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Monika Szłapka-Polok[1], Marta Dworak[1], Elżbieta Trembecka-Dubel[1], Dagmara Roszkowska-Bjanid[1], Maria Szczepańska[2]

[1]Department of Pediatric Nephrology, Pediatric Dialysis Division, Public Clinical Hospital no. 1 in Zabrze, Zabrze, Poland
[2]Department of Pediatrics, Faculty of Medical Sciences in Zabrze, Medical University of Silesia in Katowice, Katowice, Poland

Peritoneal dialysis in a newborn with complex defects of the urinary and gastrointestinal tract

Abstract

Peritoneal dialysis is the method of choice in the treatment of chronic kidney disease in young children. Renal replacement therapy aims to achieve proper development of the child and, if possible, to quickly prepare the patient for kidney transplantation. The decision to start peritoneal dialysis is made on a case-by-case basis; the main criterion is GFR dropping below 15 mL/min/1.73 m^2. The authors describe a case of a boy with chronic kidney disease CKD in whom peritoneal dialysis was performed starting from the neonatal period and present the associated challenges and complications.

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Key words: children, renal replacement therapy, peritoneal dialysis, complications of dialysis therapy, nutrition

INTRODUCTION

The most common causes of chronic kidney disease (CKD) in children are congenital anomalies of the kidney and urinary tract (CAKUT) and genetically determined conditions such as structural kidney diseases [1]. The primary indication for the initiation of chronic renal replacement treatment is the glomerular filtration rate dropping below 15–10 mL/min/1.73 m^2, which is characteristic for stage 5 CKD. The decision regarding earlier initiation of renal replacement therapy is further influenced by psychomotor retardation (particularly lack of weight gain), difficulties in compensation of metabolic acidosis, arterial hypertension, and profound ionic-profile disorders, including mineral and bone metabolism disorders [2].

Data reported in the available registry studies usually pertain to infants under 2 years of age, with the reported prevalence ranging from 7 to 12 cases per 1 million individuals in the age-matched population. Thus, neonatal CKD requiring renal replacement therapy in this early period of life should be considered an ultra rare disease [3].

Neonatal CKD requiring renal replacement therapy in this early period of life should be considered an ultra rare disease

Peritoneal dialysis is the main method of dialysis in the youngest children and is usually chosen as the first treatment option [4]. It is a continuous, technically simple procedure that ensures regular, slow dehydration and detoxification of the body. It can be carried out at home and at night while the baby is asleep. Pediatric patients may benefit from a less restrictive diet than that required in hemodialysis. In addition, peritoneal dialysis has a better impact on the maintenance of the patient's own diuresis. It also allows the child to be prepared for the renal transplantation procedure. An additional indication for peritoneal dialysis is the difficulty to establish permanent vascular access as required for hemodialysis in small children due to the small size of the blood ves-

Address for correspondence:
Maria Szczepańska
Oddział Nefrologii Dzieci z Pododdziałem Dializoterapii Dzieci
Samodzielny Publiczny Szpital Kliniczny nr 1
Śląski Uniwersytet Medyczny
ul. 3 Maja 13–15, 41–800 Zabrze, Poland
phone: +48 32 370 43 05
e-mail: mszczepanska@szpital.zabrze.pl

sels. This also facilitates "saving" of the vessels for possible future hemodialysis. At the same time, the available hemodialysis equipment is mostly suitable for the treatment of children weighing more than 8–10 kg [2]. Data from the ESPN/ERA-EDTA registry covering a total of 1063 infants aged 12 months or less, in whom dialysis treatment had been initiated between 1991 and 2013, indicated that children treated by peritoneal dialysis were more likely to present with congenital anomalies of the kidney and urinary tract (CAKUT, 48% vs. 27%), while infants treated by hemodialysis were more likely to present with metabolic disorders (12% vs. 4%) [5].

On the other hand, however, peritoneal dialysis therapy is associated with the risk of complications, the most serious of which include dialysis-related peritonitis and infection of the external outlet of the catheter lumen. Neonates and infants are particularly vulnerable to these infections due to the proximity of diapers or the presence of gastrostomies/vesicostomies [6]. Other complications include the formation of hernias, protein/energy malnutrition (the lack of appetite, the loss of proteins in the dialysis fluid), anemia, hypercoagulability, lipid disorders, and premature atherosclerosis [7, 8].

Another major problem is that the efficacy of the method decreases after 2–3 years of therapy due to the peritoneal remodeling which leads to peritoneal sclerotization, and further to polyhydramnios, particularly in patients without residual diuresis [9].

When starting peritoneal dialysis in neonates and infants, the primary requirement is to use a special pediatric peritoneal catheter, as well as automatic dialysis equipment (cyclers) with appropriate pediatric software facilitating the delivery of small inlet volumes of the dialysis fluid. Automated dialysis can be applied only starting with inlet volumes of 80–100 mL. In the youngest children (body weight < 10 kg), lower inlet volumes, corresponding to 10 ml/kg body weight, are used together with shorter dwell times, more frequent exchanges, and longer procedure times compared to those used in older children. Biocompatible fluids with neutral pH and the lowest glucose concentrations are also recommended. Calcium levels should be adjusted on a case-by-case basis depending on the parameters of the calcium-phosphate metabolism [2].

In this article, the authors report a case of a boy with CKD in whom peritoneal dialysis was performed starting from the neonatal period and present the associated challenges and complications.

CASE REPORT

The boy, currently aged 12 months, born with a number of congenital malformations, received peritoneal dialysis from the first days of his life and remained under the care of the Department of Pediatric Nephrology with the Pediatric Dialysis Division.

The obstetric history was complicated, including the suspected presence of a posterior urethral valve due to oligohydramnios and giant urinary bladder observed in prenatal imaging examinations. Amnioinfusions had been used in the treatment starting from gestation week 32. Human immunodeficiency virus (HIV) infection, hepatitis B and C, toxoplasmosis, and syphilis were excluded.

The boy, born from second gestation and second childbirth with vaginal delivery, presented with a bodyweight of 2,700 g and an Apgar score of 2/3/5. Due to a respiratory failure, the child had been intubated in the delivery room. Deviations diagnosed upon physical examination after birth included *caput succedaneum*, deformations of limbs and chest, skin bruising, prune belly (underdevelopment of abdominal muscles and cryptorchidism), and anal atresia. Abdominal ultrasound revealed a polycystic right kidney (sized 3.9 × 1.7 cm, the largest of the cyst with the diameter of 1.6 cm), blurred corticomedullary differentiation of the left kidney (sized 3.1 × 1.4 cm), and urinary bladder filled with a small quantity of urine.

The child was transferred to the neonatal intensive care unit (NICU) for further treatment. Additional radiological examination performed at the NICU revealed a non-aerated rectum and abnormal structure of sacral vertebrae with suspicion of vertebral clefts. In addition, a cranial ultrasound scan revealed subependymal cysts within the lateral ventricle horns, while a consulting cardiologist diagnosed the child with congenital heart malformation manifested by a type II atrial septal defect and moderate features of gradually resolving pulmonary hypertension. The boy was qualified for surgical treatment with prophylactic antibiotic therapy, with cystostomy and cystostomy and artificial anus on the descendingt colon formation. Intraoperative findings included intestinal malrotation and sigmoid stenosis.

▶▶Neonates and infants are particularly vulnerable to these infections due to the proximity of diapers or the presence of gastrostomies/vesicostomies◀◀

Table 1. Manual exchanges, diuresis, and arterial blood pressure values

Age of the patient	Number of exchanges/ /inlet volume	Duration of dialysis	Dwell time	Ultrafiltration	Diuresis	Blood pressure [mm Hg]
1st month of life	24 × 30–60 mL Manual exchanges	Continuous dialysis (24 h)	45 min	30–200 mL	3.2–4.8 mL/kg/h	97/67 83/45
2nd month of life	24 × 60–80 mL Manual exchanges	Continuous dialysis (24 h)	45 min	70–200 mL	Mannitol/furosemide/ /theophylline mix- -stimulated diuresis	100/78 75/43
3rd month of life	24 × 80 mL Manual exchanges	Continuous dialysis (24 h)	45 min	250– 3aa00 mL	Mannitol/furosemide/ /theophylline mix- -stimulated diuresis	74/42 98/59

Oliguria and dynamic expansion of edema were observed starting from the first days of life. The management involved diuresis being stimulated with furosemide followed by the mannitol/furosemide/theophylline mix, with no satisfactory outcomes. Despite the treatment, only small quantities of urine were collected, and the creatinine levels were increasing. In addition, metabolic acidosis and electrolyte disorders persisted, initially in the form of hyperkalemia, followed by hypokalemia, and hyponatremia. Due to the rise in inflammatory markers, early infection was diagnosed, and antibiotic therapy was modified, with creatinine and vancomycin levels being monitored. In spite of the delivered treatment, signs of renal failure were increasing in intensity, with the child being transferred, on the tenth day of his life, to a treatment facility capable of delivering chronic renal replacement therapy.

Upon being transferred to the NICU and then to the pediatric intensive care unit within our institution, the neonate was in grave general condition, intubated and mechanically ventilated, with jaundice, abdominal bloating, with hepatomegaly and inaudible peristalsis, generalized edema, as well as all the aforementioned features conditions. Laboratory investigations revealed elevated inflammatory markers and renal function indices, hypoproteinemia with hypoalbuminemia, hypertriglyceridemia, cholestasis, slightly disturbed coagulation profile parameters, moderate anemia, metabolic acidosis, and dyselectrolytemia. Treatment at the department included continued mechanical ventilation, antibiotic therapy, protein and albumin deficiency compensation, electrolyte disturbances and acidosis management, intensive stimulation of diuresis, as well as several platelet and red blood cell product transfusions. Due to the persistent edema and increasing creatinine concentrations (max. 548 μmol/L), a decision to start peritoneal dialysis was made on the 14th day of the patient's life.

After the parents' consent was obtained following the indications for renal replacement therapy, and the risks and potential adverse effects of peritoneal dialysis were presented to the patient's parents, a Tenckhoff catheter was implanted under general anesthesia and prophylactic antibiotic therapy.

Initially, during the first 2.5 months of the child's life, peritoneal dialysis was performed by manual exchanges due to low body weight (3,370–4,165 g). The dialysis regimen is presented in Table 1.

The treatment resulted in gradual resolution of edema and improvement in renal function parameters and respiratory efficiency. Consequently, the child was extubated and remained under non-invasive ventilation on the following days. After the inlet volume had been increased to 80 mL, peritoneal dialysis was continued using a cycler according to the regimen presented in Table 2 below.

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An accidental evacuation of the cystostomy catheter occurred on the 25th day of NICU stay. A Foley catheter was placed under ultrasound guidance through the cystostomy orifice by a consulting surgeon after numerous failed attempts at urethral catheterization.

At this stage of the treatment, hospitalization was complicated by two episodes of sepsis (*Enterococcus faecalis*, *Citrobacter freundii* ESBL) and three episodes of urinary tract infection (*Candida glabrata* and *Enterococcus faecalis*; *Stenotrophomonas maltophilia* and *Enterococcus faecium* VRE and HLAR; *Citrobacter freundii* ESBL) subjected to treatment as per antibiogram results.

At 3 months, the circulatory- and respiratory-efficient patient was finally transferred for further treatment from the NICU to the

Table 2. Outline of the cycler regimen, diuresis, and arterial blood pressure values

Age of the patient	Number of exchanges/ Inlet volume	Cycler time setting	Dwell time	Ultrafiltration	Diuresis	Blood pressure [mm Hg]
3rd month of life	18 × 80 mL 21 × 80 mL 20 × 80 mL 19 × 85 mL 19 × 85 mL 19 × 90 mL 18 × 90 mL	24 h 24 h 22 h 22 h 21 h 21 h 20 h	1.09 h 52 min 52 min 52 min 50 min 50 min 50 min	250–200 mL 200–330 mL	Trace	64/39 (< 90 c)
4th month of life	18 × 90 mL 17 × 95 mL 17 × 100 mL 17 × 100 mL	20 h 19 h 19 h 18 h	50 min 50 min 50 min 47 min	 205–305 mL	Trace	62/36 (< 90 c)
5th month of life	17 × 100 mL 16 × 110 mL 15 × 110 mL	18 h 17 h 16 h	47 min 47 min 48 min	200–200 mL 150–350 mL	Trace — 60 mL	95/57
6th month of life	15 × 110 mL	16 h	58 min	168–256 mL	Trace	94/75
7th month of life	15 × 110 mL	16 h	58 min	200–260 mL	5–10 mL	97/53
8th month of life	12 × 120 mL 12 × 130 mL	13 h 13 h	1 h 3 min 1 h 3 min	Approx. 200 mL 100–160 mL	220–280 mL 200–280 mL	80/42
9th month of life	12 × 130 mL 12 × 140 mL	13 h 13 h	1 h 3 min 1 h 3 min	110–120 mL 123–260 mL	170–180 mL 10–170 mL	79/49 108/86
10th month of life	12 × 140 mL	13 h	1 h 3 min	113–230 mL	Trace — 10 mL	93/78 116/93
11th month of life	12 × 140 mL	13 h	1 h 3 min	130–230 mL	5–10 mL	81/59 87/70

Department of Pediatric Nephrology with the Pediatric Dialysis Division.

At the Department, the range of diagnostic examinations of the urinary tract was extended to include voiding cystourethrography, which revealed the abnormal shape of the bladder and left-sided vesicoureteral reflux with left ureter dilatation (Figure 1).

During hospitalization, gastroenterological consultation was carried out due to the increased levels of hepatic transaminases and gamma-glutamyltranspeptidase (GGTP), most likely due to long-term antibiotic therapy. Poor weight gain was also noticeable. At the first stage of the treatment period, the child received complete parenteral nutrition via a Broviac-type catheter, which was subsequently followed by Bebilon pepti MCT whey hydrolysate administered by naso-gastric tube. Oral feeding was started at the 6th week of life. Starting from week 7, the diet was modified due to hyperphosphatemia, and the patient was nourished with Renastart — a specialized, high-energy preparation with low protein, phosphorus, potassium, calcium, chloride, and vitamin A content. An attempt was also made to increase the calorific content of meals by adding cereals, e.g. Sinlac, to each portion of modified milk. The weight gain, as observed in the patient, is presented in Figure 2.

The stay at the Department was complicated by central catheter infection (*Klebsiella pneumoniae*) with accompanying high inflammatory parameters. The infection was treated as per antibiogram results. Consequently, a decision to surgically install a Babyport-type V-port was made, with the procedure being complicated by local hematoma due to spontaneous needle withdrawal.

Rehabilitation efforts were continued throughout the child's stay at the hospital.

Following clinical improvement, normalization of inflammatory markers, and positive results of maternal training in home-based peritoneal dialysis, the patient was discharged home in the 5th month of his life with recommendations for outpatient renal replacement therapy and frequent follow-up visits at our Department (1 × week to 1 × 2 weeks).

At the age of 7 months, the boy returned to our Department with suspected dialysis-related peritonitis. The history included fever on

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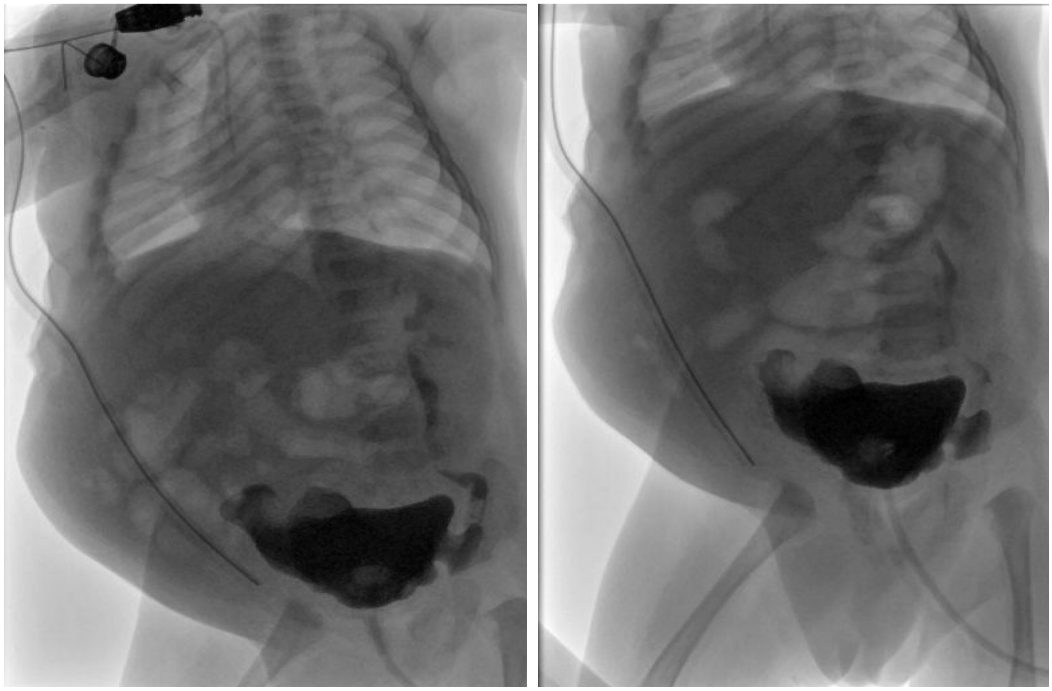

Figure 1. Voiding cystourethrography. Tenckhoff catheter tubing can be seen on the right, as well as the VascuPort chamber within the right subclavian area

Chronic peritoneal dialysis in the youngest children requires special skills and collaboration within a multidisciplinary team

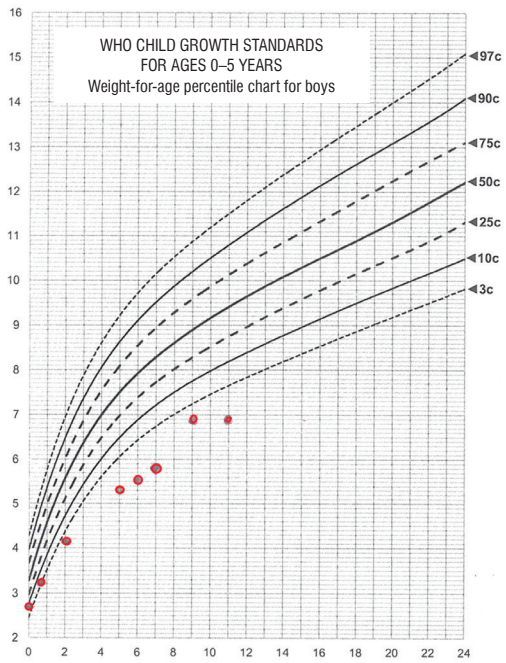

Figure 2. Body weight: results of regular monthly measurements; WHO — World Health Organizations

the day before readmission, loss of appetite, and very high levels of inflammatory markers. The initial diagnosis was confirmed, and an additional urinary tract infection (pyuria, bacteriuria — *Stenotrophomonas maltophilia*) was diagnosed on the basis of clinical presentation and the results of the additional investigations (significantly elevated levels of C-reactive protein and procalcitonin, as well as dialysate pleocytosis). The treatment was based on empirical broad-spectrum antibiotic therapy administered intravenously and intraperitoneally with additional antifungal prophylaxis. The patient required an emergency transfusion of red blood cells due to anemia. The peritoneal dialysis regimen was modified on an as-needed basis, with the extension tube being replaced after the treatment.

During hospitalization, the patient was entered into the transplant register waiting list. Qualification for a kidney transplant was temporarily delayed until the body weight would exceed 8 kg.

At present, the patient remains in outpatient care and requires frequent follow-up visits at our Department. Figure 3 presents the current anatomical status of abdominal wall.

DISCUSSION

Chronic peritoneal dialysis in the youngest children requires special skills and collaboration within a multidisciplinary team. Renal replacement therapy aims to achieve proper development of the child, and quickly prepare the patient for kidney transplantation [3, 9]. Appropriate and comprehensive treatment of infants with renal insufficiency usually allows

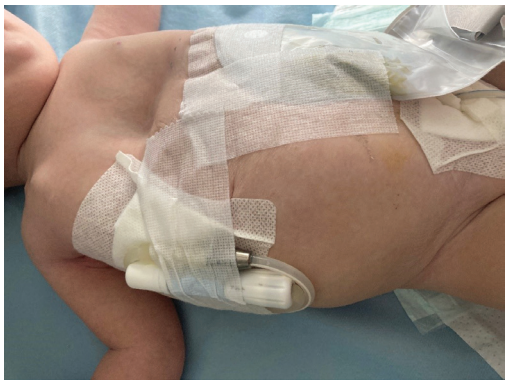

Figure 3. A photograph of abdominal integuments with peritoneal catheter, stoma bag, and cystostomy

the weight of 9 to 10 kilograms to be achieved before the patient is qualified for kidney transplant in the second year of their life. However, each case should be considered on an individual basis, with the potential presence of aggravating factors such as lung hypoplasia, oliguria, or total absence of residual kidney function, as well as the presence of comorbid defects/conditions [2, 10]. Numerous babies requiring peritoneal dialysis after birth are simultaneously struggling with other organ dysfunctions, as in the case of the patient described in this report. Ultrasound examination may facilitate prenatal diagnosis of renal diseases in most neonates born with CKD, particularly in cases of oligohydramnios [1]. Also in the presented case, the diagnosis had been made on the basis of prenatal examinations.

An important aspect associated with chronic kidney disease in the youngest patients consists of extensive nutritional treatment, involving increased calorific intake, and selection of an additional route for administration of nutrients (gastrostomy, gastric tube) [4, 11]. Adequate nutrition requires considerable effort from caretakers; frequently, its burden is greater than that of dialysis therapy. Follow-up visits are initially held every 1–2 weeks, depending on the child's health status; the intervals may be then gradually increased. Monitoring includes anthropometric measurements, assessments of patients' hydration status, verification of laboratory test outcomes, and possible modifications of drug dosages.

A multicenter Polish study presented by Jander et al. revealed that the average weight standard deviation score (SDS) at the start of the peritoneal dialysis was –2.0, as compared to –1.7 at the age of 1 year, as well as that 40% of the analyzed infants were adequately nourished at the age of 1. The authors have also demonstrated that the treatment of infants with renal impairment remains a challenge for the medical team and carers, but the overall results are encouraging [12]. In their study carried out on the data from the International Pediatric Peritoneal Dialysis Network (IPPN) Registry, Schaefer et al. showed that the most important single factor associated with higher body mass index (BMI) values, both at the initiation of dialysis therapy and during long-term follow-up, consisted of the presence of gastrostomy [11].

Due to concomitant congenital defects, the mortality rate in neonates and infants requiring dialysis at the age of less than one month is much higher than in older infants, albeit the percentage of patients in whom the kidney function is restored is also much higher [9].

The case of the neonatal patient described above shows that despite a complex defect within the gastrointestinal tract and recent surgery on abdominal wall, it is possible to continue peritoneal dialysis for several months in a chronic setting. Dialysis therapy of neonates and small infants remains a challenge for healthcare professionals and the patient's family. Nonetheless, efforts should be made to provide this treatment because literature data suggest that remote prognosis in patients receiving dialysis therapy and subsequent kidney transplantation rate was similar to those observed in older children [3, 13].

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