



## Unravelling the Cocoon: A Case Report, Review of the Literature, and Journey in Rediscovering Congenital Peritoneal Encapsulation

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### Abstract

Congenital peritoneal encapsulation is an uncommon and poorly understood condition. It usually presents as a diagnostic dilemma with eventual unexpected laparotomy findings and occasional surgical misadventure. We describe a rare case of an elderly gentleman presenting with acute intestinal obstruction, with typical radiographic findings raising pre-operative suspicion of an abdominal cocoon. Surgery was mandatory due to non-resolving mechanical obstruction, and concerns that there may have been segments of bowel in closed loops. Recognition of the condition is critical in directing clinical decision-making and successful application of appropriate surgical techniques, particularly for patients of “idiopathic” or congenital etiology.

**Keywords:** Abdominal cocoon; Congenital peritoneal encapsulation; Sclerosing encapsulating peritonitis; Intra peritoneal fibro sclerosis; Peritonitis chronica fibrosa incapsulata

### Background

Definition itself has raised controversy in the rare condition ‘congenital peritoneal encapsulation’. More plainly described as ‘congenital abdominal cocoon’, it has been argued to be both interchangeable and distinct from terms like ‘sclerosing encapsulating peritonitis’, ‘intra-peritoneal fibrosclerosis’, and ‘peritonitis chronica fibrosa incapsulata’, all of which encompass a wider spectrum of conditions broadly classified as primary or secondary encapsulating peritonitis.

The congenital variety is usually asymptomatic and discovered serendipitously at unrelated surgery or autopsy. However, intestinal obstruction may also result from encasement of variable lengths of bowel by dense fibrocollagenous membranes, visually resembling a cocoon. The rare and poorly understood condition usually presents itself as a diagnostic dilemma. More often seen in younger females [1] and males [2], to the best of our knowledge, this is the first elderly male patient from Singapore and the 7<sup>th</sup> reported in international literature.

### Case Presentation

A 60 year old Chinese male presented with colicky central abdominal pain, vomiting and lack of bowel movement for two days. Prior to this he did not have longstanding gastrointestinal symptoms. He had no significant medical history, specifically autoimmune conditions, tuberculosis, connective tissue disorders, beta-blocker usage, peritoneal catheters, prior abdominal infection, trauma, nor surgery. On examination, his abdomen appeared distended and distorted by a mildly tender, diffuse mass in the right lower quadrant.

Haematological investigations revealed mild leucocytosis. Plain radiographs showed a distended gastric bubble and multiple air-fluid levels. Contrast enhanced computed-tomography of the abdomen and pelvis revealed gross dilatation of the stomach and duodenum, transitioning into a conglomeration of segmentally dilated loops of jejunum and ileum. A thickened well enhancing layer surrounded it, likely fibrotic peritoneum. There was subtle crowding of the mesenteric vessels, but the superior mesenteric vessels remained in a normal anatomic configuration with no malrotation (Figure 1).

Surgery was mandatory due to non-resolving mechanical obstruction and concerns that there may have been segments of bowel in closed loops. Intra-operatively, the bowel was wrapped in a near-opaque membranous sac in a concertina-like fashion from the duodenojejunal to ileocaecal junction (Figure 2a). The entire cocoon was mobile and the sac itself appeared to be free from

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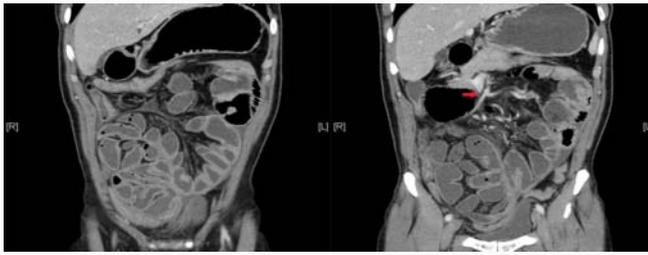
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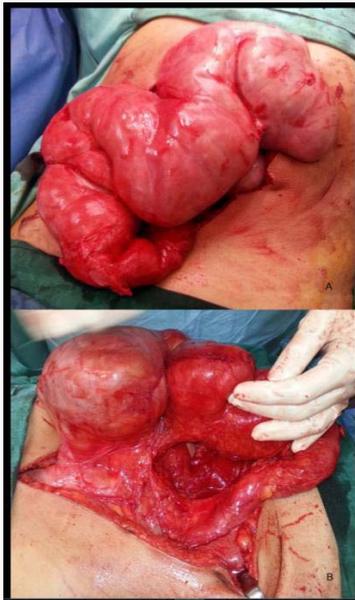
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**Figure 1:** Contrast enhanced computed-tomography of the abdomen and pelvis displaying the typical small bowel cocoon with crowding of the mesenteric vessels.



**Figure 2:** Intra-operative view of the cocooned bowel with its shortened mesentery.

the parietal peritoneum. Resolution of mechanical obstruction was achieved by resection of the encasing membrane and adhesiolysis with a combination of sharp and blunt dissection. This parted the small bowel, while preserving its serosa. Apart from mild congestion, the bowel was otherwise normal. The root of the mesentery was assessed to be shortened (Figure 2b), contributing to the crowding of the entire length of small bowel within the membranous sac. The small bowel was decompressed before closure of the abdomen.

Our patient made an uneventful recovery after a week of post-operative ileus, and was discharged well nine days later. He remains healthy and asymptomatic after a year's follow up.

## Discussion

First described by Cleland [3] and named "peritonitis chronica fibrosa incapsulata" by Owtschinnikow [4], the idiopathic form of encapsulating peritonitis was formally identified by Foo et al. [1]. Three decades on, the disease remains too uncommon to quantify its incidence nor qualify its pathophysiology and natural history. Early clinical features are non-specific, but established congenital peritoneal encapsulation may present as either acute or chronic extramural cause of intestinal obstruction. Patients may also exhibit variable degrees of weight loss and occasionally there may be a 'misleading' abdominal mass. Complications include malnutrition, ischemia, perforation, or enterocutaneous fistulae.

To date, there exist questions that possess unsatisfactory answers.

### What is the aetiology and pathophysiology?

Abdominal cocoon may be classified as primary or secondary. The development of primary sclerosing encapsulating peritonitis remains 'idiopathic' to date. It was first documented in adolescent females within 2 years of menarche, originating from warm tropical and subtropical climates. Hence authors had initially suggested that this could be related to retrograde menstruation or gynaecological infection from transvaginal peritonitis [1]. Theories behind such a disease process have little supportive evidence. With more reports of male patients, some postulate instead that there may be congenital defective return of the embryonic midgut loop to the intraperitoneal abdominal cavity [5].

The development of secondary sclerosing encapsulating peritonitis may be better explained. Risk factors include chronic ambulatory peritoneal dialysis [6], peritonitis [7], previous abdominal surgery [8], and prolonged use of beta blockers [9]. There also exist case reports of disease related to benign gynaecological conditions, ovarian malignancy, gastrointestinal malignancy, patients presenting along with abdominal trauma, peritoneal shunts (ventriculoperitoneal and peritoneal-venous), liver cirrhosis, systemic lupus erythematosus, tuberculosis, familial Mediterranean fever, *ascaris lumbricoides*, and one reporting consumption of cereals infected with fungus containing biologically active amines. The common underlying disease process points towards low-grade intra-abdominal sepsis, resulting in sclerosis and fibrosis. This is consistent with reports of success in treating early stages of the disease with immunosuppressants, corticosteroids and tamoxifen.

Many terms have been utilized to describe the same disease. Perhaps the clinical entity would benefit from realignment of terminology with clear stratification by proposed aetiology.

### What factors will allow us to consider a congenital cause?

The most peculiar anatomical clue is that the prime majority of 'idiopathic' cases only affect the small bowel. Only isolated cases describe involvement of the small intestine plus other organs (e.g. appendix, ascending colon, ovaries). This may be consistent with a developmental defect resulting in the small intestine being covered by the original dorsal mesentery, which forms the accessory peritoneal sac.

### Are there typical and distinctive radiological features that can confirm the diagnosis preoperatively?

Anecdotally, the use of radiological imaging modalities has been the single clearest aid in attaining non-invasive pre-operative diagnosis. Sonography may reveal small bowel loops arranged in concertina shape with a narrow posterior base, in a cauliflower appearance, only if there were concurrent ascites [10]. Typical features of dilated but congregated bowel loops encased in a thick membrane of soft tissue density may be evident on computed-tomography scan [10]. Barium follow-through may also show similar findings along with delayed transit of contrast media. There have also been recent developments in using dynamic cinematographic magnetic resonance, coupled with advanced image analysis. Current studies involve patients with suspected PD-related encapsulating peritoneal sclerosis, where areas of altered movement might suggest frank adhesions [11] although diagnosis may be suspected with the assistance of such radiological findings, true pre-operative diagnosis still requires a high index of suspicion in the informed clinician.

Surgery is indicated in non-resolving mechanical obstruction, when there are concerns of bowel viability or complications such as perforation. Incision or excision of the encasing membrane is the ideal management option. When indicated, bowel resection or stoma creation should be considered. More extensive operations, including unnecessary bowel resections and bypass procedures, were performed when the condition was not recognised [1]. Only a small number of cases utilizing laparoscopic approach have been reported [12]. Some authors suggested prophylactic appendectomy to avoid difficult appendectomy should the patients develop appendicitis later [1].

Histologically, the membrane typically exhibits proliferation of fibro-connective tissue and inflammatory infiltrates, and no foreign body granulomas, giant cells, or birefringent material [13].

Long term prognosis appears to be excellent. There have been no reports of recurrence to date.

## Conclusion

Recognition of congenital peritoneal encapsulation is fundamental in insightful management to avoid surgical misadventure which may result in unwarranted enterotomies, bowel resection or stoma creation.

## Chief Educational Objective

Congenital peritoneal encapsulation is an uncommon and poorly understood condition. This article endeavors to revisit the topic and provide clarity to the issues surrounding the abdominal cocoon.

1. Congenital peritoneal encapsulation has been described to present as all of the following, except:

- a. Intestinal obstruction
- b. Abdominal mass
- c. Loss of appetite and loss of weight
- d. Enterocutaneous fistula
- e. Tenesmus

**Correct answer:** e. Tenesmus.

Explanation: Congenital peritoneal encapsulation results in the encasement of variable lengths of bowel by dense fibrocollagenous membranes, which may be asymptomatic, result in vague symptoms such as early satiety, loss of appetite, loss of weight, intestinal obstruction, an abdominal mass, or complications such as malnutrition, ischemia, perforation, or enterocutaneous fistulae. Tenesmus has not been described.

2. Described risk factors for secondary sclerosing encapsulating peritonitis include all of the following, except:

- a. Beta blockers
- b. Peritoneal dialysis
- c. Radiotherapy
- d. Tuberculosis
- e. Previous abdominal surgery

**Correct answer:** c. Radiotherapy.

Explanation: While there have been various associated risk factors for secondary sclerosing encapsulating peritonitis, radiotherapy has

not been described to be amongst them.

3. Typical radiological features of congenital peritoneal encapsulation include:

- a. Dilated bowel with a transition point
- b. Football sign
- c. Silver's sign
- d. Congregated bowel loops encased in a thick membrane of soft tissue density
- e. Rigler's sign

**Correct answer:** d. Congregated bowel loops encased in a thick membrane of soft tissue density.

Explanation: Typical features of dilated but congregated bowel loops encased in a thick membrane of soft tissue density may be evident on computed-tomography scan.

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