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Prenatal diagnosis of Scimitar syndrome associated with duplicated superior vena cava in fetal echocardiography: A case report

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Case Report

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Abstract

Background

Scimitar syndrome is a rare congenital cardiopulmonary anomaly featuring partial anomalous pulmonary venous return from the right lung to the inferior vena cava, right lung hypoplasia, and cardiac dextroposition. Prenatal diagnosis remains challenging due to subtle imaging findings and variable presentations, with early second-trimester detection and association with duplicated superior vena cava being exceptionally uncommon. This case underscores the value of recognizing such rare combinations through fetal echocardiography to refine diagnostic criteria.

Case presentation

A 33-year-old gravida 3 para 1 Iranian woman with unremarkable history was referred for fetal echocardiography at 18 weeks and 6 days gestation after routine ultrasound suggested abnormal cardiac position. Examination revealed a four-chambered heart with mesocardia and right lung hypoplasia. Right pulmonary veins drained anomalously via a collector into the inferior vena cava-right atrium junction. Additional findings included duplicated superior vena cava (left draining to dilated coronary sinus), mild-to-moderate tricuspid valve dysplasia with regurgitation (velocity ~ 70 cm/s), and prominent fossa ovalis consistent with secundum atrial septal defect. No extracardiac anomalies were noted. Multidisciplinary counseling addressed risks of neonatal respiratory distress, pulmonary hypertension, and need for postnatal interventions.

Conclusions

Comprehensive fetal echocardiography enables early detection of complex Scimitar syndrome variants, facilitating perinatal planning, timely postnatal evaluation, and potential surgical or catheter-based repairs to mitigate right heart overload and improve outcomes.

Introduction

Scimitar syndrome (SS) is an uncommon form of partial anomalous pulmonary venous return (PAPVR), with an estimated incidence of approximately 1–3 per 100,000 live births [1]. It is classically defined by anomalous venous drainage of the right lung into the inferior vena cava (IVC), right lung and right pulmonary artery hypoplasia, and cardiac displacement toward the affected side. The term “Scimitar” refers to the curved sword-like appearance of the anomalous draining vein on chest imaging [2].

Clinically, SS exhibits a broad spectrum ranging from a severe infantile form, frequently associated with pulmonary hypertension and multiple cardiopulmonary anomalies, to a milder adult form often detected incidentally. Prenatal diagnosis is difficult and has been only rarely reported, largely because key signs such as abnormal pulmonary venous drainage and systemic arterial supply to the hypoplastic lung can be challenging to visualize on routine obstetric scans. The most important prenatal clue is abnormal

cardiac position with mediastinal shift toward a hypoplastic right lung, which should prompt a systematic search for anomalous pulmonary venous connections [3, 4].

Duplicated superior vena cava (SVC), most often presenting as a persistent left SVC draining into a dilated coronary sinus, is a recognized but infrequently documented association of SS [5]. Documenting such rare combinations is valuable for refining prenatal diagnostic criteria and optimizing postnatal management strategies. This report describes an early second-trimester diagnosis of SS associated with duplicated SVC and multiple right heart anomalies, emphasizing the echocardiographic features and implications for perinatal care.

Case presentation

A 33-year-old gravida 3, para 1 Iranian woman was referred for fetal echocardiography at 18 weeks and 6 days of gestation following a routine anomaly scan that raised concern for abnormal cardiac position. Her medical history, family history, and previous pregnancy were unremarkable, and there was no known consanguinity or exposure to teratogens.

Fetal biometry was appropriate for gestational age, and the fetal heart rate was 150 beats per minute. A four-chamber view demonstrated a structurally four-chambered heart with levo-apex but positioned in the midline of the thorax, consistent with mesocardia and associated with a reduced right hemithoracic volume suggestive of right lung hypoplasia. Color Doppler evaluation showed partial anomalous pulmonary venous return: two right pulmonary veins coalesced into a venous collector draining into the junction of the IVC and the right atrium, confirming a Scimitar-type anomalous connection (Fig. 1).

Systemic venous assessment revealed duplicated SVC, with a right SVC draining normally into the right atrium and a left SVC draining into a markedly dilated coronary sinus. The tricuspid valve appeared mildly to moderately dysplastic, with echogenic, thickened leaflets and tricuspid regurgitation at a velocity of approximately 70 cm/s, indicating early right heart volume overload. The interatrial septum showed a prominent fossa ovalis measuring 5 mm in diameter, compatible with a secundum atrial septal defect (type II). No significant extracardiac anomalies or additional structural cardiac defects were identified at this gestational age.

The constellation of mesocardia, right lung hypoplasia, partial anomalous pulmonary venous return to the cavo-atrial junction, duplicated SVC, tricuspid valve dysplasia, and secundum atrial septal defect supported the diagnosis of SS with complex associated cardiac anomalies. Multidisciplinary counseling was provided, including discussion of the potential for neonatal respiratory compromise, right heart failure, pulmonary hypertension, and the likely need for early postnatal imaging and consideration of surgical or catheter-based interventions.

Discussion and Conclusions

The prenatal presentation of our case, characterized by mesocardia, right lung hypoplasia, partial anomalous pulmonary venous return to the cavo-atrial junction, duplicated SVC with a left SVC draining into a dilated coronary sinus, tricuspid valve dysplasia, and a prominent secundum atrial septal defect, represents a complex and rare constellation within the spectrum of SS. This anomaly pattern is consistent with the infantile, more severe form of the disease, in which multiple associated cardiovascular lesions and significant right-sided volume overload are common and prognostically relevant. The timing of diagnosis at 18 weeks and 6 days is notably early compared with most published prenatal series, in which detection is more often reported in the late second or third trimester [6,7].

From an echocardiographic standpoint, the most important prenatal clue in this and other reports is abnormal cardiac position with ipsilateral mediastinal shift in the context of unilateral right lung hypoplasia [6-9]. Once cardiac malposition is recognized, systematic interrogation of pulmonary venous return using coronal and sagittal views, supplemented by color and power Doppler, is essential to demonstrate anomalous drainage to the IVC or its junction with the right atrium and to distinguish SS from other causes of dextroposition, such as bronchopulmonary sequestration or congenital diaphragmatic hernia [8-10,6,7]. In line with previous series, the confluence at the cavo-atrial junction in our case corresponds to a typical site for the scimitar vein, reinforcing the diagnostic value of careful imaging of this region when a right-sided mediastinal shift is present [6-8].

Several recent reports have expanded the prenatal spectrum of SS and provide a useful framework for interpreting the present case (Table 1). Wang et al. [6] described six fetuses diagnosed between 26 and 32 weeks' gestation, all with right lung hypoplasia, rightward cardiac displacement, and right-sided anomalous pulmonary venous drainage, frequently associated with secundum atrial septal defect, ventricular septal defect, right aortic arch, and, in some cases persistent left SVC; most infants underwent postnatal surgical repair with favorable short-term outcomes. Recker et al. [7] similarly reported a six-case series in which all fetuses had right lung hypoplasia, mediastinal shift, and anomalous venous drainage to the IVC. However, the cohort was characterized by a high burden of additional major anomalies (including coarctation, diaphragmatic hernia, and VACTERL association) and substantial early mortality, underscoring the prognostic impact of associated defects. Kochanski et al. [11] presented a single fetus with a particularly complex Scimitar variant, including two right-sided scimitar veins draining to the suprahepatic IVC, horseshoe lung, right pulmonary hypoplasia, sequestrated lung tissue with an aortopulmonary collateral, and perimembranous ventricular septal defect, where multimodality prenatal imaging like echocardiography and fetal magnetic resonance imaging enabled targeted neonatal catheter-based and surgical interventions with good short-term outcome. Sliwka et al. [12] reported two prenatally diagnosed cases with right lung hypoplasia, mediastinal shift, and abnormal pulmonary venous drainage identified from the first or early second trimester onward, one requiring early neonatal surgery for associated aortic coarctation and arch hypoplasia, again emphasizing that prognosis largely depends on concomitant cardiovascular pathology and pulmonary hypertension.

Against this background, the case presented herein appears to represent an extremely rare prenatal phenotype. The diagnosis made at 18 weeks and 6 days is far quicker than in most of the published series; and the associated Scimitar-type partial anomalous pulmonary venous return, mesocardia, right lung hypoplasia, duplicated SVC with the left SVC draining into a markedly dilated coronary sinus, tricuspid valve dysplasia with significant regurgitation, and a prominent secundum atrial septal defect represent an anomaly complex not described in the reports above [6,7,11,12]. These features, taken together, extend the known prenatal phenotypic spectrum of SS and point to the necessity of systematically assessing the anatomy of the systemic and pulmonary venous systems when cardiac malposition and unilateral lung hypoplasia are detected on fetal echocardiography.

The hemodynamic profile inferred from our findings suggests substantial right-heart volume overload even in utero, driven by left-to-right shunting at the atrial level and through the anomalous pulmonary venous connection, with tricuspid valve dysplasia and significant regurgitation serving as early markers of right-sided stress. Similar associations between significant tricuspid regurgitation, pulmonary hypertension, and poorer early outcomes have been highlighted in clinical series of infantile SS, supporting the need for close perinatal surveillance in such complex variants [9,13,14]. Perinatal management of fetuses with prenatally suspected SS, therefore, centers on detailed postnatal imaging, including echocardiography and, when indicated, CT angiography, to fully delineate pulmonary venous drainage, systemic arterial supply, and associated intracardiac defects. In symptomatic infants or those with significant left-to-right shunt, pulmonary overcirculation, or pulmonary hypertension, early interventions may include embolization of systemic aortopulmonary collaterals and surgical redirection of anomalous pulmonary veins, together with repair of associated cardiac lesions. Outcomes vary and are strongly influenced by the severity of pulmonary hypoplasia, the presence of major associated anomalies, and the timing and success of interventions, with infantile presentations generally carrying a poorer prognosis than isolated, milder variants [6-8,4,15].

Practically, this case highlights several important points for fetal cardiologists and obstetric sonographers. First, any unexplained rightward mediastinal shift with preserved lung echogenicity and an intact diaphragm should prompt a focused search for anomalous pulmonary venous return, including careful assessment of the IVC–right atrial junction and the posterior venous confluence behind the left atrium [6-8]. Second, the venous survey should routinely include evaluation for persistent or duplicated SVC and coronary sinus dilatation, as systemic venous variants can coexist with SS and complicate postnatal surgical planning [6,7,1]. Finally, early identification of complex Scimitar variants supports multidisciplinary counseling, delivery planning in tertiary centers, and preparation for staged catheter-based and surgical management during infancy [14,4].

Our case highlights the necessity for precise and prompt prenatal diagnosis during standard obstetric ultrasound assessments. Early identification of such a complicated variant yields essential prognostic information and facilitates prompt intervention planning. Because there are serious associated defects (ASD type II) and signs of right heart volume overload (tricuspid dysplasia/TR), it is very important to perform an early postnatal work-up to confirm the full vascular and cardiac anatomy and to make it

easier for possible early surgical or catheter-based interventions. This will improve the prognosis for the affected newborn.

Abbreviations

ASD
Atrial septal defect
CT
Computed tomography
GA
Gestational age
IVC
Inferior vena cava
MRI
Magnetic resonance imaging
PAPVR
Partial anomalous pulmonary venous return
PDA
Patent ductus arteriosus
RA
Right atrium
SS
Scimitar syndrome
SVC
Superior vena cava
TR
Tricuspid regurgitation
VACTERL
Vertebral–anal–cardiac–tracheo-esophageal–renal–limb association
VSD
Ventricular septal defect

Declarations

Ethics and Consent to Participate

Ethical approval was not applicable. Written informed consent was obtained from the patient for publication of this case report and accompanying images. We confirmed the patient’s anonymity.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare no competing interests.

Data availability

No datasets were generated or analysed during the current study.

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Author contributions

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by all authors. The first draft of the manuscript was written by R.G. and all authors commented on previous and subsequent versions of the manuscript. All authors read and approved the final manuscript.

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Table

Table 1: Comparative overview of prenatally diagnosed Scimitar syndrome cases in the literature and the present fetus

Study	Summary of cases
Wang et al.	Six fetuses diagnosed at 26–32 weeks with right lung hypoplasia, rightward cardiac displacement, and right-sided Scimitar-type venous drainage, often with ASD, VSD, or right aortic arch; most underwent postnatal repair with good short-term outcomes.
Recker et al.	Six fetuses with right lung hypoplasia, mediastinal shift, Scimitar vein to the IVC, systemic arterial supply, and frequent major anomalies (ASD, coarctation, diaphragmatic hernia, VACTERL); half died within six months, survivors had significant morbidity.
Kochanski et al.	Single late-gestation fetus with right lung hypoplasia, dextroposition, two scimitar veins to suprahepatic IVC, horseshoe lung, sequestration with aortopulmonary collateral, and VSD; prenatal echo and MRI enabled targeted neonatal embolization and surgery with good early outcome.
Sliwka et al.	Two fetuses with early suspected Scimitar syndrome, right lung hypoplasia, mediastinal shift, and abnormal venous drainage; one isolated and observed, one with aortic coarctation requiring neonatal surgery; both had favorable early outcomes.
Present case	Single fetus diagnosed at 18+6 weeks with right lung hypoplasia, mesocardia, Scimitar-type PAPVR to the IVC–RA junction, duplicated SVC with left SVC to a dilated coronary sinus, tricuspid dysplasia with TR, and prominent secundum ASD, representing a rare early-gestation variant.

ASD, atrial septal defect; GA, gestational age; IVC, inferior vena cava; MRI, magnetic resonance imaging; PAPVR, partial anomalous pulmonary venous return; PDA, patent ductus arteriosus; RA, right atrium; SVC, superior vena cava; TR, tricuspid regurgitation; VACTERL, vertebral–anal–cardiac–tracheo-esophageal–renal–limb association; VSD, ventricular septal defect.

Figures

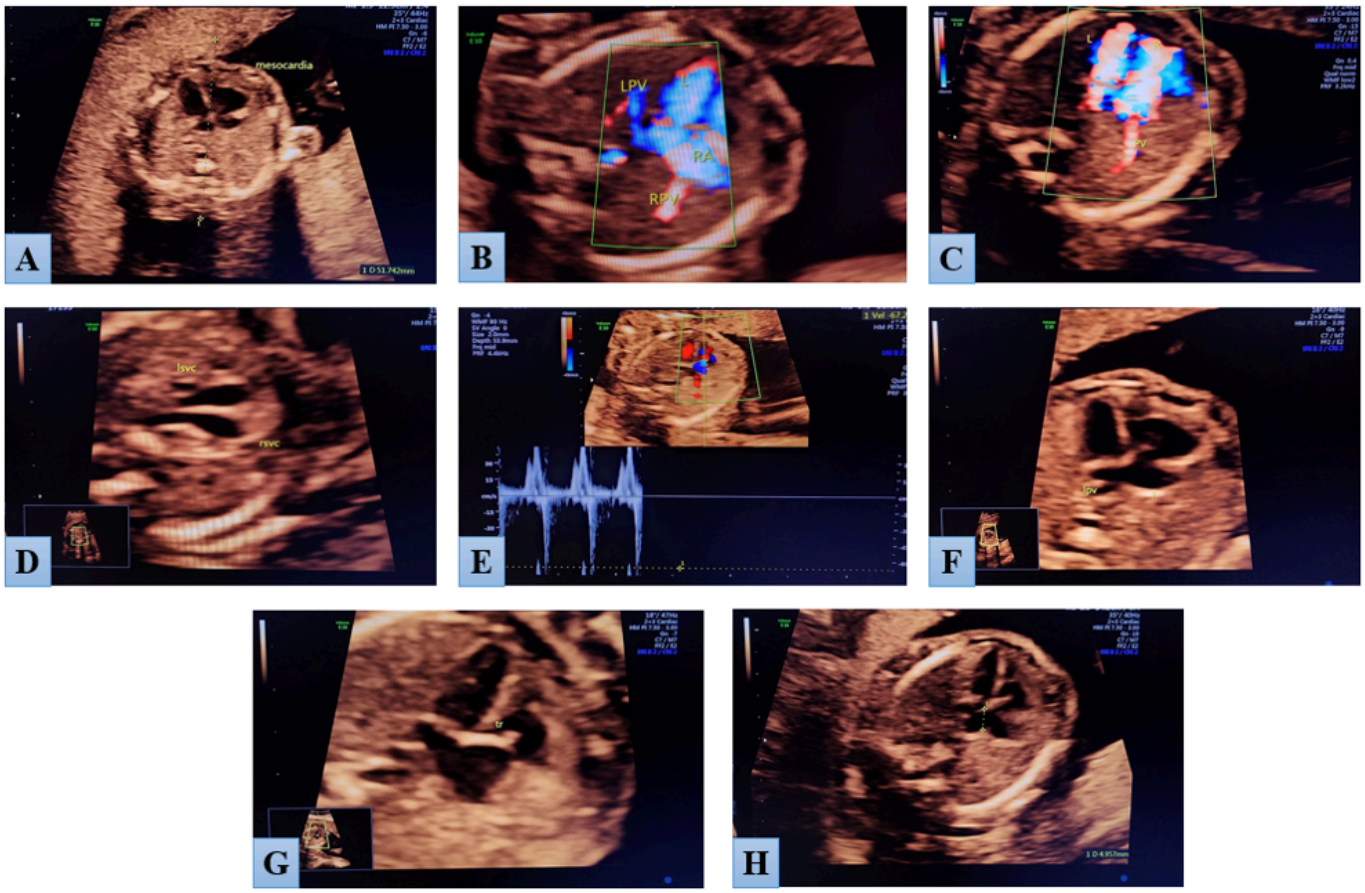


Figure 1

Fetal echocardiography. (A) Evidence of mesocardia is evident as the apex of the heart is located in the midline of the thorax, which is in the context of hypoplasia of the right lung. (B & C) Evidence of partial anomalous pulmonary venous return is seen as drainage of the two right pulmonary veins into the right atrium (by a venous collector to the junction of the IVC and the right atrium). (D) Double SVC is also seen with drainage of the left SVC into the coronary sinus and dilatation of the coronary sinus. (E, F & G) Mild-to-moderate tricuspid valve dysplasia is also seen as echogenic and relatively thick flaps with TR at a velocity of approximately 70 cm/s. (H) The fossa ovalis was also observed to be 5 mm in diameter and prominent.

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