Anomalous Pulmonary Venous Connection (APVC)

The aim of this information sheet is to explain what APVC is, what effect it will have on a child and how it can be treated.

What is APVC?

Anomalous means 'not normal'. Pulmonary means of the lungs and venous 'means of the veins'. Anomalous Pulmonary Venous Connection means some or all of the veins bringing red blood from the lungs to the heart are not connected into the left atrium.

APVC is also called:

- APVD - Anomalous Pulmonary Venous Drainage
- APVR - Anomalous Pulmonary Venous Return
- If none of the veins connect into the left atrium, the condition is referred to as Total – i.e. TAPVC
- If some of the veins connect into the left atrium,the condition is referred to as Partial – i.e. PAPVC

Instead of flowing into the left atrium, the veins bringing the blood from the lungs may be connecting:

- Into the Superior Vena Cava – the large vein that brings blue blood back to the heart from the upperbody. (Supracardiac)
- Into the Coronary Sinus – the vein that takes blueblood from the heart muscle itself into the right atrium (Intracardiac)
- Into the Inferior Vena Cava – the vein that brings blue blood back to the heart from the lower body (Infracardiac).

Wherever the veins are connecting the result is that there is a shortage of oxygenated blood for the body, and a bigger flow of oxygenated blood to the lungs.

Children suffering from APVC normally also have an Atrial Septal Defect (a hole in the wall between the right and left atriums- which is normally a problem). The hole allows some oxygenated blood from the right atrium to flow into the left atrium. The resulting mix of oxygenated and deoxygenated blood then flows into the left ventricle and the aorta. The bigger the Atrial Septal Defect the more oxygenated blood can get through and thehealthier the child will be.

Scimitar Syndrome is a form of PAPVC. If your child has this condition, then his or her right lung may be underdeveloped (hypoplasia) and the pulmonary vein from that lung will connect up to the inferior vena cava. His or her heart may be on the right of the chest (dextrocardia), in the space that the lung would normally fill.

Diagnosis

Babies with APVC may be diagnosed before they are born, during ante-natal scans. However, APVC is a difficult condition to detect before birth.

Most babies are diagnosed with APVC after birth. Symptoms include breathlessness and turning a bluish colour. They may also have problems feeding and may be slow to put on weight. Frequent chest infections are another symptom and the sound of the extra blood being pushed towards the lungs and leaking through the valve can be heard as a heart murmur.

If your baby has Partial APVC there may not be any symptoms early in life, other than a heart murmur. When a heart disorder is suspected the tests used can be:

- pulse, blood pressure, temperature, and number of breaths a baby takes a minute
- listening with a stethoscope for changes in the heart sounds
- an oxygen saturation monitor to see how much oxygen is getting into the blood
- a chest x-ray to see the size and position of the heart
- an ECG (electrocardiogram) to check the electrical activity
- an ultrasound scan (echocardiogram) to see how the blood moves through the heart
- checks for chemical balance in blood and urine
- a catheter or Magnetic Resonance Imaging test may be needed.

Treatment

The treatment a child needs will depend on the kind of APVC they have.

If the veins are not allowing blood to move freely into the right atrium (obstructed) surgery will be needed at the first opportunity.

Surgery is almost always needed at some point, but can be a few weeks after diagnosis.

About this document:
Published: June 2013
Reviewed: Feb 2024

During open heart surgery – the heart will need to be stopped and opened to repair it. A heart-lung bypass machine will have to take over the job that the heart normally does.

The aim of the operation is to make the circulation of blood through the heart and lungs normal, so a patch is put over the hole between the right and left atrium and the veins are connected to the left atrium.

For most children this surgery is low risk, but it will depend on how well your child is otherwise. The cardiologist or surgeon should discuss the risks with you in detail before asking you to consent to the operation.

The length of time in hospital after surgery will usually be 10 to 12 days, of which one or two will be spent in the intensive care and high dependency unit. Of course this depends on how well your child is before and after the surgery, and whether any complications arise.

A common complication is pulmonary hypertension which can be detected at the time of surgery or may occur much later.

How it affects your child

If the surgery is straightforward, and your baby does not have other health problems, he or she should be completely well shortly after surgery. There will be a scar down the middle of the chest, and there may be small scars where drain tubes were used. These fade very rapidly in most children, but they will not go altogether. Smaller scars on the hands and neck usually fade away to nothing.

Most parents are amazed at how quickly their baby recovers from surgery and starts to gain weight. In some cases though, the child may need to be on ventilation for as long as it takes for their lungs to recover. There is also the risk that scar tissue obstructs the veins.

Children with heart conditions are more likely to have an infection called endocarditis (see Endocarditis information sheet).

Evidence and sources of information for this CHF information sheet can be obtained at:

National Institute for Health & Care Excellence. NICE Guidance CG64. London: NICE; 2017. Available at: www.nice.org.uk/guidance/cg64

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