

## **Definition of Peritoneal Dialysis related Encapsulating Peritoneal Sclerosis**

used by the Scottish Renal Registry

Encapsulating Peritoneal Sclerosis, previously known as sclerosing encapsulating peritonitis, is a relatively uncommon but very serious complication of peritoneal dialysis that was first described in 1980<sup>1</sup>. EPS is characterized as a progressive intra-abdominal inflammatory process that results in sheets of fibrous tissue that encapsulate, bind and constrict the viscera, compromise the motility and function of the bowel and may induce ascites. The diagnosis is based on a combination of clinical and radiological features of subacute obstruction and/or ascites in patients who are or have been on PD. The definition of a diagnosis of EPS was debated and reported by an ad hoc committee on ultrafiltration management of the International Society of Peritoneal Dialysis in 2000<sup>2</sup>.

Definition used for the diagnosis of EPS.

The literature refers to a number of terminologies, often used interchangeably, for peritoneal fibrosing syndromes such as peritoneal sclerosis or fibrosis, sclerotic thickening of the peritoneum, calcific peritonitis, abdominal cocoon or sclerotic obstructive peritonitis as well as sclerosing peritonitis<sup>3-5</sup>. The more recent EPS terminology is more appropriate than sclerosing encapsulating peritonitis since an inflammatory component is often not present in the fully developed syndrome.

There are three components to the diagnosis of EPS:

a. Clinical diagnosis.

1. Non-specific symptoms such as anorexia, nausea, vomiting, abdominal fullness and weight loss and non-specific signs such as anaemia, fever or raised CRP often develop insidiously before the diagnosis is first suspected.

2. The diagnosis is most commonly considered when the patient develops symptoms and signs of obstructive ileus due to loss of intestinal motility and encapsulation of the bowel.

3. Less commonly EPS may present clinically as ascites, recurring or non-resolving peritonitis, bloody dialysate effluent or poor ultrafiltration capacity.

4. Subacute obstruction and ascites are the major clinical features of the patients who develop EPS after PD has already been discontinued.

b. Radiological diagnosis.

1. Ultrasound - intraperitoneal echogenic strands, dilated loops of bowel matted together and tethered posteriorly.

2. Computed tomography - peritoneal thickening and calcification, loculated ascites, narrowing of the bowel lumen, adherent bowel loops.

c. Pathological diagnosis.

1. Gross interstitial thickening which may be cellular (presumably fibroblasts) or acellular (interstitial collagen production) associated with loss of the mesothelium.

A definite diagnosis of EPS requires that a clinical diagnosis, based on at least two of the above 4 clinical features, is supported by pathognomonic radiological and/or pathological criteria. Several features on plain abdominal film (dilated loops of small bowel, air fluid levels, peritoneal calcification) and water-soluble contrast studies (proximal small bowel dilatation, obstruction accompanied by increased transit time) are highly suggestive of EPS but further investigations by abdominal CT, ultrasound if on PD and/or peritoneal histology are required to demonstrate the pathognomonic findings of peritoneal thickening and encapsulation before a definitive diagnosis is reached.

A number of conditions unrelated to PD can cause EPS and should be excluded before EPS is attributed to prior PD therapy.

Exclusion criteria of EPS related to PD

1. Post-surgical adhesions
2. Beta-blockers
3. Cirrhosis with ascites
4. Intraperitoneal chemotherapy or malignancy
5. Intra-abdominal infections (TB) or sarcoidosis
6. Auto-immune disease (SLE) or familial Mediterranean fever
7. Peritoneovenous or ventriculoperitoneal shunts
8. Peritoneal lavage with disinfectants or talc contamination
9. Haemodialysis
10. Idiopathic

References

1. Gandhi VC, Ing, TS, Daugirdas JT et al. Failure of peritoneal dialysis due to peritoneal sclerosis. Arch. Int. Med 1980; 140: 1201-1203.
2. Kawaguchi Y, Kawanishi H, Mujais S, Topley N, Oreopoulos DG for the International Society for Peritoneal Dialysis Ad Hoc Committee on Ultrafiltration Management in Peritoneal Dialysis. Encapsulating peritoneal sclerosis: Definition, aetiology, diagnosis and treatment. Perit Dial Int 2000; 20 (S4): S43- S55.
3. Mactier RA. The spectrum of peritoneal fibrosing syndromes in peritoneal dialysis. Adv Perit Dial 2000; 16: 223-228.
4. Kawaguchi Y, Saito A, Kawanishi H et al. Recommendations on the management of encapsulating peritoneal sclerosis in Japan, 2005: diagnosis,

predictive markers, treatment, and preventive measures. Perit Dial Int 2005; S4: S83-95.

5. Perks FJ, Murchison JT, Gibson P, Jackson SHL. Imaging findings in sclerosing encapsulating peritonitis. J R Coll Physicians Edinb 2004; 34: 116-119.

File	SRR_Encapsulating_Peritoneal_Sclerosis_definition.doc		
ver	date	author	comment
01	21032006	RM	
02	28062009	KS	Minor formatting