



# Sclerosing encapsulating peritonitis: a latent threat. A change in the approach to surgical treatment

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## SUMMARY

*Sclerosing Encapsulating Peritonitis (SEP) is a rare but serious complication of continuous ambulatory peritoneal dialysis (CAPD) with a high morbi-mortality. We describe our experience with patients was diagnostic of SEP, their characteristics in CAPD and their clinic evolution after diagnosis. 190 CAPD patients were follow-up during 17 years. Eight patients (4,2%) developed SEP. Average age  $45 \pm 14$  years (range 29-64 years), four was male. Time in CAPD was  $72 \pm 29$  months (range 24-120 months). All patients have peritonitis previously (mean  $3 \pm 1$ ). We observe a change in peritoneum characteristics (D/P Cr 4), with an average of  $0.6 \pm 0.1$  at one year of CAPD, versus  $0.82 \pm 0.08$  at the end of CAPD, with statistic significance ( $p < 0.001$ ). There are increases in use of hypertonic bags:  $53\% \pm 28$  at beginning versus  $91\% \pm 27$  at end, with statistic significance ( $p < 0.009$ ). All patients show tendency to hyperphosphoremia (mean  $6.7 \pm 0.7$  mg/dl), with product calcium-phosphorus  $68.4 \pm 8.3$ . Five patients (62.5%) have a previous renal transplant, one lost due to early graft thrombosis and two lost due to acute rejection. Six patients (75%) have a previous abdominal surgery, although was extra peritoneal in all cases. The diagnosis of SEP was clinic suspicion in all cases, suggestive radiological data (intestinal handle group) and laparoscopy showing SEP (cocoon) with histological confirmation (fibrosis and peritoneal calcification) in four cases. The treatment was medical in six cases associated with surgery in four of them. The medical treatment was tamoxifen and/or corticosteroids, associated with total parenteral nutrition in two patients and enteral nutrition in one. Surgery in six patients: three as urgent surgery (all died) and three as programme surgery (two live still). Etiology of died was: three for sepsis, one for peritonitis after bowel perforation, one for severe problems of nutrition. The average survival of three patients alive was  $38 \pm 17$  months, two of them had programme surgery, and one with functioning transplant we opt for conservative treatment. The actuarial survival at 24 months was 51%. **Conclusion:** The SEP is a serious entity with high mortality. Although our short experience doesn't can indicate a concrete treatment, our personal impression is that early surgery associated with corticosteroids treatment may improve the prognostic*

Key words: **Sclerosing encapsulatin peritonitis. Peritoneal dialysis. Surgical treatment.**

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## PERITONITIS ESCLEROSANTE: UNA AMENAZA LATENTE. CAMBIO DE ACTITUD EN EL TRATAMIENTO QUIRÚRGICO

### RESUMEN

*La Peritonitis Esclerosante (PE) es una entidad grave que puede aparecer en pacientes en Diálisis Peritoneal (DP) con una morbi-mortalidad elevada. Describimos nuestra experiencia con pacientes diagnosticados de PE, sus características y evolución clínica. De 190 pacientes en DP durante un periodo de 17 años, hubo ocho casos de PE. Edad media  $45 \pm 14$  años (rango 29-64), cuatro eran varones. Tiempo en DP  $72 \pm 29$  meses. Todos presentaron episodios de peritonitis previa (media  $3 \pm 1$  episodios). Se observó un cambio en las características de transporte peritoneal (D/P Cr 4); media de  $0,6 \pm 0,1$  al año de DP, frente a  $0,82 \pm 0,08$  al final de DP ( $p < 0,001$ ). Incremento en el uso de bolsas hipertónicas:  $53\% \pm 28$  al inicio frente a  $91\% \pm 27$  al final ( $p < 0,009$ ). Cinco pacientes (62,5%) recibieron un injerto renal previo: uno con pérdida de función precoz por trombosis y dos por rechazo agudo. Seis pacientes (75%) tuvieron cirugía abdominal previa, en todos fue extraperitoneal. El diagnóstico de PE fue clínico en todos los casos, con datos radiológicos sugestivos y confirmación laparoscópica e histológica (fibrosis y calcificación peritoneal) en cuatro casos. Seis pacientes fueron intervenidos: tres de forma urgente (éxito en todos) y tres de forma programada (uno falleció). En seis pacientes se realizó tratamiento médico (tamoxifeno y/o esteroides), asociado con nutrición parenteral en dos y nutrición enteral en uno. Cuatro de esos seis pacientes necesitaron además tratamiento quirúrgico. Causas de éxito: tres por sepsis, uno por peritonitis post-perforación intestinal y uno por malnutrición severa. Media de supervivencia de los tres pacientes que viven es de  $38 \pm 17$  meses, dos de ellos habían sido intervenidos y el tercero era una trasplantada que se optó por tratamiento conservador. **Conclusión:** La PE es una entidad severa con mortalidad elevada. Aunque nuestra escasa experiencia no permite recomendar una pauta terapéutica concreta, nuestra impresión es que la cirugía precoz cuando existe obstrucción (PE encapsulante) seguida de tratamiento esteroideo puede mejorar el pronóstico.*

Palabras clave: **Peritonitis esclerosante. Diálisis peritoneal. Tratamiento quirúrgico.**

### INTRODUCTION

Sclerosing peritonitis (SP) is a rare condition seen in peritoneal dialysis (PD), and many times with a fatal course.<sup>1</sup> In sclerosing encapsulating peritonitis (SEP), the most severe form of the disease, the bowel gets entrapped within the fibrous tissue, leading to intestinal obstruction<sup>2</sup> and conditioning the clinical presentation.

SP is a clinical entity described in the 1970s in patients with no renal impairments and related with the use of b-adrenergic blockers.<sup>3</sup> In 1980, Gandhi *et al.* described in the United States the presence of thickening and marked sclerosis of the peritoneal membrane in five patients submitted to PD.<sup>4</sup> The etiology is unknown, although it is believed to be multifactorial, standing out the time from onset of peritoneal replacement therapy, greater use of hypertonic solutions to keep adequate ultrafiltration rates, and the presence of previous peritonitis.<sup>5-7</sup>

Several previous studies have shown a generally low incidence for this condition.<sup>7-10</sup> The prognosis is poor, with a high mortality rate up to 56%.<sup>8</sup> The main mortality causes are complications derived from intestinal obstruction or surgery, such as malnourishment or sepsis.

The treatment has not been clearly defined. Withdrawal of peritoneal dialysis is generally recommended, with or without immediate elimination of the peritoneal catheter and heparin washing.<sup>11,12</sup> Besides, therapy with corticosteroids<sup>13,14</sup> and/or tamoxifen,<sup>2,15,16</sup> and the option of surgical lysis of the adhesions, or debridement of encapsulated bowels.<sup>17,18</sup>

We describe the experience with patients diagnosed with SP at our Peritoneal Dialysis Unit during the last 17 years. We highlight the difference in their clinical presentation and course. We wanted to assess treatment options and, essentially, the change towards an earlier surgical approach.

## MATERIAL AND METHODS

We have analyzed those patients diagnosed with SP that were or had been on peritoneal dialysis program at our Center. The study period was from January of 1990 to May of 2006.

Diagnostic criteria for sclerosing peritonitis were both clinical and radiological. From the clinical point of view: the presence of signs of intestinal obstruction (pain and abdominal distension, vomiting) and its complications (fever, malnourishment). From the radiological perspective, the presence of intestinal loops grouping, air-fluid levels, absence of gas at the rectal ampoule, and peritoneal calcification. The confirmation was done, in those cases submitted to surgery, by the presence of abdominal «cocoon» sclerosis, peritoneal thickening, and widespread or generalized adhesions of the small bowel.

General epidemiological characteristics were analyzed in every patient: age, gender, etiology of renal failure, previous abdominal surgery, immunosuppressive therapies, number of renal transplants and cause of loss. Aiming at analyzing the influence of the dialysis technique on SP, we studied: the time on PD, time to occurrence of SP, number of peritonitis episodes and causing organisms, permeability characteristics of the peritoneum (determined by the simple peritoneal equilibrium test, or D/P Cr4), dialysis dose (Kt/v), liters of fluid in PD, percentage of the hypertonic bags (glucose concentration > 2.27%), use of icodextrin, and withdrawal of PD technique before the occurrence of the clinical picture of SP. We analyzed the surgical technique, as well as the time of surgery, each patient's clinical course, and cause of death.

The results obtained were analyzed by the SPSS (Statistics Program for Social Science) statistical package, version 12.0. The data are expressed as mean  $\pm$  SD (standard deviation). Qualitative data are expressed as frequency of occurrence. The survival study was done by using the Kaplan-Meier curves of actuarial survival. P values < 0.05 were considered significant.

## RESULTS

During the 17 years studied of our PD program, with a total of 190 patients, we found eight cases of SP, with a prevalence of 4.2%. Tables I and II show the main results for each patient.

Mean age was  $45 \pm 14$  years, range 29-64 years (Table I). For patients were males. The causes of CRF were diverse. The average time on PD program was  $72 \pm 29$  months (range 24-120 months), with perito-

nititis being the main cause for switching to hemodialysis (five patients).

During their stay on PD, all patients had had previous peritonitis episodes, with an average number of  $3 \pm 1$  (range 2-5). The most common causing organism was *coagulase-negative Staphylococcus*, followed by *Staphylococcus aureus* and *Pseudomonas aeruginosa* (Table III). Antibiotic therapy was established according to our protocol with vancomycin and cef-tazidime, and then following the culture sensibilities.

En six (75%) patients, there was an abdominal surgery, although it was extraperitoneal. Five patients had received some sort of transplant. The causes for graft loss were acute severe rejection (two cases), and thrombosis (one case), implicating early discontinuation of immunosuppressive therapy. Patient #4 had had an extraperitoneal surgery consisting in sequential bilateral nephrectomy due to uroepithelial carcinoma.

Only one case had received beta-blocker therapy for 30 months. At some time, 62.5% (five patients) had received ACEIS, with average treatment duration of  $16 \pm 8$  months.

The average time from the end of PD to the occurrence of SP was  $7.5 \pm 5$  months (range 2-14 months). In all patients the initial diagnosis was done by the presence of the symptoms listed in Table I. Then, it was confirmed by radiological techniques, with increasing sensitivity, and surgery. Intestinal obstruction was the main clinical presentation in five cases. At least two patients had hemoperitoneum throughout the course of PD, with little specific previous signs of diarrhea, nausea, and unspecific abdominal pain. In patient #7, the clinical picture of SP occurred when she had a functioning renal transplantation.

About the evolution parameters during the time on PD (Table II), we observed a change in the permeability characteristics of the peritoneum (D/P Cr4). The peritoneal equilibrium test performed during the first year on PD indicated a low transport profile, changing towards the end of PD to high-transporter profile. The weekly urea Kt/v did not change with time ( $2.45 \pm 0.4$  at the beginning, and  $2.2 \pm 0.37$  at the end of PD), but at the expense of increasing the dialysis dose ( $14.4 \pm 3$  versus  $8.6 \pm 2.6$  L/day at the beginning,  $SS < 0.002$ ), likely reflecting the progressive decrease in residual renal function. Throughout the stay on PD, an increase in the use of hypertonic bags was observed (mean  $53\% \pm 28$  at the beginning versus  $91\% \pm 27$  at the end,  $SS < 0.009$ ), more than 50% of them requiring at some point bags of 3.86%. Sixty-two point five percent (five cases) had received treatment with icodextrin.

All patients had very high phosphate levels throughout their course ( $6.7 \pm 0.7$ ), in spite of taking high

**Table 1.** Clinical characteristics

Case	Age (years)	Gender	Etiology of CRF	Time on PD (months)	Time end of PD to SP (m)	Num. of peritonitis episodes	Causa switching to HD	Previous Tx (Num) Cause of loss	Clinical picture	Treatment	Progression Time (m)	Etiology of exitus
1	52	V	IgA mesangial GN	84	2	4	Peritonitis (a)	1 Thrombosis	Pain, abdominal silence	Surgery (u)	Exitus 1.5 months	Peritonitis post-perforation
2	38	M	Chronic GN	120	8	3	Peritonitis (b)	1 severe RA	Intestinal obstruction	Steroids Surgery (SS)	Alive 18 months	
3	64	V	Diabetic nephropathy	72	3	3	Peritonitis (c)	0	Intestinal obstruction	T + S + PN	Exitus 5 months	Malnourishment
4	58	V	Bilateral nephrectomy	84	10	5	UF failure	0	Diarrhea, fever Hemoperitoneum	T + S Surgery (u)	Exitus 1 month	Sepsis
5	56	M	Unknown	48	12	3	UF failure	0	Abdominal pain	Surgery (u)	Exitus 2 months	Sepsis
6	29	M	LES	84	2	2	SP	2 (GCN, severe RA)	Malnourishment, sub-occlusion Hemoperitoneum	1° T + S 2° Switch to HD, Surgery (SS)	Exitus 2 months	Dehiscence of anastomosis Sepsis
7	29	M	Familial nephropathy	60	14 (in Tx)	3		2 (1° FCN, 2° functions)	Intestinal obstruction	Conservative Tamoxifen	Alive 48 months	
8	38	V	Diabetic nephropathy	24	9	3	Post Tx	1 (Tx P-K, GCN)	Intestinal obstruction	1° T + S + PN Surgery (SS)	Alive 48 months	

Abbreviations: m = months; DP = peritoneal dialysis; SP = sclerosing peritonitis; M/F = male/female; u = urgent; SS = elective; AR = acute rejection; GCN = graft chronic nephropathy; T = tamoxifen; S = steroids; PN = parenteral nutrition; Tx P-K = transplant of pancreas-kidney; UF = ultrafiltration; Peritonitis: (a) = pseudomonas aeruginosa; (b) = Staph aureus; (c) = escherichia coli.

doses of phosphate chelating agents, with a calcium-phosphate product of  $68.4 \pm 8.3$ .

The average total number of PD catheters per patient was  $1.8 \pm 0.6$  (range 1-3). In all cases, the catheter was taken out when transferred to hemodialysis.

Six patients received medical treatment, in four with associated surgery, and in two without it (Table I). The medical treatment was diverse: tamoxifen and/or steroids in six patients, parenteral nutrition was associated in two, and in one patient conservative therapy with enteral nutrition was carried out.

Seventy-five percent (six patients) received surgery. In three cases, it was urgent surgery, and in three scheduled surgery. All patients submitted to urgent surgery died within two months. Two of them from sepsis, and the third one from intestinal perforation-induced peritonitis. However, of three patients with scheduled surgery, only one died within two months due to sepsis, after requiring another urgent surgery because of anastomosis dehiscence. The remaining two are still alive. Patient #3 could not receive surgery because of several reasons, dying five months after the SP diagnosis from severe malnourishment. The surgery performed was long and laborious, consisting in lysis of the adhesions.

Three out of eight patients in our series are still alive. The mean survival for the five patients dying after the surgery was  $2.5 \pm 1.5$  months. The mean sur-

vival of the three patients still alive is  $38 \pm 17$  months. Of these, two were operated with scheduled surgery and one presented the clinical picture of SP while being with a functioning renal transplant; in this latter case, conservative therapy was chosen by increasing the steroid dose and tamoxifen. The actuarial survival of the patients for 24 months was 51%.

## DISCUSSION

SP is a rare condition with important associated complications leading to high morbidity and mortality of patients on PD program.<sup>1, 19, 20</sup>

Its prevalence ranges 0.5%-2.8% according to the literature reviewed,<sup>19</sup> with some old series reporting rates of 7.3%.<sup>10</sup> The lowest rates (0.5%-0.8%) were described in Canada by Afthentopoulos *et al.*<sup>21</sup> and those from the multicenter studies from Japan,<sup>7</sup> Australia,<sup>8</sup> and more recently Korea.<sup>22</sup> The English study by Jenkins *et al.*<sup>23</sup> shows an intermediate rate (1.4%). The highest rates (2.2%-2.8%) come from several Japanese studies.<sup>9, 24, 25</sup> In their Australian series, Rigby and Hamley<sup>8</sup> pointed out the increasing prevalence as the time spent by the patients on PD increased. Thus, at two years it was 1.9%, increasing to 19.4% at eight years of being on PD. In our center, the prevalence was 4.2%, with a mean stay on PD of almost

**Table II.** Characteristics of the PD technique

Case #	Catheter type (Num.)	Catheter withdrawal Switch to HD	D/P Cr4 Start (a)/End	Kt/V Start/End	UF (cc) Start/End	Liters of PD fluids Start/End	% bags > 2.25% Start/End	Icodextrin
1	Thenkoff (3)	Yes	0.49 (1 a)/0.64	1.98/2.01	1,300/1,080	6/17	25/100 (b)	No
2	Thenkoff (2) Tugsteno (1)	Yes	0.5 (1 a)/0.9	2.5/3	1,700/1,800	8/17	50/100	Yes
3	Thenkoff (1)	Yes	0.71 (2 a)/0.9	1.82/1.71	1,200/1,800	15/17	75/100	Yes
4	Thenkoff (2)	Yes	0.74 (1 a)/0.8	2.5/2	1,700/1,300	8/14	50/100 (c)	Yes
5	Thenkoff (2)	Yes	0.48 (1 a)/0.78	2.5/2.36	1,300/1,300	8/14	25/100	Yes
6	Thenkoff (2)		0.61 (1 a)/0.87	2.5/2.16	1,300/600	8/14	25/25	Yes
7	Thenkoff (2)	Yes (Tx)	0.66 (1 a)/0.84	2.9/2.18	1,500/1,000	8/8	100/100 (b)	No
8	Thenkoff (1)	Yes (Tx)	0.66 (1 a)/0.88	2.9/2.18	2,100/900	8/14	75/100 (c)	No
p			0.001	0.129	0.177	0.002	0.009	

Abbreviations: PD = peritoneal dialysis; Tx = transplant; a = year; SS = statistical significance: bags 3.86% at the beginning and at the end (b) or only at the end of PD (c).

six years, with five out of eight patients in our series having a longer stay than that.

The prognosis is poor, with a reported mortality rate in the largest study, the multicenter Japanese study,<sup>7</sup> of 43.5% and of 56% in the Australian multicenter study.<sup>8</sup> In the prospective study carried out by Kawanishi<sup>9</sup> in Japan, the prevalence and mortality rates were 2.5% and 38%, respectively.

The higher prevalence in recent years may be the expression of increased diagnostic suspicion and better identification of this condition, partly due to the technological advances (high-resolution CT scan, etc.). On the other hand, it could be also influenced by a reduction in the peritonitis rate, allowing so longer stay on PD and the occurrence of SP.<sup>8,9,19</sup>

The pathogenesis of SP is believed to be similar to that of another condition, peritoneal fibrosis, and due to the loss of ultrafiltration as similar predisposing factors.<sup>19</sup> The damage to the peritoneal mesothelium may be considered as the initial stimulus for developing SP in susceptible patients.<sup>20</sup> Fibrin deposition may be considered as one of the essential histological findings,<sup>26</sup> as a manifestation of an inflammatory process, both acute and essentially chronic, which occurs in this pathology.

However, some authors such as Garosi *et al.*<sup>27</sup> postulate the possibility of considering SP and peritoneal fibrosis as two different conditions. They based their hypothesis in the higher frequency and lower clinical manifestations of peritoneal fibrosis, as compared to SP, which is less frequent and has higher clinical involvement and mortality. Future studies will help making clear this controversy.

Among the multiple risk factors reported in the different publications, most of them agree on two of them.<sup>3,4,7,9,10,19</sup> In the first place, the existence of re-

peated episodes of peritonitis, and in the second place, the duration of PD.<sup>7-9,9,22,25</sup>

The loss of the mesothelium during the infectious process would induce damage to the peritoneal membrane because of bioincompatibility with PD fluids.<sup>6,8</sup> During acute peritonitis, there is an unbalance between fibrinolysis and fibrinogenesis. The loss of the fibrinolytic activity of the peritoneal membrane may contribute to the development of SP after severe peritonitis episodes.<sup>6,8,28,29</sup> In our series, all patients had some episode of peritonitis, with a mean of  $3 \pm 1$  and great variety of causing organisms (Table III). The most common were *coagulase-negative Staphylococcus*, *Staphylococcus aureus*, and *Pseudomonas aeruginosa*.

A second remarkable risk factor is the duration of the PD technique.<sup>7-9,19,22</sup> As it has already been pointed out, in the Australian series<sup>8</sup> a prevalence of 1.9% was reported among patients with two years of stay on PD, increasing to 6.4%, 10.8%, and 19.4% in patients on PD for 5, 6, and 8 years, respectively. In our eight patients diagnosed with SP, the mean time on PD was  $72 \pm 29$  months, with one patient being for 10 years and four other patients above five years.

It is worth mentioning the cases #1 and #2 (Table I) in whom the reason for switching to hemodialysis was the presence of previous episodes of severe peritonitis due to *Pseudomonas aeruginosa* and *Staphylococcus aureus*, respectively. Both had been for a long time on PD, 84 and 120 months, respectively, with a personal history of previous peritonitis episodes, and supposedly their peritoneum could already be partially damaged. The episode of severe peritonitis induced by these two microorganisms could have been the ultimate trigger for the onset of SP.

**Table III.** Peritonitis, number and type of microorganism

Germen/Patient	1	2	3	4	5	6	7	8	Total (%)
Staph coagulase (-)				2	2		2		6 (23)
Staph aureus	1	1				1		1	4 (15.5)
Pseud. aeruginosa	2	1		1					4 (15.5)
Sterile fluid			2					1	3 (11.6)
Escherichia coli			1			1			2 (7.7)
Corynebacterium					1		1		2 (7.7)
Klebsiella								1	1 (3.8)
Stenotrophomonas	1								1 (3.8)
Strept. bovis			1						1 (3.8)
Bacillus saprophyticus				1					1 (3.8)
Polymicrobial				1					1 (3.8)
<b>Total</b>	<b>4</b>	<b>2</b>	<b>4</b>	<b>5</b>	<b>3</b>	<b>2</b>	<b>3</b>	<b>3</b>	<b>26</b>

Abbreviations: Staph = staphylococcus; Pseud. = pseudomona; Strep = streptococcus.

As a result of the stay on PD, several series point out the loss of ultrafiltration as an additional factor for the occurrence of SP.<sup>22,30</sup> In our patients, the peritoneal equilibrium test shows a D/P Cr4 value during the first year in agreement with low average transport (mean  $0.6 \pm 0.1$ ), progressing throughout the observation period to a value compatible with high transport ( $0.82 \pm 0.08$ ) (Table II). This is accompanied by an increase in the number of hypertonic bags necessary to maintain ultrafiltration. Up to 50% of the patients required the use of bags with a concentration of 3.86% of glucose at the end of PD. Besides, throughout the follow-up period, as expected, it is necessary to increase the volume of dialysis fluid as residual renal function decreases in order to maintain the appropriateness parameters.

Although classically beta-blockers, especially practolol,<sup>3</sup> have been postulated as another predisposing factor in SP occurrence, in our patients only one received this kind of therapy. Most of them, five patients, received ACEIs at some time of their course. The peritoneal catheter would be another factor of peritoneal irritation. In our series, in all cases the catheter was taken out because of bad course of peritonitis, but in patient #5 in whom it was taken out because of ultrafiltration failure.

The clinical presentation is varied, the most common symptoms being those related with ultrafiltration failure and others of slower and more progressive onset, such as anorexia, nausea, and weight loss, or more evident in relation with the clinical picture of intestinal obstruction.<sup>20</sup> These findings reflect a fibrosis-induced inhibition of peristalsis, involving the bowels and leading to encapsulating sclerosis (abdominal «cocoon»). Ascites develops when lymphatic

drainage is impaired.<sup>20</sup> Many times, ascites may be hemorrhagic in nature.

In the series presented, the main symptoms were derived from the clinical picture of intestinal obstruction with associated data of malnourishment. The diagnosis was based on clinical data supported by the radiological signs of intestinal loops grouping, the presence of peritoneal calcifications, and ascites. At surgery, the typical carapace or «cocoon» could be observed, including all the intestinal loops.

Although the clinical picture secondary to intestinal obstruction is admitted by most of the authors, the presence of other manifestations is also cited in several publications postulating a classification by stages of SP according to the clinical picture and the treatment options.<sup>19,25</sup> In these articles, a first asymptomatic stage is reported, with later presence of inflammation, followed by a stage with encapsulation of the intestinal loops, and finally the stage of intestinal obstruction would follow, with disappearance of inflammation data and where the indicated treatment would be surgery with enterolysis.

We would like to emphasize the management. Most of the published series agree in the need of peritoneal rest. Therefore, PD would be withdrawn, transferring the patient to HD.<sup>20,22</sup> In the case that catheter withdrawal would not be required, peritoneal washes with heparin could be associated. In the case the inflammatory process progresses and obstruction signs ensure, the indication of corticosteroids is evident as well as the start of parenteral or enteral nutrition.<sup>13,14,19,20</sup> Besides corticosteroids, the use of several immunosuppressants has been reported, among which the most widely used and with greater experience is tamoxifen.<sup>15,16</sup> Less frequently azathioprine,<sup>8</sup> sirolimus

(both as monotherapy and associated to corticosteroids),<sup>31</sup> mycophenolate, and tacrolimus<sup>32</sup> have also been used.

The surgical management is more controversial. Classically, mortality was quite high after surgery, usually because of problems with intestinal ischemia.<sup>33,34</sup> In the year 2000 and in 2002, the mortality rate as a result of surgical complications was 45% and 82% in patients requiring enterotomy and intestinal anastomosis.<sup>33,35</sup> Other centers reported mortality rates of 31%.<sup>36,37</sup> These percentages made that most of the teams would chose a conservative approach in SP.

However, in recent years, several series have been published with more promising surgical outcomes. In the year 2005, Kawanishi *et al.*<sup>17</sup> reported an important improvement in survival with post-surgical mortality of 4%. This improvement is based on the change of the surgical technique. Given the high mortality from sepsis due to rupture of the anastomosis, presumably because of degeneration of the intestinal wall,<sup>17,33</sup> intestinal resection and anastomosis are avoided, trying the enterolysis technique. This implies the resection or lysis of adhesions and of the calcified «cocoon» or carapace that covers the peritoneum.<sup>17</sup> A differentiating circumstance of this type of surgery is being a prolonged and labor-intensive technique, requiring a certain degree of expertise.<sup>9,17,37</sup> It is recommended that this technique is done with scheduled surgery, avoiding an urgent intervention by the surgical team on call that will more likely lead to sepsis from perforation of a degenerated intestinal wall.

In our series of eight patients, only two of them did not receive surgical treatment (Table I). One case for being hemophiliac with high associated comorbidity in whom the possibility of bleeding during the surgery and the difficulty in managing it advised against surgical intervention. The second case was a woman with functioning renal transplant in whom conservative management was chosen. Besides, tamoxifen was added to the immunosuppressive therapy for transplantation (corticosteroids, tacrolimus, and mycophenolate mofetil), with good clinical course and still surviving.

In the remaining six patients of our series, three had urgent surgery, with a survival shorter than one month. In the remaining three, with scheduled surgery, two are still alive and only one died two months after surgery.

Our experience with only eight patients, but spanning a long time, has allowed us using several medical and surgical options. The option showing the best outcome has been the one implicating, in the first place, switching to hemodialysis associated with tamoxifen and corticosteroids, and parenteral or enteral

nutrition, trying to achieve intestinal rest and a decrease in intestinal inflammation. Then, elective surgery, as soon as possible, is done with enterolysis.

To conclude, SP is a rare condition that should always be kept in mind in PD patients, particularly when these patients have been for a long time on this dialysis technique, have presented repeated episodes of peritonitis, or with a poor course, or have ultrafiltration failure with changes in the peritoneal membrane permeability characteristics. Before this diagnosis, switching to hemodialysis as a measure of peritoneal rest associated with parenteral nutrition and corticosteroids or immunosuppressants that will decrease the inflammation, will prepare the patient for the surgery, as early as possible. The best surgical option would be enterolysis technique.

Finally, we would like to highlight the need for higher consensus in the diagnosis and treatment of this pathological condition, although the first steps in this sense have already been taken.<sup>4</sup>

## REFERENCES

1. Garosi G, Di Paolo N, Sacchi G, Gaggiotti E: Sclerosing peritonitis: a nosological entity. *Perit Dial Int* 25(Supl. 3): S110-S112, 2005.
2. Eltoun MA, Wright S, Atchley J, Mason JC: Four consecutive cases of peritoneal dialysis-related encapsulating peritoneal sclerosis treated successfully with tamoxifeno. *Perit Dial Int* 26(2): 203-6, 2006.
3. Brown P, Baddeley H, Read AE, Davies JD, McGarry J: Sclerosing peritonitis, an unusual reaction to a beta-adrenergic blocking drug (practolol). *Lancet* 2(7895): 1477-81, 1974.
4. Gandhi WC, Humayan HM, Ing TS, Daugirdas JT, Jablowski VR, Iwatsuki S, Geis WP, Hano JE: Sclerotic thickening of the peritoneal membrane in maintenance peritoneal dialysis patients. *Arch Intern Med* 140: 1201-3, 1980.
5. Soria M, Arrieta J, Ugarte I, Moína I, Guerra M, Escanero J: Alteraciones del pH intracelular en fibroblastos peritoneales inducida por factores implicados en la esclerosis peritoneal. *Nefrología* 24(2): 158-66, 2004.
6. Dobbie JW: Pathogenesis of peritoneal fibrosing syndromes (sclerosing peritonitis) in peritoneal dialysis. *Perit Dial Int* 12: 14-27, 1992.
7. Nomoto Y, Kawaguchi Y, Kubo H, Hirano H, Sakai S, Kurokawa K: Sclerosing encapsulating peritonitis in patients undergoing continuous ambulatory peritoneal dialysis: a report of the Japanese Sclerosing Encapsulating Peritonitis Study Group. *Am J Kidney Dis* 28: 420-7, 1996.
8. Rigby RJ, Hawley CM: Sclerosing peritonitis: the experience in Australia. *Nephrol Dial Transplant* 13: 154-9, 1998.
9. Kawanishi H, Kawaguchi Y, Fukui H, Hara S, Imada A, Kubo H, Kin M, Nakamoto M, Ohira S, Shoji T: Encapsulating peritoneal sclerosis in Japan: a prospective, controlled, multicenter study. *Am J Kidney Dis* 44: 729-37, 2004.
10. Rottembourg J, Issad B, Langlois P *et al.*: Loss of ultrafiltration and sclerosing encapsulating peritonitis during CAPD. Evaluation of the potential risk factors. *Adv Perit Dial* 1: 109-17, 1985.
11. Cancarini GC, Sandrini M, Vizzardi V, Bertoli S, Buzzi L, Maiorca R: Clinical aspects of peritoneal sclerosis. *J Nephrol* 14(Supl. 4): 39-47, 2001.

12. Poultsidi A, Liakopoulos V, Eleftheriadis T, Zarogiannis S, Bouchlariotou S, Stefanidis I: Gross calcification of the small bowel in a continuous ambulatory peritoneal dialysis patient with Sclerosing peritonitis. *Adv Perit Dial* 22: 104-7, 2006.
13. Kuriyama S, Tomonari H: Corticosteroid therapy in encapsulating peritoneal sclerosis. *Nephrol Dial Transplant* 16; 1304-5, 2001.
14. Mori Y, Matsuo S, Sutoh H, Toriyama T, Kawahara H, Hotta N: A case of a dialysis patient with sclerosing peritonitis treated successfully with corticosteroid therapy alone. *Am J Kidney Dis* 30: 275-8, 1997.
15. Del Peso G, Bajo MA, Gil F, Aguilera A, Ros S, Costero O, Castro MJ, Selgas R: Clinical experience with tamoxifen in peritoneal fibrosing syndromes. *Adv Perit Dial* 19: 32-35, 2003.
16. Evrenkaya TR, Atasoyu EM, Unver S, Basekim C, Baloglu H, Tulbek MY: Corticosteroid and tamoxifen therapy in sclerosing encapsulating peritonitis in a patient on continuous peritoneal dialysis. *Nephrol Dial Transplant* 19: 2423-4, 2004.
17. Kawanishi H, Watanabe H, Moriishi M, Tsuchiya S: Successful surgical management of encapsulating peritoneal sclerosis. *Perit Dial Inter* 25: s39-47, 2005.
18. Serafimidis C, Katsarolis I, Vernadakis S, Rallis G, Giannopoulos G, Legakis N, Peros G: Idiopathic sclerosing encapsulating peritonitis (or abdominal cocoon). *BMC Surg* 6: 3, 2006.
19. Chin AI, Yeun JY: Encapsulating peritoneal sclerosis: an unpredictable and devastating complication of peritoneal dialysis. *Am J Kidney Dis* 47: 697-712, 2006.
20. Courtney AE, Doherty CC: Fulminant Sclerosing peritonitis immediately following acute bacterial peritonitis. *Nephrol Dial Transplant* 21: 532-4, 2006.
21. Afthentopoulos IE, Pasadakis P, Oreopoulos OG, Bargman J: Sclerosin peritonitis in continuous ambulatory peritoneal dialysis patients: one center's experience and review of the literature. *Adv Ren Replace Ther* 5: 157-67, 1998.
22. Kim BS, Choi KY, Ryu DR, Yoo TH, Park HC, Kang SW, Choi KH, Ha SK, Han DS, Lee HY: Clinical characteristics of dialysis related Sclerosing encapsulating peritonitis: Multi-center experience in Korea. *Yonsei Med J* 46: 104-11, 2005.
23. Jenkins SB, Leng BL, Shortland JR, Brown PW, Wilkie ME: Sclerosing encapsulating peritonitis: a case series from a single UK center during a 10-year period. *Adv Perit Dial* 17: 191-5, 2001.
24. Kawaguchi Y, Kawanishi H, Mujais S, Topley N, Oreopoulos DG: Encapsulating peritoneal sclerosis: definition, etiology, diagnosis and treatment. International Society for Peritoneal Dialysis Ad Hoc Committee on Ultrafiltration Management in Peritoneal Dialysis. *Perit Dial Int* 20(Supl. 4): S43-S55, 2000.
25. Nakamoto H, Kawaguchi Y, Suzuki H. Encapsulating peritoneal sclerosis in patients undergoing continuous ambulatory peritoneal dialysis in Japan. *Adv Perit Dial* 18: 119-23, 2002.
26. Honda K, Nitta K, Horita S, Sukada M, Itabashi M, Nihei H, Akiba T, Oda H: Histologic criteria for diagnosing encapsulating peritoneal sclerosis in continuous ambulatory peritoneal dialysis patients. *Adv Perit Dial* 19: 169-75, 2003.
27. Garosi G, Cappelletti F, Di Paolo N: Fibrosis and sclerosis: different disorders or different stages? *Contrib Nephrol* 150; 62-69, 2006.
28. Jorres A, Gahl GM, Frei U: Peritoneal dialysis fluid histocompatibility: does it really matter? *Kidney Int* 46(Supl. 48): S79-S86, 1994.
29. Junor BJ, McMillan MA. Immunosuppression in Sclerosing peritonitis. *Adv Perit Dial* 9: 187-9, 1993.
30. Yamamoto R, Nakayama M, Hasegawa T, Miwako N, Yamamoto H, Yokoyami K, Ikeda M, Kato N, Hayakawa H, Takahashi H, Otsuka Y, Kawaguchi Y, Hosoya T: High transport membrane is a risk factor for encapsulating peritoneal sclerosis developing after long-term continuous ambulatory peritoneal dialysis treatment. *Adv Perit Dial* 18: 131-4, 2002.
31. Rajani R, Smyth J, Koffman CG, Abbs I, Goldsmith DJ: Differential effects of Sirolimus vs prednisolone in the treatment of sclerosing encapsulating peritonitis. *Nephrol Dial Transplant* 17: 2278-80, 2002.
32. Dejagere T, Evenepoel P, Claes K, Kuypers D, Maes B, Vanrenterghem Y: Acute-onset, steroid-sensitive, encapsulating peritoneal sclerosis in a renal transplant recipient. *Am J Kidney Dis* 45: e33-e37, 2005.
33. Kawanishi H: Surgical treatment for encapsulating peritoneal sclerosis. *Adv Perit Dial* 18: 139-43, 2002.
34. Kittur DS, Korpe SW, Raytch RE, Smith GW: Surgical aspects of sclerosing encapsulating peritonitis. *Arch Surg* 125: 1626-8, 1990.
35. Kawaguchi Y, Kawanishi H, Mujais S, Topley N, Oreopoulos DG: Encapsulating peritoneal sclerosis: definition, etiology, diagnosis and treatment. International Society for Peritoneal Dialysis Ad Hoc Committee on ultrafiltration management in peritoneal dialysis. *Perit Dial Int* 20(Supl. 4): S43-S55, 2000.
36. Summers AM, Clancy MJ, Syed F, Harwood N, Brenchley PE, Augustine T et al.: Single-center experience of encapsulating peritoneal sclerosis in patients on peritoneal dialysis for end-stage renal failure. *Kidney Int* 68: 2381-8, 2005.
37. Summers AM, Brenchley PE: An international encapsulating peritoneal sclerosis registry and DNA bank: why we need one now. *Perit Dial Int* 26: 559-63, 2006.