African Journal of Urology (2015) **21**, 87–95

HOSTED BY

Pan African Urological Surgeons' Association

African Journal of Urology

www.ees.elsevier.com/afju
www.sciencedirect.com

Review

Congenital obstructive uropathy – Diagnostics for optimal treatment

Christian Radmayr

Department of Paediatric Urology, Medical University Innsbruck, Austria

Received 4 March 2015; accepted 31 March 2015

KEYWORDS
Congenital obstructive uropathy;
Diagnosis;
Treatment

Abstract
Antenatal ultrasound screening techniques reveal urological abnormalities in about 1 of 500 cases with half of which belonging to hydronephrosis. Postnatal appropriate diagnostic modalities are obligatory for proper diagnosis. Nowadays, ultrasound is definitively one of the most useful imaging modalities for the upper tract in children with hydronephrosis. It is non-invasive, radiation free and can be repeated. In addition, ultrasound also offers excellent images of the lower urinary tract in the diagnosis of posterior urethral valves or ureteroceles for example.
An additional tool is isotope renal scintigraphy which is method to analyse differential renal function and drainage of the kidney as well as functioning cortical mass with an accurate image of renal parenchyma.
Today MRI studies are becoming more and more popular. This technique is particularly suited to urological imaging, because of its excellent delineation of water/urine-containing structures.
The micturating cystourethrogram is still the gold standard in imaging the bladder and the urethra. It also serves as a functional investigation while the patient is voiding. The technique consists of catheterizing the person in order to fill the bladder with a radiocontrast agent and is, therefore, an invasive method.
Since there is a risk of an increase in severity and functional deterioration with time on the one hand but also improvement and stable function on the other hand, suitable diagnosis is mandatory in order to rule out the children at risk.

Natural history of prenatally diagnosed hydronephrosis

Routine antenatal ultrasound screening was introduced 30 years ago, and rapidly gained wide acceptance in some countries. Urological abnormalities are detected in 1 in 500 pregnancies [1,2], among which half are unilateral hydronephrosis (Fig. 1).

Before the era of prenatal ultrasound, hydronephrosis was mainly diagnosed in symptomatic older children presenting with loin pain, stones, haematuria, or pyelonephritis. Antenatal diagnosis has brought into light a unique and unknown population of healthy and asymptomatic infants having mild to severe hydronephrosis, with variable outcome [3].

E-mail address: christian.radmayr@i-med.ac.at

Peer review under responsibility of Pan African Urological Surgeons' Association.

http://dx.doi.org/10.1016/j.afju.2015.03.006

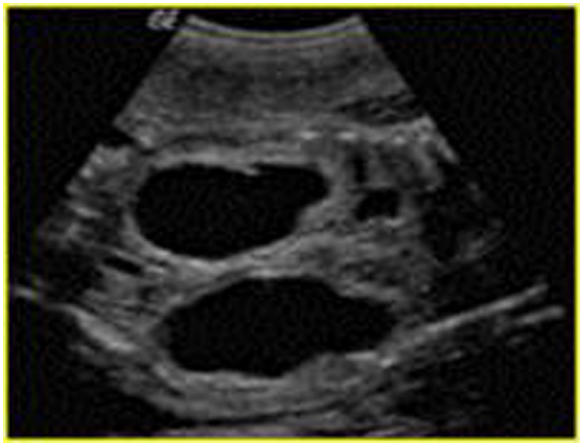

Figure 1 Antenatal ultrasound showing bilateral hydronephrosis.

It is known that prenatally diagnosed hydronephrosis may increase in severity and show functional deterioration with time, consistent with the definition of obstruction. It has also been reported that such a dilatation may remain stable (in fact the majority of cases), or improve with time, although it is unknown whether a true obstruction was present and vanished, or if the dilated but non obstructed system simply improved [4,5]. The potential for spontaneous resolution of unilateral hydronephrosis thus questions the necessity of surgical treatment, and the true nature of this dilatation if not obstruction.

Tribute must be paid to HK Dhillon and the perinatal urologists team of the Great Ormond Street Hospital for the outstanding amount of work dedicated to the follow-up of prenatally diagnosed hydronephrosis over a 25 years period. In their "natural history" series with conservative management, they showed that:

- Follow-up based on ultrasound is safe.
- Virtually all infants requiring pyeloplasty [6,7] for impaired renal function at birth had pelvic AP diameter >20 mm (Fig. 2).
- The vast majority of children born with normally functioning kidney, but who required surgery for deteriorating function had pelvic AP diameter >20 mm at birth.
- When deterioration of the function occurs, it is usually preceded by a worsening of the dilatation.

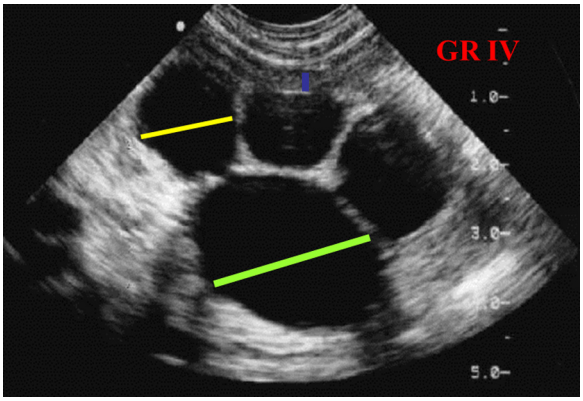

Figure 2 Grade IV hydronephrosis with an AP diameter above 20 mm.

The same authors showed that ultrasound is the most useful imaging study to differentiate infants with PUJ obstruction who (will) require surgery from those who have pelvi-calyceal dilatation of no clinical significance. Among infants with unilateral hydronephrosis and normally functioning kidney on the postnatal MAG-3 renal scan, 90% of those with renal pelvis between 30 and 40 mm, and virtually all of those with renal pelvis >40 mm will eventually deteriorate. In contrast, among those with hydronephrosis <20 mm at birth, only 11% will eventually require surgery (they usually have major calyceal involvement, intra-renal pelvis, and often early prenatal diagnosis during the second trimester). The "grey zone" lies between 20 and 30 mm, where 40% may eventually deteriorate, but the same proportion may also improve spontaneously.

Other authors have also studied the natural history of prenatally diagnosed hydronephrosis with regard to the potential deterioration of renal function [8–11]. Koff followed non-operatively 104 neonates with unilateral hydronephrosis regardless of the degree of dilatation and of the initial degree of functional impairment. A large proportion of those with initial impairment of DRF rapidly improved spontaneously. Twenty-three of 104 (22%) ultimately required surgery for deterioration of either DRF, dilatation, or both, always before 18 months of age. Of those requiring pyeloplasty, there was no permanent loss of renal function and DRF eventually returned to exceed predeterioration levels.

Natural history series have provided very useful informations regarding to prenatal counselling and postnatal management strategy. The main points arising from these natural history studies are

- The majority of prenatal unilateral hydronephrosis (75%) will either remain stable or improve spontaneously
- Some of them will show increase of dilatation and/or deterioration of the renal function, and will ultimately require surgery [12–14]
- It is probably safe to closely follow-up dilated kidneys with ultrasound alone, and perform isotope imaging if hydronephrosis worsens
- The risk of deterioration correlates with the degree of dilatation of both pelvis and calyces. The threshold above which the risk of deterioration during follow-up seems unreasonable is about 30 mm.
- When normally functioning kidneys deteriorate during follow-up, many of them will recover preoperative function level after pyeloplasty.
- Management of kidneys with initial functional impairment is debated. Some of them may see their DRF improve spontaneously with time. In contradiction, even when surgery is performed, this subgroup of congenitally damaged kidneys seem not to improve as much after pyeloplasty as the ones with sudden deterioration. It is probably safe (and easier to follow-up) to perform pyeloplasty in infants with initial renal differential function impairment, although the renal outcome may not be different.

Symptomatic hydronephrosis

Symptomatic hydronephrosis has long been the main clinical presentation leading to the diagnosis of PUJ obstruction. Different complication could be observed, like febrile UTI, symptomatic stones, or intermittent abdominal pain. In a number of cases, the latter takes part of a very specific clinical picture named "acute intermittent hydronephrosis" [15,16]. Typically, these children (usually