

# Sclerosing Encapsulating Peritonitis

## Review

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## التهاب الصفاق المحفظ المصلب

### مراجعة

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**ABSTRACT:** Sclerosing encapsulating peritonitis (SEP) is a rare chronic inflammatory condition of the peritoneum with an unknown aetiology. Also known as abdominal cocoon, the condition occurs when loops of the bowel are encased within the peritoneal cavity by a membrane, leading to intestinal obstruction. Due to its rarity and non-specific clinical features, it is often misdiagnosed. The condition presents with recurrent episodes of small bowel obstruction and can be idiopathic or secondary; the latter is associated with predisposing factors such as peritoneal dialysis or abdominal tuberculosis. In the early stages, patients can be managed conservatively; however, surgical intervention is necessary for those with advanced stage intestinal obstruction. A literature review revealed 118 cases of SEP; the mean age of these patients was 39 years and 68.0% were male. The predominant presentation was abdominal pain (72.0%), distension (44.9%) or a mass (30.5%). Almost all of the patients underwent surgical excision (99.2%) without postoperative complications (88.1%).

**Keywords:** Intestinal Diseases; Peritonitis; Sclerosis; Membrane Tissue; Abdominal Pain; Intestinal Obstruction.

**المخلص:** التهاب الصفاق المحفظ المصلب هو حالة من الالتهاب المزمن نادر الحدوث في الصفاق المعوي وغير معروف السبب. ويعرف كذلك بشرنقة البطن، ويحدث عندما تغطي حلقات من الأمعاء بغشاء داخل جوف الصفاق مما يؤدي إلى انسداد معوي. بسبب ندرة هذه الحالة و السمات السريرية اللانوعية، فغالبا ما يشخص بالخطأ. التهاب الصفاق المحفظ المصلب يظهر كنواب متكررة من انسداد الأمعاء الدقيقة. وقد تكون مجهولة السبب أو ثانوية، والأخير له علاقة بالعوامل المؤهبة مثل الديال الصفاقي أو السل البطني. يمكن استخدام العلاج التحفظي للمرضى في المراحل المبكرة، ومع ذلك، فإن التدخل الجراحي يعتبر ضروريا للحالات المتقدمة من انسداد الأمعاء. مراجعة الأدبيات أوضحت وجود 118 حالة من التهاب الصفاق المحفظ المصلب، متوسط عمر المرضى هو 39 عاما وكان 68.0% منهم ذكورا. كان العرض السائد هو ألم البطن (72.0%)، تمدد (44.9%) أو كتلة (30.5%). معظم هؤلاء المرضى خضعوا للاستئصال الجراحي (99.2%) بدون مضاعفات ما بعد الجراحة (88.1%).

**كلمات مفتاحية:** أمراض معوية؛ التهاب الصفاق؛ تصلب؛ غشاء نسيجي؛ ألم بطني؛ انسداد الأمعاء.

**S**CLEROSING ENCAPSULATING PERITONITIS (SEP) is a chronic inflammatory condition of unknown aetiology believed to result from recurrent low-grade or subclinical peritonitis with no specific abdominal signs; this eventually progresses to sclerosis and membrane formation with subsequent cocoon formation.<sup>1-6</sup> The condition is characterised by a thick greyish-white fibrotic membrane encasing the contents of the abdomen, predominately the small intestine.<sup>1-4</sup> Primary SEP, known also as an abdominal cocoon, has no obvious associated conditions; however, SEP can also be secondary to conditions that cause peritoneum inflammation and fibroblastic proliferation, for example peritoneal dialysis (PD)-related conditions or abdominal tuberculosis.<sup>1-4</sup> A clinical diagnosis of SEP is difficult in the early stages as symptoms are non-specific; radiology can hence play a significant role in diagnosing this condition preoperatively.<sup>1-6</sup> However,

symptoms often present as an acute emergency with an intestinal obstruction and the condition is frequently only diagnosed intra-operatively.<sup>1-6</sup> While patients can be managed conservatively in the early stages, when presenting at a later stage with intestinal obstruction, surgical intervention is essential. This review focuses on the aetiopathogenesis, diagnosis and management of this rare condition.

## Terminology and Classification

First described more than a century ago, SEP was initially termed peritonitis *chronica fibrosa incapsulata* to describe the membrane encasing the intestine; it has since also been named 'icing sugar' bowel and fibroplastic peritonitis.<sup>7-9</sup> In 1868, an entity known as peritoneal encapsulation was described.<sup>2</sup> In 1921, Winnen reported the first case of SEP, terming

**Table 1:** Types and features of encapsulating membrane conditions

| Condition                | Aetiopathogenesis   | Pathology  | Symptoms/findings  | Location   | Other   |
|--------------------------|---|--|--|--|---|
| Peritoneal encapsulation | <ul style="list-style-type: none"> <li>• Developmental anomaly</li> <li>• Derived from the yolk sac peritoneum in fetal life</li> <li>• Non-inflammatory</li> </ul> | <ul style="list-style-type: none"> <li>• Mesothelium membrane</li> <li>• Membrane is usually thin</li> </ul>   | <ul style="list-style-type: none"> <li>• Usually asymptomatic</li> <li>• Often detected incidentally during a laparotomy</li> </ul>  | <ul style="list-style-type: none"> <li>• Between the mesocolon, <i>omentum</i> and small intestine</li> </ul>  | -   |
| Primary SEP              | <ul style="list-style-type: none"> <li>• Unknown</li> </ul>   | <ul style="list-style-type: none"> <li>• Thick fibrocollagenous membrane</li> <li>• Inflammatory cells</li> <li>• Complete mesothelium loss</li> </ul> | <ul style="list-style-type: none"> <li>• Initially asymptomatic</li> <li>• Inflammatory phase: non-specific, including pain, nausea, vomiting, loss of appetite and malnutrition</li> <li>• Fibrosclerotic (advanced) phase: intestinal obstruction</li> </ul> | <ul style="list-style-type: none"> <li>• Type I: part of small intestine</li> <li>• Type II: whole small intestine</li> <li>• Type III: small and large intestine, ovary, liver and stomach</li> </ul> | <ul style="list-style-type: none"> <li>• Occurs in two forms: adolescent and adult</li> </ul> |
| Secondary SEP            | <ul style="list-style-type: none"> <li>• Several*</li> </ul>  | <ul style="list-style-type: none"> <li>• See above</li> </ul>  | <ul style="list-style-type: none"> <li>• See above</li> </ul>  | <ul style="list-style-type: none"> <li>• See above</li> </ul>  | -   |

SEP = sclerosing encapsulating peritonitis. \*See Table 2.

it *Zuckergussdarm*, which translates literally as ‘icing gut’, due to the intestinal surface appearing white from the membrane covering.<sup>8</sup> In 1978, Foo *et al.* coined the expression ‘abdominal cocoon’ to describe encapsulation of the abdominal contents.<sup>10</sup>

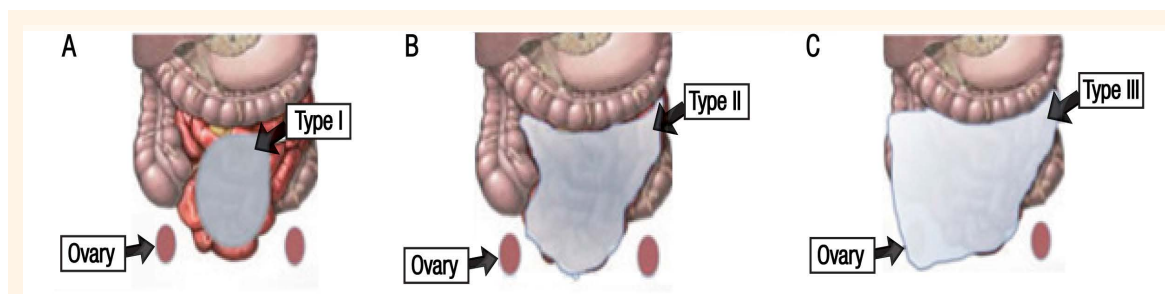
Several terms are currently used to describe various conditions in which a membrane encases the gut, including peritoneal encapsulation (PE), abdominal cocoon and idiopathic and secondary SEP [Table 1].<sup>2</sup> PE is a developmental anomaly presenting as an accessory peritoneal membrane, derived from the yolk sac peritoneum in the early stages of fetal life.<sup>2,11,12</sup> Hence, it is not related to an inflammatory process; it is predominately asymptomatic and is generally detected incidentally during a laparotomy performed for some other purpose.<sup>2,12–14</sup> The peritoneal membrane is typically found between the mesocolon, *omentum* and most of the small intestine.<sup>13,14</sup> On the other hand, SEP is an acquired condition and is a consequence of peritoneal inflammation due to various triggering factors.<sup>1–6,12,15</sup> The SEP membrane is covered by a dull fibrous structure that contains inflammatory cells, unlike PE whereby the membrane is covered by the

mesothelium.<sup>1–6,14</sup> SEP may enclose the gut partially or completely and may occasionally involve other intraperitoneal organs, including the stomach, liver and colon.<sup>1–6</sup>

Currently, SEP is classified as either primary (idiopathic) or secondary, depending on the aetiopathogenesis and the pathological characteristics of the encasing membrane.<sup>1–6,12,14</sup> Primary SEP is often referred to as abdominal cocoon syndrome and is classified into three categories based on the extent of encasement by the membrane [Figure 1]. Type I and II involves the encasement of either part of or the complete intestine, respectively, by a fibrocollagenous membrane. In type III, the appendix, *caecum*, ascending colon, stomach, liver and ovaries are also encased in addition to the small intestine.<sup>1–6,16</sup>

### Aetiology and Incidence

The aetiology and incidence of SEP depend on its classification. Primary SEP is idiopathic and not associated with any obvious cause. However, cytokines and fibroblasts likely influence the development of



**Figure 1A–C:** Classifications of primary sclerosing encapsulating peritonitis (SEP) into (A) type I, (B) type II and (C) type III. For types I and II, the membrane (grey shading) encloses part of and the whole of the small intestine, respectively. Type III SEP involves a membrane (grey shading) which encloses the whole of the small bowel and other organs, such as the ovaries and colon.

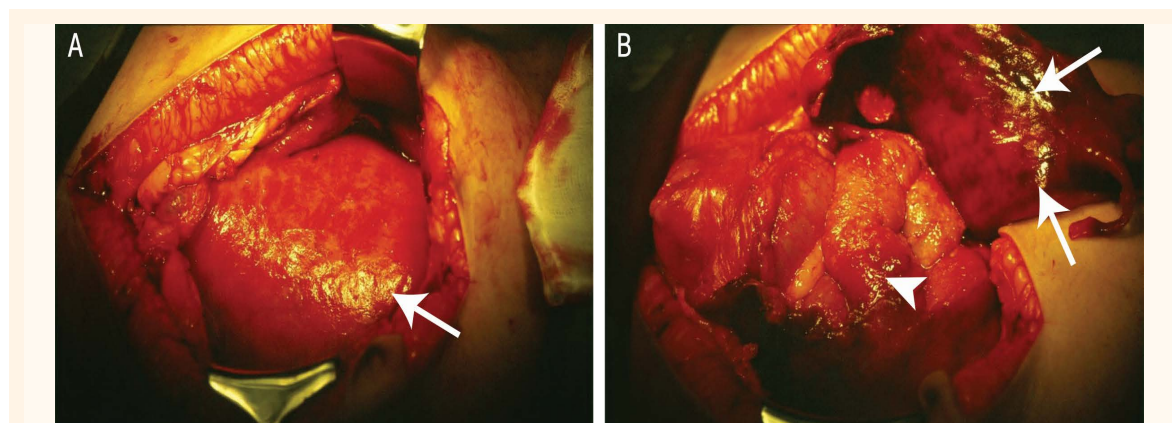
**Table 2:** Underlying causal factors of secondary sclerosing encapsulating peritonitis

| Aetiopathogenesis           | Specific causes  |
|-----------------------------|--|
| Drug-related                | <ul style="list-style-type: none"> <li>• Beta-blockers (e.g. practolol, timolol/propranolol)</li> <li>• Chemotherapy (e.g. methotrexate)</li> <li>• Asbestos exposure</li> </ul>   |
| Surgical/medical procedures | <ul style="list-style-type: none"> <li>• Peritoneal dialysis</li> <li>• Peritovenous shunts</li> <li>• Ventriculoperitoneal shunts</li> <li>• Intraperitoneal chemotherapy</li> <li>• Trauma-related</li> <li>• Liver transplantation</li> </ul> |
| Infection                   | <ul style="list-style-type: none"> <li>• Abdominal tuberculosis</li> <li>• Cytomegalovirus peritonitis</li> <li>• Granulomatous peritonitis due to parasitic infection</li> <li>• Recurrent peritonitis</li> </ul>                               |
| Inflammatory/autoimmune     | <ul style="list-style-type: none"> <li>• Sarcoidosis</li> <li>• Systemic <i>lupus</i> erythematosus</li> <li>• Familial Mediterranean fever</li> <li>• Fibrogenic foreign bodies</li> </ul>  |
| Disease                     | <ul style="list-style-type: none"> <li>• Liver cirrhosis</li> <li>• Gastrointestinal malignancy</li> <li>• Endometriosis</li> <li>• Ruptured dermoid cyst</li> <li>• Luteinized ovarian <i>thecomas</i></li> </ul>                               |
| Systemic                    | <ul style="list-style-type: none"> <li>• Protein S deficiency</li> </ul>   |

peritoneal fibrosis and neoangiogenesis in some way.<sup>2,17,18</sup> Several aetiopathogeneses have been proposed to explain primary SEP, including retrograde menstruation with a viral infection, retrograde peritonitis via the fallopian tubes and gynaecological infection-inducing cell-mediated immunological tissue damage.<sup>1-6,11-19</sup> However, these hypotheses do not explain the aetiopathogenesis for all patients, as 75% of patients with primary SEP are men,

premenstrual women or children.<sup>2,5,16</sup> Other theories proposed for the aetiopathogenesis of primary SEP include developmental disorders related to vascular anomalies and omental hypoplasia.<sup>2,16,19</sup>

In contrast, secondary SEP is associated with several causes and is therefore more common [Table 2]. The predominant cause of secondary SEP is PD, due to both its frequency worldwide and the associated peritoneal inflammation that the dialysis fluid induces.<sup>2-4</sup> Patients on PD are predisposed to developing peritoneal deterioration after prolonged exposure to PD fluids and subsequent bacterial peritonitis.<sup>3</sup> Long PD duration, acetate-buffered or hypertonic solutions and recurrent episodes of peritonitis may also predispose patients to infection; *Staphylococcus aureus*, various fungi or *Pseudomonas* sp. can also be contributory to the development of SEP.<sup>20</sup> Patients with these infections are at risk of developing intestinal obstructions, in addition to ineffective ultrafiltration caused by the presence of an encasing membrane.<sup>2,3,20</sup> A prospective multicentre study in Japan reported the overall incidence of SEP in patients undergoing PD to be 2.5%.<sup>21</sup> Interestingly, the incidence increases with prolonged periods of PD; another study reported the incidence as 1.9%, 6.4%, 10.8% and 19.4% among patients undergoing PD of two, five, six and eight years' duration, respectively.<sup>22</sup> Several other causes may contribute to secondary SEP, including abdominal tuberculosis [Figure 2]; autoimmune conditions like systemic *lupus* erythematosus; recurrent peritonitis; drug use such as chemotherapy or beta-blocker treatment; ovarian disorders such as dermoid cyst rupture or luteinized ovarian *thecomas*; abdominal surgery; peritoneal shunts; abdominal sarcoidosis; and fibrogenic foreign material.<sup>1-6,10-20</sup>



**Figure 2A–B:** Laparotomy images revealing (A) the thick membrane (arrow) cocooning the intestine of a patient with sclerosing encapsulating peritonitis and abdominal tuberculosis and (B) multiple exposed tubercles (arrowhead) on the intestinal surface after the membrane has been excised (arrows).

## Histopathological Features

Characteristic histopathological features of biopsied peritonea may include fibroconnective tissue proliferation, inflammatory infiltration and dilated lymphatic vessels.<sup>2-4</sup> While these non-specific features are not pathognomonic of SEP, they support the diagnosis when combined with operative findings. However, foreign body granulomas, giant cells and befringement foreign material are distinctly absent, thus excluding other diagnoses such as tuberculosis.<sup>1-3,5</sup> As a condition, SEP derives its name from characteristic macro- and microscopic pathological findings, including progressive formation of dense collagenous tissue sheets ('sclerosing'), sheaths of new fibrous tissue which contain and constrict the small bowel ('encapsulating') and the ongoing inflammatory process and presence of mononuclear inflammatory infiltrates within the new fibrosing tissue ('peritonitis').<sup>1-4</sup> One proposed pathological classification of SEP progression follows four phases: the pre-SEP, inflammatory, progressive and fibrotic phases.<sup>23,24</sup>

## Clinical Presentation

The clinical course of SEP usually includes episodes of intermittent and partial small bowel obstruction as a consequence of the kinking and compression of the intestine within the encasing membrane.<sup>1-6,10-25</sup> In the initial phase, the symptoms of SEP are non-specific and include fever, *ascites*, weight loss, loss of appetite and altered bowels; as the disease progresses, intestinal obstruction sets in.<sup>1,2,5</sup> The development of the membrane usually occurs over several years; however, it also has been reported to occur rapidly, within 12 weeks of the onset of symptoms.<sup>2,23,24</sup>

Idiopathic SEP typically presents in young adolescent girls in tropical and subtropical countries such as those in the Indian subcontinent as well as China, Malaysia, Singapore, Nigeria, Kenya and South Africa.<sup>1-6,25</sup> However, it has also been reported in more temperate zones.<sup>2</sup> As patients are generally asymptomatic and primary SEP is not obviously associated with other conditions, this rare condition is likely to be misdiagnosed. Nevertheless, a high index of clinical suspicion is indicated for patients who present with recurrent abdominal pain which cannot be attributed to any other obvious pathology.<sup>1-6,16,22,25</sup> A considerable number of primary SEP patients present with an intestinal obstruction.<sup>2,15</sup> Biopsy results from surgical interventions may indicate SEP unexpectedly.<sup>1-5,16,26,27</sup> Li *et al.* found that 52.3% of a large series of 65 SEP patients were diagnosed during

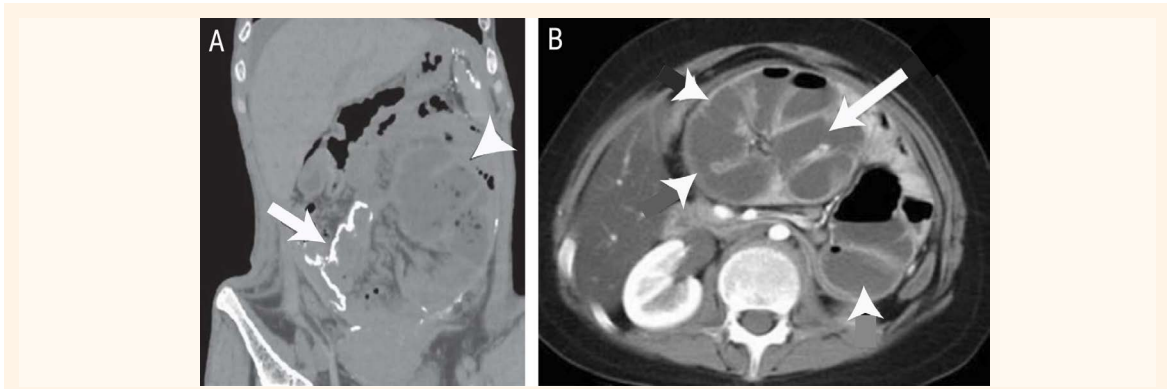
surgery in contrast to 47.7% who were diagnosed preoperatively.<sup>5</sup> While the majority of SEP patients are asymptomatic, some symptoms may include nausea, vomiting, loss of appetite, weight loss and malnutrition as a consequence of acute, subacute or chronic episodes of complete or incomplete gastrointestinal obstruction.<sup>1-25</sup> Symptomatic patients may present with tell-tale signs, including a painless soft abdominal mass and *ascites* in those with a severe form of the disease.<sup>1-6</sup> However, acute emergencies due to perforation are rare.<sup>28</sup> In patients with secondary SEP, a clinician may be alerted to the possible diagnosis due to predisposing factors such as PD, intraperitoneal shunts or autoimmune conditions.<sup>1-5</sup>

## Diagnostic Modalities

A diagnosis of SEP is facilitated by the patient's history, existing predisposing factors, various biochemical parameters, radiological imaging and, above all, a high index of clinical suspicion.<sup>1-6,10-26</sup> Clinical indicators of possible secondary SEP include predisposing factors presenting with unexplained abdominal discomfort such as abdominal tuberculosis, systemic *lupus erythematosus* or PD.<sup>1-6,16,20,26</sup> Radiological imaging can also support the diagnosis, starting with abdominal X-rays, barium studies and ultrasonography and progressing to abdominal computed tomography (CT) and, in occasional cases, contrast-enhanced magnetic resonance imaging (MRI).<sup>11,12,17,24,27,29-32</sup>

Abdominal X-ray findings are likely to be non-specific and would indicate features of bowel obstruction, including dilated loops of the small intestine with multiple air fluid levels and, occasionally, bowel wall and peritoneal calcification [Figure 3A].<sup>1,2,5,24,30</sup> On the other hand, a barium study may show central clumping of the gut—often described as a cauliflower sign or accordion pattern—as a consequence of membrane encasing.<sup>1,2,5,16,24,30,32</sup> This encasement may also affect the gut functionally, leading to prolonged intestinal transit time.<sup>1,15,32</sup> For obvious reasons, a barium study is not an option for patients presenting with intestinal obstruction. Ultrasonography has been reported to facilitate the diagnosis of SEP and may reveal dilated bowel segments encased by a dense fibrous membrane.<sup>1,33</sup> Adherent bowel segments of various diameters may be found arranged in a concertina fashion with a narrow posterior base.<sup>1,2,16,24</sup> A thickened peritoneal layer may be noted, appearing as a thick rim of echo-poor tissue with free or loculated abdominal fluid.<sup>1,3,4</sup>

Contrast-enhanced CT is the most reliable investigative method to diagnose SEP; features include a central accumulation of the small intestine encased



**Figure 3A–B:** **A:** Plain abdominal X-ray showing bowel wall calcification (arrow) and peritoneal calcification with centrally clumped dilated small bowel loops (arrowhead) in a patient with sclerosing encapsulating peritonitis. **B:** Abdominal computed tomography revealing encased loops of bowel (arrow) by a distinct membrane (arrowheads) in a patient with sclerosing encapsulating peritonitis.

Figure 3A reproduced with permission from Candido PC, Werner AF, Pereira IM, Matos BA, Pfeilsticker RM, Filho RS. Sclerosing encapsulating peritonitis: A case report.<sup>24</sup>

by a dense membrane with a contrast-free periphery [Figure 3B].<sup>1–3,17,24,27,30</sup> The presence of additional findings may further facilitate the diagnosis, including: intestinal obstruction; *ascites*; peritoneal or mesenteric thickening; thickening of the small bowel wall; a soft-tissue density mantle; calcification of the serosal bowel wall over the liver capsule, spleen or peritoneal wall; localised fluid collection; and lymphadenopathy.<sup>1–3,17,24</sup> Calcification often occurs around blood capillaries and may extend into the serosal and muscular layers; the presence of this feature is important as it could influence both the integrity of the gut and reflect the difficulty in dissecting the membrane from the bowel wall.<sup>3</sup> Severe adhesions in calcified areas between the bowel wall and membrane makes their identification and separation very hazardous, with the potential risk of perforation.<sup>3</sup> A multidetector CT scan with axial, sagittal and coronal reconstruction provides greater accuracy in detecting characteristic findings, excluding other differential diagnoses and facilitating the decision-making process with regards to surgical interventions.<sup>1–4,17</sup> As CT scans are relatively cheaper, more widely available and more reliable in detecting certain findings, they are the obvious choice for establishing a diagnosis of SEP.<sup>1–4,17,24,27</sup> Reportedly, CT scans are able to reveal characteristic findings of SEP including peritoneal thickening, loculated fluid collection, calcification, congregated small bowel loops in the centre of the abdomen and peritoneal enhancement, with sensitivity rates of 100%, 90%, 70%, 60% and 50%, respectively.<sup>24</sup> The role of MRI in the diagnosis of SEP is limited, although some research indicates that this modality may be marginally superior to CT imaging.<sup>29</sup>

Certain biochemical parameters have been reported as indicators for SEP among patients undergoing

PD with a catheter *in situ*. These include haemorrhagic effluent, elevated levels of anti-inflammatory mediators and markers of coagulation-fibrinolysis such as interleukin-6 and fibrin/fibrinogen degradation products.<sup>3</sup> A definitive diagnosis is achieved by macroscopically confirming the encapsulation of the intestines via biopsy. In addition, diagnostic laparoscopies are recommended, particularly for patients undergoing PD prior to catheter removal.<sup>1–3,16</sup> Moreover, a pathological examination will reveal a complete loss of the mesothelium associated with significant interstitial thickening composed of fibroblasts and collagen deposition within the peritoneal membrane.<sup>34</sup> Inflammatory cells are invariably present but leukocyte infiltration is not a critical part of the diagnosis.<sup>34</sup>

## Differential Diagnosis

Recurrent abdominal pain, a predominant feature suggestive of acute, subacute or chronic obstruction, may mimic several other conditions.<sup>1,2,5,16</sup> Although postoperative adhesions are the most common cause of intestinal obstruction, the absence of other predisposing conditions in cases of idiopathic SEP often leads to diagnoses of internal herniation, congenital PE, intestinal malrotation or secondary peritonitis, among others.<sup>1–5,11,18,30</sup> Patients with secondary SEP warrant investigations to confirm the presence of predisposing conditions; these should include measurements of erythrocyte sedimentation rate, *sputum* tests for tuberculosis, ascetic fluid tests for adenosine deaminase levels, examinations for suspected abdominal tuberculosis and, in some cases, laparoscopies and biopsies.<sup>1,2,35,36</sup> Similar investigations would be required to rule out other possible diagnoses

including autoimmune conditions or pelvic ovarian inflammatory pathologies.<sup>1-4</sup> Other differential diagnoses include retractile mesenteritis, sclerosing malignant lymphomas, malignant primary mesenteric tumours and other metastatic neoplasms.<sup>1-5,16</sup>

Features that may help to differentiate SEP from other causes of intestinal obstruction are its chronic course, a palpable abdominal mass due to the clustering of the intestines from encapsulation and indolent peritonitis in the absence of a positive peritoneal culture. Moreover, among patients undergoing PD, bloody solute, declining small solute clearance and ultrafiltration failure may be noted.<sup>3</sup> However, despite various clinical observations, radiological findings and investigations, a preoperative diagnosis of SEP may still elude the clinician. In such cases, a definitive diagnosis of SEP may only be achieved by laparoscopy or laparotomy and histological confirmation.<sup>5,16,26</sup>

## Management

### CONSERVATIVE TREATMENT

Evidence in the literature indicates that it is prudent to manage patients with minimal abdominal symptoms conservatively, with bowel rest, nasogastric decompression and either enteral or parenteral nutritional support.<sup>1-6,10-24,37</sup> As a considerable number of patients with recurrent abdominal complaints have nutritional problems, addressing these issues is an important component of management.<sup>1-4,24</sup> Enteral feeding is recommended for any patient who can tolerate oral nutrition; when this is not feasible, parenteral nutrition should be considered.<sup>1-3,37</sup> Improving the nutritional status of these patients is of paramount importance as it may improve the response to conservative management or avoid subsequent surgical complications such as infection and *fistulae*.<sup>1-3,5,37</sup>

Drug therapy may be initiated for patients who fail to respond to conservative treatment, including tamoxifen, steroids, colchicine, azathioprine and mycophenolic acid.<sup>1,2,5,15,37-40</sup> Tamoxifen acts as an oestrogen receptor modulator that inhibits the fibroblastic production of transforming growth factor beta, while steroids inhibit collagen synthesis and maturation by suppressing the inflammatory process within the peritoneal membrane.<sup>1,2,38,40</sup> On the other hand, colchicine inhibits the messenger ribonucleic acid expression of transforming factor beta, thereby exhibiting an anti-inflammatory action.<sup>2,38</sup> Some reports have described the effectiveness of these medications for patients with idiopathic SEP.<sup>1,2,38,40</sup> Success has also been achieved with these drugs for

patients with previous failed surgical interventions in which complete excision and adhesiolysis could not be achieved (e.g. those with type II or III SEP).<sup>38,40</sup> Furthermore, patients who continue to have recurrent postoperative symptoms may also benefit from the use of these drugs.<sup>2</sup> The role of various drugs in SEP treatment, including hormones and immunosuppressive and anti-inflammatories, requires further investigation.

### SURGICAL INTERVENTIONS

Patients with severe symptoms of intestinal obstruction, those with virgin abdomens and those who do not respond to conservative management may be candidates for surgical interventions.<sup>1-6,10-24,26,27,31,33,34</sup> Surgical options include membrane excision plus adhesiolysis or, for patients with a gut injury, resection plus *anastomosis* with or without a protective enterostomy. An integral part of surgery is the complete excision of the membrane; this ensures a reduction in the recurrence rate.<sup>1-6,16,30,31,34</sup> Fibrotic membranous sacs which envelop and encase the coiled intestinal loops like thick plastic bags may pose a technical challenge.<sup>2,3,6,16,34</sup> Separating these sacs from the underlying bowel loops may require multiple longitudinal and transverse incisions; this may facilitate the stripping of the membrane in order to allow the underlying intestines to return to their normal function and length.<sup>3,5,16,34</sup> Failure to peel off or excise these membranes, or intense difficulties during the removal process, may result in gut perforation.<sup>1-6</sup> In most instances, an enterotomy forms the primary repair, while intestinal resection is reserved for cases with extensive injuries or suspected or confirmed loss of the vascular integrity of the gut.<sup>2,3,5,16</sup> Resection, particularly when not obviously indicated, could result in increased morbidity and mortality for these patients.<sup>2,3,5,16</sup> Kawanishi *et al.* reported a mortality rate of 4% among PD-related secondary SEP patients undergoing adhesiolysis alone in comparison to 82% for those who underwent an enterectomy and *anastomosis* in a separate study.<sup>3,41</sup>

One important factor predicting postsurgical outcomes is peritoneal deterioration, the occurrence of which has been found to increase with the duration of PD, particularly in cases that have extended beyond 10 years.<sup>3</sup> In such patients, the capsules are poorly demarcated from the intestinal wall and imprecise enterolysis can hence easily result in intestinal perforation.<sup>3</sup> Peritoneal calcification around capillaries could also increase the risk of perforation during dissection.<sup>3</sup> Following complete excision of the membrane, placing anti-adhesive substances between the bowel loops before closing the abdomen

Table 3: Demographic, clinical and treatment details of cases of sclerosing encapsulating peritonitis\*

| Author and year of case series         | n   | Mean age in years (range) | Male-to-female ratio | Symptom duration | Symptoms/signs, n (%)  | Imaging modality  | Time of diagnosis, n (%)   | Treatment, n (%)   | Surgery type, n (%)   | Intraoperative findings, n (%)   | Complications, n (%)   |
|--|-----|---------------------------|----------------------|------------------|--|---|--|--|---|--|--|
| Li <i>et al.</i> <sup>5</sup> 2014     | 65  | 39 (14–79)                | 57:8                 | 3.9 ± 6.7 years  | <ul style="list-style-type: none"> <li>• Ab pain, 56 (86.2)</li> <li>• Ab distension, 53 (81.5)</li> <li>• Nausea/vomiting, 35 (53.8)</li> <li>• Fever, 19 (29.2)</li> <li>• Ab mass, 21 (32.3)</li> </ul> | -   | <ul style="list-style-type: none"> <li>• Preop imaging, 31 (47.7)</li> <li>• Operative, 34 (52.3)</li> </ul>                       | <ul style="list-style-type: none"> <li>• Surgery, 65 (100.0)</li> <li>-Emergency, 19 (29.2)</li> <li>-Elective, 46 (70.7)</li> </ul> | <ul style="list-style-type: none"> <li>• ME, ADH and intestinal stenting, 42 (65.0)</li> <li>• ME and ADH, 23 (35.0)</li> </ul>   | <ul style="list-style-type: none"> <li>• Type I, 16 (24.0)</li> <li>• Type II, 16 (24.0)</li> <li>• Type III, 15 (23.0)</li> </ul> | <ul style="list-style-type: none"> <li>• Rec obstruction, 4 (6.1)</li> </ul>   |
| Singh <i>et al.</i> <sup>26</sup> 2013 | 18  | 43 (16–70)                | 8:10                 | 0.5–60 months    | <ul style="list-style-type: none"> <li>• Ab pain, 18 (100.0)</li> <li>• Ab mass, 2 (11.1)</li> <li>• Ab TB, 9 (50.0)</li> </ul>  | • CT  | -  | <ul style="list-style-type: none"> <li>• Conservative treatment, 1 (5.6)</li> <li>• Surgery, 17 (94.4)</li> </ul>                    | <ul style="list-style-type: none"> <li>• ME and ADH, 15 (83.3)</li> <li>• ME, ADH and ileostomy, 2 (11.1)</li> </ul>  | • Type II, 17 (100.0)  | <ul style="list-style-type: none"> <li>• Death due to liver failure, 1 (5.6)<sup>†</sup></li> </ul>                                |
| Yang <i>et al.</i> <sup>39</sup> 2009  | 6   | 43.7 (39–48)              | 4:2                  | 3–60 months      | <ul style="list-style-type: none"> <li>• Ab pain only, 1 (16.6)</li> <li>• Ab pain and intestinal obstruction, 5 (83.3)</li> </ul>   | <ul style="list-style-type: none"> <li>• X-ray</li> <li>• CT</li> </ul>   | -  | <ul style="list-style-type: none"> <li>• Surgery, 6 (100.0)</li> </ul>   | <ul style="list-style-type: none"> <li>• ME, ADH, mesenteric plication and intestinal stenting, 5 (83.3)</li> <li>• ME, ADH and jejunum resection, 1 (16.6)</li> </ul>  | -  | <ul style="list-style-type: none"> <li>• Rec obstruction, 2 (33.3)<sup>‡</sup></li> </ul>  |
| Wei <i>et al.</i> <sup>16</sup> 2009   | 24  | 34 (15–57)                | 9:15                 | 0.1–26 months    | <ul style="list-style-type: none"> <li>• Intestinal obstruction only, 11 (45.8)</li> <li>• Ab mass, 10 (41.7)</li> <li>• Asymptomatic Ab mass, 3 (12.5)</li> </ul>   | <ul style="list-style-type: none"> <li>• X-ray</li> <li>• Barium</li> <li>• US</li> <li>• CT</li> </ul>                 | <ul style="list-style-type: none"> <li>• Preop, 4 (16.7)</li> <li>• Operative, 20 (83.3)</li> </ul>                                | <ul style="list-style-type: none"> <li>• Surgery, 24 (100.0)</li> </ul>  | <ul style="list-style-type: none"> <li>• ME, ADH and appendectomy, 17 (70.8)</li> <li>• ME, ADH and enterotomy, 2 (8.3)</li> <li>• ME, ADH and cecofixation, 2 (8.3)</li> <li>• ME and ADH, 3 (12.5)</li> </ul> | <ul style="list-style-type: none"> <li>• Type I, 14 (58.3)</li> <li>• Type II, 6 (25.0)</li> <li>• Type III, 4 (16.6)</li> </ul>   | <ul style="list-style-type: none"> <li>• IBO, 3 (12.5)</li> <li>• Adhesive ileum obstruction, 3 (12.5)</li> </ul>                  |
| Xu <i>et al.</i> <sup>19</sup> 2007    | 5   | 37 (21–49)                | 3:2                  | 0.5–120 months   | <ul style="list-style-type: none"> <li>• Recurrent Ab pain, 5 (100.0)</li> </ul>   | <ul style="list-style-type: none"> <li>• X-ray</li> <li>• CT</li> <li>• Endo</li> </ul>                                 | <ul style="list-style-type: none"> <li>• Preop CT imaging, 4 (80%)</li> </ul>  | <ul style="list-style-type: none"> <li>• Surgery, 5 (100.0)</li> </ul>   | <ul style="list-style-type: none"> <li>• ME and ADH, 4 (80)</li> <li>• ADH and jejunum resection, 1 (20)</li> </ul>   | <ul style="list-style-type: none"> <li>• Type I, 3 (60.0)</li> <li>• Type II, 2 (40.0)</li> </ul>                                  | <ul style="list-style-type: none"> <li>• Rec obstruction, 1 (20.0)</li> </ul>  |
| Total                                  | 118 | 39.3 (14–79)              | 81:37                | 0.1–120 months   | <ul style="list-style-type: none"> <li>• Ab pain, 85 (72.0)</li> <li>• Ab distension, 53 (44.9)</li> <li>• Ab mass, 36 (30.5)</li> <li>• Nausea/vomiting, 35 (29.7)</li> <li>• Other, 43 (36.4)</li> </ul> | <ul style="list-style-type: none"> <li>• X-ray</li> <li>• Barium</li> <li>• US</li> <li>• CT</li> <li>• Endo</li> </ul> | <ul style="list-style-type: none"> <li>• Operative, 54 (45.7)</li> <li>• Preop, 35 (29.7)</li> <li>• Unknown, 29 (24.6)</li> </ul> | <ul style="list-style-type: none"> <li>• Surgery, 117 (99.2)</li> <li>• Other, 1 (0.8)</li> </ul>                                    | <ul style="list-style-type: none"> <li>• ME, ADH and other procedures, 71 (60.7)</li> <li>• ME and ADH, 45 (38.5)</li> <li>• Other, 1 (0.9)</li> </ul>  | <ul style="list-style-type: none"> <li>• Type I, 33 (43.4)</li> <li>• Type II, 24 (31.6)</li> <li>• Type III, 19 (25.0)</li> </ul> | <ul style="list-style-type: none"> <li>• None, 104 (88.1)</li> <li>• Rec obstruction, 7 (5.9)</li> <li>• Other, 7 (5.9)</li> </ul> |

Ab = abdominal; Preop = preoperative; ME = membrane excision; ADH = adhesiolysis; CT = computed tomography; Rec = recurring; TB = tuberculosis; US = ultrasonography; IBO = inflammatory bowel obstruction; Endo = endoscopy. \*Literature review including only case series with five or more cases. †Patient was managed conservatively and did not undergo surgery. ‡Over a 33-month follow-up period.

may reduce the risk of postoperative adhesions;<sup>2,5,16</sup> however, the effect of these substances for patients with partially excised membranes is debatable.<sup>2</sup>

Even though laparoscopies may have both diagnostic and therapeutic purposes, few reports indicate this role clearly.<sup>2,35,42–44</sup> Technically, laparoscopies may be challenging in patients with an advanced abdominal cocoon, such as those with type III SEP. There is a potential risk of gut injury during the *trocar* insertion and separation and resection of the membrane from the underlying intestine.<sup>2,43</sup> However, the risk of injury during *trocar* insertion can be reduced or avoided by employing an open technique during the initial insertion for insufflation.<sup>43</sup>

### Progressive Sclerosing Encapsulating Peritonitis

For patients with secondary PD-related SEP, the condition is believed to progress rapidly; hence, it is important to initiate treatment immediately after diagnosis.<sup>3</sup> Stage one is considered the inflammatory stage, with the inflammation denoted by elevated serum C-reactive protein levels, fibrin/fibrin degradation products and effluent occult blood. Treatment during this stage is with corticosteroids.<sup>3</sup> In stage two, the encapsulating stage, inflammation is less pronounced, although there is progression of adhesion and encapsulation. Symptoms of bowel obstruction usually appear at this point. Patients at this stage can be managed with total parenteral nutrition.<sup>3</sup> In stage three, the patient experiences bowel obstruction without inflammation; this stage must be managed with complete membrane excision of the membrane and adhesiolysis.<sup>3</sup>

### Postoperative Complications

Postoperative complications include early bowel obstruction, intra-abdominal infections, enterocutaneous *fistulae*, short bowel syndrome and bowel perforation.<sup>1–6,16,39,44</sup> Predisposing factors that enhance the risk of a postoperative obstruction include significant manipulation of the gut, oedema and prolonged duration of the operation.<sup>2,5,35</sup> Several methods can be employed to reduce the incidence of postoperative intestinal obstruction, including intestinal intubation through the orifice of the appendix in patients with type II or III SEP, the use of steroids to reduce oedema, intestinal *stasis* or bacterial translocation and somatostatin administration to reduce secretion and intestinal distension.<sup>1–3,45</sup> Inserting long intestinal tube splinting may prevent a potential obstruction; thus, it

would be wise to fix the bowel in a favourable position.<sup>4</sup> In one report, the postoperative complication rate among patients who underwent enterolysis alone was 9.1% compared to 6.1% among those with internal splinting; moreover, the recurrence rate of intestinal obstruction was significantly higher (40% versus 6.7%;  $P = 0.02$ ).<sup>44</sup> Spontaneous complications such as perforation and enterocutaneous *fistulae* are rare and are usually the *sequelae* of a missed iatrogenic injury.<sup>2</sup>

The prognosis of secondary SEP depends on the cause of the condition. For patients undergoing PD for end-stage renal disease, the outcome is generally poor with reported mortality rates as high as 69%.<sup>3,46</sup> The reported mortality among surgical patients is 45–82% and occurs within a few weeks or months of the interventions.<sup>3</sup> However, recent research suggests a good outcome following surgical intervention with either no or minimal perioperative mortality (survival rate: 94%) after three years of follow-up.<sup>24,26</sup>

### Summary of Literature Review

A review of case series involving five or more cases of SEP revealed a total of 118 cases [Table 3].<sup>5,16,19,26,39</sup> The mean patient age was 39 years with the majority being male (68.0%). Type I SEP was the most common finding (43.4%). The predominant symptom of presentation was abdominal pain (72.0%), followed by abdominal distension (44.9%) and abdominal masses (30.5%). The majority of the patients were diagnosed on the operating table (45.7%). Almost all of the patients underwent surgical exploration (99.2%); of these, surgery included either excision of the membrane or adhesiolysis (100.0%), along with other procedures such as resection, *anastomosis*, mesenteric plication or intestinal stenting. Outcomes were generally good, with recurrent obstruction seen in only 5.9% of patients. One patient was treated conservatively and died due to liver failure; this was the only death reported (0.8%). Patients with abdominal tuberculosis (7.6%) received adequate therapy with antitubercular drugs.

### Conclusion

SEP is a rare clinical entity and is often encountered unexpectedly in patients with acute intestinal obstruction. A high index of clinical suspicion in susceptible patients is necessary to achieve a preoperative diagnosis. Radiological imaging, particularly CT scans, plays a major role in establishing the diagnosis. Conservative management is the ideal approach for patients who present with mild



symptoms; however, those with severe intestinal obstruction are likely to require surgical intervention, usually comprising of the complete excision of the membrane, adhesiolysis and occasionally resection.

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