SCIMITAR SYNDROME

Synonyms:
- Congenital pulmonary venolobar syndrome
- Epibronchial right pulmonary artery syndrome
- Halasz syndrome
- Hypogenetic lung syndrome

Definition: 
Scimitar Syndrome is a complex and uncommon congenital malformation of the vascular structures (arteries and veins), bronchia and tissue of the right lung. It is a partial or total anomaly in the venous return from the right lung to the inferior vena cava below or above the diaphragm. This means that the right lung veins return loaded with oxygenated blood to the inferior vena cava by mistake, instead of draining into the left atrium. It can be related to:
1) Malformations of the right lung: hypoplasia (under-development), or bronchial and lobar defects.
2) Heart located on the right side of the thorax (dextroposition of the heart).
3) Anomalous pulmonary blood supply by arteries coming from the descending aorta.
4) Hypoplasia (under-development) of the right pulmonary artery branch.
5) Atrial septal defect.
Clinical Picture:
Two groups of patients are defined, based on clinical presentation (See Clinical Picture in Diagnosis and Treatment):
1) Infants: symptoms appear before the first year of life. These children typically present heart failure and excessive pulmonary blood flow. They usually suffer from shortness of breath, they do not grow properly and they have excessive sweating. They can turn blue (cyanotic) and have repeated pneumonies.
2) Adults: children over 1 year, youth and adults. Normally, they present no symptoms, however, they may show slight symptoms of heart failure (agitation and fatigue, for instance) and occasional respiratory infections.

Diagnosis:
The name “Scimitar Syndrome” refers to the special way the anomalous veins can be seen in the chest x-ray, which reminds the curved Turkish sword that carries that name (See Diagnosis in Diagnosis and Treatment).

The x-ray also shows right lung hypoplasia (under-development) and the heart located on the right side of the thorax (dextrocardia). These two elements in the chest x-ray in the context of a small child with severe respiratory stress should guide the doctor towards this diagnosis.

For older children and adults, the diagnosis is usually made accidentally after a chest x-ray with the characteristic signs of scimitar syndrome. These patients usually present a murmur when auscultated, and they have a previous history of respiratory infections.

The echocardiogram is an important diagnostic tool. The anomalous connection of the pulmonary vein is identified in the inferior vena cava. The right pulmonary artery branch can be hypoplastic. The presence of related malformations is also assessed, such as atrial septal defect and persistent ductus arteriosus.

Tomography and magnetic resonance clearly show the anatomy. Diagnostic catheterization is useful to find aortopulmonary collaterals and also to measure the pulmonary pressure, since it can be increased, above all, in young symptomatic children (See Haemodynamics in Diagnosis and Treatment).
Treatment:

Treatment should be planned according to each patient. There are patients whose symptoms are mild or absent, and there are no signs of excessive pulmonary blood flow. In these cases, only periodic checks are carried out. When the blood flow towards the lungs is excessive, it is usually necessary to treat the patient. If there were collaterals coming from the aorta towards the right lung, they should be closed by catheterization. Sometimes, symptoms disappear with this treatment and the patient is compensated.

But in the cases in which this is not enough, a surgery must be performed to deviate the blood flow from the scimitar vein to the left atrium. There are several strategies to correct this malformation. In the “classic” operation, the heart is connected to cardiopulmonary bypass (See Surgery in Diagnosis and Treatment), it is isolated from the circulation and the right atrium is opened. The atrial septal defect (if present) and the anomalous pulmonary vein draining in the inferior vena cava are observed through it. A patch taken from the own pericardium is used to connect the opening of the anomalous vein to the atrial septal defect. In this way, blood coming from the anomalous pulmonary vein goes through the tunnel constructed with the patch and ends in the left atrium. Given the fact that the tunnel is long, there is an important risk of obstruction of either the tunnel or the inferior vena cava. To avoid it, many other surgical techniques have been developed. The vein can be detached and reinserted in the right atrium to tunnel it later on (with a smaller patch) to the left atrium through the atrial septal defect. Another strategy consists in implanting the vein directly in the left atrium, with or without interposition of a tube graft.

Some surgical approaches to scimitar syndrome surgical treatment

A new technique has recently been described by the author of this page (Ignacio Lugones, MD). The anomalous vein is connected to the lateral pericardium. After cardiopulmonary bypass is established, the left atrium is wide opened. Then the pericardium is sutured around this opening, creating a tunnel made of own pericardium to redirect blood flow from the pulmonary vein to the left atrium.

This technique has proved to be safe and effective, and has many advantages. The wide connection that is achieved determines a low risk of pathway obstruction. Damage of the conduction tissue (and therefore arrhythmias) and obstruction of the inferior vena cava are unlikely, and there is no need to open the interatrial septum or
the diaphragm as in other techniques. From the technical point of view it is a straightforward procedure that can be applied in all anatomic variants without the use of synthetic grafts. It is especially useful in cases of stenosis (small size) of the scimitar vein in its connection to the inferior vena cava. The risks of this procedure are low (See Postoperative in Diagnosis and Treatment).

**Lugones operation**
(for scimitar syndrome repair)

A) The scimitar vein (SV) is closed just above the diaphragm (D). Then, its lateral aspect is opened and sutured to a hole made in the pericardium (P) that surrounds the heart. B) The vein is now draining inside the pericardial cavity. Cardiopulmonary bypass is established, the heart is arrested and the left atrium (LA) is opened. C) The pericardium is sutured to the lateral external surface of the right atrium, making a tunnel that connects the vein with the left atrium. D) The operation is finished. Blood uses this pericardial tunnel to return to the left atrium.
This technique has been published in one of the most prestigious journals of thoracic surgery (Annals of Thoracic Surgery 2014, Volume 97, page 353) and presented in the 4th Meeting of the World Society for Pediatric and Congenital Heart Surgery.

**Prognosis:**

Prognosis depends on the anatomy and the chosen treatment. In older children without severe right lung hypoplasia and with an adequate correction, prognosis is excellent. These patients typically return to their daily lives without complications. They should have regular checkups, especially to confirm that reconstructed pathways are not obstructed.

The opposite case is that of a small baby severely ill, with severe lung hypoplasia and significant heart failure. For these children, prognosis is not that good, since they suffer from pulmonary sequelae which, many times, are significant. Nevertheless, they still have a good life expectancy.

In between these two opposite conditions, there are many varieties of presentation and evolution after treatment (**Follow-up and Control** in Diagnosis and Treatment).