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Case Report

Fetal Double-Bubble Image: Is there Only Duodenal Atresia?

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Abstract

The "double bubble" sign is an ultrasonographic finding that is characterized by the presence of a double liquid-filled bubble image in the fetal abdomen. It indicates duodenal atresia which is frequently associated with other congenital anomalies including esophageal atresia. Antenatal ultrasound diagnosis may sometimes miss this association leading to postnatal surprise and insufficient care. We report the case of a double fetal bubble image identified during a routine antenatal assessment, initially diagnosed as a simple duodenal atresia, with postnatal discovery of an association with esophageal atresia which was overlooked.

Keywords: *Double Bubble; Duodenal Atresia; Esophageal Atresia; Prenatal Diagnosis*

Introduction

Congenital anomalies affecting the digestive system pose significant challenges in both prenatal diagnosis and postnatal management. The "double bubble" sign, often indicative of duodenal atresia, represents a crucial ultrasonographic finding during antenatal assessments. However, the association with other congenital anomalies, particularly esophageal atresia, can be overlooked, leading to postnatal surprises.

Case Report

We report the case of a 30 years old multiparous female patient with no significant past medical history or notion of consanguinity. She was pregnant at 25 weeks of gestation, with good clinical, biological and morphological follow-up. She was admitted for an obstetric ultrasound scan as part of the routine 2nd trimester check-up.

The obstetric ultrasound revealed a monofetal pregnancy with positive cardiac activity and an estimated gestational age of 25 weeks. Anechogenic cystic double bulla was observed in the fetal epigastric region (Figure 1). It was associated with an abundance of amniotic fluid with a large cistern measuring approximately 10 cm in anteroposterior diameter (Figure 2), and an amniotic index exceeding 25, indicating a polyhydramnios. No other associated morphological abnormalities were detected.

The diagnosis of an isolated duodenal atresia was strongly suspected, prompting the cessation of further investigations at this stage.

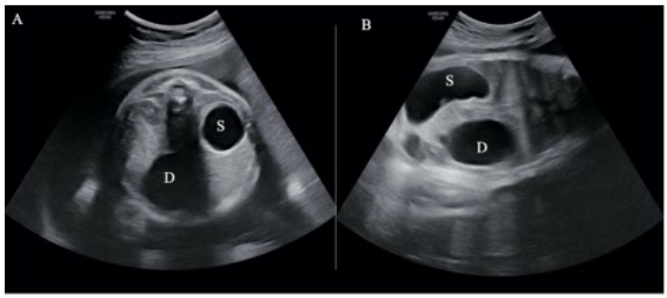

Figure 1: *Prenatal ultrasonography on axial (A) and coronal (B) sections of the fetal abdomen: Two distended cystic bubbles, producing the fetal double-bubble sign indicating the stomach (S) and the duodenum (D).*

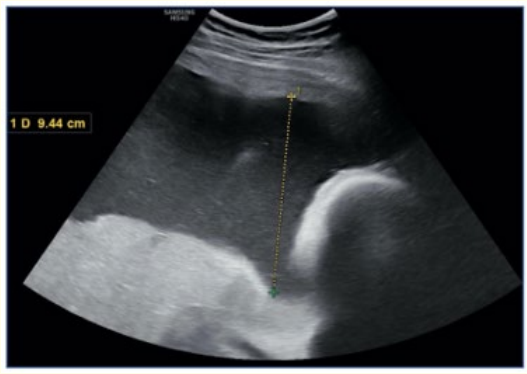

Figure 2: *Prenatal ultrasonography in an axial section of the large cistern, showing an abundance of amniotic fluid. The anteroposterior diameter of the large cistern measures 9.44 cm.*

The parents were informed about potential risks and the specialized care required for the newborn immediately after birth, with close monitoring, this pregnancy was carried to term in a hospital where multidisciplinary care can be provided, with preparation of the surgical tray for emergency surgery. Post-birth, the newborn presented respiratory distress immediately after the first feeds with failure to pass a nasogastric tube into oesophagus. It is worth noting that the introduced tube was of small caliber (6 French). A frontal thoracoabdominal X-ray film has been performed with nasogastric tube in place (Figure 3). The latter was blocked in the upper oesophagus cul-de-sac, and there were a non-aeration of the digestive tract suggesting an associated type I oesophageal atresia. The newborn was transferred to an intensive care unit for conditioning, monitoring and preoperative management. The evolution was unfortunately marked by newborn death before surgical repair.

Discussion

Duodenal atresia is the second most common type of atresia in the gastrointestinal tract [1], with an estimated incidence of 1 in 10,000 live births [2]. It may present as an isolated condition or in association with other congenital anomalies, notably trisomy 21 in 25 to 40%

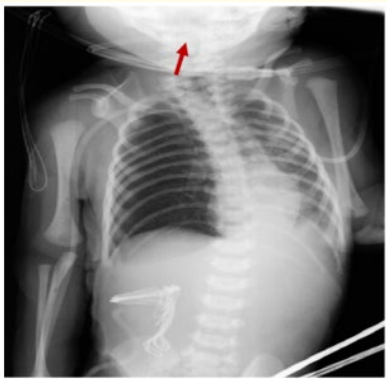

Figure 3: *Frontal thoracoabdominal X-Ray image shows orogastric tube coiled in the atretic proximal esophagus (red arrow).*

of cases [3], VACTERL syndrome (vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula, esophageal atresia, renal anomalies, and limb defects), malrotation, annular pancreas, biliary tract abnormalities, and mandibulofacial anomalies [4]. Duodenal atresia is thought to result from a failure of recanalization during the solid stage of duodenum development in the gastrointestinal tract [5,6]. However, the precise causes remain unclear, with genetic factors and intrauterine vascular ischemia being proposed as possible contributors [7].

The diagnosis of duodenal atresia is often suspected prenatally via ultrasound, especially when polyhydramnios and/or a dilated stomach with the characteristic "double bubble" sign (a large gastric bubble and a smaller proximal duodenal bubble due to fluid accumulation in the proximal duodenum) is observed in up to 80% of cases [8].

Postnatally, infants with duodenal atresia typically present with feeding difficulties and bilious vomiting. Once suspected, infants should be placed on nil per os (NPO) and a nasogastric tube should be inserted for gastric decompression. The diagnosis is confirmed through frontal and lateral thoraco-abdominal X-rays, which reveal the double air bubble. Surgical intervention is the definitive treatment following preoperative management.

Esophageal atresia, characterized by incomplete formation of the esophagus, is often associated with a tracheoesophageal fistula and is the most common type of gastrointestinal atresia, with an estimated incidence of 1 in 3,500 live births. Congenital malformations are present in up to 50% of cases, with two syndromes in particular being associated: VACTERL syndrome, as mentioned earlier, and CHARGE syndrome, which includes coloboma, heart defects, choanal atresia, developmental delays, genital hypoplasia, and ear anomalies. Approximately 19% of infants with esophageal atresia meet the criteria for VACTERL.

Esophageal atresia is classified into five main types, based on the location of the fistulas [9]:

- Type I (or A): isolated atresia with no fistula.
- Type II (or B): atresia with a proximal fistula.
- Type III (or C): atresia with a distal fistula.
- Type IV (or D): a variant of type III.
- Type V (or E): atresia with two or more fistulas.

The most common form is type III (or C). Clinical signs include excessive salivation, coughing, cyanosis during feeding attempts, and aspiration pneumonia. H-type fistulas typically present later in life and are often suspected due to recurrent aspiration pneumonia [3].

The diagnosis of esophageal atresia can be made either prenatally or postnatally. Prenatal ultrasound may suggest esophageal atresia, particularly in cases without a fistula, due to the presence of polyhydramnios and the absence of a visible stomach on repeated scans [10]. In cases of high atresia [11], a cervico-mediastinal pouch with a liquid echostructure may be observed, corresponding to the amniotic fluid-filled upper esophageal pouch [12]. However, these signs can be easily missed, as polyhydramnios is not specific to esophageal atresia, and the stomach may still be visible due to passive filling from a tracheoesophageal fistula or gastric secretions, complicating prenatal diagnosis [12].

Antenatal diagnosis, suspected in about 10% of cases, is typically confirmed by MRI, which may also reveal associated anomalies [13]. According to the literature, duodenal obstruction is more reliably diagnosed prenatally due to the high predictive value of the double bubble sign [14]. In the case we present, prenatal diagnosis of duodenal atresia was made easily. The double bubble sign is rarely seen in other congenital gastrointestinal malformations, including esophageal atresia, consistent with the case of our patient.

Postnatally, esophageal atresia is suggested by the inability to pass a nasogastric or orogastric tube into the stomach. Urgent frontal and lateral thoracoabdominal X-rays should be performed, typically confirming the diagnosis by showing the tube coiling in the upper esophageal pouch. A radiopaque catheter can help localize the atresia on imaging.

In our case, while the antenatal diagnosis of duodenal atresia was straightforward, esophageal atresia was not diagnosed antenatally due to the limitations of obstetric ultrasound. It was only after postnatal clinical signs suggestive of esophageal atresia that the diagnosis was confirmed by a frontal X-ray with the nasogastric tube in place. Due to the unstable condition of the newborn, a lateral radiograph could not be performed.

Following diagnosis, oral feeding must be immediately discontinued, and continuous aspiration should be initiated as part of preoperative management. Once the newborn is stable, surgical repair of the esophageal atresia, typically via an extrapleural approach, can be performed, involving the reconnection of the esophagus and closure of the tracheoesophageal fistula [3].

In cases where surgery must be delayed due to prematurity or clinical instability, the infant should be fed via gastrostomy. When duodenal atresia is also present, the surgical management becomes more complex and may carry a higher risk of severe complications.

Conclusion

The prenatal detection of the double bubble sign is a reliable indicator of duodenal atresia, though other digestive anomalies may also be present. With advancements in prenatal diagnostics, digestive malformations should increasingly be anticipated rather than presenting as unexpected neonatal findings. In some cases, prenatal ultrasound alone may not provide enough information. Therefore, a fetal digestive MRI should be considered to complement the ultrasound, offering a more detailed and comprehensive evaluation of the malformations. Esophageal atresia is one such anomaly that should be included in the differential diagnosis during prenatal assessments. This thorough diagnostic approach helps not only in confirming the presence of malformations but also in planning the immediate care and management of the newborn at birth.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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