Pediatr Surg Int (1997) 12: 497–500

ORIGINAL ARTICLE

A. J. A. Holland · J. K. Freeman · G. W. Le Quesne · T. Y. Khong

Idiopathic focal foveolar hyperplasia in infants

Accepted: 26 September 1996

Abstract Idiopathic focal foveolar hyperplasia (FFH) is a rare cause of gastric outlet obstruction in infants. We present two cases, including the first reported association with infantile hypertrophic pyloric stenosis. The diagnostic ultrasound appearance of FFH is described. Optimal treatment is surgical excision of the involved mucosa with pyloroplasty or pyloromyotomy.

Key words Focal foveolar hyperplasia · Infant
Infantile hypertrophic pyloric stenosis · Ultrasound scan

Introduction

Non-bilious vomiting in infants is a common clinical problem for which a surgical opinion may be sought. In cases secondary to gastric outlet obstruction (GOO), the most frequent surgical pathology is infantile hypertrophic pyloric stenosis (IHPS). Other lesions much less frequently encountered include gastric antral web, pyloric atresia, and gastric duplication cysts.

We describe two patients with non-bilious vomiting secondary to idiopathic focal foveolar hyperplasia (FFH). This condition results in an elongated mucosal polyp of the antrum and pyloric canal, producing partial GOO. The presentation and management of this condition are reviewed, together with the diagnostic ultrasound (US) appearance.

A. J. A. Holland · J. K. Freeman (✉)
Department of Paediatric Surgery,
Women's and Children's Hospital, 72 King William Road,
North Adelaide, SA 5006, Australia

G. W. Le Quesne
Department of Paediatric Ultrasound,
Women's and Children's Hospital, 72 King William Road,
North Adelaide, SA 5006, Australia

T. Y. Khong
Department of Histopathology, Women's and Children's Hospital,
72 King William Road, North Adelaide, SA 5006, Australia

Case reports

Case 1

A 6-week-old male was admitted initially for investigation of intermittent upper airway obstruction. He was conceived by in vitro fertilisation and had been delivered by elective Caesarean section after an otherwise normal pregnancy. Following delivery, he was noted to have a cleft palate and micrognathia suggestive of Pierre-Robin syndrome. The patient experienced feeding difficulties from birth, requiring supplementary nasogastric feeds of expressed breast milk to ensure adequate nutrition. A polysomnogram indicated an obstructive pattern with multiple episodes of regurgitation. The obstruction was satisfactorily treated with a nasopharyngeal airway. Despite satisfactory weight gain, the infant's regurgitation became increasingly severe and was not relieved by cisapride. Twenty-four-hour ambulatory pH monitoring indicated a moderate frequency of gastro-oesophageal reflux (GOR), with a reflux index of 3.6%. In view of this low index but continuing unsatisfactory oral intake, an open gastrostomy was performed at the age of 10 weeks.

Following this procedure, the frequency of vomiting increased with an associated fall in weight gain. A complete blood picture (CBP) was normal, and screening for cytomegalovirus (CMV) infection was negative. A repeat ambulatory pH study showed a reflux index of 16.4% with a close correlation between symptoms of irritability and reflux. In view of the continued severe vomiting, a barium meal was performed at the age of 14 weeks. This revealed a prolonged delay in the passage of contrast from the stomach to the duodenum and a longitudinal filling defect within an elongated pyloric canal (Fig. 1). An US scan indicated a linear redundancy of the mucosa of the distal antrum and pylorus, which folded upon itself to almost completely occlude the pyloric canal (Figs. 2–4). There was no evidence of muscle hypertrophy. The diagnosis of FFH producing GOO and exacerbating GOR was made.

Upper gastrointestinal (GI) endoscopy showed no evidence of oesophagitis, but confirmed the US diagnosis of FFH. At operation, the external appearance of the pylorus was normal, although the muscle layer felt slightly thickened. The duodenum was mobilised and the pylorus opened via a longitudinal incision to reveal a prominent mucosal fold. This was excised and the mucosal layer repaired. A formal pyloroplasty and Nissen fundoplication were performed. The patient made a satisfactory recovery and was discharged tolerating normal feeds 14 days after surgery. Histopathology of the mucosal fold showed elongated and cystically dilated foveolae in keeping with the diagnosis of FFH. Twelve months after surgery the patient has had no recurrence of pyloric obstruction.

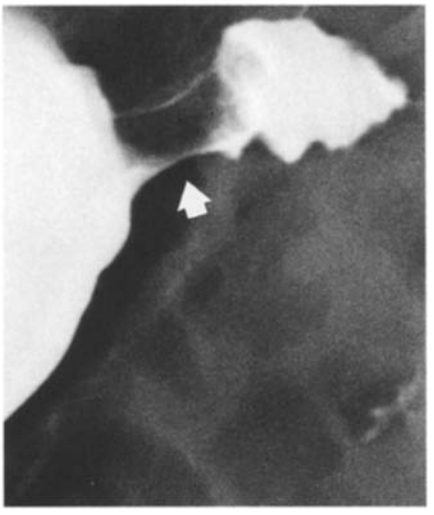


Fig. 1 Barium meal showing delayed gastric emptying with a longitudinal filling defect (*arrow*) within elongated and distorted pyloric canal

Case 2

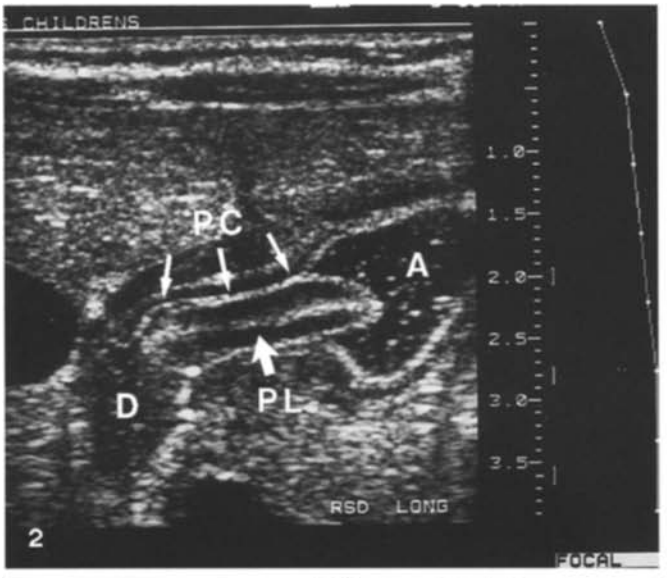
A 13-week-old girl presented with a 4-week history of intermittent, effortless projectile vomiting following an upper respiratory tract infection. The vomiting affected 2 to 3 feeds per day and was not bile-stained. Despite the frequency of vomiting, satisfactory weight gain had been maintained. Abdominal examination was unre-

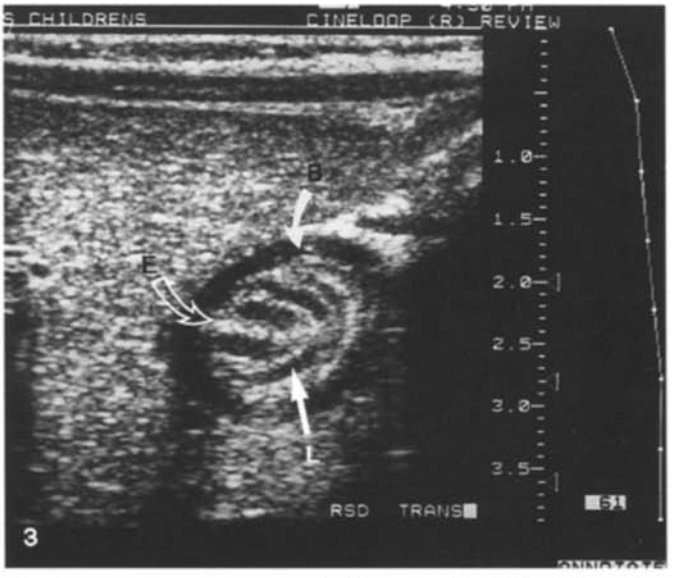
Fig. 2 Longitudinal US section of gastric antrum and pyloric canal. 'Sausage-shaped' polypoidal lesion (*PL*) with central echogenic fold extends from antrum (*A*) to duodenal cap (*D*), occluding elongated pyloric canal (*PC*)

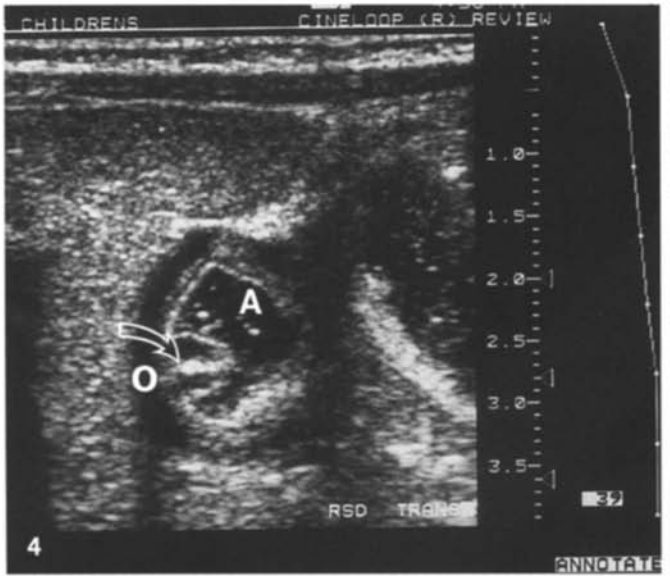
Fig. 3 Transverse US section through mid-pyloric canal showing multilayered 'horseshoe' pattern of redundant mucosa with central echogenic fold (*E*), compromised lumen (*L*), and normal-thickness bowel wall (*B*)

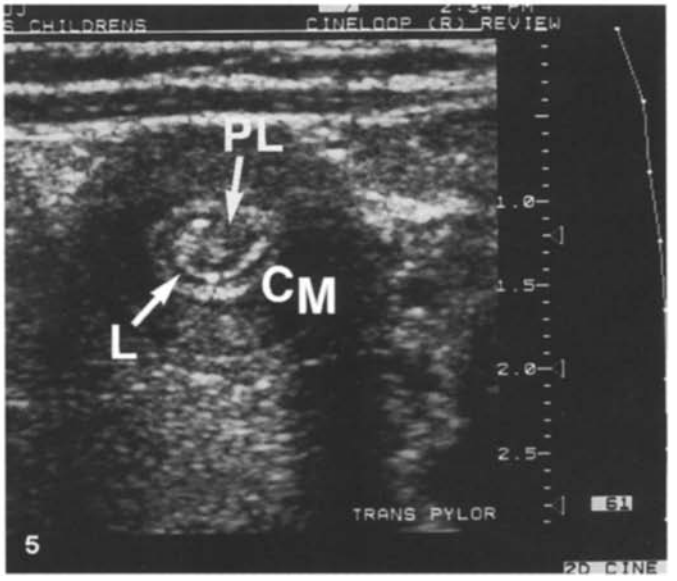
Fig. 4 Transverse US section through distal gastric antrum demonstrates proximal origin of elongated mucosal redundancy (*O*), which arises in distal antrum (*A*) and extends into pyloric canal. *Arrow* indicates central echogenic fold

Fig. 5 Transverse US section through pyloric canal. Polypoidal lesion (*PL*) of redundant mucosa significantly reduces lumen (*L*) and demonstrates characteristic horseshoe pattern of FFH. Significant hypertrophy of circular muscle of pylorus (*CM*), consistent with IHPS









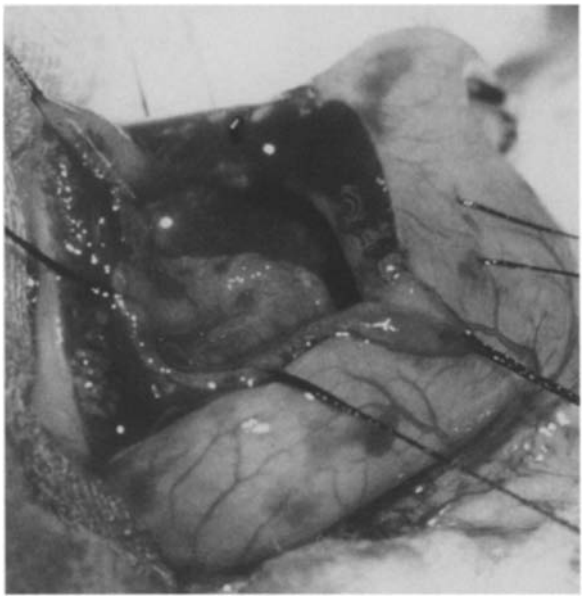


Fig. 6 Intraoperative photograph. Pyloromyotomy has been performed and anterior mucosa incised longitudinally to reveal polypoidal lesion of FFH

markable. The infant had been delivered by emergency Caesarean section at 36 weeks' gestation for pre-eclampsia and had been breast-fed since birth. A CBP was normal, with no evidence of peripheral eosinophilia or acute viral infection. US revealed active peristalsis in the stomach, but very little forward passage of stomach contents into the duodenum. The pyloric canal was elongated, with redundant mucosa giving rise to an elongated filling defect within the lumen diagnostic of FFH. In addition, there was hypertrophy of the circular muscle of the pylorus (muscle thickness 4.0 mm) suggestive of an associated IHPS (Fig. 5). Upper GI endoscopy revealed marked oesophagitis and polypoidal mucosal hypertrophy in the gastric antrum, extending into the pyloric canal.

At operation, a moderate-sized pyloric 'tumour' was found. A pyloromyotomy was performed and the anterior mucosa incised to reveal a polypoidal lesion projecting into the pyloric canal (Fig. 6). The lesion was excised and the resultant posterior mucosal defect repaired. The anterior mucosal incision was closed longitudinally, but the muscular layer was not approximated. The patient made a good recovery and was discharged tolerating normal feeds 3 days after surgery. Histopathology indicated foveolar hyperplasia with a sparse eosinophilic infiltrate within the lamina propria, consistent with a diagnosis of FFH (Fig. 7). The patient has remained well with no further feeding difficulties 6 months after surgery.

Discussion

Idiopathic infantile FFH is a rare condition. The first clearly reported case was described in 1985 in a 7-week-old male who presented with persistent vomiting and failure to thrive [3]. Since then, there have been only two further idiopathic cases clearly identified in the English literature [6, 10]. Histologically, the condition is characterised by enlargement of the gastric pits or foveolae of the distal antrum and pylorus, which become tortuous and dilated [3, 9]. The mucosal thickening that results leads to the development of an unusual polyp, which causes incomplete GOO and is responsible for the characteristic US features of this condition. The redundant mucosa produces a sausage-shaped structure (Fig. 1) that arises from a narrow, elongated base, commencing in the distal portion of the antrum (Fig. 3) and extending through an elongated pyloric canal to the base of the duodenal cap. The double layer of redundant mucosa bulges into the lumen, partially or completely occluding the pyloric canal. It contains an echogenic central fold where the two layers of mucosa are opposed. In cross-section a multi-layered horseshoe pattern is seen, the open side of the horseshoe representing the site of linear attachment of the polyp (Fig. 2). This pattern was evident in both of our cases and is suggested in the only other published US image [6]. The pattern appears to be characteristic.

The aetiology of infantile FFH is unknown and may represent an abnormal response to earlier injury [6]. The finding of a submucosal eosinophilic inflammatory reaction in a proportion of cases might suggest an allergic

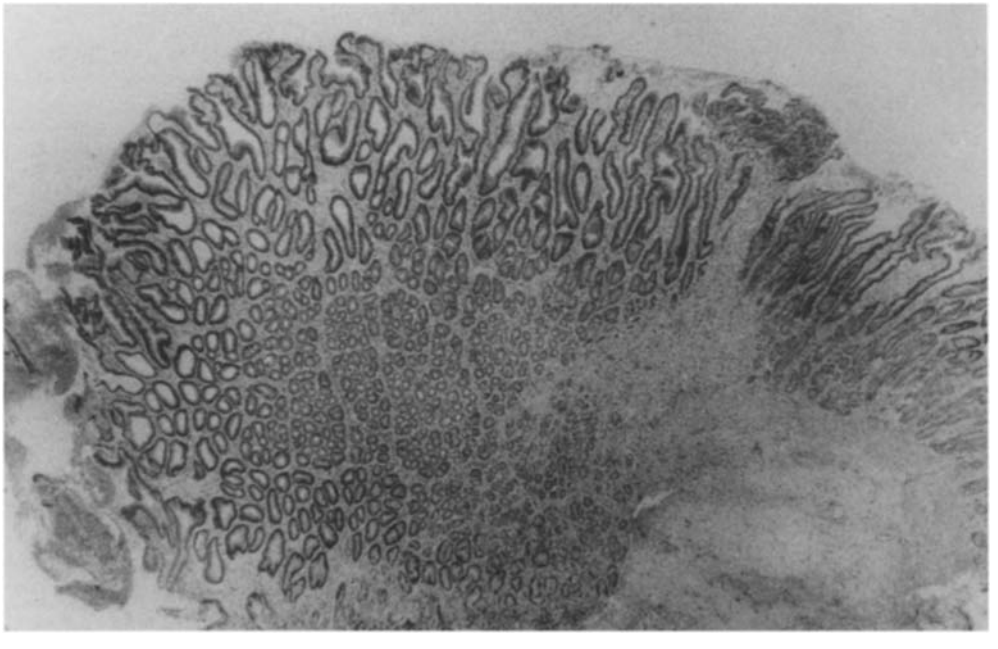


Fig. 7 Photomicrograph of FFH. Polypoidal mucosa with elongation and focal cystic dilatation of foveolar portion of mucosa

reaction, although neither of our patients received formula milk prior to the onset of symptoms [3]. There have been a number of cases of FFH related to treatment with prostaglandin E_1 used to maintain patency of the ductus arteriosus in infants with cyanotic congenital heart disease [7, 9]. This seems to be related to the duration of treatment and cumulative dose, and may resolve on cessation of the drug [9].

Infantile FFH shares some of the histological features of Menetrier's disease (MD), namely, foveolar hyperplasia in association with an eosinophilic inflammatory submucosal infiltrate [2, 5]. While the aetiology of MD is also poorly understood, a similar infective or allergic phenomenon is thought to be the most plausible explanation in children [2]. Certainly, MD has been described in a young child with acute CMV infection, although this association has not been described in FFH [4]. There are, however, a number of key differences. The hallmark of MD, marked gastric mucosal hypertrophy in association with a protein-losing enteropathy, is absent in FFH [13]. The antrum and pylorus, the site of the lesion in FFH, are typically spared in MD, which is a more generalised condition of the stomach [2, 13]. Although MD seems to run a more benign course in children than in adults, with spontaneous resolution in many cases, this favourable outcome without surgical intervention has only been reported in prostaglandin-induced FFH [2, 9, 13]. Further, MD usually seems to affect older children [2]. There has only been one case involving an infant, an 8-week-old male born to a consanguineous couple [5].

The finding of an eosinophilic inflammatory response might suggest the alternative diagnosis of eosinophilic gastroenteropathy (EG) [11]. This rare condition is characterised by the association of GI symptoms with an eosinophilic infiltration of the GI tract, in the absence of parasitic infection or extraintestinal disease [12]. EG may involve any part of the GI tract, but the duodenum, proximal jejunum, and stomach are most commonly involved [14]. Whilst adults seem to be more frequently affected, there have been at least ten children reported with EG who experienced GOO secondary to polypoidal mucosal enlargement [8, 15]. There are a number of important features, however, which usually allow FFH and EG to be distinguished from each other. EG is frequently but not invariably associated with a peripheral eosinophilia, which has not been reported in FFH [1, 12]. The presentation of EG is typically with abdominal plain, nausea, diarrhoea, and weight loss rather than GOO [14, 15]. Infants are rarely affected; in the single case reported in this age group the mucosal polyp projected through the pylorus from the duodenal bulb rather than from the antrum as in FFH [8]. Finally, GOO that occurs in EG is usually secondary to involvement of the muscular layer: there may be secondary mucosal oedema, but foveolar hyperplasia is not seen [11].

A formal pyloroplasty and Nissen fundoplication, in addition to excision of the FFH, were performed in our first patient because of the importance of ensuring effective gastric feeding, and therefore optimal growth, in the presence of Pierre-Robin syndrome. Excision of the hyperplastic polyp and pyloromyotomy alone in the absence of IHPS might not have achieved this aim. The relative contribution to GOO of IHPS and FFH seen in our second patient is difficult to determine. This association has not previously been documented. If a pyloric 'tumour' had been palpable and an US scan not performed, it is uncertain whether a standard pyloromyotomy alone would have been effective, as the natural history of idiopathic FFH is unknown.

Paediatric surgeons should consider FFH in the differential diagnosis of infants who present with a history suggestive of GOO. Often these patients are older than the typical age at which IHPS presents. The diagnosis can be made by the characteristic features on US examination. Critical assessment of the US appearance should determine the need for pyloromyotomy in the presence of an associated IHPS. Surgical excision of the redundant mucosa in isolated FFH would seem to be definitive treatment.

References

1. Blackshaw AJ, Levision DA (1986) Eosinophilic infiltrates of the gastrointestinal tract. J Clin Pathol 39: 1–7
2. Chouraqui JP, Roy CC, Brochu P, Gregoire H, Morin CL, Weber AM (1981) Menetrier's disease in children: report of a patient and review of sixteen other cases. Gastroenterology 80: 1042–1047
3. Katz ME, Blocker SH, McAlister WH (1985) Focal foveolar hyperplasia presenting as an antral-pyloric mass in a young infant. Pediatr Radiol 15: 136–137
4. Leonidas JC, Beatty EC, Wenner HA (1973) Menetrier disease and cytomegalovirus infection in childhood. Am J Dis Child 126: 806–808
5. Marcus P, Verp MS (1993) Menetrier disease in a child of a consanguineous union. Am J Med Genet 47: 1231–1232
6. McAlister WH, Katz ME, Perlman JM, Tack ED (1988) Sonography of focal foveolar hyperplasia causing gastric obstruction in an infant. Pediatr Radiol 18: 79–81
7. Mercado-Deane MG, Burton EM, Brawley AV, Hatley R (1994) Prostaglandin-induced foveolar hyperplasia simulating pyloric stenosis in an infant with cyanotic heart disease. Pediatr Radiol 24: 45–46
8. Mezoff AG, Hyams JS (1995) Radiological case of the month. Arch Pediatr Adolesc Med 149: 1169–1170
9. Peled N, Dagan O, Babyn P, Silver MM, Barker G, Hellmann J, et al (1992) Gastric-outlet obstruction induced by prostaglandin therapy in neonates. N Engl J Med 327: 505–510
10. Sanna CM, Loriga P, Dessi E, Frau G, Congiu G, Corrias A, et al (1991) Hyperplastic polyp of the stomach simulating hypertrophic pyloric stenosis. J Pediatr Gastroenterol Nutr 13: 204–208
11. Seidman E (1995) Eosinophilic gastroenteritis. In: Roy CC, Silverman A, Alagille D (eds) Pediatric clinical gastroenterology. Mosby Year Book St.Louis, pp 384–387
12. Spry CJF (1984) Eosinophilic gastroenteritis. In: Bouchier IAD, Allan RN, Hodgson HJF (eds) Textbook of gastroenterology. Balliere Tindall, London, pp 596–598
13. Sundt TM, Compton CC, Malt RA (1988) Menetrier's disease – a trivalent gastropathy. Ann Surg 208: 694–701
14. Talley NJ, Shorter RG, Philips SF, Zinsmeister AR (1990) Eosinophilic gastroenteritis: a clinicopathological study of patients with disease of the mucosa, muscle layer, and subserosal tissues. Gut 31: 54–58
15. Whitington PF, Whitington GL (1988) Eosinophilic gastroenteropathy in childhood. J Pediatr Gastroenterol Nutr 7: 379–385