# Hypertrophic pyloric stenosis in a preterm infant

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Hypertrophic pyloric stenosis is one of the most common conditions requiring surgery in the first months of life. The etiology is unknown but several risk factors have been described, for example, male sex with a preponderance of 5:1, maternal smoking during pregnancy, low birth weight and prematurity (1). The incidence of hypertrophic pyloric stenosis is 2 to 5 per 1000 births per year in the Western world (2).

Hypertrophic pyloric stenosis in term infants presents with typical clinical signs including projectile non-bilious vomiting following feeds, a palpable olive-like epigastric mass, visible peristalsis and dehydration with metabolic abnormalities (hypochloremic and hypokalemic metabolic alkalosis) (2). These classical symptoms are uncommon in infants born prematurely. Most cases are diagnosed between 2 and 11 weeks of age, with a median age at diagnosis of 3 to 5 weeks for term babies and 6 weeks for preterm babies (1-3). Pyloric stenosis is treated by pyloromyotomy and has a good prognosis (4).

A preterm male infant was born by Caesarean section to a 40-year-old G2/P2 at 32 weeks of gestation. The pregnancy had been uneventful until shortly before delivery when the mother developed preeclampsia. The mother had been treated for depression throughout pregnancy with quetiapine, trazodone hydrochloride and a herbal sedative (Redormin©).

Apgar scores were 9, 9 and 10 at 1, 5 and 10 minutes, respectively. Birth weight was 1470 g (P10), length 40 cm (P10) and head circumference was 30 cm (P40). The baby was admitted to an intermediate care ward. Enteral feeds were started on the first day of life and supplemented withintravenous dextrose and amino acids. On the second day of life, he had bilious gastric residuals, probably secondary to delayed meconium passage. Following a rectal enema with NaCl 0.9%, he had no further problems. Intravenous fluids could be stopped after 8 days and the clinical course was unremarkable with good weight gain.

On day of life 28, at a corrected gestational age of 36 weeks, he developed gastric residuals and repeated episodes of non-bilious emesis. The baby was not ill appearing, well hydrated and had stable vital signs. The clinical examination was normal apart from mild abdominal distension but no mass was palpable. Laboratory findings were all normal including electrolytes and blood gas analysis. An abdominal ultrasound showed hypertrophy of the pylorus with a pyloric muscle thickness of 3.5 mm (upper limit of normal 3 mm) (Fig. 1) (5), a pyloric length of 14 mm (upper limit of normal 11.7 mm) (Fig. 2) (5) and the target appearance of the pylorus showed an external diameter of 8.6 mm (Fig. 3). The abdominal radiograph showed a distended stomach bubble, but no other signs of mechanical obstruction (Fig. 4).

Enteral feeds were stopped and the pediatric surgeons were consulted. A pyloromyotomy was performed the following day and a white glistening pyloric mass was found intraoperatively; the width of the pyloric musculature was 3.5 mm on both sides (Fig. 5). Nasogastric feeds were restarted on the first postoperative day and could be advanced to full feeds over the next 7 days. No further episodes of projectile vomiting occurred and the patient was transferred to another hospital for further care on the 7th postoperative day.
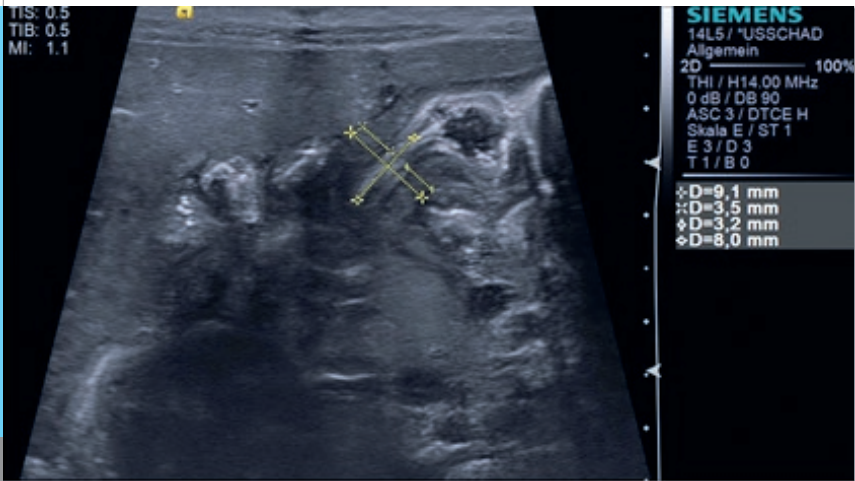
*Ultrasonography of the pylorus: hypertrophic pyloric wall with a  thickness of 3.5 mm.*
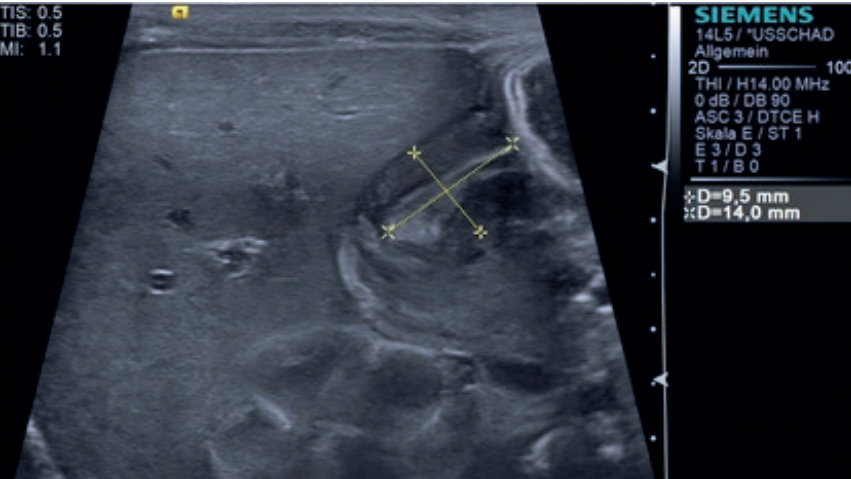
Fig. 2

*Ultrasonography of the pylorus: hypertrophic pyloric wall with a length of 14 mm, and external diameter 9.5 mm.*
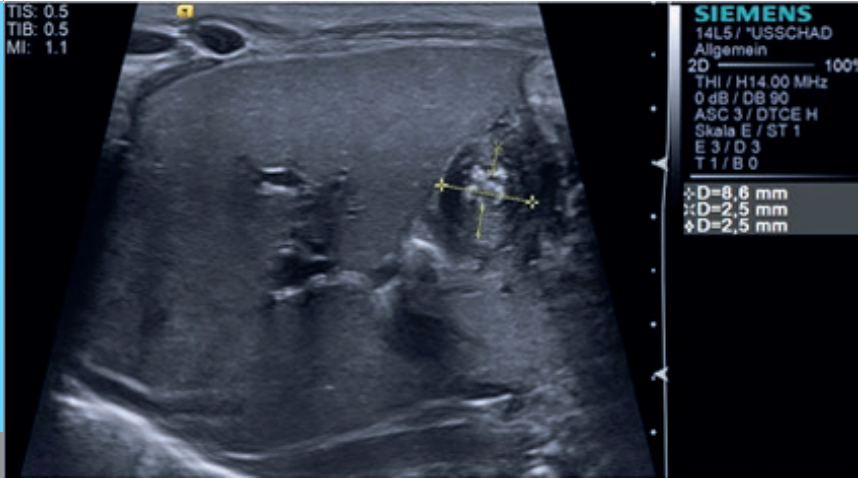
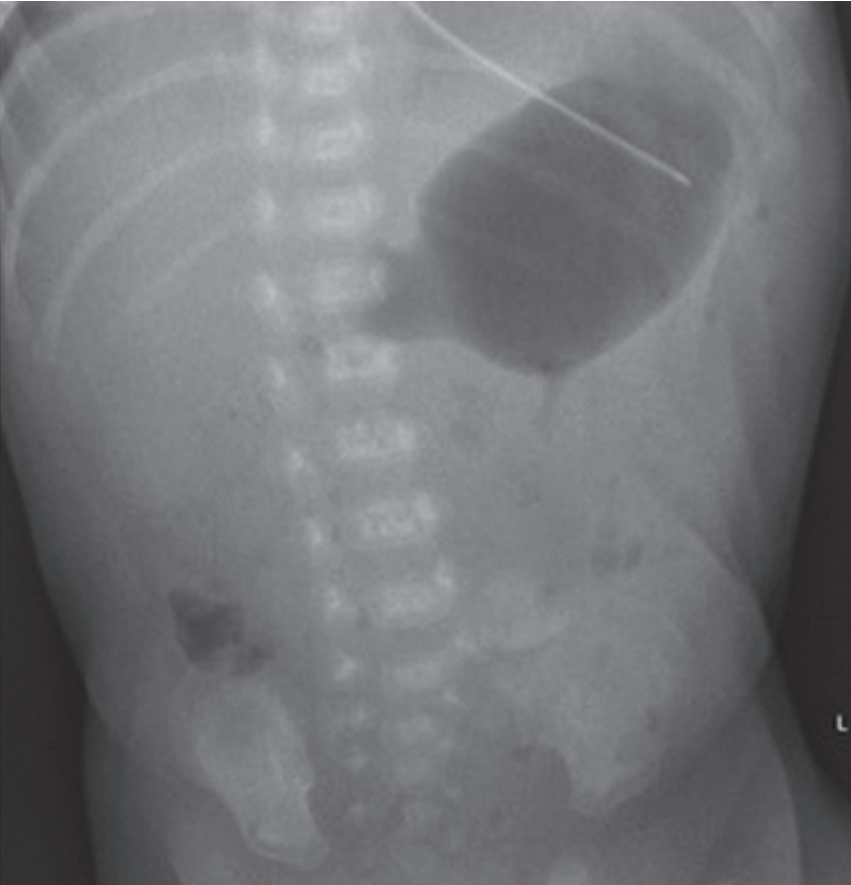*Ultrasound of the pylorus with a target sign of the pylorus.*

*Plain abdominal X-ray: distended stomach bubble.*

Fig. 4

*Intraoperative appearance of the pyloric mass.*

The first case of pyloric stenosis in a baby was reported in 1627 by Fabricius Hildanus (4). The first description of a preterm infant with hypertrophic pyloric stenosis appeared in 1907 (6). The presentation of hypertrophic pyloric stenosis is often atypical in preterm infants. Between 3.1% (7, 8) and 19% (2) of patients diagnosed with hypertrophic pyloric stenosis are preterm infants.

The etiology of hypertrophic pyloric stenosis is unknown but many risk factors have been described, such as a positive family history in up to 15% of cases, male sex, first-born, smoking during pregnancy, low birth weight and prematurity (1). Transpyloric feeding has also been reported as a possible cause (9). Early and prolonged (> 14 days) erythromycin therapy has been implicated in certain studies but this association has not been confirmed in subsequent reviews (10, 11). In our patient, several risk factors were present: maternal smoking, low birth weight, prematurity and male sex.

The onset of symptoms in a premature infant is usually delayed compared to a term infant, probably reflecting postmenstrual age (2). Symptoms can be atypical and it is important to consider hypertrophic pyloric stenosis in the differential diagnosis of emesis. Increased awareness of the condition and the advent of ultrasound diagnosis may lead to earlier diagnosis before the classic signs and metabolic decompensation appear (2).

Abdominal ultrasound is useful for confirming the diagnosis. There are well-established values for muscle thickness, length and width of the pylorus for term infants (12). For preterm infants, it is more difficult to find reference values. Haider et al. showed that pyloric length correlates with weight and not the age of the infant (5). In our case, pyloric length and muscle thickness were above the normal values expected for the baby's weight.

The definitive treatment of hypertrophic pyloric stenosis is pyloromyotomy or longitudinal splitting of the pyloric muscle, usually done via a periumbilical approach and often laparoscopically (4). The short and long-term outcomes are excellent.

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