THE OUTCOMES OF NEONATAL AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD)

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Objectives:

The frequency of ARPKD is 1 : 20000 birth. ARPKD manifested in early neonatal period is often complicated by respiratory insufficiency caused by lung hypoplasia.

Methods:

We retrospectively evaluated the results of the treatment in 8 children (5 females and 3 males) with neonatal manifestation of ARPKD who was treated in our dialysis unit in 2008-2015. 4 anuric children were admitted at 2-5 days of life, 4 with normal diuresis – on 11-49 days. All children were born prematurely, at gestation term 34-37 weeks (median 36 weeks). In all patients ARPKD was suspected at II-III trimesters of gestation, but prenatal observation wasn't carried out. Olygohydroamnion was revealed in 5 cases, Potter syndrome in 2. In 7 children mechanical ventilation was started just after birth. All patients presented with very large kidneys (10.5-13.0 cm length by ultrasonography), were edematous and their arterial blood pressure was upper 95 per centile. In 4 patients who needed renal replacement therapy continuous veno-venous hemodiafiltration (CVVHDF) was performed from the admission to the unit.

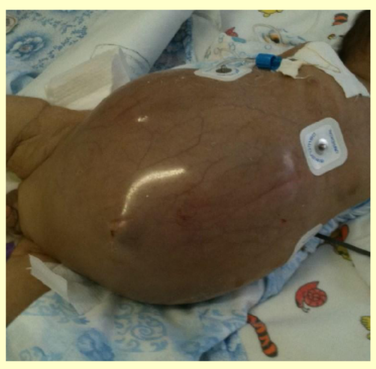

Fig.1. Newborn with enlarged abdomen.

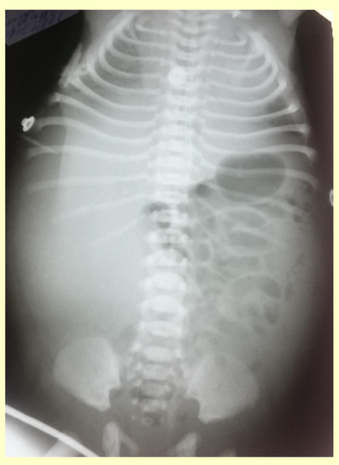

Fig.2. X-ray chest and abdomen of neonate on 8 day of birth with pulmonary hypoplasia requiring ventilation. The abdomen is enlarged. No renal function.

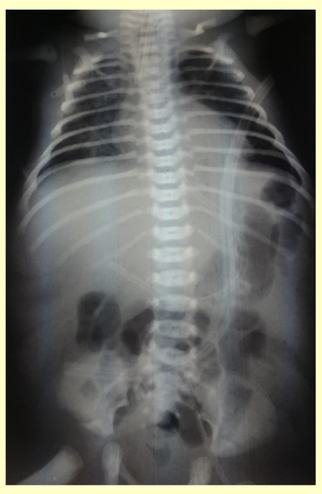

Fig.3. X-ray chest and abdomen of neonate on 18 day of birth after bilateral nephrectomy and started peritoneal dialysis.

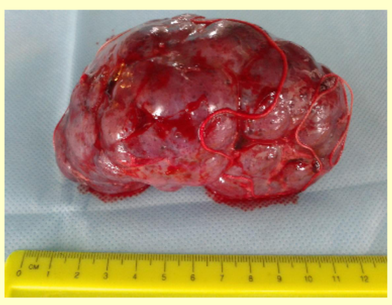

Fig.4. The right kidney after nephrectomy (length-120mm; weigt-0,4kg)

Results:

One patient (with Potter's syndrome) died from pulmonary insufficiency at 20th day of life before any surgery was conducted. The unilateral nephrectomy by retroperitoneal access was performed in 3 children at the age of 38, 40 and 58 days. The weight of remote kidneys was 400-700g. In two of them CVVHDF after surgery was conducted, in one renal function preserved. All patient died at 3, 4 and 10 days after surgery because of severe pulmonary and heart failure. In 2 children at the age of 15 and 10 days bilateral nephrectomy was made and CVVHDF was performed subsequently. They died on 6 and 9 days postoperatively due to pulmonary and heart failure. In 2 patients two-stage bilateral nephrectomy was carried out with 7 days interval. The surgery was performed at 10th and 17th days of life in the first child, at the 5th and 12th in the second. CVVHDF was conducted before and after surgery, and at the age of 45 and 32 days peritoneal dialysis was started. During all the treatment patients were on mechanical ventilation. The attempts to decrease respiratory support led to deterioration of the patients' condition. These patients had been living for 70 and 40 days, respectively. The lethal outcome owed to respiratory distress syndrome caused by lungs' hypoplasia in both cases. In one patient lipoblastoma of the chest and neck soft tissues developed.

Conclusions:

In our small group of patients with the neonatal manifestation of ARPKD the 100% lethality was revealed. The main cause of death was respiratory insufficiency due to lungs' hypoplasia. Nevertheless, early two-stage bilateral nephrectomy in combination with continuous renal replacement therapy and respiratory support offer a chance of surviving to such patients.

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