Pneumatosis cystoides intestinalis (PCI) is a rare condition which is characterised by the presence of multiple thin walled gaseous cysts in the bowel wall. We report a patient who presented to the Emergency Department with features of intestinal obstruction and peritonitis and was operated upon. The histopathological diagnosis was pneumatosis cystoides intestinalis. The likely reason for PCI in this patient was pyloric stenosis.

Keyword: Pyloric stenosis, intestinal obstruction, terminal ileum, Pneumatosis cystoides intestinalis (PCI), mechanical theory

Case report:
A sixty year old male with no known medical comorbidity presented to our Emergency Department with severe right upper and lower quadrant abdominal pain of twenty day duration. The pain was colicky in nature and was associated with few episodes of vomiting. There was no history of any previous abdominal operations, altered bowel habits, jaundice or gastrointestinal bleeding. He gave history of having taken medication for acid peptic disease in the past. On examination he was dehydrated and tachycardic. The abdomen was distended with marked tenderness in the right hypochondrium and lumbar regions. Routine blood investigations including CBC, electrolytes, liver function test and bleeding profile were within normal limits. The plain x-ray abdomen showed multiple dilated small bowel loops suggestive of small bowel obstruction. A provisional diagnosis of intestinal obstruction was considered and was taken up for exploratory laparotomy. At operation mass lesion was seen in the terminal ileum around twenty centimetres proximal to the ileocaecal junction causing luminal obstruction. This patient also had Meckels diverticulum and the serosa over the diverticulum and the diseased ileum showed what appeared to be bubbles of air. Resection of the affected segment of terminal ileum along with the diverticulum was done. The histology showed multiple cystic spaces in the subserosa. The cystic spaces were lined with flattened macrophages and multinucleated giant cell were found (Figure 1).

The IHC staining revealed CD68 positive histiocytes around the cystic spaces (Figure 2). Post operatively he had prolonged ileus which settled with conservative management.

The patient presented to the Emergency Department a week later with symptoms of postprandial vomiting. Intestinal obstruction probably secondary to postoperative adhesions was considered. Upper GI endoscopy showed scarring of the pyloroduodenum with luminal narrowing. He underwent truncal vagotomy and gastrojejunostomy. He had an uneventful recovery. He was doing well at his last follow up six months from the time of operation.
Discussion:
Pneumatosis cystoides intestinalis (PCI) is a rare clinical entity which is characterised by the presence of multiple thin walled gaseous cysts containing nitrogen, hydrogen and carbon dioxide in the bowel wall.  
Duo Vernoi in 1730 used the term "pneumatosis intestinalitis" and in 1754 wrote the first report of PCI, which was published in the French literature.  
The diagnosis of PCI in surviving patients was first established by Hahn in 1899.  
Diagnosis of PCI by means of preoperative radiological findings was first described by Baumann-Schender in 1939. 
The exact incidence of PCI is difficult to ascertain as majority of patients with this pathology are asymptomatic and may not seek medical attention. 
Based on an autopsy series the overall incidence of PCI was found to be 0.03% in the general population. PCI is seen more commonly in males than females. The peak incidence is seen at 30-50 years of age.  
The cysts are commonly located in the subserosal or submucosal plane of the bowel wall. The subserosal cysts are common in small intestinal pneumatosis while in colonic pneumatosis the cysts are often found to be submucosal. 
In a large series by Jamart on a study of 919 cases of PCI 42% of the patients had involvement of small bowel, 36% had colonic involvement and 22% present had involvement of both small and large bowel. It is likely that multiple mechanisms are involved in the development of PCI and exact aetiology is unknown.  
Different theories have been proposed to explain the pathogenesis of PCI. Mechanical, bacterial and biochemical are among them. The mechanical theory suggests that gas dissects into the wall of the bowel from either the luminal surface through breaks in the mucosa or through the serosal surface by tracking along mesenteric blood vessels. This happens in patients, who have had blunt abdominal trauma, after bowel anastomosis and with gastric outlet obstruction. The bacterial theory suggests that gas forming bacteria gain access to the submucosa through breaches in the mucosa. The biochemical theory proposes that luminal bacteria produce excessive amounts of hydrogen gas through fermentation of carbohydrates and other foodstuffs. As the pressure of the gas within the intestinal lumen increases, gas may be forced directly through the mucosa and get trapped within the submucosa. PCI is of two types - primary or idiopathic type which accounts to 15% of cases and the secondary type which accounts to 85% of cases. 
The secondary causes of PCI may be classified under two categories: the ones that are benign and those that are life threatening. The benign causes include (i) gastrointestinal conditions like pyloric stenosis, inflammatory bowel diseases, bowel obstruction (ii) pulmonary conditions like asthma, bronchitis, emphysema (iii) systemic diseases like scleroderma and AIDS (iv) medications like steroids, alpha glucosidase inhibitors (v)iatrogenic post endoscopy, post-surgical bypass/anastomosis and (vi) Post organ transplant. The life threatening causes are mesenteric ischemia (most common), toxic megacolon, bowel obstruction, caecal ileus and acute graft versus host reactions. Most of the patients with PCI are asymptomatic. The signs and symptoms of PCI are nonspecific and often lead to erroneous diagnosis. The common symptoms of small intestinal pneumatosis are vomiting, abdominal distension, weight loss, abdominal pain, and diarrhoea. The symptoms of colonic pneumatosis are diarrhoea, hematochezia, abdominal pain, abdominal distension, and constipation. Abdominal plain X ray will show the characteristic finding intestinal wall in two thirds of patients. Intramural gas can be linear, curvilinear, or circular in appearance. Pneumoperitoneum develops when the subserosal blebs rupture. CT scan is more sensitive than plain X ray and can help in detection of underlying cause for PCI. Characteristic findings of PCI include circumferential collections of air adjacent to the lumen of the bowel that run in parallel with the wall of the bowel, but without the air-fluid levels. 
The important decision in the management of patients with PCI is to make a decision as to treat these patients conservatively or proceed with emergent exploratory laparotomy. 

To determine the need for surgical therapy Knechtle et al. found a correlation between the clinical presentation, the need for surgery and the final outcome. The following clinical features predicted the presence of bowel necrosis: a history and physical examination that suggested an acute abdominal process, arterial pH <7.3, HCO3 <20 mmol/L, an elevated lactate level, an elevated serum amylase level, or the presence of portal venous gas. If a patient lacks above clinical findings they can be managed conservatively with antibiotics, special diets, high-flow oxygen therapy, hyperbaric oxygen therapy, and endoscopic therapy. However surgery should be considered if patients remain symptomatic despite medical therapy or who develop complications. Our patient did not have any history of trauma to his abdomen or any recent abdominal operation. Also the diagnosis of PCI was not made preoperatively. The finding of curvilinear radiolucency suggestive of air in the bowel wall in the right lower quadrant. (Figure 3) and left lower quadrant (Figure 4) was picked up when the x-ray was reviewed retrospectively. It is possible that the likely cause for our patient to develop PCI was the pyloric stenosis which the patient became symptomatic after the first operation. Similar it has been reported in the literature that 60% of cases of pneumatosis are unrelated to pyloric stenosis. 

Conclusion:
Pneumatosis cystoides intestinalis is a rare clinical entity which could be primary or secondary due to various causes. It important to know and be aware of this condition, and also to understand the various causes and associations of this uncommon finding so that the condition is diagnosed correctly and appropriate management can be offered to the patient.
References: