Reminder of important clinical lesson

Can you be too premature to develop pyloric stenosis?

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Summary

Diagnosis of Pyloric stenosis is rare in preterm babies. A case of pyloric stenosis diagnosed at 33⁺⁶ postmenstrual age is reported. To our knowledge, this is the youngest postmenstrual age in the literature for a preterm infant to have a confirmed diagnosis of pyloric stenosis. The report also discusses issues that could delay the diagnosis of this condition in this patient population.

BACKGROUND

Pyloric stenosis is rarely diagnosed in premature babies. While preterm infants can subsequently develop pyloric stenosis, the clinical presentation is frequently around fetal maturity and is often delayed and atypical.¹ This report discusses the details of a preterm baby who was diagnosed with this condition during the third week of life at a postmenstrual age of 33⁺⁶ weeks. This, to our knowledge, is the youngest postmenstrual age at the diagnosis of confirmed pyloric stenosis reported in the literature. The report also looks at some of the factors that may delay the diagnosis of this condition, and emphasises on the importance of considering pyloric stenosis in the differential diagnoses of persistent vomiting in preterm infants.

CASE PRESENTATION

A male infant was born at 30⁺⁶ weeks gestational age and weighed 1640 g, following an emergency caesarean section for ante-partum haemorrhage. The baby was born in a good condition and did not require respiratory support.

The parents were non-consanguineous Caucasians and had two elder male children, both born at term. One of the siblings had a pyloromyotomy at 6 weeks of age for pyloric stenosis and the other sibling had duplex kidneys. Feeds were started with expressed breast milk and Cow and Gate Nutriprem1 (Nutricia Ltd, Wiltshire, UK) on the first day of life and were gradually increased to 180 ml/kg by the seventh day. He was also started on vitamins and phosphate supplementation according to the local protocol. Apart from occasional possetting and vomiting (about one episode/day), there were no feeding concerns initially and he gained weight satisfactorily. By day 19, he developed recurrent early postprandial vomiting. Feed intolerance was suspected, and the volume of feeds was reduced initially, followed by changing the milk type to Neocate (Nutricia Ltd). Apart from these measures, no medications were given for his symptoms. On the 21st day of life (33⁺⁶ weeks of gestation), clinical examination revealed a 'tumour' in the typical position. Capillary gas analysis at this stage showed hypochloraemic metabolic alkalosis. Ultrasound of the abdomen confirmed pyloric stenosis,

Figure 1 Ultrasound image of hypertrophied pylorus. Long arrows indicate the length of pyloric canal and short arrows indicate the thickness of pylorus.

with pyloric muscle thickness of about 3 mm and length of 14 mm (figure 1).

He was subsequently transferred to the regional paediatric surgical centre on day 22 of life. The diagnosis was confirmed here clinically and sonographically (pylorus measured 4 mm in thickness and 16 mm in length on repeat ultrasound imaging). A standard 'open' Ramstedt pyloromyotomy was performed on the 23rd day of life. The procedure was uneventful. Enteral feeds were restarted on the day after surgery and gradually increased in volume. He was transferred back to the referring hospital on day 24 of life. He was discharged home on 43rd day of life on full bottle feeds and demonstrated good weight gain.

INVESTIGATIONS

Abdominal ultrasound scan.

TREATMENT

Pyloromyotomy.

DISCUSSION

Late (term or post-term) occurrence of pyloric stenosis in infants born prematurely is well recognised, and accounts for a reported incidence of up to 10%, of total cases, in a large series.² It appears that the development of pyloric stenosis in preterm infants requires a certain degree of maturation of the gastrointestinal tract.³ In general, preterm babies tend to become symptomatic during the fifth week of life, in contrast to the onset during the third week in term babies.¹ On analysis of a cohort of 214 infants with pyloric stenosis, it was observed that the mean age of symptom onset in preterm babies was 32.11+/-4.47 days, but the diagnosis was made at a mean age of 45.94 days.³ To our knowledge, the earliest post-menstrual age reported for a preterm infant to have a confirmed diagnosis of pyloric stenosis is greater than 35 weeks.⁴ In the above clinical scenario, the diagnosis was established clinically and sonographically at 33 postmenstrual weeks of age. This was later confirmed intraoperatively.

Several studies have attempted to characterise the differences between preterm and full-term infants with pyloric stenosis.¹ Most notably, preterm infants may not display projectile vomiting, visible gastric peristalsis or metabolic alkalosis.³ This, along with the following factors, may contribute to a delay in the diagnosis of this condition. (1) Feeding intolerance manifesting as recurrent emesis with or without abdominal distension is often attributed to gastro-oesophageal reflux, 'immaturity' of the gut or causes other than pyloric stenosis.^{5 6} (2) The non-specific nature of abdominal x-ray findings: gastric dilatation on abdominal x-rays is often observed during mechanical ventilation and placement of nasogastric tube,² thus causing confusion. (3) Absence of age-adjusted ultrasonic diagnostic criteria for preterm infants.¹

The results of previous population-based cohort studies suggest that pyloric stenosis is an inherited disease with strong familial aggregation.⁷ Analysis of a large series of preterm infants with pyloric stenosis showed a family history in 5.56% cases.³ There was a high index of suspicion in this case from the onset of symptoms because of the positive family history. However, with initial absence of weight loss, clinical findings and abnormal biochemistry, a trial of change of formula and reduction of feed volume was wise to exclude the relatively common so-called 'gut immaturity syndrome'. Persistence of symptoms and subsequent evolution of a palpable pyloric mass led to sonographic confirmation of the diagnosis.

Proposed risk factors of pyloric stenosis include birth rank, gender, social class, seasonal variation, maternal smoking, feeding practices, birth-weight and blood group.⁸ Erythromycin, an antibiotic commonly used for treating pertussis in infants, has also been implicated in the causation of pyloric stenosis. Infants exposed to antimicrobial doses of erythromycin in the first few weeks of life appear to be at a greater risk for developing hypertrophic pyloric stenosis.⁹ The risk was reported to be substantially higher (8-fold to 10-fold) in the first 2 weeks of life, in term or near-term infants. It has been hypothesised that erythromycin, through its ability to interact with motilin receptors, can induce strong gastric and pyloric bulb contractions, resulting in pylorus hypertrophy.

At present, there are two valid and successful methods for the imaging diagnosis of infantile hypertrophic pyloric stenosis—sonography and upper gastrointestinal contrast study. With either of these modalities, the diagnosis is made unequivocally by the demonstration of a hypertrophied pylorus.¹⁰ In general, a pyloric muscle thickness of 4 mm or above and a pyloric length of 14 mm or above are used as accepted criteria for the diagnosis of infantile pyloric stenosis in term babies.¹¹ Two factors should however be considered while interpreting the results; first, pyloric stenosis is an evolving condition and the pyloric measurements may vary with time, as was demonstrated in our case. Second, pyloric dimensions in preterm infants with pyloric stenosis are found to be significantly less, compared with their term counterparts.¹² Pyloric stenosis has been diagnosed in preterm babies with a pyloric muscle thickness of 2–2.4 mm, in upper gastrointestinal contrast studies.¹ It is thus not surprising that an analysis of a cohort of infants with pyloric stenosis observed a higher percentage of diagnoses made by contrast study among preterm babies, when compared with term babies.³

In patients with pyloric stenosis, there is a failure of relaxation of the prepyloric antrum, typically described as 'elongation' of the pyloric canal.¹⁰ As is the case on upper gastrointestinal contrast study, real-time sonographic observation for a few minutes usually identifies this finding.¹³

Patients in whom the antropylic canal relaxes to a normal morphology do not have pyloric stenosis. It has been recommended that patients in whom the muscle is 2–3 mm thick and does not relax throughout the examination warrant careful monitoring and follow-up examination, particularly if they are at the younger end of the age spectrum at the time of presentation.^{14 15} A contrast study should be considered for cases where there is a high index of clinical suspicion but inconclusive ultrasound scan findings.

This case occurred at what appears to be the earliest postmenstrual age at which a preterm baby has had a confirmed diagnosis of pyloric stenosis. Pyloric stenosis should be considered as a possible diagnosis in preterm neonates with persistent vomiting and 'feed intolerance'. A contrast study may be required in order to confirm the diagnosis in this group of patients if the sonographic findings are inconclusive.

Learning points

- Pyloric stenosis should be considered as a possible diagnosis in preterm neonates with persistent vomiting and 'feed intolerance'.
- A contrast study may be required in order to confirm the diagnosis in this group of patients if the sonographic findings are inconclusive.

Competing interests None.

Patient consent Obtained.

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