

## Laterality Disturbance and Heart Defects

The information which is used to plan our bodies is carried by our genes, half of which we inherit from our mother and half from our father. This hereditary information can carry errors which can ultimately lead to errors in the way parts of our body, such as the heart, develop.

### **What does laterality disturbance mean?**

#### **We should not be symmetrical**

Although our bodies are mostly symmetrical, several of our internal organs are not: for example, the **heart** is on the centre-left of the chest, one side pumps blood to the lungs and the other to the body, and the **stomach** is on the left, the **liver** is on the right, and the spleen on the left.

Very early in pregnancy - about the fourth or fifth week - the cells which make our various organs, receive signals which program them to produce structures for the left or right side of the body. If the cells are programmed incorrectly the body's left-right plan or 'laterality' is disturbed.

#### **Mirror image**

About one of every ten thousand people has a mirror image arrangement, with the heart, spleen and stomach on the right and the liver on the left side. This is called situs inversus and results in normal relationships between the left-right positions of the organs - for example the arteries carrying red blood from the heart will travel towards and reach the correct organ.

#### **Symmetrical**

Occasionally some or all of the organs are symmetrical - this is called 'heterotaxy'. The most extreme form of heterotaxy is 'isomerism' (pronounced eyes-om-er-ism) where two right sides or two left sides develop.

### **Why can laterality disturbance cause heart defects?**

#### **Asymmetrical heart**

Because the normal heart is not symmetrical, laterality disturbance may affect its development. The two sides of the heart differ in structure, and in function. The right side of the heart pumps blood to the lungs to pick up oxygen whereas the left side pumps oxygenated blood around the body.

The heart develops from cells which come from both the left and right sides of the body. These cells join together to form a single tube, and if these cells

have not been programmed correctly the development of the single tube into an organ with four chambers may go wrong.

Only 3% of congenital (born with) heart defects are associated with laterality disturbance. A confusing number of names are used to describe this condition: isomerism sequence, asplenia syndrome, Ivemark syndrome, polysplenia syndrome, situs ambiguous, heterotaxia, partial situs inversus and laterality sequence!

The particular type and severity of heart defect that can result will depend on the particular laterality disturbance condition, and will vary from child to child.

In right isomerism sequence - sometimes called asplenia syndrome, Ivemark syndrome, or right atrial isomerism - the right side structures of the heart are duplicated on the left.

The heart defects which result are usually severe and complex, and may be picked up on a scan during pregnancy. The commonest abnormality is atrioventricular septal defect (AVSD) - a hole between the two top chambers of the heart, a single valve between the two top and two bottom chambers, and a hole between the bottom chambers.

The AVSD may be combined with a single ventricle (pumping chamber), the blood vessels may not be correctly connected to the heart (transposition of the great arteries and anomalous pulmonary venous drainage). Because these children have no left-side organs they have no spleen (asplenia). This leaves them susceptible to meningitis and pneumonia, and they need to take antibiotics.

In left isomerism sequence (also known as polysplenia syndrome) the left side structures are duplicated. The heart defects are less severe than for right isomerism. The commonest are complete heart block, resulting in a slow heart beat, atrial septal defect (a hole between the top two chambers) and AVSD. The child may have multiple small spleens - polysplenia) but this will not affect his or her health.

In either of these conditions, twisting of the bowel (intestinal obstruction) occurs in around 10% of children. This is caused by incorrect positioning of the intestines. About one in ten children will have other congenital abnormalities such as a kidney problem, or a spinal curvature.

### **The chances of a second affected child**

Most cases of laterality disturbance occur as a one-off, but a small proportion of cases is inherited. For a couple with an affected child there is a 5%, or 1 in 20, risk of it happening again.

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Please contact CHF if you have suggested amendments or changes as we like to keep our information sheets relevant and up-to-date.

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