Antenatally Detected Hydronephrosis

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The strategy of management of children with hydronephrosis has considerably changed as a result of the development of ultrasound techniques, allowing a prenatal detection. Hydronephrosis is defined as a dilation of the renal collecting system, and several entirely different clinical entities can be considered this general heading, whereas early detection may have a different impact depending on the entity considered. The present work aims to describe a certain number of these clinical entities, to discuss the strategic options of management that are proposed, and to evaluate the role of medical imaging, in particular the radionuclide approach. Congenital ureteropelvic junction anomaly, vesicoureteral reflux, posterior urethral valves, and duplex kidney will be successively considered. Multicystic dysplastic kidney disease, although not classified as hydronephrosis, will be mentioned because it may be mistaken for hydronephrosis.

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The strategy of management of children with hydronephrosis has considerably changed as a result of the development of ultrasound techniques, allowing a prenatal detection. Hydronephrosis is defined as a dilation of the renal collecting system, and several entirely different clinical entities, such as posterior urethral valves, megaureter, vesicoureteral reflux (VUR), or pelviureteric junction stenosis can be considered under this general heading. Fetal pelvic dilation is a frequent abnormality that has been observed in 1% to 5% of pregnancies.1-4

Early detection may have a different impact depending on the entity considered. Although in the times before ultrasound time, sepsis and renal failure were the usual signs, allowing a diagnosis of posterior urethral valves to come too late, the relief of obstruction in the very first days of life has now considerably improved the functional prognosis.

Being able to detect major VUR before the appearance of any clinical symptom suggestive of infection has led the clinicians to change their views about the pathogenesis of renal lesions associated to reflux. The concept of first urinary tract infection in the early months of life resulting in extensive irreversible lesions—the big bang theory3—has lost some credit in favor of congenital structural abnormalities. Moreover, the functional outcome of dilated reflux in adulthood probably essentially depends on the initial level of function, which can now be accurately evaluated in the very early weeks of life. Indeed, it has been shown that as many as 56% of children having at entry a reflux associated with low glomerular filtration rate (GFR) values will develop end-stage renal disease by 20 years of age.6

In another clinical entity, the pelviureteric junction stenosis, much debate has resulted from the antenatal detection. It is clear that the clinician has the possibility of organizing, from the beginning on, a close follow-up of both the anatomical aspects and the functional evaluation. If the clinician has chosen a conservative nonsurgical approach, he or she is now able to react rapidly in case of unfavorable evolution and operate. The drawback of the antenatal detection of this abnormality, discovered in entirely asymptomatic children, has led to a non-negligeable number of examinations and/or to early surgical interventions which, in a certain number of cases, might have been unnecessary.

The present work is aimed at describing a certain number of clinical entities grouped under the general heading of antenatally detected hydronephrosis, discussing the strategic options of management that are proposed, and evaluating the role of medical imaging, in particular the radionuclide approach. Congenital ureteropelvic junction anomaly, VUR, posterior urethral valves, and duplex kidney will be successively considered. Multicystic dysplastic kidney disease, although not classified as hydronephrosis, will be mentioned because it may be mistaken for hydronephrosis.

Techniques

Ultrasound

Antenatal Ultrasound

The introduction of routine fetal ultrasonographic examination has increased the detection rate of fetal anomalies,
whereas the technical developments of ultrasound (US) equipment have improved the diagnostic accuracy. This has led to the discovery of many fetal anomalies, and among them, upper renal tract dilation represents one of the largest groups amenable to neonatal management. Fetal renal pelvis dilation can be an early sonographic sign of fetal hydronephrosis, or a marker of other abnormalities such as renal duplication or VUR, which can, however, not be easily identified with US during pregnancy. Therefore, the patient is now presenting to the urologist or pediatric nephrologist before the baby is even born, with a presumptive diagnosis rather than with a symptom.

The majority of authors use the anteroposterior (AP) renal pelvis diameter system for classification of fetal renal pelvis dilation. An upper limit of 7 mm for the AP diameter in the third trimester of gestation has been convincingly validated as a predictor of renal pathology. MRI is more likely to display very dilated and poorly functioning systems. Compared with US, the coronal images provided by MRI seem to be particularly helpful. MRI is promising in the estimation of functional parameters such as split renal function and excretion, but it still requires technical consensus and extensive evaluation before compete with the isotopic studies. Limitations of MRI are mainly related to the need of sedation in infants and to the accessibility of the equipment.

Nuclear Medicine Techniques

99mTc Dimercaptosuccinic Acid Scintigraphy

This technique aims to provide visualization of the renal cortex and to detect regional or diffuse impairment, related mainly to acute pyelonephritis or late sequelae. The details concerning the technique itself have been extensively described and several guidelines are now available. However, because this review is devoted to antenatal hydronephrosis, some technical points should be raised. The renal uptake of the tracer, in the very early days of life, is low, and the acquisition time for obtaining images can be long, with the risk of getting poor-quality images because of the infant moving or crying. Because of the small size of the child, zoom images are mandatory, with the drawback of increasing the acquisition time once again. In hydronephrosis, there might be accumulation of tracer within the collecting system, which may be difficult to distinguish from true tubular cell uptake (Fig. 1) and may result in an overestimation of the function of the hydronephrotic kidney. However, the technique can be useful in these antenatally detected children who are, in most cases, asymptomatic. Any lesion detected will by definition be considered as congenital and not related to early infection. According to Nguyen and coworkers, parenchymal defects detected by dimercaptosuccinic acid (DMSA) scintigraphy often were not identified with renal US.

99mTc MAG3 Renography

Much has been written about the renographic technique, which in children has specific characteristics and pitfalls. We refer to recent overviews on that matter. Let me simply summarize here some of these particular points. In infants up to 6 months of age, physicians should give preference to tracers with high extraction rates, such as 99mTc MAG3 or 123 hippuran. Split function with 99mTc DTPA is not precise in this age group. The child should be adequately hydrated, but the need for intravenous fluids is considered unnecessary in the majority of patients. Similarly, placing a bladder catheter is an invasive procedure that should be avoided because spontaneous voiding is the rule in almost all patients having undergone a diuretic renogram. A late gravity assisted and spontaneous voiding image, properly scaled, is then necessary at acquisition, adapted to the size of the child. Furosemide injection together with the tracer (F0) has become more and more popular, thus reducing the gamma camera time and the
number of venipunctures. As far as the final result on drainage is concerned, this technique offers no additional advantage compared with the classical F + 20 test. However, in case of known hydronephrosis, there is no need for a basic renogram because in almost all cases the diuretic will be necessary. An important pitfall in case of F0 renogram is the shortening of transit and a time to the maximum of the renogram (Tmax), which may be less than 2 min. The time interval available for calculation of split function should then be adapted. Calculation of split function can be obtained using either the integral method and the Patlak-Rutland method. Identical results obtained with both methods represent a good quality control. In young infants with an immature function, important divergences between both methods can be observed, often because of the uncertainties in determining the Patlak fit (Fig. 3). Despite its theoretical advantage of a better correction of the vascular part of the background, the preference should probably be given, for those immature kidneys, to the integral method because of better counting statistics. Estimation of renal transit on both basic renogram and F0 renogram can easily be obtained by means of Tmax, which will discriminate in most of the cases between normal, moderately altered, or extremely altered (continuous ascending curve) transit.

The deconvolution technique has a theoretical advantage to allow a more objective assessment of renal transit. The practical drawbacks of the deconvolution method have been previously underlined and offer no special advantage for both the whole kidney and the cortical transit time. Moreover, defining the cortical area in young infants is particularly hazardous, and there is no single report in the literature demonstrating the advantage of the technique in the strategy of hydronephrosis. Output efficiency and normalized residual activity are probably the best quantitative parameters of transit, being more independent of the renal input. They can be calculated at the end of the basic or the F0 renogram, at the end of a F + 20 test and on the late postmicturition image, the latter representing the best drainage available. The T1/2 of the furosemide curve is traditionally used to estimate the

**Figure 1** A 6-week-old boy with posterior urethral valve, cystostomy, bilateral grade IV reflux, altered plasma creatinine. In the upper panel, for the evaluation of split function, a DMSA scintigraphy (performed 4 h after injection of the tracer) was chosen because of the patient’s young age, immature renal function, and altered overall renal function. The percentage relative function left is 60%, but the precise contours of the kidneys are rather difficult to estimate. In the lower panel, MAG3 scintigraphy was performed a few weeks later. The early 1 to 2 minute image looks clearly different from the DMSA image, with a relative left function of 70%. The late MAG3 images reveal the huge hydro-ureteronephrosis, explaining the misleading DMSA image, summing up true tubular uptake and retention in the collecting system.
response to the diuretic. In case of nonequivocal steep slope, it excludes any significant problem of drainage. In the other cases, this parameter is not recommended, in particular because the late post erect and postmicturition views will often change completely the interpretation of the test. Moreover, it is remarkable that $T_{1/2}$ is now applied to F0 test without changing the criteria of interpretation as defined on a F+20 test. Finally, the main pitfall of the renogram is the significance of a poor drainage. The furosemide test is aimed to separate the “lazy” collecting systems from the real impairment of urinary flow. Unfortunately, the “reservoir” effect of a dilated cavity may result in a poor drainage, even if the flow through a narrowed junction is rather acceptable.

**Plasma Sample Clearance**

The GFR can now be accurately determined using simplified 2- or 1-blood sample plasma clearance methods. Minor controversies related to the technical aspects of these methods concern principally some correction factors, the quality control, the normal values in children. Normal values for Cr-EDTA clearance have been recently revised and can now be expressed as percentile values for all ages between 1 month and 15 years. However, the main problem is the reluctance of the clinician to apply these methods, despite the accuracy and precision, which are greater than with the traditional chemical methods. The easiest and probably the most accurate method to determine the absolute single kidney GFR is to combine the split function obtained by means of MAG3 renogram and the measurement of overall GFR using plasma samples. Changes in split function values during follow-up should be taken with caution, a decrease on the abnormal side might indeed be the result of an increase of functional compensation on the contralateral side, to a normal maturation on that side while no maturation occurs on the abnormal side, or to a real decrease of the absolute value on the abnormal side. Determination of absolute single kidney allows to discriminate between these three conditions (Fig. 4).

**Radioisotopic Cystography**

Direct RN cystography is based on the same principle as MCUG, and its invasiveness is identical. The bladder is pro-
gressively filled through a bladder catheter and both the filling phase and the voiding phase are entirely recorded.\textsuperscript{59} The sensitivity in detecting reflux is therefore greater than with MCUG. However, the urologist, being familiar with the radiological grading system and the capacity to distinguish dilated from nondilated systems, will generally give the preference, at least at entry, to the radiological MCUG. Any further control of a known reflux should theoretically occur by means of direct radionuclide technique.

Indirect cystography at the end of a \textsuperscript{99m}Tc MAG3 renogram is not feasible in patients younger than 3 years of age. However, it is not rare that a reflux episode can be detected in infants during the renogram, either because of a full bladder, or as a consequence of a spontaneous micturition. Such a detection may spare this young child the aggressive direct cystography, which often is planned during conservative follow-up of antenatally detected hydronephrosis. A sudden increase of activity in the late phase of the renogram is indicative of reflux. However, images should be carefully checked to exclude any movement artifact which could give rise to the same curve pattern.

\textbf{Clinical Entities and Medical Imaging}

\textbf{Multicystic Dysplastic Kidney Disease}

Distinguishing hydronephrosis from multicystic dysplastic kidney disease (MCDK) is essential because the ap-
The approach of therapy and indications for surgery differ in each. Typical MCDK consists of multiple variably sized cysts in a “bunch-of-grapes” type of cluster, with very little stroma, loss of reniform configuration, and an absence of a caliceal collecting system. The renal artery and vein usually are hypoplastic or absent. The ureter may be absent but is usually atretic. The etiology remains unclear. Many investigators have proposed that obstruction is the primary factor involved in the development of dysplasia. However, MCDK secondary to obstruction could not be simulated in an animal model. Another theory that predominates is the failure of ureteral bud-metanephros interaction. The incidence of MCDK is high, between 1 in 3,100 and 1 in 4,300. It is one of the most common causes of abdominal mass in infants. The diagnosis may be made as early as 15 weeks of gestation and bilateral cases have been reported with a frequency as high as 19% to 34%. Segmental MCKD may appear in horseshoe kidney and in single moieties of kidneys with duplicated collecting systems. In 20% to 43% of unilateral MCDKs, contralateral upper or lower urinary tract abnormalities are encountered, with the most common being contralateral VUR, occurring in 15% to 28% of cases. Contralateral ureteropelvic junction stenosis is found in 3% to 12% of patients. Confusion between MCDK and hydronephrosis is essentially in cases with a single predominant cyst. The natural history of MCDK reveals no association with urinary tract infection. The association with hypertension is questionable in most of the cases. Although in a review of the literature, multiple reports of malignancy arising in MCDKs were found to exist, one may question whether there is truly an increased risk above that of the normal population. As far as treatment is concerned, the high grade of spontaneous regression of the disease and the low rate of complications support the conservative management of MCKD. However, the patient’s family should be informed of the life-long need for regular follow-up.

**Imaging Procedures**

**Ultrasound.** Antenatal routine sonography can make the diagnosis at a mean gestational age of 28 weeks, depending on severity. The US pattern is characteristic: multiple noncommunicating cysts of varying size and nonmedial location of the largest cyst, in addition to the absence of normal renal parenchyma. The use of US can help the clinician to evaluate the morphology of the contralateral kidney and rule out most of the possibly associated anom-
Postnatal US may be required to differentiate MCDK from hydromeplasia.

**99mTc DMSA Scintigraphy or 99mTc MAG3 Renography.** Suspicious sonographic findings should be correlated with renal scintigraphy findings of an absence of renal function. On the contrary, the presence of a peripheral ring of activity will suggest the diagnosis of hydronephrosis. MAG3 renography provides the additional information regarding impaired drainage. On occasion, renal scintigraphy (either DMSA scintigraphy or MAG3 renogram) may demonstrate a functional segment associated with a MCDK in a duplicated system. The determination of overall GFR by means of blood sampling, combined with the scintigraphic study, may provide useful information about the contralateral kidney and the functional compensation one may expect in case of a unique normal functioning kidney.

**Cystography.** Complete evaluation of MCDK also includes MCUG. The reason for that is that “early detection of contralateral VUR is vital to prevent pyelonephritic scarring and deterioration of renal function in the only functioning renal unit.” Is this reasonable, taking into account the very low rate of infection occurring in patients with MCKD? Furthermore, the majority of those cases of contralateral VUR are low grade with a high rate of spontaneous resolution at 2 years. It should be noted that, according to the Multicystic Kidney Registry, only 15% of the total patients had undergone voiding cystourethrography.

**Posterior Urethral Valves**

Congenital urethral obstruction (posterior urethral valves; PUVs) creates a spectrum disease. The timing and degree of the urethral obstruction as well as the associated anatomy of the upper tracts are responsible for the range of abnormalities found in valve patients. The degree of urethral obstruction is variable with the anteroposterior diameter of the urethral lumen, at the level of the valves, measuring from 3 mm to greater than 1 cm. In the second trimester of fetal life, the most common cause of lower urinary tract obstruction in boys is posterior urethral valves, which are tissue leaflets fanning distally from the prostatic urethra to the external urethral sphincter. The failure of the bladder to empty during an extended examination, the presence of abnormal kidneys and oligohydramnios must raise the suspicion of posterior urethral valves.

With fetal bladder outlet obstruction by PUV, high intravesical pressures develop and are transmitted to the upper tracts, resulting in ureteral dilation and vesico-ureteral reflux, urinary stasis, parenchymal compression and damage of renal parenchyma that may be primarily abnormal. Those fetuses with the most high-grade obstruction often do not survive in utero. Historically, severe PUVs that remained undetected after birth resulted in urinary sepsis, renal failure and death in infancy. It is therefore clear that ablation of the urethral obstruction should be performed in the first days of life.

VUR is the retrograde flow of bladder urine into the upper tracts. We refer for a detailed description of the ureterovesical junction, pathogenesis of VUR and experimental studies to Godley’s review. In clinical practice, VUR describes a common primary disorder of childhood associated with urinary tract infection and renal scarring, traditionally called “reflux nephropathy.” The reflux-associated nephropathy can be acquired from reflux of infected bladder urine, or it can be congenital. One can distinguish mild forms of VUR with normal or mildly dilated ureters and severe VUR into dilated upper tracts. An abnormal ureterovesical junction may arise from maldevelopment or delayed maturity, resulting in so-called primary reflux, or it may be distorted by changes in the bladder wall secondary to other pathology, the so-called secondary reflux. The distinction has relevance in that there may be contributing pathology, anatomical abnormalities, that need to be identified or treated.

For example, neurogenic bladder and elevated intravesical pressure may play a major role in the etiology of reflux.
Transient infravesical obstruction during fetal development has been cited to account in part for the predominance of boys with VUR identified after prenatal hydronephrosis, some of whom have urological features reminiscent of posterior urethral valves, such as thickened bladder wall. Although the reflux event most usually disappears during growth, the disorder is of clinical concern because of the morbidity from ascending urinary infection and the risk of hypertension and progressive functional deterioration of the kidney. It has been shown that there is a strong association between the severity of reflux and the frequency of pyelonephritis episodes as well as the frequency and extension of the renal scarring (Fig. 5). However, it is unclear what the causal relationships are among VUR, urinary infection, and the associated nephropathy. Most clinical studies have concerned older children with acquired renal scarring, and it is now clear that the contribution of congenital nephropathy in association with VUR has been largely underestimated.

The antenatally detected VUR is by definition asymptomatic. Thus, any kind of associated nephropathy detected at this stage is necessarily congenital, corresponds to some kind of maldevelopment, and is not related to urinary tract infection. In young infants, abnormal lower urinary tract function often coexists with VUR without any identified neurogenic or other causal pathology. The spontaneous resolution of VUR during childhood accounts for its greater prevalence among neonates compared with older children. In a screening study of 2,384 healthy normal neonates, VUR was diagnosed from MCUG with a prevalence rate of 1.26% after postnatal urinary tract dilation was detected with US. The indications for MCUG were moderate or severe hydronephrosis. Nevertheless, reflux is the cause of fetal renal pelvis dilation in 11% to 30% of cases. Some authors have made a case for performing MCUG in all infants presenting with antenatally detected renal pelvis dilation, irrespective of the degree of renal collecting system dilation on postnatal US. Nevertheless, a more recent prospective study demonstrated that a normal-appearing urinary tract on 2 successive postnatal US (5 and 30 days) rarely coexists with abnormal findings at MCUG.

The opinion that routine MCUG is not justified seems to be shared by more and more authors. Furthermore, current evidence suggests that only patients with grade IV to V disease are at high risk of serious adverse outcome and delayed resolution. In contradiction with a widely held belief, VUR in infants with antenatally diagnosed renal pelvis dilation was found, in a large and prospective study, to be of low grade in 74% of cases, with a 91% of 2-year spontaneous resolution. US abnormalities were found in 100% of major reflux and in 79% of lower grade reflux. Dysplasia was the only specific US finding in case of major reflux and was associated with low split function. Other US findings were not predictive for alteration of split function but, as mentioned previously, repeated normal US practically excludes the presence of severe reflux. MCUG could therefore be limited to those cases with abnormal US, whereas a more precise determination of single kidney function by means of radioisotopes would be indicated in cases with major reflux. Follow-up of single kidney function is particularly indicated in those cases with renal dysplasia detected on US, since maturation of function is generally absent.

**Pelviureteric Junction Stenosis**

Pelviureteric junction stenosis occurs in 13% of children with antenatally diagnosed renal pelvis dilation and is characterized by obstruction at the level of the junction between the renal pelvis and the ureter. The anatomical basis for obstruction includes intrinsic stenosis/valves or crossing vessels. Sonographic diagnosis depends on the demonstration of a dilated renal pelvis in the absence of any dilation of ureter or bladder. It should particularly be suspected when moderate (10-15 mm) or severe (>15 mm) dilation is seen in these circumstances. Prognosis may be poor in bilateral cases associated with oligohydramnios. Postnatal management of these children still remains a controversial topic among the nephrourologic community.

Expectancy and close follow-up have progressively gained wide acceptance, although the surgical attitude, either systematic within the first months of life, or on the basis of variable morphological or functional parameters, is still the present attitude for many clinicians. The degree of hydronephrosis in postnatal period is undoubtedly important to evaluate. Indeed, spontaneous resolution takes place in approximately 50% of the cases with mild hydronephrosis, whereas spontaneous resolution is much less frequent in case of more pronounced hydronephrosis. Consequently, no intervention seems to be required in the majority of cases. However, in a recent review of the literature, it was established that 0% to 51% of the cases were operated at study entry or at the first evaluation. Approximately 25% underwent surgery after a period of observation, without consistency in the criteria of treatment. Decreased split function at initial assessment is often used as an absolute indication for surgery, but this has been questioned in a prospective conservative follow-up. Moreover, the overall impression is that improvement does not occur when surgery is performed because of initial decreased function. A hydronephrotic kidney with a split function less than 40% may have developed like this in utero and no surgical intervention can be expected.
to produce a normal kidney. It remains also unclear whether the chance of postoperative functional restitution is greater in the younger age compared with later intervention. The time interval between functional deterioration and surgical intervention is generally lacking and might be more important than age at surgery for the functional outcome.

Can diuretic renography helps in the decision for surgery? We have already underlined the pitfalls in performing the test, such as the lack of postcontact and postmicturition views. However, even if the test is correctly performed, there is no straight relation between the degree of emptying and the indication of surgery, since poor or no emptying can reflect almost exclusively a dilated reservoir. Entities such as dilated VUR, obviously not related to obstruction, may reveal a similar degree of dilation. One can probably reasonably exclude an obstructive phenomenon in case of good response. Absence of significant drainage can only be described and quantified and should not be interpreted as representing true obstruction. The often observed improvement of drainage after surgery is not surprising because, during pyeloplasty, part of the reservoir has been taken off.

Can we then define obstruction in those patients with ureteropelvic stenosis? Stratifying those patients in 2 categories, the obstructed ones and the nonobstructed ones, is undoubtedly too simplistic. Some degree of partial obstruction exists in all cases but lies in a continuum between slight and much more pronounced narrowing of the junction. The main question is probably whether or not one can predict deterioration of function and anatomy at initial evaluation. There are unfortunately no solid data demonstrating that the size of the renal cavity, the initial differential renal function, the level of pelvic pressure, or the response to furosemide can elucidate an obstructive phenomenon in case of good response. Absence of significant drainage can only be described and quantified and should not be interpreted as representing true obstruction. The often observed improvement of drainage after surgery is not surprising because, during pyeloplasty, part of the reservoir has been taken off.

In summary, ultrasound performed at day 4 and 30 postnatally will define the grade of hydronephrosis. Once VUR and dilated ureter are excluded, pelviureteric junction stenosis becomes the most probable diagnosis. A MAG3 renographic study when the patient is approximately 1 month of age is indicated in case of significant hydronephrosis and will estimate the level of renal function and the quality of drainage. In well-selected cases, the use of MRI may offer additional morphological information, such as the presence of huge dilation of calices contrasting with a rather noncompliant pelvis, which may constitute an additional factor of risk. In the absence of predictive factors of further deterioration, a conservative approach seems reasonable at the price of close follow-up. It has indeed been shown that surgery, performed soon after sudden decrease of function, will result in restoration of function. On the contrary, loss of function will be irreversible if the acute total or subtotal obstruction complicating the partial chronic obstruction is not rapidly relieved.

Ultrasound is certainly the instrument of choice to detect any significant alteration of pelvic size and should be repeated frequently, at least during the first 2 years of life. Renographic control is mandatory in case of significant increases in the ultrasonographic diameter. Early surgery is not the solution to avoid this kind of complication. Although not frequent, clinical postsurgical complications such as severe recurrent infections caused by multiresistant bacteria, leakage, secondary obstructions, or vascular ureteral damage leading to loss of renal function are not excluded. Randomized standardized clinical trials are still missing despite 20 years of publishing mostly retrospective investigations related to congenital hydronephrosis. The experience on follow-up of conservatively treated patients is still limited and is not more than 10 to 15 years. However, in the past, when patients were diagnosed with pelviureteric junction as a result of symptoms, split function was often acceptable or almost normal, suggesting that the effect of symptoms on function was relatively modest.

Long-term follow-up of these patients is desperately needed to evaluate more precisely the frequency and consequences of clinical symptoms or the occurrence of complications such as severe tubular disease or stones.

**Duplex Kidneys**

Duplication of the renal collecting system is characterized by the presence of a kidney having 2 pelvic structures with 2 ureters that may be completely or partially formed. Many cases have no renal impairment and should be considered as normal variants. However, a proportion of duplex kidneys will be associated with significant pathology, usually attrib-
utable to the presence of VUR or obstruction. Fetal urinary tract dilations are related to complicated renal duplication in 4.7% of cases. VUR usually involves only the lower-pole ureter in 90% of cases. Compared with single-system reflux, duplex-system VUR tends to be of a higher grade with a high incidence of lower pole dysplasia. A ureterocele consists of a cystic dilation of the intravesical submucosal ureter. It is 80% of the time associated with the upper pole of a kidney with complete ureteral duplication. Renal tissue associated with ureters joining ureteroceles frequently is dysplastic, with accompanying hydroureteronephrosis. On occasion, ureteropelvic junction obstruction may be found in the lower moiety, in association with incomplete duplex systems. In utero, duplex kidneys are highly suspected in the presence of 2 separate noncommunicating renal pelvis, dilated ureters, cystic structures within one pole, and echogenic cyst in the bladder, representing ureterocele.

After an infant is born, the classical radiological workup of abnormal duplex kidneys is based on US and MCUG. The aim of US is to confirm the diagnosis, whereas MCUG is performed to detect VUR and to evaluate the ureterocele. MRI can be useful in selected difficult cases, allowing a better definition of the 2 moieties, the shape of the ureters, and their possible ectopic implantation. Isotope studies are mandatory to determine renal function that remains in the abnormal renal moiety and the functional compensation in the normal moiety. Most people agree that the surgical approach to complicated duplex systems is largely determined on the presence or absence of function of the affected renal moiety. Regardless of the nature of the diseased moiety, however, the evolution of the functioning moiety seems very favorable through time with remarkable stable split renal function and the absence or absence of function of the affected renal moiety.

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