

What is An A-V Canal Defect?

An A-V Canal Defect is a family of defects of the heart related to a malformation of the center of the heart. It is a very primitive defect, meaning that it occurs early in the development of the heart. It may be represented by something as simple as a small cleft (opening) in the Mitral valve to the complete absence of the center of the heart. It may be associated with a hole between the upper chambers of the heart (ASD), a hole between the lower chambers (VSD), or both. Usually these defects will need to be surgically repaired. This type of defect is usually silent when a baby is first born and may not be noticed for several weeks. Most of the time these children will have an abnormal EKG.

The most common form is called a "complete A-V canal defect". This includes an ASD and VSD, also the mitral and tricuspid share common tissue (normally they are two separate valves). The result of this is that one or both of the valves leak. This causes the heart to work less efficiently, and often will lead to heart failure requiring medical therapy with digoxin, Captopril and Lasix. Parents will sometimes recognize heart failure because the baby may breathe rapidly, may sweat a lot and may not feed as well as before. They also tend to not gain weight adequately. This type of defect is seen in about 25% of children with Down's Syndrome. It can also occur in normal children as well, and tends to be more complex in them. On occasion one of the ventricles (pumping chambers) may be bigger than the other and this can make the repair more complex. Coarctation of the Aorta is sometimes seen with this as well.

All of these defects will need to be surgically repaired, as they are almost always lethal. We normally like to have this done sometime between the 3rd and 10th month of life. The reason for this is that most of these children develop heart failure early in life and their growth is somewhat stunted. Also, there is a higher risk of damage to the arteries of the lungs if repair is delayed beyond one year of life. This is because the lung arteries (Pulmonary Arteries) are exposed to a much greater than normal amount of blood flow and higher pressures. On occasion it may not be possible to repair the Mitral valve and it may need to be replaced with a mechanical (artificial) valve. When this happens the child will need to be on a blood thinner (coumadin) for life. Sometimes it may be necessary to replace the artificial valve again as the child becomes larger depending on the size of the original valve. For the most part these children can do well and have active and near normal lives assuming the repair goes well.

Sometimes there may just be a hole between the upper chambers of the heart (ASD) associated with a cleft (or hole) in the Mitral valve. This is called a Primum ASD. These will usually need to be surgically repaired but this can be delayed for 2 - 3 years because the pressure in the lungs are low and there is less chance of permanent damage. With these children the mitral valve can usually be repaired and do not require replacement. It is unusual for these children to develop heart failure and require medication. These defects are usually picked up by the doctor as a heart murmur during the latter part of the first year of life. The good news is that these children usually do well after surgery and can lead normal lives. All children who have an A-V Canal defect will need to have SBE prophylaxis for dental work and surgery to prevent an infection of the heart. If you have any questions, please ask one of the doctors.

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