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Antenatally diagnosed right-sided stomach (dextrogastria): A rare rotational anomaly

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ABSTRACT

Aim: Antenatal detection of right-sided stomach (dextrogastria) is rare, and its significance in regards to intestinal rotation is unclear. We aimed to review all cases of antenatally-diagnosed dextrogastria in our regional fetal medicine unit over 10 years.
Methods: A retrospective case-note review of patients identified from a prospectively-maintained database was performed.
Results: Twenty cases of antenatally-diagnosed dextrogastria were identified from 2004 to 2014. There were 8 terminations and 1 intra-uterine death. One patient has no post-natal information obtainable. Ten infants were live-born, and 2 died secondary to cardiac disease in the neonatal period. All had significant cardiac/vascular anomaly on postnatal assessment, including the 3 neonates in whom dextrogastria was the only antenatal finding. Two neonates developed bilious vomiting and underwent Ladd's procedure. Operative findings were dextrogastria/malrotation in both. A third child had gastro-oesophageal reflux, and contrast demonstrated stable duodenal/midgut position. This child has not developed symptoms attributable to malrotation and not undergone surgery. All 3 of these infants had asplenia or polysplenia and were managed with antibiotic prophylaxis/immunisation. Five children in the series were not investigated for malrotation and have not come to surgical attention (one is known to be asplenic).
Conclusion: Antenatally-detected dextrogastria, even if apparently isolated, was always associated with postnatal significant cardiovascular anomaly, splenic abnormality or *situs inversus*. This may be important for antenatal counselling. We currently recommend postnatal echocardiography and splenic assessment, but reserve GI investigation/intervention for symptomatic malrotation owing to potential significant cardiac comorbidity.

A right-sided stomach (dextrogastria) is a rare finding with only 80 cases presented in the literature, mainly as a part of the heterotaxy syndrome or *situs inversus* [1,2]. Heterotaxy syndrome, also known as isomerism of the atrial appendages, has a wide variety of subtypes, with an estimated incidence of 1 per 10,000–40,000 live-born [3,4]. Children with isolated dextrogastria have been reported, but this rotational anomaly is rarely reported without any intracardiac anomaly, and has not been described as an isolated antenatal finding [5–9]. Diagnosis may be on antenatal ultrasound, or subsequently as a consequence of clinical symptoms after birth.

Controversy exists as to whether patients with heterotaxy syndrome should undergo elective investigation for malrotation and surgical correction if present [10]. It is currently unknown what the implications of making an antentatal diagnosis of apparently isolated dextrogastria are, in terms of possible associated anomalies and importantly, whether the midgut is likely to be in an unstable position, thus predisposing to volvulus.

In this study, we aimed to review our regional fetal medicine and paediatric surgery centre's experience of antenatally-diagnosed dextrogastria from the past 10 years. We explore the issues involved in establishing an accurate diagnosis and the subsequent dilemmas in managing these patients.

1. Methods

Our prospectively maintained regional antenatally detected anomalies register was searched for 'right-sided stomach', 'abnormally-sited stomach' and 'dextrogastria', and patient notes were reviewed retrospectively. This was registered as a service evaluation with institutional approval; formal ethical approval was not required. Data were collected on antenatal findings, subsequent postnatal abnormalities, in particular splenic and cardiac anomalies. Gastrointestinal symptoms, imaging and surgery were documented.

Current literature was reviewed up to July 2014 using the PubMed database and with the use of the following search terms; ('antenatal' OR 'antenatally-diagnosed') AND ('dextrogastria' OR 'right-sided stomach' OR 'atrial isomerism' OR 'heterotaxy') AND 'stomach').

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Table 1
Details of the outcomes of the 10 live-born children with dextrogastria.

Antenatal US	Postnatal Cardiac Diagnosis	Postnatal GI Diagnosis	Outcome
Dextrogastria only	Absent IVC, Small ASD	Dextrogastria and malrotation. Asplenia	Ladd's procedure age 4 days for symptomatic malrotation.
Dextrogastria only	Heterotaxy syndrome. Right-sided SVC	Dextrogastria and malrotation. Polysplenia,	Ladd's procedure age 12 days for symptomatic malrotation.
Dextrogastria only	Right-sided IVC & SVC	GOR. 1 cm splenunculus. No GI surgery	Prokinetic for GOR. Upper GI contrast — stable DJ/midgut position.
Dextrogastria. AVSD/Univentricular	Transposition of Great Arteries, Tricuspid atresia, VSD, Pulmonary Stenosis	Dextrogastria. Asplenia	Pulmonary Artery Band performed, awaiting atrial septectomy.
Dextrogastria. Situs Inversus	Situs inversus, VSD, Pulmonary Atresia, Aortic origin from RV	No contrast study or GI surgery	Corrective cardiac surgery
Dextrogastria. Right atrial isomerism	Right Atrial Isomerism, AVSD, Situs Inversus	No contrast study or GI surgery	Corrective cardiac surgery
Dextrogastria. Left atrial isomerism	Unbalanced AVSD	No contrast study or GI surgery	Corrective cardiac surgery
Dextrogastria. Situs Inversus	Primary Cilliary Dyskinesia, Situs inversus totalis. Structurally normal heart	No contrast study or GI surgery	No cardiac surgery.
Dextrogastria. Situs Inversus	TAPVD, RA isomerism, VSD and TGA	No contrast study or GI surgery	**Postnatal Death.** Spontaneous labour at 28 weeks, died prior to cardiac sugrery
Dextrogastria. Situs Inversus	Situs Inversus, hypertrophic cardiomyopathy	No contrast study or GI surgery	**Postnatal Death.** Term delivery, idiopathic cardiomyopathy.

2. Results

Twenty cases of antenatally-diagnosed dextrogastria were identified in the period 2004–2014. The first 3 children discussed were referred for paediatric surgical opinion on the basis of the isolated antenatal finding of dextrogastria. Of the further 17 fetuses diagnosed with right-sided stomach in this period 15 had significant cardiac anomalies (5 left atrial isomerism, 4 right atrial isomerism, 2 transposition of the great arteries, 3 undefined cardiac anomalies, 1 pulmonary atresia with ventriculoseptal defect), 1 had trisomy 13 and 1 had spina bifida. One patient was demonstrated to have *situs inversus totalis* with a structurally normal heart.

Eight of the pregnancies were terminated, one was an intrauterine death, and one had no post-natal details. Ten were live-born but two died in the early neonatal period. Details of the live-born babies are summarised in Table 1. The three infants with dextrogastria only are reviewed in detail.

3. Cases

In the first, a right-sided stomach was the only abnormality seen on routine anomaly ultrasound scan at 22 weeks gestation. After term delivery, the baby had two bilious vomits on day 1. Ultrasound and upper gastrointestinal contrast study confirmed the right-sided stomach, a midline saddle liver, absence of the inferior vena cava (IVC), absent spleen and a small atrioseptal defect. The rotation of the midgut was difficult to ascertain on the contrast examination; however, there was no evidence of volvulus. Vomiting recurred on day 3 and the patient underwent laparotomy. Right-sided stomach was confirmed, with midline liver, intrahepatic gallbladder, normal pancreas and asplenia. The duodenum was in the left upper quadrant with the duodenal–jejunal (DJ) flexure also situated to the left of the midline, confirming midgut malrotation. The caecum was located in the right lower quadrant. The colonic fixation was abnormal (Fig. 1). A modified Ladd's procedure was carried out. Postoperative recovery was uneventful and the infant was discharged at day 4 after surgery. Prophylactic penicillin has been prescribed to manage the asplenia.

The second infant was antenatally-diagnosed with a right-sided stomach only on routine anomaly ultrasound at 20 weeks gestation. After term delivery, early post-natal abdominal ultrasound confirmed dextrogastria, with midline saddle liver, a left-sided gallbladder and no spleen. On cardiac ultrasound and MRI, the hepatic veins were seen to drain directly into the right-atrium, there was no IVC, and the azygos vein drained into a right-sided superior vena cava. Six small splenules were located behind the right-sided stomach. The duodenal loop was seen in reversed position with D3 seen to cross the midline. The patient was initially asymptomatic and discharged at day 3. However, on day 10 he developed bilious vomiting, was re-admitted and underwent emergency laparotomy. The small bowel mesentery was narrow-based and there was a non-critical mid-gut volvulus. A modified Ladd's procedure was carried out with normal post-operative course. At latest follow-up age 15 months, the patient remained well. He has been commenced on prophylactic penicillin and vaccinated to cover potentially nonfunctioning polysplenia.

The third infant was diagnosed with dextrogastria only on antenatal anomaly ultrasound scan and post-natally was asymptomatic. On day 5, she underwent ultrasound studies that, besides the known dextrogastria, showed midline saddle liver and an 11 mm splenunculus in the right upper quadrant. Echocardiogram demonstrated a right-sided IVC which was a continuation of the left renal vein. Interruption of the IVC was seen towards the heart, with azygos continuation into the right superior vena cava. Upper GI series showed right-sided stomach, with segments 1 and 2 of the duodenum also on the right side and the 3rd segment crossing the midline with the DJ-flexure just left of the pedicle of L1. The patient was discharged on day 6 without undergoing any surgical procedure. In the first 6 months, the patient suffered from 5 significant apnoeic attacks after feeding, possibly associated with nonbilious

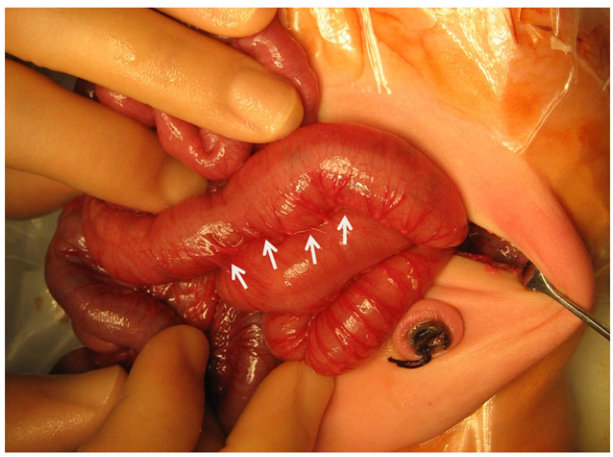

Fig. 1. Right colon unfixed, transverse colon and left colon adherent to each other (white arrows) and left colon attached to the posterior peritoneum in the left upper quadrant.