Pediatr Nephrol (2013) 28:315–319
DOI 10.1007/s00467-012-2303-9

ORIGINAL ARTICLE

Ventriculoperitoneal shunts in children on peritoneal dialysis: a survey of the International Pediatric Peritoneal Dialysis Network

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Received: 1 May 2012 / Revised: 8 July 2012 / Accepted: 10 July 2012 / Published online: 14 September 2012

Abstract
Objective The aim of this study was to inform best evidence-based practice by collating and disseminating the experiences of members of the International Pediatric Peritoneal Dialysis Network with children having concurrent ventriculoperitoneal shunts (VPS) and peritoneal dialysis catheters (PDC).

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Methods An online questionnaire was created and distributed to all 135 centers participating in the International Pediatric Peritoneal Dialysis Network; the overall response rate was 56 %.
Results A total of 18 patients with a concurrent VPS and PDC were reported. The children were 0–12 (mean 6.8) years old at the time of placement of the second indwelling device (PDC or VPS). In 15 cases, the PDC was inserted post-VPS. On average, the two catheters were present concurrently for 23 (range 1–60) months. There were 20 episodes of peritonitis observed in 11 of the 18 patients during a period of 392 months at risk, which is a peritonitis rate of 1/19.6 months. Only one patient developed both a VPS infection and an episode of peritonitis, and these events were temporally unrelated. No episodes of an ascending shunt infection or meningitis occurred in association with any episode of peritonitis, and no other complications of catheter dysfunction were described.
Conclusions The rate of peritonitis, the absence of any documented ascending or descending infections and the lack of catheter dysfunction during the period of observation suggests that the presence of, or need for, a VPS should not preclude PD as a safe option for children requiring renal replacement therapy.

Keywords Pediatric · Renal replacement therapy · Spina bifida · Hydrocephalus · Intraperitoneal pressure

Introduction

End stage renal disease (ESRD) is a rare condition in childhood, with a reported incidence of only nine per million age-related population [1]. Peritoneal dialysis (PD) remains the preferred treatment modality worldwide for children requiring chronic dialysis support [2]. That being said, there are a number of absolute or relative contraindications against the use of PD which include the presence of a large omphalocoele, gastroschisis, bladder extrophy, diaphragmatic hernia, obliterated peritoneal cavity and known or suspected peritoneal membrane failure [2]. Little experience exists with the performance of PD in children with a ventriculoperitoneal shunt (VPS), resulting in a lack of evidence upon which to base practice recommendations [3–9].

The rarity of children with both a PD catheter (PDC) and VPS may be due to pediatric dialysis centers assuming that the presence of a foreign body in the peritoneal cavity is a contraindication to both safe and effective PD. It is our impression that such centers are more concerned with the perceived risks of ascending infection or shunt malfunction when both devices are in situ concurrently.

In order to enhance the limited information on the topic currently available in the literature, the objective of this study was to collect the experiences of members of the International Pediatric Peritoneal Dialysis Network (IPPN) pertaining to children having concurrent VPS and chronic PDC in order to address the likelihood of shunt dysfunction or the risk of ascending (meningitis from peritonitis) or descending (meningitis to peritonitis) infections.

Materials and methods

A questionnaire (Table 1) was developed and distributed twice via an online survey tool (Survey Monkey™) through an email link to all 135 centers participating in the IPPN from April–May 2011. All centers that responded were requested to answer as many questions as possible and to provide their contact information to facilitate follow-up on the cases identified. A follow-up request for additional information was made to all centers that responded that they had cared for one or more children with a concurrent VPS and PDC.

Statistics

Only descriptive statistics are reported due to the small number of patients.

Results

Of the 75 centers responding to the online questionnaire (56 % response rate), 13 (17 %) reported having provided

Table 1 Questionnaire distributed to all centers participating in the International Pediatric Peritoneal Dialysis Network

1. Age commencing peritoneal dialysis?
2. Cause of renal failure?
3. Reason for ventriculoperitoneal shunt insertion?
4. Ventriculoperitoneal shunt placed before, after or with peritoneal dialysis catheter?
5. Patients current treatment status:
   (a) Remains on peritoneal dialysis
   (b) Renal transplant
   (c) Switched to hemodialysis due to ventriculoperitoneal shunt ventriculoperitoneal shunt related problem
   (d) Switched to hemodialysis due to peritoneal dialysis related problem
   (e) Dead
6. Any peritonitis episodes while ventriculoperitoneal shunt and peritoneal dialysis catheter in place?
7. Number peritonitis episodes with ventriculoperitoneal shunt in situ?
8. Any shunt infection/meningitis while ventriculoperitoneal shunt and peritoneal dialysis catheter in situ?
9. Did the meningitis/shunt infection occur in temporal relationship with a peritonitis episode?
10. Was the organism the same, if YES please identify?
11. Any urinary/intestinal stomata present?

care for 18 patients with both a functional VPS and PDC in situ. This represents 0.6 % of all patients enrolled in the IPPN registry at the time of data collection. At the time of the data collection, three patients of the 460 enrolled in the 75 IPPN centers who responded had concurrent VPS and PDC for a point prevalence of 1 in 153 patients (personal communication, F. Schaefer, IPPN data). The characteristics and demographics of the patients are provided in Table 2. Of the 18 patients reported with concurrent VPS and PDC, 15 (83 %) had the PDC inserted post-VPS placement. On average, patients had both catheters present for 23 (range 1–60) months.

Eleven of the 18 patients, ten of whom had PDC placement following VPS, developed a total of 20 episodes of peritonitis, with a maximum of four per any individual patient, during a total of 392 patient-months at risk. The calculated peritonitis rate for this time period was 1/19.6 months. Table 3 presents the organisms isolated during each of the episodes of peritonitis.

No ascending VPS infections (meningitis) occurred during or immediately following an episode of peritonitis. In one patient who had a PDC placed subsequent to their VPS, a single remote and isolated episode of meningitis developed during the presence of both catheters without evidence for extension into the peritoneal cavity/peritonitis.

Although four patients were transferred from PD to hemodialysis (HD), the PDC malfunction in three patients and the infection in the remaining patient that prompted transfer were unrelated to a VPS-related infection or malfunction. Of these four patients, one transferred to HD for only 4 months then recommenced PD, one received a renal transplant, one died and one remains on HD at the time of this report.

Table 2 Patient characteristics

Patient characteristics	Data
Centers Reporting (*n*)	13/135
Patients Identified (*n*)	18
Male gender (*n*)	7
Mean age at PDC insertion, years [range]	6.8 [0–12]
Primary dialysis modality[a]	CAPD (7)
	CCPD (11)
Urinary/intestinal stomata[a]	Mitrofanoff (4)
	Vesicostomy (1)
	None (13)
Patient status at time of report[a]	Recovery of renal function (2)
	Transplanted (6)
	Converted to hemodialysis (1)
	Remains on PD (3)
	Death (6)
Cause of renal failure[a]	Neuropathic bladder (9)
	Renal dysplasia (3)
	Recurrent urinary tract infections[b] (2)
	Hemolytic uremic syndrome (1)
	Nephronopthisis (1)
	Membranoproliferative glomerulonephritis (1)
	Cortical necrosis (1)
Need for VPS due to[a]:	Myelomenigocoele (11)
	Congenital hydrocephalus (3)
	Benign Intracranial hypertension (1)
	Cerebral venous thrombosis (1)
	Cerebral edema (1)
	Subdural hematoma (1)

PDC, Peritoneal dialysis catheter; CAPD, continuous ambulatory peritoneal dialysis; CCPD, continuous cycling peritoneal dialysis; VPS, ventriculoperitoneal shunt

[a] Number of patients given in parenthesis

[b] Secondary to complex urological malformations

Discussion

Despite an early report from Warady et al. in 1990 of two patients who successfully performed chronic PD in the presence of a VPS [5], there seems to be great reluctance in both the Pediatric Dialysis and Pediatric Neurosurgery communities to adopt this practice. In fact, while the long-term survival of spina bifida patients is increasing [8], and renal damage and the potential need for renal replacement therapy in this population is not entirely uncommon, the published reports of PD use in these patients remain exceedingly rare. Unpublished data from the IPPN would seem to confirm the rarity of these patients, with an estimated prevalence of only one in 153 children (0.6 %) on PD having a concurrent VPS (personal communication, F. Schaefer).

While only anecdotal evidence can be offered to support this statement, we believe that the major reason for the

Table 3 Microorganisms isolated during episodes of peritonitis from patients with concurrent VPS and PDC

Organisms cultured (peritonitis)	*n* (%)
Negative	5 (25)
Streptococcus salivarius	1 (5)
Staphylococcus epidermidis	2 (10)
Staphylococcus aureus	5 (25)
Seratia marcecens	1 (5)
Escherichia coli	2 (10)
Streptococcus viridans	2 (10)
Pseudomonas aeruginosa	1 (5)
Proteus spp.	1 (5)

VPS, ventriculoperitoneal shunt; PDC, peritoneal dialysis catheter

apparent avoidance of the concurrent use of both VPS and PDC is due to nephrologist, surgeon and potentially family anxieties regarding the perceived risk of developing either an ascending or descending infection along the VPS and/or VPS dysfunction.

With respect to the risk of ascending or descending infections, it is difficult to assign a true risk of such an event due to a paucity of published cases, thus limiting evidence-based decisions as to the suitability of PD as an option for children with, or needing, a VPS. The published literature on children with concurrent and functional VPS and PDC is extremely small, with only 11 such patients previously described in the literature [3–9], plus two others referenced by Grunberg [9]. As expected, given the complex nature of such patients, many complications occurred in these children, and a number of the described cases had multiple episodes of peritonitis (including fungal). Nevertheless, there were no reported occurrences of meningitis/ascending infections, and only one proven [4] and one suspected [6] concurrent VPS infection requiring externalization and shunt replacement during the presence of both catheters.

We report a peritonitis rate of 1/19.6 months among children with both a VPS and PDC in situ. This is comparable to the rate of 1/18.8 months in the 2011 annual report of the North American Pediatric Renal Trials and Collaborative Studies (NAPRTCS) [10] and a rate of 1/ 21.6 months among children who have a PDC in situ, based upon unpublished data obtained following a dedicated search of the IPPN registry (personal communication, F. Schaefer). As such, the infection risk would not in itself appear to be a contraindication to PD in this select patient group. Most importantly, the absence of any episodes of meningitis in both the patients reported here as well as those reported previously who experienced peritonitis is reassuring, although not conclusive proof, that the risk of an ascending ventriculitis or meningitis is extremely small.

The concern regarding VPS dysfunction when patients are concurrently on PD is reasonable given the recognized increase in intraperitoneal pressure (IPP) seen in children on PD and its possible effect on cerebrospinal fluid (CSF) drainage through the VPS. Current VPS devices have either a static or adjustable one-way valve, which allows for drainage of CSF from the ventricles to the peritoneal cavity based on the pressure differential between the two cavities. In the situation of a supine patient, this pressure differential is dependent on volume/pressure relationships in the two cavities, whereas in the upright or seated patient the effect of gravity on the CSF column increases the effective pressure gradient and facilitates drainage of the CSF. A recent study by Avery et al. examined the CSF pressures in nearly 200 children felt to be 'normal' at the time of lumbar puncture [11]. From this cohort the authors defined the overall mean opening pressure to be 19.8 ± 6.8 cmH_20, with the 10th and 90th percentiles at 11.5 and 28 cmH_2O, respectively. Fischbach and Warady's work on IPP measurements in children >2 years of age and on PD demonstrate that when the dwell volumes fall within the generally accepted range of 1,000–1,400 ml/m^2, one can expect to see IPPs of between 7 and 14 cmH_20 and maximal IPP, in terms of patient tolerability, occur in the range of 18 cmH_2O [12]. Taking these numbers together, and recognizing that most children currently perform automated cycler dialysis in the supine position overnight and are generally filled with less than 1,000 ml/m^2 for any day dwell, it seems very unlikely that the IPP would ever persistently exceed the CSF pressure and inhibit VPS function. However, it should be noted that—to the best of our knowledge—this presumption has never been formally investigated and proven to be accurate.

There are a number of important limitations to this study. Firstly, data was obtained from a voluntary electronic survey, and we cannot be certain that in the 44 % of centers that did not respond there were in fact no cases. We also asked for centers to describe any current or past patients they remembered caring for with both VPS and PDC in situ; thus, we need to allow for the possibility of under-reporting and for skewing of the reported outcomes due to the non-prospective nature of the survey. Nevertheless, we believe that the need to dialyze children as described here is rare and that even in the larger centers each such case is memorable. Likewise, if such a child were to suffer complications as serious as meningitis or peritonitis during the presence of the two catheters, it is our feeling that this would be remembered as well.

In conclusion, based on the results from previous small case series as well as the contribution of this experience from the IPPN, we propose that pediatric nephrology and neurosurgery programs consider the concurrent use of a VPS and PDC to be a safe and acceptable option in the rare child requiring dialysis and a cerebral fluid shunt. At present, there is no evidence to support an increased risk of peritonitis, ascending ventricular infections, or shunt dysfunction in these patients. The continued collection of data on this unique patient population by registries such as the IPPN should be encouraged so as to further inform the pediatric nephrology community about the risks and benefits associated with this rare chronic care scenario.

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