

URINARY COMPLEX CONGENITAL MALFORMATION

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Abstract

The urinary congenital anomalies are frequent in pediatric pathology and their presence is often pointed out early by an acute symptom. We present a case report of a 3 month girl hospitalized with severe hydronephrosis and urinary infection, who presented ureteral segmental stenosis and ureterocel. The prenatal diagnosis was unknown, because the mother did not make an ultrasound exam during pregnancy. By an endourological approach we found a left ureterocel and a close narrowing of the superior half of the left ureter. We removed the ureterocel by electroresection. By an open approach we removed the stenotic ureteral segment. From a pelvic flap we obtained a new superior ureteral segment and we made an anastomosis between the 2 ureteral segments. After 10 months a follow up revealed a narrowing of the inferior segment of the left ureter. By the endourological approach we inserted into the ureter a 3 CH JJ stent for a long term. It is a good long term evolution. The follow up evaluation (by clinical examination, ultrasound exam and laboratory investigations) reveals no clinical signs and the urine sterility.

Key words: hydronephrosis, congenital urinary anomalies, early diagnosis, surgical treatment.

Introduction

Hydronephrosis is a dilation of the inside or collecting part of the kidney. It often results from a blockage in the ureter where it joins the kidney that prevents urine from draining into the bladder. Urine is trapped in the kidney and causes the kidney to stretch. Hydronephrosis may also be due to abnormal backwash or reflux of urine from the bladder. It is often detected on an ultrasound test during pregnancy (prenatal ultrasound). The blockage that produces hydronephrosis is usually the result of a narrowing at the top of the ureter near the kidney. The severity of hydronephrosis depends on the extent of the blockage and the amount of stretching of the kidney. It may range from mild to severe. Children with mild and moderate hydronephrosis usually do not have symptoms, the kidneys are minimally affected and the problems may disappear in the first year of life. In extremely severe cases of hydronephrosis, damage to normal kidney function may occur. In addition to affecting the child's kidney function, this condition may also cause infections, pain and bleeding. These effects may take months or even years to occur or may never occur. To determine the amount of hydronephrosis present, nuclear medicine and radiologic tests to measure kidney function and structure may be

recommended. In children with mild hydronephrosis, observational therapy has been shown to be safe and has become the accepted method of treatment. However, in children with moderate to severe hydronephrosis, the answer is not as clear. The surgical procedure (pyeloplasty) involves removing the obstructed part of the ureter and then reattaching the healthy ureter to the collecting part of the kidney. The success rate of the surgical therapy in infants is 90-95%. Prenatal testing for hydronephrosis has permitted early detection and treatment. In the past most children were found to have hydronephrosis at the time of urinary infection or pain. Surgery was almost always performed in many children after 3-4 years of age. Those children that improved by itself without ever causing infection or pain, were probably never diagnosed or treated for hydronephrosis. Children who are diagnosed prenatally with moderate to severe hydronephrosis are now being seen at such an early age that they have not had a chance to improve on their own. Current testing cannot accurately predict which patients might or might not get better on their own. Therefore, today there is no standard treatment for all children. Many centers are choosing to watch and carefully monitor children with moderate to severe hydronephrosis while others continue to use surgery as treatment.

Case report

A three month girl presented in our clinic with urinary tract infection. She presented fever, vomiting, increasing urinary frequency and a general bad condition. Laboratory exam of the urine revealed the infection with E.coli. Antibiotic therapy was started and we obtained a short remission of the clinic symptoms. The abdominal ultrasound exam and the intravenous urography revealed a left severe hydronephrosis and a left ureterocel. By a 7 CH cystoscope we explored the urinary bladder and found the left ureterocel. We removed this ureterocel by electroresection. The left ureter exploration reveals a strong stenosis in its superior half. In this situation we decided the open surgery is necessary and we found an important pelvic dilation, a partial kidney rotation with a fetal aspect, a reduced renal mass and a strong stenosis of the superior segment of the left ureter. We removed the narrowed segment of the ureter (3 cm length). We created a new superior ureteral tube from a flap obtained from the stretched pelvic wall. We performed an anastomosis between the two ureteral segments (fig. 1). For 10 days, a 3 CH tube protected the anastomosis. We used 6-0 monofilament resorbable sutures (Surgicryl). Antibiotic therapy continued for three months.



Fig.1. Postoperative aspect with the JJ stent.

The evolution was good after operation. A follow up evaluation consisted in clinic, ultrasound exam and urine exam in every month in the firsts 6 months, and at three months after this period, and no problems reveal. In the second year we found an ureterohydronephrosis (by ultrasound examination). By cystoscopy we found the narrowing of the anasthmosis between the two ureteral segments. We performed to incise this stenosis and inserted a 3 CH JJ stent for long term. Antibiotics used for three months. After this operation follow up evaluation continued (at every three months) and no clinical problems reveal and urine is sterile.

Discutions

In the absence of maternal ultrasound exam, the diagnosis delaied since the severe urinary infection occurs. The early diagnosis is esential (better during pregnancy) and it permit to adopt a correct therapy. The associated urinary malformations needs a complex therapeutical approach (endourologically tehncis and open surgery tehncis). Short term prognosis is good in the presented case because the urine flow is good and no functional troublesof kidney occurs. Long term prognosis depend by the urinary flow and the development of the affected kidney.

References

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