

## Scimitar Syndrome: Pathology, Clinical Presentation, Radiographic Features, and Treatment

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### Abstract

Scimitar syndrome is characterized by partial or total anomalous pulmonary venous return from the right lung along with pulmonary hypoplasia. We searched the mail databases such as Medline (via PubMed), Scopus and EMBASE and Google Scholar. Diagnosing infantile scimitar syndrome requires meticulous attention and high suspicion of the early referral and management. The association of the syndrome with pulmonary hypertension leads to recurrent and prolonged hospitalization. Scimitar syndrome can be initially suspected from a chest X-ray, but it is typically confirmed via Computed Tomography (CT) angiography.

The clinical spectrum of Scimitar syndrome ranges from severely ill infants to asymptomatic adults, which may present respiratory or cardiac failure, hemoptysis and pulmonary hypertension, tachypnea, chest infection, and failure to thrive. The diagnosis can be made by transthoracic or transesophageal echocardiography, angiography, or by CT or MR angiography. Considering the wide clinical spectrum of scimitar syndrome, the medical intervention depends on the severity of presentation and the amount of blood flowing to the Inferior Vena Cava (IVC) from completely or partially anomalous pulmonary veins. In the presence of significant left to right shunting and pulmonary hypertension, surgical intervention should be considered.

**Key Words:** Congenital Anomaly, Pulmonary Hypertension, Scimitar Syndrome.

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## 1- INTRODUCTION

Scimitar syndrome was first reported by Naill et al. in 1960, as the radiographic appearance of the anomalous vein, which manifests as a tubular opacity parallel to the right heart border, which simulates a curved Turkish sword or a Scimitar, and hence the term Scimitar sign (1). It is a kind of partially anomalous pulmonary venous return and one of the several symptoms of congenital pulmonary venolobar syndrome. Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies consisting of a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, right lung hypoplasia, heart dextroposition, and anomalous systemic arterial supply to the right lung (2, 3).

Scimitar syndrome occurs exclusively on the right side and it is essentially a mixture of pulmonary hypoplasia and partial anomalous pulmonary venous return (PAPVR). Hemodynamically, there is an acyanotic left to right shunt. The anomalous vein usually drains into either inferior vena cava (most common), right atrium or portal vein. Draining a part or the entire of right lung into the inferior vena cava (IVC) occurs with an anomalous right pulmonary vein, which is the hallmark of scimitar syndrome (4, 5). There are two different types of scimitar veins, first simple classic vein that runs from the middle of the right lung to the cardiophrenic angle and the second type is double arched vein in the upper and lower lung zones, with drainage into the left atrium and inferior vein cava (4, 5).

## 2- MATERIALS AND METHODS

We searched the main databases including Medline (via PubMed), Scopus, Embase, and Google Scholar to November 2019. Also, the references list of selected records was reviewed. Following keywords were used independently and in combination: Scimitar Syndrome AND

(Pathology OR Clinical Presentation OR Radiographic OR Features OR Treatment). Database search was done for possible studies, abstracts of the studies were screened for identification of eligible studies, full text articles were obtained and assessed and a final list of included studies was made. This process was done independently and in duplication by two reviewers and any disagreement was resolved by the 3<sup>rd</sup> reviewer. The kappa statistic between two authors was 78% for selecting the articles.

## 3- RESULTS

### 3-1. Epidemiology

This anomaly has an occurrence rate of about 1 to 3 per 100,000 live births. Since many patients do not show any symptoms, true occurrence may be higher. Generally, the symptoms of this syndrome appear 7 months after birth, but they can occur in different ages (6). Many patients do not demonstrate any symptoms during childhood except recurrent respiratory tract infection. Despite the lack of a definite genetic cause for this syndrome, it is described at five different families, which are noted in this article (2, 3, 7).

### 3-2. Associated congenital cardiac anomalies

It has been reported that 19–31% of patients with scimitar syndrome have associated congenital cardiac anomalies, and atrial septal defect, with a probability of 70%, is the most common (4). Less frequent associations include tetralogy of fallot, ventricular septal defects, patent ductus arteriosus, coratriatum, bicuspid aortic valve and sub-aortic stenosis. Coronary artery abnormalities are relatively rare associations. This paper reports some classic and special forms of scimitar syndrome (7, 8). The infantile scimitar reported in our population is similar to the previous studies and includes the classic scimitar, the bilateral scimitar,

scimitar syndrome with scimitar vein stenosis and contralateral left upper pulmonary vein stenosis, and scimitar syndrome with right and left pulmonary arterial stenosis at multiple levels (8, 9): Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA), accessory diaphragm, eventration or partial absence of the diaphragm, phrenic cyst, horseshoe lung, esophageal and gastric lung, congenital cardiac malformations (25% of cases) including Atrial Septal Defect (ASD), ventricular septal defect and coarctation of the aorta (9). Ruggieri et al. and Kahraman et al. reported the association of dysmorphic features, craniofacial and central nervous system anomalies and renal agenesis with scimitar syndrome, besides associations with Turner syndrome and VATER association (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities) reported in other studies. The fact that half of the patients were the result of consanguineous marriage and some malformation did not fit with any syndrome may indicate a complex genetic cause (10, 11). A rare pulmonary malformation associated with 8% of reported cases of scimitar syndrome is horseshoe lung. An isthmus of the pulmonary parenchymal tissue that arises from the right lung base is the typical feature of horseshoe lung. Pulmonary arterial and bronchial supply extends from the right lower lobe pulmonary artery and bronchus (12, 13). The combination of scimitar syndrome and aortic coarctation is rare and the presence of a left-to-right shunt may lead to a development of pulmonary hypertension and Eisenmenger physiology (14, 15).

### **3-3. Pulmonary hypertension in Scimitar syndrome**

Pulmonary hypertension, as one the best indicators of Scimitar syndrome, is often identified by severe symptoms and it

shows poor prognosis during infancy. There are many factors contributing to pulmonary hypertension including large left to right shunt via the anomalous pulmonary vein, right lung hypoplasia with reduced pulmonary vascular bed, pulmonary vein stenosis and obstruction, and persistent pulmonary hypertension of the newborn (4, 16). Studies have suggested multiple factors for pulmonary hypertension including large left to right shunting by the anomalous pulmonary vein or other intra-cardiac lesions such as ASD or Ventricular Septal Defect (VSD). The main source of pulmonary hypertension in some patients with Scimitar syndrome and in infants with total anomalous pulmonary venous connection has been identified as pulmonary venous stenosis (4, 5). Pulmonary arterial hypertension with infantile-type scimitar syndrome has been recognized as a cause of severe symptoms and poor outcomes. It is attributed to the following factors: 1) stenosis of the anomalous pulmonary veins, 2) the presence of systemic arterial supply in the right lung, 3) reduction of the pulmonary vascular bed on the right side, and 4) increased pulmonary blood flow from anomalous drainage or the intra-cardiac lesion (17, 18). There is an increased risk of heart failure when Scimitar syndrome is accompanied with other cardiac defect and pulmonary hypertension. The etiology of pulmonary hypertension in infants with scimitar syndrome is variable and may be related to a decreased right-sided pulmonary vascular bed, scimitar vein stenosis, and unappreciated systemic collateral blood flow along with intra-cardiac shunts (19).

### **3-4. Clinical manifestation**

Classic findings on physical examination include a shift in heart sounds and cardiac impulse to the right and a systolic murmur. Auscultation of the lung is usually normal, although breathing sounds may be reduced on the right (20, 21). Patients are

diagnosed either early with severe symptoms (infantile type) including tachypnea, chest infection, heart failure and failure to thrive or late with minimal symptoms (childhood/adult type) as a result of accidental findings (22-24). The patients may have different presentations of this syndrome, ranging from asymptomatic infants to those with heart failure and severe pulmonary hypertension. Those who present early in life usually have associated congenital heart disease. The triad of respiratory distress, right lung hypoplasia, and dextroposition of the heart should warn the clinician about the possibility of this syndrome (3, 24). The presentation of the syndrome could be as early as immediately after birth, and the diagnosis should be suspected when the signs of respiratory distress and/or heart failure are present in association with radiological findings of cardiac dextroposition and haziness or suspicion of atelectasis of the right lung (25). In the scimitar syndrome that presents in infants, large blood flow into the IVC from the anomalous pulmonary veins and the presence of hydrocephalus are negative prognostic factors. Clinical symptomatology of the scimitar syndrome is based on the age at which the syndrome presents. Infants with scimitar syndrome present cyanosis, poor growth, Pulmonary Arterial Hypertension (PAH), and often complex cardiac defects, many of which need surgical intervention with a high mortality rate (24). In older children and adults, it commonly presents with recurrent pulmonary infections and/or exertional dyspnea. Hemoptysis as a presenting symptom is exceptionally rare in patients with scimitar syndrome (3). Interestingly, according to the literature, the youngest patient presenting with hemoptysis was seven years old. The possible mechanisms of hemoptysis in scimitar syndrome include rupture of hypertrophied systemic pulmonary anastomosis (3, 9). PAH is a problem of

infancy and it rarely presents in a patient aged 14 years. It has been shown that pulmonary arterial pressure greater than 50 mm Hg is exceedingly rare in patients who present beyond the infancy. Further, it has been demonstrated that PAH in this population may be associated with cardiac anomalies. This could be explained by larger degree of anomalous pulmonary venous drainage, greater extent of hypoplasia of right lung and pulmonary artery, and increased systemic blood flow to the hypoplastic lung (3, 24, 26). In the neonatal period with respiratory and/or cardiac failure, which is most commonly caused by pulmonary hypertension due to cardiac and/or right lung abnormalities. It is treated with surgery and result is dependent on the nature and severity of the abnormalities. Heart failure may also be caused due to a large arterial supply from the abdominal aorta to a sequestered lobe (15, 26). At any stage of life presenting with hemoptysis and pulmonary hypertension due to recurrent respiratory infections. At any stage in life as a result of accidental finding, e.g. due to the detection of murmur or evident Chest X-ray (CXR) abnormalities (16).

### **3-5. Radiographic features**

The first-line imaging investigation in infants is chest radiography and echocardiography. The diagnosis can be made by transthoracic or transesophageal echocardiography, angiography, or by Computed Tomography (CT), Angiography (CTA), and Magnetic Resonance Angiography (MRA) CT or MR angiography. Chest radiographic findings include a small lung with ipsilateral mediastinal shift, and in one-third of cases, the anomalous draining vein, which is seen as a tubular structure paralleling the right heart border in the shape of a Turkish sword ("scimitar") (27). Important differential diagnosis of small lung and small hilar shadow on conventional chest radiograph includes

congenital hypoplastic lung, congenital absence of pulmonary artery, partial atelectasis of the lung, and Macleod's syndrome. The congenital hypoplastic lung and absent/hypo plastic pulmonary artery disease can be excluded from scimitar syndromes based on the normal venous drainage to the left atrium (25). The unusual venous return is the main feature of Scimitar syndrome, which appears as a characteristic, unusual radiographic shadow descending along the right cardiac border. Because of the associated dextrocardia, this radiologic sign may seem obscured. The most striking radiographic feature in each of these patients is the dextroposition of the heart along with varying degrees of opacity in the right hemithorax. A small, opaque hemithorax generally indicates volume loss (28). Mediastinal shift toward the opacified hemithorax with compensatory hyperinflation of the abnormal lung can emerge with atelectasis or pulmonary agenesis. Pulmonary hypoplasia can also result in a unilateral small lung with the heart shifted toward the affected side. The scimitar sign is usually absent in infants (29). General imaging and differential considerations include pulmonary sequestration, right middle lobe atelectasis (on AP plain radiograph) and unilateral absence of pulmonary artery (UAPA) (30).

When anomalous pulmonary venous drainage is suspected, the diagnosis can be made easily by transthoracic echocardiography delineating the abnormal anatomy with the use of the "crab view" coupled with subcostal imaging to reveal the anomalous pulmonary venous flow into the inferior vena cava below or just above the diaphragm (31). Echocardiography could be used to determine whether common cardiac defects such as ASD and PDA are present and whether there are significant pulmonary hypertension and drainage of the pulmonary veins (32). Bronchoscopy

can be used in cases of scimitar syndrome to distinguish the presence of airway hypoplasia (17). Doppler examinations may reveal the union of the scimitar and systemic veins. Fetal echocardiography allows prenatal diagnosis in which spectral and color Doppler provides clues to the presence of an obstructed pulmonary venous pathway (33, 34). The flow velocity pattern in the Scimitar vein is different from the normal pulmonary venous flow (35). The former is biphasic or triphasic with one or two peaks in systole and one peak in diastole (peak velocity of about 0.5 m/s), and reverse flow at atrial contraction (peak velocity of about 0.2 m/s). The flow pattern in Scimitar vein is monophasic extending throughout the cardiac cycle with no reverse flow at atrial contraction (36). This syndrome is chiefly diagnosed by the presence of characteristic radiological sign (scimitar sign) on conventional chest radiography. However, when scimitar vein is masked by cardiac shadow, diagnosis can be made using one or more traditional modalities such as angiography, CT scan and echocardiography. The MR technology also shows excellent visualization of vascular anatomy of this complex congenital defect noninvasively (22, 36). CT and MRI allow direct visualization of the anomalous vein and angiographic techniques and multi-planar reconstructions allow radiologists to determine arterial and bronchial anatomy in detail (37). Cine MRI and Three Dimensional (3D) enhanced MR angiography provide a non-invasive diagnostic technique for the assessment of anomalous pulmonary venous return. Gadolinium enhanced 3D MR angiography (38, 39) that provides concurrent non-invasive complete anatomical (arterial and venous supply) and functional (calculation of left to right shunt using phase contrast MRI) diagnosis eliminates the need for more traditional invasive techniques (40). Cardiac catheterization and angiography

are probably the most useful procedures for confirming the diagnosis and clarifying the exact anatomy and degree of pulmonary hypertension. An aortogram should also be performed to visualize the presence or absence of an anomalous systemic artery entering the right lower lobe (41). Three-dimensional CT and cardiac-gated MRI (83) are useful for visualizing the anomalous pulmonary vein. They can be particularly helpful in finding an associated horseshoe lung, in which there is a posterior fusion of portions of the right and left lungs behind the heart and before the esophagus and spine. Approximately 80% of infants with horseshoe lung also have scimitar syndrome (42). The features of combined scimitar syndrome-ALCAPA on both conventional angiography and cardiovascular CT are surprisingly consistent. When these features are present and specifically sought, it is possible to identify an ALCAPA in scimitar syndrome patients with precision. Ischemia should increase suspicion about this dual diagnosis (43). It should be noted, however, that the absence of ischemic signs and symptoms does not necessarily exclude the diagnosis as scimitar syndrome is often associated with pulmonary hypertension, and the raised pulmonary pressure may provide sufficient perfusion of the circumflex artery to hamper the development of ischemia. CT is increasingly performed as non-electrocardiogram, high-pitch spiral or volumetric acquisitions. It is important that to examine any patient presenting with scimitar syndrome for the signs of ALCAPA and other associations, so cardiovascular CT is the most appropriate first-line investigation for patients with suspected scimitar syndrome on echocardiography (41, 44-46).

### **3-6. Treatment and prognosis**

In the presence of significant left to right shunting and pulmonary hypertension,

surgical intervention should be considered. This involves developing an interatrial baffle to redirect the pulmonary venous return into the left atrium. Alternatively, the abnormal vein can be re-implanted directly into the left atrium. Considering the wide clinical spectrum of scimitar syndrome, the medical intervention depends on the severity of presentation and the amount of blood flowing to the IVC from completely or partially anomalous pulmonary veins. When there is a small amount of drainage, therapy may not be required. Management is often supportive and may include the prescription of cardiac medication in the event of volume overload (47). The use of antibiotics for chest infections, the promotion of good nutrition, oxygen supplementation and the prescription of Sildenafil for pulmonary hypertension could be effective. The respiratory symptoms are typically one of the main indications of surgical correction. Simple ligation or coil embolization of abnormal arterial vessels has been advocated as the most effective form of treatment, particularly in symptomatic infants. In contrast, many researchers recommended the repair of the anomalous venous return and the ligation of collaterals in symptomatic patients. Thrombosis and fibrosis of the redirected pulmonary veins represent serious complications of the surgical re-implantation procedure, which often requires re-thoracotomy with resection of the remaining lung. Pneumonectomy (either as a primary therapy or after repair failure) has yielded similar results (48, 49). Its severity and frequency is related to the degree of pulmonary hypoplasia. Lobectomy or even right Pneumonectomy may be required to deal with bronchiectasis and prevent further respiratory infections (3). A two-staged approach for the interventional management of scimitar patients has been proposed. The occlusion of systemic collaterals in the first stage showed clinical

improvement and reduced left to right shunt and pulmonary artery pressure. Yet, there is no consensus on the management guidelines for this syndrome. The other approach is coiling with diuretics, which produces a drop in pulmonary blood pressure in periods 3 months compared to patients receiving advanced pulmonary hypertension medications (sildenafil and or iloprost) with or without coiling. Although the review was retrospective and the sample was small to draw conclusions, we observed that advanced pulmonary hypertension medications were useful as the first-line treatment (50, 51). Gastroesophageal reflux disease was another risk factor of hospital admission. This comorbidity should be addressed to enable better control of symptoms and reduce hospital admissions in a select subset of infants with scimitar syndrome. Surgical approaches to the Scimitar syndrome vary according to the anatomic and pathologic features presented in each case (25).

#### 4- CONCLUSION

Scimitar syndrome is an infrequent disease with diverse presentations and associations, as described in this paper. Thus, diagnosing infantile scimitar syndrome requires meticulous attention and high suspicion of the early referral and management. Scimitar syndrome has associated congenital cardiac and pulmonary anomalies which should be considered. The association of the syndrome with pulmonary hypertension leads to recurrent and prolonged hospitalization. Considering the wide clinical spectrum of scimitar syndrome, the medical intervention depends on the severity of presentation and the amount of blood flowing to the IVC from completely or partially anomalous pulmonary veins. This condition can be initially suspected from a chest X-ray, but it is typically confirmed via CT angiography.

**5- CONFLICT OF INTEREST:** None.

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