Case Number 11
Hypertrophic Pyloric Stenosis

Andrea Vella Baldacchino & Trevor Tabone
Reviewed by: Dr. Raymond Parascandalo

Case summary:

Demographic details:
Ms. KP, female,
Referred from: clinic

A 6-week old baby was brought to A&E by her parents, after a 3 week history of frequent vomiting, not tolerating feeds, and weight loss. She was well for the first 3 weeks of life, before she stopped tolerating any feeds. Despite several changes with the feeds given, the baby's symptoms did not improve. Moreover, she started losing weight, while she became increasingly irritable and inconsolable. Her consultant paediatrician decided to admit her for a trial of other formula feeds, and further investigations. In hospital, examination was unremarkable, except for an intermittently palpable mass in the RUQ. An ultrasound of the abdomen confirmed the diagnosis of pyloric stenosis. She was kept nil by mouth, an NG tube inserted for drainage, and intravenous fluids administered, with close monitoring of fluid input and output. Pyloromyotomy was performed and the patient was discharged after ensuring that she was tolerating feeds.

Presenting complaint:

Vomiting, not tolerating feeds: 3 weeks
Recent weight loss

History of presenting complaint:

KP was well for the first 3 weeks, during which she was growing adequately, reaching a weight of 3.78kg. She was initially fed on breast milk and formula milk (Aptamil). At 3 weeks, she started vomiting frequently with excessive, inconsolable crying. The vomiting was non-bilious, and at variable times after feeds, sometimes right after feeds or 1-2 hours later. At this time, she was seen by a paediatrician and feeds were changed, first to Novalac AC (anti-colic) and then to Novalac AR (anti-reflux), but with minimal improvement. Consequently, she was started on ranitidine and since she was failing to thrive, having lost weight down to 3.56kg, she was admitted for trial of HA formula and additional investigations. By the time of admission, she had visibly lost weight, and was becoming increasingly irritable and inconsolable.

Past medical and surgical history:

Born: 39^{+2}/40 weeks
Birth weight: 2.86kg
Occipitofrontal circumference: 33cm
Mode of Delivery: Normal vaginal delivery; no complications
No vaccinations yet

Drug history:

Drug	Dosage	Frequency	Type	Reason
Ranitidine	1mL (15mg)	BD	H2-receptor antagonist	To reduce acidity of gastric contents in gastro-oesophageal reflux
Simethicone		PRN	Anti-foaming agent	Reduces colic caused by gas in GIT

NKDA

Family history:

No family history of note

Systemic inquiry:

- General Health: Lost weight since stopped tolerating feeds
- Gastrointestinal System: Passes stools twice daily, parents claim it has been looser than usual
- Afebrile

Discussion of results of general and specific examinations:

General examination and vital signs: Patient appeared alert, not cyanosed (pink in air). A papular erythematous rash on the face and trunk was noted, and she had little subcutaneous fat stores. The following parameters were taken:

Admission weight: 3.56kg (3rd centile)
Capillary refill time: <2seconds
Heart rate: 140bpm
Blood pressure: 116/80 mmHg
O2 saturations: 98% in air
Temperature: 36.7ºC

General examination:
It was established that the patient was haemodynamically stable on admission, especially given her history of recurrent vomiting. Her alertness, normal capillary refill time, heart rate and blood pressure indicate that there is no clinical dehydration. Absence of fever suggests that there is no significant inflammatory process or infection contributing to the presenting symptoms. The poor subcutaneous adipose tissue is consistent with the history of weight loss and failure to thrive.

Respiratory examination:
Clear chest with good air entry. These findings are relevant given the history of frequent vomiting, since pulmonary aspiration of gastric contents can cause recurrent pneumonia, cough and wheezing.

Abdominal examination:
Abdomen was soft on palpation, and a mass was intermittently palpable in the right upper quadrant. Bowel sounds and hernial orifices were normal, and there was no organomegaly. The ‘olive’ mass in the RUQ is characteristic of pyloric stenosis, felt on gentle, deep palpation halfway between the midpoint of the anterior border of the right ribcage, and the umbilicus. It must be differentiated from other abdominal masses in infants, such as the ‘sausage shaped’ mass found in intussusception (although this condition is typically found in older age groups). Normal bowel sounds confirm that there is no bowel obstruction and normal bowel motility.

Differential diagnosis:

- Pyloric stenosis
- Cow's milk protein allergy
- Gastro-oesophageal reflux
- Infant colic
- Infection (urinary tract, otitis media)

Diagnostic procedures:

Laboratory exams:

Test: Complete blood count
Justification: Anaemia e.g. deficiency anaemias secondary to repeated vomiting or due to oesophagitis complicating reflux; neutropenia could indicate sepsis; neutrophilia and thrombocytosis could indicate an inflammatory process; baseline blood counts to monitor disease process or as part of preoperative assessment if surgery needed.
Result: Normal values

Test: Urea, electrolytes and creatinine; venous blood gases
Justification: Since history of recurrent vomiting, essential to determine if there is significant dehydration (high urea), and electrolyte disturbances such as hypokalaemia, hypochloraemia and alkalosis. eGFR values can help in excluding pre-renal acute kidney injury due to hypovolaemia.
Result: Normal values
Conclusion: No metabolic derangement

Test: Blood glucose
Justification: Hypo- or hyperglycaemia in neonatal sepsis; increased susceptibility to develop hypoglycaemia due to poor fat depots; Hyperglycaemia with DKA (diabetic ketoacidosis) consistent with vomiting and weight loss.
Result: 4.3mmol/dL (normal range)
Conclusion: Normoglycaemic

Test: IgE & RAST (radioallergosorbent test)
Justification: Cow's milk allergy
Result: Negative
Conclusion: Cow's milk allergy unlikely to be the underlying cause for the presenting symptoms.

Instrumental investigations:

Test: Abdomen Ultrasonography
Justification: To confirm of refute the diagnosis of hypertrophic pyloric stenosis
Result: - Thickened and elongated pylorus (Pyloric width: 17mm; Length: 18mm - both exceed normal values)
- Hyperechoic gastric content, stomach distended despite having had last feed more than 3hours before.
- Pylorus not seen to open

Conclusion: All measurements and observations are in keeping with a diagnosis of pyloric stenosis.

Therapy:

Resuscitation and hydration	Dosage	Frequency	Type	Reason
Dextrose and KCl in saline.	5% dextrose in 0.45 saline and 2.7mLs of 20% KCl in each 500mL	19mL/hr IV	Crystalloid	Fluid resuscitation as a pre-operative measurement.

Surgical therapy:

Pre-operative: Following diagnosis of pyloric stenosis on ultrasound, the paediatric surgeon was informed. KP was subsequently kept on a nil by mouth regimen, and an intravenous infusion at 19mL/hr of 5% dextrose in 0.45 saline and 2.7mLs of 20% KCl in each 500mL saline was started. Strict input/output charting and daily weight measurements were recorded. A nasogastric tube was left on open drainage.

Operation: An open pyloromyotomy was performed (Ramstedt's procedure). Two control trials of passing air through pylorus were carried out – no leaking was reported. After ensuring adequate haemostasis, the wound was closed in layers with vicryl 4/0 and 6/0.

Post-operative: Day 1: Early morning occasional retching with posseting of small amounts of clear fluid was noted. KP slept comfortably and did not appear in pain according to mother. The patient was also afebrile, with a slightly elevated serum K+ level of 5.65.
Day 2: The nasogastric tube was removed and half-strength feeds were started, which were tolerated. On examination, the abdomen was found to be soft and non-tender whilst the wound was noted to be healing well.
Day 3: Full strength feeds started at 60mL/3hr.
Day 4: All feeds were now being tolerated, hence it was decided that KP is deemed fit for discharge, after all post-operative parameters were found to be normal. Paracetamol 60mg 8 hourly per rectum was prescribed.

Diagnosis:

A definitive diagnosis of hypertrophic pyloric stenosis (HPS) was made. HPS is characterized by diffuse hypertrophy and hyperplasia of the muscular layers at the gastric antrum and pylorus, leading to a narrowing of the pyloric channel and a functional gastric outlet obstruction[1,2]. This rather common condition has an incidence of 2-4 per 1,000 births in white populations, being slightly less prevalent among Black, and rare in Asian populations. It is far more common in males than in in females, with a ratio of approximately 4:1. Curiously, there appears to be a greater risk for offspring of mothers who had HPS, than of fathers who had the condition, and HPS has been associated with B and O blood groups, as well as certain congenital anomalies, namely tracheo-oesophageal fistula and hypoplasia/agenesis of the inferior labial frenulum[3].

The aetiology of HPS remains unknown. However, immunocytochemistry techniques have determined a number of abnormal features within the muscular layers including a deficiency in nerve terminals, markers for nerve supporting cells, peptide-containing nerve fibres, nitric oxide synthase (NOS) activity (and mRNA for NOS) as well as interstitial cells of Cajal. Moreover, increased expression of insulin-like growth factor (IGF-1) mRNA and platelet-derived growth factors (PDGF) have been found. One hypothesis postulates that this abnormal innervation of the muscular layers leads to reduced muscular relaxation, with increased growth factor production, which promote muscular hyperplasia, hypertrophy and obstruction[2].

This case presented with symptoms which began at three weeks of life, and this is typical of HPS (average age of presentation 3/4 weeks). Almost all cases are diagnosed within the range of 1 to 12 weeks of life, and is exceedingly rare in stillbirths and preterms, hence the belief that HPS develops after birth[1,4]. The 'hypergastrinaemic hypothesis' attempts to justify this belief by proposing that an inherited increase in the parietal cell mass causes hyperacidity and decreased gastrin control, which in turn leads to repeated contractions of the pylorus and secondary work hypertrophy. This could explain why pyloric stenosis develops after initiation of feedings. In a study by Krogh et al.[6], a strong correlation of pyloric stenosis with bottle-feeding was recorded, with bottle-fed infants experiencing a 4.6-fold higher risk of developing PS than those who were exclusively breast-fed. In our case, we note that the baby was both breast and bottle-fed for the first 3 weeks before initiation of symptoms.

Pyloric stenosis usually presents with either intermittent or regular non-bilious vomiting after each feeding, with symptoms typically starting at 3 weeks of age. The vomitus may be blood stained with protracted vomiting, presumably due to underlying gastritis. The classical clinical picture described for pyloric stenosis is that of progressively projectile vomiting, hypochloraemic metabolic alkalosis, a hard "olive-shaped" mass palpated in the mid-epigastrium and visible gastric peristaltic waves, more pronounced after feeding. This traditional description of the clinical presentation of HPS has become increasingly uncommon. Glatstein et al.[6] reported drastic reductions in the frequencies of HPS cases in which an "olive" was palpated (from more than 50% in previous studies down to 13.4%) and also cases with electrolyte disturbances at presentation. The same trends were recently echoed in a study by Taylor et al.[8] in which frequencies of palpation of an "olive", hypochloraemia, visible peristalsis and haematemsis were all shown to be reduced when compared to a study evaluating patients treated between 1984 and 1995. These studies point to the fact that pyloric stenosis is being diagnosed earlier due to the increase reliance on ultrasound rather than clinical examination. This earlier diagnosis accounts for the decreasing number of patients presenting with metabolic derangements and the classical signs and symptoms. Imaging by ultrasound is now firmly established as the chief diagnostic method as is reflected in the vast majority of cases being diagnosed by sonography, given its accuracy with a specificity of 100% and sensitivity of 98%, ease of repeating the test and non-invasiveness[9].

Our case is consistent with these current trends of presentation and diagnosis. At the time of diagnosis, the patient did not demonstrate any strongly suggestive symptoms. Palpation of an 'olive' mass was not consistently reported by all medical professionals, and was only first noted after several previous examinations had failed to pick up the sign, which shows the unreliability of clinical examination.

The definitive treatment for pyloric stenosis is by pyloromyotomy, in which the hypertrophied muscle, but not the mucosa, is divided. Nowadays, the major treatment modalities available are either as an open procedure, also known as Ramstedt's procedure, which was done in our case (see below), or laparoscopically. Since its introduction, laparoscopic pyloromyotomy (LP) has gained popularity with the advances being made in laparoscopic technology. Current controversy exists about which approach is the more superior and most effective. A study by Oomen et al.[10] revealed that the time needed postoperatively to start full feeds and later hospital stay were both statistically significantly shorter when compared to patients that had undergone an open pyloromyotomy. Moreover, the major postoperative complication rate (such as incomplete pyloromyotomy and perforation – needing reoperation) after LP was found to be no different from that following an open procedure. While this might point towards the laparoscopic approach as being superior to the traditional open one, there is still no unanimous agreement. Some studies have even reported more complications following LP, particularly with regards to mucosal perforation and incomplete pyloromyotomy (Hall et al.[11], Adibe et al.[12]). Furthermore, while most studies agree that LP is associated with shorter times to full feeds and hospital stays postoperatively, these improvements only amount to a few hours at most. Therefore, it must be acknowledged that LP is associated with a steep learning curve, with the implication that this approach should only be considered as a standard of care and as a safe alternative to the open approach if done by experienced professionals with specific expertise in LP.

Final treatment and follow ups:

The definitive treatment was by pyloromyotomy, which resulted in complete resolution of signs and symptoms. The patient was discharged on the basis of having remained hydrated, tolerating feeds, and on adequate enteral intake.

A follow-up appointment at paediatric surgical outpatients was booked. The main reason for this is to check that the patient is thriving adequately for her age – which is a sign that feeds are being tolerated. Thus it is important that an accurate weight measurement is obtained and plotted on the growth chart.

Fact Box 11:

Title: Hypertrophic Pyloric Stenosis

Epidemiology:

- Prevalence: 2-4 per 1000 live births
- More common in caucasians
- More prevalent in males than in females, ratio 4:1
- Family history, especially maternal history, increases risk
- Associated with O and B blood groups, and congenital anomalies.
- Bottle-fed infants at higher risk

Symptoms:

Frequent vomiting, starting at around 4 weeks, and progressively more forceful, resulting in the pathognomonic projectile vomiting.

Signs:

- Gastric Peristaltic waves visible especially after feeds
- Pyloric mass, palpated as an 'olive' in the right upper quadrant
- Signs of dehydration: tachycardia, low urine output, mucosal dryness, sunken eyes, cold peripheries and, if decompensated dehydration, hypotension

N.B. The classical clinical presentation of projectile vomiting, visible peristalsis and a palpable 'olive' is much less commonly seen, with the advent of ultrasonography which has superseded clinical examination as the chief diagnostic method, allowing diagnosis to be made at a younger age, and institution of treatment before metabolic derangements. Therefore, clinicians should have a high degree of suspicion and consider pyloric stenosis with presentations of recurrent vomiting in infants of the appropriate age, in the absence of the later classical signs.

Treatment:

- Following diagnosis, patient is kept nil by mouth, while intravenous fluids are given, to correct electrolyte disturbances and dehydration.
- Pyloromyotomy (open or laparoscopic) provides an excellent outcome and minimal complications

References:

1. http://emedicine.medscape.com/article/929829-overview - accessed on 21/12/2012
2. Hernanz-Schulman M. Infantile hypertrophic pyloric stenosis. Radiology. 2003; 227(2):319-31.
3. Kliegman RM, Stanton BF, St. Geme JW, et al. 2011 Nelson Textbook of Pediatrics Saunders, Philadelphia ISBN: 978-1-4377-0755-7 (Chapter 321.1 – Pages 1274-1275).
4. Chan SM, Chan EK, Chu WC, et al. Hypertrophic pyloric stenosis in a newborn: a diagnostic dilemma. Hong Kong Med J. 2011;17(3):245-7.
5. Rogers Im. The true cause of pyloric stenosis is hyperacidity. Acta Paediatr. 2006;95(2):132-6.
6. Krogh C, Biggar RJ, Fischer TK, et al. Bottle-feeding and the Risk of Pyloric Stenosis. Pediatrics. 2012;130(4):e943-9.
7. Glatstein M, Carbell G, Boddu SK, et al. The changing clinical presentation of hypertrophic pyloric stenosis: the experience of a large, tertiary care pediatric hospital. Clin Pediatr (Phila). 2011;50(3):192-5.
8. Taylor ND, Cass DT, Holland AJ. Infantile hypertrophic pyloric stenosis: Has anything changed?. J Paediatr Child Health. 2012; J Paediatr Child Health. 2012 Dec 2. doi: 10.1111.
9. Niedzielski J, Kobielski A, Sokal J, et al. Accuracy of sonographic criteria in the decision for surgical treatment in infantile hypertrophic pyloric stenosis. Arch Med Sci. 2011 Jun;7(3):508-11.
10. Oomen MW, Hoekstra LT, Bakx R, et al. Open versus laparoscopic pyloromyotomy for hypertrophic pyloric stenosis: a

systematic review and meta-analysis focusing on major complications. Surg Endosc. 2012;26(8):2104-10.
11. Hall NJ, Van Der Zee J, Tan HL, et al. Meta-analysis of Laparoscopic Versus Open Pyloromyotomy. Ann Surg. 2004;240(5):774–778.
12. Adibe OO, Nichol PF, Flake AW, et al. Comparison of outcomes after laparoscopic and open pyloromyotomy at a high-volume pediatric teaching hospital. J Pediatr Surg. 2006 Oct;41(10):1676-8.