4 Perinatal Urology

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The increased use of maternal-fetal ultrasound has led to the development of the field of perinatal urology. Antenatal hydronephrosis (ANH) is identified in approximately 1–3% of all pregnancies and is one of the most common birth defects detected (1–4). Other urologic abnormalities have been diagnosed prenatally as well, including renal cystic disease, renal agenesis, stones and tumors. For the pediatric urologist, these prenatal findings have created numerous clinical dilemmas that challenge our understanding of normal and abnormal renal embryology and physiology. In this chapter, we discuss the diagnosis of prenatal urologic abnormalities and the postnatal implications, the rationale behind prenatal intervention, and our clinical experience in managing children with prenatal urologic abnormalities.

Diagnosis

In a large prospective Swedish, study, the incidence of prenatally detected renal anomalies was 0.28% in which two-thirds (0.18%) were hydronephrosis (5). A British study, in which 99% of the pregnant population in Stoke-on-Trent were scanned at 28 weeks' gestation, demonstrated hydronephrosis prenatally in 1.40% of cases, which was confirmed postnatally in 0.65% (3). These authors defined prenatal hydronephrosis as an anteroposterior (A–P) diameter of the renal pelvis greater than 5 mm but noted the lack of consensus on the definition of antenatal hydronephrosis (6–8). Many variations in the definition and management of ANH exist in the literature and clinical practice, including method and frequency of in utero testing, radiographic documentation, classification, or postnatal management (9–15).

When an abnormality of the urinary tract is determined by maternal-fetal ultrasound, several questions should be raised by the ultrasonographer and consulting urologist. Combinations of specific findings direct the differential diagnosis and permit more accurate prognosis and tailoring of postnatal evaluation. The principal findings and their implications are listed in *Table 4-1*.

Diagnostic Accuracy

As both ultrasound and MRI technology improve, more accurate radiographic information is obtainable (16). However, predicting accurate postnatal diagnosis and outcome, regardless of the prenatal information, still remains challenging. The importance of accurate diagnosis is particularly critical in cases where fetal intervention is considered, such as posterior urethral valves (PUV). In other cases, the ability to identify some degree of ANH is adequate to permit a postnatal evaluation if deemed clinically appropriate.

A recent systematic review of the ANH literature attempted to determine the risk of a pathological diagnosis for patients with varying severity of ANH (15). In this review of 1,308 patients with any ANH and postnatal radiographic follow-up, 36% had a postnatal pathological diagnosis. The degree of ANH was defined by the anterior posterior diameter (APD) identified in a particular trimester (*Table 4-2*) (15). The overall risk for any pathology increased with the degree of hydronephrosis; except for vesicoureteral reflux which remained consistent regardless of the degree of ANH (*Table 4-3*) (15).

Although the risk of pathology with degrees of ANH appears to be increased, accurately determining the diagnosis remains difficult (15). An early report by Hobbins et al. suggested that the correct prenatal identification of the site of obstruction could be confirmed postnatally in 88% of cases (17). Subsequent studies reported fairly high false-positive rates ranging from 9 to 22% (6). The majority of false positives in these studies were nonobstructive causes of hydronephrosis, such as high-grade reflux, large, nonobstructed, extrarenal pelves, or transient hydronephrosis. Similarly, the diagnosis of vesicoureteral reflux is extremely challenging to make in-utero, as evidenced by the fact that the risk for vesicoureteral reflux is the same regardless of the degree of ANH (15).

As another example, the accurate diagnosis of posterior urethral valves (PUV), in which intervention might be considered, has proven difficult. In one series, the false-positive rate was as high as 58%, but the criteria for

Table 4–1

Major diagnostic findings in prenatal imaging

	Finding	Comment
Kidney	Hydronephrosis	Assess degree
	Unilateral & bilateral	May be different degrees
	Parenchymal echogenicity	Should be less than spleen or liver; if increased and organ enlarged, suggests autosomal recessive polycystic kidney disease
	Duplication	Often with dilation of upper pole; may be lower pole dilation
	Cysts	Small cysts associated with dysplasia; simple cyst of upper pole suggests duplication with ureterocele or ectopic ureter; genetic cystic disease
	Urinoma	Perinephric or subcapsular
Ureter	Dilation/tortuosity	Obstruction or reflux
Bladder	Distended	Variation with time
	Wall thickness	In relation to filling status
	Intravesical cystic structure	Ureterocele
	"Keyhole" pattern	Dilated posterior urethra; PUV
	Not visible	Exstrophy
Amniotic fluid	Absence; oligohydramnios	Impaired urine output
	Polyhydramnios	May be seen with mild-moderate hydronephrosis
Gender	Penis/scrotum/ testes	Sex-associated conditions (e.g., PUV)
Spine	Meningocele	Neural tube defect

PUV, posterior urethral valves

Table 4–2

Classification of antenatal hydronephrosis (ANH) by anterior posterior diameter (APD)

ANH Classification	APD	
	2nd Trimester (mm)	3rd Trimester (mm)
1. Mild	≤7	≤9
2. Mild/Moderate	<10	<15
3. Moderate	7–10	9–15
4. Moderate/Severe	≥7	≥9
5. Severe	≥10	≥15

diagnosing valves were quite liberal and perhaps inappropriate (18). In another population-based series, the sensitivity in detecting valves was as low as 23% (6). Increased renal echogenicity and decreased amniotic fluid have been suggested to be indicative of obstructive conditions (19). Although the hallmark signs of an in-utero diagnosis of posterior urethral valves have been described (oligohydramnios, dilated posterior urethra, thickened bladder, and hydroureteronephrosis), there are very few studies that have prospectively examined the clinical urologic implications of these findings alone or in combination (15).

Ureteropelvic Junction Obstruction

The basic features of ureteropelvic junction obstruction (UPJO) in the fetus include dilation of the renal pelvis and collecting system with no evidence of ureteral dilation (*Fig. 4-1*). The best way to detect ureteral dilaion is at the level of the bladder, preferably in transverse view. The threshold for recommending postnatal follow-up is largely arbitrary and currently there are no long-term prospective studies to best determine the degree of postnatal evaluation. Lee et al. demonstrated that increasing severity of ANH increased the chance of identifying postnatal UPJO (15). Others have recommended that unilateral APD over 7 or 8 mm in the third trimester

Table 4-3

Risk of pathology by degree of antenatal hydronephrosis

Postnatal Pathology [% (95% CI)/[a]]	Degree of Antenatal Hydronephrosis					
	Mild (*N* = 587)	Mild-Moderate (*N* = 213)	Moderate (*N* = 235)	Moderate-Severe (*N* = 179)	Severe (*N* = 94)	Trend P-value[b]
Any Pathology	11.9 (4.5, 28.0)	39.0 (32.6, 45.7)	45.1 (25.3, 66.6)	72.1 (47.6, 88.0)	88.3 (53.7, 98.0)	<0.001
UPJ	4.9 (2.0, 11.9)	13.6 (9.6, 18.9)	17.0 (7.6, 33.9)	36.9 (17.9, 61.0)	54.3 (21.7, 83.6)	<0.001
VUR	4.4 (1.5, 12.1)	10.8 (7.3, 15.7)	14.0 (7.1, 25.9)	12.3 (8.4, 17.7)	8.5 (4.7, 15.0)	0.10
PUV	0.2 (0.0, 1.4)	0.9 (0.2, 3.7)	0.9 (0.2, 2.9)	6.7 (2.5, 16.6)	5.3 (1.2, 21.0)	<0.001
Ureteral Obstruction	1.2 (0.2, 8.0)	11.7 (8.1, 16.8)	9.8 (6.3, 14.9)	10.6 (7.4, 15.0)	5.3 (1.4, 18.2)	0.025
Other[c]	1.2 (0.3, 4.0)	1.9 (0.7, 4.9)	3.4 (0.5, 19.4)	5.6 (3.0, 10.2)	14.9 (3.6, 44.9)	0.002

[a]Pointwise 95% confidence intervals were estimated by logistic regression with robust standard errors based on generalized estimating equations with a working independence correlation structure to adjust for clustering by study for all degrees of antenatal hydronephrosis except mild-moderate. Because only one study had subjects with mild-moderate antenatal hydronephrosis, the pointwise 95% confidence intervals had to be estimated using logistic regression with unadjusted standard errors

[b]Testing for trend in risks with increasing degree of antenatal hydronephrosis using logistic regression with robust standard errors based on generalized estimating equations with a working independence correlation structure

[c]Includes prune belly syndrome, VATER syndrome, solitary kidney, renal mass, and unclassified

(5–6 mm with bilateral dilation) warrants postnatal follow-up (20). Some have recommended that all children with any ANH should be investigated postnatally (21). Nevertheless, in the case of significant unilateral hydronephrosis, there is little rationale for in utero intervention. In a few cases with massive dilation, therapeutic aspiration has been recommended for dystocia. In the case of bilateral UPJO, the efficacy of in utero intervention is difficult to assess.

Attempts to correlate prenatal ultrasound appearance with postnatal outcomes have been complicated by the long-standing controversy regarding postnatal evaluation and management of UPJO. Grignon et al. developed a system of grading hydronephrosis secondary to UPJO based on the APD and degree of calyceal dilatation (22, 23). Mandell et al. attempted to correlate the degree of APD relative to gestational age with subsequent need for postnatal surgical intervention (24). They found the "at risk" diameter to be greater than or equal to 5 mm at 15–20 weeks' gestation, greater than or equal to 8 mm at 20–30 weeks' gestation, and greater than 1 cm at over 30 weeks' gestation. An alternative system proposed by Kleiner et al. defined hydronephrosis as the ratio of APD to A-P diameter of the kidney as being greater than 0.5 cm (25); caliectasis was later added as an additional indicator of significant hydronephrosis. Mild degrees of renal pelvic dilatation may resolve in utero. Mandell et al. noted this occurred in 23% of cases, with 66% remaining stable and 9% worsening over the course of the pregnancy (24). Similarly, Lee et al. noted that 88.1% of mild ANH were found to have no postnatal pathology (15). Severe forms of UPJO may be associated with urinary ascites or perinephric urinomas, which often precede nonfunction of the kidney (26).

Cystic Kidneys

The distinction between severe unilateral hydronephrosis and a multicystic dysplastic kidney may occasionally be unclear. The findings of multiple noncommunicating cysts, minimal or absent renal parenchyma, and the absence of a central large cyst are diagnostic of a multicystic dysplastic kidney (MCDK) (*Fig. 4-2*). Bilaterally enlarged echogenic kidneys, particularly if associated with hepatobiliary dilatation or oligohydramnios, suggests autosomal recessive polycystic kidney disease (*Fig. 4-3*). A more challenging finding is normal-sized, diffusely echogenic kidneys that are not associated with other urologic

Figure 4–1

Unilateral renal pelvis dilation with no ureteral dilation in 37-week old fetus.

lesions. Estroff et al. described 19 cases (14 bilateral), including 10 with normal function who survived and 4 with autosomal recessive polycystic kidneys who died (14).

Ureterovesical Junction Obstruction

Less common than UPJO, ureterovesical obstruction (UVJ) is characterized by ureteral dilation along with varying degrees of renal pelvic and calyceal dilation (*Fig. 4-4*). More extreme cases may be confused with single system ectopic ureters, particularly in males. In general, the differentiation is made postnatally.

Duplication Anomalies

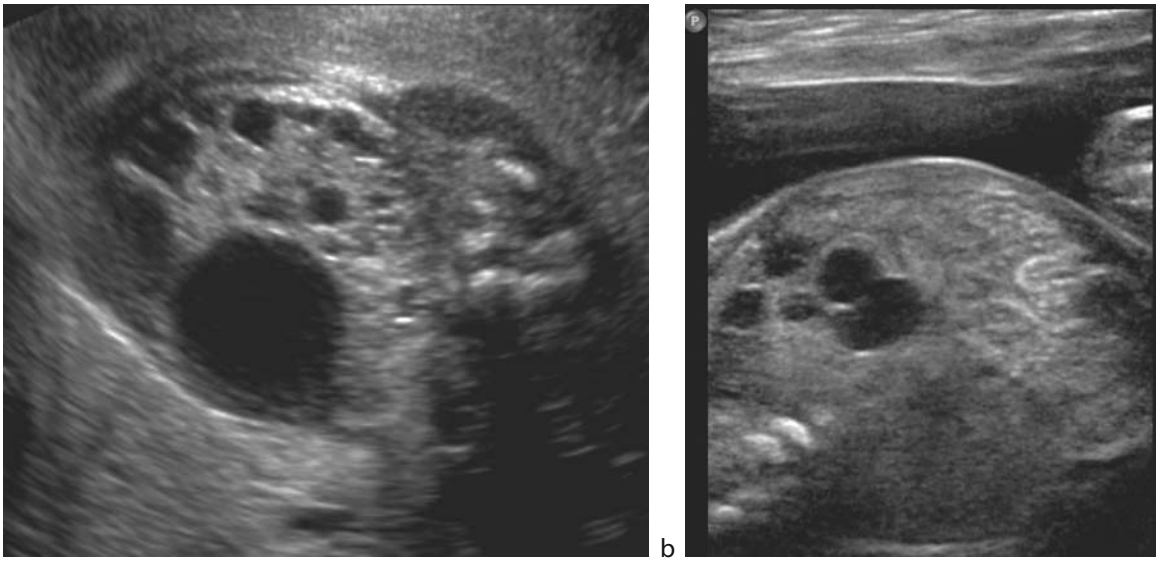
Among the most interesting prenatal urologic findings are duplication anomalies. These are often recognized on the basis of upper pole hydroureteronephrosis, associated with either an obstructing ureterocele within the bladder, or ectopic ureter inserting outside of the bladder (27) (*Fig. 4-5*). Lower pole hydronephrosis may be present as a result of vesicoureteral reflux or more rarely a lower pole UPJO. Occasionally, lower pole dilation is due to obstruction of both the upper and lower pole ureter by the large ureterocele. In some cases of a very large ureterocele, the ureterocele may be mistaken for the bladder.

Vesicoureteral Reflux

One cannot make a firm diagnosis of vesicoureteral reflux (VUR) based on prenatal ultrasound, although intermittent hydronephrosis or hydroureter is highly suggestive. Vesicoureteral reflux may be present in as many as 38% of children with prenatal hydronephrosis (28). Reflux occurred in 42% of children in whom postnatal imaging revealed persistent upper tract abnormalities and in 25%

Figure 4–2

Two different fetal images (a) at 19 weeks) and (b) at 26 weeks of a multicystic dysplastic kidney with multiple noncommunicating cysts.



of those with normal findings on postnatal ultrasound but having a history of prenatal dilation. Tibballs and Debrun reported that in patients with prenatal dilation and postnatally normal renal units by ultrasound, 25% had grade III-V reflux (29). The incidence of high-grade reflux was greater in males than in females as noted in previous studies. In two systemic reviews of the ANH literature a 10–15% incidence of VUR was identified regardless of the degree of ANH (15, 30) indicating that ANH is not indicative of VUR and may not be the appropriate trigger for postnatal evaluation. In a neonate with prenatally detected hydronephrosis, the importance of diagnosing vesicoureteral reflux remains controversial. While, several studies have demonstrated that a high incidence of reflux is associated with prenatally detected hydronephrosis, its clinical significance is unclear.

Posterior Urethral Valves

Perhaps the most important diagnosis to be made prenatally is that of PUV in the male fetus. PUV, at the very least, mandates prompt postnatal intervention and in some cases, prenatal intervention may be warranted. Fetal sonographic findings of PUV include bilateral hydroureteronephrosis, a thick-walled bladder with dilated posterior urethra, and, in more severe cases, dysplastic renal parenchymal changes with perinephric urinomas and urinary ascites (*Fig. 4-6*) (31). When characteristic sonographic findings are present, the differential diagnosis includes prune belly syndrome (with or without urethral atresia), massive vesicoureteral reflux, and certain cloacal anomalies (in genetic females) (32, 33). Prenatal diagnostic accuracy for PUV is far from perfect but is probably better than the 40% figure previously reported (18).

Rationale for Prenatal Intervention

The scientific rationale for prenatal treatment of hydronephrosis is to maximize normal development of renal and pulmonary function. These two aspects of fetal development are closely linked because urine comprises 90% of amniotic fluid volume, and oligohydramnios during the third trimester has been causally related to pulmonary hypoplasia.

Before embarking on prenatal surgical intervention for obstructive uropathy, it is critical to assess the risk-benefit ratio. The most widely accepted indicator of salvageable renal function is analysis of fetal urine. When the urinary sodium is less than 100 mg/dL and urine osmolarity less than 200 mOsm/dL, renal function appears to be salvageable with in utero intervention (*Table 4-4*) (34). The accuracy of these predictors has been challenged (35, 36). More recently, serial aspirations of fetal urine have been reported to yield more valuable results (37). Guez et al. reported ten fetuses who underwent multiple urine samplings and in whom severe obstruction reduced sodium and calcium reabsorption (38).

Figure 4–3

Bilateral markedly enlarged echogenic (“bright”) kidneys (a) with a small cystic lesion (b) in a fetus with oligohydramnios, consistent with autosomal recessive polycystic kidney disease.

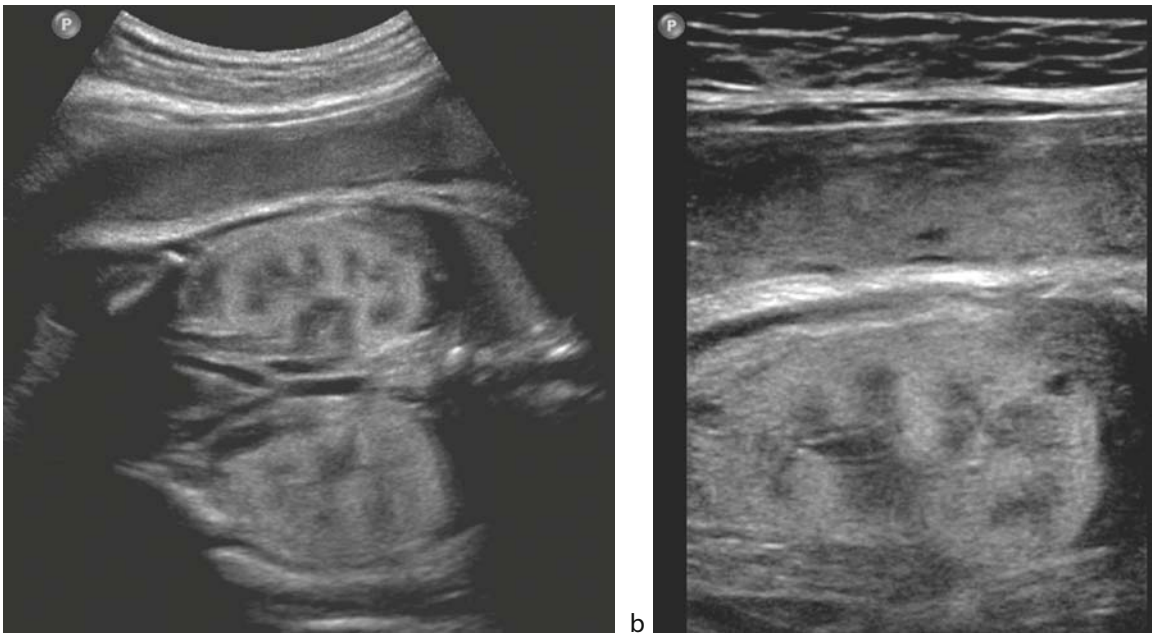
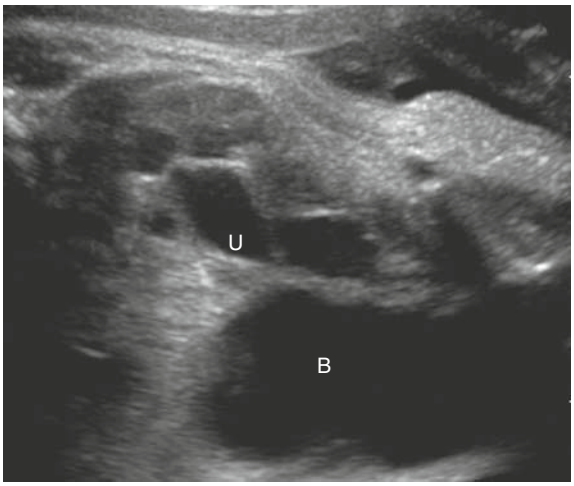


Figure 4–4

Unilateral hydroureteronephrosis at 35 weeks. Note the dilated ureter (U) and bladder (B). Postnatal imaging confirmed this to be a ureterovesical junction obstruction.



They concluded that fetal urinary chemistries were reasonably predictive of severe but not moderate postnatal renal impairment. Other investigators have suggested the use of fetal urinary beta-$\tilde{}_{22}$ microglobulin as an indicator of tubular damage. Using this parameter, poor renal-outcome has been predicted with a specificity of 83% and sensitivity of 80% (39).

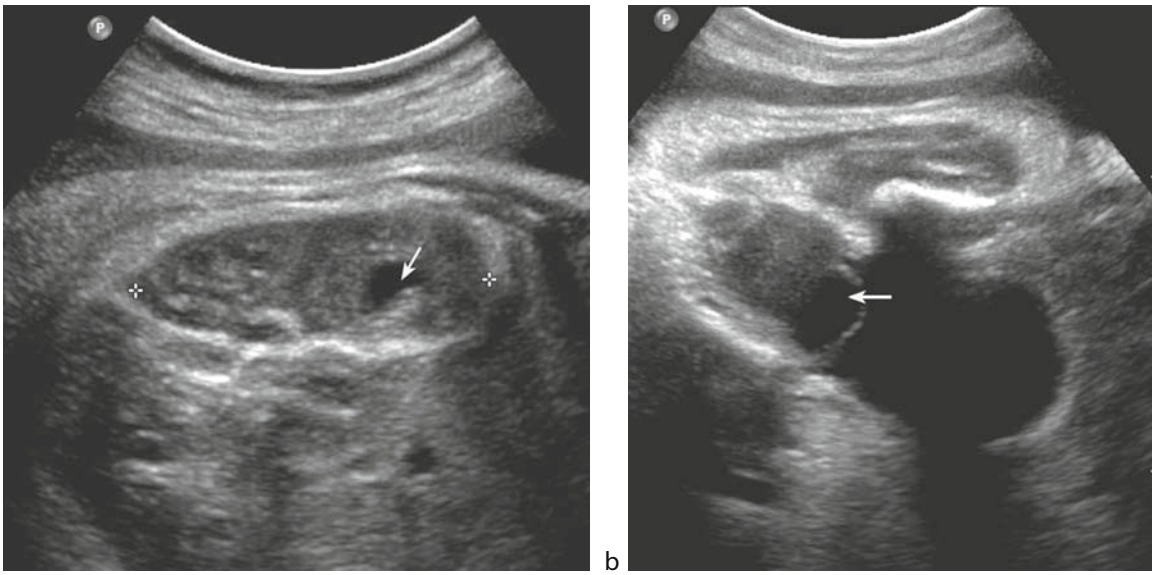
The time of onset of oligohydiamnios has been shown to be an important determinant of outcome (40, 41). In fetuses in which adequate amniotic fluid was documented at up to 30 weeks’ gestation in association with a urologic abnormality, pulmonary outcomes were satisfactory, and postnatal clinical problems were related to renal disease. It seems inappropriate to recommend late urinary tract decompression from a pulmonary or renal basis. It is unclear whether early delivery, to permit earlier postnatal urinary decompression, is beneficial.

Clinical Experience with Intervention for Prenatal Hydronephrosis

The ability to diagnose severe prenatal hydronephrosis and advances in fetal intervention helped develop prenatal surgery for obstructive uropathy. In 1982, Harrison et al. described the initial report of fetal surgery in a 21-week-old fetus with bilateral hydroureteronephrosis secondary to PUV (42). After the 1986 report of the International Fetal Surgery Registry in which outcomes did not seem to justify risk, a de facto moratorium on in utero urinary tract shunting evolved (43). More recently, with improved technology and renewed interest in fetal shunting, most

Figure 4–5

Fetal image of duplex kidney with marked upper pole hydronephrosis (arrow) in contrast to a normal lower pole (a). Associated with this image is the finding of a ureterocele (arrow) within the bladder (b).



cases have been referred to a small number of highly specialized centers actively engaged in prenatal surgery. The initial method of decompression with open surgery has largely been replaced by in utero shunt placement, although this has been complicated by technical problems of shunt dislodgement and, in the case of the double-J shunt, bowel herniation (44). Some investigators have explored the use of fetoscopic methods for direct intervention to provide prolonged bladder drainage, whereas others have attempted direct endoscopic valve ablation (45–49).

Harrison et al. have clearly outlined the indications and contraindications of intervention for prenatal obstructive uropathy (*Table 4-5*) (50). Additionally, serial bladder sampling over 3 days has been used to help determine if the fetus is a viable candidate. The serial nature of the procedure allows one to see the response of the fetal kidneys to bladder decompression (37). The principal reason for considering vesicoamniotic shunting is to prevent early neonatal pulmonary insufficiency and death. The risks that one accepts with intervention include induction of premature labor, perforation of fetal bowel and bladder, and fetal and/or mother hemorrhage and infection.

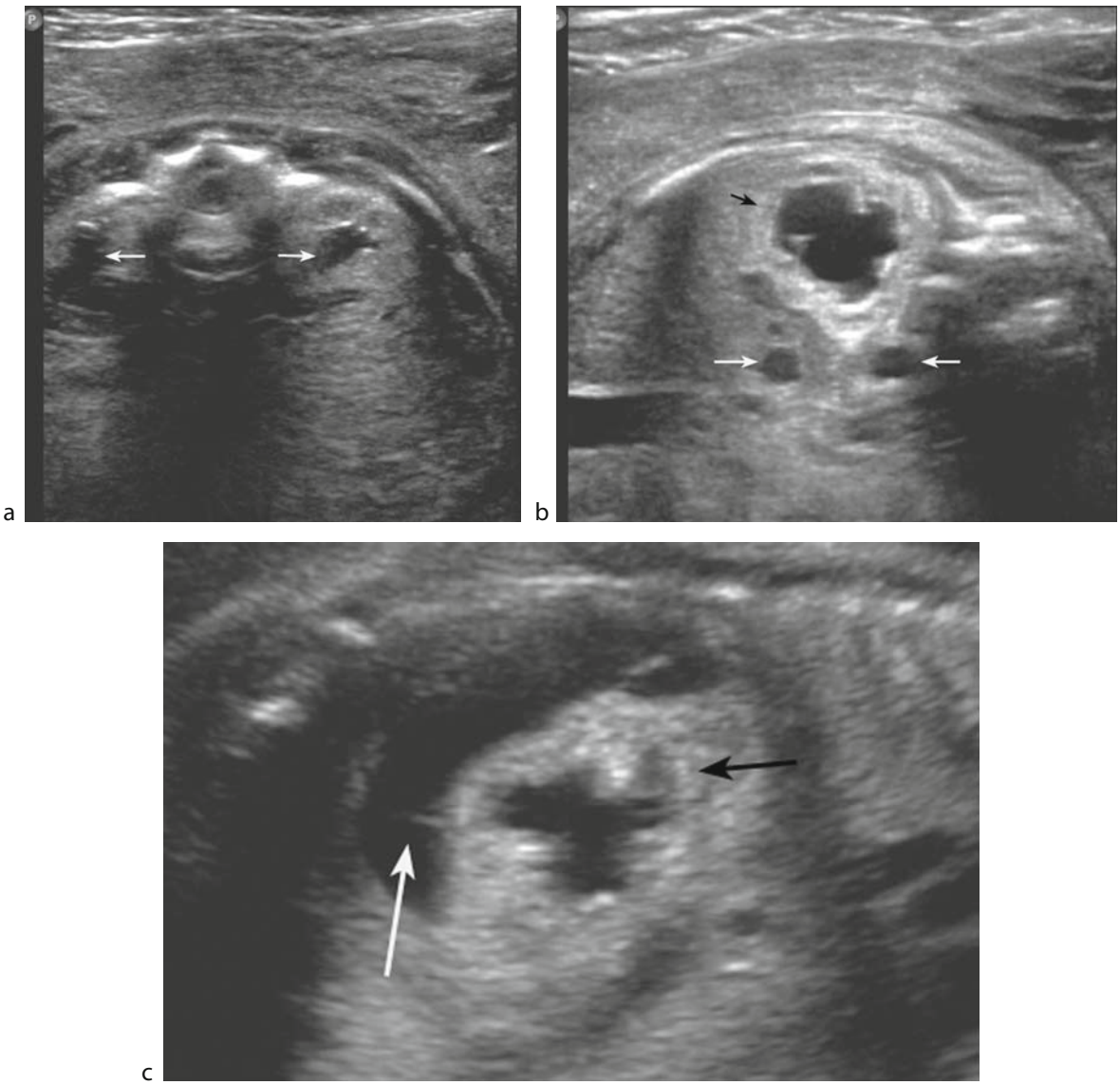
More recently, the ability to influence renal outcome in male patients with PUV but without oligohydramnios has been suggested as a possible indication for in utero intervention. The principal goal of intervention is not to prevent pulmonary hypoplasia and deaths but to prevent or delay end-stage renal failure. Although some reports have shown promise in the ability to distinguish those fetuses with likely early renal failure from those with later-onset failure, the specificity and accuracy of methods using a combination of ultrasound and urinary chemistries (sodium, beta$_2^{\sim}$ microglobulin, and calcium) has not been well defined (51–53). In summary, precise identification of those situations in which intervention may benefit the fetus with obstructive uropathy remains unclear.

Overall, the need to consider in utero intervention for obstruction is not common. In one study, only 9 of 177 fetuses with a diagnosis of hydronephrosis were considered to have PUV and only 3 warranted serious consideration for intervention (24).

To date, the reported long term outcomes of antenatal intervention for severe obstructive uropathy (e.g. PUV, prune belly syndrome, urethral atresia) are mixed (54–62). Variability in patient selection and assessment of outcome within these studies has limited the ability to determine if prenatal intervention has altered the postnatal course. A large systematic review of the prenatal intervention for obstructive uropathy showed a statistically significant perinatal survival advantage with shunting (60). Of the studies that have reported long term outcomes of in-utero vescioamniotic shunting, many of the children have renal insufficiency (57%), and growth impairment (86%) (54, 56, 57). Recently, Baird et al., reported on long-term follow-up (5.8 years) of patients who have survived in-utero shunting (54). They noted

Figure 4–6

Images of a fetus with posterior urethral valves. (a) depicts bilateral echogenic kidneys with hydroureteronephrosis (arrow). (b) demonstrates the associated thickened bladder wall (black arrow) and hydroureter (white arrow). (c) further imaging revealed a perinephric urinoma (white arrow) surrounding the hydronephrotic kidney (black arrow).



acceptable renal function in 44%, mild impairment in 22%, and renal failure in 33%. Prune belly patients had the best renal outcome (57%), followed by PUV (43%), then urethral atresia (25%). Overall, it appears that in-utero intervention for the appropriate patient may reduce the risk of neonatal mortality and may potentially improve renal function. To further improve outcomes, more sensitive and specific markers to better identify which fetus will benefit from in-utero shunting need to be defined.

Postnatal Management of Infants with Prenatally Diagnosed Urologic Renal Abnormality

A child with a prenatal diagnosis of a urologic renal abnormality such as ANH should be carefully evaluated and followed by a pediatric urologist from birth. The vast majority of these children appear entirely healthy and, in the absence of prenatal ultrasound findings, would not have

Table 4–4

Prenatal assessment of renal functional prognosis

	Good		Poor	
Amniotic fluid	Normal to moderately decreased		Moderate to severely decreased	
Sonographic appearance of kidneys	Normal to echogenic		Echogenic to cystic	
Fetal urine	Glick et al. (21)	Johnson et al. (24)	Glick et al. (21)	Johnson et al. (24)
Sodium (mEq/L)	<100	<100	>100	>100
Chloride (mEq/L)	<90	–	>90	–
Osmolarity (mOsm/L)	<210	<200	>210	>200
Calcium (mg/dL)	–	<8	–	>8
β_2-Microglobulin (mg/L)	–	<4	–	>4
Total protein (mg/dL)	–	<20	–	>20
Output (mL/h)	>2	–	<2	–
Sequential improvement in urinary values	–	X	–	–

X, only in this series was the criterion used

Table 4–5

Prenatal intervention for hydronephrosis

Indications (prerequisites)	Contraindications
Presumed obstructive hydronephrosis, persistent or progressive, bilateral or insolitary unit	Unilateral hydronephrosis with an adequately functioning contralateral kidney
Otherwise healthy fetus	Chromosomal abnormalities or presence of associated severe anomalies
Oligohydramnios	Bilateral hydronephrosis without oligohydramnios
No overt renal dysplasia	Severely dysplastic kidneys
Adequate renal functional potential based on urinary indices (see text)	Evidence or uretheral atresia
Informed consent	Presence of a normal twin

any indications for regular urologic follow-up. Parental anxiety is common and should be addressed directly with prenatal counseling and education.

Unilateral Hydronephrosis

The presence of unilateral dilation of the kidney warrants postnatal evaluation in a timely but non urgent fashion (3–8 weeks of life) with an ultrasound (51). The most common diagnoses associated with this finding are UPJO, VUR and UVJO/megaureter. Early ultrasound is unlikely to miss a significant abnormality. A normal postnatal ultrasound indicates that obstructive uropathy is not present; however, it does not determine whether or not the child has VUR (29).

The decision to perform a voiding cystourethrogram (VCUG) or initiate prophylactic antibiotics in the newborn period is unclear. Although some groups advocate postnatal VCUG in any child with a history of prenatal hydronephrosis, others have questioned the value of this approach (63). Infants with severe ANH should be placed on prophylactic antibiotics (amoxicillin, 10 mg/kg/day or 50 mg/day) and undergo a VCUG. Severe ANH may be associated with an increased risk of febrile urinary tract infection (64). As for mild ANH, a recent prospective study of 192 infants with ANH noted that the majority of patients with mild ANH had no significant events during infancy (65). In another study, female infants with a history of ANH and postnatal uropathy had a higher risk of febrile urinary tract infection (66). Regardless, no appropriate prospective studies with coordinated and comprehensive postnatal follow-up have examined this question in a rigorous fashion to provide consensus guidelines (15, 30).

At our institution, children with moderate or severe ANH are placed on prophylactic antibiotics at birth. They undergo renal bladder ultrasound and VCUG postnatally. Diuretic renography is reserved for those with persistent moderate or severe postnatal hydronephrosis not related to VUR. Infants with persistent mild on serial US or no postnatal hydronephrosis are observed and followed clinically. Infants with any degree of antenatal or postnatal

ureteral dilation undergo ultrasound, VCUG and possibly diuretic renography (after 3 months) if clinically indicated.

Perhaps the most challenging aspect of managing prenatal hydronephrosis is determining if and when postnatal surgical correction for obstruction is appropriate. Some have suggested that regardless of the degree of ANH, moderate or severe postnatal hydronephrosis with evidence of decreased renal function should be the indications for surgical intervention (67). Despite the improved anatomic detail afforded by real-time ultrasound and the increasing experience with functional nuclear medicine studies (mercaptotriglycylglycine) no radiographic or clinical gold standard for physiologically significant obstruction exists. Over time, some kidneys have been seen to improve, whereas others appear to lose function. The natural history of prenatal hydronephrosis is not clearly defined.

The debate over the appropriate management of infants with unilateral ANH continues and may ultimately be determined by a combination of epidemiologic, radiographic, and new innovative biomarker discoveries. More accurate and reproducible prenatal and postnatal radiographic documentation of the degree of hydronephrosis and function combined with appropriate natural history data are needed to better categorize the infants. Finally, new serum or urine biomarkers indicative of ongoing renal damage will be critical in helping to further define which infants are truly at risk.

Bilateral Hydronephrosis

Infants with bilateral hydroureteronephrosis may have PUV, bilateral VUR, bilateral UPJO or UVJO, or a combination of the above. For the child with bilateral hydroureteronephrosis suggestive of bladder outlet obstruction, an ultrasound and VCUG should be performed promptly. In boys, PUV is the most important diagnosis to be ruled out. In girls, an obstructing ectopic ureterocele would be the most likely cause for bladder outlet obstruction. In the event that an obstructive lesion is discovered, it should be corrected promptly. For children with suspected lower urinary tract obstruction (e.g. PUV), prompt bladder decompression and antibiotic prophylaxis (amoxicillin 10 mg/kg/day or 50 mg/day) should be initiated prior to radiographic intervention.

Renal Agenesis, Renal Ectopia and Unilateral Multicystic Dysplastic Kidney

Infants born with solitary kidneys (renal agenesis), renal ectopia or unilateral multicystic dysplasia should be evaluated postnatally by US and VCUG. Functional studies such as a dimercaptosuccinic acid study (DMSA) are occasionally needed to confirm the diagnosis. The need for further screening is controversial. It has been reported that among infants with a solitary kidney, 30% have VUR, 11% UPJO, and 7% UVJO (68, 69). Similarly, those with renal ectopia (simple or crossed fused ectopia) may also be at risk for VUR in the ectopic or contralateral kidney (30%) (70–72). However, there are others that report a very low incidence of associated urologic anomalies and do not recommend screening (73).

Multicystic dysplastic kidneys (MCDK) are primarily unilateral, isolated and associated with a good prognosis. If at birth the US findings are not absolutely diagnostic of a classic MCDK, a DMSA study can be used to confirm the diagnosis with the absence of uptake. Patients with MCDK are often thought to be similar to those born with a solitary kidney. Additionally, patients with MCDK have an increased frequency of VUR and UPJO in the contralateral normal kidney (74, 75).

Summary

With the increased use of maternal-fetal ultrasound, more genitourinary abnormalities are being detected prenatally. Although advances in imaging have increased the detection and characterization of these abnormalities, further work is needed to identify which abnormalities are clinically significant. Research directives should focus on identifying which infants require postnatal diagnostic imaging and intervention. Developments in the fields of imaging, proteomics, and genomics may provide the necessary information to not only detect the abnormality, but to also prognosticate which ones require further testing and medical intervention.

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References

1. Blyth B, Snyder HM, Duckett JW. Antenatal diagnosis and subsequent management of hydronephrosis. *J Urol* 1993;149(4):693–698.
2. Gunn TR, Mora JD, Pease P. Antenatal diagnosis of urinary tract abnormalities by ultrasonography after 28 weeks' gestation: incidence and outcome. *Am J Obstet Gynecol* 1995;172(2 Pt 1):479–486.

3. Livera LN et al. Antenatal ultrasonography to detect fetal renal abnormalities: a prospective screening programme. *BMJ* 1989;298 (6685):1421–1423.
4. Sairam S et al. Natural history of fetal hydronephrosis diagnosed on mid-trimester ultrasound. *Ultrasound Obstet Gynecol* 2001;17(3): 191–196.
5. Helin I, Persson PH. Prenatal diagnosis of urinary tract abnormalities by ultrasound. *Pediatrics* 1986;78(5):879–883.
6. Scott JE, Renwick M. Urological anomalies in the Northern Region Fetal Abnormality Survey. *Arch Dis Child* 1993;68 (1 Spec No):22–26.
7. Scott JE, Renwick M. Screening for fetal urological abnormalities: how effective? *BJU Int* 1999;84(6):693–700.
8. Scott JE et al. Measuring the fetal kidney with ultrasonography. *Br J Urol* 1995;76(6):769–774.
9. Benacerraf BR et al. Fetal pyelectasis: a possible association with Down syndrome. *Obstet Gynecol* 1990;76(1):58–60.
10. Corteville JE, Dicke JM, Crane JP. Fetal pyelectasis and Down syndrome: is genetic amniocentesis warranted? *Obstet Gynecol* 1992;79(5 (Pt 1)):770–772.
11. Adra AM et al. Fetal pyelectasis: is it always "physiologic"? *Am J Obstet Gynecol* 1995;173(4):1263–1266.
12. Thompson MO, Thilaganathan B. Effect of routine screening for Down's syndrome on the significance of isolated fetal hydronephrosis. *Br J Obstet Gynaecol* 1998;105(8):860–864.
13. Chudleigh PM et al. The association of aneuploidy and mild fetal pyelectasis in an unselected population: the results of a multicenter study. *Ultrasound Obstet Gynecol,* 2001;17(3):197–202.
14. Fernbach SK, Maizels M, Conway JJ. Ultrasound grading of hydronephrosis: introduction to the system used by the Society for Fetal Urology. *Pediatr Radiol* 1993;23(6):478–480.
15. Lee RS et al. Antenatal hydronephrosis as a predictor of postnatal outcome: a meta-analysis. *Pediatrics* 2006;118(2):586–593.
16. Laifer-Narin S et al. Fetal magnetic resonance imaging: a review. *Curr Opin Obstet Gynecol* 2007;19(2):151–156.
17. Hobbins JC et al. Antenatal diagnosis of renal anomalies with ultrasound. I. Obstructive uropathy. *Am J Obstet Gynecol* 1984;148(7): 868–877.
18. Abbott JF, Levine D, Wapner R. Posterior urethral valves: inaccuracy of prenatal diagnosis. *Fetal Diagn Ther* 1998;13(3):179–183.
19. Kaefer M et al. Increased renal echogenicity: a sonographic sign for differentiating between obstructive and nonobstructive etiologies of in utero bladder distension. *J Urol* 1997;158(3 Pt 2): 1026–1029.
20. Kent A et al. A study of mild fetal pyelectasia - outcome and proposed strategy of management. *Prenat Diagn* 2000;20 (3):206–209.
21. Aksu N et al. Postnatal management of infants with antenatally detected hydronephrosis. *Pediatr Nephrol* 2005;20(9):1253–1259.
22. Grignon A et al. Ureteropelvic junction stenosis: antenatal ultrasonographic diagnosis, postnatal investigation, and follow-up. *Radiology* 1986;160(3):649–651.
23. Grignon A et al. Urinary tract dilatation in utero: classification and clinical applications. *Radiology* 1986;160(3):645–647.
24. Mandell J et al. Structural genitourinary defects detected in utero. *Radiology* 1991;178(1):193–6.
25. Kleiner B, Callen PW, Filly RA. Sonographic analysis of the fetus with ureteropelvic junction obstruction. *AJR Am J Roentgenol* 1987;148(2):359–363.
26. Mandell J et al. Prenatal findings associated with a unilateral nonfunctioning or absent kidney. *J Urol* 1994;152(1):176–178.
27. Vergani P et al. Accuracy of prenatal ultrasonographic diagnosis of duplex renal system. *J Ultrasound Med* 1999;18(7):463–467.
28. Zerin JM, Ritchey ML, Chang AC. Incidental vesicoureteral reflux in neonates with antenatally detected hydronephrosis and other renal abnormalities. *Radiology* 1993;187(1):157–160.
29. Tibballs JM, De Bruyn R. Primary vesicoureteric reflux–how useful is postnatal ultrasound? *Arch Dis Child* 1996;75(5):444–447.
30. van Eerde AM et al. Vesico-ureteral reflux in children with prenatally detected hydronephrosis: a systematic review. *Ultrasound Obstet Gynecol* 2007;29(4):463–469.
31. Peters CA. Lower urinary tract obstruction: clinical and experimental aspects. *Br J Urol* 1998;81(Suppl 2):22–32.
32. Kaefer M et al. The sonographic diagnosis of infravesical obstruction in children: evaluation of bladder wall thickness indexed to bladder filling. *J Urol* 1997;157(3):989–991.
33. Oliveira EA et al. Predictive factors of fetal urethral obstruction: a multivariate analysis. *Fetal Diagn Ther* 2000;15(3):180–186.
34. Glick PL et al. Management of the fetus with congenital hydronephrosis II: Prognostic criteria and selection for treatment. *J Pediatr Surg* 1985;20(4):376–387.
35. Elder JS et al. Evaluation of fetal renal function: unreliability of fetal urinary electrolytes. *J Urol* 1990;144(2 Pt 2):574–578, discussion 593–594.
36. Wilkins IA et al. The nonpredictive value of fetal urinary electrolytes: preliminary report of outcomes and correlations with pathologic diagnosis. *Am J Obstet Gynecol* 1987;157(3):694–698.
37. Johnson MP et al. Sequential urinalysis improves evaluation of fetal renal function in obstructive uropathy. *Am J Obstet Gynecol* 1995;173 (1):59–65.
38. Guez S et al. Shortcomings in predicting postnatal renal function using prenatal urine biochemistry in fetuses with congenital hydronephrosis. *J Pediatr Surg* 1996;31(10):1401–1404.
39. Tassis BM et al. In fetuses with isolated hydronephrosis, urinary beta 2-microglobulin and N-acetyl-beta-D-glucosaminidase (NAG) have a limited role in the prediction of postnatal renal function. *Prenat Diagn* 1996;16(12):1087–1093.
40. Mahony BS, Callen PW, Filly RA. Fetal urethral obstruction: US evaluation. *Radiology* 1985;157(1):221–224.
41. Mandell J et al. Late onset severe oligohydramnios associated with genitourinary abnormalities. *J Urol* 1992;148(2 Pt 2):515–518.
42. Harrison MR et al. Correction of congenital hydronephrosis in utero II. Decompression reverses the effects of obstruction on the fetal lung and urinary tract. *J Pediatr Surg* 1982;17(6):965–974.
43. Manning FA, Harrison MR, Rodeck C. Catheter shunts for fetal hydronephrosis and hydrocephalus. Report of the International Fetal Surgery Registry. *N Engl J Med* 1986;315(5):336–340.
44. Robichaux AG III et al. Fetal abdominal wall defect: a new complication of vesicoamniotic shunting. *Fetal Diagn Ther* 1991;6 (1–2):11–13.
45. Luks FI et al. Access techniques in endoscopic fetal surgery. *Eur J Pediatr Surg* 1997;7(3):131–134.
46. Estes JM et al. Fetoscopic surgery for the treatment of congenital anomalies. *J Pediatr Surg* 1992;27(8):950–954.
47. Kohl T et al. Percutaneous fetal access and uterine closure for fetoscopic surgery. Lessons learned from 16 consecutive procedures in pregnant sheep. *Surg Endosc* 1997;11(8):819–824.
48. Luks FI et al. A model for fetal surgery through intrauterine endoscopy. *J Pediatr Surg* 1994;29(8):1007–1009.
49. Quintero RA et al. Successful in utero endoscopic ablation of posterior urethral valves: a new dimension in fetal urology. *Urology* 2000;55(5):774.

50. Harrison MR et al. Fetal hydronephrosis: selection and surgical repair. *J Pediatr Surg* 1987;22(6):556–558.
51. Clautice-Engle T et al. Diagnosis of obstructive hydronephrosis in infants: comparison sonograms performed 6 days and 6 weeks after birth. *AJR Am J Roentgenol* 1995;164(4):963–967.
52. Muller F et al. Fetal urinary biochemistry predicts postnatal renal function in children with bilateral obstructive uropathies. *Obstet Gynecol* 1993;82(5):813–820.
53. Dommergues M et al. Fetal serum beta2-microglobulin predicts postnatal renal function in bilateral uropathies. *Kidney Int* 2000;58 (1):312–316.
54. Biard JM et al. Long-term outcomes in children treated by prenatal vesicoamniotic shunting for lower urinary tract obstruction. *Obstet Gynecol* 2005;106(3)503–508.
55. Crombleholme TM et al. Fetal intervention in obstructive uropathy: prognostic indicators and efficacy of intervention. *Am J Obstet Gynecol* 1990;162(5)1239–1244.
56. Freedman AL et al. Long-term outcome in children after antenatal intervention for obstructive uropathies. *Lancet* 1999;354 (9176):374–377.
57. Holmes N, Harrison MR, Baskin LS. Fetal surgery for posterior urethral valves: long-term postnatal outcomes. *Pediatrics* 2001;108 (1):E7.
58. McLorie G et al. Outcome analysis of vesicoamniotic shunting in a comprehensive population. *J Urol* 2001;166(3):1036–1040.
59. Salam MA. Posterior urethral valve: Outcome of antenatal intervention. *Int J Urol* 2006;13(10):1317–1322.
60. Clark TJ et al. Prenatal bladder drainage in the management of fetal lower urinary tract obstruction: a systematic review and meta-analysis. *Obstet Gynecol* 2003;102(2):367–382.
61. Johnson MP et al. In utero surgical treatment of fetal obstructive uropathy: a new comprehensive approach to identify appropriate candidates for vesicoamniotic shunt therapy. *Am J Obstet Gynecol* 1994;170(6):1770–1776, discussion 1776–1779.
62. Coplen DE et al. 10-year experience with prenatal intervention for hydronephrosis. *J Urol* 1996;156(3):1142–1145.
63. Yerkes EB et al. Does every patient with prenatal hydronephrosis need voiding cystourethrography? *J Urol* 1999;162(3 Pt 2): 1218–1220.
64. Song SH et al. Is antibiotic prophylaxis necessary in infants with obstructive hydronephrosis? *J Urol* 2007;177(3):1098–1101, discussion 1101.
65. Coelho GM et al. Outcome of isolated antenatal hydronephrosis: a prospective cohort study. *Pediatr Nephrol* 2007;22(10):1727–1734.
66. Coelho GM et al. Risk factors for urinary tract infection in children with prenatal renal pelvic dilatation. *J Urol* 2008;179(1):284–289.
67. Chertin B et al. Conservative treatment of ureteropelvic junction obstruction in children with antenatal diagnosis of hydronephrosis: lessons learned after 16 years of follow-up. *Eur Urol* 2006;49 (4):734–738.
68. Atiyeh B, Husmann D, Baum M. Contralateral renal abnormalities in patients with renal agenesis and noncystic renal dysplasia. *Pediatrics* 1993;91(4):812–815.
69. Cascio S, Paran S, Puri P. Associated urological anomalies in children with unilateral renal agenesis. *J Urol* 1999;1621081–1083.
70. Arena F et al. Is a complete urological evaluation necessary in all newborns with asymptomatic renal ectopia? *Int J Urol* 2007;14 (6):491–495.
71. Gleason PE et al. Hydronephrosis in renal ectopia: incidence, etiology and significance. *J Urol* 1994;151(6):1660–1661.
72. Guarino N et al. The incidence of associated urological abnormalities in children with renal ectopia. *J Urol* 2004;172(4 Pt 2): 1757–1759, discussion 1759.
73. Calisti A et al. The risk of associated urological abnormalities in children with pre and postnatal occasional diagnosis of solitary, small or ectopic kidney: is a complete urological screening always necessary? *World J Urol* 2008;26(3):281–284.
74. Kaneko K et al. Abnormal contralateral kidney in unilateral multicystic dysplastic kidney disease. *Pediatr Radiol* 1995;25(4):275–277.
75. Miller DC et al. What is the fate of the refluxing contralateral kidney in children with multicystic dysplastic kidney? *J Urol* 2004;172(4 Pt 2): 1630–1634.