

## Case Report

# Scimitar syndrome: a rare congenital venolobar anomaly

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### ABSTRACT

Scimitar syndrome is a rare congenital anomaly presenting with partial anomalous pulmonary venous drainage from right lung to inferior vena cava. This is the rarest anomaly seen in 1 to 3 in 100000 births. Embryologically it is thought to be a primary developmental anomaly of lung with secondary anomalous venous drainage. This is seen commonly in right lung and is common in females. Most of the cases are asymptomatic or minimally symptomatised. If symptoms are present patient usually presents with pulmonary hypertension and symptoms of left to right shunt. Clinically this syndrome is diagnosed by chest x-ray by the presence of "Scimitar sign". Surgical treatment effective in this syndrome.

**Keywords:** Scimitar syndrome, Congenital anomaly, Anomalous pulmonary venous drainage

## INTRODUCTION

Scimitar syndrome or congenital pulmonary venolobar syndrome is an unique and rare form of lobar hypoplasia associated with other anomalies of the pulmonary vessels. This is a very rare anomaly and has reported incidence of 1-3 cases per 100000 live births. Radiologically the shadow extends from the lateral superior position of right lung to cardiophrenic angle. The appearance closely resembles that of Turkish sword or Scimitar.<sup>1</sup> The scimitar syndrome is most commonly seen in right side. There are rare cases reported on left side and rarely on both sides.<sup>2</sup> This syndrome is associated with lung hypoplasia with partial anomalous pulmonary venous drainage to right atrium, coronary sinus, Inferior Vena Cava (IVC), or hepatic circulation.<sup>3-8</sup> Sometimes this is associated with cardiac anomalies like Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), Patent Ductus Arteriosus (PDA), tetralogy of Fallot, coarctation, hypoplastic left heart or endocardial cushion defects, diaphragmatic hernia, and anomalies of the bony thorax

or thoracic soft tissues. Embryologically it is thought to represent a primary developmental anomaly of the right lung with secondary anomalous venous drainage. Diagnosis of this syndrome is made by "Scimitar sign" in chest X-ray. This syndrome has varied presentation from asymptomatic state to severe pulmonary hypertension<sup>9</sup> and heart failure.<sup>10</sup>

**History:** George Cooper and Raoul Chassinat are credited with the earliest description of the anomaly in the medical literature<sup>11</sup> In 1836, they individually described single case of a rare congenital malformation that included venous drainage from the lung beneath the diaphragm. The first authors to use the term 'scimitar' were Halasz et al. in 1956.<sup>12</sup> They referred to a vertical curvilinear vein resembling a scimitar, draining all or a portion of the right lung to the right atrium or more commonly to the IVC. The first detailed radiographic description of the 'scimitar sign' (the presence of a scimitar vein on a chest radiograph) was by Dotter et al. in 1949.<sup>13</sup> The term 'scimitar syndrome' first appears in

the 1960 article by Neill et al. The name of the syndrome has subsequently been refined, and the preferred term 'congenital pulmonary venolobar syndrome' was coined by Dr Ben Felson in 2003. The first report of surgical therapy for Scimitar syndrome was in 1950 by Drake and Lynch who performed right lobectomy.<sup>14</sup>

**Embryology:** Scimitar syndrome arises during the embryologic development of one or more of the pulmonary veins maintain systemic venous connection, instead of connecting to the common pulmonary vein along with lung hypoplasia. As a result, one or more pulmonary veins will drain to a location other than the left atrium - to the systemic veins (superior vena cava (SVC), inferior vena cava (IVC), subclavian vein (SCV), brachio cephalic vein (BCV), azygos vein, right atrium or coronary sinus. This anomaly results in left-to-right shunt. The development of the respiratory system entails both structural development of the lung and lung maturation in order to have normal lung function. Lung development occurs during fetal growth in five phases. The embryonic period occurs when the lung first appears as a ventral bud off the esophagus. The lung bud then elongates to form the two main stem bronchi. Subsequent branching gives rise to the conducting airways. Lobar segments are formed by 37 days then progress to segmental airways by 42 days and further division into sub segmental bronchi by 48 days. The pulmonary vasculature branches off the sixth aortic arch to form a vascular plexus within the mesenchymal of the lung bud. Many of the severe tracheal and pulmonary abnormalities occur during this early embryogenesis period.

The second stage involves further branching of about 15 to 20 generations of the airways and occurs during the 7<sup>th</sup> to 18<sup>th</sup> week of gestation. This stage is called the pseudoglandular stage. There is some epithelial differentiation that occurs with the appearance of ciliated cells, goblet cells and basal cells. Pulmonary arteries grow in conjunction with the airways with the principal arterial pathways being present by 14 weeks. Pulmonary venous development occurs in parallel but with a different pattern that demarcates lung segments and subsegments. By the end of the pseudoglandular stage, airways, arteries and veins are similar in pattern to an adult. The third stage is the canalicular stage that occurs between 16 and 25 weeks gestation. This represents the transformation of the pre-viable lung to the viable lung that can exchange gases. The three major changes that occur during this stage are the appearance of the alveolar air sacs, epithelial differentiation with the appearance of an air- blood barrier, and the presence of surfactant secreting type II cells. The formation of a capillary network to occur in tandem with saccular branching is critical for air exchange. Failure for this to occur can result in alveolar-capillary dysplasia.

The fourth and fifth stages of lung development are the saccular and alveolar stages, respectively. The saccular stage encompasses the period of development from 25

weeks until term. The saccular stage involves the terminal or distal airway that elongates branches and widens until alveolarization is complete. Alveolarization is initiated in the terminal saccules by the appearance of septa in association with capillaries, elastin fibers, and collagen fibers. Alveoli are increasing in number with the most rapid increase from 32 weeks gestation until the first few months after term delivery. New alveoli continue to form until 7-8 years of age. In the human embryo, primordial lung bud initially drains into the cardinal venous system, without having any connection with the heart. By days 32-33, the common pulmonary vein arises from the left atrium, establishing a connection with the pulmonary vascular bed.

Once a direct connection with the heart is established, connections to the cardinal venous system begin to involute, with blood draining from the developing lung to the common pulmonary vein and then to the left atrium through four individual pulmonary veins.

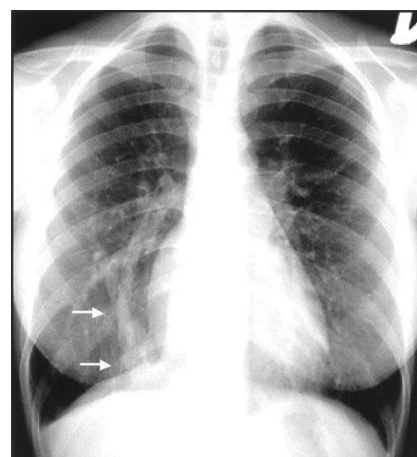
Finally, the common pulmonary vein incorporates into the left atrium and the four pulmonary veins. In normal conditions, four pulmonary veins carrying oxygenated blood drain into the left atrium. The right superior pulmonary vein drains the right upper and middle lobes, the left superior pulmonary vein drains the left upper lobe and lingula, and the right and left inferior pulmonary veins drain the lower lobe.

## CASE REPORT

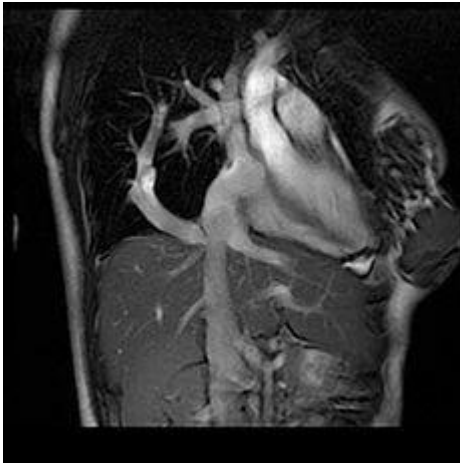
This case report describes 21 year old female with known scimitar syndrome undergoing treatment for recurrent respiratory infection and dyspnoea.

Chest radiography showed dextrocardia, elevated right diaphragm and the Scimitar sign.

She was with partial anomalous pulmonary venous connection to inferior vena cava.



**Figure 1: X-ray showing Scimitar sign.**



**Figure 2: MRI scan showing pulmonary vein draining in to inferior vena cava.**

## DISCUSSION

Scimitar or congenital venolobar syndrome is the rarest congenital anomaly of lung with partial anomalous drainage of pulmonary vein. Though it is a rare anomaly the patient is minimally symptomatic state to severe pulmonary hypertension and heart failure. The presentation is different in different age groups.

Mostly it is characterized by a combination of cardiopulmonary anomalies including partial anomalous pulmonary vein draining to the IVC, as this appearance resembles a scimitar, a curved Turkish sword venous return connection of the right lung to the inferior vena cava leading to the creation of a left-to-right shunt. Females are more frequently affected than males. In the majority of cases, the disease manifests in the first months of life. In the neonatal period, the disease presents with congestive cardiac failure, most commonly due to pulmonary hypertension and respiratory distress. The right lung is most frequently involved. Not a single spectrum of disease is seen but variable and different degrees of hypoplasia and malformations of the pulmonary arteries are found in the affected lung, as well as arterial supply from the aorta, which can also arise above or below the diaphragm. An association with congenital heart disease (Coarctation of aorta, Tetralogy of Fallot, Patent arterial duct or Ventricular septal defect) is seen. Pulmonary venous drainage into the right atrium, superior vena cava, the azygos system, the hepatic vein, or the left atrium has also been described. Rarely, a scimitar syndrome of the left lung has been reported. In most of the cases there is a combination of hypoplastic lung and partial anomalous pulmonary venous return. The anomalous vein drains into right atrium, IVC or even portal vein. It can be present in early life as well as in adults. Partial Anomalous Pulmonary Venous Return (PAPVR) has a prevalence of 0.4-0.7% in the general population, with prevalence abruptly rising in patients with atrial septal defects (10-15% in ostium secundum type and nearly 100% in sinus venosus type).

PAPVR arises during the embryologic development. One or more of the pulmonary veins maintain systemic venous connection, instead of connecting to the common pulmonary vein. As a result, one or more pulmonary veins will drain to a location other than the left atrium - to the systemic veins (SVC, IVC, SCV, BCV, azygos vein), right atrium or coronary sinus.

This anomaly results in left-to-right shunt, with partial admixture of deoxygenated and oxygenated blood. However this shunt is usually hemodynamically insignificant, with most patients being asymptomatic or mildly symptomatic. As such, it is commonly detected incidentally on imaging studies.

As the above described systemic thoracic anomalies, PAPVR may be associated with other congenital heart diseases, namely atrial, ventricular or atrioventricular septal defects, tetralogy of Fallot and coarctation of aorta.

Three types of PAPVR are described, with right-side PAPVR being more frequent than the left.

1. Right upper lobe venous drainage into the low SVC or superior cavoatrial junction.
2. Left pulmonary venous drainage to the left brachiocephalic vein.
3. PAPVR may be associated with complex pulmonary anomalies in the spectrum of congenital pulmonary venolobar syndrome.

Though the anomalous vein most frequently drains to the IVC below the right hemidiaphragm, it may rarely drain into the suprahepatic portion of the IVC, to the hepatic veins, to the portal vein.

## CONCLUSION

The disease itself is a rarity and few clinicians come across such cases and that too when they do not suspect the disease and is usually an incidental finding. But a detailed investigation can reveal the disease which otherwise can lead to fatal outcome. Fortunately in our case the female had no associated defects which can occur as a part of the syndrome.

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