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SITUS INVERSUS TOTALIS IN NEWBORN

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Abstract

Situs inversus totalis [SIT] is a rare congenital abnormality and a complex disorder of embryological morphogenesis. It is characterized by a mirror-image transposition of the abdominal and the thoracic organs. Frequently it is associated with other congenital anomalies, congenital heart malformations being found in 3-9% of patients with SIT. Isolated SIT is also reported.

We report a preterm newborn treated with non-invasive ventilation [CPAP] for respiratory distress. The X-ray of the lungs revealed the expected respiratory distress syndrome [RDS], but surprisingly and accidentally dextrocardia and left sided liver. Heart ultrasound showed dextrocardia without congenital anomaly. Abdominal ultrasound confirmed left-sided liver, and right sided spleen. After a lengthy 30 days stay the baby was discharged at home without any complication.

SIT is an extraordinary rare condition in neonates. Timely diagnosis in-utero is important for prenatal and postnatal monitoring of newborns. Most of the patients with SIT without congenital anomalies do not have any complications in their life. However, appropriate information about the condition is important to prevent complications during some invasive and surgical interventions.

***Keywords*:** situs inversus totalis, newborn.

Introduction

The term situs refers to the position or location of an organ in relation to the midline of the body [1]. Three types of situs are described: solitus, inversus, and ambiguous, corresponding to normal arrangement of the organs, a mirror image of the normal position of internal organs (SIT)[1], and is the random arrangement of internal organs [situs ambiguous] [2].

Situs inversus totalis [SIT] is a rare congenital abnormality [3], first described by Aristotle [BC. 384–322][3, 4], followed by the report SIT in a humans Fabricius in 1600a.d. [5]. Vehsemayer [6] reported the first case of transposition of the viscera by X-rays was in 1897 [6].

It is of note that six risk factors have been identified which influence to the development of SIT: family history of heart defects, family history of noncardiac anomalies, maternal diabetes, antitussive use, paternal smoking, and low socioeconomic status [7].

Left-right asymmetry occurs as a result of rotation in the opposite direction of the internal organs during the organogenesis of the embryo [8, 9]. Laterality is influenced by a cascade of signaling molecules and genes [3].

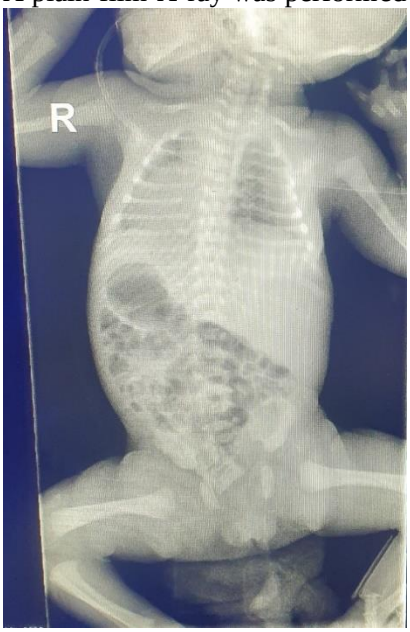
Dextrocardia occurs when the heart fails to migrate to the left chest. The exact mechanism that controls the rotation and migration of internal organs is unknown. More than 100 genes are associated with laterality, including genes for primary ciliary dyskinesia [PCD] [3].

In this article we present a newborn with SIT in a premature baby without congenital anomaly.

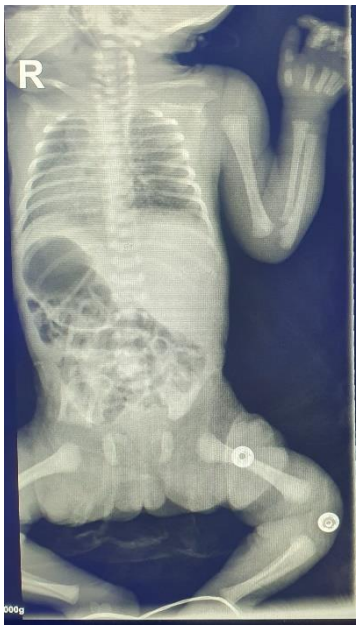
Case report

This was the first pregnancy of a 23-year-old patient. Prenatal cortical maturation of the fetal lung has been done. Family history was uneventful. Fetal distress was diagnosed and she was delivered by

emergency caesarean section. A girl was born in 30+6 week of gestation, with birth weight 1600 g, Apgar score 6/7/7 in first, fifth and tenth minute. After primary resuscitation with a T-resuscitator with FiO2 90% to achieve target saturation, baby was admitted to NICU. A non-invasive CPAP ventilation was started. Lung X-ray showed respiratory distress syndrome [RDS], dextrocardia and liver on the left side. A plain film X-ray was performed and situs inversus totalis and RDS was confirmed [Figure 1A].



A.



B.

Figure 1: Situs inversus totali and RDS before surfactant (A) and after surfactant(B)

Surfactant 200mg/kg by LISA [less invasive surfactant administration] was applied twice. The blood gas analises and lung X-ray improved [Figure 1B].

Abdominal ultrasoun was performed, which confirmed SIT: liver on the left side, spleen in the right hemiabdomen. Heart ultrasound confirmed the dextrocardia without congenital heart anomaly. Cranial ultrasound observed intracranial hemorrhage grade II on both side. The neonate required non-invasive CPAP ventilation for 6 days. After that continued with oxygen support by high flow nasal cannula for 8 days. It was done continuous monitoring of vital parameters during the stay in NICU. At the age of 30 days, the newborn was discharged home in good condition. It is regularly monitored by a pediatrician

Discussion

The incidence of situs inversus totalis is reported to be 1:6500 to 1:25,000 [3], with a male-to-female ratio of 3:2 [9]. In hospitals SIT is observed in 28.8% aged 19 - 45 years and in 29.9% aged 46 - 65 years [10]. The first suspicion of situs inversus totalis can be raised after a careful physical examination, as well as by widely used imaging techniques.

The simplest imaging techniques are ultrasound [US] and plain film X-ray. Typical findings are dextrocardia, left-sided liver and right-sided spleen [11-13]. Dextrocardia is confirmed by electrocardiography, showing inversion of electrical waves [10]. Computed tomography [CT] or fetal

MRI provide a detailed description of situs anomalies well before delivery [14]. SPECT/CT labeled with 99mTc can be used to differentiate polysplenia and abdominal masses [15].

The diagnosis of SIT is rarely confirmed in the neonatal period. In newborns, SIT is most often incidental during radiographic evaluation for respiratory distress [16].

In our patient the diagnosis of SIT was accidental. We performed a lung X-ray due to respiratory distress in a premature infant and noted dextrocardia and left-side liver. It is of note that the mother had regular prenatal monitoring, but no SIT was detected during pregnancy. The diagnosis of SIT and congenital heart disease was verified prenatally by fetal ultrasound. We confirmed SIT with plain film X-ray [dextrocardia, left-placed liver and right-placed spleen] and heart ultrasound [dextrocardia with complex congenital heart disease].

The fact that the patient has a diagnosis of SIT is an important finding. Even in situations where there are no other congenital anomalies and the patient has a normal life, the presentation of common diseases in these patients can be difficult to diagnose due to mirror anatomy [17].

The diagnosis of SIT is especially important in emergency situations. In cases of acute abdominal diseases, such as cholecystitis, acute appendicitis or spleen injury, the manifestation of symptoms and signs is unusual [18, 19].

Oftentimes technical modifications are needed in surgical interventions. Like the general population, patients with SIT can develop malignant or benign neoplasms [20, 21] and accurate diagnosis is required for localization and treatment. In patients with SIT, organ transplantation is more complicated because the donor organs are almost always from a situs solitus donor and there is a problem in placing the organ in a mirror-image cavity because the heart and liver are chiral [22].

Most people with situs inversus live normal lives without symptoms or disability [10]. SIT can be isolated or associated with other congenital abnormalities. The most common is intestinal malrotation, which affects 40–90% of patients [23].

Other conditions that can cause SIT include: duodenal atresia, biliary atresia, gastrochisis, congenital coronary abnormalities, ventricular septal defect [24, 25]. SIT can also be part of some syndromes, such as Kartagener's syndrome [situs inversus totalis, abnormal paranasal sinuses and bronchiectasis] [26] and Ivemark's syndrome [SIT and asplenia] [27].

In polysplenia, SIT is reported in 20% of cases [28]. The rate of congenital heart disease is around 0.6% in situs solitus [normal anatomy], 3–9% in situs inversus totalis, and almost 80% in situs ambiguous [3]. SIT with dextrocardia and congenital heart defect is observed in 3-5%. The most common congenital heart defect in SIT is transposition of the great vessels [29].

Situs inversus totalis with left heart is a rare condition [30] and is mostly associated with congenital heart disease [31].

The presented case is without congenital anomalies. The infant is continuously monitored by the pediatrician for recurrent respiratory infections and bronchiectasis due to the risk of developing Kartagener syndrome.

Conclusion

SIT is a rare condition. The detection of anatomical alteration of the fetus in-utero is important for an appropriate strategy in prenatal and postnatal monitoring. Diagnostics should always be expanded to reveal any accompanying congenital anomalies. A multidisciplinary approach is necessery in diagnosis and treatment, as well as in monitoring the patients. Although most of the patients with SIT do not have any complications in their life, it is necessary to inform their clinicians to prevent complications during some surgery interventions.

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