Clinics in Diagnostic Imaging (73)

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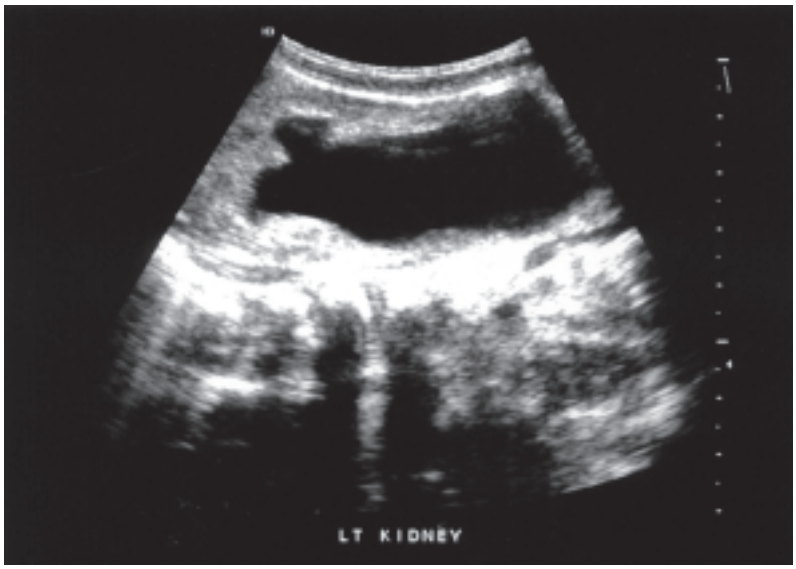

Fig. 1a Longitudinal US scan of the left kidney.

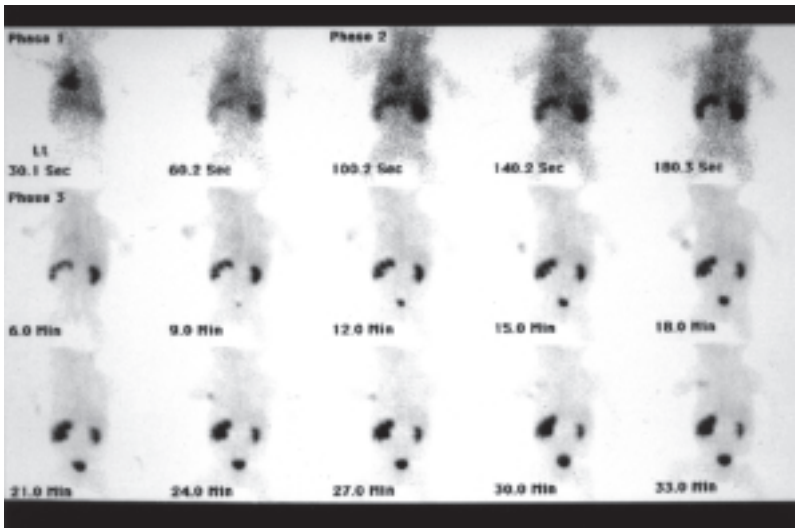

Fig. 1b MAG-3 diuretic renogram (posterior projection).

CASE PRESENTATION

A full-term male infant was born via Caesarean Section for placenta praevia. Antenatal ultrasonography (US) done at 32 weeks gestation demonstrated a left renal pelvis dilatation of 3.2 cm with normal-looking kidney parenchyma and normal bladder volume. On physical examination, his abdomen was soft and non-tender. No mass was palpable. Post-natal US was done on the 2nd day of life (Fig. 1a). What does it show? A Technetium (Tc)-99m MAG-3 renogram was performed on the 5th day of life (Fig. 1 b). What does it show? What is the diagnosis?

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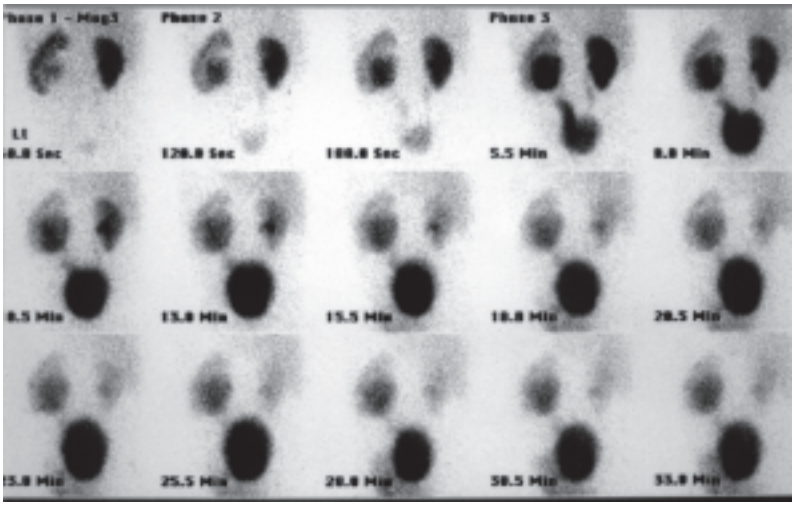

Fig. 2 Same patient as Fig. 1a, b. MAG-3 diuretic renogram (posterior projection) five months post-pyeloplasty shows much improved tracer drainage from the left kidney.

IMAGE INTERPRETATION

The post-natal renal US (Fig. 1a) showed an enlarged left kidney measuring 7.1 cm in length. The pelvicalyceal system was very dilated with a mid-transverse diameter of 3 cm. There was no hydroureter. The lower pole renal cortex was thinned. The right kidney measured 4.3 cm in length and showed a normal configuration and echogenicity. These US findings were consistent with the diagnosis of left pelvi-ureteric junction (PUJ) obstruction.

Tc-99m MAG-3 diuretic renography (Fig. 1b) showed that the right kidney was normal, displaying good uptake of tracer material with no obstruction to urine flow from the pelvicalyceal system into the bladder. Excretion half-time was nine minutes. The left kidney showed a dilated pelvicalyceal system with cortical thinning. There was little flow of tracer material from the hydronephrotic kidney into the bladder, even after the intravenous administration of frusemide. The excretory half-time of the left kidney was 33 minutes. These appearances were consistent with a diagnosis of incomplete left PUJ obstruction.

DIAGNOSIS

Left hydronephrosis due to pelvi-ureteric junction obstruction.

CLINICAL COURSE

The patient's biochemical renal function test was normal. Urine cultures were negative. He was discharged with prophylactic oral amoxicillin. A micturating cystourethrogram (MCU) was performed on an outpatient basis on the 10th day of life. There was no vesico-ureteric reflux or posterior urethral valve detected. He was followed up by the Paediatric Surgery service. At six weeks of age, the patient developed a fever and was admitted to the paediatric ward. Urine culture grew *Escherichia coli* in significant quantities. This was treated with intravenous antibiotics and the fever resolved. The patient was discharged with oral cephalexin prophylaxis.

The patient was followed up in the outpatient clinic with serial US and MAG-3 renography, without improvement in the hydronephrosis. The left kidney still showed a delay in isotope excretion. A decision for surgery was made and the patient underwent a cystoscopy, left retrograde pyelogram and a left Anderson-Hynes pyeloplasty at nine months of age. The cystoscopy showed normal urethral and ureteric orifices. The retrograde pyelogram showed gross hydronephrosis of the left kidney with a serpiginous configuration of the ureter at the pelvi-ureteric junction. Pyeloplasty was performed and a double-J stent was left in situ. The surgery was uncomplicated and the patient was well at discharge. The double-J stent was subsequently removed. A follow-up MAG-3 renogram five months post-surgery showed satisfactory drainage from the left kidney and normal differential function (Fig. 2). Renal US performed 11 months post-surgery showed a normal post pyeloplasty appearance of the left kidney with no evidence of hydronephrosis. The latest MAG-3 renogram two years post-surgery was normal. The patient has been taken off prophylactic antibiotics and is undergoing yearly outpatient follow-up.

DISCUSSION

The widespread use of the antenatal US in foetal-maternal screening has led to an explosion in the number of neonates with antenatally-diagnosed hydronephrosis. The most common causes of hydronephrosis in order of frequency are: PUJ anomalies, vesicoureteric junction anomalies, vesicoureteric reflux, multicystic kidney and posterior urethral valve. Other causes include

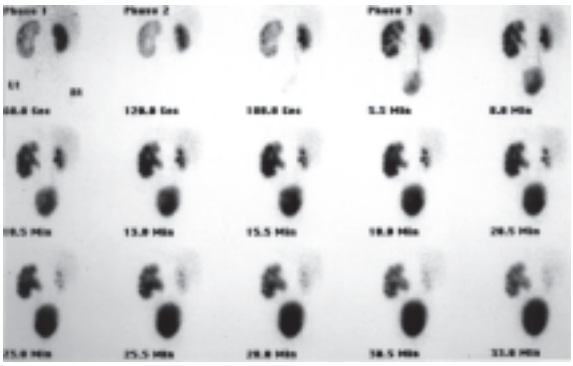

Fig. 3a MAG-3 diuretic renogram (posterior projection) shows delayed uptake and tracer hold-up in an 11-month-old boy with left pelvi-ureteric junction obstruction.

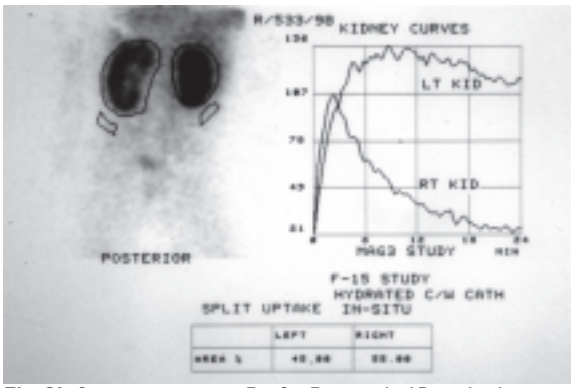

Fig. 3b Same patient as in Fig. 3a. Frusemide-15 study shows an obstructed left renogram curve.

megaureter, ureterocoele, neurogenic bladder, prune-belly syndrome and urethral atresia. Of all these causes, PUJ obstruction is by far the most common[1]. In cases where the aetiology is understood (for instance, in posterior urethral valve and ureterocoele), conventional treatment to re-establish optimal urinary drainage is accepted. It is the population of neonates with hydronephrosis consistent with a radiological diagnosis of PUJ obstruction that poses a challenge to the clinician. The majority of these infants are asymptomatic with no abnormal physical signs. The challenge lies in differentiating between neonates who would benefit from early surgery to prevent subsequent symptoms or deterioration of renal function, and those whose hydronephrosis is inconsequential.

Hydronephrosis is not synonymous with obstruction. Hydronephrosis is merely an anatomical entity defined as an enlargement of the capacity of the collecting system of the kidney, calyces and pelvis. Obstruction, on the other hand, has been defined clinically as any restriction to urinary outflow which if untreated will injure the kidney[2]. The two most common tests to assess the significance of hydronephrosis are US and diuretic renography. However, these tests do not accurately assess whether obstruction is present[3]. Therein lies the challenge for the clinician differentiating those who have asymptomatic hydronephrosis per se from those with obstruction, using the limited diagnostic tests available today. Obstruction needs surgical correction whereas non-obstructive hydronephrosis does not[4].

The post-natal treatment of children with antenatal hydronephrosis has been a controversial topic over the recent decade. During this time, there has been a gradual paradigm shift from early surgical intervention to a more considered conservative one. Yet, the indications and timing of surgery are still debated, and disagreement exists over the ability of different diagnostic tests to define obstruction accurately or predict which kidney would benefit from surgical intervention. The clinician must develop his or her own "comfort level" for treating hydronephrosis. This "comfort level" is dependent on three main criteria, namely (1) the amount of hydronephrosis present on US, (2) the relative renal function as measured by renal scintigraphy, and (3) the rate of radionuclide tracer washout with frusemide. All of these measurements are helpful parameters but are associated with limitations. There is thus far no "gold standard" for prospectively identifying those who would benefit from surgery.

Ultrasonography provides the best evaluation of hydronephrosis, both for screening and for follow-up. Important features to note on US are renal length, anteroposterior diameter of the renal pelvis, cortical thickness and the echogenicity of the cortex. The most meaningful way to utilise US is probably to obtain serial measurements. Progressive worsening of hydronephrosis usually indicates an obstruction, whereas improvement in hydronephrosis suggests the opposite.

Nuclear renography is currently the best means of determining the relative renal function and of following renal function over time (Fig. 3a). The potential of nuclear renography is fully achieved when certain concepts are used in the interpretation. First, one must compare the dilated kidney with the non-dilated kidney. Secondly, differential and absolute renal function must be determined. Lastly, these determinations must be followed over time. The radiopharmaceutical agents used are all bound to Tc-99m, an isotope that has excellent imaging characteristics and that is associated with a relatively low radiation dose. The difference in characteristics between agents depends on the specific compound bound to the Tc-99m. Diethylenetriaminepentaacetic acid (DTPA) is excreted by glomerular filtration and is not secreted or reabsorbed by the renal tubules. Because of this, it provides an indirect means of measuring the glomerular filtration rate (GFR)[6]. Differential GFR is obtained by comparing the amount of uptake in each kidney during the first one to three minutes after intravenous injection[6]. Mercaptoacetyltriglycine (MAG-3) is cleared by the kidneys primarily by tubular secretion and to a much lesser extent, by glomerular filtration. Tc-99m-labelled

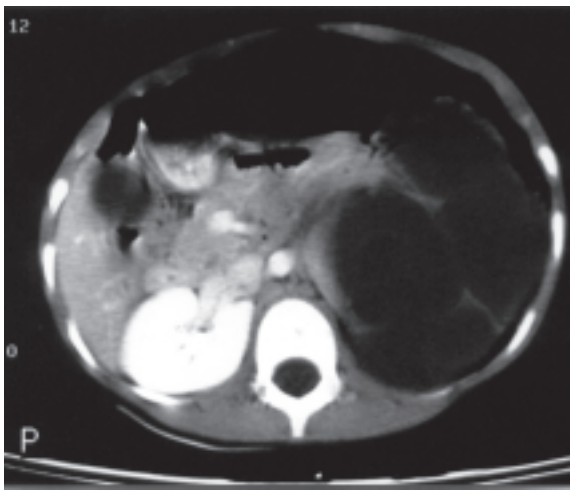

Fig. 4 Axial CT scan of the abdomen of a seven-year-old boy shows gross left hydronephrosis from long-standing obstruction.

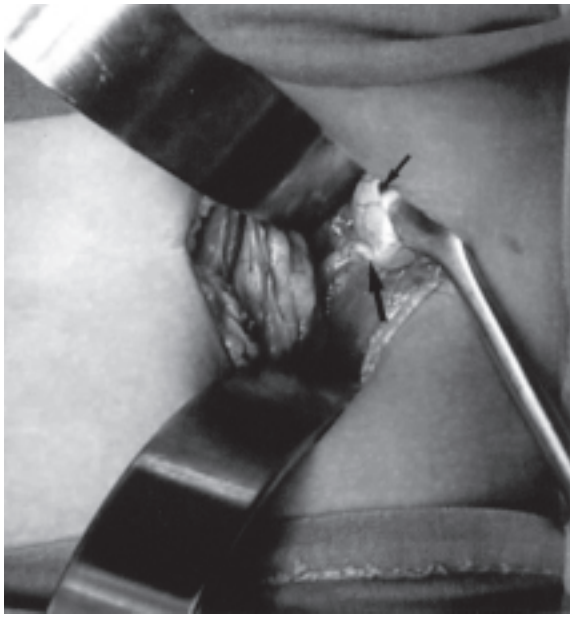

Fig. 5 Intra-operative photograph shows hydronephrosis (small arrow) and a serpiginous configuration of the pelvi-ureteric junction (large arrow).

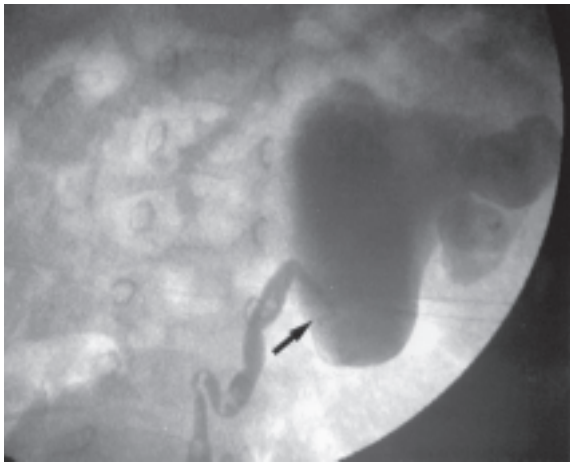

Fig. 6 Retrograde pyelogrm of a patient with a large left pelvi-ureteric junction obstruction shows a patent ureter with a serpiginous configuration at the pelvi-ureteric junction (arrow).

MAG-3 is thus an excellent agent for estimating the effective renal plasma flow, which itself corresponds to differential renal function, because it is not retained in the parenchyma of the normal kidney for very long, MAG-3 also provides excellent imaging characteristics[7]. Moreover, it may be more effective than DTPA in cases of renal function impairment because of its relatively high degree of renal tubular secretion.

Dimercaptosuccinic acid (DMSA) affords an accurate calculation of differential function, as well as excellent imaging of the renal cortex. In DMSA renography, the kidneys are imaged at least two to six hours after intravenous administration of the radioisotope. Because the individual measurements of differential function are subject to inaccuracies, serial measurements are most meaningful. If serial measurements indicate that the affected kidney function is improving rapidly and appropriately as expected for age, obstruction does not exist and surgery is not indicated. On the other hand, if sequential measurements indicate that renal function is deteriorating, failing to improve or not improving as expected, obstruction is most likely present and should be corrected immediately with surgery.

Diuretic renography is the third tool used to diagnose obstruction (Fig. 3b). Although a seemingly straightforward test, the kidney's response to diuretic "washout" of a radiopharmaceutical has proved controversial. The underlying idea of diuretic renography is simple, i.e. in the absence of obstruction, the radionuclide will be washed out by the diuretic effect of frusemide. If obstruction is present, the radionuclide will not wash out. However, there are many factors that can affect the results of diuretic renography. It is a heavily operator-dependent test, and the techniques and standards vary from one institution to another. It is subject to so many variables that it has been denounced as unacceptable in the diagnosis of obstruction[3,8]. The state of hydration, renal function, volume and contractility of the renal pelvis, patient position, bladder filling, timing and dose of diuretic administration, and type of radioisotope may all result in misinterpretation. It has been suggested that the washout pattern should only be considered valid in the newborn with a hydronephrotic kidney when it shows brisk washout and thereby excludes obstruction[4].

Micturating cystourethrogram is indicated in all cases of neonatal hydronephrosis because vesicoureteric reflux has been found in up to 14% of children with suspected PUJ obstruction[9]. Abdominal computed tomography (CT) is sometimes requested in children presenting with a flank mass and shows hydronephrosis (Fig. 4)

The ideal management of the infant with unilateral PUJ obstruction is currently unresolved.

Management paradigms range from early pyeloplasty to aggressive observation. Dhillon has proposed following-up patients based on the degree of hydronephrosis as measured by the anteroposterior diameter and cortical thinning on US[10]. Neonates with mild hydronephrosis (less than 12 mm) pre- and post-natally require US at the ages of three months and one year. If the dilatation persists, this can be repeated at two, five and 10 years of age. The group with moderate hydronephrosis (13-19 mm) and good function should undergo follow-up US with the addition of nuclear renography at three months, as well as one, two, five and 10 years of age. Children with severe hydronephrosis (more than 20 mm) will have functional levels ranging from good to poor. Neonates with good renal function and a dilatation of 20-50 mm require further long-term studies to identify those who will benefit from surgery. After the initial functional assessment, management should be conservative with follow-up by US and nuclear renography at three months, six months, one year and yearly thereafter.

In all cases, surgery is indicated if there is worsening hydronephrosis, deteriorating differential renal function, or the appearance of symptoms such as urinary tract infection, haematuria, pain or flank mass. Kidneys with dilatation more than 50 mm on anteroposterior diameter should almost always have a pyeloplasty by six months of age. The subgroup of kidneys with very poor function (less than 10%) would warrant a nephrectomy.

Open Anderson-Hynes pyeloplasty (Fig. 5) has been widely accepted as the surgical treatment of choice for PUJ obstruction in children, with a success rate of more than 90% in most reports[11]. The procedure is usually preceded by an intra-operative retrograde pyelogram (Fig. 6). It is a dismembered pyeloplasty in which a microsurgical technique is used to provide a watertight anastomosis[12] and remains the "gold-standard" in the surgical management of PUJ obstruction. However, debate still persists on the merits of a transanastomotic stent placed intraoperatively[13]. It has been suggested that placement of a stent lowers the incidence of urinary extravasation and urinoma formation after repair of the obstruction. The decision to place a stent remains largely a matter of individual preference.

In conclusion, the management of PUJ obstruction is controversial. The pendulum has swung from an initially aggressive surgical approach to a more considered conservative one. Imaging modalities continue to be modified and improved in the hope of identifying a parameter which could pick up those infants whose hydronephrosis represents an obstruction detrimental to their well-being. However, it is mandatory when dealing with a group who are initially healthy and asymptomatic that a balance is achieved between the risk of surgery and excessive imaging.

ACKNOWLEDGEMENT

The authors would like to record a note of thanks to Professor Wilfred CG Peh, Senior Consultant, Department of Diagnostic Radiology, Singapore General Hospital, for his help.

ABSTRACT

A full-term male infant was antenatally diagnosed to have left hydronephrosis. Post natal ultrasonography (US) and diuretic renography confirmed the diagnosis of left pelvi-ureteric junction obstruction. His clinical course was complicated by one episode of urinary tract infection. Serial US and diuretic renography showed no improvement in the obstruction. The patient underwent an Anderson-Hynes pyeloplasty at nine months of age with no post-operative complications. The diagnosis and management of antenatally-diagnosed hydronephrosis are discussed.

Keywords: pelvi-ureteric junction obstruction, ultrasonography, nuclear renography, pyeloplasty

Singapore Med J 2002 Vol 43(5):265-269

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