Aortic Stenosis

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

What is Aortic Stenosis?

Aortic stenosis is a congenital heart defect (present at birth) that develops abnormally during the first eight weeks of pregnancy. In a healthy heart, the left ventricle, one of the two pumping chambers of the heart, pumps blood in to the main body artery (aorta) that takes oxygenated blood to the different parts of the body.

The aortic valve is the valve between the left ventricle (one of the pumping chambers) of the heart called the aorta, which distributes the blood to the head and brain, and the rest of the body. Under some circumstances the aortic valve becomes narrower than normal, restricting the flow of blood coming out of the left pumping chamber. This is known as Aortic Valve Stenosis.

How does Aortic Stenosis happen?

Significant aortic stenosis is relatively uncommon, affecting about 6 in every 1000 babies born. It can occur alone or in combination with other heart abnormalities. Aortic stenosis occurs due to improper development of the aortic valve during the early part of fetal growth. The normal aortic valve has three thin and flexible leaflets. In cases of aortic stenosis, the valve leaflets are thickened or become less pliable and fuse together. Most commonly, the abnormality occurs when the aortic valve has two instead of three leaflets (bicuspid aortic valve). 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”) which looks more closely at the genetic make-up of the fetus. You might be able to consult with a Geneticist or Genetic Counselor, who are specialists in genetic conditions.

What are the things to watch for during the pregnancy?

Babies with aortic stenosis require frequent ultrasound by a fetal medicine specialist and a paediatric cardiologist to monitor the progression of the condition. When the aortic valve does not open normally, the muscles of the left ventricle have to work harder to pump blood into the aorta. As the disease progresses, the valve obstruction becomes significant. To compensate for this increased workload the muscles of the left ventricle gradually thickens. If the valve is severely obstructed, the muscles of the left ventricle may not be able to compensate and may fail to pump blood into the aorta. In some cases, with progression, the left ventricle becomes

small and non-functioning. Knowing these findings in advance can help your doctor and the care team to decide what delivery plan will be best for you and your baby. One in 10 babies with aortic stenosis is born with significant narrowing that requires emergency treatment at birth.

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

A baby with aortic stenosis should be delivered in a tertiary center with neonatal intensive care, paediatric cardiology, and paediatric cardiothoracic facilities. This allows coordination of care and permits access to emergency services in the immediate newborn period if needed. Vaginal delivery is usually recommended and caesarean section is reserved for obstetric indications.
Depending on the severity of the baby's condition, the paediatric cardiologist will make a decision what treatment baby would require after birth. Some of these possible treatment pathways would have been discussed with you during pregnancy. Treatment would depend on the size and function of the left pumping chamber. In some cases, the baby might require a small operation to relieve the obstruction. If the left pumping chamber is very small in size then the baby would

require a series of operations in order to increase the blood flow to the body and bypass the poorly functioning left pumping chamber.
Overall outcomes for isolated aortic 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.

Will it happen again?

There is a slightly higher chance of this condition happening again in the next pregnancy. Genetic studies have shown that there is a 13%-15% recurrence risk of aortic stenosis in a baby if the mother is affected, a 5% risk if the father is affected, and a 2% risk if one child in the family is affected. When the heart defect recurs in another child, it is not always the same heart defect: it may be something minor or more severe.

In your next pregnancy, you can benefit from a detailed fetal heart examination early in the pregnancy (if facilities are available) or at the time of a routine fetal anomaly scan at 19-20 weeks.

What other questions should I ask?

- Does the baby's heart look normal?
- How often will I have ultrasound examinations done?
- What is the severity of the aortic stenosis?
- What are the chances of this condition to progress in the course of pregnancy?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can you give an indication of the duration of the hospital stay after surgery?
- Can I meet in advance with the team of doctors that will be looking after my baby when it is born?

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