

## Prenatal diagnosis of Scimitar syndrome: a series of six cases

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### Objective

Scimitar syndrome is characterized by hypoplasia of the right lung and abnormal pulmonary venous drainage to the inferior vena cava with an incidence of 2: 100,000 live-born infants. Abnormal arterial supply from the descending aorta to the hypoplastic lung is often present. Here we present six prenatally diagnosed cases of Scimitar syndrome.

### Methods

All cases with a prenatal diagnosis of Scimitar syndrome with or without associated malformations in an 18 year period (2000–2018) in two large tertiary referral centers (University of Bonn and University of Cologne, Germany) were retrospectively reviewed for intrauterine course and outcome.

### Results

We reviewed 6 pregnancies with 6 affected fetuses. All 6 fetuses presented with the typical hypoplasia of the right lung and abnormal pulmonary venous drainage to the inferior vena cava. Right-sided mediastinal shift was present at the time of referral in all cases. Despite intensive searches for an abnormal feeding vessel, systemic blood supply to the right lung could not be demonstrated prenatally in any of the cases.

### Conclusion

The primary pointer to prenatal diagnosis of Scimitar syndrome is the abnormal position of the heart in the chest. If searched for, abnormal venous drainage can be identified prenatally, but demonstration of the anomalous arterial supply is difficult. Therefore, whenever this diagnosis is considered, early postnatal work-up is important to outline the details of vascular anatomy and to proceed to interventional cardiac catheterization, if necessary.