile CHD infantile 33 (75%) 11 (25%)** Age at diagnosis (m) 1.7 (0-41) 0.3 (0.1-30) Asymptomatic 18 4 CHF 2 4 Respiratory symptoms 15 5 Qp:Qs 1.45 (1-3) 1.95 (1-2.8) mPAP (mmHg) 20 (13-34) 34 (18-50) Coil-MAPCA 11 4 Pulmonary hypoplasia 17 8 Surgery for other CHD 0 8 Deaths 0 2 Respiratory symptoms 2 1	9 Italian centers **Isolated infantile CHD infantile 33 (75%) 11 (25%)** Age at diagnosis (m) 1.7 (0-41) 0.3 (0.1-30) Asymptomatic 18 4 CHF 2 4 Respiratory symptoms 15 5 Qp:Qs 1.45 (1-3) 1.95 (1-2.8) mPAP (mmHg) 20 (13-34) 34 (18-50) Coil-MAPCA 11 4 Pulmonary hypoplasia 17 8 Surgery for other CHD 0 8 Deaths 0 2 Respiratory symptoms 2 1	

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
40.	Najm HK et al, 2018[E62-64]	2018-2019	6	3 months-65 years, median 40 years	Scimitar syndrome, PAPVR-RUPV, RMVSVC, no ASD (3) Heterotaxy unbalanced AVSD, mixed obstructed TAPVR (2)	In situ pericardial roll directing SV to SV with repair of concomitant cardiac anomalies	No mortality Median hospital stay 23 days (4-60 days), median follow-up 20 months (1-36 months), widely patent PVLA anastomosis, all asymptomatic NYHA-I
41.	Kowatari R et al, 2016[E65]	2016	1	15 years	Recurrent RTI, CXR-scimitar sign, CT-SV infradiaphragmatic IVC, PAP 28/9 (15 mmHg) cath Qp:Qs 3.2, PVr 0.7u/m^2	CPB, cardioplegia, , SVC with RA, IVC flap anastomosed directly to right-sided LA wall	Postoperative Echo, CT-no stenosis / kinking scimitar vein

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
42.	Jensen H et al, 2009[E66]	2009-2012	6	17 years, 2 years, 6 months, 11 years, 1 year, 7 months	SS, MAPCA (2), previous failed baffle (1), moderate PAH (2), ASD (4), PFO (1), bilateral SVC (2)	CPB, cardioplegia, SV transected at the junction of IVC. Caval end-sutured, SV-tunneled through a large pericardial opening posterior to the phrenic nerve-anastomosed to the right-side of posterior LA with access via an existing or surgically created ASD. CPB median 88 minutes, ACCT median 38 minutes	No deaths, follow-up median 28 months (8-41 months), no reoperation, no PVO

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
43.	Lugones I et al, 2017[E67]	2017	1	2 years male	SS, CXR-destrocardia, cardiomegaly, increased PBF, hypoplasia right lung, Echo-RV, RA dilatation, no ASD, SV draining whole right lung infradiaphragmatic, IVC-stenotic opening, Cath Qp:Qs 1.8:1, MAPCAs-occluded	CPB, cardioplegia, in situ pericardial rerouting in which the SV was detached from its insertion line and the atrial septum was repositioned to create a large opening into the LA. SV drained to a pericardial bag connected to the LA	Surgived TEE, Cath- unobstructed anastomosis, discharged on 4th postoperative day

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
43.	Geggel RL et al, 2020[E68]	2011-2018	11	-	22-SV surgery- baffling/reimplantation (11), new multipatch technique (11).	A double or triple patch technique depending on the distance between the SV and atrial tissue. Autologous pericardium harvested. CPB, cardioplegia. Resection of the atrial septum with removal of the muscular limbus. LA-pulled down towards the SV- V shcpaed incision at the SV-atrial junction-space filled with pulmonary homograft- a pericardial patch to septate the atrium-additional patch anteriorly to augment the vena cava	5/11-conventional baffle/reimplantat pulmonary vein obstruction 45 days-9.5 months postoperative Follow-up 3.6 years, no obstruction with new operation

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
44.	Sun Y et al, 2018[E69]	2011-2017	9	Male 4 Age: 3.1±3.6 mean (range 0.3-12 years), Infantile form (4), childhood adult (4)	Vertical form of SV-IVC, CT (9), Echo (9_), Cath (5); PDA (4), ASD (9), VSD (1), MAPCA (3), destroposition (9), right lung hypoplasia (7)	SV-transected from IVC-attached to the right lateral pericardium-left atriotomy. The pericardial flap is turned to the left with its anterior margin sutured on the free wall of the RA anterior to the IA groove to create a tunnel.	No hospital death, no residual SV obstruction, 1 lost to follow-up, 8 survivors follow-up 33.4±19.2 (2-72) months, no PH, No SV obstruction

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
45.	Vida L et al 2020[E72]	-	485 (51 European Centres)	SS patients, overall (485), neonates/infants (282), children >1-10 years (113), adolescents/adults >10 years (90).	SS, dextrocardia 240, mesocardia (83), levocardia (163), isolated (186), associated CHD (299), right pulmonary hypoplasia (346), PA hypoplasia-severe (46), preoperative cardiac symptoms (277), respiratory symptoms (24), pH (157), embolization MAPCAs (177)	STP (279), CMP (206), follow-up 72 years (range 2.2-14 years)	STP, hospital death 29 (10%), late mortality 11 (3.9%), symptoms at last follow-up 172 (37%).

[Abbreviations: ACCT: Aortic cross clamp time; ALCAPA: Anomalous origin of left coronary artery from the pulmonary artery; ASD: Atrial septal defect; ATL: Anterior tricuspid leaflet; CHF: Congestive heart failure; COA: Coarctation of aorta; CPB: Cardiopulmonary bypass; CPBT: Cardiopulmonary bypass time; CXR: Chest x-ray; DORV: Double outlet right ventricle; ECMO: Extracorporeal membrane oxygenation; ESM: Ejection systolic murmur; HLHS: Hypoplastic left heart syndrome; HMD: Hyaline membrane disease; ICR: Intracardiac repair; IQR: Interquartile range; IVC: Inferior caval vein; LA: Left atrium; LV: Left ventricle; MAPCAs: Major aortopulmonary collateral arteries; MRA: Magnetic resonance angiography; MRI: Magnetic resonance imaging; MV: Mitral valve; NYHA: New Yori Heart Association; OS: Ostium secundum; PAH: Pulmonary artery hypertension; PAP: Pulmonary artery pressure; PDA: Persistent arterial duct; PTFE: Polytetrafluoroethylene; PV: Pulmonary vein; PVR: Pulmonary vascular; resistance PVR: Pulmonary vascular resistance; Qp:Qs: Pulmonary blood flow / systemic bloow flow; RA: Right atrium; RAA: Right aortic arch; RALT: Right anterolateral thoracotomy; RPA: Right pulmonary artery; RSPV: Right superior pulmonary vein; RTI: Respiratory tract infection; RVMB: Right ventricular muscle bundle;

SCPC: Superior cavopulmonary connection; SS: scimitar syndrome; STP: Surgically treated patients; SV: scimitar vein; TA: Tricuspid atresia; TCPC: Totally cavopulmonary connection; TEE: Transesophageal echocardiography; TGA: Transposition of the great arteries; TOF: Tetralogy of fallot; UVH: Univentricular heart; VACTERL: Vertebral anal cardiac tracheal esophageal rectal laryngeal; VSD: Ventricular septal defect]

Table E2: Summary of the published investigations documenting the surgical anatomy, associated cardiac lesions, indications for different types of operation for patients with scimitar syndrome

S. No.	Operations performed	Indications, surgical anatomy, associated anomalies, surgical details	References
1.	Division and reimplantation of the scimitar vein into the right atrium with CPB	Short length scimitar vein Not long enough for direct anastomosis to left atrium SVRA anastomosis plus Bailey;s atrioseptopexy	18,E3
2.	Division and translocation of the scimilar vein into the right atrium	scimitar vein division and translocation to posterolateral wall – RA adjacent to pre-existing ASD or newly created ASD Short baffle Associated stenosis- SV if present, spatulated and caval cuff IVC- patch to avoid stenosis	13,19,E5,E6
3.	Division and translocation of the scimitar vein into left atrium	Right anterolateral thoracotomy, no CPB Direct anastomosis to LA after division and translocation scimitar vein divided with caval cuff Not associated ASD Both infants and adults	E2,E6,E8
4.	Division and translocation of the scimitar vein into LA using PTFE graft interposition	Short length of the scimitar vein Median sternotomy under CPB	39
5.	Division and translocation into left atrium using Dacron graft interposition	Right thoracotomy No ASD 20 mm Dacron graft- short length scimitar vein	7,19

S. No.	Operations performed	Indications, surgical anatomy, associated anomalies, surgical details	References
6.	Division and translocation of the scimitar vein into LA via pericardial tunnel	Short length scimitar vein Division of the scimitar vein at SV-IVC confluence with caval cuff CPB-SV to pericardial anastomosis Circulatory arrest - Left atriotomy-pericardial margins sutured to the left atriotomy and freewall of the RA	E10
7.	Partitioning of the inferior caval vein and rechanneling of the scimitar vein into the LA	No associated ASD DHCA at 18°C Inferior cavoatrial junction – transected Pericardial patch partitioning into anterior and posterior compartments Posterior receiving SV blood, anterior compartment – caval blood Infantile variety	E11
8.	Intra-atrial baffle	Useful in adults Long, intra-atrial baffle of PTFE or right atrial wall or pericardium From the orifice of the scimitar vein, across the atrial septal defect Under circulatory arrest	E12
9.	Atrial septectomy and intra-atrial baffling	scimitar syndrome with intact interatrial septum	E14
10.	Scimitar vein cut back technique	SV to RA cut back along with venoatrial junction Angle away from the anastomosis Decreasing the risk of local stenosis	
11	In situ pericardial roll technique for distant anomalous pulmonary venous return	Using both anterior and posterior configuration for anomalous SV that are distant from the LA either because of remote insertion, because of complex cardiac anatomy	E64

S. No.	Operations performed	Indications, surgical anatomy, associated anomalies, surgical details	References
12	Division and translocation of the SV into LA with SV flap	Median sternotomy, CPB, Cardioplegic arrest SV was harvested with the surrounding IVC and RA wall as SV flap The SV flap was anastomosed directly to the LA	E65
13	Modified in situ pericardial rerouting and repositioning of the atrial septum	Useful in case of SS with dextrocardia or mal rotation of the heart with or without pulmonary venous obstruction	E67
14	Directly implantation of the SV to the LA via sternotomy	Under cardioplegic arrest, SV was transected and brought through a large pericardial opening posterior to the phrenic narve Anastomosed to LA with access via ASD	E66
15	Repair by a novel multipatch technique	Under cardioplegic arrest, resection of the atrial septum, a double or triple patch technique depending on the distance between the SV and atrial tissue Avoids circuitous pathways or tension on the scimitar pulmonary vein	E68
16	Pericardial tunnel technique	Under cardioplegic arrest, SV was divided at ifs insertion site on the IVE The proximal and was oversewn The distal end was filleted open longitudianally on ifs medial aspect The SV was attached to the right lateral pericardium using 7-O polydioxanone suture Blood flow was directed to LA through a left atritomy inside the pericardial tunnel	E69

S. No.	Operations performed	Indications, surgical anatomy, associated anomalies, surgical details	References
17	Divison and translocation of the SV into LA using a ringed PTFE conduit	Under CPB, cardioplegic arrest Distal and of SV was transected and mobilized A ringed PTFE interposition graph was sewn ent-to-end the divided SV and the other end was anastomosed end to side to the LA	E70
18.	Pneumonectomy	scimitar vein coursing posterior to the hilum Post repair- stenosed, fibrosed scimitar vein Recurrent stenosis of the pulmonary vein following dilatation / stenting	E15,E17
19.	Lobectomy / lung resection	Persistent right-sided pneumonia on a grossly hypoplastic lung Sequestration of the pulmonary parenchyma	E17,E18
20.	Lung transplantation	Persistent postoperative pulmonary hypertension Recurrent pulmonary vein stenosis following surgery for scimitar vein	9, E2
21.	Orthotopic cardiac transplantation	scimitar syndrome with a functionally univentricular heart with severely hypoplastic pulmonary artery	13,E19
22	Coil embolization / surgical ligation / balloon angioplasty	Sizeable MAPCAs Infants via axillary artery, juvenile / adults-femoral artery Mini balloon Balloon expandable stainless steel Palmaz stent – for stenotic pulmonary vein or stenosed baffle	9,13,19,E2,E20-E24

[Abbreviations: ASD: Atrial septal defect; CPB: Cardiopulmonary bypass; DHCA: Deep hypothermic circulatory arrest; IVC: Inferior caval vein; LA: Left atrium; MAPCAs: Major aortopulmonary collateral arteries; PTFE: Polytetrafluoroethylene; RA: Right atrium; SV: scimitar vein]

Table E3: Early and late postoperative outcomes of the patients undergoing different types of operation for patients with scimitar syndrome in the published investigations

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
Short-term outcomes	**Short-term outcomes**	**Short-term outcomes**	**Short-term outcomes**	**Short-term outcomes**	**Short-term outcomes**	**Short-term outcomes**	**Short-term outcomes**	
Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	Overall operative mortality: 4.8%-5.9% Causes of death: Infantile presentation with associated complex CHDs, severe pulmonary hypertension, major associated extracardiac anomalies, severe pulmonary hypoplasia with sequestration, hypoplastic left heart syndrome	
Long-term outcomes	**Long-term outcomes**	**Long-term outcomes**	**Long-term outcomes**	**Long-term outcomes**	**Long-term outcomes**	**Long-term outcomes**	**Long-term outcomes**	

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
Schramel et al, 1995	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	N=7, no surgery (3), reimplantation of SV into LA, resection of the sequestered lobe (2), postoperative fibrosed, stenosed SV needing pneumonectomy (2)	E17

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
Dusenbery et al, 2013	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	N=80 (<1 year=56), MAPCA ipsilateral lung (56), coil embolization (31), surgical ligation (12), no intervention (13), SV surgery: 13/28 with baffle procedure, 5/8 with reimplantation procedure had postoperative SV stenosis	E22

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
Eckhauser et al, 2013	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	N=5, SS with HLHS between 3-6 years, 4 patients were alive: 2 after cardiac trans-planta-tion, 1 under-went a Nor-wood opera-tion, 1 survived at staged pallia-tion culmi-nating in a Fontan operation	E10

Variables	**Results**	**Results**	**Results**	**Results**	**Results**	**Results**	**Results**	**Refere**
Brink J et al, 2015	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	20 surgical patients of SS were followed for 1.3-38.5 years (mean 13.7±10.3 years) with 5 patients developing postoperative pulmonary vein stenosis, 3 (14.3%) patients were reoperated	E6

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
Wang H et al, 2018	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	Out of 47 patients, 28 non-operative medical management, 18 (38.3%) patients underwent surgery: Intra-atrial baffle (12), reimplantation (5), pneumonectomy (1), median follow-up 3.55 years, overall mortality baffle/reimplantation 37.5%, medical management (46.7%), Median follow-up 2.74 years (IQR: 0.46-9.68), 11/16 (68.8%) had post repair stenosis of SV.	13

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
Vida VL et al, 2018	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	Out of 485 SS patients, 279 (57%) were surgically treated and 206 (43%) were clinically monitored. Median age at follow-up was 11.6 years (IQR 4-22 years), among surgically treated patients (279), HD hospital mortality was 6% (n=17), late mortality was 4% (n=11) and 90% (n=243) survived. Among CM patients (n=206) late mortality 6.5% (n=13), 93% (n=184) survived. Sixty-three (25%) surgical patients had stenosis/occlusion of the scimitar veins. 42 (67%) underwent reoperation/reintervention 0.8 years after repair	5

Variables	Results	Results	Results	Results	Results	Results	Results	Refere
	No. of patients	Overall mortality	Medical treatment	MAPCA occlusion	PDA/COA	Complete repair	Pulmonary resection	
Huddleston CBC et al, 1999	12	4	2 (2)	1 (1)	-	7 (1)	5 (0)	E2
Najm HK et al, 1996	32	5	7	5 (3)	2 (2)	17 (0)	1 (0)	19
Dupuis C et al, 1993	25	16	10 (7)	6 (1)	5 (5)	0	3 (2)	11
Torres AR et al, 1993	14	1	0	3 (1)	0	10 (0)	1 (0)	E5
Brown JW et al, 2003	10	0	0	3 (0)	0	10 (0)	0	E7

[Abbreviations: CM: Clinically monitored; COA: Coarctation of aorta; CHD: Chronic heart disease; HD: Hospital death; HLHS: Hypoplastic left heart syndrome; IQR: Interquartile range; IVCLA: Left atrium; MAPCAs: Major aortopulmonary collateral arteries; PDA: Persistent arterial duct; SS: scimitar syndrome; SV: scimitar vein]