

# **Encapsulating Peritoneal Sclerosis: The Abdominal Cocoon**

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**Abbreviations:** EPS = encapsulating peritoneal sclerosis, H-E = hematoxylin-eosin

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## **SA-CME LEARNING OBJECTIVES**

After completing this journal-based SA-CME activity, participants will be able to:

- Discuss the nomenclature and etiopathogenesis of encapsulating peritoneal sclerosis (EPS).
- Identify the multimodality imaging appearance and complications of EPS.
- Describe the differential considerations and management of EPS.

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Encapsulating peritoneal sclerosis (EPS) is a rare but serious condition that results in (a) encapsulation of bowel within a thickened fibrocollagenous peritoneal membrane and (b) recurrent episodes of bowel obstruction. Although described by various names in the literature, the preferred term is encapsulating peritoneal sclerosis because it best describes the morphologic and histologic changes in this disorder. The etiology of EPS is multifactorial, with a wide variety of implicated predisposing factors that disrupt the normal physiologic function of the peritoneal membrane—prime among these factors being long-term peritoneal dialysis and bacterial peritoneal infections, especially tuberculosis. The clinical features of EPS are usually nonspecific, and knowledge of the radiologic features is necessary to make a specific diagnosis. The findings on radiographs are usually normal. Images from small-bowel follow-through studies show the bowel loops conglomerated in a concertina-like fashion with a serpentine arrangement in a fixed *U*-shaped configuration. US demonstrates a "cauliflower" appearance of bowel with a narrow base, as well as a "trilaminar" appearance depicted especially with use of high-resolution US probes. CT is the imaging modality of choice and allows identification of the thickened contrast material-enhanced abnormal peritoneal membrane and the encapsulated clumped bowel loops. In addition, CT can potentially help identify the cause of EPS (omental granuloma in tuberculosis), as well as the complications of EPS (bowel obstruction). Conservative medical treatment and surgical therapy early in the course of EPS have been used for management of the condition. The purpose of this article is to review the nomenclature and etiopathogenesis of EPS, describe the multimodality imaging appearances of EPS, including differentiating its features from those of other conditions mimicking EPS, and give an overview of management options.

Online DICOM image stacks are available for this article.

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#### Introduction

Encapsulating peritoneal sclerosis (EPS) is a potentially life-threatening condition that causes fibrocollagenous cocoonlike encapsulation of the bowel. EPS occurs in response to a wide variety of inciting factors and in various other clinical conditions spanning the entire domain of medicine. Although traditionally described as rare, EPS has been a growing concern, especially in relation to peritoneal dialysis. Although little is understood about the pathophysiology of this condition, and even less is understood about how to prevent or cure EPS, advances in imaging have allowed us to routinely make a reliable preoperative diagnosis. In this article, the various aspects that a radiologist must know regarding the etiopathogenesis of EPS are highlighted. Also, the imaging appearances of EPS with various modalities are discussed in detail. Knowledge

## **TEACHING POINTS**

- The term *encapsulating peritoneal sclerosis* is considered to be the most appropriate term to describe fibrocollagenous peritoneal envelopment of the bowel, because this term more accurately correlates with the morphologic changes in this condition; and this term is the preferred term that is used throughout this article.
- EPS is a well-recognized complication of continuous ambulatory peritoneal dialysis. The inciting factor for EPS in this setting is the bioincompatibility of the dialysis fluid.
- The ileal loops appear conglomerated in a concertina-like fashion named because of the resemblance to the bellows of the musical instrument of the same name. A serpentine arrangement of dilated small-bowel loops in a fixed U-shaped configuration is characteristic.
- The "trilaminar" membrane appearance at US has been described as characteristic of EPS. The three laminae in this US trilaminar appearance, from superficial to deep, are formed by (a) a superficial hyperechoic membrane, (b) a middle hypoechoic layer of the bowel wall, and (c) a deep hyperechoic layer of bowel gas and/or bowel contents.
- In EPS, the peritoneum is thickened, with marked continuous enhancement.

of the imaging findings and a high index of suspicion can help in the early diagnosis of EPS and in early intervention for management of EPS, which currently offers the only hope for a better outcome.

# Terminology

Considerable ambiguity exists in the terminology and nomenclature of EPS. The first report of this condition came from Owtschinnikow (1) in 1907, and he named it "peritonitis chronica fibrosa incapsulata." Henceforth, it has been indiscriminately and interchangeably called by various names: peritoneal fibrosis, peritoneal sclerosis, sclerotic thickening of the peritoneal membrane (2), sclerosing peritonitis (3), sclerosing obstructive peritonitis (4), encapsulating peritonitis (5), chronic encapsulating fibrous peritonitis (6), calcific peritonitis (7), or, most commonly, sclerosing encapsulating peritonitis (8). Foo et al (9) used the term abdominal cocoon to describe a primary or idiopathic form of this condition that they noted in adolescent girls from tropical or subtropical countries. However, the condition has been recognized to occur in various geographically and ethnically diverse locations, as well as occurring secondary to a multitude of causes; and the term abdominal cocoon is currently used synonymously with "sclerosing encapsulating peritonitis." Although the most commonly used term in the literature in recent years is sclerosing encapsulating peritonitis, the popularity of the term does not necessarily validate its continued use, because the term is marred by its inaccuracy

in its reference to an inflammatory component, which is frequently absent in this condition. The term encapsulating peritoneal sclerosis is considered to be the most appropriate term to describe fibrocollagenous peritoneal envelopment of the bowel, because this term more accurately correlates with the morphologic changes in this condition (10); and this term is the preferred term that is used throughout this article.

# Causes

Causes of this enigmatic condition can be broadly divided into primary (or idiopathic) causes and secondary causes. Secondary causes are usually considered as either associated with peritoneal dialysis or not associated with peritoneal dialysis.

## **Peritoneal Dialysis**

EPS is a well-recognized complication of continuous ambulatory peritoneal dialysis (Fig 1). The inciting factor for EPS in this setting is the bioincompatibility of the dialysis fluid. The overall prevalence of EPS in patients undergoing continuous ambulatory peritoneal dialysis is approximately 0.7% (11). The propensity to develop EPS is in direct proportion to the duration of treatment, with the prevalence increasing to 19.4% in patients receiving therapy with continuous ambulatory peritoneal dialysis for more than 8 years (11). Although the exact cause and the etiopathogenesis of EPS in the setting of peritoneal dialysis are still unknown, multiple possible contributing factors have been implicated, including the duration of peritoneal dialysis, recurrent episodes of bacterial peritonitis, use of a glucose-based dialysate or hypertonic solution (12), use of an acetate dialysis solution (13), the presence of endotoxin from bacterial filters (14), plasticizers (15), and use of chlorhexidine (16). Most of these possible contributing factors have been recognized and have been discontinued from routine usage during continuous ambulatory peritoneal dialysis. A genetic propensity to develop EPS after exposure to any one of the etiologic factors has also been considered.

#### **Tuberculosis**

Tuberculosis as a cause of EPS is well documented in the literature (17–20). However, it is unclear if tuberculosis triggers peritoneal sclerosis directly or if the cocoon formation is a rare fibroadhesive form of tuberculous peritonitis (Fig 2).

The histopathologic findings in patients with EPS (interstitial fibrin deposition and fibroblast swelling, loss of the mesothelial layer, and the presence of immunohistochemical markers for

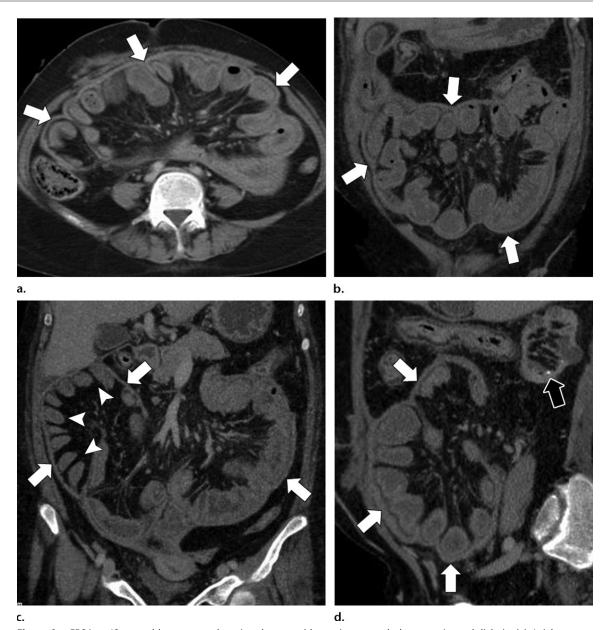


Figure 1. EPS in a 43-year-old woman undergoing therapy with continuous ambulatory peritoneal dialysis. (a) Axial contrast material—enhanced CT image shows a thick soft-tissue mantle of EPS encasing the bowel loops (arrows). (A full DICOM image stack is available online.) (b, c) Coronal contrast-enhanced CT images (b obtained more anteriorly than c) show a thick soft-tissue mantle of EPS encasing the bowel loops (arrows) and the characteristic pattern of arrangement of the small-bowel loops (arrowheads on c). (d) Sagittal contrast-enhanced CT image again shows a thick soft-tissue mantle of EPS encasing the bowel loops (white arrows). Note also the atrophic kidney (black arrow) in this patient with end-stage renal disease.

peritoneal fibroblast activation and proliferation) are expected to be different from those in patients with tuberculous peritonitis (epithelioid giant cell granulomas, caseous necrosis with or without acid-fast bacilli) (21). Both of these histopathologic features have been documented in the literature (17,22). However, a definite difference exists in the clinical manifestations of a tubercular EPS, compared with those of EPS owing to other causes, a difference that has a bearing on the clinical management as well. Clinically, the incidence of acute small-bowel obstruction is higher in patients with tubercular

EPS. Conservative management generally fails, likely owing to the higher incidence of interbowel adhesions and fibrosis in tuberculosis, in addition to the fibrocollagenous cocoon. Surgery entails a higher rate of iatrogenic complications after attempted adhesiolysis of these interbowel adhesions (22). Caseating lymph nodes and serosal tubercles are also seen at surgery, and postoperative administration of antituberculous therapy is warranted. The rate of recurrence of intestinal symptoms after surgery is also higher in patients with tubercular EPS than in those with EPS that is due to other causes.

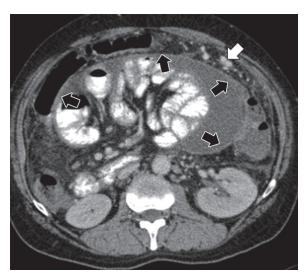


Figure 2. EPS secondary to intra-abdominal tuberculosis in a 54-year-old man. Axial contrast-enhanced CT image shows the characteristic abdominal cocoon (black arrows) with the encased bowel loops and fluid. Note the omental thickening with omental nodules (white arrow). The findings at histopathologic examination helped confirm caseating tubercular granulomas.

EPS has been reported in association with or as a consequence of a multitude of other conditions (Table) (23–52). Despite the myriad causes, the inciting event of EPS is not known in a number of patients.

## **Idiopathic or Primary Causes**

The initial observation that most cases of EPS occur in adolescent girls led to the hypothesis that EPS was secondary to subclinical peritonitis that was caused by a viral infection superimposed on retrograde menstruation (9). Poor perineal hygiene leading to retrograde subclinical bacterial infection was also considered. However, the findings in later reports of idiopathic EPS in young premenarchal female patients (53), as well as in male patients (54,55), along with the fact that viral or bacterial markers could not be consistently demonstrated, have reduced the credibility of this assumption (Fig 3). Ethnic or geographic predisposition could have also played a role, because many case reports were from the tropics or Asia. Dietary factors such as improperly cooked fish have also been implicated (56).

# **Pathogenesis**

Two distinct and opposing hypotheses exist with regard to the pathogenesis of EPS. According to the European school of thought, "simple sclerosis" and EPS are two different conditions and distinct entities (57,58). Simple sclerosis occurs in virtually all patients who undergo peritoneal dialysis, causing moderate fibrosis without inflammation; and simple sclerosis can be reliably replicated in animal models of peri-

#### Causes of and Associations with EPS

#### Causes

Primary (idiopathic)

Secondary

Related to peritoneal dialysis

Unrelated to peritoneal dialysis

#### Associations

Surgery or surgical shunts

Laparotomy for carcinoma or benign disorders (23)

Abdominal lavage with povidone iodine (24)

Ventriculoperitoneal shunt (25)

LeVeen peritoneovenous shunt (26)

Peritonitis

Bacterial peritonitis (27), including tubercular peritonitis

Meconium peritonitis (28)

Malignancies

Intra-abdominal-gastric (29), pancreatic (30), renal (31), and midgut neuroendocrine (32)

Pelvic (ovarian) (33,34)

Other: lymphoma (35,36)

Diseases of the female reproductive tract

Luteinized thecoma of the ovary (37)

Endometriosis (38)

Adenomyosis, leiomyoma (39)

Teratoma (40)

Administration of  $\beta$ -blockers (41,42)

Cirrhosis (43)

Bowel perforation (44)

Autoimmune disease (systemic lupus

erythematosus) (45)

Peritoneal sarcoidosis (46)

Familial Mediterranean fever (47)

After organ transplantation

Liver (48)

Kidney(49)

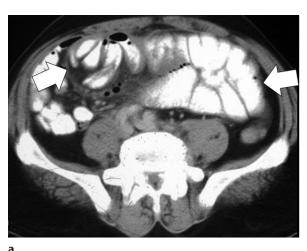
Intestine (50)

Intraperitoneal drug administration (51)

Radiation enteritis (52)

Note.—Numbers within parentheses are reference citations.

toneal dialysis. EPS, on the other hand, is rare and manifests with marked fibrosis with some component of inflammation. In addition to the association with peritoneal dialysis, the development of EPS needs a "second hit" by triggering factors (chemicals, toxins, infectious agents, etc). An element of probable genetic susceptibility also exists, because only small numbers of the exposed population actually go on to develop EPS.





**Figure 3.** Idiopathic EPS in a 43-year-old man. Axial contrast-enhanced CT images (**a** obtained lower than **b**) show the characteristic clumped appearance of bowel loops within a thin cocoon (arrows). No obvious cause was found at surgery or at histopathologic examination. No risk factors or associations could be identified clinically. EPS was hence deemed to be idiopathic.

Simple sclerosis is virtually asymptomatic, but a 50% mortality rate for EPS has been reported in a few series (59).

On the other hand, the Japanese school of thought considers simple sclerosis and EPS to be the two extremes of the same disease that is caused by peritoneal irritation (60).

Whatever the inciting agent, the end result is transformation of fibrocytes by the transforming growth factor  $\beta$  (TGF- $\beta$ ). This transformation triggers collagen production that leads to formation of the encapsulating sheet of fibrotic membrane that hampers intestinal motility (61).

## **Clinical Manifestations**

Yip and Lee (62) described four cardinal features of the abdominal cocoon: (a) occurrence in young women, (b) subacute obstruction with no clear cause, (c) previous similar episodes that had resolved spontaneously, and (d) the presence of a palpable abdominal mass with pain. However, we now know that primary and secondary forms of EPS exist, and EPS has been reported in a wide range of ages, with the youngest patient being a 2-day-old neonate (28) and the oldest patient being an 82-year-old man (63).

The presenting signs and symptoms of EPS are usually vague and nonlocalizing. The patient usually presents with vomiting, abdominal pain or heaviness, and other features of subacute intestinal obstruction. Usually, patients have had prior episodes with similar symptoms that have resolved spontaneously. Malnutrition may occur owing to recurrent episodes. Clinically, the abdomen is soft at palpation. A soft nontender mass may be palpable in the central part of the abdomen, which actually represents the clumped-up bowel loops.

# Findings at Imaging

# **Radiographic Findings**

Abdominal radiographs are usually normal in patients with EPS, with a normal bowel gas pattern. Calcification, which is a frequent finding in the setting of peritoneal dialysis, may be depicted on radiographs (Fig 4). Because the obstruction is usually subacute, dramatic dilatation of bowel loops or air-fluid levels are not usually depicted. Colonic and rectal gas is usually seen. However, tubercular EPS has a propensity to cause acute obstruction. In such cases, dilated bowel loops, air-fluid levels, and an absence of colonic and rectal gas may be seen. Air under the diaphragm from perforation is rare.

## **Barium Study Findings**

Barium studies were commonly used in the past for evaluation of recurrent episodes of spontaneously resolving subacute intestinal obstruction, which is common in this condition. Mild dilatation of the ileal loops may be seen. However, usually, no well-defined transition point is noted. The ileal loops appear conglomerated in a concertinalike fashion named because of the resemblance to the bellows of the musical instrument of the same name (Fig 5). A serpentine arrangement of dilated small-bowel loops in a fixed *U*-shaped configuration is characteristic (64). This cluster of loops appears rigid and fixed. They cannot be separated by applying pressure on the abdominal wall with a spatula. Kinking and tethering of bowel loops may be noted in cases with excessive interbowel adhesions. Usually, some motility disturbance exists, which is appreciated at real-time fluoroscopy. Varying degrees of obstruction and hypermobility of a few loops are seen. Overall

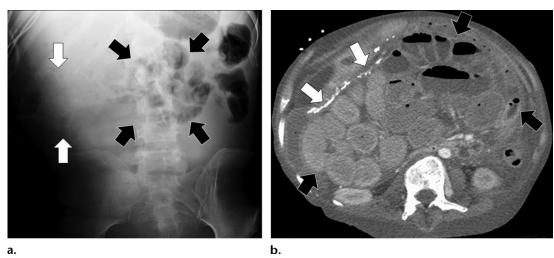


Figure 4. EPS in a 26-year-old man after lung transplantation (same patient as shown in Fig 10). (a) Anteroposterior radiograph shows calcifications in the right flank (white arrows) and a nonspecific pattern of small-bowel loops (black arrows) clustering in the mid portion of the abdomen. (b) Axial contrast-enhanced CT image helps confirm the thickened enhancing peritoneal membrane (black arrows), a finding that is in keeping with EPS. The membrane is calcified (white arrows) on its anterior aspect. (A full DICOM image stack is available online.)

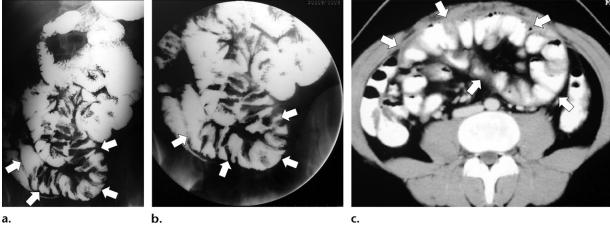


Figure 5. EPS in a 22-year-old man. (a, b) Anteroposterior small-bowel followthrough images obtained at earlier (a) and later (b) barium studies show the characteristic rigid cluster of bowel loops arranged in a concertina-like fashion (arrows). (c) Axial contrast-enhanced CT image shows the thickened peritoneal membrane (arrows) of EPS. (d) Drawing shows a model of a concertina, with the arrangement of the instrument's bellows resembling the arrangement of the bowel loops in EPS.

intestinal transit is delayed. However, the colon is eventually opacified, because the obstruction is usually not complete.

## **US Findings**

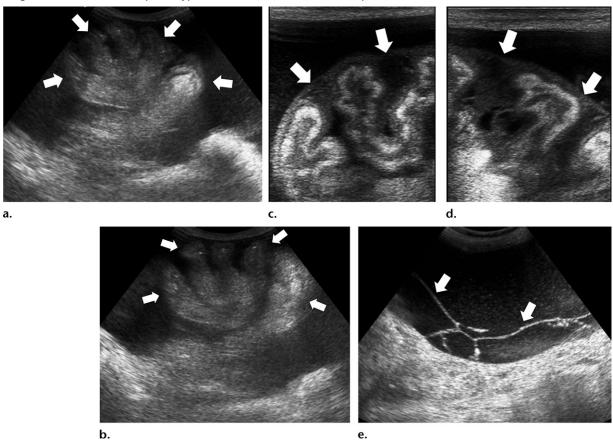
d.

US shows clumping of small-bowel loops in the center of the abdomen. The bowel loops may show thickening or may be hyperechoic. However, the vascularity at color Doppler flow imaging is usually preserved. A characteristic arrangement of loops in a concertina-like fashion with a narrow

base posteriorly is highly suggestive of EPS. Thus, the overall appearance resembles that of a cauliflower (65). This appearance is best demonstrated when there is moderate coexistent ascites. This cauliflower arrangement cannot be disturbed by applying probe pressure or trying to disperse the bowel loops, because the loops are fixed (Fig 6).

The "trilaminar" membrane appearance at US has been described as characteristic of EPS (66,67). The three laminae in this US trilaminar appearance (Fig 7), from superficial to deep, are formed by (a) a superficial hyperechoic membrane, (b) a middle hypoechoic layer of the bowel wall, and (c) a deep hyperechoic layer of bowel

**Figure 6.** EPS in a 39-year-old man. **(a)** US image obtained with a curvilinear transducer shows the characteristic central clumping of small-bowel loops with a narrow base resembling a cauliflower (arrows). **(b)** US image obtained after applying probe pressure shows that this arrangement could not be altered by applying probe pressure (arrows). **(c, d)** High-resolution US images obtained to the right **(c)** and left **(d)** of the cocoon with a linear transducer show the encasing thickened peritoneal membrane (arrows). **(e)** US image shows that additional septa or hyperechoic strands (arrows) are also depicted in the ascitic fluid.



gas and/or bowel contents. This US trilaminar appearance in fact merely represents peritoneal thickening or formation of a membrane over the bowel surface. This US trilaminar sign should not be confused with the CT trilaminar sign, in which a hypoattenuating layer of submucosal edema is sandwiched between enhancing mucosal and muscular-serosal layers on either side. This CT trilaminar sign has been described in various diseases such as Crohn disease, intestinal ischemia, radiation enteritis, and graft versus host disease.

The membrane encapsulating the bowel loops may be directly depicted, especially with a high-resolution high-frequency linear US probe, as a hypoechoic structure stretched over the anterior surface of the bowel loops. Adhesions to the anterior or posterior abdominal wall may be seen. Intraperitoneal hyperechoic strands are often depicted. Disturbances in peristalsis are common. Peristalsis may be reduced in a few of the bowel loops, with hyperperistalsis in the others. Loculated fluid collections can be demonstrated, and the degree of internal septation can be ascertained. Guided aspiration can be helpful in the analysis of peritoneal fluid. US is better than CT for the

evaluation and management of loculated collections. Although the membrane itself is usually not amenable to biopsy, US can aid in the biopsy of associated omental nodules in tubercular EPS or in mimics such as peritoneal carcinomatosis.

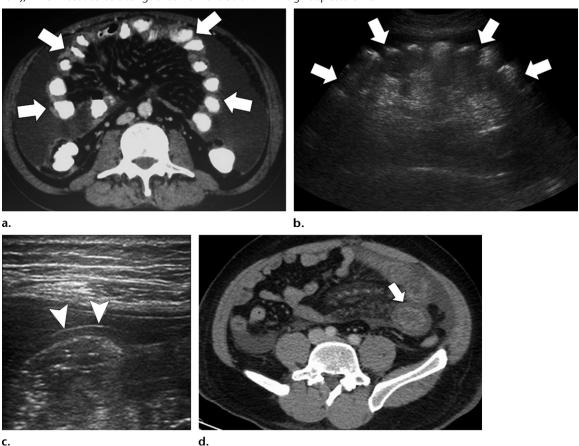
One shortcoming of using US for the diagnosis of EPS is that all of the findings are best demonstrated only in the presence of mild to moderate ascites. Although the incidence of ascites in patients with EPS is not known, some amount of peritoneal fluid is usually seen in settings such as peritoneal dialysis or tuberculosis.

## CT Findings

Currently, CT has become the mainstay for diagnosis of EPS. Although nonenhanced CT may depict calcification slightly better, routine portal venous phase CT after intravenous administration of contrast material is usually sufficient to give adequate information. Administration of positive oral contrast material may help in better bowel delineation.

With thin isotropic voxels and multiplanar reformation, it has now become possible to routinely depict the encapsulating membrane. The normal peritoneum is a thin, smooth, barely perceptible

Figure 7. US trilaminar sign in a patient with EPS, compared with the CT trilaminar sign in a different patient. (a-c) EPS in a 43-year-old man. (a) Axial CT image shows the characteristic clumped appearance of the bowel loops (arrows). However, the thickened peritoneal membrane is not well depicted. (b) US image obtained with a 5-MHz curvilinear transducer shows the characteristic cauliflower arrangement of the bowel loops (arrows). (c) US image obtained with a high-frequency linear transducer perpendicular to the membrane, with exclusion of anisotropy, shows the characteristic US trilaminar appearance (arrowheads) of EPS. (d) CT trilaminar sign: Axial CT image of a different patient with enteritis shows the CT trilaminar sign (arrow), which needs to be distinguished from the US trilaminar sign depicted on c.



structure with discontinuous enhancement. In EPS, the peritoneum is thickened, with marked continuous enhancement. Assessment of peritoneal thickening is usually subjective, and no optimum threshold has been described, although in our experience, a thickness greater than 2 mm appears to be a reasonable cutoff. The small-bowel loops appear congregated toward the center of the abdominal cavity and are encased by a mantle that demonstrates soft-tissue attenuation. In our personal experience, we find volume-rendered CT images depicting the intraintestinal positive oral contrast material to be useful in identifying the altered bowel configuration (Fig 8).

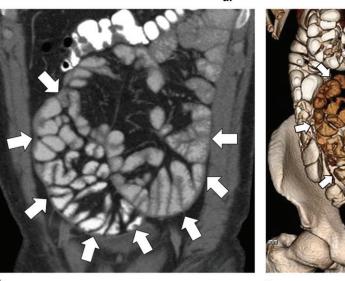
In addition to the altered position and placement of the bowel in the abdomen, CT also helps depict changes in the contour and caliber of the bowel. Bowel thickening may or may not be demonstrated. A few of the bowel loops may appear dilated or prominent. Usually, no obvious transition point is seen. Angulation, kinking, and tethering of the bowel loops indicate extensive interbowel adhesions, which portend a poor prognosis. This

information about interbowel adhesions is also important to the surgical team, as well as the anesthesia team, because the surgery is expected to be difficult and prolonged. Such interbowel adhesions are usually common in EPS with a tuberculous cause, in which case additional findings such as omental caking, mesenteric and retroperitoneal necrotic lymph nodes, and serosal tubercles may be identified at CT (Fig 2).

Calcification, if any, either on the membrane or the lymph nodes, can be reliably depicted with CT. The calcification usually manifests on the visceral surface of the bowel, although calcification of the parietal peritoneum has also been described. The calcification can be focal, diffuse with a fine linear pattern, or extensive and conglomerate (68). Ascites, especially interbowel ascites, is common in EPS, and loculated fluid collections can be demonstrated and quantified.

CT can be used to identify complications of EPS such as bowel gangrene (lack of normal bowel enhancement) and bowel perforation (pneumoperitoneum, oral contrast material







**Figure 8.** EPS in a 32-year-old man. **(a, b)** Axial **(a)** and coronal **(b)** contrast-enhanced CT images obtained after administration of oral contrast material show the EPS encasing the small-bowel loops (arrows). **(c)** Volume-rendered three-dimensional CT image in the anteroposterior orientation obtained after administration of oral contrast material allows better appreciation of the clumped nature of the small-bowel loops (arrows).

extravasation). These complications are relatively rare because the thickened peritoneal membrane encapsulates and compresses the bowel from the exterior, with preservation of the central vascular pedicle (as opposed to mimics such as internal hernia, in which the pedicle is twisted, leading to early ischemia). CT can reliably depict these complications when they occur and can help determine management. Owing to its excellent depiction of the anatomic abnormality and the complications, contrast-enhanced CT is regarded as the modality of choice in the imaging of EPS.

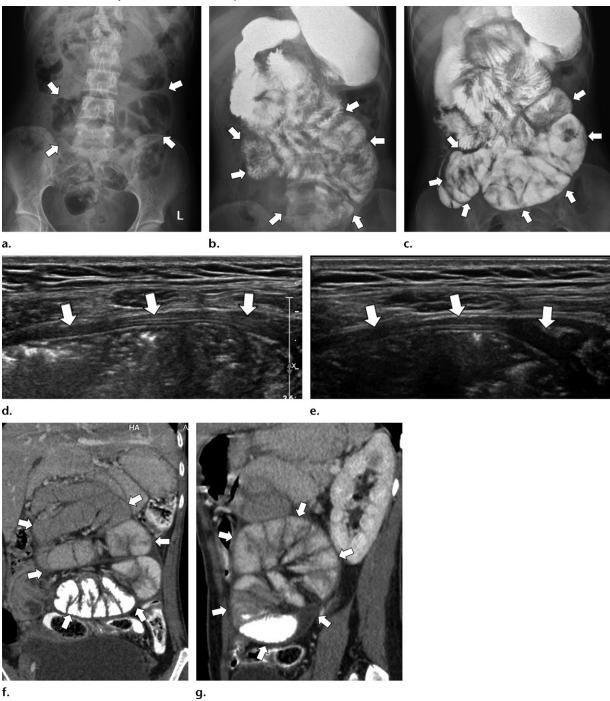
Although scoring systems that are based on CT findings have been proposed (69,70), they are more useful as academic tools to assess interobserver variation and cannot be used in prognostication, because the CT scores do not correlate with the outcome. Parameters used for scoring include peritoneal thickening, bowel wall thickening, calcification, and tethering.

Benefits from CT usually outweigh potential concerns of contrast agent–induced nephropathy (because the patient is already receiving therapy with dialysis) and concerns about radiation exposure. Often, a combination of modalities is used to raise suspicion regarding EPS and to arrive at the diagnosis before surgery (Fig 9).

## **MRI Findings**

MRI shows features similar to those depicted at CT, with demonstration of the thickened membrane and bowel abnormalities (71,72) (Fig 10). Normal peritoneal enhancement is less than or equal to that of the liver. Abnormal enhancement greater than that of the liver is better depicted with the high contrast conspicuity of MRI but is not easily appreciable at CT (73). Cine sequences at MRI that demonstrate peristalsis can potentially help to better differentiate the bowel wall from the peritoneal membrane. MRI has, however, been

Figure 9. Features of subacute bowel obstruction in a 36-year-old woman with intermittent abdominal pain (same patient as shown in Fig 11). (a) Anteroposterior radiograph shows nonspecific clumping of small-bowel loops (arrows) without bowel dilatation. (b, c) Small-bowel follow-through images obtained earlier (b) and later (c) from a barium study show a clustered serpentine arrangement of bowel loops in a fixed *U*-shaped configuration (arrows). (d, e) Transverse (d) and longitudinal (e) US images obtained with a high-frequency transducer raise suspicion for a possible thickened peritoneal membrane (arrows). However, owing to the absence of ascites, this possibility is suboptimally assessed. (f, g) Coronal (f) and sagittal (g) CT images show the thickened membrane of EPS, with encasement of mildly dilated small-bowel loops (arrows).



infrequently used because of its low availability and the cost constraints, along with a reluctance to use MRI for bowel imaging owing to respiratory artifacts. The lack of radiation exposure, along with the refinement of enterographic techniques, makes MRI an attractive option. Caution may be

warranted in the use of contrast-enhanced MRI in the setting of renal insufficiency.

## **PET Findings**

Although the uptake of fluorine 18 fluorodeoxyglucose (FDG) at PET may demonstrate EPS in

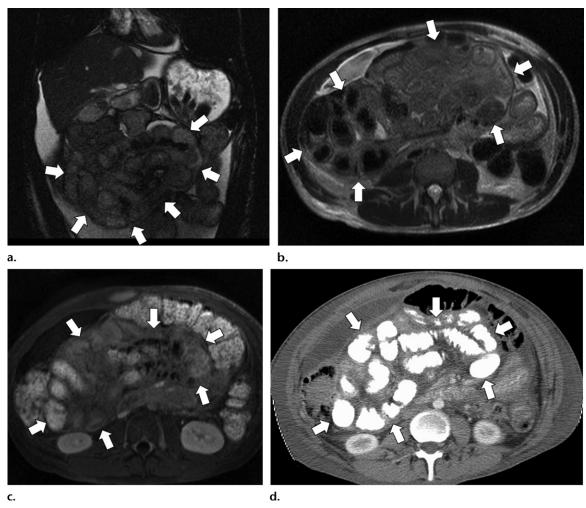


Figure 10. EPS in a 26-year-old man with vague abdominal symptoms who was receiving therapy with long-term dialysis (same patient as shown in Fig 4). (a–c) Coronal balanced steady-state precession MR image (a), axial single-shot fast spin-echo MR image (b), and axial gadolinium-enhanced T1-weighted gradient-echo MR image (c) show clustered small-bowel loops and the encasing thickened peritoneal membrane (arrows). (d) Axial contrast-enhanced CT image obtained after administration of oral contrast material helps confirm these findings of EPS (arrows).

the inflammatory phase or help diagnose acute peritonitis (74), PET does not have an appreciable advantage in the diagnosis of EPS, compared with the other modalities, especially because active inflammation is frequently absent in this condition.

## **Intraoperative Findings**

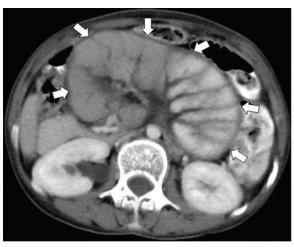
At surgery, the usually thin translucent-appearing peritoneum appears whitish, opaque, and thickened. The peritoneum may have a "tanned" or "leathery" appearance (Fig 11). This abnormal peritoneum encases or encapsulates the small-bowel loops. The findings at histopathologic examination reveal fibrous tissue with or without mononuclear inflammatory cells. Patients with tubercular EPS can potentially have epithelioid giant cell granulomas, which can give a clue to the cause of the EPS (Fig 12).

# **Differential Diagnosis**

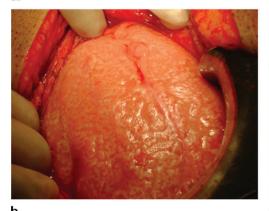
Congenital peritoneal encapsulation is a benign condition characterized by a thin accessory peritoneal membrane surrounding the small bowel (75). This condition is, however, asymptomatic and is usually diagnosed during unrelated surgery or radiologic examinations.

Peritoneal carcinomatosis (Fig 13a) can demonstrate thickening and abnormal enhancement of the peritoneum, findings mimicking EPS. However, it is generally easy to differentiate the two conditions, because the thickening in peritoneal carcinomatosis is nodular (compared with a smooth thickening in EPS), with associated nodules in the omentum, the pouch of Douglas, and the serosal surfaces, with or without lymphadenopathy. Evidence of a primary malignancy (ovarian, gastric, etc) may also be depicted.

Internal hernias (Fig 13b) may demonstrate abnormal clustering of bowel loops that may mimic the centrally displaced bowel loop pattern of EPS. However, it is usually possible to differentiate the two conditions at CT because of the absence of the soft-tissue mantle in internal



a.



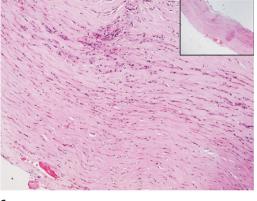


Figure 11. EPS in a 36-year-old woman (same patient as shown in Fig 9). (a) Axial contrast-enhanced CT im-

age obtained after administration of intravenous and oral contrast materials shows the classic features of EPS (arrows). (b) Intraoperative photograph shows a characteristic thickened peritoneum with a tanned, leathery appearance. (c) High-power photomicrograph shows the fibrocollagenous membrane with fibrin deposition and a mild inflammatory infiltrate. (Hematoxylin-eosin [H-E] stain; original magnification, ×100.) Inset: Low-power photomicrograph shows the same findings. (H-E stain; original magnification, ×40.)

hernias as well as the relatively fixed anatomic regions in which they occur. Complications such as ischemia are more common with internal hernias owing to the compromise of the vascular pedicle, as discussed previously.

# Management

### Surgery

Traditionally, most patients with EPS were treated surgically, because the diagnosis was usually made only intraoperatively when the patient underwent surgery for recurrent unexplained episodes of obstruction (Fig 14). In addition to dissection of the abdominal cocoon, extensive interbowel adhesiolysis may be needed for symptom relief. Bowel resection is resorted to only in the case of nonviability of the bowel. However, owing to the increased propensity for complications such as fistulas, abscesses, sepsis, and recurrence, limited surgeries have been tried, such as multiple releasing incisions in the membrane without attempting

complete dissection. The best results have been achieved when surgery was performed early in the course of EPS (76), likely owing to the ease of membrane resection, fewer interbowel adhesions, and fewer iatrogenic complications.

### Conservative Management

A shift in management protocols has occurred recently, with attempts at conservative management, because routine preoperative diagnosis is now possible. Patients without overt signs of obstruction or ischemia can be given a trial of conservative management. Complete reversal of the membrane formation may even occur with conservative therapy (77). Cessation of peritoneal dialysis and a shift to hemodialysis and total parenteral nutrition are usually the mainstay of conservative therapy in patients with peritoneal dialysis-associated EPS. Renin-angiotensin-aldosterone system inhibitor therapy and therapy with immunosuppressants (corticosteroids, cyclophosphamide, azathioprine, and mycophenolate mofetil) have been found to

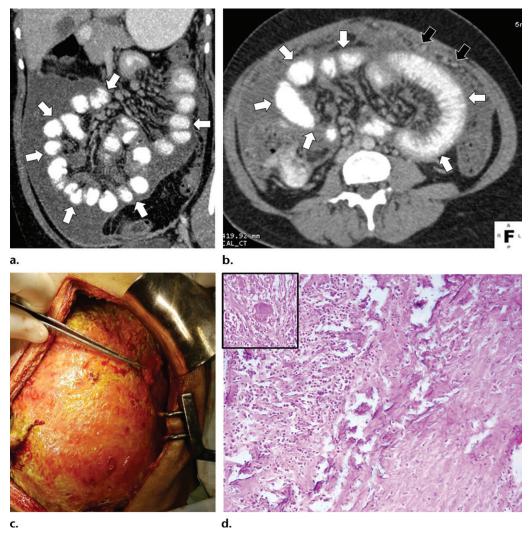


Figure 12. Tubercular EPS in a 23-year-old woman with known tuberculosis. (a, b) Coronal (a) and axial (b) contrast-enhanced CT images show the characteristic features of EPS: the cocoon with fluid and the serpentine appearance of the bowel loops (white arrows), with omental thickening and nodularity (black arrows on b). (c) Intraoperative photograph shows a characteristic thickened peritoneum with fibrinous exudates on the surface. (d) Low-power photomicrograph shows fibrocollagenous tissue with a mixed inflammatory infiltrate including eosinophils, lymphocytes, and plasma cells. (H-E stain; original magnification,  $\times 100$ .) Inset: Highpower photomicrograph shows a well-formed epithelioid cell granuloma with giant cells. (H-E stain; original magnification,  $\times 400$ .)

be efficacious, especially in cases without inflammation (78). Recently, tamoxifen therapy has also been used with some success (79).

# **Prognosis**

Despite advances in diagnosis and treatment, EPS, when symptomatic, has a high mortality rate that has been reported to vary between 4% and 82% (80,81). On average, the mortality rate is about 35%; and in more than 60% of patients with more severe disease, death occurs within 4 months of diagnosis, either owing to bowel obstruction or iatrogenic complications (82).

## Conclusion

EPS is a benign enigmatic condition with a variety of proposed causes. This condition is still consid-

ered life threatening, because it has a high fatality rate. Best outcomes have been reported for patients in whom early surgery has been performed. Thus, early diagnosis is of paramount importance. A high index of suspicion and a familiarity with the multimodality radiologic findings enable early diagnosis, which could markedly affect patient outcome.

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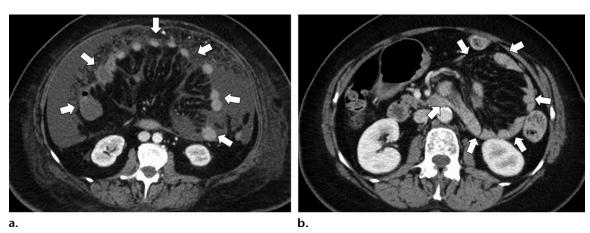


Figure 13. Mimics of EPS in two different patients. (a) Axial CT image of a 61-year-old woman with known peritoneal carcinomatosis secondary to an ovarian malignancy shows a characteristic centrally clustered arrangement of small-bowel loops in a cauliflower-like pattern (arrows). Confluent omental caking is demonstrated, a finding in keeping with carcinomatosis, but no thickened enhancing peritoneal membrane is depicted encasing the small-bowel loops, which would be seen in EPS. (b) Axial CT image of a 63-year-old woman with a nonobstructed left paraduodenal hernia (internal hernia) shows a characteristic clustered arrangement of small-bowel loops (arrows) in a paramidline location posterior to the inferior mesenteric vein. No thickened enhancing peritoneal membrane encases the small-bowel loops, which would be seen in EPS. The findings at surgery helped confirm the internal hernia.

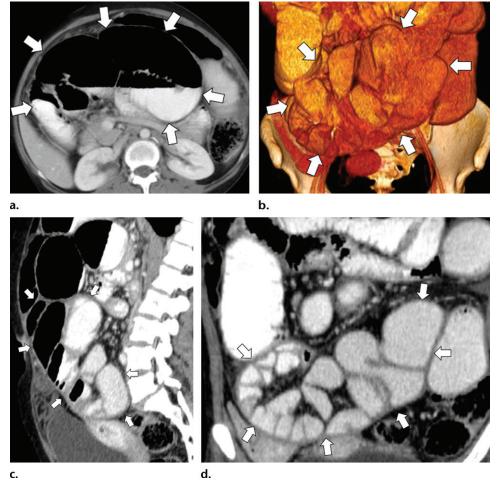


Figure 14. EPS in a 35-year-old woman with bowel obstruction. (a) Axial CT image shows dilated bowel loops with fluid levels in keeping with obstruction (arrows). (b) Volume-rendered three-dimensional CT image in the anteroposterior orientation better shows the clumped pattern of the bowel loops (arrows). (c) Sagittal reformatted CT image better shows the thickened peritoneal membrane enveloping the bowel loops, resulting in small-bowel obstruction with dilated bowel loops (arrows) and fluid levels. (d) Coronal reformatted CT image also shows the thickened peritoneal membrane enveloping the bowel loops, resulting in small-bowel obstruction with dilated bowel loops (arrows) and fluid levels.

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