

CASE REPORT

Challenges in the management of an anuric child with end-stage renal disease since the neonatal period: a case report

Joanna Bieniek, Katarzyna Zachwieja, Monika Miklaszewska, Dorota Drożdż

Department of Pediatric Nephrology and Hypertension, Jagiellonian University Medical College, Krakow, Poland

ABSTRACT

End-stage renal disease requiring renal replacement therapy (RRT) since the neonatal period is a very rare condition. Peritoneal dialysis is the primary RRT in infancy. The case report describes a patient with congenital renal failure and anuria, abdominal wall laxity, small atrial septal defect, cryptorchidism, but without pulmonary hypoplasia or other disqualifying severe congenital defects. The patient had been dialysed since the fourth day of his life, until he received a kidney transplant at the age of 3.5 years. During RRT there were numerous complications related to dialysis catheter, infections, poor nutrition, and electrolyte abnormalities. It is a medical challenge to dialyse anuric children. Success requires a multidisciplinary team working towards the child's proper growth as well as giving them a chance for a good quality of life.

KEY WORDS:

child, peritoneal dialysis, peritonitis, Tenckhoff catheter.

INTRODUCTION

End-stage renal disease (ESRD) receiving paediatric renal replacement therapy (RRT) is a very rare condition in children and occurs with a frequency of 5.5 cases per million children aged 0–14 years, according to the European ESPN/ERA-EDTA registry. Congenital anomalies of the kidney and urinary tract (CAKUT) are the most common cause (41%) of ESRD in children [1]. In recent years, the number of children initiating dialysis therapy in the first year of their lives has clearly increased, accounting for about 11% of the paediatric population receiving RRT [2, 3]. However, only 264 patients from 32 countries started RRT between 2000 and 2011 in the neonatal period [4]. Peritoneal dialysis (PD) is the main method of RRT in the youngest age group. In newborns, PD is chosen as the first method in approximately 90% [4].

An early kidney transplant is difficult to receive because of patient weight limitations [5]. During the 5-year follow-up of newborns starting dialysis, 54% received kidney transplants at a median age of 25 months [4]. Optimal care for newborns without any residual diuresis is particularly difficult. Important treatment targets are maintaining water and electrolyte balance, nutritional management, and prevention of infection [6].

CASE REPORT

The patient was admitted to the neonatal intensive care unit in the first 24 hours of his life due to left-sided pneumothorax. The boy was born in the natural way as the fourth childbirth with normal birth weight (3300 g) and with 6 and 7 points on Apgar score within the first and fifth minute, respectively. He was classified as a full-

ADDRESS FOR CORRESPONDENCE:

Joanna Bieniek, Department of Pediatric Nephrology and Hypertension, Jagiellonian University Medical College, 265 Wielicka St., 30-663 Krakow, Poland, e-mail: avalkosz@o2.pl

TABLE 1. Results of selected laboratory tests

Patient's age	Urea [mmol/l]	Creatinine [μmol/l]	K ⁺ [mmol/l]	Na ⁺ [mmol/l]	Ca ²⁺ [mmol/l]	Mg ²⁺ [mmol/l]	P [mmol/l]	Hgb [g/l]	pH	HCO ₃ ⁻ [mmol/l]	PTH [pg/ml]
1 day	1.9	101.3	4.63	136	2.32	0.83		20.0	7.23	24.0	
2 days	5.8	291.9	6.35	128					7.25	21.4	
4 days	16.4	456.6	6.70	104	2.21	0.74		20.8	7.29	16.2	
Starting dialysis											
7 days	19.2	384.9	3.88	109	1.18	16.1	7.34	18.6			
14 days	16.6	371.3	5.47	113	1.90	0.63	1.73	14.0	7.38	19.1	
30 days	11.9	302.4	4.80	117	1.16	0.48	2.67	8.7	7.35	17.5	13.1
3 months	9.0	253.5	3.31	135	2.61	1.04	1.51	10.1	7.39	24.8	47.5
6 months	10.9	480.9	4.10	134	2.01	0.75	1.45	11.3	7.47	27.1	
1 year	13.1	435.4	4.71	130	2.00	0.68	1.01	9.0	7.46	29.3	764.9
2 years	7.8	392.1	6.06	141	2.28	1.21	1.18	11.5	7.51	24.9	868.2
3 years	14.9	508.6	6.00	135	2.51	0.70	1.35	12.2	7.43	23.9	1316.4
3.5 years	17.1	530.4	5.30	135	2.43	0.92	1.34	8.3	7.46	32.3	
Kidney transplantation											
5.5 years	9.4	44.4	5.33	141	2.45	0.81	1.73	11.6	7.33	17.8	

term newborn, but the pregnancy was not controlled. After decompression of the pneumothorax, he was respiratorily efficient. The first laboratory tests showed a high serum creatinine concentration. The child did not urinate. On ultrasound, both kidneys were small, cystic, and dysplastic, and the urinary bladder was empty. Despite pharmacological treatment (furosemide), diuresis was not accomplished. For the further survival of the child, it was necessary to implement RRT. Together with the parents we decided to start PD. At that time, he was the youngest child who had started RRT in our hospital.

DIALYSIS CATHETER

Tenckhoff peritoneal catheter insertion was performed on the fourth day of the patient's life. PD was started immediately due to persistent anuria, increasing parameters of renal failure, electrolyte disturbances, and metabolic acidosis (Table 1). From the beginning, there were problems with pericatheter leakage of dialysis fluid and good healing in the flaccid abdominal wall and sealing the catheter insertion site. These problems repeated after almost every PD catheter insertion and made effective dialysis impossible (Table 2).

INFECTIONS

The first hospitalisation of the patient lasted continuously for 17 months. The clinical course was complicated by many infections (Table 3). He was treated with multiple courses of parenteral and intraperitoneal antibiotics. Chronic significant deficiency of IgG was found. Immunoglobulin infusions were administered ad hoc during severe infections. Finally, at the end of 2 years of age, the boy was qualified for regular immunoglobulin substitution every 4-6 weeks. During each serious infection the patient's development was stopped and the patient's development and proper weight gain was delayed (Figure 1).

NUTRITION

The adequate nutrition of the boy was a challenge from the beginning. As a newborn, he was fed parenterally (an additional central line was needed), then enteral feeding by a nasogastric tube was introduced. His milk formula was changed many times. Eventually, an elemental diet based on the amino-acid formula was introduced due to the appearance of rectal bleeding. The child was confirmed as having a cow's milk allergy. Despite supplying an adequate diet, it was not possible to achieve oral nutrition tolerance. Persistent vomiting was observed, which limited the amount of enteral feeding. It was necessary to carry out parenteral hyperalimentation until the child was 16 months old. Nevertheless, the child's weight at 16 months was 7.670 g. The boy was consulted by gastroenterologists many times, and additional images

TABLE 2. Complications related to dialysis catheter

Age of the patient	Reason	Type of intervention
5 day	Pericatheter leakage of dialysis fluid	Topical sealing at the exit site
9 day	Progressive pericatheter leakage of dialysis fluid	Revision of the catheter
10 months	Too short catheter – poor drainage dialysate	Catheter removal and reinsertion below
10–12 months	Increasing leakage of dialysis fluid	Few surgical attempts to obtain the tightness
12 months	Persistent leakage of dialysis fluid	Removal and reinsertion of the catheter on the other side of the abdominal wall
21 months	Persistent exit site and tunnel infections (skin necrosis and fistula over the catheter)	Catheter removal and reinsertion
34 months	Recurrent peritonitis caused by colonization by <i>Staphylococcus aureus</i> of catheter	Catheter removal and reinsertion
34 months	Leakage of dialysis fluid	Switch to hemodialysis for 10 days to allow healing and sealing at the exit site of the catheter

TABLE 3. Characteristic of patient's infections

Age (months)	Infection	Cultured pathogen
0.5	Sepsis	<i>Staphylococcus epidermidis</i> MRSE, MLS
2.5	Sepsis – respiratory failure, mechanical ventilation for 11 days	<i>Staphylococcus epidermidis</i> MRSE, MLS
3	Peritonitis	<i>Acinetobacter baumannii</i>
5	Sepsis	<i>Staphylococcus epidermidis</i> MRSE, MLS + <i>Corynebacterium</i> spp.
9	Sepsis – fulminant G-negative	<i>Enterobacter cloacae</i>
13	Sepsis	<i>Staphylococcus epidermidis</i> MRSE, MLS
15	Sepsis – significant dyspnoea	<i>Klebsiella pneumoniae</i>
16	Sepsis	<i>Staphylococcus aureus</i> MRSA
19	Peritonitis	<i>Staphylococcus aureus</i> MRSA
21	Exit-site infection	<i>Staphylococcus aureus</i> MRSA
24	Peritonitis	<i>Klebsiella pneumoniae</i>
32	Peritonitis	<i>Staphylococcus aureus</i> MRSA
33.5	Peritonitis	<i>Staphylococcus aureus</i> MRSA
35	Peritonitis	<i>Staphylococcus aureus</i> MRSA
38	Peritonitis	<i>Staphylococcus aureus</i> MRSA

like ultrasound of the abdominal cavity, intestinal passage, pH measurement, and gastroscopy were performed, which did not reveal any cause of vomiting or oral feeding intolerance. Until the age of 8 years, the patient was fed mainly through a nosogastric tube that he inserted himself. He also developed symptoms of severe atopic dermatitis.

HOMEOSTASIS

The applied PD turned out to be a very effective method of RRT for the patient. PD on manual mode administration was started at 10 ml/kg/h and gradually increased with the biocompatible and non-biocompatible dialysate solutions with glucose concentrations of 1.36%

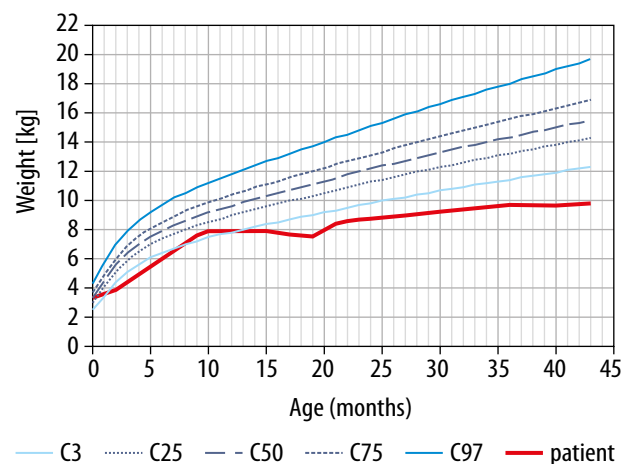
**FIGURE 1.** The weight gain shown in centile boy chart

TABLE 4. The patient's regimen of PD

Age of the patient	Number of exchanges and using of method	Inlet volume	Duration of dialysis	Dialysate solutions with glucose concentrations
4 days – start dialysis	24 manual	30 ml	Continuous dialysis (24 hours)	Physioneal 1.36%
7 days	24 manual	40 ml	24 hours	Physioneal 1.36%
30 days	12 manual	55 ml	24 hours	Physioneal 1.36%
3 months	12 manual	60 ml	24 hours	Physioneal 1.36%
6 months	8 cyclor	200 ml	16 hours	Physioneal 2.27%
1 year	8 cyclor	230 ml	16 hours	Physioneal 2.27%
2 years	6 cyclor	400 ml	15 hours	Dianeal 1.36%
3 years	7 cyclor	450 ml	13 hours	Dianeal 1.36%

or 2.27% (Table 4). After 2 years of age the patient used conventional lactate-buffered PD fluid. This was also due to better metabolic and fluid homeostasis (higher dehydration was observed when this type of dialysate fluid was implemented). There were no problems with adequacy of ultrafiltration and maintaining of acid-base balance. From the beginning he received prophylaxis of metabolic consequences of ESRD in the form of supply of calcium, preparations of vitamin D, sodium chloride, bicarbonate, iron, and erythrocyte stimulating agents. Nevertheless, severe electrolyte abnormalities were seen (episodes of hypocalcaemia, hypomagnesaemia, hyponatraemia, and hyperphosphataemia). Hemiconic seizures were observed once, probably due to hypocalcaemia (0.66 mmol/l). Electrolyte abnormalities had to be chronically corrected by intravenous infusions. During blood pressure monitoring, hypotension was recorded periodically despite an appropriate fluid status. Diagnosis of transient adrenocortical insufficiency was made, and dexamethasone and fludrocortisone were administered with good clinical response.

The patient at the age of 3 years (weight almost 10 kg) was entered on the urgent list of kidney recipients. At age 3 years and 7 months the patient received a renal allograft from a deceased donor.

Currently, 7 years after the kidney transplantation, the patient weights 26 kg (3 pc) and is 126 cm (< 3 pc) tall. He attends school, and his physical and mental development is only slightly delayed.

DISCUSSION

Conducting long-term RRT in the youngest children remains one of the greatest challenges of paediatric nephrology. Currently, most serious kidney defects are detected prenatally. Early detection enables the initiation of intrauterine therapy and early specialised treatment. In the presented case, prenatal ultrasound screening was not performed, and the serious kidney defect was revealed after birth. The decision to initiate neonatal dialysis should be taken by the parents together with the healthcare team in the best interests of the child. Short and long-term

prognosis, comorbidities, technical possibilities, and anticipated quality of life of the child as well as the family acceptance should be taken into account and discussed [3, 7]. At the time of qualifying for dialysis, our patient had no additional serious disorders diagnosed. Over time, it turned out that he had significant comorbidities, like atrial septal defect, abdominal wall laxity, cryptorchidism, cow's milk allergy, and hypogammaglobulinaemia. This high number of comorbidities influenced the management of dialysis therapy. Despite numerous older siblings and not the best material status, the parents declared their readiness to take long-term care of the child. The International Society for Peritoneal Dialysis (ISPD) guidelines recommend that PD catheters should be placed at least 2 weeks prior to the initiation of dialysis to allow healing [8]. The patient was anuric at birth, and soon a life-threatening fluid overload developed. He required urgent start of PD. American scientists showed that waiting even 3 days prior to catheter use conferred better outcomes compared to when the catheter was used immediately [9]. The need to revise the catheter in children who were initiated PD within the first year of life is more than twice as common as in the older age group [10]. In infants, dialysate leak from the catheter's exit site is the most common complication reported. The reason seems to be the delay of healing, decreased subcutaneous tissue and poor nutrition [9]. All these features were seen in our patient. Moreover, the repeated leakage of fluid seems to be particularly related to the laxity of the abdominal wall musculature (like in incomplete prune belly syndrome – type II or III described by Woodard [11]). The 5-year survival rate among children undergoing dialysis from a newborn is worse than in older children and is up to 76% according to the European registry [4] compared with a rate of 74.6% reported in the USA [12], while in older children it is around 95% [1]. The main cause of death in young children on RRT (35%) is infection [4]. The frequency of peritonitis and catheter exit-site infections (ESI) is higher in newborns than in older children. Gram-positive organisms are accounted for the greatest percentage of infections (about 39%), culture-negative peritonitis accounted for 28%, and

gram-negative bacteria accounted for 23% of peritonitis episodes [13]. In our case the *Staphylococcus aureus* MRSA was the main cultured pathogen. Factors such as younger age, no residual renal output, G-tube insertion after the catheter placement, and an upwardly directed catheter exit site have been proposed as potential risk factors associated with the development of peritonitis in infancy [4, 12, 13]. Despite maintaining standards of hygiene, our patient had a lot of infections. This prompted us to search for an immunodeficiency. Diagnosed hypogammaglobulinaemia was a possible additional cause of recurrent infections. One of the major clinical issues of an infant on dialysis is adequate nutrition for maintaining linear growth. Multiple factors contribute to oral nutrition intolerance in children with ESRD, including vomiting, gastroesophageal reflux, fluid restriction, acidosis, alteration in the gut hormonal balance, altered peristalsis, and psychosocial deprivation related or uraemia. Enteral feedings via nasogastric tubes or gastrostomy and supplemental feedings are necessary for most infants to achieve adequate nutrition, but it is not very frequently implemented [14]. In our patient, due to numerous complications, recurrent infections, persistent vomiting, and food allergy, it was not possible to improve nutrition despite adequate dialysis and enteral feedings via nasogastric tubes. He also has persistent oral nutrition intolerance. Therefore, early rehabilitation of oral feeding difficulties should be recommended. In a Polish multi-centre analysis of PD in infants, it was calculated that only 40% of the analysed infants were adequately nourished at the age of one year, and the mean standard deviation score of body weight was very low (-2.0 at the start of PD and -1.7 at the age of one year) [15]. Management of fluid status, and correction of metabolic acidosis and electrolyte abnormalities require daily adequacy prescription [16]. In neonates and infants, for optimal dialysis and fluid removal, more frequent exchanges on manual mode, lower fill volumes, and longer duration of dialysis (16–24 h) are required. The frequency of intervention can place a great burden on team workers, and hospitalisation times are long. Automated PD on a cycler is achieved when the fill volumes are greater than 100 ml [17]. In our case, despite effective dialysis, electrolytic balance was achieved only with supplementation of adrenal hormones. Recent studies confirm that most neonates and infants on early starting PD can be successfully transplanted and can have a favourable growth outcome [18]. However, the quality of life of these children is severely impaired by prolonged and frequent hospitalisations, comorbidities, and neuro-developmental delay.

DISCLOSURES

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3. Financial support and sponsorship: None.
4. Conflicts of interest: None.

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