

# Abnormal location of umbilical venous catheter due to Scimitar syndrome

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## ABSTRACT

**Scimitar syndrome is a rare congenital anomaly where the right pulmonary veins return to the inferior vena cava (IVC) just below the diaphragm. On chest X-ray (CXR), an IVC catheter will be in a bizarre location outside the heart if it inadvertently passes into the scimitar vein rather than into the right atrium.**

**Keywords:** Scimitar, syndrome, X-ray

## CLINICAL SUMMARY

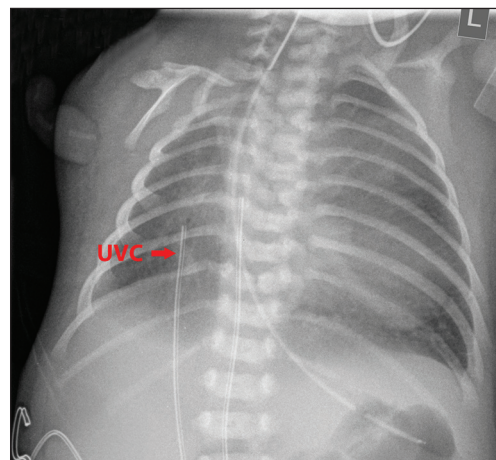
A 37-week-old female infant with multiple congenital anomalies (absent right arm with finger-like appendage at the shoulder, hypoplastic right lung, and multiple skeletal anomalies) was intubated immediately after birth for apnea. On CXR, the umbilical arterial catheter was in the normal position but the umbilical venous catheter was in a very abnormal location — lateral and to the right of the cardiac border [Figure 1]. An echocardiogram demonstrated an obstructed right pulmonary vein returning to the inferior vena cava (IVC) consistent with Scimitar syndrome — a large secundum atrial septal defect [Figure 2, Video 1], hypoplastic right pulmonary artery, and a patent ductus arteriosus. A magnetic resonance imaging (MRI) scan confirmed the presence of the narrowed (arrow) scimitar vein (SV), [Figure 3]. The unusual location of the umbilical venous catheter is due to its passing from the IVC into the scimitar vein, which courses to the right of the cardiac border.

## DISCUSSION

During normal development,<sup>[1]</sup> the lungs are surrounded by a vascular plexus of the foregut called the splanchnic plexus,

which gives rise to the pulmonary venous confluence. Early in gestation, the splanchnic plexus/developing pulmonary venous confluence is connected to the umbilicovitelline and cardinal venous systems. The common pulmonary vein grows out of the back wall of the left atrium and becomes connected to the developing pulmonary venous confluence. The connections between the developing pulmonary venous confluence and the umbilicovitelline and cardinal venous systems then involute as the pulmonary venous confluence becomes incorporated into the back wall of the left atrium. This results in all of the pulmonary veins returning to the left atrium.

When development of the pulmonary venous confluence is disrupted and the only connection of the right pulmonary veins is to the umbilicovitelline venous system, they ultimately enter the IVC just below the diaphragm. This



**Figure 1: Chest X-ray showing abnormal location of the inferior vena cava (IVC) catheter**

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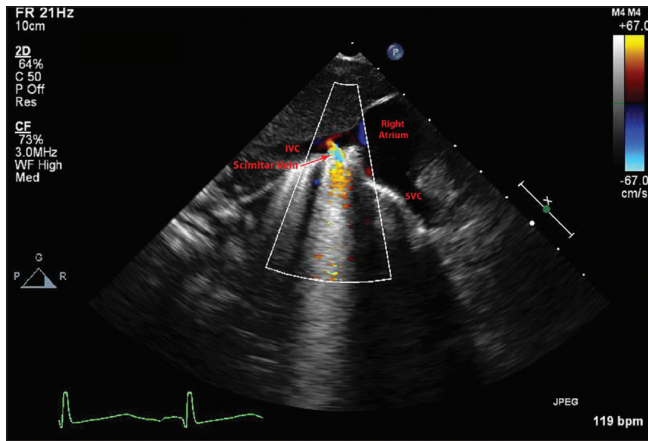
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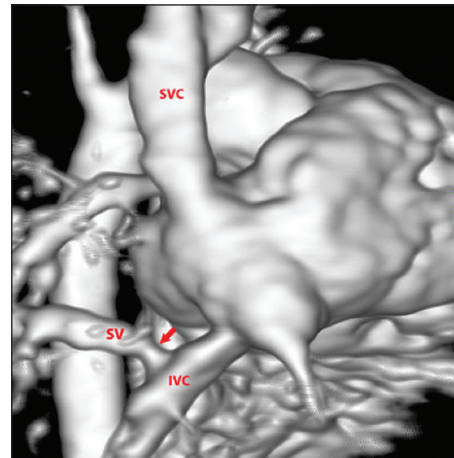
**Figure 2: Echocardiogram still frame showing scimitar vein entering the inferior vena cava (IVC) just below the diaphragm**

gives an abnormal “fir tree” pattern to the right pulmonary veins, which Neill *et al.*<sup>[2]</sup> called Scimitar syndrome because the abnormal pulmonary veins resembled a curved sword originating in southwest Asia called a scimitar.

In addition to the pulmonary venous anomaly, Scimitar syndrome is associated with an intact atrial septum (usually), right lung and right pulmonary artery hypoplasia, bronchial anomalies, horseshoe lung, secondary dextrocardia, anomalous arterial connection from the aorta to the right lung, and pulmonary sequestration.<sup>[3]</sup> Diverse congenital thoracic anomalies may also be associated with this anomaly. Scimitar syndrome can be suspected on CXR and confirmed with echocardiography and computerized tomography (CT)/MRI scanning.<sup>[4,5]</sup> Surgical repair is accomplished by connecting or baffling the anomalous pulmonary veins to the left atrium.<sup>[4]</sup>

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**Figure 3: MRI scan showing the SV (arrow) entering the IVC. SVC: Superior vena cava, IVC: Inferior vena cava, SV: Scimitar vein**

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