

# Pulmonary Stenosis

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

## **What is Pulmonary Stenosis?**

Pulmonary stenosis is a congenital heart defect (present at birth) that develops abnormally during the first eight weeks of pregnancy. In a healthy heart, the right ventricle, one of the two pumping chambers of the heart, pumps blood into the pulmonary artery that takes blood to the lungs. The pulmonary valve is a one-way door located between the right ventricle and main pulmonary artery. The valve of the pulmonary artery is made up of three thin leaflets, and its job is to control the flow of blood from the heart to the main pulmonary artery.

Pulmonary stenosis occurs when there are problems with the pulmonary valve: two of the leaflets may be stuck or too thick, and, sometimes, there are fewer leaflets than usual. As a result, the pulmonary valve is small and narrow. The heart must work harder to pump enough blood into the body. Pulmonary stenosis can range from mild to severe. Narrowing at the valve is called pulmonary valve stenosis. When narrowing is below the valve, this is called subvalvular pulmonary stenosis. Narrowing above the valve is called supra-valvular pulmonary stenosis.

## **How does Pulmonary Stenosis happen?**

Pulmonary stenosis can be caused by many factors. It is commonly seen with other heart defects. The exact cause as to why this happens is not known. Most of the time this heart defect occurs by chance with no apparent reason for its development. However, sometimes it can have a genetic link and occur more often in certain families.

## **Should I have more tests done?**

Many women will choose to have more tests done to know more about the condition of the fetus. You should also ask if fetal echocardiography, a specialised ultrasound of the heart of the baby during the pregnancy is available or request a detailed fetal scan by a fetal medicine specialist. Additional testing includes an amniocentesis (where a thin needle is used to take some of the amniotic fluid in the womb) to look for chromosomal abnormalities, or other genetic tests, such as Chromosomal Microarray (CMA, or “chip”) or whole exome sequencing, which are advanced genetic tests to look more closely at the genetic make-up

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of the fetus. You might be able to consult with a Geneticist or Genetic Counselor, who are specialists in genetic conditions. In some cases of severe pulmonary stenosis that is causing problems with the development of the right ventricle (pumping chamber), a very specialised intervention, called fetal pulmonary valvuloplasty, may be performed on the fetus during the pregnancy. You can ask your caregiver whether your fetus may be a candidate for such a procedure.

## **What are things to watch out for during my pregnancy?**

Babies with congenital heart defects are at risk of problems during pregnancy. Specialists recommend regular ultrasounds to help identify swelling or fluid retention in or around the baby. This condition is called hydrops fetalis. A paediatric cardiologist will routinely look at the heart and evaluate the narrowing and the leaflets of the pulmonary valve with an ultrasound. Pulmonary stenosis can worsen over time, developing into a severe form requiring surgery. Knowing this in advance can help your doctor and the care team decide what delivery plan will be best for you and your baby.

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

A postnatal ultrasound of your baby's heart will be done to evaluate the pulmonary stenosis further, as your baby adjusts to life outside the womb. Mild cases of pulmonary stenosis often do not need any treatment and these babies are usually able to go home from the hospital with only outpatient follow-up. In cases of severe pulmonary stenosis, some babies will be very sick after delivery, requiring a stay in the NICU (neonatal intensive care unit). Treatment options such as a balloon valvuloplasty, where a thin tube with a balloon on the tip is inserted to dilate the valve in the heart, or surgery may be needed to repair the pulmonary valve. Supplemental oxygen and medications will be given to your baby as necessary to help provide sufficient oxygen to your baby's organs.

Overall outcomes for isolated pulmonary stenosis are excellent. Children should receive follow-up over time, into adulthood, to make sure the narrowing within the pulmonary artery does not get worse.

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## What other questions should I ask?

You might ask your caregiver these questions regarding your pregnancy:

- Does this look like severe pulmonary stenosis?
- How often will I have ultrasound examinations done?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet the team of doctors that will be assisting my baby when it is born, in advance of my delivery?

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