

Patient Report

Family with three daughters, two of them (6 and 3 years old) PH Type I

When our eldest daughter was two years old and had a urinary tract infections (UTI), an X-ray examination (MCUG test) confirmed a vesicoureteral reflux grade I-II (VUR). It was then recommended to treat her for one year with oral antibiotics as prophylaxis. She didn't have any more urinary tract infection after that and the antibiotic treatment was stopped.

Our second daughter suffered her first inflammation of the renal pelvis when she was one year old. This time we were only advised to do an MCUG test in case of a further UTI. The second UTI soon followed and she too had to undergo that test. A grade II VUR was then observed. She was then also treated with daily antibiotic treatment.

Recurring UTI's, disappointing experiences and a deep intuition caused us to change hospitals. In retrospect, it was a very good decision. Ultrasound examinations showed deposition of calcium salts (nephrocalcinosis) on both kidneys.

Surgical intervention of VUR to correct urinary reflux were performed endoscopically when she was three years old. At the same time the nephrology department carried out further investigations. An increased excretion of oxalate in the urine was proven. A 24-hour urine sample was taken.

The nephrologist suspected primary hyperoxaluria (PH). A genetic investigation confirmed the suspicion. The diagnosis PH type I was confirmed.

After a few months another infection of the renal pelvis occurred. In addition the urine continuously carried bacteria from the very beginning, including the pathogen *Pseudomonas*. We opted for open surgery (ureteral reimplantation UCNST). Several kidney stones were also removed. The hospital stay was used to simultaneously try to prevent the *pseudomonas* by an intravenous course of antibiotics. However, to this day the *Pseudomonas* persists in urine analysis. A year later she suffered another severe infection, which needed another hospitalization. Three months later, she spontaneously passed a large kidney stone.

She is now six years old and has been taking potassium daily for three years (to inhibit the formation of kidney stones) and pyridoxine. We pay attention to a low oxalate Nutrition. Her drinking quantity amounts to approx. 2 - 2 1/2 L per day for a body size of 128 cm and 26 kg weight. Since she is one year old, she has had a restless sleep and therefore wakes up regularly and drinks on this occasion. After school has started, it got a little bit more tricky with continuous drinking.

Between two hospital stays of the sister, our then nine month-old third daughter got sick. She also suffered from infection of the renal pelvis. The MCUG test confirmed a reflux, the ultrasound images showed nephrocalcinosis, the urine an increased oxalic acid excretion and the gene test also PH type I. When she was 11 months old, kidney stones had to be surgically removed twice and another surgery followed to remove a double J-catheter.

Based on our experience, and considering her underlying condition, we decided to treat her reflux with surgery instead of antibiotic prophylaxis. She was 1 1/2 years old at the time of surgery.

She's now 3 1/2 years old. For the last two years, everything has been going well. However, from the beginning it was very difficult for her to change her drinking behaviour. She drinks about 1.2 - 1.3 litres per day for a height of 107 cm and 19 kg weight.

She also takes pyridoxine and potassium daily and has a low oxalate diet. We are very grateful that our children are responding to the pyridoxine. Because of the low oxalate diet, nuts were avoided. To an unwanted ingestion of peanuts (we strongly suspect), an allergic reaction occurred. At the moment, clarifications are being made in this regard.