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Bulletin de la Dialyse à Domicile

Outcome of patients with polycystic kidney disease initially treated with peritoneal dialysis. Experience of our home dialysis unit since 1997

(Devenir des patients atteints de polykystose rénale traités en première intention par dialyse péritonéale. Expérience du pôle de dialyse à domicile depuis 1997)

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Résumé

Nous avons, à l'aide du Registre de Dialyse Péritonéale de Langue Française, extrait et étudié rétrospectivement les infections péritonéales et le devenir des trente patients atteints de polykystose rénale autosomique dominante de l'adulte pris en charge en dialyse péritonéale depuis 1997 dans notre unité.

Il s'agissait de 15 hommes et 15 femmes, âgés en moyenne de 54 ans. L'atteinte hépatique était présente chez 85% d'entre eux. Dix patients n'avaient aucune morbidité. Le score de comorbidité Charlson était supérieur ou égal à 4 chez cinq patients. La majorité des patients était traitée par dialyse péritonéale automatisée nocturne. Un seul patient était non autonome en dialyse. Le temps cumulé de suivi pour l'ensemble des patients était de 836 mois soit en moyenne 28 mois par patient. Onze patients ont présenté au moins une infection péritonéale. Le nombre total d'infections péritonéales était de 24 dont 9 à bacille gram négatif. L'incidence des infections péritonéales était d'un épisode tous les 35 mois-patient. L'infection péritonéale a été responsable du transfert en hémodialyse de deux patients et du décès d'un patient. Quinze patients (50%) ont bénéficié d'une transplantation rénale. Un seul patient a nécessité une néphrectomie préparatoire à la transplantation. Sept patients ont été transférés en hémodialyse avec une médiane de traitement en dialyse péritonéale de 36 mois. Quatre patients sont décédés. Quatre patients sont actuellement traités en dialyse péritonéale.

En conclusion, ce travail rétrospectif montre que la dialyse péritonéale, particulièrement la dialyse péritonéale automatisée nocturne, est une bonne option de traitement en dialyse pour les patients atteints de polykystose rénale.

Mots clés : polykystose rénale, dialyse péritonéale, infection péritonéale, transplantation rénale

Summary

We extracted data from the French Language Peritoneal Dialysis Register (RDPLF) and retrospectively studied peritonitis and the outcome of 30 patients with polycystic kidney disease initially treated with peritoneal dialysis within our dialysis unit since 1997.

There were 15 men and 15 women with a mean age of 54 years. Eighty-five percent of the patients had hepatic involvement. Ten patients did not suffer from comorbidities. The Charlson comorbidity index was greater than or equal to four in five patients. Most of the patients were treated with automated peritoneal dialysis during the night. Only one patient was not self sufficient with peritoneal dialysis. The entire medical monitoring lasted 836 months, representing an average of 28 months per patient. Eleven patients had one or more peritonitis. There were a total of 24 peritonitis, nine with gram negative bacillus. Incidence of peritonitis was one episode every 35 patient months. Peritonitis was responsible for sudden admission to the hemodialysis unit in two cases and death in one case. Fifteen patients (50%) benefited from renal transplantation. Only one patient had to undergo nephrectomy prior to renal transplantation. Seven patients were admitted to the hemodialysis unit (the median duration time on peritoneal dialysis was 36 months). Four patients died. Four patients are currently being treated with peritoneal dialysis.

In conclusion, this retrospective study points out that peritoneal dialysis, especially nocturnal automated peritoneal dialysis, is a good treatment option for patients with polycystic kidney disease necessitating dialysis.

Key words : polycystic kidney disease, peritoneal dialysis, peritonitis, renal transplantation

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INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is the main hereditary kidney disease worldwide, responsible for an average of 10% of cases of end-stage chronic kidney disease in France [1]. It is characterized by a constant increase, sometimes significantly, in the volume of the two kidneys carrying innumerable cysts, but also often of the liver which can, in certain cases, occupy an important place in the peritoneal cavity. It is associated with increased frequency of diverticular colonic pathologies and classically hernias of the abdominal wall due to abdominal hypertension generated by the volume of the liver and kidneys [2].

According to the recommendations of the National Authority of Health in 2008 [3], ADPKD is not, despite this particular clinical phenotype, a contraindication for the initiation of treatment by peritoneal dialysis, but a certain reluctance of nephrologists concerning this dialysis technique is still encountered. The main issue, above all, is that of kidney transplantation program (in particular the management of increased kidney size) in this relatively young population on dialysis.
We report our experience in the management of patients with ADPKD and treated with peritoneal dialysis (PD) as first-line treatment since 1997 in our home dialysis unit.

METHODS

Using the French Language Peritoneal Dialysis Register (RDPLF), we extracted data from the regularly updated main module «survival and infection» and retrospectively studied the course and fate of the 30 patients with ADPKD for whom PD has been prescribed as a first-line treatment since 1997 in our home dialysis unit. The computerization of the dialysis medical record in our center appeared in 2005 after the start of our participation in the RDPLF (1997), which explains why certain clinical and therapeutic information may be absent in six patients treated between 1997 and 2005. The diagnosis of ADPKD is based on the usual criteria, in particular radiological, and a family history of kidney disease [4]. Finally, we do not have a sufficient number of peritoneal clearance measurements in this cohort of patients to report results regarding the evolution of renal function.

RESULTS

1. Clinical characteristics and treatments of patients

Table 1 describes the clinical characteristics and the PD follow-up time of the 30 ADPKD patients extracted from the RDPLF since 1997. These patients were on average 54 years old when they received PD and had little comorbidity (in particular diabetes). The Charlson comorbidity index was greater than or equal to four in five patients. Only three patients had a history of cardiovascular disease. One patient was a carrier of inflammatory disease of the digestive tract. Hepatic involvement was present at the time of dialysis treatment in 85% of patients. The cumulative follow-up time for this cohort of patients was 836 months, or 28 months on average per patient.

Table 2 describes the main treatments and the treatment modalities for peritoneal dialysis, continuous ambulatory peritoneal dialysis (CAPD) or nighttime automated peritoneal dialysis (APD). The vast majority of patients were self sufficient in the dialysis method and were treated with APD. Erythropoietin treatment was used in 43% of patients. Seventy-five percent of patients

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were receiving antihypertensive therapy (angiotensin-converting enzyme inhibitor, angiotensin 2 antagonist, or calcium channel blocker).

Table I. Clinical characteristics of patients

Sex	15 M / 15 F
Average age on dialysis	54 years
	(min/max: 33 - 75 years)
Hepatic involvement	22/26 patients
Comorbidities	
None	10/26 patients
Diabetes	1/26 patients
Severe cardiopathy	3/26 patients
Inflammatory colon pathology	1 patient
BMI > 30	3/25 patients
Charlson index ≥ 4	5/27 patients
Follow-up	
Total follow-up in months (30 patients)	836
Average follow up in patient months	28
	(min/max: 4 - 91 months)

Table II. Treatments

Anti-hypertensive drugs	18/24 patients (75%)
Erythropoïetin	10/23 patients (43%)
Peritoneal Dialysis (PD)	
CAPD	7 patients
APD	23 patients (77%)
Autonomous on PD	29/30 patients

2. Peritoneal infections

Peritoneal infections are shown in Figure 1. Eleven patients presented at least one peritoneal infection. The total number of peritoneal infections for all patients was 24. The incidence of infectious peritonitis was one episode every 35 patient months, all peritoneal dialysis methods combined. Gram positive bacteria were as follows: 5 Staphylococcus Epidermidis, 3 Staphylococcus Aureus, 2 Streptococcus and 1 Bacillus Cereus. In nine cases it was a gram negative bacillus: 4 E. Coli, 2 Moraxella, 1 Pantoea spp, 1 Pseudomonas, and 1 Citrobacter. Finally, in the last four cases the peritonitis was sterile.

Peritoneal infection led to the termination of PD (with transfer to hemodialysis) in two cases: patient 2 in Figure 1 - infection with Staphylococcus Epidermidis and patient 10 in Figure 1 - infection with Bacillus Cereus. Peritoneal infection led to the death of one patient (patient 4 in Figure 1 - multi-resistant E. Coli infection). This patient had the sixth peritonitis, four of which were gram negative (2 E. Coli and 2 Moraxella).

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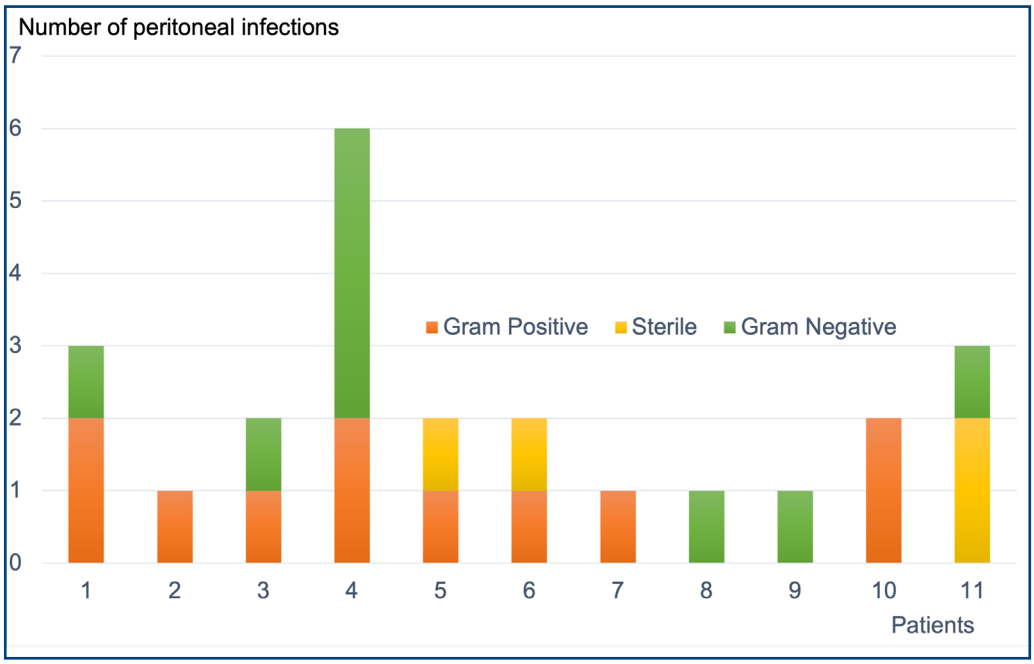

Figure 1. Peritoneal infections in eleven patients.

3. Fate of patients; Renal transplantation and transfers in hemodialysis

The fate of all 30 ADPKD patients is shown in Table 3. Fifteen patients (50%) underwent renal transplantation in a relatively short time, less than two years after starting PD for the majority of patients (Figure 2). Only one out of 30 patients required nephrectomy to be prepared for surgical transplantation, with no change in dialysis method.

Seven patients (23.3%) were transferred to hemodialysis. Figure 3 represents the duration time on PD treatment in months (from 11 to 71 months) for patients transferred to hemodialysis and the causes of transfer (inadequate dialysis and/or loss of ultrafiltration, peritoneal infections and dysfunction of the PD catheter). The median duration of PD treatment was 36 months for these seven patients.

Finally, four ADPKD patients are currently being treated (7, 8, 11 and 34 months) with PD. One of these four patients underwent kidney embolization in preparation for the kidney transplant.

Table III. Fate of 30 patients (on PD)

Renal transplantation	15 patients	50.0%
Previous nephrectomy	1 patient	
Hemodialysis transfers	7 patients	23.3%
Median duration on PD	36 months	
Death	4 patients	13.3%
Mean age	62 years	
Mean duration on PD	53 months	
Related to peritoneal infection	1 case	
Currently on PD	4 patients	13.3%

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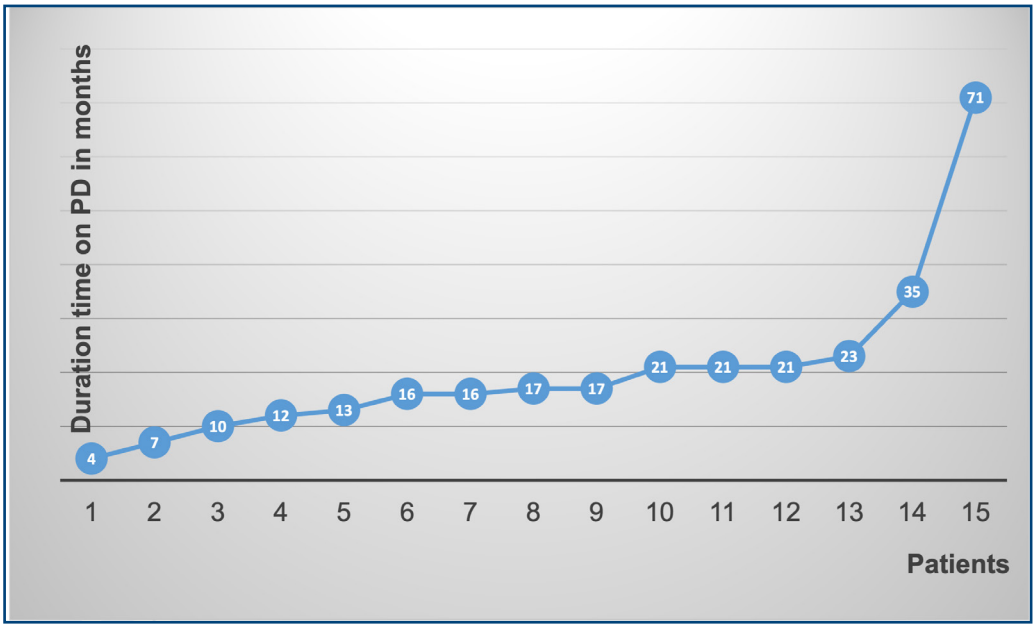

Figure 2. Renal transplantation in fifteen patients.

Causes of transfers to hemodialysis	Patients
1. Inadequate dialysis and/or loss of ultrafiltration	2, 5, 6, 7
2. Peritoneal infections	3, 4
3. Catheter dysfunction	1

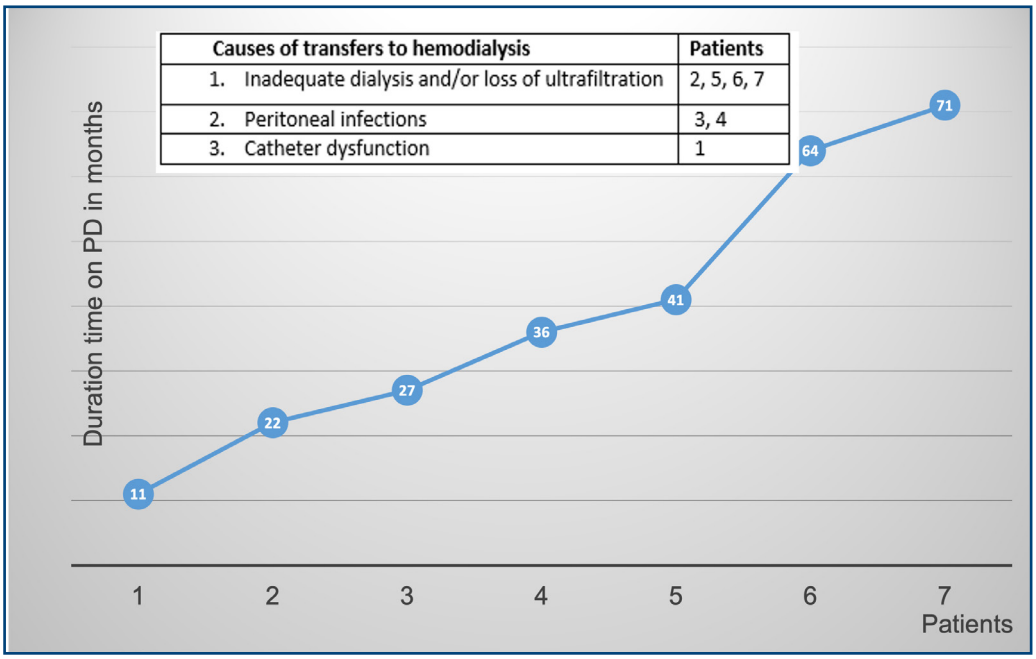

Figure 3. Transfers to hemodialysis in seven patients.

DISCUSSION

The results of our monocentric retrospective study involving 30 patients are favorable for the use of peritoneal dialysis in ADPKD patients, with possible selection of ADPKD patients according to the size of the kidneys when choosing a modality dialysis. We compared our experience with the results of the two French dialysis registries, the REIN network for all dialysis patients and the RDPLF for home dialysis (years 2000 - 2010), which were published jointly in September

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2017 [1], and those of two recent meta-analyses grouping, respectively, 9 and 12 studies worldwide [5,6], mostly retrospective and often monocentric. These two meta-analyses were primarily concerned with patient survival and that of the dialysis technique as well as peritoneal infections, all PD methods combined. Access to transplantation is studied in the first meta-analysis.

The age of commencing dialysis in our cohort corresponds to that described in the literature [4]. The frequency of liver damage (85%) is also similar to that described in the literature [2]. The majority of patients were autonomous and treated on APD.

Half of the patients received a kidney transplant in a relatively short time. Only one patient required surgical nephrectomy prior to kidney transplantation. Renal volume measurements are not available in our study. This information is not available in any of the studies referenced in the two recent meta-analyses on PD. It cannot be excluded that the renal volume (low volume), in our center, oriented the choice of the dialysis method towards PD. Note that a French study involving 60 ADPKD patients observed that intraperitoneal pressure was not influenced by renal volume but was dependent on the body mass index [7]. In our population it turns out that only three out of 25 patients had a BMI > 30.

For patients who have not been able to benefit from a kidney transplant, transfer to hemodialysis appears on average after three years of treatment with PD (median 36 months). The causes of transfer are represented by catheter dysfunction in only one case, inadequate dialysis and/or loss of ultrafiltration in four cases, and peritoneal infection in two cases. Four patients are undergoing PD treatment.

The first publication concerning PD (CAPD) as a dialysis method in 26 ADPKD patients dates back to 1998 [8]. Recent data from French registries show that only 11% of ADPKD patients are treated with PD, which probably reflects some reluctance of nephrologists to use this dialysis technique in this kidney disease. It is important to remember that PD remains globally a victim of "brakes" since the percentage of patients (with ADPKD or not) treated with PD has remained <10% over the years [9]. In French registries, PD management of ADPKD patients is associated with a better probability of access to renal transplantation [1]. The two meta-analyses published in PD [5,6] grouping, respectively, 882 and 931 ADPKD patients show similar results in favor of PD management of these patients, in particular for survival (better in the event of ADPKD) and survival of the dialysis method, which is not different from the control groups (non-ADPKD patients). Better survival of ADPKD patients in PD is also observed in French dialysis registries [1]. The better survival of ADPKD patients with PD, observed in all the studies, is most likely explained by a younger age during dialysis treatment and the associated low comorbidities. It should be noted that in these two meta-analyses, the French experience (data from the RDPLF) represents an important part of the results since it includes 344 ADPKD patients [10].

Most of our patients are self-sufficient on peritoneal dialysis and we have opted for nocturnal APD as much as possible (just over 75% of them) given the potential risks of increased intraperitoneal pressure. This does not exclude, in our experience, the possibility of doing CAPD. APD also allows for relatively young patient population to have a professional life that has little impact on dialysis, but also a better social life. If we exclude the first publication from 1998 [8], information concerning APD is available in seven publications [6]. APD coverage varies between 7% in China and 54.9% in France [10]. In the most recent studies (2015-2016) APD represents 42 to

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45% of patients.

The frequency of peritoneal infections in our cohort of 30 ADPKD patients is calculated, all PD methods combined, as one episode every 35 patient months. This peritoneal infection rate agree with the most recent international recommendations [11]. Gram-negative peritoneal infections do not appear to be predominant. Gram-negative bacilli make up just over 37% of our cases. Peritoneal infection remains a severe complication since it was the cause of a patient's death. In our center, the rate of peritonitis varies from year to year between an episode every 31 to 35 patient months and the frequency of gram-negative bacillus infections is estimated at 25% in non-ADPKD patients (unpublished data). In the two meta-analyses [5,6] the frequency of peritonitis was no greater in the population of ADPKD patients than in the population without ADPKD.

The issue of nephrectomy is not addressed in these two meta-analyses. One of our patients underwent nephrectomy before kidney transplantation and another underwent kidney embolization for transplantation program; the dialysis method did not change for either patient. Very little information is available on the management of nephrectomy in patients with PD. A recent French study examined the number of nephrectomies in a cohort of 24 ADPKD patients treated with PD. Six of these 24 patients underwent a pre-kidney transplant nephrectomy. Five patients were able to be maintained on PD postoperatively [12].

Finally, it should be remembered that two other recent meta-analyses [13,14], grouping together more than 7,500 ADPKD patients, were interested in comparing the mortality of patients treated with PD and hemodialysis. It turns out that the mortality does not differ between the two dialysis techniques. Logically, hemodialysis appears to be associated with increased incidence of cystic renal bleeding.

The limitations of the studies available on PD in this hereditary kidney disease are significant. First of all, the number of studies related to this dialysis technique is small. The number of ADPKD patients included in these studies is also often quite low: fewer than 40 patients in six of 12 studies were included in the two meta-analyses. As in our study, the data in the literature available are mainly retrospective, some taken from national dialysis registries. The majority of them, like ours, have come from mono-centric studies possibly involving patient selection biases such as taking into account the volume of the kidneys, or infectious cystic or digestive diverticular complications. Finally, it clearly appears that the better survival of ADPKD patients with PD is explained by a younger age at the time of dialysis treatment and low comorbidities, in particular diabetes.

CONCLUSIONS

This retrospective study tends to confirm that PD, especially the nocturnal APD technique, is a good treatment option (patient survival, technical survival, incidence of peritonitis) in patients with polycystic kidney disease, in particular for those awaiting renal transplantation.
Nevertheless, it cannot be excluded that the renal low volume was an important element in the decision to refer these patients to PD. The management of nephrectomy, if necessary, remains a major difficulty in the care process of these patients on PD. Renal embolization probably offers an interesting solution in this area.

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CONFLICT OF INTEREST

The authors declare no conflict of interest for this article.

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