



Laterality Disturbances and Heart Defects

This information sheet summarises what a Laterality Disturbance is and how it can cause congenital heart defects

What does laterality disturbance mean?

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 genetic information can carry errors which can ultimately lead to errors in the way parts of our body, such as the heart, develop. In this instance, to disrupt the body's normal left and right layout of internal organs.

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.

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). Rarely these children have malformed or absent left-side organs and may 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 usually 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 10% of 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 oneoff, but in a small proportion of cases it's inherited.

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

(1) BMC Medical Genetics BMC series 2017 18:77 *Functional study of DAND5 variant in patients with Congenital Heart Disease and laterality defects.* Fernando Cristo, et al. Available at:

<https://doi.org/10.1186/s12881-017-0444-1>

(2) European Heart Journal. EHJ314 2017 Familial co-occurrence of congenital heart defects follows distinct patterns. Sabrina G. Ellesøe, et al. Available at:

<https://doi.org/10.1093/eurheartj/ehx314>

(3) Contact A Family. Ivemark Syndrome. Contact A Family. 2017. Available at:

www.cafamily.org.uk

<https://www.cafamily.org.uk/medical-information/conditions/i/ivemark-syndrome/>

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