

Single Cardiac Ventricle

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

What is a single cardiac ventricle?

A single cardiac ventricle is a congenital heart defect that occurs when a developing fetus has only one functional ventricle (a pumping chamber) in the heart instead of the usual two (a left ventricle and a right ventricle). This condition is also known as "single ventricle heart" or "single ventricle defect". In a typical heart, the left ventricle pumps oxygen-rich blood to the brain and body, while the right ventricle pumps oxygen-poor blood to the lungs for oxygenation. However, in a single ventricle defect, the heart is not fully formed with two separate ventricles, and one of the ventricles is underdeveloped or non-functional. The remaining functional ventricle may be the right or left ventricle. Single ventricle defect may occur in association with other cardiac defects such as atrio-ventricular canal defect, tricuspid atresia, mitral atresia, or pulmonary atresia with intact ventricular septum.

What causes single ventricle cardiac defect?

Single ventricle defects are complex and their causes are not always well understood. Genetics may play a role in some cases, for example, DiGeorge syndrome (22q11.2 deletion syndrome) or other chromosomal abnormalities, can affect the development of the heart and other organs and may be found in some cases of single ventricle defects. In many cases however, no cause can be identified.

Should I have more tests done?

In addition to your regular prenatal care and monitoring throughout the pregnancy to track the baby's growth and overall well-being, you might have fetal echocardiography, a specialized ultrasound of the heart that provides detailed images of its structure and function. Genetic counseling and testing can help to determine if there is a genetic component to your baby's condition. Another imaging test that provides detailed information about the structure and function of the fetal heart is Cardiac MRI. You might consult with a maternal-fetal medicine specialist, who specializes in high-risk pregnancies, and a pediatric cardiologist, who specializes in congenital heart disease, to advise you on your individual baby's condition, development, and prognosis.

What are things I should watch for during pregnancy?

Your healthcare team can counsel you on how to watch for any signs of complications during pregnancy, such as reduced fetal movements or changes in your health, as well as advise you regarding regular antenatal check-ups and fetal monitoring. Fetal ultrasound exams and specialized echocardiography can be used to monitor the baby's heart structure and function, blood flow, and overall development. You may be advised to plan for your delivery in a facility equipped to handle high-risk pregnancies and that has the necessary resources for immediate care of newborns with heart

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defects. This often includes having a neonatal intensive care unit (NICU) and pediatric cardiac surgery availability.

What does a diagnosis of single ventricle mean for my baby after it is born?

Every baby born with single cardiac ventricle is unique. This condition is a complex and serious congenital heart defect that requires medical intervention and surgical correction shortly after birth. Treatment typically involves a series of surgical procedures known as the Fontan procedure, which redirects blood flow in a way that allows the single ventricle to adequately oxygenate and pump blood to the body. The goal of these surgeries is to improve blood circulation and oxygenation, but lifelong medical management and monitoring are often required.

Will it happen again?

The risk of having another child with a single ventricle defect or any other congenital heart defect is influenced by multiple factors, and it can vary from one family to another. Genetic counseling and consultation with a pediatric cardiologist can provide valuable insights into your specific situation and help you make informed decisions about family planning and prenatal care.

What other questions should I ask?

- Where should I deliver my baby?
- What is the specific diagnosis and the severity of the single ventricle defect in my baby?
- Are there any additional tests or evaluations that need to be performed during the pregnancy to assess my baby's condition further?
- Can you explain the expected course of treatment and surgeries that my baby will need after birth?
- What are the potential complications and risks associated with this heart defect and its treatment?
- What are the long-term implications of this heart defect for my child's health, development,

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