

Prenatal ultrasound scans showing isomerism: information for families

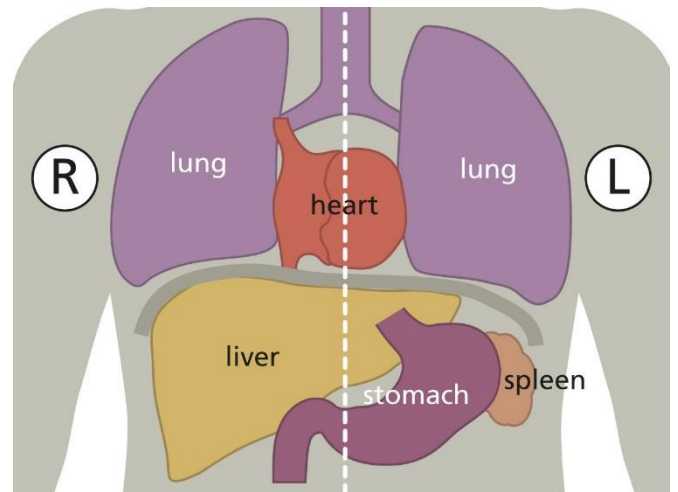
Isomerism is a congenital (present at birth) disorder where organs develop on the ‘wrong’ or opposite side of the body to usual. This information sheet from Great Ormond Street Hospital (GOSH) explains about isomerism detected during a prenatal ultrasound scan and what this might mean for your child. It will support the information discussed with you by your doctor and nurse at your appointment and it is important to remember that every case is slightly different.

You have been given this information sheet as the scan has shown that your baby has some features suggestive of an underlying diagnosis of right or left isomerism. In addition to the detailed report from your doctor, this information sheet gives general information to help you understand the other features that can be seen when this diagnosis is made.

What is isomerism?

The human body is not symmetrical. In early development, there are certain structures and organs that develop on one side rather than the other. For example, normally the heart and stomach are on the left and the liver is more to the right.

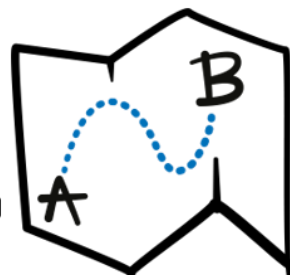
In isomerism, an abnormality occurs very early in the development of the embryo where there is ‘confusion’ over whether structures develop on the left or the right. This may result in some of the organs being on the opposite side to usual.

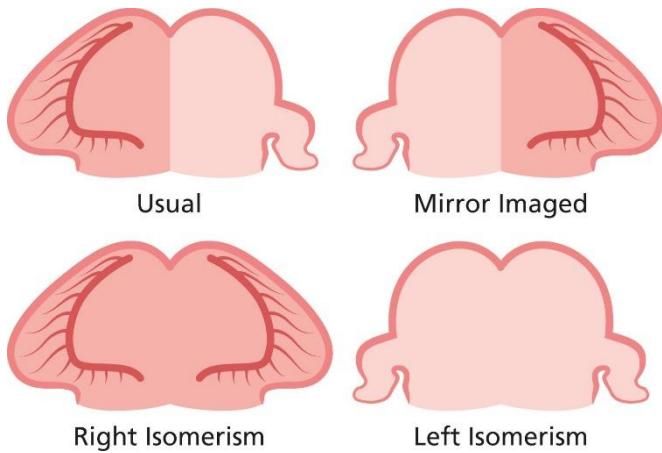


Abnormalities within the heart in isomerism

Normally in the heart, there are two filling chambers (atria) at the top of the heart. The left and the right atria are not identical – they each have a characteristic appearance and features.

In isomerism there are either two copies of a left atrium (left isomerism), or two copies of a right atrium (right isomerism).





In some cases, all the rest of the heart structures are normal, but often with isomerism there are also abnormalities in the way the chambers and vessels join together and abnormalities in the size of chambers.

The details and severity of your baby's heart abnormality will have been discussed with you by your doctor and be written in your report from clinic.

In some cases of left isomerism, the heart beat is abnormally slow (heart block). If the slow heart beat is tolerated during pregnancy then this may require a procedure to place a pacemaker during childhood. If it is very slow, the baby may become unwell in the womb and develop a condition called hydrops where fluid collects within the baby due to poor heart function.

Abnormalities of other organs

As well as the abnormal atria in the heart, isomerism can also affect other areas of the body.

Increased risk of twisting (malrotation) of the bowel

The abnormal position of the bowel increases the risk of twisting, which can result in the baby having features of bowel obstruction before or after birth. If this occurs surgery is needed to untwist the bowel and fix it in position. It may be recommended, after birth, to fix the bowel in

position to reduce the risk. Information about malrotation is available at www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/malrotation-and-volvulus

Abnormalities of the size and function of the spleen

The spleen can be important in protecting the baby against some types of infection. If the spleen is absent or not working normally, a daily dose of antibiotics (penicillin) would be needed to protect against bacterial infection. The information sheet about removal of the spleen (splenectomy) also explains the implication of having a missing or dysfunctional spleen -

www.gosh.nhs.uk/conditions-and-treatments/procedures-and-treatments/splenectomy

Underdevelopment of the channels that drain bile from the liver (biliary atresia)

This is relatively rare but an important condition sometimes seen with left isomerism. This would require an operation over the first few months of life and long term follow up by the liver team. Information about biliary atresia is available from the Children's Liver Disease Foundation at childliverdisease.org/liver-information/childhood-liver-conditions/biliary-atresia/

After birth, your baby will be referred to relevant specialists for related conditions which don't involve the heart.

What is the risk of having another child with isomerism?

The risk of having another pregnancy with isomerism is approximately 5 to 10 per cent. Your local hospital can refer you to a genetic counsellor before planning another pregnancy. This would be particularly important if another family member has any abnormality of the position of the heart as this may increase the risk.



Further information and support

Before birth, please call the Fetal Cardiac Nurse Specialists on 020 7762 6711 or send them an email to gosh-tr.fetalcns@nhs.net More information about the Fetal Cardiology service is at <https://www.gosh.nhs.uk/wards-and-departments/departments/clinical-specialties/fetal-cardiology-service-information-parents>

Heartline Families is a support group for anyone affected by a child's heart problem. Call them on 0330 022 44 66 or visit their website at www.heartline.org.uk.

Antenatal Results and Choices (ARC) is a registered charity that offers continued support and advice to parents who face difficult decisions regarding fetal abnormalities. Call them on 020 7631 0285 or visit their website at www.arc-org.uk

Children's Heart Federation (CHF) is a national charity and the umbrella body for voluntary organisations working to meet the needs of children and young people with congenital and acquired heart conditions and their families. Call their free helpline on 0808 808 5000 (Monday to Friday from 9.30am to 4.30pm) or visit their website at www.childrens-heart-fed.org.uk

British Heart Foundation (BHF) is a national charity providing information and support to anyone of any age affected by heart problems. Call their helpline on 0300 330 3311 or visit their website at www.bhf.org.uk

