

COARCTATION OF THE AORTA

What is coarctation of the aorta?

Coarctation of the aorta is a congenital heart defect. It is a narrowing in the aorta, which is the large blood vessel that delivers blood from the heart to the rest of the body. The aorta delivers blood to the upper body, then curves like a candy cane and dives down to the lower body. The narrowing in coarctation occurs in the area after the blood vessels go to the upper body and before the aorta goes down to the lower body.

What causes coarctation of the aorta?

We do not know exactly why patients develop coarctation of the aorta. It can be found in patients with genetic syndromes, such as Turner syndrome. It can also be seen in patients who have more complex heart disease. The most common abnormality seen with coarctation is a bicuspid aortic valve, or an abnormality of the valve between the left ventricle (pumping chamber) and the aorta. Bicuspid means that there are 2 leaflets of the valve instead of the normal 3 leaflets. Patients with coarctation may have relatives with similar heart defects.

How is coarctation diagnosed?

The most common method of diagnosis is an echocardiogram (ultrasound of the heart). Coarctation can be a difficult diagnosis before birth because of differences in the blood flow patterns. There may be some findings on a fetal echocardiogram which suggest that there may be a coarctation (right side of the heart is larger than the left, aorta is smaller than the pulmonary artery), but sometimes we can only tell for sure with an echocardiogram after the baby is born.

How will your pregnancy be managed?

If your baby is diagnosed with coarctation of aorta, a high-risk obstetrician will participate your obstetric care. Overall care should be transferred to a specialized center, where multidisciplinary care is available. The team should include a perinatologist, fetal and pediatric cardiologists, a genetic counselor, a neonatologist and a pediatric cardiac surgeon. The fetal well being will be followed closely by fetal ultrasound and nonstress tests. Towards the end of pregnancy, visits may be as often as two to three times a week. If there is no specific maternal or fetal reason for a C-section, vaginal delivery is often possible. Induction of labor is often scheduled for pregnancies affected with coarctation of the aorta to make sure that all of the team members are available at the time of delivery.

Why does coarctation make babies sick?

The narrowing in the aorta limits the amount of blood that can get to the lower body. Before birth, there is a connection that allows blood to bypass the area of narrowing. This connection is between the pulmonary artery (artery to the lungs) and the aorta; it is called a patent ductus arteriosus (PDA). Every fetus has a PDA, and it typically closes by itself in the first few days of life. When there is a coarctation, the narrowing will worsen when the PDA closes. The heart will pump hard to push the blood through the narrowed area, but eventually it becomes too difficult for the heart to overcome. At that time, babies will start to breathe fast, have difficulty feeding and become cool and pale in their legs. If nothing is done, babies can become very sick.

What can I expect after my baby is born?

Most babies do very well right after birth. The PDA is still open, so they are usually able to get enough blood to their lower body and don't have difficulty breathing. Your baby will be brought to the neonatal intensive care unit. Depending on how the baby is doing, he or she may need some large IV's. An echocardiogram will be done to confirm the diagnosis. Much like before babies are born, it may be difficult to tell for sure if there is a coarctation while the PDA is still open. Those babies may need to be monitored closely as the PDA closes. Other times it is very obvious that there is a coarctation. Once the diagnosis has been confirmed, the baby will be started on a medicine called prostaglandins. This will keep the PDA open while the baby is transitioning.

In some cases there may be a coarctation which is only a mild narrowing. Those babies may not need surgery right away. They would be able to go home and be followed closely by a cardiologist. Because the care for children with congenital heart disease is complex, your baby will be cared for by a team of skilled clinicians. This starts before birth with maternal-fetal medicine and fetal cardiologists and continues with pediatric cardiologists and nurse practitioners, a pediatric cardiac surgeon, neonatal and pediatric intensive care physicians, pediatric anesthesiologists, pediatric cardiac operating room staff, pediatric nurses and many others.

What is the treatment/surgery for coarctation?

Although prostaglandins keep the PDA open, this medicine is not a good long term solution. The baby will need surgery within the first week or so of life. The surgeon will cut out the narrowed area and sew the aorta back together. Babies typically need about 7-10 days in the hospital to recover from the procedure.

What other procedures or follow up will my baby need?

Because the surgery leaves an area of scar on the blood vessel, it is possible for this area to narrow again in the future. Less than 10% of patients need to have another procedure. Many times when there is a narrowing in the aorta, it can be fixed in the cardiac catheterization laboratory. This means that the narrowing may be fixed with a balloon or stent (metal mesh that holds open a narrowed area) instead of surgery.

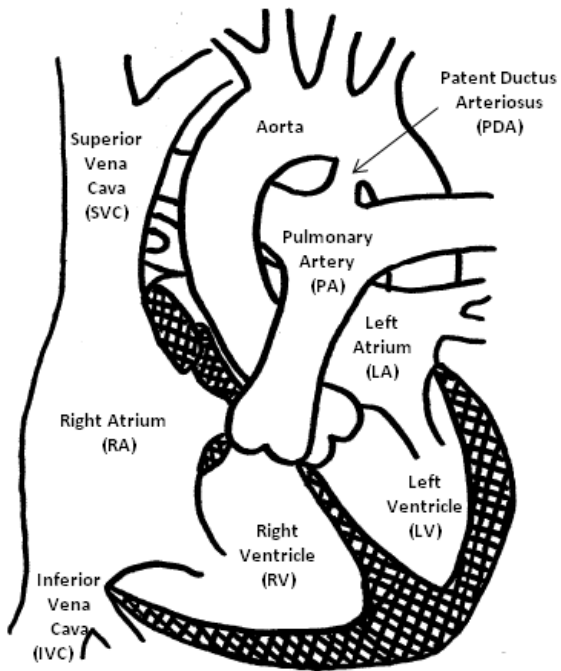
Infants after surgery may need more frequent follow up, but as children grow, they only need to be followed yearly. They will be monitored with echocardiograms, electrocardiograms (EKG's) and sometimes CT scans or cardiac MRIs.

What is the long term prognosis for coarctation?

The long term prognosis for coarctation is very good. The survival after surgery is very high. Children are allowed to participate in normal activities, including sports. Some patients with coarctation develop high blood pressure and may need medications to control it. Patients who also have a bicuspid aortic valve may have complications from the valve abnormality.

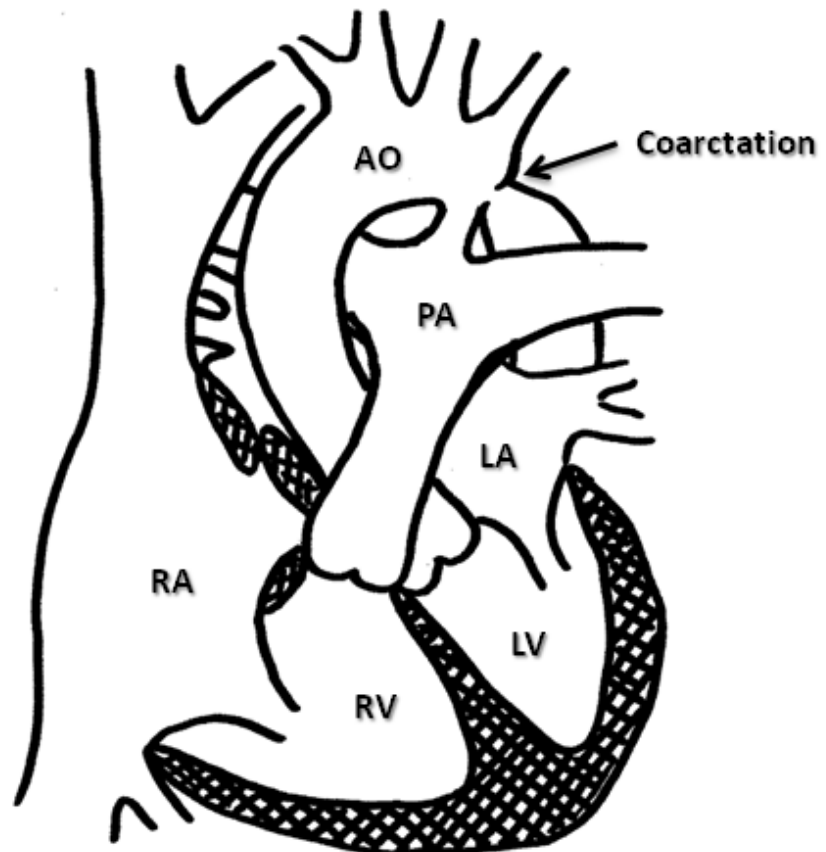
If I have one baby with coarctation of the aorta, are my future children more likely to have coarctation?

If your baby's coarctation is related to a chromosome abnormality (like Turner's syndrome) or a genetic syndrome, a genetic counselor can tell you what the chances are that a future pregnancy would have the same condition. Studies show that when coarctation of the aorta is not associated with an underlying genetic problem, the chance that future children will have any heart defect is about 2%. In future pregnancies, nuchal translucency ultrasound (at the end of the first trimester), targeted anatomy ultrasound (between 18-20 weeks) and fetal echocardiography are recommended.

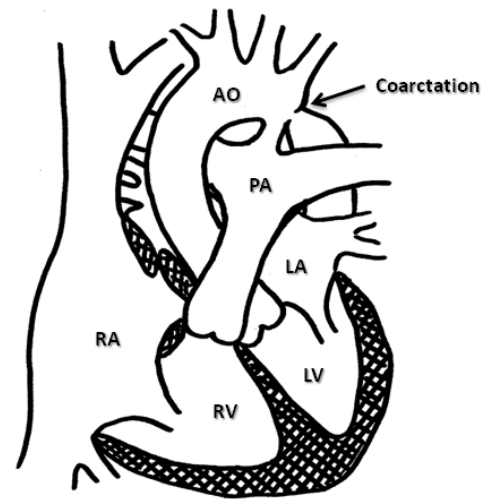


Normal Newborn Heart

Coarctation of the Aorta



Before Surgery



After surgery

