Double Inlet Single Ventricle (DISV)

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

What is a double inlet single ventricle (DISV)?

The normal heart is divided into four chambers:

- Two atria (upper chambers): right and left.
- Two ventricles (lower cavities): right and left.

The two atria are separated by the atrial septum, which presents a small communication that closes after birth during the fetal period. The two ventricles are separated by the interventricular septum. Blood normally passes from the atria to the ventricles, then to the lungs and the rest of the body.

DISV is a form of severe congenital heart disease in which the two upper cardiac chambers (atria) connect totally or predominantly to a single lower chamber (ventricle).

How does a Double inlet single ventricle happen?

DISV is a rare congenital cardiac anomaly with an incidence of 0.05 to 0.1 per 1000 live births. It accounts for 1% of all congenital cardiac anomalies. It develops because there has been a failure in the division of the ventricles in early embryonic life. This defect's origin is unknown.

Why this anomaly is important?

While the baby is inside the uterus, there is no risk for the baby. However, after birth, the baby's oxygenated and non-oxygenated blood mix in the heart's single ventricle. Therefore, the blood that reaches the whole body is the blood that does not have the optimal oxygen content necessary for the proper development of a child.

Should I have more tests done?

- A detailed anatomical survey should be performed to assess whether this disease is isolated or associated with other anomalies.
- This cardiac defect is usually an isolated finding and is not often associated with chromosomal or genetic abnormalities. Although the risk of associated chromosomal abnormalities is low, the possibility of invasive genetic testing for the fetus should be considered. To carry out a fetal genetic study, an
- amniocentesis is performed, a technique where a needle is used to take some of the amniotic fluid from the womb for testing.
- Follow-up ultrasounds will be performed during pregnancy to assess the course of the disease.

What are the things to watch for during the pregnancy?

Fetuses with DISV may have associated heart defects that can develop throughout gestation, so serial ultrasound monitoring is recommended. The presence of this disease does not modify the usual obstetric management except when it is associated with heart failure.

Where should I deliver? Where will the baby receive the best care after it is born?

Delivery should be planned in a tertiary centre to receive adequate management with access to neonatology, cardiology, and pediatric cardiac surgery.

What does it mean for my baby after it is born?

The anatomical spectrum of this anomaly is very diverse; hence symptoms in the new-born may vary widely. However, the new-born usually has no symptoms in the absence of hypoplasia (small size) or significant obstruction of the main blood vessel leaving the heart (aorta) and/or pulmonary artery (artery supplying the lungs).

The prognosis is generally poor in this condition, as these patients are candidates for non-curative surgery, which consists of multiple surgeries in three stages- in the new-born period, in the first year of life and after one year of life. Despite significant surgical advances in recent decades, this type of cardiac disease continues to be associated with a high rate of complications and mortality, and cardiac transplantation is the only definitive treatment. The 10-year survival rate is around 70% and 80%.

Will it happen again?

As the cause of the DISV is unknown, the risk of recurrence is unknown at this time.

Last updated August 2023