ISOLATED DUODENAL PATHOLOGY IN MONOCHORIONIC DIAMNIOTIC TWINS INVOLVING SAME SEX

Junaid Ashraf, Rakesh Thakur, Ruth Hallows, Varadaraja Kalidasan

1Department of Paediatric Urology, Leeds General Infirmary, Leeds, UK
2Department of Paediatric Surgery, Royal Alexandra Children’s Hospital, Brighton, UK

Abstract
We are reporting first case of duodenal atresia occurring in both members of premature low birth weight monochorionic diamniotic twins of same sex. The twins were born at 28 weeks of gestations with no dysmorphic features and normal chromosomes. One of the twins had type II duodenal atresia in the second part of the duodenum along with meckel’s diverticulum which was also excised. The other presented with duodenal web at the junction of second and third part of duodenum. He also underwent an elective left sided inguinal hernia repair at five months of age. They underwent delayed repair at 3rd and 4th weeks of age respectively. Both babies had an uneventful post-operative course.

Keywords: duodenal atresia, duodenal web, monozygotic twins

Introduction
The incidence of duodenal atresia is about 1.4 per 10,000 live births in a singleton pregnancy [1] and there have been reports of occurrence in the same family [2]. However, duodenal atresia in identical twins is exceedingly rare. Duodenal atresia includes a range of anomalies from a simple duodenal diaphragm, duodenal stenosis to complete discontinuity. Associated conditions include Down’s syndrome (30%) and VACTERL anomalies (Vertebral, Anal, Cardiac, Tracheo Esophageal, Renal and Limb anomalies) in about 50% cases. We present a previously unreported case of isolated duodenal atresia and duodenal web in same sex mono-zygotic identical twins.

Family and Antenatal History
There is no history of consanguineous marriage in the family pedigree. Their mother was only 17 years old primipara white Caucasian. There is no history of any maternal ingestion of drugs, infections or irradiation exposure. Her antenatal scans at 14 weeks diagnosed monochorionic diamniotic pregnancy. At 21 weeks, polyhydramnios along with double bubble were seen in one foetus (case 1). Throughout the pregnancy, the umbilical vascular Doppler flow was reported to be normal. At 28 weeks of gestation, owing to failure to progress of spontaneous labour with breech presentation, an emergency caesarean section was carried out.

Case 1
A male baby weighing only 1010 grams was born at Gestation 28+4 weeks. Baby had to be ventilated because of respiratory distress of newborn; however he was extubated onto continuous positive airway pressure (CPAP) on day 4 of life. An echo done found a structurally normal heart with a small patent ductus arteriosus (PDA), though, the later scans showed no evidence of PDA. A plain abdominal X ray taken on 1st day of life showed a large stomach air bubble and dilatation of the first portion of the duodenum (Fig. 1). No air was seen distal to the proximal duodenum. An ultrasound abdomen was normal, ultrasound of brain prominent lateral ventricles...
and possibly small old intraventricular haemorrhage in left ventricle. Laparotomy was performed using transverse supraumbilical incision on the right side. Duodenal atresia with annular pancreas involving the junction of first and second part of duodenum was found and also a Meckel’s diverticulum. A side to side wide duodeno-duodenostomy was carried out along with excision of Meckel’s diverticulum and end to end bowel anastomosis. A feeding tube was passed through the anastomosis for post operative jejunal feeding.

Case 2

The twin brother of case 1 baby weighed 1130 grams at birth. This baby was also ventilated at birth due to respiratory distress of newborn but extubated next day and weaned to optiflow by day 9. An echo done on day 1 showed a small PDA but a repeat echo after a week was absolutely normal. During first few days he did not tolerate enteral feeds and was developing abdominal distension with increased biliary aspirates. An abdominal X ray showed normal gas pattern with air up to the rectum (Fig. 2). An upper gastrointestinal contrast study showed grossly distended descending limb with a liner filling defect (Fig. 3) at the junction of second and third segment of the duodenum raising the possibility of a partially obstructing duodenal web. Contrast was seen to pass through the narrowed segment and opacify the proximal jejunum.
He underwent laparotomy 5 days after his brothers surgery through the similar supraumbilical right transverse incision. A duodenal diaphragm with a central hole identified. The duodenum was mobilized, opened transversely in its first segment, duodenal diaphragm was identified and resected, a feeding tube passed into the jejunum before closing the duodenotomy. He also underwent an elective left sided inguinal herniotomy before discharge. Both babies had an uneventful postoperative course and were on full feeds via bottle at discharge. Chromosomal studies in both babies were normal, had no dysmorphic features, no signs of Down's syndrome or associated congenital abnormalities of the genitalia or anorectum and no limb deformities. The twins are monozygous and monochorionic on the basis of a single, shared placenta, same sex and same blood group as well. They were oxygen dependent initially and stayed on optiflow or oxygen via nasal cannula but at discharge both maintained oxygen saturations without oxygen supplementation. Also, at discharge, both babies had resolving conjugated bilirubinaemia most likely secondary to parental nutrition. There was also evidence of metabolic bone disease requiring supplementation of phosphate and alfa calcidol at discharge.

Discussion

We are reporting first ever case of duodenal atresia (DA) in monochorionic diamniotic twins of same sex, one with unlinked type duodenal atresia in the second portion of duodenum and other with duodenal web between the second and third part of duodenum. Fonkalsrud et al. [2] reported duodenal atresia in six twin patients but they did not state whether both members of the pair had duodenal atresia. There is only one other report of monochorionic diamniotic twins with duodenal atresia by Enrico De Grazia [3], but their twins had intestinal atresia along with duodenal atresia. Also, the member twins were of different sexes, one male and other female. Takahide et al. [4] have also reported duodenal atresia in both members of the twins but their twins were diamniotic dichorionic gestation. There are at least 2 other reports of duodenal atresia in twins [5, 6] but they do not state whether the twins were identical or dizygotic. Our case therefore is the first reported case report of isolated duodenal pathology in monochorionic diamniotic twins involving same sex.

Duodenal atresia is an extremely uncommon congenital malformation. In a review by Cragan et al. [1] the incidence of duodenal atresia in singleton pregnancy has been estimated to be 1.4/10,000 but the incidence of DA in twin pregnancy is higher, 2.4 pre 10,000 (relative risk, 1.8; 95% CI, 0.6, 5.6). This higher incidence in familial DA supports the theory that this anomaly may be determined by genetic factors. Structural malformations that are not genetically determined obviously are seen in only one member of the monozygotic twins. It has been estimated that overall in around 15% of the cases both twins are affected by at least one structural anomaly while mostly only one twin is affected [7]. The etiology of duodenal atresia still remains unclear. Tandler J [8] has suggested that duodenal atresia is secondary to failure of recanalization of epithelia plugging by vacuolar coalescence in the solid developing duodenum. On the other hand, various authors including Louw and Barnard [8, 10, 11] have postulated a vascular theory which states that intestinal atresia in the mid gut is a result of interference of blood supply to a segment of the fetal bowel. At the same time, there are certain environmental factors which are generally implicated in relation to congenital anomalies including various drugs ingestion during pregnancy, maternal infections and exposure to irradiation. Esterly JR et al. [12] have reported an association between jejunal atresia in both members of twins following interruption of the blood supply to the fetal jejunum secondary to maternal rubella infection. Several clinical reports have been reported suggesting a genetic cause of congenital DA as well as other intestinal atresia with an autosomal recessive inheritance [13]. These occurrences of DA in both members of our identical twins may suggest that they may be expression of a single gene responsible for such rare condition. However, no specific gene has been identified so far that can be linked to the development of DA in humans.

Fairbanks TJ et al. [14] have published a study evaluating the role of Fibroblast growth factor receptor 2b (Fgfr2b) which is a critical developmental regulator of proliferation and apoptosis in multiple organ systems including the gastrointestinal tract (GIT). Fgfr2b invalidation results in an autosomal recessive intestinal atresia phenotype. The study done on pregnant mice suggests an innovative idea that absence of embryonic GIT Fgfr2b expression results in decreased proliferation and increased apoptosis resulting in GIT atresia including DA.

Fairbanks [15] also showed that vascular occlusion does not lead to all intestinal atresia but they can result from a genetic defect in the development as well as vascular occlusion; the
REFERENCES