

Congenital Duodenal Stenosis: Prenatal Evaluation by Three-dimensional Ultrasound HDlive Silhouette Mode, Magnetic Resonance Imaging, and Postnatal Outcomes

Pedro Teixeira Castro^{1,2}, Ana Paula Pinho Matos¹, Heron Werner², Edward Araujo Júnior^{3*}

¹Department of Radiology, Imaging Diagnostic Clinic, Federal University of Rio de Janeiro, Rio de Janeiro-RJ, Brazil, ²Department of Radiology, Federal University of Rio de Janeiro, Rio de Janeiro (RJ), Rio de Janeiro-RJ, Brazil, ³Department of Obstetrics, Federal University of São Paulo (EPM-UNIFESP), Paulista School of Medicine, São Paulo-SP, Brazil

Abstract

The “double bubble” sign is a common sign of congenital duodenal obstruction (CDO), which has been detected during prenatal diagnosis for over 40 years. CDO is strongly associated with chromosomal and structural abnormalities and encompasses a wide spectrum of diagnoses. Here, we describe a case of duodenal stenosis, a rare cause of duodenal obstruction, which was suspected using conventional two-dimensional ultrasound and three-dimensional reconstruction with the HDlive silhouette mode at the 28th prenatal week. The suspicion was further supported by magnetic resonance imaging performed at the 32nd prenatal week and confirmed by postnatal surgery.

Keywords: Congenital duodenal obstruction, magnetic resonance imaging, postnatal outcome, prenatal diagnosis, three-dimensional ultrasound

INTRODUCTION

Congenital duodenal obstruction (CDO) and its most common imaging finding – the “double bubble” sign – have been described during prenatal diagnosis for over 40 years and may occur as early as the beginning of the 2nd trimester.^[1] During the intrauterine and neonatal periods, CDO has a poor prognosis, which is mostly related to its common association with chromosomal abnormalities, such as trisomy 21, and associated malformations, such as congenital heart diseases. Therefore, the prenatal diagnosis of CDO presents several benefits for the neonate because it decreases the rate of preoperative complications and allows to adequately plan and prepare neonatal-related operations, thereby reducing the time to begin postoperative oral feeding and the neonate’s hospital stay.^[2]

We describe a case of CDO suspected using conventional two-dimensional (2D) ultrasound and three-dimensional (3D) reconstruction with HDlive silhouette-rendering mode at the 28th week of gestation, which was supported by magnetic resonance imaging (MRI) study performed at the 32nd week and confirmed by postnatal surgery.

CASE REPORT

A 36 years old woman, G3 P0 who presented a normal 1st trimester scan exhibited an enlarged stomach during the 2nd scan at 23 weeks. No other malformations or signs were observed during the examination. A follow-up scan was recommended, and at the 28th week, the “double bubble” sign was observed. The lumen of the stomach and the duodenum, which was obstructed at the angle of Treitz, could be completely reconstructed in the 3D ultrasound using the HDlive silhouette-rendering mode [Voluson E10, General Electric, Zipf, Austria; Figure 1]. The distal intestine was apparently normal, with a visualization of the rectum and anus, and no other malformation was observed. An MRI study was performed at the 32nd week to analyze the distal portion of the intestine, and the T2 sequences demonstrated a

Address for correspondence: Prof. Edward Araujo Júnior,
Rua Belchior de Azevedo, 156 Apto. 111 Torre Vitoria,
São Paulo-SP, CEP 05089-030, Brazil.
E-mail: arauojred@terra.com.br

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“double bubble” sign and a distal duodenal obstruction. The colon appeared normal. The T1 sequence showed that the colon was filled with meconium, demonstrating a continuous communication of the intestinal lumen [Figure 2]. MRI findings supported the hypothesis of duodenal stenosis.

The pregnancy followed without complications, and the delivery occurred at the 38th week by cesarean section. The neonate had a body weight of 2980 g, a height of 46 cm, and Apgar scores of 9 and 10 at the 1st and 5th min, respectively. Breastfeeding was offered immediately after birth. On the 1st day of life, the neonate showed good tolerance to breastfeeding and normal stools. However, subsequent recurrent episodes of regurgitation required radiological studies to confirm obstruction and evaluate its degree. An abdominal radiograph was performed, which demonstrated the presence of gas in the distal intestines, including the colon. A fluoroscopic study of the upper gastrointestinal region demonstrated an important

distension of all duodenal segments, with an obstruction at the level of the 4th portion. 90 min postexamination, there was no passage of contrast to the jejunum [Figure 3]. A diagnosis of incomplete obstruction was confirmed by the presence of gas in the distal parts of the intestine. On the 5th day of life, the child underwent an enterectomy and was transferred to the intensive neonatal care unit [Figure 4]. After 1 year, the child presented normal physical and cognitive development.

DISCUSSION

CDO results from deficient intestinal canalization during embryonic life, occurring around the 7th week, and genetic predisposition is a suggested pathogenesis mechanism. Duodenal atresia is the most common duodenal malformation, with a prevalence of 0.9 per 10,000 live births.^[3] Although the incidence of duodenal stenosis remains unknown, intraluminal pathologies (duodenal webs) are rarer, with estimated

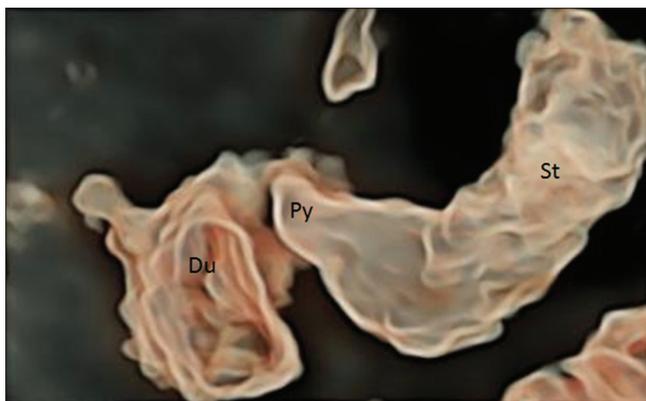


Figure 1: Three-dimensional ultrasound with the HDlive silhouette-rendering mode showing fetus at the 28th week of gestation presenting the “double bubble” sign. The image shows dilated stomach (St), pylorus (Py), and dilated duodenum (Du)

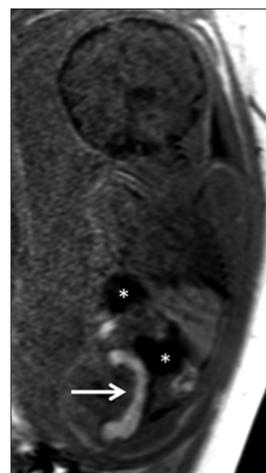


Figure 2: Fetal magnetic resonance imaging at the 32nd week of gestation at T1 sequence: “double bubble” sign (asterisks) and colon replete with the meconium (arrow)

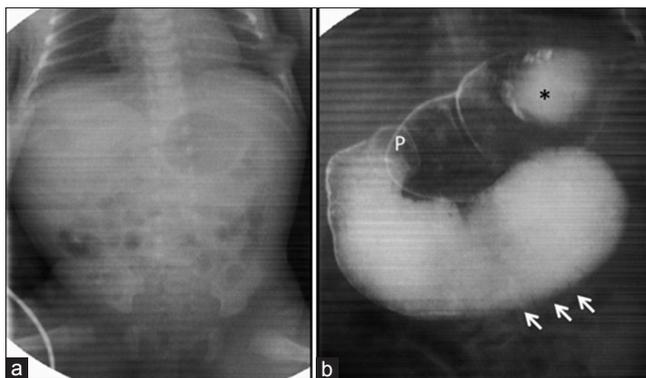


Figure 3: (a) Abdominal radiograph demonstrating two air-filled portions of the bowel in the region of the stomach and distal duodenum. The presence of distal air suggests incomplete obstruction. (b) Fluoroscopic upper gastrointestinal study demonstrates a normal stomach (asterisk), with an important distension of all duodenal portions (arrows) P: Pylorus. No contrast passed to the jejunum after 90 min of examination. The presence of gas in the distal bowel suggests the diagnosis of duodenal stenosis

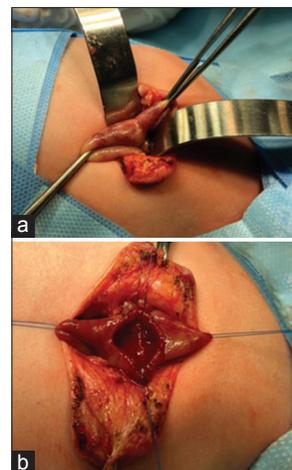


Figure 4: (a) Laparotomy demonstrating duodenal stenosis. (b) Enterostomy demonstrating partial obstruction and the absence of duodenal webs

incidences of 1 in 10,000 and 1 in 40,000 live births. However, in a recent study with 92 fetuses presenting the “double bubble” sign, the incidence of chromosomal abnormalities was 12%. This reduction in the incidence of image findings may be related to the development of 1st and 2nd trimester screening tests, early diagnosis, and pregnancy interruption.^[4]

In most cases, the diagnosis of CDO is made during the late 2nd trimester, with the mean gestational age of diagnosis ranging between 29 and 32 weeks, as revealed using conventional 2D ultrasound.^[5,6] Fetal MRI presents an important feature in intestinal abnormalities because in cases of obstruction, the meconium constitutes a natural contrast in T1 sequences, allowing fetal MRI to identify the distal bowel position and to evaluate malrotation pathologies, such as midgut volvulus and Ladd’s band.^[7] In the present case, MRI allowed to study the distal intestine, demonstrating duodenal permeability through the presence of meconium.

The HDlive silhouette-rendering mode is a novel technology that demonstrates structures with a vitreous-like clarity. Its capacity to visualize the inner structures resembles that of the hologram technology.^[8] This mode allows the assessment of spatial relationship among the stomach, duodenum, and jejunum and delineation of their outer contours, thereby enabling the diagnosis of proximal jejunal atresia.^[9] In the present case, the HDlive silhouette-rendering mode allowed a clearer visualization of the dilated stomach, pylorus, and dilated duodenum.

As previously described, in fetal intestinal obstructions, the 3D reconstruction can show the different parts of intestinal obstruction anatomy, and the visualization of the anatomical spatial relationships of the malformation became easier than 2D.^[9] Furthermore, the 3D images of the intestinal contour and its peristaltic movements, added by the HDlive silhouette-rendering mode, also assist in the evaluation of the site of obstruction and the exclusion of other differential diagnosis and complications.^[10]

Despite the common presence of chromosomal abnormalities and other malformations, CDO has an early postoperative survival rate of over 90%, which is mostly related to advances in operative techniques and neonatal intensive care. Late complications may occur in 12% of patients, with an associated mortality rate of 6%. The most common late complications are gastroesophageal reflux disease and duodenal pathologies related to reflux, motility, and megaduodenum. Complex congenital heart diseases are the most frequent causes of early and late postoperative mortality.^[11]

In summary, fetal intestinal obstructions can be analyzed and reconstructed in 3D using the HDlive silhouette-rendering mode and MRI, with both techniques playing an important role in the diagnosis and surgical planning of intestinal obstructive pathologies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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