Anomalies of the Renal Pelvis and Ureter

Fred E. Avni, Michelle Hall, Frank Collier, and Claude Schulman

CONTENTS

5.1 Introduction 89
5.2 Imaging the Pelvis and Ureter 90
5.3 Anomalies of the Pelvis and Ureter in Single and Bifid Collecting Systems 91
  5.3.1 Calyceal Diverticulum 91
  5.3.2 Hydrocalyx, Fraley's Syndrome and Infundibular Stenosis 91
  5.3.3 Megacalycosis 93
  5.3.4 Ureteropelvic Obstruction 95
    5.3.4.1 Diagnosis of Obstruction 95
    5.3.4.2 Etiology of Ureteropelvic Junction Obstruction 98
    5.3.4.3 Clinical Presentations 98
    5.3.4.4 Particular Forms of UPJ Obstruction 98
    5.3.4.5 Differential Diagnosis of UPJ Obstruction 101
    5.3.4.6 The Natural History and Treatment of Neonatal UPJ Obstruction 101
    5.3.4.7 Progression of Obstruction 102
  5.3.5 Megaureter and Hydroureter 102
    5.3.5.1 Primary Megaureter 102
    5.3.5.2 Refluxing Megaureter 103
    5.3.5.3 Nonrefluxing Nonobstructive Megaureter 103
    5.3.5.4 Secondary Hydroureter 103
    5.3.5.5 The Natural History of Primary Megaureter 106
  5.3.6 Ureteral Wall Lesions and Look-Alike 106
    5.3.6.1 Ureterocele 107
    5.3.6.2 Bifid Collecting Systems 107
  5.4 Duplex Collecting Systems 109
    5.4.1 Etiology and Epidemiology 109
    5.4.2 Presentation and Circumstances of Diagnosis 109
    5.4.3 The Work Up of Duplex Kidneys: General Considerations 110
    5.4.4 Duplication and VUR 110
    5.4.5 Ureteral ectopia 110
    5.4.6 Ureterocele 111
    5.4.7 (Cystic) Dysplasia of the Upper Pole 116
    5.4.8 Other Types of Obstruction in Duplex Kidney 116
  5.5 Triplication and Quadruplication of the Ureter 117
  5.6 Conclusion 117
5.7 References 118

5.1 Introduction

Congenital anomalies may involve any level of the collecting system; the most usual presentation is urinary tract dilatation that may already be detected during fetal life. The role of imaging is to determine the origin of the dilatation, i.e., obstructive versus non-obstructive (Table 5.1 lists the causes of urinary tract dilatation). Other useful information includes the level of the obstruction and its impact on renal function. All these data are important in order to determine the best therapeutic approach.

Two imaging techniques have been classically used in order to demonstrate the morphology of the collecting system: ultrasound (US) and intravenous urography (IVU). Newer techniques have merged these last years: computed tomography (CT) and MR urography; their aim is to complete the information brought by US and to replace IVU. In many indications, the newer techniques have progres-
sively replaced the older ones. The work-up of most anomalies has to be completed by voiding cystourethrography (VCU) and functional isotope studies.

Table 5.1. Causes of urinary tract dilatations

<table>
<thead>
<tr>
<th>Causes of urinary tract dilatations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Congenital etiologies</strong></td>
</tr>
<tr>
<td>Ureteropelvic junction obstruction</td>
</tr>
<tr>
<td>Ureterovesical junction obstruction</td>
</tr>
<tr>
<td>Vesicoureteric reflux (grades III–V)</td>
</tr>
<tr>
<td>Nonobstructive nonrefluxing megaureter</td>
</tr>
<tr>
<td>Duplex collecting system</td>
</tr>
<tr>
<td>Posterior urethral valves</td>
</tr>
<tr>
<td>Megacalycosis</td>
</tr>
<tr>
<td>Pelvi-infundibular stenosis</td>
</tr>
<tr>
<td><strong>Secondary etiologies</strong></td>
</tr>
<tr>
<td>Tumoral involvement</td>
</tr>
<tr>
<td>Extrinsic compression</td>
</tr>
<tr>
<td>Retroperitoneal fibrosis</td>
</tr>
<tr>
<td>Constipation</td>
</tr>
<tr>
<td>Megabladder</td>
</tr>
<tr>
<td>Lithiasis</td>
</tr>
</tbody>
</table>

5.2 Imaging the Pelvis and Ureter

Historically, IVU was the first imaging technique that allowed the visualization of the collecting system; after iodinated contrast injection, the pyelocalyceal system, the ureter and the bladder opacify, and this allows a morphological evaluation of the collecting system. The best results are obtained when the collecting system is not too dilated and when good renal function is maintained. This technique is rapidly disappearing because of its drawbacks; it is an irradiating technique, and iodinated contrast material has to be injected. Also, if the renal function has deteriorated, opacification will not be optimal and the information provided will be insufficient (Almen and Mattson 1995; Hilton and Kaplan 1995).

In pediatric urology, US has maintained a central position; whatever the anomaly, it will be performed first and will determine the subsequent work up. US is very efficient for the demonstration of dilatation of the urinary tract and the level of obstruction; yet, the method cannot differentiate between obstructive and nonobstructive dilatation. Also, the degree of dilatation is influenced by the state of hydration of the patient; therefore, some teams advocate the use of furosemide and measurement of the resistive index in order to diagnose obstruction. Another interest of US is that the technique also provides information on the renal parenchyma (Chopra and Teele 1980; Patriquin 1991; Gilbert et al. 1993; Haller and Cohen 1987; Bude et al. 1992; Bude et al. 1994; Palmer 2006).

CT has proved informative in many pathological or doubtful situations involving the pyelocalyceal and ureteral system; it may demonstrate the connections of atypical cystic parenchymal lesions with the collecting system, and it may determine the primitive or secondary origin of an obstruction. CT completes the information given by US; if necessary 2D or 3D reconstructions or urographic images post-contrast injection may also be obtained. The technique is irradiating, and therefore its use must be well thought-out and cautious (Berdon 1991; Hilton and Kaplan 1995; Palmer 2006).

These last years, the use of MR urography has been gaining popularity for the visualization of the urinary tract, both the parenchyma and collecting system. Its best indications are the morphological assessment of the very dilated urinary tract, ectopic ureteral insertion, and assessment of renal parenchymal damage. The combination of hydrographic sequences and gadolinium-enhanced sequences provides information on both morphology and (indirect) function. In the near future, further studies will determine whether MR imaging can be considered as an “all in one” examination and will replace both IVU and isotopes (Roy et al. 1998; Borthne et al. 1999; Wille 2003; Jones 2004; Rohrschneider 2003; Boss 2006; Nolte-Ernstig et al. 1998).

Till then, the best functional examination of renal function and of the degree of renal obstruction is obtained by isotope studies with furosemide injection (Berdon 1991; Roarke and Sandler 1998; Piepz and Ham 2006; Palmer 2006). Other techniques such as ascending pyelography have almost completely been abandoned.

Conclusion

US is the central imaging technique for the visualization of a dilated collecting system. Morphology of the urinary tract is best assessed by MR imaging or CT (IVU is more and more abandoned). Function is best assessed by isotope studies.