Vascular Rings

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Vascular rings are developmental anomalies that affect the aortic arch and the major blood vessels that arise from it. The aorta is the biggest artery (blood vessel) that arises from the heart. Usually, these problems cause compression of the trachea (windpipe) and the esophagus or both. Sometimes these blood vessel abnormalities form a complete ring around the trachea and esophagus, and in other cases the ring may be incomplete but still be enough to cause symptoms. There are many types of vascular rings.

During normal development there are two arches to the aorta as it leaves the heart, and the right arch usually goes away. When it does not, this leaves a double aortic arch and if only parts of either of the two arches don't regress as they are supposed to, many of the various types of ring anomalies arise.

Other types of vascular rings include pulmonary artery slings. This occurs when the left pulmonary artery (blood vessel from the heart to the lung) originates from the right pulmonary artery instead of the main pulmonary artery as it comes out of the heart. The abnormal left pulmonary artery passes between the trachea and esophagus and causes compression.

Signs and Symptoms - "What symptoms will my child have?"

Early signs/symptoms: Vascular rings usually cause symptoms because the abnormal blood vessels press on the trachea, esophagus or both and cause narrowing and partial blockage of that structure. This narrowing may cause problems breathing or swallowing. A child may stop breathing (apnea), have wheezing if the airway is compressed, or problems with vomiting or persistent spitting up if the esophagus is involved. Some infants may have recurrent respiratory infections. Other problems include difficulty feeding, failure to gain weight and grow.

Later signs/symptoms: Sometimes, an infant will be fine when taking only formula and liquids and then have problems with swallowing when more solid food is introduced. Most of the time, symptoms occur sometime in the first year of life, often in the first several weeks or months. Surgical correction may be needed at that time. Double aortic arches can cause symptoms early on. Some children with mild symptoms may outgrow the symptoms.

Some children may never have symptoms only to become symptomatic as adults when they have hardening of the arteries (atherosclerosis) and symptoms become more apparent.

Diagnosis - "What tests are done to find out what my child has?"

A variety of studies may be used to make the diagnosis of a vascular ring.

Esophagram: An esophagram is a test where the patient is made to drink liquid contrast with a X-ray taken during the swallowing process looking for changes in the shape of the esophagus that suggest a vascular ring.

Ultrasound: is used to look at the urachus. Sound waves are used to image the urachus or its remnants. Ultrasound is accurate for diagnosis in over 90% of cases.

Magnetic resonance Imaging (MRI): uses a catheter to instill dye to see a contrast look at the blood vessels themselves to make the diagnosis. This test uses a magnet, radio waves, and computer to obtain images of organs in the body. MRI does not use radiation

Computer tomography (CT scan): Detailed pictures of the chest reconstructed in different views to get a better picture of blood vessels and the heart.

Bronchoscopy: This study requires anesthesia. A telescope is placed through the vocal cords to look at the windpipe and the airways.

Cardiac catheterization: may be necessary if the anatomy of the vessels are difficult to show on the other imaging modalities, then direct injection of contrast into the heart will show the flow of blood from the heart to the blood vessels.

Breathing and swallowing problems can be caused by a wide variety of other problems most of which are more common than vascular rings. Making the diagnosis of a vascular ring requires someone to think of the problem as a potential cause of the child's symptoms and then order the appropriate tests.

Treatment - "What will be done to make my child better?"

Surgery: is the only way to take care of a vascular ring and is indicated for any patient who has one and is symptomatic. Failure to treat the ring can lead to serious problems including inability to eat well, causing weight loss or poor weight gain. Respiratory symptoms may be as severe as apnea (not breathing for a short duration) or hypoxia (lack of adequate oxygen). Sudden death has been reported. The type of surgery needed depends on the vascular ring diagnosed and to some extent the preference and experience of the surgeon. Many of these require an open thoracotomy (incision in the chest) between the ribs. However, there are several types that are amenable to thoracoscopic repair. In thoracoscopy, several small cuts (incisions) are made.

Through one of the cuts, a video camera is placed. The surgery itself is done using small instruments placed through the other incisions. The usual number of incisions (cuts) for thoracoscopic surgery vary.

Risks / Benefits:

Surgery is very safe when performed by an experienced surgeon and surgical team. Some of the risks include injury to the nerves that control the vocal cords (voice box). These nerves are specifically looked for during surgery but may have to be moved a bit in order to take care of the vascular ring. This can cause a soft voice or cry in babies after surgery, but this will get better in several weeks, if it happens. Another possible problem is a lymph leak in the chest after surgery. This is due to drainage of clear fluid from lymphatic ducts in the chest. While this is not common, it can be a difficult problem to take care of and involve dietary measures, medicine, or even further surgery.

Preoperative preparation: Nothing to eat or drink for eight hours prior to surgery. If the operation is a scheduled (not emergency), a bath or shower the night prior or the morning of the procedure is recommended.

Home Care - "What do I need to do once my child goes home?"

The care at home will, to some extent, depend on the type of surgery that was needed and the age of the child when the surgery was done.

Activity:

- For repairs done thoracoscopically, children can return to normal activity as soon as they are able.
- For open thoracotomy, activity may be more limited in older children as their wound heals and pain medicine may be needed longer.

Diet: There is no special diet.

Medicines:

- Pain medication may be needed for only a short period of time.
- If narcotics are used, constipation may occur. Stool softeners should be used if narcotics are being taken.
- Acetaminophen (Tylenol®) or Ibuprofen (Motrin® or Advil®) may also be used for mild to moderate pain

When to call your doctor: Fevers, trouble breathing, redness of the wound, drainage from the wound.

Follow-up: Your surgeon will need to see your child in clinic a few weeks after surgery to make sure the wound is healing well.

Long Term Outcomes - "Are there future conditions to worry about?"

Sometimes, respiratory symptoms take months to resolve completely because the trachea was narrowed by the vessels and, in a sense, molded to the narrowed shape. Reshaping of the trachea requires growth with time to resolve the narrowing. Sometimes even swallowing problems take some time to fully resolve.

When corrected, vascular rings do not shorten life span, and children should make a full recovery with excellent long-term function.

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