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Unraveling the enigma of sclerosing encapsulating peritonitis: a comprehensive review

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Sclerosing encapsulating peritonitis (SEP) is a rare but serious condition characterized by the progressive formation of a dense fibrous sheath encasing the small bowel within the peritoneal cavity. This review provides a comprehensive overview of the current understanding of SEP, focusing on its etiology, clinical presentation, diagnostic modalities, and management strategies. SEP can be classified into primary and secondary forms, each with distinct etiologies and treatment approaches. Primary SEP typically presents with acute or subacute bowel obstruction symptoms, necessitating surgical intervention to excise the fibrous sheath and relieve the obstruction. Secondary SEP often occurs in patients undergoing peritoneal dialysis, with cessation of dialysis being a key component of management. Medical treatments, including corticosteroids, immunosuppressive agents, and nutritional support, may complement surgical intervention, particularly in cases of secondary SEP. Advanced imaging techniques and personalized medicine approaches show promise in improving diagnostic accuracy and tailoring treatment strategies to individual patients. Future research directions include investigating targeted pharmacological therapies, exploring minimally invasive surgical techniques, and conducting long-term follow-up studies to evaluate treatment efficacy and disease recurrence. Multidisciplinary care teams play a crucial role in the comprehensive management of SEP, emphasizing collaboration among various specialties to optimize patient outcomes.

Keywords: Peritoneal fibrosis; Abdominal cocoon; Peritoneal dialysis; Peritoneal diseases; Immunosuppressive agents

INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) is a chronic inflammatory disorder with an unknown etiology that is thought to arise from recurrent episodes of low-grade or subclinical peritonitis presenting without specific abdominal signs. Over time, this condition progresses to sclerosis and membrane formation, leading to the development of a cocoon-like structure [1].

The hallmark of SEP is a thick, grayish-white fibrotic membrane that partially or completely encases the small bowel and may extend to other intra-abdominal organs [2]. SEP is categorized into primary and secondary forms. Primary SEP, also referred to as abdominal cocoon syndrome, occurs in the absence of any identifiable associated conditions. In contrast, secondary

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SEP develops due to underlying conditions that lead to peritoneal inflammation and fibroblastic proliferation [3].

Early diagnosis of SEP is challenging due to the nonspecific nature of its symptoms. Therefore, radiological imaging is essential for the preoperative diagnosis of this condition [1]. Treatment approaches for SEP vary depending on the disease stage and type, ranging from conservative management to interventional procedures [4, 5]. This article offers a comprehensive review of the literature, including case series and reports, to explore the definition, etiology, clinical presentation, radiological manifestations, diagnostic methods, treatment options, prognosis, and histopathological features of SEP.

DEFINITIONS

Peritoneal encapsulation

Peritoneal encapsulation, first described by Cleland in 1868, is a developmental anomaly marked by a peritoneal membrane [1]. This membrane, originating from the yolk sac peritoneum in early fetal life [6], typically lies between the mesocolon and omentum, with most of the small intestines located behind it. A distinctive aspect of peritoneal encapsulation is that it is not associated with inflammatory processes [1]. Cleland's 1868 description highlighted peritoneal encapsulation's developmental origin, anatomical features, and noninflammatory nature [7].

Sclerosing encapsulating peritonitis

SEP is caused by peritoneal inflammation due to various factors [1, 5, 8]. Unlike peritoneal encapsulation, SEP is characterized by a dull, fibrous membrane with inflammatory cells encasing the intestines [9–11]. This thick fibrocollagenous membrane may partially or completely surround abdominal organs. The term "peritonitis chronica fibrosa incapsulata" was first used by Owtschinnikow in 1907 to describe SEP, which can be either primary (idiopathic) or secondary [12]. Idiopathic SEP, also known as "abdominal cocoon syndrome," was initially named by Foo et al. [13] in 1978. There are 3 types of abdominal cocoon syndrome, categorized based on the extent of the encasing membrane: types I and II involve part or all of the small intestine, while type III extends to other organs such as the appendix, colon, stomach, liver, and ovaries (Fig. 1) [8, 14].

ETIOLOGY AND PATHOPHYSIOLOGY

Primary SEP

Primary SEP, often considered idiopathic, lacks a clear mechanism but is relatively common among young men in tropical and subtropical regions [1, 5, 12]. It may involve cytokines and fibroblasts in the development of peritoneal fibrosis and angiogenesis. Potential causes include retrograde menstruation, viral infections, retrograde peritonitis, and immune system damage, all of which can lead to inflammation and fibrosis [1, 15]. However, the presence of SEP in men, premenopausal women, and children poses challenges to these theories [2, 11]. Some researchers have suggested a possible link to vascular anomalies and omental hypoplasia [8].

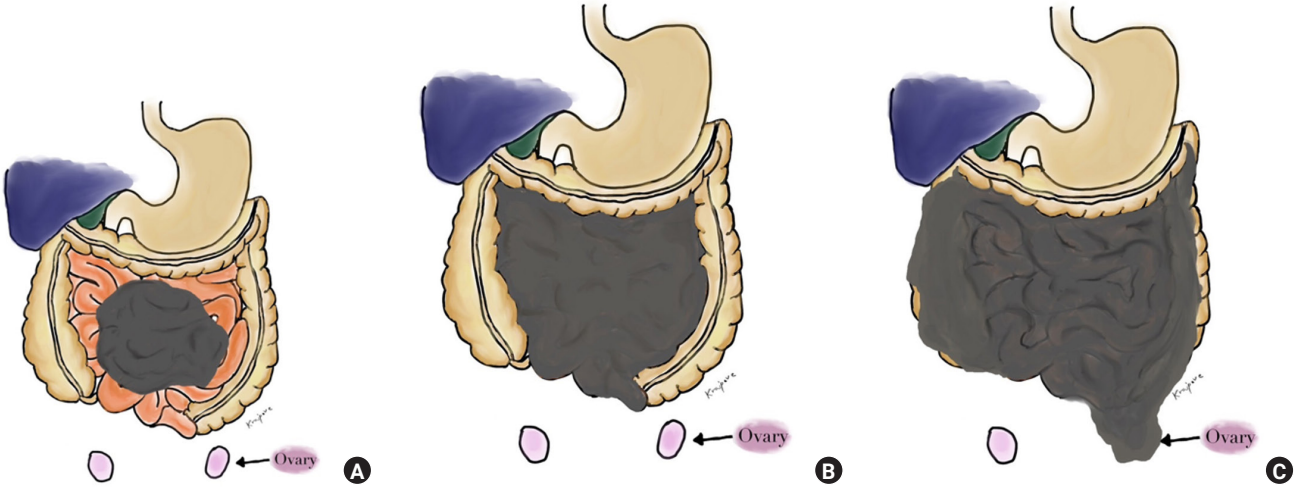

Fig. 1. The categorizations of primary sclerosing encapsulating peritonitis (SEP) as (A) type I, (B) type II, and (C) type III. In type I and II cases, the membrane (depicted in gray) encapsulates a portion and the entirety of the small intestine, respectively. Type III SEP is characterized by a membrane (depicted in gray) encompassing the entire small bowel along with additional organs, such as the ovaries and colon. Illustration by Kraipop Wongwaiyut.

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Secondary SEP

Secondary SEP is more prevalent than idiopathic SEP and is associated with specific triggers that lead to peritoneal inflammation. The primary cause is peritoneal dialysis (PD), during which the peritoneum is damaged by prolonged exposure to PD fluids and bacterial peritonitis [16]. Some studies support the "two-hit" hypothesis for secondary SEP [5]. According to this model, the "first hit" involves noninflammatory peritoneal sclerosis due to repeated dialysis sessions [17]. Evidence for this is seen in the cumulative incidence of structural and functional changes in the peritoneal membrane, which are typically linked to SEP and show a significant increase as PD exposure continues [18]. The "second hit" is characterized by the action of proinflammatory and proangiogenic cytokines that progressively harm the peritoneum [17, 19]. The following cytokines may trigger secondary SEP:

- Transforming growth factor β1 (TGF-β1) [17]
- Interleukin-6 (IL-6) [17]
- Cellular communication network factor 2 (CCN2) [17]
- Vascular endothelial growth factor (VEGF) [19]
- Endothelial nitric oxide synthase (eNOS) [19]

Mesothelial cells may undergo a mesothelial-to-mesenchymal transition, causing a loss of their polarized cytoskeletal organization and cell-to-cell contacts [20]. As a result, they acquire a myofibroblast-like phenotype characterized by increased motility and the secretion of extracellular matrix compounds, and profibrotic and angiogenetic cytokines [20]. This cascade of events leads to the depletion of mesothelial cells, increased production of extracellular matrix components such as collagen type 1, alpha 1, fibrogenesis, and reduced fibrolytic activity of mesothelial cells [20]. Ultimately, these processes culminate in the formation of a fibrocollagenous cocoon [19]. In PD, genetic variations in the receptor for advanced glycation end products may predispose individuals to peritoneal deterioration [5]. Additional contributors to secondary SEP include abdominal tuberculosis, autoimmune disorders, drug use, ovarian disorders, abdominal surgery, peritoneal shunts, and fibrogenic foreign materials, as detailed in Table 1 [5, 16].

EPIDEMIOLOGY

Due to its rarity and diverse causes, the overall incidence and prevalence of SEP are not well-documented. An analysis of 169 primary SEP cases described in 67 journals (Table 2) shows that most cases have been reported in tropical and subtropical regions, especially in Asia (73.4%) (Fig. 2). The average age of patients was 40.2 years, ranging from 7 to 90 years, with a male to female ratio of 3:1 (Fig. 3) [4, 6, 7, 9–12, 14, 15, 21–78].

For secondary SEP caused by PD, the annual incidence ranges from 0.5% to 7.3%, potentially reaching 17.2% in patients on dialysis for 15 years or more [16, 18]. Rigby and Hawley [79] observed incidence rates of 1.9%, 6.4%, 10.8%, and 19.4% in patients undergoing PD for 2, 5, 6, and 8 years, respectively. Recent studies have suggested that improvements in dialysis techniques may reduce the incidence of SEP [79].

Table 1. Etiology of sclerosing encapsulating peritonitis

Etiology	Description
Primary (idiopathic)	Male to female ratio, 3:1
	Most commonly reported in Asia
Secondary	
Medication	β-Blockers: practolol, timolol, propranolol
	Methotrexate
	Antiepileptic drugs
Infection	Tuberculosis
	Nontuberculous mycobacteria
	Bacterial peritonitis
	Cytomegalovirus
	Fungus
	Parasite
Cirrhosis	
Protein S deficiency	
Organ transplantation	Liver
	Small intestine
	Renal
Rheumatologic/systemic inflammatory condition	Sarcoidosis
	Systemic lupus erythematosus
	Familial Mediterranean fever
Gastrointestinal tract neoplasm	
Gynecologic neoplasm	Luteinized thecoma
	Luteinizing granulosa cell tumor
Mechanical or chemical irritation	Peritoneal dialysis
	Intraperitoneal chemotherapy
	Ventriculoperitoneal shunt
	Peritoneovenous shunt
	Intraperitoneal iodine
	Abdominal trauma
	Intra-abdominal surgery
	Foreign body
	Talcum powder
	Asbestos
	Silica
	Endometriosis
	Dermoid cyst rupture
	Recurrent peritonitis

Adapted from Danford et al. [5], available under the Creative Commons Attribution NonCommercial license.

Table 2. Distribution of articles and number of cases with primary sclerosing encapsulating peritonitis according to countries

Country	No. of publications (%) (n = 67)	No. of patients (%) (n = 169)
Asia		
China	11 (16.4)	97 (57.4)
India	4 (6.0)	4 (2.4)
Pakistan	4 (6.0)	4 (2.4)
Iran	3 (4.5)	3 (1.8)
Nepal	3 (4.5)	3 (1.8)
Thailand	1 (1.5)	3 (1.8)
Qatar	1 (1.5)	2 (1.2)
Saudi Arabia	1 (1.5)	2 (1.2)
Bangladesh	1 (1.5)	1 (0.6)
Iraq	1 (1.5)	1 (0.6)
Korea	1 (1.5)	1 (0.6)
Palestine	1 (1.5)	1 (0.6)
Singapore	1 (1.5)	1 (0.6)
United Arab Emirates	1 (1.5)	1 (0.6)
Europe		
Türkiye	12 (17.9)	21 (12.4)
Greece	5 (7.5)	6 (3.6)
Portugal	1 (1.5)	2 (1.2)
Belgium	1 (1.5)	1 (0.6)
Denmark	1 (1.5)	1 (0.6)
North Macedonia	1 (1.5)	1 (0.6)
Spain	1 (1.5)	1 (0.6)
UK	1 (1.5)	1 (0.6)
America		
USA	4 (6.0)	4 (2.4)
Brazil	2 (3.0)	2 (1.2)
Africa		
Tunisia	1 (1.5)	2 (1.2)
Nigeria	1 (1.5)	1 (0.6)
Somalia	1 (1.5)	1 (0.6)
Sudan	1 (1.5)	1 (0.6)

Percentages may not total 100 due to rounding.

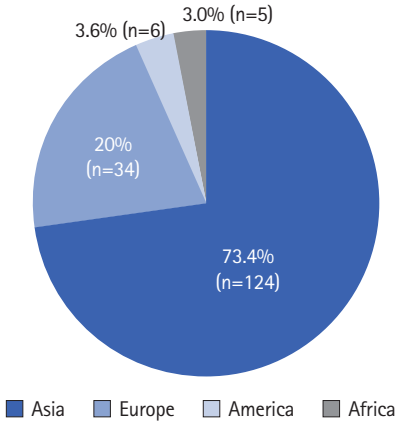

Fig. 2. Global distribution of primary sclerosing encapsulated peritonitis.

CLINICAL PRESENTATION

Common presentation

SEP symptoms vary depending on the affected area and the organs involved. Common symptoms include intermittent pain (33.1%) and small bowel obstruction (57.9%), which results from the intestine being compressed by fibrous tissue (Fig. 4) [4, 6, 7, 9–12, 14, 15, 21–78]. This compression leads to symptoms akin to ileus, including abdominal pain, nausea, loss of appetite, vomiting, constipation, and weight loss. The diagnosis is supported by a history of symptoms that resolved spontaneously [16, 54, 58, 67, 77].

In the most extensive case series, symptoms persisted for an average of 3.9 years before a diagnosis was made, with many patients exhibiting signs of malnutrition [65]. Despite its chronic nature, 59.2% of the cases necessitated emergency surgery due to acute complications such as obstruction, ischemia, or perforation [70].

Case series data

Palpable masses and ascites may be detected in certain cases [48, 65, 75]. SEP remains asymptomatic in the majority of cases or is incidentally discovered during laparotomy [14, 32]. In pubescent female patients, presentations may include infertility or tube obstruction [6, 76, 77]. In rare instances, patients may present with a groin mass, mimicking a groin hernia [38, 46]. Since SEP is rare and lacks specific symptoms, physicians should be particularly cautious when patients have recurring abdominal pain without a clear cause.

DIAGNOSTIC MODALITIES

SEP is a rare condition, both in its primary and secondary forms, and is frequently overlooked by physicians. Diagnosing it prior to surgery is challenging due to the nonspecific nature of early symptoms [8, 12, 15, 21]. A high degree of clinical suspicion and the use of imaging techniques are essential for an accurate diagnosis. In cases of secondary SEP, contributing factors may include abdominal tuberculosis, systemic lupus erythematosus, or PD [1, 37]. A review of 67 studies revealed that only 31% of primary SEP cases were correctly diagnosed before surgery [4, 6, 7, 9–12, 14, 15, 21–78]. Imaging studies play a crucial role in the diagnostic process. The use of biomarkers for secondary SEP remains a topic of ongoing debate [16, 18].

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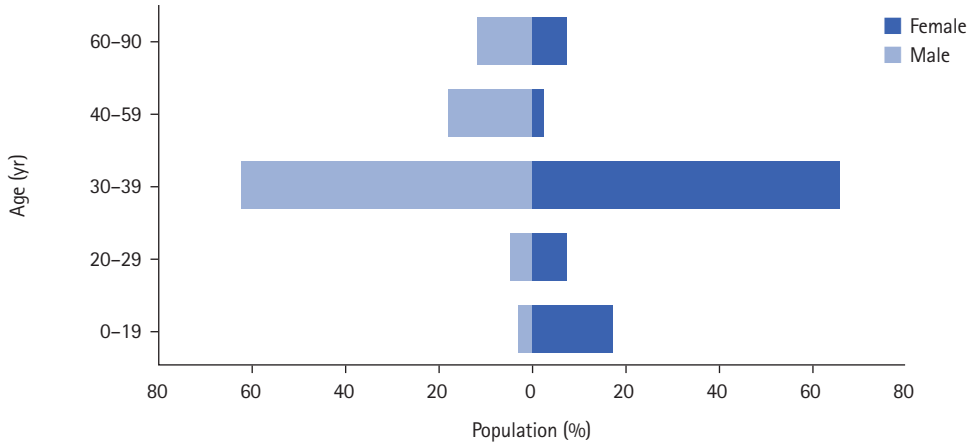

Fig. 3. Sex and age distribution of primary sclerosing encapsulated peritonitis.

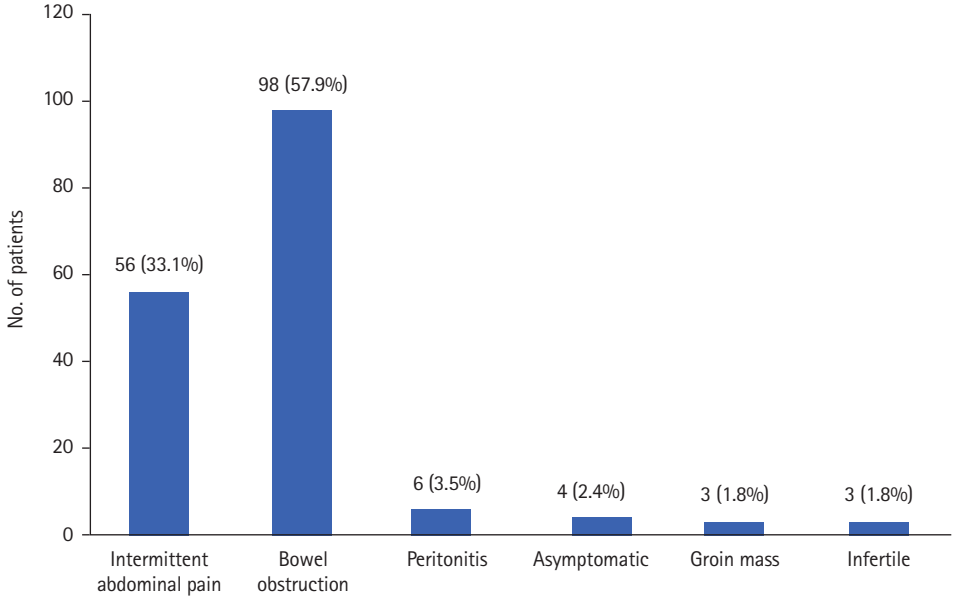

Fig. 4. Primary symptoms of primary sclerosing encapsulated peritonitis in 169 cases.

Imaging

Abdominal x-rays are frequently the initial choice for imaging due to their simplicity and ability to reveal nonspecific indicators of bowel obstruction, such as dilated loops, air-fluid levels, and occasional calcifications [1, 2, 8–10, 16, 37]. Barium studies can illustrate the small bowel as clumped and encased within a thickened peritoneum, producing a "cocoon" appearance and occasionally displaying a cauliflower sign [37]. However, these studies are impractical in cases of obstruction.

Ultrasonography is valuable in diagnosing SEP, as it reveals specific features such as a trilaminar bowel wall, bowel tethering, dilated and fixed loops, ascites, and membrane formation [8, 14, 61]. Although it can sometimes mimic an intestinal mass, in other instances, it may only display diffuse intestinal distention (Fig. 5) [7, 9, 22, 78].

Computed tomography (CT) scans significantly improve the diagnosis of SEP by providing a noninvasive and reliable method for early detection and management planning. Noncontrast CT imaging reveals small bowel loops clumped in the center of the abdomen, encased by a thick membrane, with features such as peritoneal calcification, thickening, fluid collections, and ascites [24, 26, 29]. Contrast-enhanced CT reveals a central accumulation of the small intestines surrounded by dense fibrous tissue (Fig. 6) [10, 43], with additional signs including intestinal obstruction, peritoneal or mesenteric thickening, small bowel wall thickening, fluid collection, and lymphadenopathy [10, 30, 41,

73]. Peritoneal thickening is often subjective, but a thickness over 2 mm is a useful threshold [37].

Recent research by Wang et al. [80] introduced a CT scoring system designed to predict surgical outcomes in SEP. This system evaluates 6 parameters: peritoneal thickening, peritoneal calcification, fluid loculation, tethering of the small bowel, and bowel wall thickening. A CT score of 15 or higher is associated with increased surgical mortality (area under the curve, 0.93; sensitivity, 88.9%; specificity, 82.1%). Patients with scores of 15 or above also exhibit higher mortality rates (61.5% vs. 4.2%, $P<0.001$), greater blood loss (400 mL vs. 50 mL, $P=0.007$), and a higher incidence of bowel perforation (61.5% vs. 12.5%, $P=0.006$) compared to those with lower scores.

Although magnetic resonance imaging (MRI) can yield results similar to other imaging modalities, it is less commonly utilized due to its longer duration and impracticality in urgent scenarios, such as cases of clinical bowel obstruction presenting in the emergency room. The advantages of MRI include the avoidance of ionizing radiation and enhanced imaging of bowel encasement and peritoneal thickening [54, 80].

Biomarkers in SEP

There are no specific biomarkers for predicting primary SEP [5, 37]. Laboratory findings associated with primary SEP are nonspecific and can be influenced by various factors. In contrast, secondary SEP in PD patients is characterized by specific biochemical indicators, such as hemorrhagic effluent and elevated levels of anti-inflammatory mediators and coagulation markers, including IL-6 and fibrin/fibrinogen degradation products [81]. Proposed biomarkers for secondary SEP include serum β_2 microglobulin (β_2MG), IL-1, dermatopontin (DPT), gelsolin (GSN), and retinol binding protein-4 (RBP 4) [18].

Yokoyama et al. [82] found that using serum β_2MG for SEP had a sensitivity of 64% and specificity of 80% at a cutoff level of 37 mg/dL. Lowering the cutoff to 30 mg/dL resulted in an odds ratio of 1. However, the direct pathophysiological link remains uncertain. To enhance clinical applications, future work should prioritize validating prognostic tools and biomarkers and developing streamlined diagnostic protocols that incorporate the most effective imaging techniques.

STAGING OF SEP

In primary SEP, there is no specific disease staging system; instead, the same staging system used for secondary SEP is applied. A staging system for PD-associated SEP has been proposed, which incorporates clinical, laboratory, and radiographic findings [83]. Nakamoto [83] categorized SEP patients into 4 stages: stage 1 (pre-SEP), stage 2 (inflammatory), stage 3 (encapsulating), and stage 4 (chronic). This classification is based on abdominal symptoms, inflammation, encapsulation, and intestinal observations (Table 3). Treatment strategies differ depending on the stage [18,

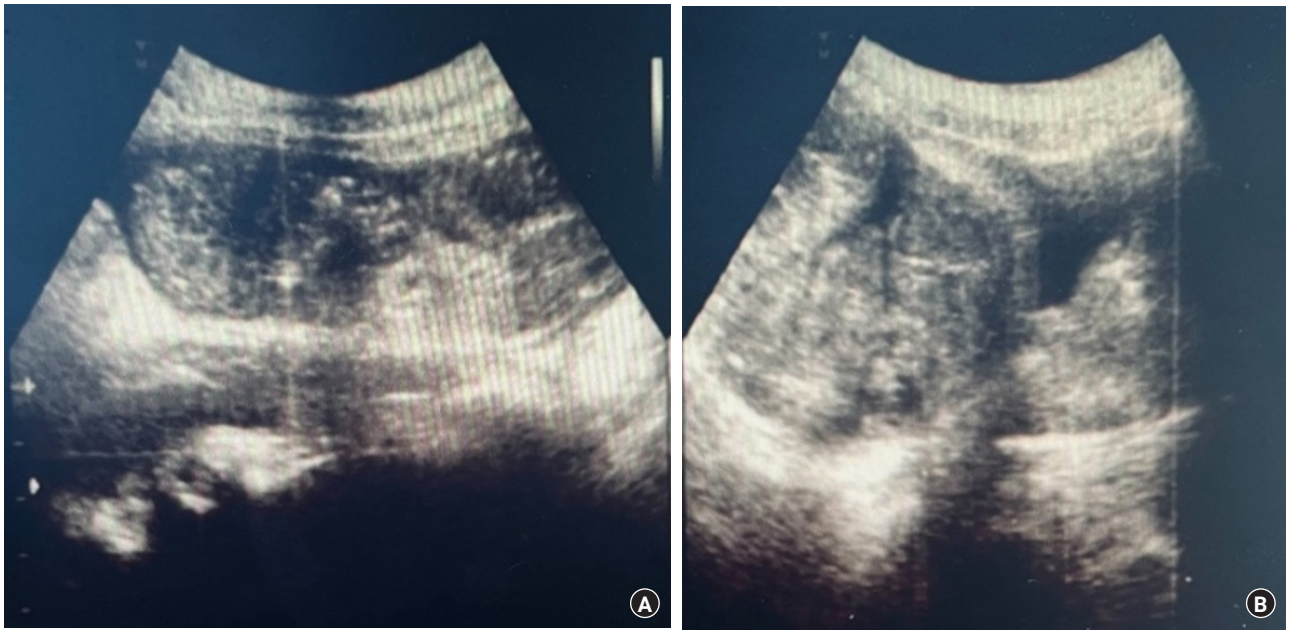

Fig. 5. Ultrasound examinations of the abdominal region in cases of primary sclerosing encapsulated peritonitis reveal widespread intestinal distension accompanied by the presence of ascites. (A) Distended small intestine. (B) Interloop ascites.

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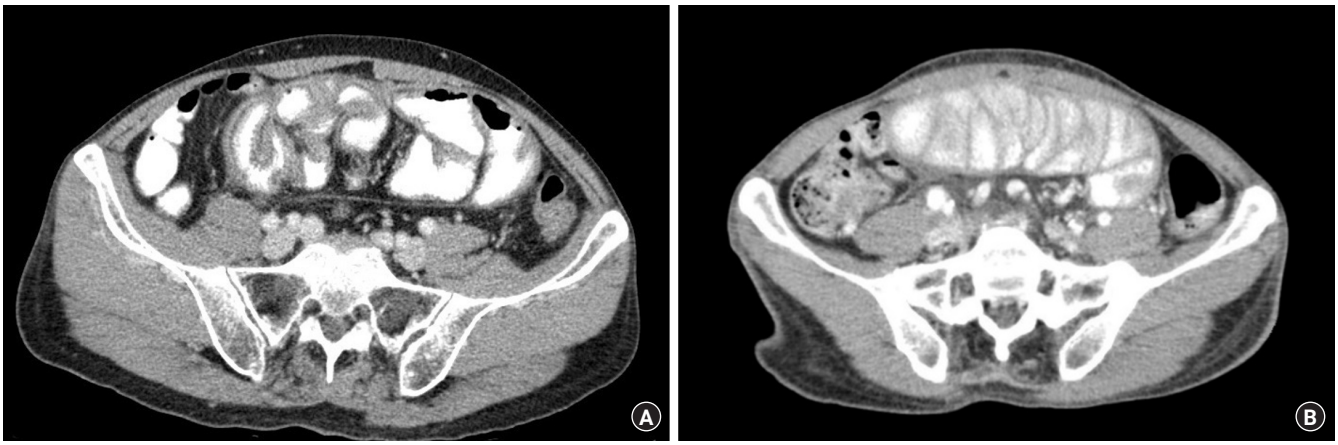

Fig. 6. Whole-abdomen computed tomography with oral and intravenous contrast. (A) Thick membrane surrounding the small intestine. (B) Stacking dilated small bowel loops inside a thick membrane-like sac.

Table 3. Stages of secondary sclerosing encapsulated peritonitis with associated clinical, laboratory, and radiographic profiles

Stage	Manifestation
Stage 1 (pre-SEP)	
Abdominal symptom	Mild abdominal discomfort
Laboratory finding	Low albumin level
Intra-abdominal finding	Bloody dialysate (hemoperitoneum)
	Ascites (mild to moderate)
	Calcifications in the peritoneum
	Mild inflammation
	Encapsulation none present
Stage 2 (inflammatory)	
Abdominal symptom	Mild to severe abdominal pain and encompass nausea, vomiting, anorexia, diarrhea, weight loss and systemic inflammation (fever)
Laboratory finding	Increased CRP, leukocytosis
Intra-abdominal finding	Ascites (mark)
	Moderate to severe inflammation
	Partial encapsulation
Stage 3 (encapsulating)	
Abdominal symptom	Intermittent ileus
Laboratory	Restoration of normal inflammatory markers
Intra-abdominal finding	Ascites with forming mass process
	Mild to moderate inflammation
	Encapsulation present
Stage 4 (chronic/ileus)	
Abdominal symptom	Persistent ileus, anorexia, and abdominal mass
Laboratory finding	Normal inflammatory markers
Intra-abdominal finding	Rigidity of the abdomen, fibrosis
	None to mild inflammation
	Encapsulation present

SEP, sclerosing encapsulating peritonitis; CRP, C-reactive protein.

83]. In primary SEP, the progression can occur in a few months [45]. For instance, a case series from Türkiye documented 2 patients who developed clinical SEP shortly after undergoing surgery for other conditions: one developed SEP 2 months post-left hemicolectomy for sigmoid cancer, and another 1 month post-transabdominal hysterectomy with bilateral salpingo-oophorectomy for endometrial cancer [45].

HISTOPATHOLOGY

Histopathological analysis is essential for diagnosing SEP. Peritoneal biopsies generally reveal fibroconnective tissue proliferation, inflammatory infiltration, and dilated lymphatic vessels [1, 65]. Although these features are not unique to SEP, their presence, in conjunction with surgical findings, supports the diagnosis [69]. The absence of foreign body granulomas and giant cells is instrumental in distinguishing SEP from conditions such as tuberculosis [8, 19]. SEP is marked by dense sheets of collagenous tissue that encapsulate and constrict the small bowel, along with mononuclear inflammation [17, 19]. For instance, one case displayed ileal hemorrhagic infarction due to adhesions, fibrocollagenous fiber proliferation in the serosa, mesentery, and appendix, and chronic inflammatory cells (Figs. 7, 8). While these histological features may overlap with other peritoneal diseases, the identification of a thickened fibrocollagenous membrane within the clinical context usually confirms SEP. Nevertheless, the growing use of CT imaging and clinical assessments has reduced the dependence on histopathology for diagnosing SEP in contemporary medical practice.

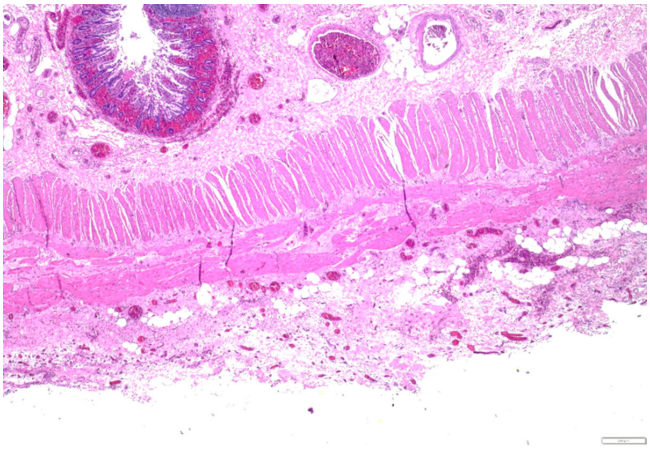

Fig. 7. The ileum shows acute transmural hemorrhagic infarction (hematoxylin-eosin, original magnification ×20). The mucosa loses its epithelial cells with hemorrhage. The submucosal layer shows congested vessels. The muscularis propria shows smooth muscles with degenerated nuclei. The serosa shows fibrosis.

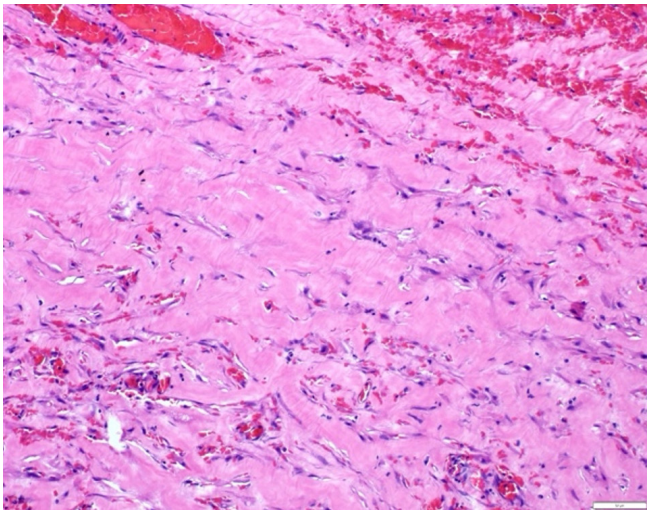

Fig. 8. Areas of fibrosis with hemorrhage are observed in the mesentery, serosa of the vermiform appendix, mesoappendix, and peritoneum (hematoxylin-eosin, original magnification ×100). These areas are composed of fibroblasts, spindle-shaped cells with oval-shaped nuclei, with small new vessel formations are demonstrated among the thick pink collagenous fibers and scattered a small number of chronic inflammatory cells.

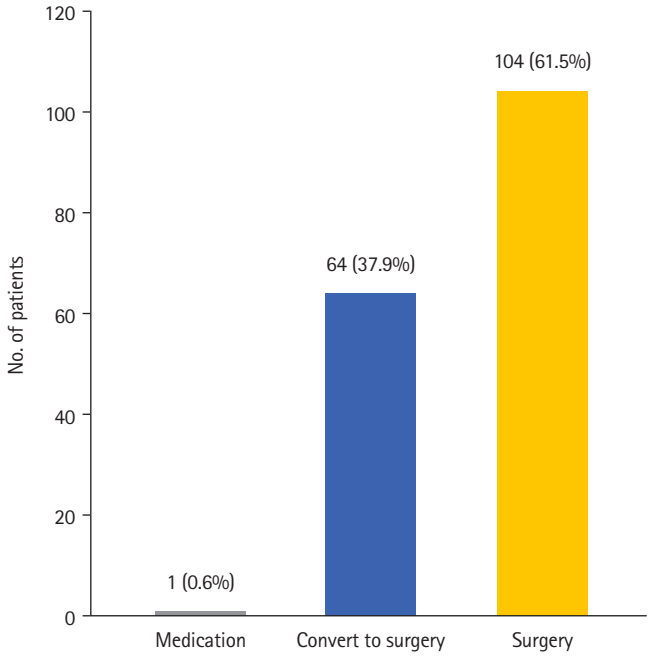

Fig. 9. Treatment options for primary sclerosing encapsulated peritonitis in the 169 symptomatic cases.

TREATMENT

Primary SEP

Surgical intervention

Most patients with primary SEP underwent surgery due to the risk of complications, with the goal of removing the fibrous sheath causing obstruction. In our analysis of 169 symptomatic cases, 104 (61.5%) underwent immediate surgery, while 64 (37.9%) initially attempted conservative management, which was unsuccessful, resulting in 168 patients (99.4%) eventually requiring surgery (Fig. 9).

Primary SEP demonstrated more favorable outcomes than secondary SEP, with a mortality rate of 3.6% compared to 45% to 82% in secondary cases [84]. Among the 168 patients who underwent surgery, 6 (3.6%) died, primarily due to sepsis and multiorgan failure, which were likely associated with preexisting conditions such as end-stage renal disease [4, 6, 7, 9–12, 14, 15, 21–78].

Noteworthy findings emerged regarding surgical techniques. Within the group of 168 operated patients, 7 (4.2%) did not undergo fibrous sheath excision. This was due to 2 primary factors. First, in 3 instances, the fibrous sheath was unexpectedly found during surgeries that were intended to address different medical issues [6, 38, 77]. Second, in 4 cases, severe adhesions made fibrous dissection unsafe, or the patient's overall condition did not allow for a lengthy procedure. Consequently, these operations were limited to histopathological examination or palliative interventions, such as the creation of an ileostomy [7, 45, 54, 55]. However, all patients in this latter group eventually succumbed to their conditions. Notably, 4 of the 6 deaths occurred in the group that did not receive fibrous sheath excision, representing 66.7% of the mortality cases. In contrast, only 3 of the 162 survivors (1.9%) were from this group. Therefore, surgical excision of the fibrous

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sheath should be considered when symptoms warrant it.

Postoperative complications included ileus, early postoperative small bowel obstruction [65], and surgical site infections (21.5%). Additional issues such as pulmonary infections, urinary tract infections, and small bowel obstructions were also reported, though none necessitated reoperation. Other documented morbidities encompassed intestinal fistula [14, 22], iatrogenic bowel injury [6, 31, 61], anastomosis leakage [46], and the creation of ostomies (ileostomy or gastrostomy) [28, 45, 50, 55, 58, 61, 72]. A technique developed by Li et al. [65], termed "intestinal stenting," involves the retrograde insertion of a tube from the appendix stump into the intestine. This method has been shown to significantly reduce both early postoperative small bowel obstruction and small bowel obstruction.

Follow-up

The duration of surveillance remains unclear. In this review, the mean surveillance period was 8 months, during which no recurrences were reported among the patients. However, 1 patient did experience a recurrence of primary SEP, having previously undergone surgery 2 years prior to the subsequent episode [61]. Most patients underwent clinical follow-up, with only 5 opting for gastrointestinal studies or CT scans. Therefore, a longer period of clinical follow-up appears to be prudent.

In conclusion, due to the progressive nature of primary SEP, surgical excision of fibrous tissue remains the primary option to prevent recurrence and associated complications. Further research into medical interventions such as immunotherapy is warranted.

Secondary SEP

Cessation of PD in PD patients

The initial step in managing secondary SEP among PD patients involves discontinuing PD to prevent further damage to the peritoneal membrane [85]. Although this approach seems rational, its effectiveness in reversing peritoneal fibrosis is still debated, possibly due to the absence of peritoneal lavage to remove fibrin, profibrotic factors, and cytokines [86]. An alternative method, as some authors suggest, includes keeping the catheter in place and performing regular peritoneal lavage for patients who have stopped PD; however, there is currently no conclusive evidence to support its positive effect on SEP progression [3, 85]. Withdrawing patients from PD after a SEP diagnosis poses challenges, especially considering the link between the duration of PD and SEP progression. We recommend transitioning from PD to hemodialysis and simultaneously removing the PD catheter [18, 87]. Although this approach seems logical, its efficacy is a matter of debate, as it does not consistently reverse the progression of peritoneal fibrosis and may lead to worsening symptoms [88]. The decision to discontinue PD demands careful evaluation of the benefits and risks, as most patients transition to hemodialysis, which involves its own set of complications, risks, and adjustments to lifestyle.

Nutrition improvement

Effective nutritional support is crucial in managing SEP, especially in advanced cases where there is a risk of malnutrition [89]. Patients who experience weight loss exceeding 10% are at increased risk [65]. Despite this, more than 60% of SEP patients struggle with reduced appetite and gastrointestinal symptoms [2, 85]. Nutritional support, whether provided parenterally or enterally, has been shown to improve postoperative outcomes [85]. The decision to initiate such support is critical, with research highlighting the significance of total parenteral nutrition (TPN) and dietary counseling. However, TPN alone does not have curative effects and increases the risk of infections, thus requiring careful monitoring of nutritional status through markers like albumin [89]. Assessment at the time of SEP diagnosis is vital, and although bowel rest and TPN alone may not be curative, they play a significant role in ensuring adequate nutrition. In situations where enteral feeding is not feasible, TPN is essential, as studies have shown that preoperative TPN administration can reduce complications and shorten hospital stays [5, 85, 87].

Medication for secondary SEP

Currently, there are no established protocols for treating secondary SEP, as most approaches are based on observational case reports [3, 5, 84–89]. Although some outcomes have been favorable, the evidence is limited to observational data.

Early intervention is recommended, focusing on glucocorticoids, tamoxifen, and immunosuppressive agents [3, 5, 84–89]. In cases of complications or delayed diagnosis, surgery is preferred. However, surgery does not stop peritoneal deterioration. Furthermore, symptoms may recur within 6 to 12 months [87] due to new fibrotic capsule formation, and surgery can also lead to new adhesions.

(1) Immunosuppressive drugs

The use of immunosuppressive drugs for treating SEP is not well-supported by evidence and lacks consensus, primarily due to the absence of targeted therapies and large-scale clinical trials. Immunosuppressive agents such as corticosteroids, azathioprine (AZA), mycophenolate mofetil (MMF), and mammalian target of rapamycin (mTOR) inhibitors are frequently used, often in conjunction with corticosteroids [86, 90, 91]. However, these drugs carry a risk of infections, making it essential to confirm that pa-

tients are infection-free before initiating treatment. Additionally, the time required for these drugs to take effect renders them unsuitable for severe cases of SEP.

Corticosteroids are the primary treatment for secondary SEP, supported by numerous case reports with varying formulations, dosages, and durations [86, 90, 91]. They work by suppressing inflammation, preventing fibrin deposition, and inhibiting collagen synthesis, as well as by impeding the glucose-mediated induction of monocyte chemoattractant protein-1 (MCP-1) [18], which is crucial in reducing fibrosis in peritoneal sclerosis. Steroids also help prevent intraperitoneal fluid accumulation and the formation of ascites [85, 90]. Whether used alone or with other drugs, such as immunosuppressants, corticosteroids have shown success [86, 90, 91].

For example, Mori et al. [90] reported successful outcomes using corticosteroids alone, while Kuriyama and Tomonari [92] observed that patients treated with prednisolone experienced better outcomes than those who did not receive this treatment. However, corticosteroids alone may not be adequate for all cases; a 2004 multicenter study by Kawanishi et al. [93] revealed that only 38.5% of patients with secondary SEP could be managed with steroids alone.

The effectiveness of steroid treatment in SEP seems to vary depending on the disease stage, showing better outcomes in the early stages. In contrast, surgical intervention may be preferable in the advanced stages of SEP, where the inflammatory tissue tends to evolve into fibrosis, reducing the efficacy of medical therapy [87, 92]. While there are no established guidelines from controlled trials regarding the optimal dose and duration of steroid therapy, most literature suggests starting with a regimen of prednisolone at 0.5 to 1.0 mg/kg/day or administering a pulse dose of 500 to 1,000 mg of methylprednisolone for 2 to 3 days. This initial phase is typically followed by a continuation of 0.5 to 1 mg/kg/day for 1 month, then tapering the dosage based on clinical symptoms, aiming for a total treatment duration of at least 1 year [90–92].

Nonsteroid immunosuppressive drugs include AZA, MMF, and calcineurin inhibitors (CNIs). AZA, an immunosuppressive antimetabolite, has limited evidence supporting its use in SEP treatment compared to other drugs. Wong et al. [91] reported success using AZA in combination with corticosteroids. In a similar vein, Fagugli et al. [94] administered an initial dose of 100 mg of AZA, which was reduced to 50 mg after 2 months, in conjunction with prednisolone and colchicine. This regimen resulted in reduced symptoms and fibrosis. Additionally, Pepels et al. [95] found that combining AZA with prednisolone could decrease complications such as ascites following laparotomy. Despite these positive outcomes, AZA is not recommended as a standalone treatment but may be useful as an adjunct to steroids.

MMF, another immunosuppressant, exhibits antifibrotic properties by inhibiting specific cellular pathways without inducing TGF-β, a fibrogenic factor [86]. Recent studies involving rat models and patient cases indicate that MMF reduces peritoneal thickness, inflammation, and fibrosis, although the evidence is still limited [86].

CNIs, commonly used after transplantation, may exacerbate SEP by upregulating TGF-β1 and VEGF, which contribute to fibrosis and neoangiogenesis [18]. The mTOR inhibitors, such as sirolimus and everolimus, which regulate cellular metabolism and growth, have demonstrated some effectiveness in reducing peritoneal thickness and fibrosis [96]. Although case reports underscore their effectiveness when used in conjunction with steroids, a case series by Ghadimi et al. [96] reported clinical improvement in only 25% of SEP patients treated with mTOR inhibitors. The mTOR inhibitors are recommended as alternative treatments for SEP in post-transplant patients, particularly when transitioning from CNIs [18, 96].

(2) Tamoxifen

Tamoxifen was first successfully used in secondary SEP patients in 1992 [18], with subsequent reports indicating favorable outcomes when used alone or in combination with corticosteroids or immunosuppressants [18, 97]. A Dutch multicenter study reported significantly lower mortality rates in tamoxifen-treated SEP patients (45.8%) compared to those not treated with tamoxifen (74.4%) after 130 months, along with longer adjusted survival (hazard ratio, 0.39; $P = 0.056$) [97]. However, a 2007 UK study involving 111 SEP cases from PD patients found no survival benefit, likely due to variability in treatment [98]. del Peso et al. [99] suggested that tamoxifen might prevent SEP, as none of the 23 patients who took it developed SEP, while 4 cases occurred in those who did not. A recent study by Liakopoulos et al. [100] reported the safety of long-term tamoxifen use (20 mg/day) for symptom prevention over 10 years, despite persistent SEP calcifications on CT scans.

Dosing and duration of tamoxifen treatment are not well-defined due to the lack of large clinical trials. Most studies administer daily doses ranging from 10 to 40 mg, with a clinical response typically observed within a year. Treatment should continue for at least one year and then be tapered, depending on the control of underlying conditions and the adequacy of the clinical response [18, 85, 97, 98, 100]. Potential side effects, such as hot flushes, nausea, fatigue, endometrial carcinoma, ischemic stroke, pulmonary embolism, and deep venous thrombosis, should be discussed with patients before initiating therapy [100].

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CONCLUSION

SEP presents distinct challenges in both primary and secondary forms, necessitating tailored treatment strategies.

Primary SEP primarily necessitates surgical intervention to relieve intestinal obstruction caused by encasement in a fibrous sheath. The surgical removal of this fibrous sheath is essential, as it is associated with improved outcomes. Medical management options are limited, with only a few cases demonstrating success using drugs such as MMF and prednisolone. Follow-up data are inconclusive, highlighting the necessity for extended surveillance periods to prevent recurrence.

Secondary SEP management primarily aims to halt PD to protect the peritoneal membrane from further damage. Transitioning to hemodialysis and removing the catheter might be an option, though its effectiveness remains a topic of debate. Enhancing nutrition is essential, with nutritional support being critical for successful postoperative outcomes. Medical management typically involves the use of glucocorticoids, tamoxifen, and immunosuppressive agents, reserving surgery for cases of complications or delayed diagnoses. Histopathology is crucial for diagnosis, as it identifies distinctive features such as fibrous tissue proliferation and inflammatory infiltration.

Overall, treatment strategies for both primary and secondary SEP necessitate a multidisciplinary approach that takes into account individual patient characteristics and the severity of the disease. Further research into medical interventions is particularly warranted for secondary SEP, due to the limited evidence currently available.

ARTICLE INFORMATION

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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Author contributions

Conceptualization: PS, SS, ES, KW, RR; Data curation: WV, TJ; Investigation: WV, TJ; Methodology: SC; Supervision: PS, SS, ES, KW, RR; Validation: PS, SS, ES, KW, RR; Writing–original draft: WV, TJ; Writing–review & editing: all authors. All authors read and approved the final manuscript.

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