# Scimitar Syndrome Patient Information

*Patient Information Series – What you should know, what you should ask.*

**What is Scimitar Syndrome?**
Scimitar Syndrome, also known as hypogenic lung syndrome or pulmonary venolobar syndrome, is a rare congenital abnormality affecting 1 – 3 in 100,000 babies born. It affects more females than males. In this syndrome, the right lung of the fetus is poorly developed and there is abnormal drainage of the lung vein. There may also be an abnormal blood supply to the abnormal lung from a major blood vessel called the aorta. The heart is predominantly shifted to the right chest.

**How does it happen?**
The specific cause of Scimitar syndrome is unknown but it is a developmental abnormality that occurs very early in fetal life. There are risk factors for congenital heart diseases, and these include genetic abnormalities, some medications and environmental factors like smoking in pregnancy.

**How are chromosomes relevant to Scimitar Syndrome?**
There is a very low risk of association with genetic and chromosomal associations unless associated with other structural abnormalities. A few isolated cases in families showed evidence of segmental loss in some chromosomes.

**Should I have more tests done?**
Your baby will need a detailed heart ultrasound (echocardiogram) to screen for other heart conditions which may be associated with Scimitar syndrome. Extensive ultrasound assessment for other associated abnormalities in other parts of the body of your baby will also be required. A fetal magnetic resonance imaging may be useful for the confirmation of the findings suspicious of Scimitar syndrome. There is a low association of this disorder with chromosomal syndromes however in the presence of additional structural problems your doctor may offer you genetic testing.

**What are the things to watch for during pregnancy?**
Scimitar syndrome usually does not result in significant consequences in the unborn baby except when it is associated with other complex heart abnormalities.

**What does it mean for my baby after it is born?**
The outcomes after a diagnosis of Scimitar syndrome generally depend on the presence of associated abnormalities, which might dictate the need for intervention. Only very rarely does the neonate with an isolated condition need an early intervention. Complications in infancy are generally related to the function of the lungs and the heart. Infants with Scimitar syndrome may exhibit lethargy, dusky colour (cyanosis), fast breathing (tachypnea,) poor eating, and failure to thrive. Patients who are only diagnosed later in infancy or as adults have a better prognosis than do babies with symptoms.

**Will it happen again?**
Scimitar syndrome has a very low risk of chromosomal abnormalities and is unlikely to recur in subsequent pregnancies.

**What other questions should I ask?**

- Can I carry this pregnancy to term?
- Do I have to deliver in a tertiary center?
- Will my baby require a surgery?
- What are the long-term outcomes for my baby like?

*Last updated February 2024*