Persistent Left Superior Vena Cava (PLSVC)

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

What is a Persistent Left Superior Vena Cava (PLSVC)?

Persistent left superior vena cava (PLSVC) is diagnosed when the large vein returning blood from the upper body to the right side of the heart, the superior vena cava, is seen on the left instead of the right side. In some individuals two vessels may be seen, on the right and left, while in others only the left one is present. PLSVC occurs in about 1 in 300 babies. Many people with this variant vessel are unaware of it. In some cases, however, PLSVC may occur with other congenital heart defects, such as a ventricular septal defect or coarctation of the aorta, or others.

How does a PLSVC happen?

The PLSVC is termed “persistent” because it originates from an embryonic vessel that normally shrinks and disappears during the first weeks of development in the womb. However, for reasons that are not fully understood, in some fetuses the left SVC remains or “persists”. PLSVC may be isolated, that is no other anatomic defects are present, but almost half of the babies with a PLSVC will have other problems, most often with their heart. Seven out of 100 have a change in the number of chromosomes. Chromosomes are where most of our genetic information is kept. We usually have 46: 23 come from one parent and 23 from the other parent. They are matched in pairs. As an example, people with Down syndrome have an extra chromosome number 21.

Should I have more tests done?

Many women will choose to have more tests done to know more about the condition of the baby. The tests available depend on where you are. Tests to ask about include an amniocentesis (where a needle is used to take some of the amniotic fluid from the womb) to look for problems with the number of chromosomes and some of the problems within the chromosomes. You should also ask if a fetal echocardiography, a specialised ultrasound of the heart of the baby during the pregnancy, can be done. You should also request a detailed fetal scan by a fetal medicine specialist.

What are the things to watch for during the pregnancy?

Babies with PLSVC are at risk of some problems during the pregnancy. That is why most specialists will recommend regular ultrasound examinations. The ultrasound will help identify if the baby’s heart is developing toward coarctation of the aorta. This is a narrowing within the main arterial vessel carrying blood from the heart.

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

The biggest problem for babies with PLSVC is the development of aortic narrowing. The baby should undergo an ultrasound scan in order to assess the progress of narrowing occurring.

Babies who also have a problem within the chromosomes or within the heart may have other difficulties after birth, depending on their individual diagnosis. When the baby is grown up, isolated PLSVC will not carry any problem at all.

Will it happen again?

When no other genetic reason is found to explain the PLSVC, the risk of this happening again is extremely low. If there is a genetic reason, then the risk depends on the risk of recurrence of that diagnosis. Consultation with a specialist may be helpful to sort this out.

What other questions should I ask?

- Does the baby's heart look normal?
- How often should I have ultrasound examinations done?
- Is coarctation of the aorta during the pregnancy predictable?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet in advance the team of doctors that will be looking after my baby when it is born?

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