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Prenatal diagnosis of intestinal pseudo-obstruction

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KEYWORDS: intestinal pseudo-obstruction; prenatal diagnosis; ultrasonography

ABSTRACT

The appearance of polyhydramnios and dilated bowel loops on prenatal sonographic examination usually implies mechanical obstruction. The prognosis is variable, depending on the etiology. Congenital pseudo-obstruction, a potentially lethal disease, comprises a group of disorders characterized by intestinal obstruction in the absence of an anatomic lesion. This report focuses on the prenatal diagnosis of intestinal pseudo-obstruction, and two cases of transient congenital intestinal pseudo-obstruction in one family are described. In both, the prenatal sonographic presentation was of small bowel obstruction. In one case there was postnatal suspicion of neurogenic bladder, and in the other there was unilateral hydronephrosis. The sonographic appearance of intestinal pseudo-obstruction is similar to that of mechanical obstruction. The clues to the prenatal diagnosis of pseudo-obstruction include associated urinary tract abnormalities and a family history of pseudo-obstruction. Copyright © 2007 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORTS

Case 1

A 28-year-old gravida 3, para 2 woman, was referred to our unit at 30 weeks' gestation with suspected fetal bowel obstruction. The patient and her husband were first cousins. Their medical histories were unremarkable and the woman had two healthy girls. Ultrasonography showed severe polyhydramnios with a deepest amniotic fluid pocket of 19 cm and multiple loops of dilated small bowel with visible peristalsis and no ascites or bowel-wall thickening. The scan was otherwise normal. Small bowel obstruction was suspected and the patient was counseled accordingly. She was offered and declined screening for cystic fibrosis. At 36 weeks' gestation she was admitted in labor, and a Cesarean section was performed on maternal request; the female neonate weighed 3145 g.

The neonate had a severely distended abdomen with multiple dilated loops of small bowel on plain abdominal X-ray. An explorative laparotomy was performed with a preoperative diagnosis of small bowel obstruction. On laparotomy, multiple dilated loops of small bowel were observed with no mechanical obstruction. Appendectomy and rectal biopsy were performed. These were both normal, with normal ganglion cells on pathological examination. During the immediate postoperative period the patient remained obstructed, the urinary bladder was persistently enlarged and a neurogenic bladder was suspected. No other urinary tract abnormalities were observed on postnatal investigation. With a diagnosis of primary congenital intestinal pseudo-obstruction (CIPO) and possibly megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) the patient was put on total parenteral nutrition and gastrointestinal prokinetic medication. The obstruction gradually subsided, as did the bladder dysfunction. Within a month the infant was discharged requiring no medication and is currently 7 years old and in excellent health.

Case 2

A 31-year-old gravida 5, para 4 woman – sister to the first patient – was referred to our unit at 30 weeks' gestation with polyhydramnios. Like her sister, she was married to a first cousin. She had previously given birth to four healthy children. On admission the ultrasound scan showed moderate polyhydramnios, the deepest amniotic fluid pocket being 13 cm. A detailed anatomy sonogram performed at this time revealed a normal stomach bubble and multiple hypoechogenic dilated small bowel loops (Figure 1). The dilated loops had a maximal diameter of 18 mm and visible peristaltic waves were recorded. Additionally, right pyelectasis was observed

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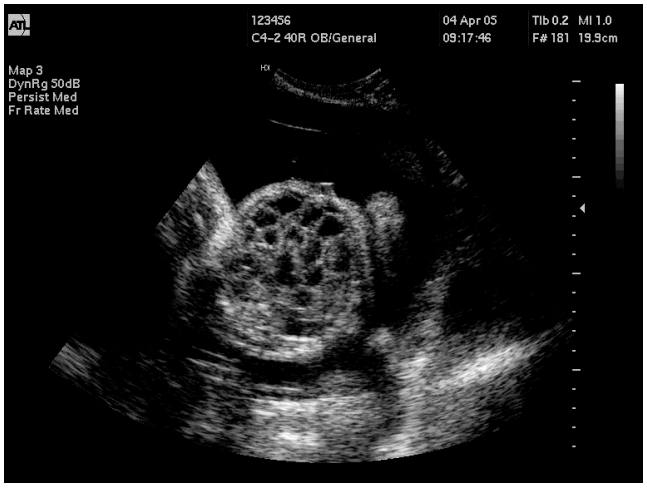

Figure 1 Case 2: prenatal sonogram at 30 weeks' gestation showing dilated bowel loops.

(anteroposterior diameter 11 mm) with an otherwise normal urinary tract. The mother was negative for the relevant cystic fibrosis mutations.

In view of the family history made available to us by the relatives, and considering the extensive intermarriage, we considered an antenatal diagnosis of intestinal pseudo-obstruction, counseled the couple accordingly, and decided on follow-up with no additional investigation. The fetal intestinal pathology remained unchanged on follow-up scans. At 37 weeks' gestation, after premature rupture of membranes, a male neonate was delivered weighing 3370 g. The baby had a distended abdomen, but otherwise appeared normal. A plain abdominal X-ray showed severely dilated small bowel. Conservative treatment was initiated, consisting of intravenous fluids and nasogastric drainage. Meconium was passed during the first postnatal day. The abdominal distention gradually subsided and eventually feeding through a tube was started. The baby was discharged at the age of 3 weeks with no signs of obstruction and in excellent health. The clinical diagnosis was transient CIPO. At 3 months of age ultrasound scan of the kidneys was normal.

DISCUSSION

Intestinal pseudo-obstruction comprises a group of disorders characterized by signs and symptoms of intestinal obstruction in the absence of an anatomic lesion[1]. Pseudo-obstruction may occur as a primary disease or may be secondary to a large number of conditions that may transiently or permanently alter bowel motility. Pseudo-obstruction represents a wide spectrum of pathologic disorders from abnormal myoelectric activity to intestinal neuropathy or myopathy. The organs involved may include the entire gastrointestinal tract or may be limited to certain components, such as the stomach or colon. In CIPO, abnormalities of the muscles or nerves can be demonstrated in most cases. Urinary tract involvement is not uncommon and may present as recurrent urinary tract infection or mimic obstructive symptoms. Clinical manifestation typically begins in the first few months after birth, and may range in severity from mild self-limiting disease, as seen in our cases, to a severe life-threatening form with high rates of infant mortality[2]. Treatment modalities include parenteral nutrition, motility agents, surgical decompression and bowel transplantation.

In utero, bowel obstruction may occur because of a large number of conditions. The most common etiology is mechanical obstruction most frequently due to ileojejunal atresia or cystic fibrosis-related meconium ileus. Functional, non-obstructive, dilated small bowel with polyhydramnios has been described in congenital chloride diarrhea, a rare autosomal recessive trait[3], in fetal paralytic ileus secondary to maternal benzodiazepam ingestion[4] and, without polyhydramnios, as a manifestation of transient fetal bowel ischemia[5]. The diagnosis of CIPO has been suspected in the context of MMIHS[6,7]. MMIHS is usually lethal during the first postnatal year, and can be suspected on the prenatal sonogram when a megabladder is demonstrated. Associated sonographic features may include dilated bowel loops[8], enlarged stomach and hydronephrosis[9]. Low amniotic fluid digestive enzyme levels may support the diagnosis. CIPO is potentially a severe and not infrequently lethal disease. In most cases the ultimate outcome is unpredictable, making the prenatal consultation difficult. This family presented with a mild form of CIPO with an excellent outcome. Having a high index of suspicion owing to a relevant family history may affect the prenatal consultation, as well as obviate the need for surgery, as happened in Case 2.

In both fetuses, the sonographic appearance of the bowel was no different from that found with mechanical obstruction. There was no bladder abnormality on the prenatal scan that might have suggested MMIHS. Furthermore, the obvious bowel activity might have suggested mechanical obstruction. Being aware of the possibility of pseudo-obstruction, we attempted in Case 2, on the prenatal sonogram, to characterize a possible dysfunctional pattern of peristalsis, different from that commonly observed in cases with mechanical obstruction. Such a pattern was not found. Other than the family history, one might consider the associated hydronephrosis a possible clue to the diagnosis, given the known association between bowel and urinary tract dysfunction. In the study of Heneyke *et al.*, 16/44 (36%) of the patients had coexisting urinary tract abnormalities, primarily obstructive uropathy[2].

The possibility of CIPO should be considered on prenatal evaluation when signs of intestinal obstruction are seen in association with urinary tract dilatation, and especially in the presence of a relevant family history. Without such a family history, a prenatal diagnosis of CIPO is unlikely.

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