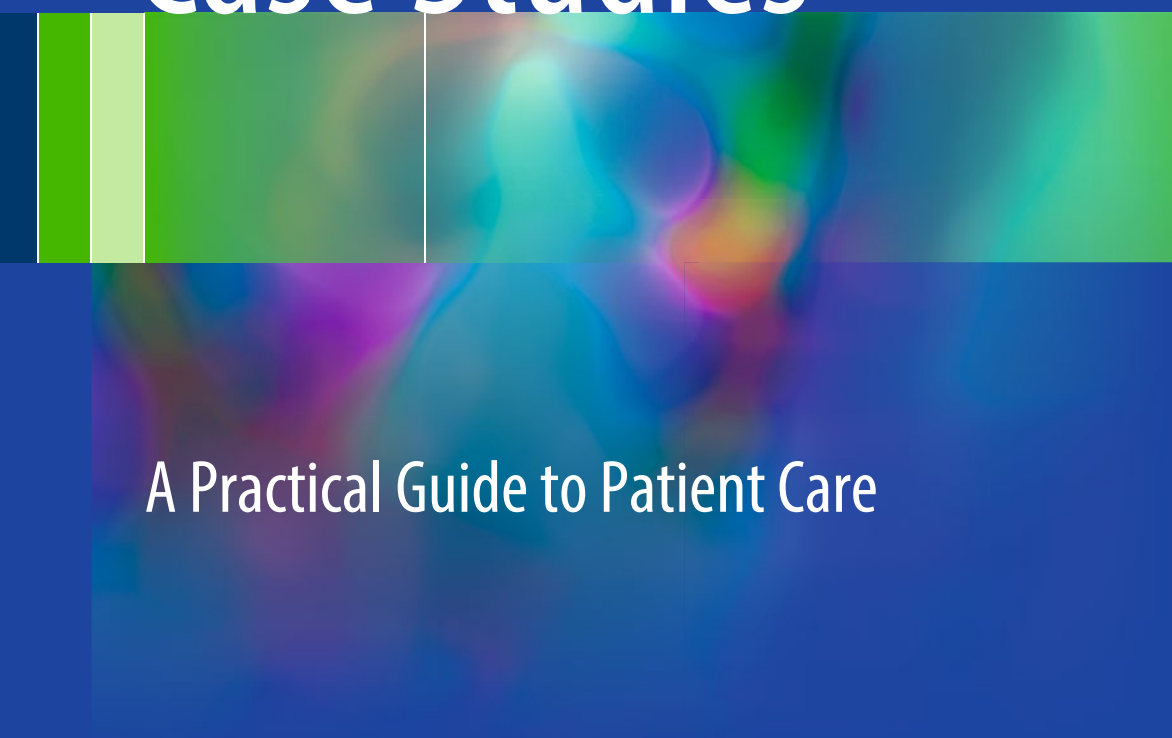
Bradley A. Warady · Franz Schaefer
Steven R. Alexander *Editors*

Pediatric Dialysis Case Studies

A Practical Guide to Patient Care



Springer

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Preface

The management of chronic dialysis therapy in children is a complex, all-consuming, and ultimately rewarding discipline. As members of a small, yet international subspecialty, pediatric dialysis practitioners have learned to turn to one another for clinical advice when faced with difficult clinical problems. It is, in fact, the clinical experience of our colleagues that continues to be a truly invaluable resource. Whereas bedside rounds remain the "gold standard" of clinically based instruction, writing about clinical situations using real-life cases employs the same teaching technique by directly applying clinical management principles at the patient level.

The worldwide success of *Pediatric Dialysis* and its second edition, which remain the only textbooks available that are entirely devoted to dialysis in children, led us to consider a companion text in which cases would be used to reinforce the material contained in those publications. To that end, we have had the great pleasure of working with a team of international experts in pediatric dialysis care to develop the book *Pediatric Dialysis Case Studies*. In this unique text, each chapter is introduced by a case presentation that serves as the basis for key learning points that are clinically applicable and presented in a succinct manner by authors who have a wealth of knowledge and clinical expertise. Whereas some chapters address frequently noted complications with evidence-based recommendations for prevention and treatment, other chapters highlight less common events and provide unique perspectives on disease management. The topics that we have included in *Pediatric Dialysis Case Studies* cover virtually all aspects of pediatric dialysis care and, in turn, represent the efforts of individuals with firsthand clinical expertise in virtually every discipline that is represented in the pediatric dialysis team.

As was the case in the development of the first and second editions of *Pediatric Dialysis*, the goal of this book is to create a resource that is worthy of a place on the bookshelves of all busy dialysis clinicians and would be frequently consulted for some "bedside" expertise. In *Pediatric Dialysis Case Studies*, we believe that we have achieved that goal.

We conclude by sincerely thanking our co-authors, all of whom are experts in their own right, and all of our patients and their families who are our best teachers. We also want to thank Springer developmental editor Michael Wilt for his patient

guidance and unfailing support. Together, we have created what we believe is a special textbook about clinical challenges associated with pediatric dialysis that we all have encountered or will likely encounter in the future. Most importantly, we hope that this textbook becomes a resource that will lead to improved outcomes for the sometimes complex, but always special, patients we care for.

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Chapter 1
Peritoneal Access

Richard J. Hendrickson and Walter S. Andrews

Case Presentation

A previously healthy one-year-old male presented to the emergency room with a several day history of decreased oral intake, cough, and decreased urine output. His past medical history was significant for poor oral intake with failure to thrive being evaluated for surgical gastrostomy placement. He had no past surgical history. After a comprehensive evaluation and initial fluid resuscitation in the emergency room, he was admitted to the pediatric intensive care unit with dehydration and septic shock. He was intubated for respiratory insufficiency and diagnosed with rhino/enterovirus infection. He remained hemodynamically stable after initial fluid resuscitation. His initial laboratory values demonstrated an elevated creatinine of 8.0 mg/dL. His electrolytes were maintained within normal ranges with medical therapy.

Despite aggressive fluid resuscitation, he remained anuric, consistent with acute kidney injury (AKI). Due to persistent anuria and worsening anasarca over the next 24 h, nephrology was consulted for further management. Despite continued maximal medical therapy, his renal function did not improve, and a surgical consult was obtained for dialysis access. At our center, we prefer peritoneal dialysis (PD) as the initial mode for both acute and chronic dialysis. If urgent dialysis is needed, hemodialysis may be instituted temporarily and possibly continuous renal replacement therapy (CRRT) if the patient cannot tolerate hemodialysis. Once stabilized, we convert to PD if possible.

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Once this patient was stable, he was taken to the operating room for PD catheter placement. In addition, as he had been scheduled to have a gastrostomy tube inserted in the near future, this procedure was added to PD catheter placement. In the OR, cefazolin and fluconazole were given within 60 min before incision, and the patient's abdomen was prepped. The PD catheter that was selected was a double-cuffed, curled, swan neck catheter (Argyle™, Peritoneal Dialysis Catheters, Covidien, Mansfield, MA). The catheter was sized by measuring the distance between the umbilicus and the symphysis pubis. The location of the exit site of the catheter was marked on the patient's right side, halfway between the midclavicular and anterior axillary lines and lateral and inferior to the initial incision located just lateral to the umbilicus.

The catheter was inserted laparoscopically with the curled portion positioned deep in the pelvis. No skin exit site sutures were used. A MIC-KEY (Ballard Medical Products, Draper, UT) gastrostomy tube was then inserted laparoscopically utilizing the LEFT upper quadrant access port incision, and the stomach wall was secured to the anterior abdominal wall with internal retention absorbable sutures. Catheter function and the absence of leaks were confirmed intraoperatively by using two passes of 10 cc/kg of dialysate. Low-volume dialysis was initiated postoperatively and over the next several days was increased to full volume without incident.

One month later, his renal function returned and his PD treatment was held. During this time off PD, his PD catheter was routinely inspected with regular dressing changes and flushing of the catheter.

Two months later his renal function deteriorated and he required PD treatment again. Unfortunately, the PD catheter would not flush easily and did not drain. An abdominal radiograph demonstrated the PD catheter to be in the pelvis. Therefore, a surgical consult was obtained and the patient was taken to the operating room for laparoscopic evaluation where intraluminal fibrin plugs were identified and removed. His PD catheter continues to function well, and the patient is undergoing evaluation for kidney transplantation.

Clinical Questions

1. What are the available modalities for pediatric dialysis?
2. When PD is the preferred method, what PD catheters are available for pediatric patients?
3. What antibiotics are routinely used for PD catheter insertion or revision?
4. How are the PD catheters surgically inserted?
5. What are the options if a PD catheter does not drain satisfactorily?

Diagnostic Discussion

1. In acute situations, hemodialysis or PD may be used to help stabilize the patient who requires fluid and toxin removal. If it is apparent that the patient will require chronic treatment, PD is the preferred modality in our center.
2. There are various types of pediatric PD catheters available (Tenckhoff single and dual cuff, Tenckhoff curl catheter, Tenckhoff Swan Neck, Tenckhoff Swan Neck curl catheter) which come in various lengths. We currently prefer Argyle catheters (Argyle™, Peritoneal Dialysis Catheters. Covidien. Mansfield, MA) (Fig. 1.1). We routinely use the dual cuff Swan Neck curl catheter for both acute (e.g. patients who have Hemolytic Uremic Syndrome) and chronic dialysis situations.

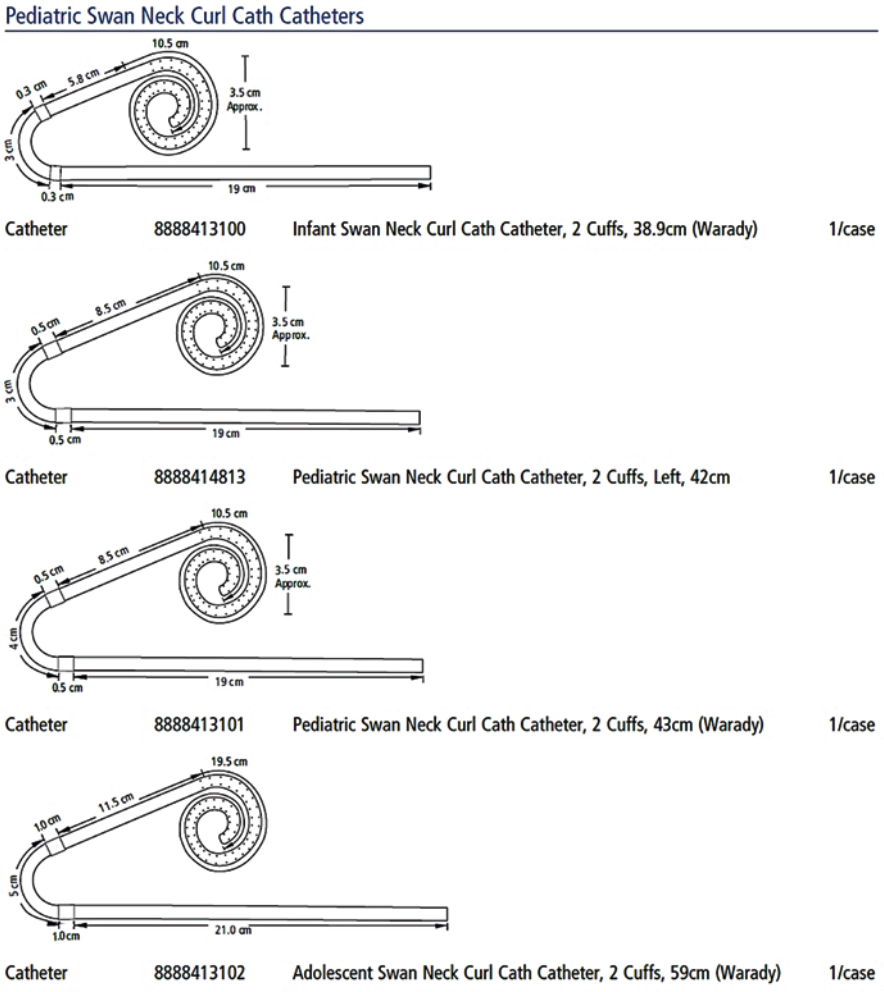

Fig. 1.1 Various types of PD catheters available for pediatric patients (Image copyright © 2016 Medtronic. All rights reserved. Used with the permission of Medtronic)

3. When a PD catheter is inserted or revised, we routinely use perioperative administration of a first-generation cephalosporin such as cefazolin, per ISPD guideline 2.2 [1]. Vancomycin is an alternative in patients hypersensitive to cephalosporins. Of note, if a gastrostomy tube will be placed simultaneously, an antifungal agent such as fluconazole is administered perioperatively as well, per ISPD guideline 7.4 [1].
4. We routinely gain access to the peritoneum via the umbilicus with a 5 mm STEP (STEP™ Instruments, Medtronic, Covidien. Mansfield, MA) port and establish a pneumoperitoneum of 12–15 mm Hg with carbon dioxide. Laparoscopy allows for a complete inspection of the peritoneal cavity for any pathology and to also identify patent internal inguinal rings that should be repaired prior to initiation of PD treatment to avoid development of inguinal hernias. Preexisting inguinal hernias can be repaired at this point via an open or laparoscopic technique at the discretion of the surgeon.

   An additional 5 mm port is placed in the LEFT upper quadrant to help facilitate visualization, as the camera can be switched between these two ports. Additionally, a 5 mm instrument can be utilized with the additional port.

   Next, we perform an omentectomy in an attempt to prevent the omentum from clogging the side holes of the PD catheter. The available omentum is retrieved via the umbilicus and sequentially ligated with Vicryl ties and electrocautery (Fig. 1.2). We are reluctant to perform an omentopexy (i.e., fixation of the omentum to the anterior abdominal wall) for fear of a potential midgut volvulus [2].

   Next, we select the appropriate length dual cuff swan neck curl catheter based upon the patient's size. The ideal catheter length should be approximated by placing the internal cuff lateral and 1 cm above the umbilicus and then measuring to the level of the symphysis pubis. We usually place the exit site on the RIGHT side of the abdomen, since some of these patients may need a gastrostomy tube in the LEFT upper quadrant.

   After marking out the anticipated catheter track on the skin of the abdomen, an incision is made in the skin to the RIGHT of midline, and the dissection is carried down to the anterior fascia of the rectus muscle where a transverse incision is made. A purse-string monofilament suture is placed in the anterior fascia. Next, a STEP access needle and sheath are inserted through this opening and carefully advanced to the preperitoneal space with laparoscopic guidance. The sheath is tilted and carefully advanced in the preperitoneal space downward toward the pelvis where it is allowed to enter the peritoneum just above the dome of the bladder (Fig. 1.3). Once fully deployed within the peritoneal cavity, the needle is removed and the sheath left in place.

   Ultimately, a peel-away introducer sheath set (Peel Away® Intoducer Set. Cook Medical; Bloomington, IN) is inserted, starting with placment of the included guidewire through the STEP sheath and advancing it into the pelvis under direct visualization. The STEP sheath is removed and the dilator and sheath advanced over the guidewire down into the pelvis (Fig. 1.4). The guidewire and dilator are removed leaving the 20 French sheath down in the pelvis.

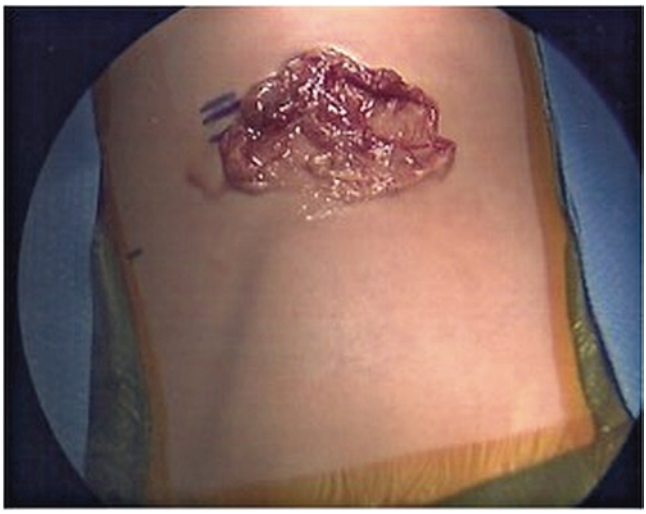

Fig. 1.2 Omentum retrieved via the umbilicus. Pen markings show anticipated medial incision to the RIGHT of the umbilicus and the more LATERAL and DOWNWARD incision for the EXIT site

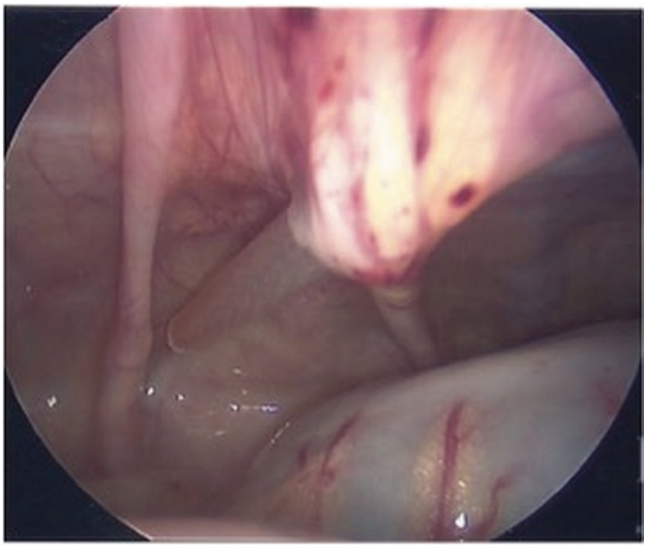

Fig. 1.3 STEP needle and sheath in a preperitoneal tunnel entering the deep pelvis above the dome of the bladder

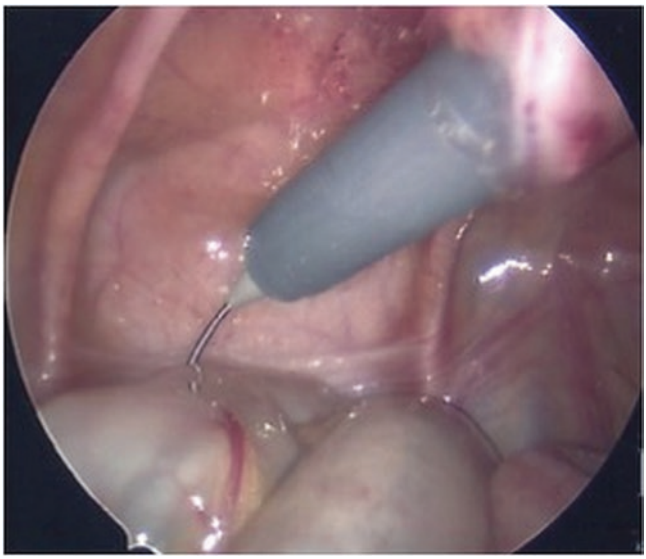

Fig. 1.4 Peel-away sheath with dilator and guide wire down deep in the pelvis

Next, the PD catheter is inserted and advanced through the peel-away sheath. When the first cuff is under the anterior fascia level, the PD catheter is usually curled within the pelvis. The peel-away sheath is then removed while holding the cuff with a pair of forceps below the level of the anterior rectus sheath. Once the sheath is removed, the cuff will remain below the rectus muscle. If needed, a 5 mm instrument can be used to steer the curl end of the PD catheter into an optimal position (Fig. 1.5). Laparoscopy is used to confirm that the radiopaque stripe is not twisted and that the inner cuff has not migrated into the peritoneum.

Next, a second incision is made out LATERAL and INFERIOR to the initial incision so that the second cuff will be positioned within the tunnel about 2 cm from the exit site, and the catheter is facing in a DOWNARD position. The catheter is then tunneled from the first or medial incision, out toward the DOWNWARD and LATERAL incision, ensuring the radiopaque strip is not twisted (Fig. 1.6).

We routinely have a dialysis nurse in the operating room to flush our newly placed PD catheter. We watch the peritoneum fill and then drain under direct laparoscopic visualization. A dwell and drain are also performed without insufflation to ensure proper function.

Once good flow is documented, the anterior fascia purse string suture is tied around the catheter to help anchor the cuff under the anterior rectus sheath to prevent leakage. The subcutaneous tissue is closed in layers followed by skin closure. The LEFT upper quadrant 5 mm incision is closed as well to ensure no leakage. Of note, this incision can be used for a gastrostomy tube placement if needed, as described in this case. The umbilical port is removed last, and the

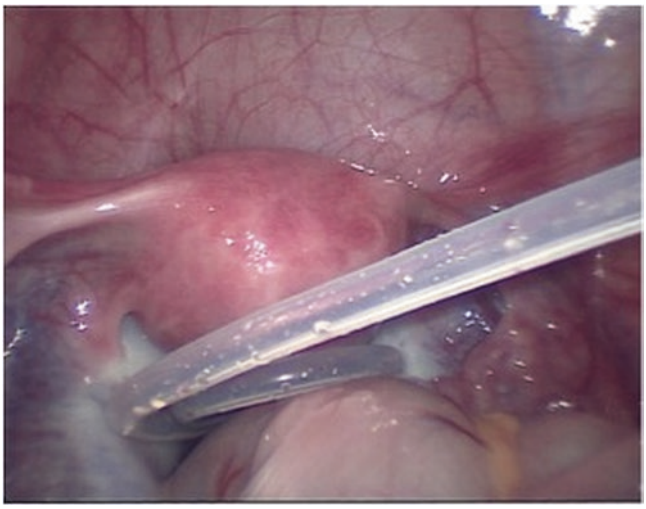

Fig. 1.5 PD catheter curl in satisfactory position deep in the pelvis

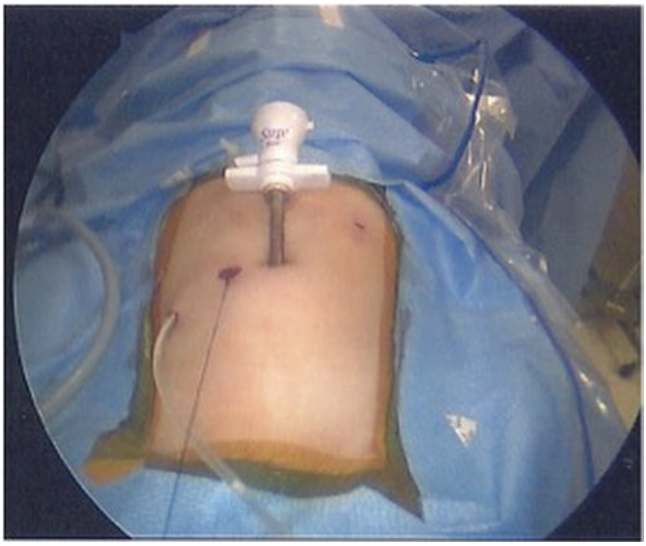

Fig. 1.6 PD catheter EXIT site is LATERAL and DOWNWARD facing. The RIGHT medial incision has the anterior fascia purse string and a 5 mm STEP port within the umbilicus. Also note the 5 mm incision in the LEFT upper quadrant where a 5 mm STEP port was utilized

fascia and skin closed in layers. NO sutures are placed at the PD catheter exit site.

Once the sterile drapes are removed, the PD catheter exit site is dressed by the dialysis nurse.

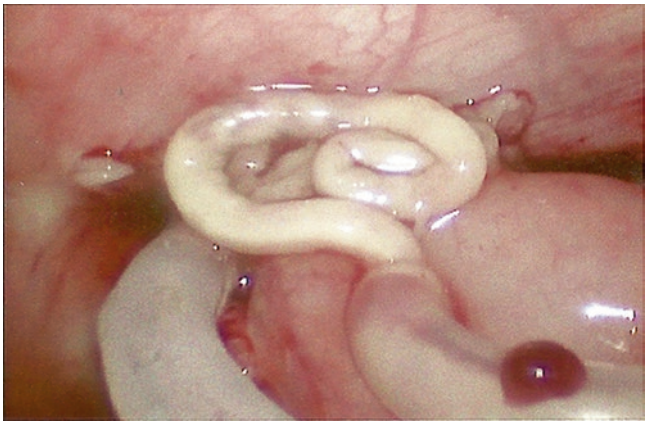

Fig. 1.7 Intraluminal fibrin plug occluding the PD catheter

5. If a peritoneal catheter is not functioning after all medical attempts to reestablish PD catheter function (i.e., flushing, positioning) have failed, a laparoscopic revision is warranted. Entrance is gained via the umbilicus, and an additional 5 mm port can be placed in the LEFT upper quadrant to help facilitate the operation. If the catheter has "flipped" out of the pelvis, it can be repositioned and tested for function. If the catheter has been encased in a fibrous peel, it can be dissected free, repositioned, and tested. If the internal lumen of the catheter is clogged (Fig. 1.7), it can be grasped via the umbilical incision and externalized to help remove the debris; note the camera will need to be in the LEFT upper quadrant port for this maneuver. If none of these maneuvers help restore function, or if the patient has outgrown the initial PD catheter, then removal and replacement can be performed as described above.

Clinical Pearls

1. Laparoscopy via the umbilicus allows for excellent peritoneal visualization for PD catheter insertion, avoiding injury to surrounding structures such as the intestines and bladder. It also allows visualization of the internal rings to ensure they are closed. If the internal rings are patent, they can be repaired with either an open or laparoscopic approach.
2. Omentectomy is routinely performed to help prevent clogging of the side holes within the catheter. Omentopexy, which is described in adults, is not our common practice for fear of a midgut volvulus.
3. STEP needle and sheath tunneling along the posterior sheath allows for deeper placement of the guide wire and sheath for the PD catheter.

4. Creation of a preperitoneal tunnel from the anterior fascia incision to the midline cephalad to the bladder helps prevent catheter dislodgement.
5. Utilization of an additional 5 mm port in the left upper quadrant helps facilitate proper catheter placement into the pelvis and omental removal. This incision can also be used for laparoscopic placement of a gastrostomy tube if needed.
6. Intraoperative testing of the peritoneal dialysis catheter with the dialysis nursing staff is very beneficial to ensure satisfactory dwell and drainage prior to leaving the operating room.
7. If PD catheters are not used for treatment, weekly inspection, and regular dressing changes and catheter flushing will help ensure continued catheter patency.

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Chapter 2
Peritoneal Equilibration Testing and Application

Francisco J. Cano

Case Presentation

FW, a recently diagnosed patient with CKD Stage 5, is a 6-year-old boy who has been recommended to initiate chronic dialysis. His primary renal disorder is renal dysplasia. His nutritional evaluation reveals a weight of 18.1 kg (SDS −1.08), height 102 cm (SDS −2.64), and BSA 0.8 m^2. His residual renal Kt/V is 0.3. A predialysis biochemical evaluation showed BUN 70 mg/dl, creatinine 6.5 mg/dl, hemoglobin 9.4 g/dl, serum calcium 9.2 mg/dl, phosphorus 7.7 mg/dl, PTH 580 pg/ml, $25(OH)D_3$ 14.5 ng/ml, and serum albumin 3.8 g/L; electrolytes were Na 138 meq/L, K 5.4 meq/L, Cl 101 meq/L, and serum CO_2 19.2 meq/L. Echocardiography showed a left ventricular mass index (LVMI) value of 45 $g/m^{2.7}$.

Peritoneal dialysis (PD) was initiated several weeks after PD catheter placement, with the fill volume reaching 700 ml/exchange (900 ml/m^2) 3 weeks after dialysis initiation. The PD modality used was continuous ambulatory peritoneal dialysis (CAPD), and FW's initial dialysis prescription consisted of Dianeal® 1.5%, four exchanges per day, with each exchange lasting 6 h. During the second month of PD, a 4-h peritoneal equilibration test (PET) was performed.

During the night prior to the test, an 800 ml (1,100 ml/m^2) exchange of 2.5% dextrose dialysis solution was instilled for 8 h. On the day of the test, the overnight exchange was drained, and another exchange with Dianeal 2.5% was infused. Dialysate samples for creatinine and glucose were obtained at 0, 2, and 4 h of dwell time, and a blood sample for creatinine was obtained at 2 h. The 4-h results were as follows:

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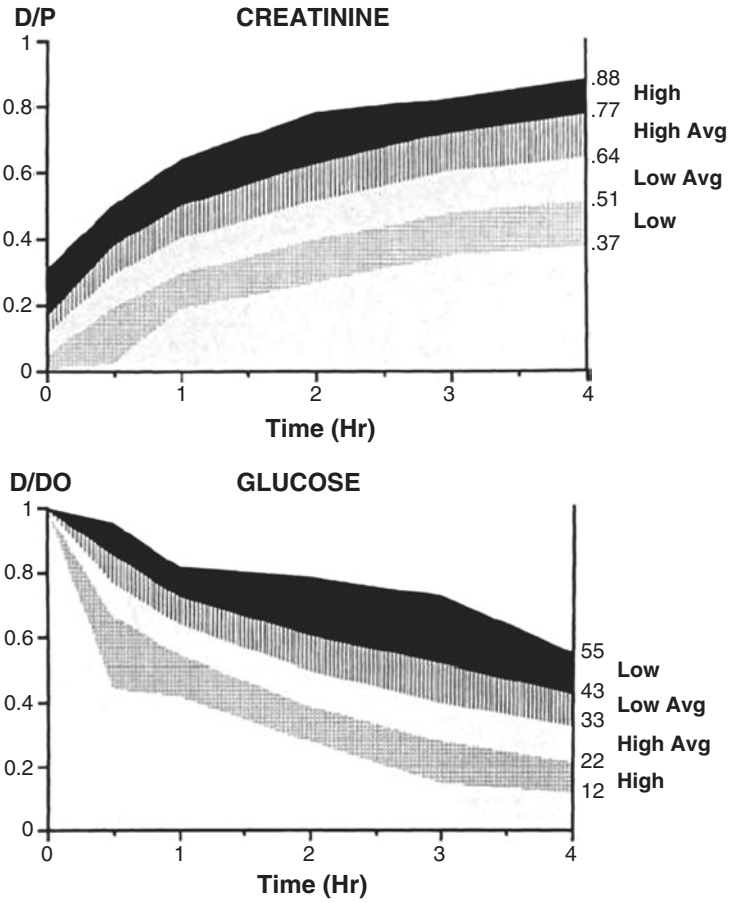

Fig. 2.1 Peritoneal equilibration test categories

D/P creatinine, 0.64, and D/D_0 glucose, 0.38. These results were compatible with a high-average transporter status (Fig. 2.1).

In view of these PET results, the PD modality was changed to nocturnal intermittent PD (NIPD). The prescription consisted of seven, 1-h exchanges nightly, with an 800 ml fill volume using Dianeal 1.5% peritoneal dialysis solution. Over the initial 18 months of PD, the patient experienced a single episode of peritonitis with a good response to antibiotic treatment. The PET was not repeated after this peritonitis episode.

After 2 years of PD, the patient's blood pressure was 110/76 mmHg (95th percentile), and the residual renal Kt/V decreased to a value of 0.2. Echocardiography demonstrated an increased LVMI with a value of 54 $g/m^{2.7}$. As a result of the clinical evidence of hypervolemia and the desire to provide the best PD prescription for both

solute and fluid management, a repeat PET was performed. The treating physician chose not to conduct a short PET. Results showed a 4-h D/P creatinine of 0.45 and a 4-h D/Do glucose of 0.58, findings now compatible with a low transporter status. Based on this result, FW had his PD prescription changed to a long dwell PD schedule, specifically the use of CAPD with a 1,000-ml fill volume and four, 6-h exchanges daily.

Clinical Questions

1. Is the PET a useful tool in pediatric peritoneal dialysis?
2. What is the importance of the duration of the exchange preceding the PET?
3. What is the importance of the fill volume in the PET?
4. How should the results of the PET be used to help select the PD modality and prescription?
5. Are both the Short PET and the Classical PET appropriate for use in children?
6. When should the PET be repeated?

Diagnostic Discussion

1. The success of peritoneal dialysis therapy is based on the ability of the peritoneal membrane to serve as a semipermeable membrane for solute transport and ultrafiltration. The properties of this membrane are also key determinants of the patient's outcome [1–4].

   The peritoneal equilibration test (PET) represents a semiquantitative means to assess the peritoneal membrane permeability in dialyzed patients, and the resultant data aids in the individualized prescription of peritoneal dialysis therapy. In pediatrics, a considerable experience with the PET has been accumulated during the past 20 years [4, 5]. The PET helps tailor the PD prescription to meet the specific needs of the patient in terms of

   (a) Fill volume
   (b) Length of each exchange
   (c) Number of daily cycles
   (d) Dextrose concentration of peritoneal dialysis solution

   - *The PET is performed in children in the following manner*:

     1. An overnight 3–8h exchange is performed.
     2. The overnight exchange is drained upon arrival to the PD unit the following morning.
     3. A transfer Y-type set is installed.
     4. A 1,100-ml/m_2 fill volume, 2.5% glucose peritoneal dialysis solution is infused, and patient is rolled from side to side during the infusion.