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RESEARCH ARTICLE

DEXTROCARDIA SITUS INVERSUS TOTALIS

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Abstract

This article was aimed at studying the occurrence of dextrocardia situs inversus totalis. It is a very rare genetic disorder which is inherited by an individual from both parents. The incidence is seen when a fertilised embryo divides later than usual during pregnancy. The abnormal genes cause the primitive or the cardiac tube to reverse directions while a foetus is developing in the womb. This study refers to the initiation of this condition, different types, symptoms, diagnosis, complications / consequences, and treatment.

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Introduction:-

Dextrocardia situs inversus is a genetic condition that is characterised by abnormal positioning of the heart. In people affected by dextrocardia, the apex (tip of the heart) points towards the right side of the chest instead of the left side. Dextrocardia situs inversus totalis involves complete transposition (right to left reversal) of the thoracic and abdominal organs. The heart is not in its usual position in the left chest but it is on the right. The stomach which is normally in the left upper abdomen, is on the right. In patients with situs inversus totalis, all of the chest and the abdominal organs are reversed and appear in mirror image when examined by tests such as X- rays.

Causes:

In most common situations, situs inversus totalis involves complete transposition (right to left reversal) of all the abdominal organs. The heart is not in its usual position in the left chest but is on the right, a condition known as *dextrocardia* which literally means right hearted. Dextrocardia in isolation without any heart defects is unusual. Non-dominant, also called as the autosomal recessive genes cause dextrocardia.

These abnormal genes cause the primitive or the cardiac tube to reverse direction while a foetus is developing in the womb. Depending on the extent and timing of the reversal, the heart and the abdominal organs may also develop in a reversed form, which is called totalis. As it is a recessive gene that causes dextrocardia, an individual must inherit a copy of the abnormal gene from both parents to develop the condition. Dextrocardia situs inversus totalis affects approximately 1 out of every 10,000 children. Ethnicity, race, gender do not seem to have any impact whether or not a person develops this condition.

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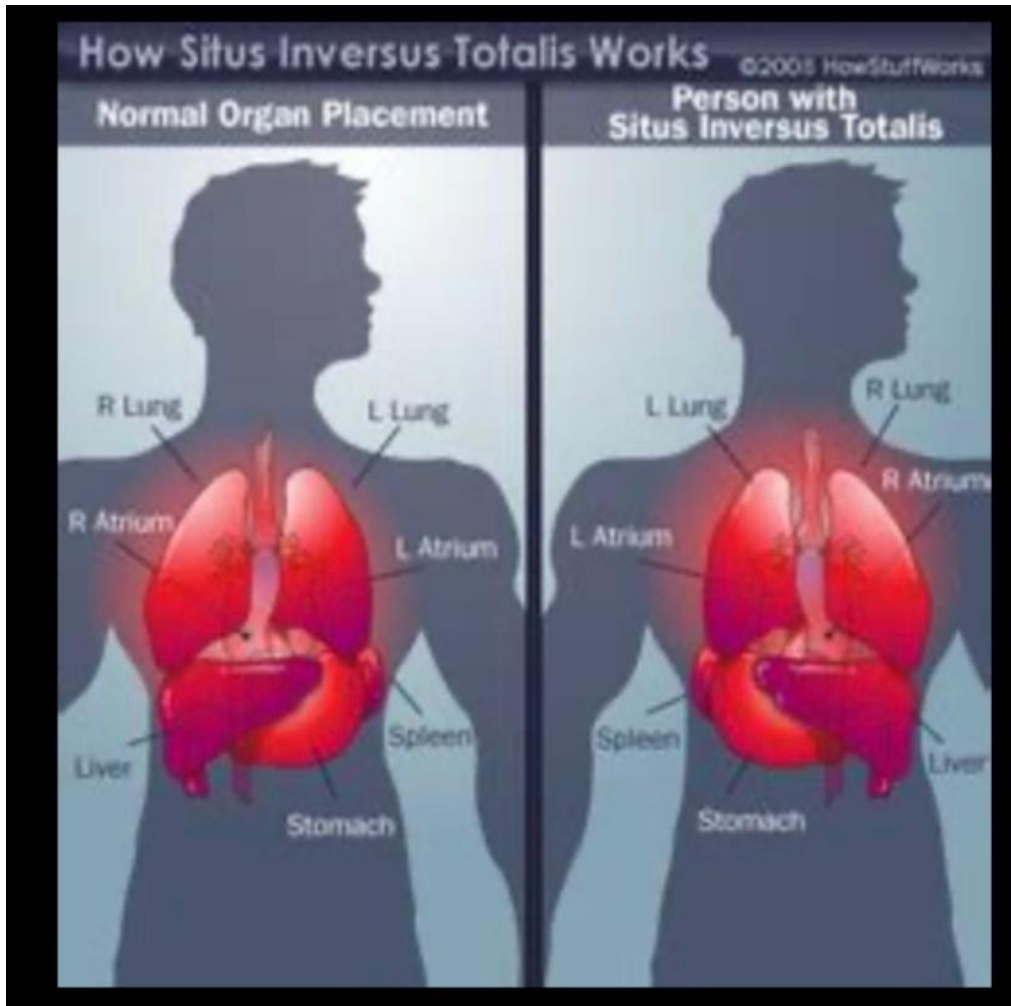


Figure 1:- How situs inversus works.

Type Of Situs Inversus

Situs inversus or situs transversus is a rare condition. There is a total transposition of abdominal and thoracic viscera. People with Situs inversus are asymptomatic of their unusual anatomy until they are evaluated medically for unrelated conditions or diagnosed incidentally during laparotomy or autopsy. But when associated with midgut volvulus or artresias, it would present early in the neonatal age. The normal anatomical position of internal organs is termed as situs solitus. Situs inversus has 2 subtypes - dextrocardia and levocardia. A person is said to have dextrocardia if the point of the heart is on the right side. A person is said to have levocardia if the heart is on the left side yet the other organs are flipped. It is associated with other cardiac abnormalities. Situs inversus abdominus also known as situs inversus with levocardia or left located heart is a condition with right to left reversal limited to the abdomen. In Situs inversus totalis there is a complete right to left reversal of all of the viscera including dextrocardia, the morphologic right atrium is on the left and the left atrium is on the right. The normal pulmonary anatomy is reversed such that the left lung has 3 lobes and the right lung has 2. The liver and the gallbladder are located on the left and spleen and stomach are on the right side. Associated anomalies with situs anomaly are congenital heart disease or splenic malformations.

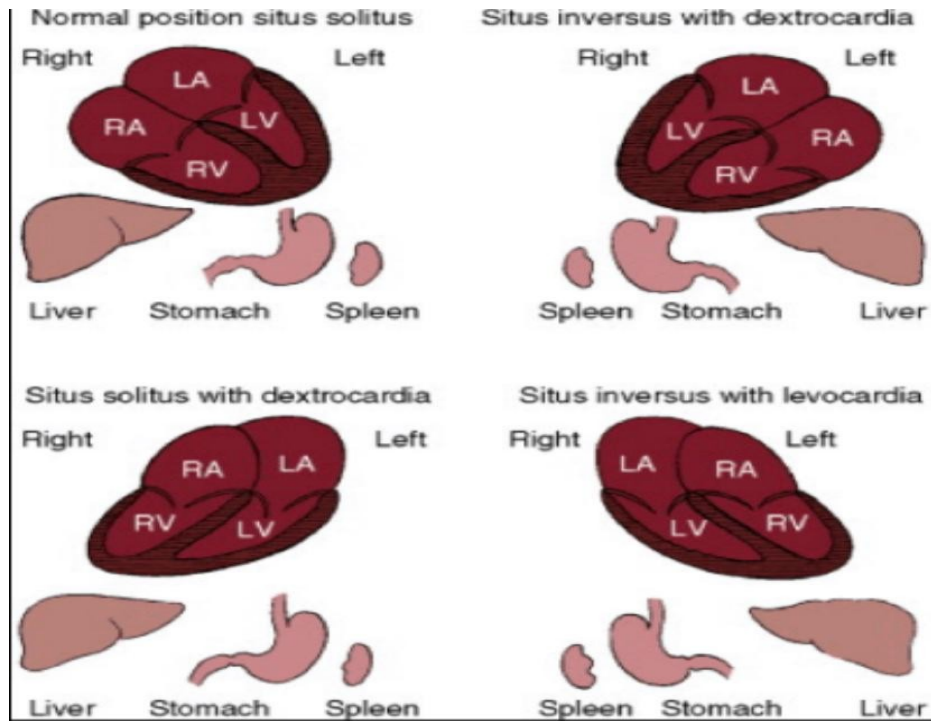


Figure 2:- Types of situs inversus.

Symptoms Of Situs Inversus

It is possible that a person may not have any complications because the organs are still functional normally and has situs inversus. Few patients experience a lung condition called primary ciliary dyskinesia which causes mucus buildup in lungs which leads to bronchitis. People with situs inversus are said to have Kartagener's syndrome. Symptoms that require medical attention include unexplained exhaustion, inability to gain weight, chronic infection of sinus and lungs, difficulty in breathing, yellow skin, blue tinted skin around fingers and toes.



Figure 3:- Xray of a dextrocardia.

Situs inversus is usually associated with dextrocardia with only 3-5% incidence of congenital heart disease, most commonly transposition of the great vessels. Of these patients 80% have a right sided aortic arch. Situs inversus with levocardia which is much rarer to 0.0005% congenital heart disease is found in 95% of patients. Upto 20% of the patients with situs inversus have Kartagener syndrome which comprises a subgroup of primary ciliary dyskinesia. Situs inversus imaging features on chest radiograph to be evaluated are:

1. Location of heart apex
2. Location of aortic arch
3. Location of stomach bubble / liver

The typical clinical phenotype of primary ciliary dyskinesia includes any or all of the following. Neonatal respiratory distress, chronic persistent lower respiratory symptoms, early onset and persistent wet cough, chronic upper respiratory symptoms, nasal congestion and otitis media. The presence of any 2 of these features provide a strong clinical phenotype of primary ciliary dyskinesia. At least 12% of primary ciliary dyskinesia patients have situs ambiguus and these patients have a 200 fold increased probability of having structural congenital heart disease as compared to the general population with heterotaxy.

The recognition of situs inversus is important for preventing surgical mishaps that result from the failure to recognize reversed anatomy or an atypical history. For example, in a patient with situs inversus, cholecystitis typically causes left upper quadrant pain, and appendicitis causes left lower quadrant pain. A trauma patient with evidence of external trauma over the ninth to eleventh ribs on the right side is at risk for splenic injury. If surgery is planned on the basis of radiographic findings in a patient with situs inversus, the surgeon should pay careful attention to image labeling to avoid errors such as a right thoracotomy for a left lung nodule.

Diagnosis Of Situs Inversus

Situs inversus can be diagnosed by looking at the organs by x-ray, CT scan, MRI scan and ultrasonography. This condition seldom causes symptoms and is so rare a person may not know they have it. It can also be discovered when a doctor checks the heartbeat. The heartbeat is typically loudest at the lower point of the heart on the person's left side. Since the persons with situs inversus have a heart that points to the right side, the beat would be loudest on that side. Surgery to reverse the organs positioning is usually not recommended. Situs abnormalities may be recognized first by using radiography or ultrasonography. However, computed tomography (CT) scanning is the preferred examination for the definitive diagnosis of situs inversus with dextrocardia. CT scanning provides good anatomic detail for confirming visceral organ position, cardiac apical position, and great vessel branching. Magnetic resonance imaging (MRI) is usually reserved for difficult cases or for patients with associated cardiac anomalies. Most patients with situs inversus with levocardia require additional imaging to evaluate the associated cardiac anomalies. When radiation exposure is a concern, MRI or ultrasonography may be preferred.

The differential diagnosis includes appendicitis, asplenia/polysplenia, congenital coronary abnormalities, sinusitis, and ventricular septal defect. Other conditions to be considered are primary ciliary dyskinesia, heterotaxy, left isomerism (ie, Ivemark syndrome), right isomerism (ie, asplenia syndrome), situs solitus, and transposition of the great arteries. If radiologic intervention is to be performed in a patient with situs inversus, the condition should be known from earlier diagnostic imaging. A question of improper image labeling must be resolved before any procedure is initiated. Failure to recognize situs inversus before performing a radiologic procedure may result in intervention on the incorrect side of the patient. Attention to the left and right sides of the patient and the left and right labeling of images is helpful to prevent mistakes in diagnosis and/or surgical intervention. Discordance between the direction of the cardiac apex and the abdominal situs suggests congenital heart disease. Situs ambiguus and situs inversus with levocardia have this discordance between the direction of the cardiac apex and the abdominal situs thus, further imaging is usually needed.

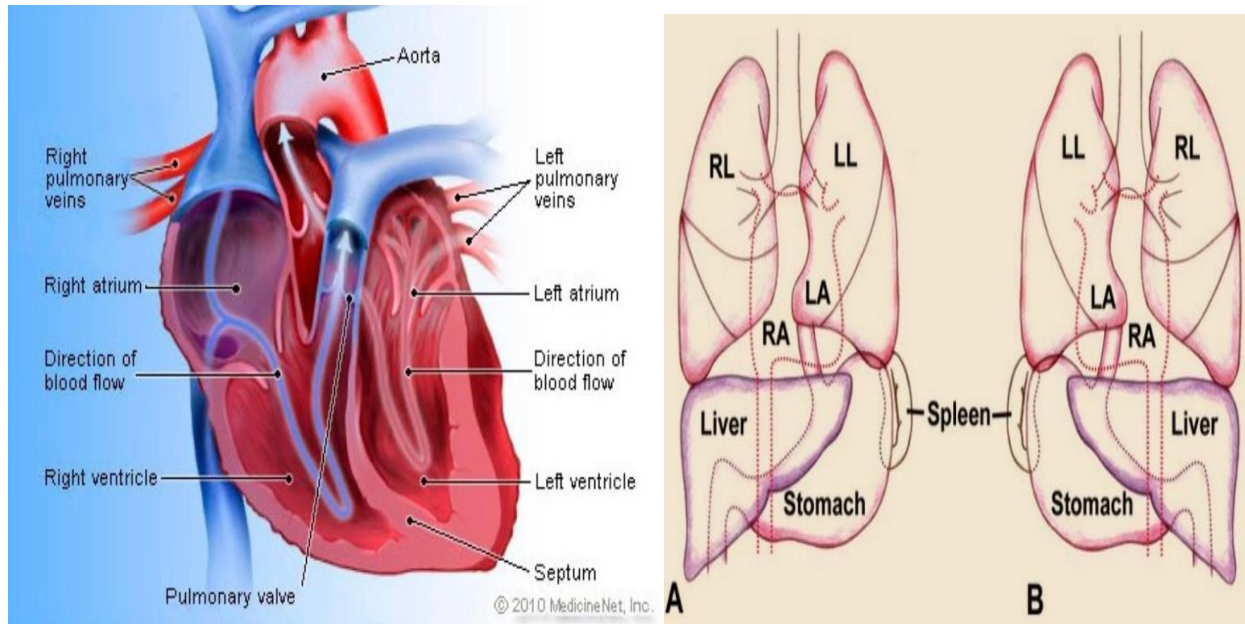


Figure 4:- Positioning of heart with dextrocardia.

Figure 5:- Complete transversal of organs (A- normal person).

Complications

All the reverse organs themselves function normally; their irregular positioning makes the diagnosis difficult. For example, in someone with dextrocardia situs inversus appendicitis will cause sharp pain in the lower left part of the abdomen instead of the right. When the anatomical difference occurs they make the surgery difficult. Other complications associated may include bronchial diseases like pneumonia associated primarily with the loss of cilia, cardiovascular disorders and infection.

Related Conditions

Dextrocardia may be associated with other related conditions which include kartagener syndrome, dextroversion, dextro position, transposition of the great vessels tricuspid atresia heterotaxy, endocardial cushion defect.

Treatment And Outlook

Most people with dextrocardia do not display any symptoms and so go untreated. In infants with dextrocardia which is accompanied by heart defects may require surgery. Many children will be given medication that increases the force of the heart beat and lower blood pressure before surgery. For those with Kartagener syndrome treatment of sinus in lungs symptoms may help lessen the chances of dextrocardia. Treatments include expectorant for mucus clearing medications, antibiotics for or bacterial infections, treating cases of bronchitis and sinusitis. Genetic counselling may help those with dextrocardia who are looking to start a family. People with this condition have a normal life expectancy.

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