

## Scimitar syndrome associated with ventricular septal defect and left pulmonary vein stenosis

*Ventriküler septal defekt ve sol pulmoner ven stenozu ile ilişkili Scimitar sendromu*

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Scimitar syndrome is a rare congenital cardiac anomaly, which is characterized by anomalous right pulmonary venous return to the inferior vena cava. In this article, we report a six-month-old girl with the diagnosis of Scimitar syndrome associated with ventricular septal defect, left pulmonary venous stenosis and severe pulmonary hypertension. The patient underwent a successful surgical treatment of all pathologies at a single session.

**Key words:** Left pulmonary venous stenosis; Scimitar syndrome; ventricular septal defect.

Scimitar syndrome is a rare congenital cardiac anomaly characterized by anomalous pulmonary venous return of the right lung to the inferior vena cava (IVC). The anomaly is usually associated with right lung hypoplasia with dextroposition of the heart and abnormal arterial supply of a variable degree to the right lung.<sup>[1-4]</sup> The presence of additional cardiac anomalies generally aggravates the clinical picture by causing significant left-to-right shunt, congestive heart failure, and severe pulmonary hypertension. Herein, we present a patient with scimitar syndrome associated with ventricular septal defect (VSD), left pulmonary venous stenosis (PVS), right pulmonary arterial stenosis (PAS), and abnormal arterial supply from the abdominal aorta to the right lung.

### CASE REPORT

A six-month-old girl was admitted to our clinic with dyspnea and recurrent respiratory tract infections.

Scimitar sendromu, anomalili sağ pulmoner venöz dönüşün inferior vena kavaya olduğu, nadir görülen doğuştan bir kalp anomalisidir. Bu yazıda, ventriküler septal defekt sol pulmoner ven stenozu ve ciddi pulmoner hipertansiyon ile ilişkili Scimitar sendromu tanısı olan altı aylık bir kız olgu sunuldu. Hasta tüm patolojilerin tek seansta tedavi edildiği başarılı bir cerrahi tedavi geçirdi.

**Anahtar sözcükler:** Sol pulmoner ven stenozu; Scimitar sendromu; ventriküler septal defekt.

Chest radiography showed right lung hypoplasia and dextrocardia, and an echocardiographic evaluation revealed partial anomalous pulmonary venous return (PVR) from the right lung to the IVC, right PAS, inlet muscular-type VSD (12 mm), and stenosis at the entrance of the left pulmonary veins to the left atrium (LA). In addition, a subsequent computed tomography scan detected an abnormal collateral artery originating from the abdominal aorta which was supplying the lower lobe of the right lung (Figure 1). Cardiac catheterization confirmed the initial diagnosis and revealed that the patient's pulmonary arterial pressure (PAP) was at a systemic level (Figure 2). After the establishment of the definitive diagnosis, we first decided to close the aberrant artery by percutaneous intervention. The patient then underwent coil embolization at the catheterization laboratory and had surgery two days later.



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The operation was carried out through a median sternotomy. Cardiopulmonary bypass (CPB) was initiated in the usual manner, but the IVC was cannulated directly below the entrance point of the anomalous pulmonary vein. The procedure was performed at moderate hypothermia, and cardiac protection was achieved by cold intermittent antegrade blood cardioplegia. After applying a cross-clamp, the anomalous vein was found and dissected from the surrounding tissue. We then separated it from the IVC and reimplanted it in the back wall of the LA. Next, the VSD was closed with a Dacron patch via the right atrium (RA), and the left PVS was repaired by a sutureless technique using the in situ pericardium.<sup>[5]</sup> Finally, the proximal aspect of the right pulmonary artery was enlarged with an appropriate-sized, glutaraldehyde-treated pericardium. The patient was weaned off of CPB with moderate doses of inotropic support. The CPB and cross-clamp times were 85 and 72 minutes, respectively. Afterwards, the mean PAP was measured at 55 mmHg, and nitric oxide (NO) inhalation was initiated (15 ppm). Although the early postoperative period was uneventful, the patient could not be extubated. At the end of two weeks, we decided to perform a tracheostomy to achieve more effective feeding and mobilization, and the patient then had a slow, but smooth recovery period. After two weeks, the patient was completely weaned off of the mechanical ventilator and was decannulated, and a postoperative echocardiographic evaluation revealed normal cardiac function and the PAP was near normal levels.



**Figure 1.** Computed tomography showing the abnormal collateral artery originating from the abdominal aorta, which was supplying the lower lobe of the right lung.

## DISCUSSION

Scimitar syndrome is a rare anomaly with an incidence of 1-3/100000 live births.<sup>[6]</sup> The clinical presentation of the patients varies widely from those who are asymptomatic to those with significant heart failure, and the age at presentation can range from infancy to adulthood.<sup>[1]</sup> Patients for whom the diagnosis is made during the first year of life generally have more severe symptoms and a higher incidence of heart failure and pulmonary hypertension.<sup>[6]</sup> This may be attributable solely to the obligatory shunt from the anomalous pulmonary venous drainage, but it is more likely the result of an additional congenital cardiac lesion, such as an atrial septal defect (ASD) or VSD. Our patient was six months old when she was diagnosed with scimitar syndrome associated with VSD, left PVS, and an abnormal arterial supply to the right lung. Her PAP was at a systemic level preoperatively but was nearly normal during the postoperative follow-up period.

The techniques for intracardiac repair include direct implantation of the anomalous vein to the LA, as performed on our patient, an intracardiac patch baffling the anomalous vein to the LA through an ASD, and reimplantation of the anomalous vein into the RA and baffling it to redirect flow to the LA.<sup>[6]</sup> The main concern with this type of surgery is the occurrence of pulmonary venous stenosis on follow-up, which can lead to pulmonary hypertension and hemoptysis. In addition, intracardiac baffle repair reportedly causes postoperative pulmonary venous obstruction more frequently than other techniques.<sup>[1,2]</sup> Therefore, we decided to perform a direct reimplantation since it offers superior long-term results.



**Figure 2.** Cardiac catheterization demonstrated partial anomalous pulmonary venous return from the right lung to the inferior vena cava.

The role played by the anomalous systemic arterial collaterals in this syndrome is unclear. It has been suggested that their occlusion may prove to be significant in the treatment of both pulmonary hypertension and heart failure.<sup>[2]</sup> On the other hand, Huddleston et al.<sup>[1]</sup> reported that occlusion of the collaterals alone is not sufficient and does not replace the need for repair. In our case, complete repair was carried out following coil embolization of the abnormal arterial supply.

In conclusion, scimitar syndrome presenting in infancy is more challenging than when it is diagnosed in the elderly. In case of early presentation, the surgical outcome is highly dependent on the precise and accurate correction of these anomalies.

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