

LATE THIRD TRIMESTER ULTRASOUND DIAGNOSIS OF DUODENAL ATRESIA - THE IMPORTANCE OF DETAILED PRENATAL ULTRASOUND SCREENING

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ABSTRACT:

Duodenal atresia (DA) is the commonest type of congenital small bowel obstruction usually presenting in the neonatal period. About half of fetuses with duodenal atresia have other associated anomalies, and these associations often contribute to morbidity and mortality. DA can be fatal unless promptly diagnosed and treated surgically. In experienced hands and in countries where prenatal ultrasound screening for anomalies is routine, DA can be confidently diagnosed prenatally, which can help in reducing the perinatal morbidity and mortality associated with diagnosis after delivery.

We report a case of DA diagnosed by ultrasonography at 36 weeks gestation in a 34 year old multiparous woman in a hospital where targeted prenatal ultrasound screening for fetal anomalies was recently introduced, and reviewed relevant literature

Keywords: Prenatal Ultrasound screening, Duodenal Atresia.

INTRODUCTION

Duodenal atresia (DA) is a congenital anomaly usually presenting in the neonatal period with upper gastrointestinal obstruction¹. Although detailed statistics are not available in much of Africa, the incidence of duodenal obstruction is reported to be 1 in 5,000 – 10,000 births². Duodenal atresia is the commonest cause of congenital small intestinal obstruction^{1,2}.

DA is associated with polyhydramnios in 45% of cases and with symmetric growth retardation in 50% of cases. Importantly, nearly 50% of patients with DA have associated congenital malformations,² of these, 30% have Trisomy 21, 22% have malrotation of the gut while 20-30% have congenital heart disease^{3,4}. The presence of associated anomalies and delay in diagnosis contribute to morbidity and mortality in children with neonatal intestinal obstruction. This is more obvious in developing countries, especially in those lacking formal birth defect screening programs.

The prenatal diagnosis of duodenal atresia can be made sonographically by the demonstration of the fluid filled dilated stomach and duodenum, which gives the typical “double bubble” sign on ultrasound³. Postnatally, the imaging diagnosis of duodenal atresia is usually first made on neonatal radiograph by demonstrating the same “double bubble” sign, which is due to a gas filled distended stomach and duodenal bulb⁵. Prenatal detection of DA helps in reducing the morbidity and mortality associated with late presentation^{5,6}.

We report a case of DA diagnosed prenatally by ultrasonography at 36 weeks of gestation in a 34year old multiparous woman who had corrective surgery within 48 hours of life.

CASE STUDY

A 34-year-old Gravida 4 Para 3 primary school teacher presented at the ultrasound unit of the Antenatal Clinic (ANC) of the University College Hospital (UCH) Ibadan for a routine third trimester ultrasound scan. She booked at the hospital at 26 weeks of gestation and she had two previous ultrasound scans before then



Figure 1: Prenatal B-mode transverse ultrasound of the fetal abdomen showing two fluid filled connecting fluid filled structures, which are the fetal stomach on the left (white arrow) and the duodenum on the right (black dotted arrow).



A



B

Figure 2A: Postnatal B-mode ultrasound scan of the baby's abdomen showing a fluid filled stomach (arrow) with low level echoes seen within it.

Figure 2B: Postnatal B-mode ultrasound scan of the baby's abdomen showing a fluid filled thick walled duodenum, which is blind ending

at private facilities, both of which revealed no obvious abnormalities. She has no child with birth defect. She was not a known hypertensive or diabetic and she had no known medical illness.

The ultrasound scan showed a single live intra uterine fetus in longitudinal lie and cephalic presentation. The

gestational age on ultrasound was 36 weeks and 2 days. The fetal abdomen (Figure 1) showed two fluid filled anechoic connecting structures in the upper abdomen. The larger structure on the left was seen in the region of the stomach, and it was seen to communicate with another blind ending cystic structure on the right suspected to be the duodenum (Figure 1). Other fetal biometry were within normal limits for gestational age. The amniotic fluid volume was within normal limits with amniotic fluid index of 15cm. A prenatal diagnosis of duodenal atresia was made and she was managed in a multidisciplinary care setting.

A male baby, with a birth weight of 3.2kg, was delivered per vagina at 39 weeks gestational age. The baby was noticed to have feed regurgitation on the first day of life for which a naso-gastric fluid was passed. Review of systems and physical examination did not suggest associated anomalies.

Abdominal ultrasound scan done showed dilated fluid filled thick walled stomach (Figure 2a) with visible peristaltic movement. This was seen to communicate with another blind ending tubular thick walled structure distally in keeping with the duodenum (Figure 2b). The remaining visualized intra-abdominal organs appeared within normal limits and no other anomaly was seen.

The plain abdominal radiograph (Figure 3) showed two oval shaped connecting lucencies in the central



Figure 3: A plain radiograph of same patient taken postnatally showing two centrally located connecting air-filled structures in the upper abdomen giving the characteristic "double bubble" appearance (arrows). There is paucity of bowel gas in the remaining parts of the abdomen.

abdomen giving the characteristic “double bubble” appearance. There was paucity of gas in the remaining parts of the abdomen. The bony skeleton and the soft tissues were within normal limits. A diagnosis of duodenal atresia was thus confirmed.

The patient was resuscitated and had a diamond duodeno-duodenostomy on the second day of life. He did well postoperatively, commenced oral intake on the fourth postoperative day and was discharged home on the eighth day after surgery.

DISCUSSION

Congenital duodenal obstruction is the result of several embryologic defects in foregut development, canalization or rotation. In addition, abnormal embryologic relationships between the duodenum and other structures in close anatomic proximity such as pancreas and portal vein may also lead to congenital duodenal obstruction.⁷ Ladd classified these lesions as either intrinsic or extrinsic. Intrinsic lesions include duodenal atresia, duodenal stenosis or duodenal web, whereas annular pancreas, malrotation, peritoneal bands and anterior portal vein are classified as extrinsic⁸. This patient presented with features of duodenal atresia, which is a type of intrinsic duodenal obstruction.

The presence of duodenal atresia may be suspected prior to the child's birth by prenatal ultrasonography⁹, which usually shows dilated stomach and duodenum giving the “double bubble” appearance. This was found in the index patient. Prenatal diagnosis of duodenal atresia helps to plan delivery, emergency resuscitation and prompt surgical intervention for the patient. This patient benefited from such intervention by receiving prompt surgical attention.

For every diagnosis of DA made, there is a 53% chance of the child having other congenital abnormalities, with an abnormal karyotyping seen in about 46% of patients, Down syndrome being the commonest.⁹ Fortunately, this patient had no other obvious congenital abnormality after birth. These were ruled out by physical examination and post natal abdominal scan. Unfortunately the patient did not have the benefit of karyotyping to rule out genetic abnormalities although the patient has not developed any dysmorphic feature to date.

At birth, newborns are usually well hydrated with adequate liver storage of glycogen. They may present with scaphoid abdomen and occasionally an epigastric mass may be noted due to dilated stomach and second part of the duodenum. However, the condition can deteriorate rapidly due to vomiting which usually starts

within the first 24 to 48hrs of life. The vomitus is bile stained in majority of patient and this is due to the fact that 80% of obstructions are located in the postampullary region of the duodenum.^{10, 11} By 72 hours of age, the ability of the baby to respond to stress gradually decreases. A prompt diagnosis of duodenal atresia is therefore important to prevent complications related to diagnostic delay, such as dehydration, electrolyte imbalance, or aspiration pneumonia¹⁰. There was no history of vomiting in this patient because a nasogastric tube was passed immediately after birth and he was placed on parenteral fluid until the diagnosis was confirmed.

Postnatal diagnosis of DA is achieved in most cases by plain abdominal radiographs, which demonstrate dilated stomach and duodenum, giving the characteristic appearance of a “double bubble” sign. No gas is observed beyond the second bubble in instances of complete obstruction; similarly noted in the index patient. In this setting, the plain film is sufficiently diagnostic so that no further imaging of the gastrointestinal tract is necessary^{5, 9}.

Barium enema examination is suggested as an adjunct study in the evaluation of duodenal atresia.⁹ In partial duodenal obstruction, a plain radiograph of the abdomen will show a double-bubble appearance but there is usually some air in the more distal intestine.⁶ The absence of this sign suggests that a patient has duodenal atresia. Early upper gastrointestinal contrast radiography is also indicated in these patients in order to establish the cause of incomplete duodenal obstruction.

Although duodenal atresia is a relative emergency, the infant should not be rushed to the operating room until he or she is hemodynamically stable. If the clinical history and findings on physical examination indicate that the baby is in no distress, and the radiograph is consistent with the usual presentation of duodenal atresia with no air beyond the second bubble, surgery should be performed within the first two days of life^{4, 5}. This patient had duodeno-duodenostomy on the second day of life.

Pre-operative management consists of nasogastric decompression and fluid and electrolyte replacement. Care is taken to preserve body heat and avoid hypoglycemia, since these newborn patients are often premature or small for date or have immature organ-systems. Pre-operative systemic antibiotics are administered prior to the start of the operation⁴. The operative management of duodenal atresia is determined by the anatomic findings and associated anomalies noted upon laparotomy. Bypass procedures

for duodenal atresia or stenosis include duodeno-duodenostomy and duodeno-jejunostomy^{2,11}. The patient presented had a diamond shaped duodeno-duodenostomy, which helps in avoiding postoperative narrowing of the duodenum.

Long-term outcome after repair of congenital duodenal atresia is excellent with contemporary operative survival exceeding 95% and with the majority of patients reported as asymptomatic and with normal growth^{10,11}. The main factors contributing to the mortality in patients with duodenal obstruction are high incidence of associated anomalies, delayed presentation, prematurity and low birth weight. The associated complex cardiac defects continue to be the leading cause of death particularly in infants with Trisomy 21¹¹. However, recent reviews document that advances in both pediatric cardiology, and cardiac surgery in neonates and infants have reduced this mortality significantly¹¹.

CONCLUSION

Duodenal atresia is the commonest cause of congenital small intestinal obstruction and the condition could be fatal if diagnosed postnatally. Prenatal ultrasound screening is of great importance in its early diagnosis, thus reducing the morbidity and mortality associated with late presentation.

REFERENCES

1. **Chirdan LB**, Ngiloi PJ, Elhalaby EA. Neonatal surgery in Africa. *Semin Pediatr Surg* 2012; 21(2): 151–159.
2. **Badrinath R**, Kakembo N, Kisa P, *et al.* Outcomes and unmet need for neonatal surgery in a resource-limited environment: Estimates of global health disparities from Kampala, Uganda. *J Pediatr Surg*. 2014; 49(12):1825–1830.
3. **Osifo OD**. Duodenal Obstruction: Etiology, morbidity and mortality among Edo State Children. Nigeria. *Afr J Biomed Res*. 2009; 12(3):193–197.
4. **Sekmenli T**, Koplay M, Alabalik U, Kivrak AS. Duodenal atresia and hirschsprung disease in a Patient with Down Syndrome. *Eur J Gen Med* . 2011; 8(2):2–45.
5. **Chirdan LB**, Uba AF, Pam SD. Intestinal atresia: Management problems in a developing country. *Pediatr Surg Int*. 2004; 20(11-12):834–837.
6. **Pariente G**, Landau D, Aviram M, HersHKovitz R. Prenatal diagnosis of a rare sonographic appearance of duodenal atresia: report of 2 cases and literature review. *J Ultrasound, Med* 2012; 31 (11):1829–1833.
7. **DeLa'O CM**, Newton CR, Stehr W. Duodenal atresia with major duodenal papilla within duodenal atretic segment: A case report. *J Pediatr Surg Case Reports*. 2014; 2(5):261–263.
8. **Nagra S**, Cama JK. Pyloric atresia in a healthy newborn - Two stage procedure. *J Pediatr Surg Case Reports*. 2014; 2(1):12–14.
9. **Jagatiani N**, Ariwala N, Shan J, *et al.* Antenatal diagnosis of duodenal atresia. *Gujarat med J*. 2009; 64(2):77–78.
10. **Osifo OD**, Ovueni ME. Challenges in the management of neonatal surgical conditions under the absence of total parenteral nutrition. *Saudi med J*. 2009; 30(7):971–973.
11. **Mandhan P**, Rathod KJ, Sankhla D. Late Presentation of a Congenital Intrinsic Duodenal Obstruction in a Patient with Anorectal Malformation. *Surg Sci*. 2013; 4(7):329–331.