Encapsulating peritoneal sclerosis (EPS) is an uncommon but one of the most serious complications in patients on long-term peritoneal dialysis (PD). The diffuse thickening and sclerosis of the peritoneal membrane that characterizes EPS leads to decreased ultrafiltration and ultimately to bowel obstruction. Given that the prognosis of established EPS is poor, early recognition of the preceding symptoms is essential. Computed tomography of the abdomen is a reliable and noninvasive diagnostic tool. Typical computed tomography features of EPS include peritoneal calcification, bowel wall thickening, peritoneal thickening, loculated fluid collections, and tethered bowel loops. These findings are diagnostic of EPS in the appropriate clinical setting. Here we present a case report of chronic abdominal pain in a patient on maintenance PD representing a case of EPS.

Key words
Encapsulating peritoneal sclerosis, EPS, end-stage renal disease

Case report
A 57-year-old white woman with history of end-stage renal disease caused by vesicoureteral reflux treated with peritoneal dialysis (PD) had experienced 4 episodes of peritonitis over 12 years, with a progressive increase in her dialysate-to-plasma ratio of creatinine. For more than 6 months, she had symptoms of intermittent nausea, vomiting, and diarrhea associated with intermittent diffuse abdominal pain. All infectious causes, including infectious peritonitis, were excluded on several occasions.

The patient was treated symptomatically with antimotility agents, gaining minimal relief, and she was referred to our center. On examination, she had mild diffuse abdominal tenderness with no guarding or rebound. The PD catheter exit site showed no discharge. Contrast computed tomography (CT) imaging of abdomen and pelvis showed diffuse wall thickening involving the entire colon (Figure 1). The patient was transitioned to hemodialysis and started on a high-dose steroid. Subsequent CT imaging of abdomen and pelvis (Figure 2) showed increased peritoneal thickening, with calcifications and tethering of the intestine consistent with a diagnosis of encapsulating peritoneal sclerosis (EPS). Surgical stripping of the encapsulating membrane and enterolysis were not attempted because of high operative mortality. The patient was maintained on total parental nutrition with adequate nutritional support, but rapidly deteriorated and died of septic shock.

Discussion
Encapsulating peritoneal sclerosis is a rare, but life-threatening complication of PD. A major limiting step...
to successful EPS therapy is delay in recognizing the diagnosis. The condition is thought to result from chronic intra-abdominal inflammation that is multifactorial in origin.

Prolonged PD, such as that in our patient (who had been on PD for more than 12 years), represents the most consistent risk factor identified in EPS, which is associated with high morbidity and mortality related to bowel obstruction and malnutrition (1). Publications have reported variable incidences of EPS, ranging from 8% after 4–5 years of PD in the Scottish Renal Registry, to 2.1% in Australia and 5.9% in Japan. A U.K. series identified 27 EPS cases, for an incidence of 3.3% over 7 years (2). An Australian study identified 54 cases over 14 years, giving a rate of 0.7% (3). Japanese and Korean studies describe rates of 0.8%–2.5% (4). Kawanishi et al. published a 2-year follow-up of a PD cohort in 2001, suggesting an incidence of 0.8% (5). Those authors published a further analysis in 2004, reporting an incidence of 2.5%, which may reflect the longer follow-up inherent to the study design rather than a true increase in incidence (6).

Encapsulating peritoneal sclerosis represents the most advanced stage of peritoneal deterioration, with bowel obstruction and malnutrition; the condition is life-threatening and often irreversible. The risk of death is significant. There are no specific predictors for the development of EPS. Patients at high risk for EPS are those who have been on PD more than 5 years, who have high peritoneal transport, and who have experienced frequent peritonitis episodes (1,5,6).

Features of bowel obstruction and encapsulation such as nausea and vomiting can help in the diagnosis. Patients can also present with hemoperitoneum and recurrent sterile peritonitis.

The diagnostic imaging of choice is CT imaging. Laparotomy in these patients carries a high risk of morbidity and mortality (1).

Peritoneal irritation leading to intra-abdominal inflammation is believed to play a role in EPS development. A peritoneal equilibration test is not discriminatory, although it can be used to monitor membrane changes.

There are no randomized controlled trial results to guide EPS treatment. Several case series have suggested a possible role for immunosuppression (prednisolone, azathioprine, mycophenolate mofetil) as a mode of treatment. In the early stages of EPS, treatment with steroids has been shown to be partially effective. There are anecdotal reports of the beneficial use of tamoxifen, a nonsteroidal anti-estrogenic agent (7,8).

Surgery (enterolysis) as a last resort is associated with variable outcomes and high mortality. The rarity of EPS makes it difficult for surgeons to develop sufficient expertise except in specialized centers, where improved survival with the use of steroids and surgery has been reported.

**Summary**

Because the prognosis of established EPS is poor, early recognition of the preceding symptoms is essential. Features suggestive of early peritoneal sclerosis include the development of a high transport state of the peritoneal membrane, loss of ultrafiltration capacity, and a decrease in mesothelial cell mass as reflected by a low level of cancer antigen 125 in peritoneal effluent.

Patients at high risk for EPS include those on PD for more than 5 years, with high peritoneal transport, and frequent episodes of peritonitis—such as our patient, who had experienced 4 episodes of peritonitis.
and had been on PD for more than 12 years. However, those symptoms are not specific, which makes early recognition difficult. Patient in whom the index of suspicion is high should undergo CT imaging of the abdomen, a reliable and noninvasive diagnostic tool. The CT features typical of EPS include peritoneal calcification, bowel wall thickening, peritoneal thickening, loculated fluid collections, and tethered bowel loops. Such findings are diagnostic of EPS in the appropriate clinical setting.

**Disclosures**

The authors have no conflicts of interest to declare.

**References**


**Corresponding author:**
Surafel Gebreselassie, MD, Department of Nephrology and Hypertension, Cleveland Clinic Main Campus, Mail Code Q7, 9500 Euclid Avenue, Cleveland, Ohio 44195 U.S.A.

**E-mail:**
Gebress@ccf.org