Situs inversus totalis, oesophageal atresia and tracheo-oesophageal fistula

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DESCRIPTION

A term baby girl had respiratory distress on first feed. She had cardiac impulse and heart sounds on the right side. Liver was palpable below the left costal margin with liver dullness on the left side. An orogastric tube got arrested at 10 cm. Chest x-ray showed the arrest of a replogle tube with its tip at T4 in keeping with oesophageal atresia (OA) and dextrocardia (figure 1A). X-ray abdomen demonstrated gas in the gastric bubble on the right side with liver on the left side suggesting situs inversus totalis (SIT) and bowel loops indicating a distal tracheo-oesophageal fistula (TOF) (figure 1B). ECG confirmed dextrocardia with right-sided aortic arch and small atrial septal defect. She underwent left-sided extrapleural posterolateral thoracotomy, division of TOF and primary repair of OA uneventfully. She is asymptomatic and thriving well at 6 year follow-up.

OA with TOF and situs inversus of the thoracic and abdominal organs is a very rare combination of congenital anomalies to find in the same patient.1 2 More than 50% of infants with OA have associated anomalies. When a patient is noted to have congenital heart disease as part of situs anomalies including SIT, or if an atypical position of organs is noted at imaging evaluation, we recommend the patient to undergo chest radiography, abdominal ultrasonography, upper gastrointestinal study and abdominal CT.1 2

Learning points

▸ Situs inversus totalis (SIT) is a rare anomaly and requires to screen for other anomalies.
▸ The presence of SIT causes difficulty in the repair of oesophageal atresia with tracheo-oesophageal fistula (OA/TOF).
▸ SIT with OA/TOF dictates left-sided operative approach.

CONTRIBUTORS

All authors have actively contributed in the management of this patient and collection, analysis and interpretation of data and preparation, appraisal and finalisation of this manuscript of this article.

COMPETING INTERESTS

None.

PATIENT CONSENT

Obtained.

PROVENANCE AND PEER REVIEW

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REFERENCES


Figure 1 (A and B) Chest and abdominal radiographs. Note the arrest of the replogle tube at the C4 level (arrow head) and the stomach bubble on the right (S) and liver shadow on the left side of the abdomen.