

Prenatal diagnosis of scimitar syndrome

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Scimitar syndrome is a rare congenital anomaly, representing 0.5–1% of congenital heart defects. It occurs at a rate of 1–3 per 100,000 live births, being twice as common in female fetuses [1]. This syndrome is a variant of right lung hypoplasia with partial anomalous pulmonary venous connection (PAPVC). It is characterized by the abnormal drainage of pulmonary veins into the inferior vena cava, ipsilateral underdevelopment of the lung and pulmonary artery, and anomalous vascular supply to the lung from branches of the thoracic aorta. Underdevelopment of the right lung leads to a displacement of the heart silhouette to the right side of the chest. Typically, this defect occurs on the right side, although there are rare cases reported where its development has occurred on the left side.

The name of the syndrome originates from the radiographic image of the chest, where a shadow resembling a scimitar (Turkish sword) is visible. This shadow is formed by the abnormal veins of the right lung running from the diaphragm towards the inferior vena cava. The pathogenesis of the syndrome is unclear, but it seems to originate from a basic developmental disorder of the entire lung bud early in embryogenesis [2]. The Scimitar syndrome may coexist with other congenital heart defects such as sinus venosus atrial septal defect, patent ductus arteriosus, or ventricular septal defect.

Advancements in prenatal diagnosis allow for the recognition of right lung hypoplasia and suspicion of "Scimitar syndrome" as early as in the first trimester, however, typically it is diagnosed in the second or third trimester of pregnancy [3]. Common findings on the prenatal ultrasound include right lung hypoplasia, displacement of the mediastinum, and abnormal pulmonary vein drainage. It must be emphasized the prenatal diagnosis of Scimitar Syndrome is extremely difficult in pregnancy. This clinical vignette presents valuable and highly informative pictures, which illustrate typical features of this abnormality.

We present two cases of prenatally diagnosed Scimitar Syndrome. Both cases were referred to as abnormal/suspected as early as in the first trimester. In later gestation MRI was performed to confirm right lung hypoplasia and abnormal pulmonary venous drainage. Figures 1–4 present early features of Scimitar Syndrome. In both cases the outcome was favorable. Both neonates were born term. In the first case the surgical treatment was not necessary, and the neonate was discharged on day 16 of life. The patient was qualified for follow up visits.

In the second case the neonatal echocardiography confirmed abnormal pulmonary vein drainage into the superior vena cava but also revealed moderate aortic coarctation. Based on the chest X-ray the suspicion of diaphragmatic hernia was established due to abnormal view of diaphragmatic dome. However, it was not confirmed in the further investigation. Surgical intervention was required within first days of life due to CoA and aortic arch hypoplasia. The patient was discharged on day 37 of life.

CONCLUSIONS

The Scimitar syndrome is a complex heart abnormality presenting with right lung hypoplasia. Modern ultrasound techniques and high-resolution ultrasound machines give a chance for these fetuses to be diagnosed as early as the second or

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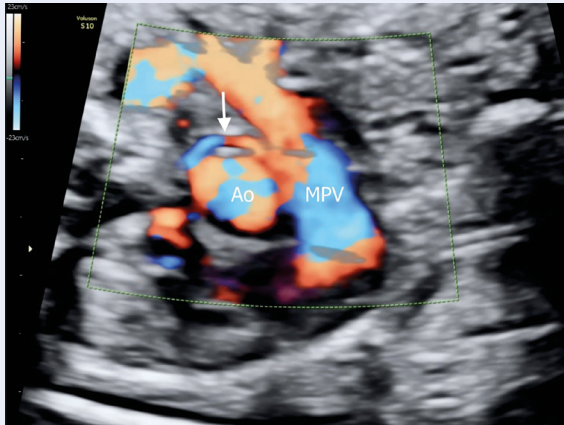


Figure 1. Axial plane of the chest in early second trimester; high Definition doppler (HD-doppler) showing both left pulmonary veins drainage to left atrium (white arrows). Right pulmonary veins are not recognized in typical location (asterisk). Note also the dextroposition of the heart when comparing to the normal position of left sided descending aorta (DA)

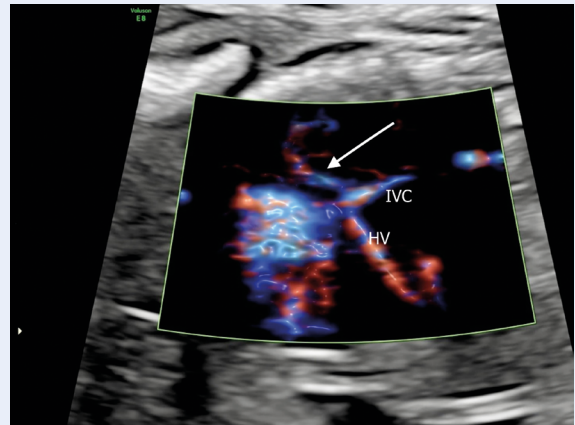


Figure 2. Longitudinal view of the fetal thorax and upper abdomen in early second trimester; slow flow doppler technique facilitates the visualization of flow within small vessels. The grey information in background are reduced to improve image contrast. White arrow shows the suspected anomalous right pulmonary vein (called scimitar vein) connected to the inferior vena cava (IVC). HV, hepatic vein

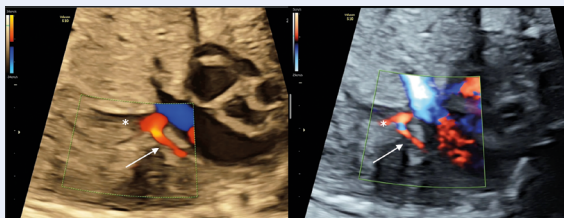


Figure 3. Second trimester; right ventricle output track plane and HD doppler showing the hypoplastic right pulmonary artery (arrow). Ao, aorta; MPV, main pulmonary artery

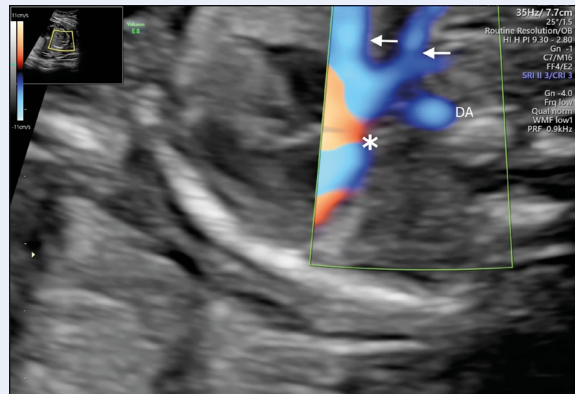


Figure 4. Early third trimester; longitudinal view in color (left) and HD doppler (right) showing the anomalous connection of the right pulmonary vein (scimitar vein) to the inferior vena cava (asterisk)

even first trimester of pregnancy. However, the Scimitar Syndrome might not be an isolated abnormality, which makes the diagnosis more complex. In isolated cases prognosis is good and depends mostly on severity of pulmonary hypertension.

Article information and declarations

Author contributions

Aleksandra Sliwka: manuscript writing, Marcelina Szytler-Krakowska: manuscript writing, Karol Bielasik: manuscript writing, Przemyslaw Kosinski: conceived of the presented idea, manuscript writing, supervision, final approval.

Conflict of interest

Authors declare no conflict of interest.

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