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ORIGINAL ARTICLE

# Encapsulating peritoneal sclerosis in children on chronic PD: a survey from the European Paediatric Dialysis Working Group

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## ABSTRACT

**Background.** Encapsulating peritoneal sclerosis (EPS) is a rare complication of peritoneal dialysis (PD) that is associated with significant morbidity and mortality in adults. There are scarce data for children. We performed a 10-year survey to determine the prevalence, risk factors and outcome for EPS in children.

**Methods.** Chronic PD patients in 14 dialysis units participating in the European Paediatric Dialysis Working Group between January 2001 and December 2010 were included in this study.

**Results.** Twenty-two cases of EPS were reported (prevalence 1.5%; 8.7 per 1000 patient-years on PD). Median PD vintage was 5.9 (1.6–10.2) in EPS and 1.7 (0.7–7.7) years in the remainder of the PD population ($P < 0.0001$). EPS patients had a

significantly higher peritonitis rate than non-EPS patients (P = 0.2). EPS was diagnosed while the child was on PD in 17 (77%), after conversion to haemodialysis (HD) in 3 and after transplantation in 2. Fifteen of 17 (88%) developed ultrafiltration (UF) failure. The median interval between UF failure and presentation with bowel obstruction was 2.8 (0.02–5.8) months. Twenty (91%) had clinical and radiological signs of bowel obstruction. Enterolysis was performed in 14 and 19 received immunosuppression or tamoxifen. Nine required parenteral nutrition. At final follow-up 4.8 (1.3–8.7) years after EPS diagnosis, 3 patients died, 11 had a functioning transplant and 8 were on HD.

**Conclusions.** The prevalence of EPS in European children on PD is comparable with that of adult PD patients, but mortality from paediatric EPS is significantly lower. A high index of suspicion is required for the diagnosis of EPS in children with longer dialysis duration, a high peritonitis rate and UF failure.

## INTRODUCTION

Encapsulating peritoneal sclerosis (EPS) is a rare but extremely serious complication of peritoneal dialysis (PD), and is associated with significant morbidity and mortality in adults. EPS has been reported in 0.7–3.3% of adult PD cohorts [1–6]. EPS may begin as small bowel dysfunction and progress, in its severest form, to complete intestinal obstruction as the bowel becomes trapped in the thickened and fibrosed peritoneal membrane [1]. The International Society of Peritoneal Dialysis (ISPD) has defined EPS as 'a clinical syndrome continuously, intermittently or repeatedly presenting with symptoms of intestinal obstruction due to adhesions of a diffusely thickened peritoneum' [1].

It is not clear as to why some individuals develop EPS, but chronic exposure of the peritoneum to PD fluids is thought to provoke activation of various inflammatory, fibrogenic and angiogenic cytokines, that eventually lead to progressive peritoneal fibrosis, vasculopathy and calcification [7]. Although most patients who have been on PD for a long duration do not develop EPS [1], the incidence of EPS increases with the length of time on PD [3, 5, 8]. The mortality from EPS is reported to be 35–69% [9], and many survivors require long-term parenteral nutrition. A high index of suspicion is required for an early diagnosis, and prompt diagnosis and management may influence outcome.

Although PD is the preferred modality of dialysis in children, and children can spend many years on dialysis before a transplant becomes available, there are only isolated case reports [10–17] and one multicentre report from the Japanese Registry [18] in the paediatric dialysis population. We conducted a 10-year survey across 14 paediatric dialysis centres from 11 European Union countries representing the European Paediatric Dialysis Working Group to review the prevalence, risk factors and outcome for EPS in children.

## MATERIALS AND METHODS

All patients receiving PD in paediatric nephrology centres between 1 January 2001 and 31 December 2010 were included, allowing at least 1-year follow-up for all patients. For inclusion in the study, the diagnosis of EPS required both clinical features of intestinal obstruction or disturbed gastrointestinal function as well as evidence of bowel encapsulation either radiologically or pathologically as defined in the ISPD position paper on EPS [1]. Children with coexisting bowel pathology that may clinically present as EPS were excluded. Patients were identified from recall and checked against the dialysis and transplant database held in each unit. Follow-up was continued in all patients (even after transfer to an adult unit) for the duration of the study period. Data were submitted to a central unit for analysis (Great Ormond Street Hospital, London) and verified by email correspondence and at meetings of the Group. Ethical approval for retrospective case-note review was obtained as per local requirements.

Anonymized retrospective data collection was performed to determine the demographics of the chronic PD population in each centre with additional detailed information on patient characteristics, specifics of PD therapy and dialysate used, presenting features of EPS including ultrafiltration (UF) failure, radiological tests, management and patient outcome for each patient in whom a diagnosis of EPS was suspected. The various dialysate solutions used were centre specific and are as follows: biocompatible [low glucose degradation products (GDP)] dialysate solutions Physioneal® (Baxter Healthcare Corporation, Deerfield, IL, USA) and BicaVera® (Fresenius Medical Care, Bad Homburg, Germany) or conventional (lactate based or high GDP) dialysate Dianeal® (Baxter Healthcare Corporation, Deerfield, IL, USA). For the purpose of this survey, UF failure was defined as reduced UF requiring either 4.25% or a mix of 2.5 and 4.25% glucose solution for ≥4 weeks, assessed on an individual basis by the patient's physician. Peritoneal equilibration tests (PETs) were performed annually as well as when UF failure was suspected in 5 of 14 centres for the duration of the study period (including $n = 3$ EPS cases), while the remainder only performed PET tests when UF failure was suspected.

### Statistical analysis

The data were tested for normality using the Kolmogorov–Smirnov test. Since most data were non-parametric, results are presented as median and range. Comparisons between the medians of the two groups were drawn using the Mann–Whitney *U*-test and correlations were tested using Spearman correlation tests for nonparametric data. A Kaplan–Meyer survival analysis was performed to determine the time to development of EPS. All P values are two-sided and $P \leq 0.05$ was considered statistically significant. Statistical analyses were performed using SPSS version 18.0.1 (SPSS, Chicago, IL, USA).

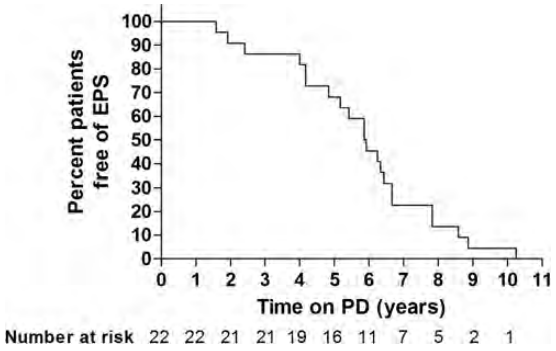


**FIGURE 1:** Kaplan–Meyer survival analysis showing the time to development of EPS.

## RESULTS

### Centre-specific details

During the 10-year study period, 1472 children received PD with a total of 2529 patient-years on PD. Twenty-two children developed EPS giving a prevalence of 1.5% or 8.7 cases per 1000 patient-years on PD. There were no EPS cases in 3 of 14 paediatric dialysis centres, one case each in eight centres, three cases each in two centres and eight cases in one centre.

### Patient demographics

The median age at the presentation of EPS was 14.8 (3.9–19.8) years. There were 15 (68%) boys. The underlying diagnoses were congenital anomalies of the kidneys and urinary tract ($n = 14$), congenital nephrotic syndrome ($n = 4$), focal segmental glomerular sclerosis (FSGS; $n = 2$) and bilateral renal venous thrombosis and post-cardiac transplantation in one patient each. Two patients were previously reported in a single-centre case report [11].

### Details of dialysis therapy

PD was the initial modality of dialysis in 17 (77%) children, 4 children had been converted from HD to PD and 1 started PD after a failed transplant. All children were on automated PD. The median age at start of PD was 2.2 (0.01–19.3) years, and the median time on PD was 5.9 (1.6–10.2) years (Figure 1). Seven (32%) had been on PD for ≤5 years, 17 (77%) had been on PD for ≤7 years and only one child was on PD for >10 years. Only one child was on PD for <2 years before EPS presentation. The duration on PD was significantly longer in those who developed EPS compared with the rest of the PD population [1.7 (0.7–7.7) years]; $P < 0.0001$.

Among those with EPS, there were a median of 4.8 (range 0–10) peritonitis episodes per patient. Peritonitis was due to Gram-positive organisms ($n = 42$, 49%; including *Staphylococcus aureus* in 14 species), Gram-negative organisms ($n = 23$, 27%; including *Pseudomonas aeruginosa* in 9 species), fungal infections ($n = 8$, 9%; all Candida species) and culture negative ($n = 13$, 15%). The annualized peritonitis rate among EPS patients was 1.9 (0.9–3.1). The peritonitis rate among EPS patients was significantly higher than in chronic PD patients in the same centres [annualized peritonitis rate 0.72 (0.3–1.2); $P = 0.02$]. Peritonitis was treated as per IPPR guidelines and infections were not refractory to treatment. PD catheter changes were required twice (0–6) per patient prior to EPS diagnosis.

Dianeal® was used in 13 (59%) and a biocompatible dialysate in 5 (22%; Physioneal® in 2 and BicaVera® in 3) children throughout their dialysis course, while 4 (18%) received a biocompatible dialysate (all Physioneal®) for a variable time since it was introduced in the renal unit. There was no difference in EPS prevalence-based exposure to conventional versus biocompatible dialysate ($P = 0.8$), nor did the duration of exposure to the different dialysates influence EPS occurrence [50 (23–122.4) versus 71 (19.2–106) years, respectively; $P = 0.14$]. There was no difference in the annualized peritonitis rate between children on conventional versus biocompatible dialysate ($P = 0.66$). Icodextrin was used in five children throughout their time on PD and in seven others when they lost residual renal function, while the remainder of the EPS patients were not exposed to Icodextrin. There was no difference in EPS prevalence based on Icodextrin use ($P = 0.9$).

We noted a high prevalence of EPS in a single centre with 8 of 22 (36%) of the total EPS cases. However, five children were referred from other centres for HD, all with UF failure and early clinical signs of bowel involvement. There was no difference in time on PD, age at start of PD, number of peritonitis episodes or age at diagnosis of EPS in this group as compared with the rest of the EPS cohort, but all children in this centre received conventional dialysate.

### Diagnosis of EPS

The clinical picture of EPS presented while the child was on PD in 17 (77%) cases, after conversion to HD in 3 cases and after transplantation in 2. UF failure was noted in 15/17 (88%) of children on PD. A PET was performed in 12 cases, and all were high transporters. For the three children who had multiple PET tests (3–5 per patient), one child was a high-average transporter for >6 years before EPS presentation, while the other two were low/low–average transporters for the duration of their dialysis course (2.7 and 4.9 years, respectively) until presentation with UF failure when PET confirmed a high-transporter status.

In 13 of 15 (87%) children, UF failure preceded the clinical signs and symptoms of bowel obstruction while two children had recurrent episodes of subacute bowel obstruction (with vomiting, abdominal distension and constipation) before developing UF failure. Three children presented with EPS within 3 (0.02–2.9) months after conversion to HD; all were converted to HD due to UF failure. Including three children on HD, the median interval between UF failure (defined as the use of 4.25% dextrose dialysate) and presentation with bowel obstruction was 2.8 (0.02–5.8) months.

Two children had functioning transplants for 2.9 and 7.5 years, respectively, before presenting with signs of subacute bowel obstruction. Neither of the transplanted patients had any intra-abdominal surgery other than PD catheter insertion and removal. These children had one and two episodes of peritonitis, respectively; all were fungal infections and PD catheters were removed each time. The renal allograft was placed

extraperitoneally in both, and both were on alternate day prednisolone and cyclosporine treatment. EPS presentation was acute with signs of intestinal obstruction and bowel perforation in one child each. The first patient underwent laparotomy with partial enterolysis: this showed matted bowel loops with 'cocooning' of the bowel in sclerosed peritoneum. The second patient developed an intestinal perforation and underwent laparotomy as well as immunosuppression with steroids and rapamycin, but died within 1 month of presentation.

The most common presenting feature was bowel obstruction and this was seen in 20 patients (91%); details are shown in Table 1A. Malnutrition (with >10% weight loss over 3 months) was seen in 16 (73%) children. The diagnosis of EPS was confirmed on abdominal ultrasound ($n = 21$) and/or computed tomography (CT scan; n = 15) and are described in Table 1B. A peritoneal biopsy was performed in four children at the time of PD catheter removal or enterolysis; this confirmed diffuse peritoneal fibrosis, sclerosis, hyalinosis and calcification in all.

**Table 1. Clinical presentation, radiological findings on ultrasound and CT scan in children with EPS**

| | Number | % |
|---|---|---|
| (A) Clinical signs and symptoms at EPS diagnosis | | |
| Bowel obstruction | 20 | 91 |
| Vomiting/abdominal distension/altered bowel habits | 20 | |
| Complete obstruction | 3 | |
| Bowel perforation | 3 | |
| Intra-abdominal mass | 8 | |
| Ascites | 4 | |
| Haemoperitoneum | 2 | |
| Malnutrition (>10% weight loss in 3 months) | 16 | 73 |
| Ultrafiltration failure | 15/17 on PD | 88 |
| (B) Radiological signs | | |
| Small bowel dilatation and abnormal peristalsis | 21 | 95 |
| Matted bowel loops with tethering to the posterior abdominal wall | 8 | 36 |
| Peritoneal thickening | 14 | 64 |
| Peritoneal calcification on CT scan | 11 | 50 |
| Loculated/septated ascites | 7 | 32 |
| Haemoperitoneum | 2 | 9 |

### Management and outcome of EPS

Seventeen (77%) children received immunosuppression (prednisolone in all, sirolimus, mycophenolate mofetil and colchicine in one each) and nine received tamoxifen. Fourteen (64%) children received surgical management with partial enterolysis in eight, complete enterolysis in three and laparotomy for bowel perforation in three. Four children needed a colostomy and one an ileostomy. Nine (41%) children received parenteral nutrition for a median of 2.3 (0.4–7.2) months. Two patients died from causes unrelated to EPS: one death was from septicaemia in a patient on HD and the other from pulmonary oedema and cardiac failure on day 15 post-transplantation. The third patient developed sepsis following intestinal perforation, and died with a functioning transplant within 1 month of EPS diagnosis. At final follow-up 4.8 (1.3–8.7) years after EPS diagnosis, 3 (13.6%) patients died, 11 had a functioning transplant and 8 were on HD (including 2 on home nocturnal HD and one on daily haemodiafiltration). None of the children had ongoing bowel problems and all were on full enteral feeds.

## DISCUSSION

In this multicentre survey of paediatric EPS cases, we report that the prevalence of EPS in European paediatric dialysis centres is comparable with that in adult PD patients. EPS was observed in children who received dialysis for a longer duration and had more frequent peritonitis episodes. UF failure preceded the presentation with signs and symptoms of bowel obstruction in the majority of children. The outcome of paediatric EPS cases was significantly better with a lower mortality and morbidity compared with adult patients. It is important that paediatric nephrologists are aware of this rare but extremely serious complication of PD so as to promptly diagnose or even prevent EPS development.

EPS is a well-recognized complication of PD in adult dialysis patients. The prevalence of EPS in different registry data is highly variable, and these results must be interpreted with caution keeping in mind the different duration of follow-up, inclusion of incident or prevalent PD patients, criteria for defining EPS using clinical and/or radiological and histological features and awareness of EPS. Thus, the prevalence of EPS has been variably reported as 0.7–3.3% in adult PD patients [2, 5, 6, 19, 20], but there is a notable difference in the follow-up period varying from 4.3 to 10 years. In order to define the true incidence of EPS, the Scottish Renal Registry has followed a cohort of adult PD patients from the start of PD for a total of 8 years and reported an incidence of 1.5% or 8.7 per 1000 person-years on PD [2]. Our study reports an identical prevalence of EPS in children with a comparable time on PD, suggesting that children are just as vulnerable to this complication as adults.

There are few data on the long-term outcome of PD in children, and most are isolated case reports describing the presentation and difficulties in management of this condition. The only published registry report on EPS in children comes from

the Japanese who report 17 (2.0%) EPS cases between 1981 and 1999 [15]. Of note, the time on PD was significantly longer than that seen in our cohort: all children had undergone PD for >5 years and the mean PD duration was 9.6 ± 3.3 years. EPS was diagnosed in 6.6 and 12% of patients dialysed for >5 and >8 years, respectively [15]. In our cohort, 8 of 22 (27%) cases with EPS were on dialysis for <5 years and the median time on dialysis was shorter (median 5.9 years). Two other important factors that differed between our cohorts is the lower peritonitis rate in Japanese children and the non-availability of biocompatible dialysate in that era. Similarly, a higher incidence of EPS has been reported in adult PD patients in the UK [2, 21] and Australia [5] as compared with Japanese studies. This may reflect a greater risk of EPS in European patients due to possible genetic predisposition, PD prescription and technique or peritonitis rates, and should be investigated in future prospective studies. Polymorphisms in genes expressing inflammatory [22], angiogenic [23] and fibrotic [22, 24] factors may influence an individual's susceptibility to EPS development, and at least in part explain geographic variations in EPS rates. A previous study has reported a 3/11 (27%) prevalence of EPS in children with FSGS [18], but this was not noted in our cohort wherein only two children (9%) had FSGS. A longer cumulative dialysis vintage, possibly due to disease recurrence post-transplantation and allograft loss or a common inflammatory signalling pathway involving TFG-β/SMAD may be involved [1].

We found a strong linear association between PD duration and prevalence of EPS that reflects findings in adult PD patients. Our cohort of prevalent PD patients had a significantly longer dialysis vintage compared with children without EPS in our centres. The Australian [5] and Japanese [3] registries report that after 4 years of PD exposure 5 and 1% of patients will develop EPS, respectively. In the Australian study, the overall rate of EPS increased progressively with duration of PD, with rates of 1.9, 6.4, 10.8 and 19.4% for patients on PD for <2, 5, 6 and 8 years, respectively [5]. This is corroborated by data from the Japanese Registry which showed a similar increase in EPS with dialysis vintage but overall lower rates: EPS was seen in 0.7% at 5 years and 2.7% at 8 years, 5.9% at 10 years and 17.2% at >15 years on continuous PD [3]. Based on these data, some authorities suggest that the PD duration should not exceed 5 years to try and avoid EPS [4, 6]. The Japanese recommend that PD may be continued for >8 years only in a select population of adult PD patients in whom there is no evidence of a high-transporter status, no regular requirement of hypertonic dialysate, absence of recurrent peritonitis, no increase in C-reactive protein, and who understand and accept the greater risks of EPS with prolonged use of PD [25]. However, no evidence-based data support a benefit of pre-emptively transferring long-term PD patients to HD, but the concept of an 'expiry date' for PD seems to be spreading among nephrologists [26]. However, there are several important reasons for resisting such an approach: the duration of PD is only one of the many risk factors for EPS, and not all PD patients are destined to develop EPS; secondly, as shown in the Scottish Renal Registry 74% of EPS cases were diagnosed after stopping PD [2], and PD by itself may play a protective role by continuously removing humoral factors related to the development of EPS from the peritoneal cavity [27]; and finally, the risks of shifting patients to HD, after a fixed time on PD in the absence of definite indications could impact negatively on their quality of life [26]. While the clinician must have a low threshold for the diagnosis of EPS in children with a longer dialysis vintage, and patients and their families should be made aware of the risks of EPS associated with long-term PD, the decision for continuation of PD needs to be individualized for each child.

It is not clear as to why some individuals can tolerate longer durations on PD: in our cohort EPS presented as early as 1.6 years and as late as 10.2 years after the start of PD, implying that other risk factors are involved. A genetic predisposition to develop EPS on exposure to dialysis fluid and modifiable risk factors such as the use of less biocompatible fluids, peritonitis episodes and angiotensin-converting enzyme inhibitors [28] need to be further explored. We found a significantly higher rate of peritonitis in children with EPS as compared with non-EPS patients in the same centres and also that reported in international registry data (median 4.8 versus 1.4 per patient) [29], with a high prevalence of *Staphylococcus aureus* and fungal peritonitis. A Japanese study has shown a similar 3-fold greater peritonitis rate in EPS patients as compared with those without EPS [30]. Peritonitis may superimpose a greater inflammatory burden on an already-damaged peritoneal membrane. These risk factors support the 'two-hit' hypothesis for EPS development [31]: advanced glycation end products, glucose degradation products and an acidic hyperosmolar dialysate create an inflammatory proangiogenetic and profibrotic milieu that degrades the mesothelium leading to peritoneal sclerosis, a precursor lesion of EPS. A second inducing event that causes an acute inflammatory response such as peritonitis episodes or possibly a genetic predisposition, then triggers the cascade of events leading to fibrosis, peritoneal encapsulation and bowel obstruction. Tamoxifen is thought to inhibit fibroblast transforming growth factor (TGF)-$\beta_1$ production, thereby preventing TGF-$\beta_1$-driven peritoneal thickening and fibrosis and is widely used in the treatment of EPS [32].

Ironically, PD itself may play a role in the progression of incipient EPS: in three (13.6%) children in our study, EPS became apparent after conversion to HD due to UF failure. A similar higher prevalence upto 30% has been reported in Japanese children [15]. Cycling the peritoneal cavity or using low-fill cycling PD while maintaining the patient on HD has been tried with short-term success by some [27], but early diagnosis and timely conversion to HD are advocated by most authorities. Similarly, EPS can present many years after transplantation, and exposure to calcineurin inhibitors, which upregulate TGF-β [33], has been identified as a potential risk factor.

Some nephrologists suggest screening patients who had a long duration of PD treatment for incipient EPS development. In our study, UF failure preceded the presentation with signs and symptoms of bowel obstruction in the majority of children. However, it is important to remember that UF failure is not pathognomonic of EPS. One study has shown that when

patients with EPS were compared with those who had UF failure alone, there were no differences in solute or fluid transport between the EPS and the UF failure groups, suggesting that a PET test alone is not effective in predicting the risk of EPS [34]. CT scans of patients with symptomatic EPS show characteristic abnormalities [35–37], and experienced radiologists can reliably diagnose EPS in over 90% of cases [35]. However, EPS has been shown to occur within a year of a normal CT scan, so CT screening of asymptomatic PD patients is not indicated [35, 37]. A Japanese study in 14 children who received continuous PD for >5 years has shown that virtually all had peritoneal sclerosis or fibrosis on biopsy, but only those with peritoneal sclerosis had UF failure and peritoneal calcifications on abdominal CT scan [10]. The authors recommend routine peritoneal biopsies in all children on PD for >5 years who have UF failure and calcifications on CT scan, but this is unlikely to become part of routine clinical practice.

The mortality in our paediatric EPS cases was significantly lower compared with that reported in adult EPS patients, and only one of the three deaths was directly attributable to EPS-related complications. The mortality in our cohort is comparable with that seen in the Japanese report wherein 3 of 11 children died [18]. This is in contrast to the extremely high mortality rate of 30–56% in adult EPS patients, with the majority of patients dying within 2 years of diagnosis [9]. The Scottish study on incident PD patients reports a mortality rate of 42% 1 year after EPS diagnosis and a median survival from diagnosis of 180 days. However, with experienced surgeons and improved survival using corticosteroids and newer immunosuppressive therapy including mycophenolate mofetil and sirolimus, the Japanese report impressive results [38, 39].

Our study has limitations common to many multicentre retrospective surveys. Patients were identified by recall and under-reporting is possible but unlikely as EPS is a major complication of PD. We do not have data on the number of new patients starting each year and are hence unable to report on the incidence of EPS in incident PD patients. It would be very interesting to compare glucose exposure as well as sequential PET data in children who developed EPS versus those who are high transporters but did not develop EPS in a future prospective study to help better delineate risk factors for EPS. Paediatric patients can be included in the European registry [40] and this will provide valuable information to inform clinicians and patients of the risks of developing EPS and the optimal management strategy for this rare complication of PD.

In conclusion, the prevalence of EPS in European children on PD is comparable with that seen in adult PD patients, but the outcome of paediatric EPS is significantly better and carries lower mortality. Risk factors for the development of EPS include a longer dialysis duration, high peritonitis rate and UF failure. We hope that this report will alert paediatric nephrologists to this rare but extremely serious complication of chronic PD and allow for prompt diagnosis and timely conversion to HD in high-risk patients with incipient UF failure.

## CONFLICT OF INTEREST STATEMENT

None declared.

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