# Posterior Urethral Valves and Ureterovesical Junction Obstruction

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**Primary aims in the diagnosis and management of posterior urethral valves (PUV):**

1. **To prevent acquired renal damage.**
2. **To improve bladder function.**

**A secondary aim is to facilitate achievement of urinary continence.**

**Summary of evidence for these goals:**

**Case series report end-stage renal disease (ESRD) in from 3 to 42 % of patients during mean follow-up ranging from 4 to 12 years:**

- **No prenatal finding (oligohydramnios, anamnios, renal hyperechogenicity, loss of corticomedullary differentiation) or fetal urine test accurately predicts postnatal renal function.**
- **Prenatal bladder drainage improves survival but not renal function.**
- **Neither prenatal versus postnatal diagnosis nor age at postnatal diagnosis predicts likelihood for ESRD.**
- **There are conflicting results from retrospective analyses regarding potential risk for CRF/ESRD by vesicoureteral reflux (VUR), recurrent febrile UTI (fUTI), bladder dysfunction, and/or pop-off mechanisms.**

**Factors predicting poor renal functional outcomes are nadir creatinine >1 mg/dL and abnormal initial postnatal renal ultrasound with hyperechogenicity, volume loss, and/or loss of corticomedullary differentiation.**

**There are no reports of longitudinal objective analysis of bladder function after valve ablation:**

- **One study compared UD in infants after ablation to control males with UTI, reporting no difference in median maximum voiding pressures.**
- **Three retrospective studies found increased bladder capacity and decreased end filling pressures in bladders after valve ablation versus urinary diversion, but did not account for selection bias.**
- **Continence is reported by age 5 in approximately 20–60 %.**
- **There are few data regarding indications and outcomes for medical bladder therapy in valve patients using anticholinergics, alpha-blockers, and/or CIC.**

**One matched cohort study using validated questionnaires reported adult men mean age 37 years with prior valves had 2× greater LUTS than controls.**

## Prenatal Diagnosis

**Despite advances in ultrasound technology, one retrospective study reported high sensitivity to diagnose PUV but low specificity to distinguish valves from bilateral VUR and PBS in male fetuses with bilateral HN. The best indicators were increased bladder thickness and dilation, whereas the keyhole sign was not diagnostic.**

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**Table 15.1** Prevalence of prenatal ultrasound signs in those fetuses that were postnatally diagnosed with posterior urethral valves (PUV Group, $n = 31$) and those that were diagnosed with other pathologies (non-PUV group, $n = 23$)[a]

| Sign | PUV group ($n$ [%]) | Non-PUV group ($n$ [%]) | $P$[b] |
|---|---|---|---|
| Keyhole sign | 16 (51.6) | 8 (34.8) | 0.27 |
| Bladder dilatation | 30 (96.8) | 11 (47.8) | <0.0001[c] |
| Thickened bladder wall | 29 (93.5) | 9 (39.1) | <0.0001[c] |
| Oligohydramnios | 19 (61.3) | 7 (30.4) | 0.024[c] |
| Dilated and thick-walled bladder | 28 (90.3) | 9 (39.1) | 0.0001[c] |
| Dilated and thick-walled bladder with the keyhole sign | 14 (45.2) | 6 (26.1) | 0.18 |

[a]Reproduced from Bernardes LS, Aksnes G, Saada J, Masse V, Elie C, Dumez Y, et al. Keyhole sign: how specific is it for the diagnosis of posterior urethral valves? Ultrasound Obstet Gynecol. 2009;34(4):419–23, with permission from John Wiley and Sons
[b]For difference between PUV-diagnosed and non-PUV diagnosed groups
[c]Statistically significant

**A second retrospective series found prenatal ultrasound to have low sensitivity to detect PUV antenatally.**

A retrospective study reviewed prenatal findings in 54 males with bilateral HN between 2000 and 2006 to determine ability to diagnose PUV versus bilateral VUR and PBS. Median gestational age at first diagnosis of HN was similar in those with PUV versus other etiologies (25 weeks [19–36] vs. 23 weeks [12–33]). Forty eight (89 %) had more than one prenatal ultrasound, with a mean of five (1–7). PUV was suspected prenatally in 42, confirmed postnatally in 29 (69 %); PUV was not suspected prenatally in 12, but found postnatally in 2 (17 %). Overall sensitivity and specificity of prenatal evaluation were 94 % (95 % CI 87–99 %) and 43 % (95 % CI 30–57 %). The keyhole sign was found in 52 % of those with PUV versus 35 % without PUV, $p = 0.27$. Combinations of ultrasonographic findings are summarized in Table 15.1.

These findings indicate prenatal diagnosis continues to have poor specificity (Bernardes et al. 2009).

Another retrospective review evaluated 34 boys with PUV diagnosed between 1992 and 2004, reporting16 (47 %) had normal prenatal ultrasound. PUV was diagnosed in the other 18 at <24 weeks in 12 and >24 weeks in 6, whereas 6 of 16 not detected had an ultrasound at >24 weeks (Harvie et al. 2009).

## Prenatal Assessment of Renal Function

**There currently is no prenatal test able to accurately predict postnatal renal function.**

**A systematic literature review found that no fetal urine test is sufficiently accurate in predicting poor postnatal renal function to be clinically useful.**

**Three retrospective studies found neither oligohydramnios/anamnios nor abnormal fetal renal appearance (hyperechogenicity, loss of corticomedullary differentiation) correlated with postnatal renal function.**

## Fetal Urine Analysis

A systematic review of publications concerning accuracy of prenatal urine chemistries to predict poor postnatal renal function in fetuses with congenital bladder outlet obstruction included 23 articles involving 572 women. The two most accurate tests were calcium >95th percentile for gestation (LR + 6.65 [0.23–190.96]; LR − 0.19 [0.05–0.74]) and sodium >95th percentile for gestation (LR + 4.46 [1.71–11.6]; LR − 0.39 [0.17–0.88]). Beta (2)-microglobulin was less accurate (LR + 2.92 [1.28–6.69]; LR − 0.53 [0.24–1.17]). Since fetal urine anlaytes vary through gestation, becoming more hypotonic, results using the 95th percentile were more accurate than absolute thresholds. Nevertheless, the authors conclude that there currently is no accurate urinary analyte to predict poor postnatal renal function (Morris et al. 2007).

## Fetal Ultrasound

A retrospective analysis was performed in 31 fetuses with proven PUV. Pregnancy was terminated at a median gestational age of 30 weeks (21–36) in six for oligohydramnios plus adverse biochemical markers. Shunting was not attempted. Of the other 25 delivered, 4 previously had urinary ascites (three undergoing prenatal shunt and the fourth delivered at 33 weeks). All patients underwent valve resection. Outcomes for these 25 with median follow-up of 40 months (12–79) included 1 death at day 16 from renal failure, and 4 (17 %) others with renal impairment (abnormal serum creatinine) at last assessment. Prenatal oligohydramnios/anamnios and timing of its onset did not predict postnatal renal function. Similarly, prenatal renal ultrasonographic features (hyperechogenicity, lack of corticomedullary differentiation) did not predict postnatal serum creatinine. Creatinine was normal in three of the four with prenatal ascites. It was also normal in 10 of 13 with oligohydramnios, including 4 with anamnios. Ten of 11 with bilateral hyperechogenicity had a normal creatinine, as did 7 of 8 with bilateral hyperechogenicity, loss of corticomedullary differentiation, and oligohydramnios (Bernardes et al. 2011).

A retrospective review of 30 patients with PUV diagnosed in utero with postnatal valve ablation and follow-up a mean of 4 years (1–8) reported 6 (20 %) had CRF (serum creatinine >2 SD for age)/ESRD. No prenatal factor predicted poor renal outcomes: diagnosis at gestational age <24 weeks (OR 0.15 [95 % CI 0.88–2.26]), oligohydramnios (OR 0.81 [95 % CI 0.11–5.63]), or hyperechogenic renal cortex (OR 0.25 [95 % CI 0.62–10.14]) (El-Ghoneimi et al. 1999).

Another retrospective review included all liveborn males with PUV and at least one documented prenatal sonogram. There was no mention of prenatal intervention. Valve ablation or vesicostomy with subsequent valve ablation was done postnatally shortly after birth or at diagnosis. There were 34 patients, 18 (53 %) with prenatal diagnosis and 16 with a normal antenatal evaluation. Of those with prenatal detection, no ultrasound parameter (oligohydramnios, HN, renal echogenicity) predicted renal functional outcomes (Harvie et al. 2009).

## Prenatal Intervention Versus Observation

**One systematic review of published interventions found prenatal bladder drainage improved postnatal survival but not renal function. Results from a RCT of prenatal shunting are pending.**

To date no published RCT compares prenatal shunting versus observation in fetuses diagnosed with bladder outlet obstruction. The multicenter PLUTO (Percutaneous Shunting in Lower Tract Obstruction) trial was initiated with the primary objective to determine if prenatal shunting improves postnatal mortality and renal function (Morris and Kilby 2009). Recruitment has closed, but results are pending.

Systematic review of published antenatal intervention for bladder outlet obstruction identified 20 articles with 369 fetuses, of which a total of 261 had antenatal intervention. Most were percutaneous vesico-amniotic shunts, but 9 were open fetal surgeries and 26 were fetal cystoscopies with 14 valve ablations. Prenatal bladder drainage improved perinatal survival (likely due to fewer deaths from pulmonary complications) versus no treatment (OR 3.86 [95 % CI 2.00–7.45]), primarily in a subgroup with predicted poor prognosis on the basis of fetal urinalysis (OR 12.85 [95 % CI 1.25–153.03]). However, improvement in postnatal renal function was not found (OR 0.50 [95 % CI 0.13–1.90]) Complications included shunt dislodgement or occlusion in 34 % with vesico-amniotic shunting, two premature rupture of membranes, four cases of chorioamnionitis, three hernias, and one bladder rupture from shunting, and three patient deaths (Morris et al. 2010).

## Prognosis Based on Prenatal Versus Postnatal Diagnosis

**The likelihood that boys with PUV will develop CRF/ESRD does not vary by prenatal versus postnatal diagnosis.**

Analysis was performed in 46 boys diagnosed with PUV between 1983 and 1997, with mean follow-up 12.5 years (5.5–20) using a prospective protocol. Antenatal diagnosis was made in 23/40 (57.5 %) at median gestational age 28 weeks (16–38). In the other 23, the diagnosis was made postnatally at median 31 days of age (1 day–1 year) for uremia/acidosis, UTI, respiratory symptoms, and/or weak urinary stream. Treatment was nearly identical in both cohorts, comprising valve ablation in 23 (50 %) or vesicostomy or supravesical diversion in 23 (50 %). CRF (GFR≤59 mL/m/1.73 $m^2$) developed in 6 (13 %) and ESRD in 8 (17 %), with no difference in antenatal versus postnatal detection (Ylinen et al. 2004).

A retrospective review of 34 boys with PUV compared prenatal versus postnatal diagnosis for risk of CRF (serum creatinine >2 SD for age)/ESRD. Sixteen of 34 had normal prenatal ultrasound with postnatal diagnosis made at a median 3.5 months (2 day–5 year). Follow-up after valve ablation was a mean of 8 years. There was no difference in poor outcomes based on antenatal versus postnatal diagnosis (5/13 vs. 2/14, $p = 0.4$) (Harvie et al. 2009).

Another retrospective review compared 30 boys with prenatally detected PUV and 10 with postnatal diagnosis at mean of 21 days (1–60). At mean follow-up of 4 years, 2/30 (7 %) and 1/10 (10 %) had ESRD. If another five with prenatal diagnosis leading to termination were included, poor outcomes remained similar (El-Ghoneimi et al. 1999).

## Prognosis Based on Postnatal Parameters

**Several reports analyzed factors potentially correlating with CRF/ESRD. All are retrospective, many involve few patients, the diagnosis of CRF varies (<90 mL/min/1.73 $m^2$ vs. <60–64 mL/min/$m^2$), and definitions of factors were not always stated.**

**Factors not predicting poor renal functional outcomes include age at diagnosis and VURD.**

**Factors correlating with poor renal functional outcomes are nadir creatinine >1 mg/dL and abnormal initial renal ultrasound (hyperechogenicity, loss of volume, loss of corticomedullary differentiation).**

**Abnormal bladder function is also associated with poor renal function, but usefulness of this observation is limited by lack of a consistent definition for diagnosis.**

**There are mixed findings concerning the relationships between urinomas, VUR, and UTI and CRF/ESRD. Although results of analyses of VUR and CRF/ESRD varied, no study reviewed found VUR grade to predict renal outcomes.**

## Age at Diagnosis

A retrospective review of 227 males with PUV undergoing primary valve ablation analyzed factors leading to ESRD. Mean age at diagnosis was 30 months (10 days–16 years), with only 2 diagnosed prenatally and 107 (47 %) during infancy. Mean follow-up was 7 years (0.5–16), during which time ESRD occurred in 27 (12 %), and was not influenced by presentation before versus after infancy (Ansari et al. 2010).

A similar retrospective analysis of 120 males undergoing primary valve ablation between 1987 and 2004 considered factors potentially associated with ESRD. Age at diagnosis was a mean of 2 years (1 day–15 years), presenting with urinary retention, UTI, stranguria/dysuria, and incontinence. With follow-up a median of 3.6 years, ESRD occurred in 18 (15 %). Age at presentation did not correlate with renal function (although statistical means to evaluate age, i.e., <1–2 years vs. >2 years mentioned in discussion, or as a continuous variable, were not described) (Sarhan et al. 2011).

Another retrospective evaluation also found age at presentation not predictive of renal functional outcomes. This study included 52 males with PUV divided into two groups, those diagnosed and treated before ($n=39$, mean age 30 days at surgery) versus after 1 year of age ($n=13$, mean age 3.8 years at surgery). Those with late diagnosis presented with UTI, voiding dysfunction, or failure to thrive. Follow-up was a mean of 7 years after valve ablation, but specific times for the two groups were not stated. ESRD did not vary significantly between the two groups (8 % vs. 0 %, $p=0.55$) (Kibar et al. 2011).

## Nadir Serum Creatinine

Retrospective analysis was performed in 227 males with PUV undergoing valve ablation between 1992 and 2008, at mean age 30 months (10 days–16 years) and mean follow-up 7 years (0.5–16). ESRD developed in 27 (12 %). Multivariable logistic regression analysis found nadir serum creatinine >1 mg/dL correlated with renal failure (OR 23.79 [95 % CI 8.20–69.05]). Twenty two of 27 patients with ESRD had nadir creatinine >1 mg/dL, although a creatinine >1 mg/dL was also found in 18/200 without renal failure (Ansari et al. 2010).

A retrospective analysis of renal functional outcomes evaluated presenting and nadir serum creatinine in a series of 120 males with primary valve ablation. Mean age at treatment was 2 years (1 day–15 years), with follow-up a mean 4.4 years. CRF was defined as GFR (determined by Schwartz formula) ≤59 mL/min/1.73 $m^2$. Patients with CRF ($n=44$) were compared to those with normal or less severe renal insufficiency ($n=76$). Mean initial serum creatinine (1.7 mg/dL vs. 0.8 mg/dL), mean nadir serum creatinine (1 mg/dL vs. 0.55 mg/dL) and mean initial calculated GFR (34 mL/min vs. 117 mL/min) were all significantly different between the two groups. Nevertheless, 24 % of patients with a nadir creatinine <1 mg/dL still developed CRF, while 16 % with nadir creatinine >1 mg/dL did not (Sarhan et al. 2010).

Retrospective analysis was done in 42 males, median age 0.97 months (interquartile range 0.03–58.49) with PUV treated between 1983 and 2009 that segregated patients into 23 with normal renal function versus 19 with GFR (using Schwartz formula) <90 mL/min/1.73 $m^2$ throughout mean follow-up of 5.2 years. Serum creatinine levels at age 1 year were available in 19/23 and 14/19 patients in the two groups, with median calculated GFR 112 mL/min/1.73 $m^2$ (IQR 96.0–134.0) in those with preserved normal renal function versus 24.5 mL/min/1.73 $m^2$ (IQR 14.0–62.3) in those with renal insufficiency, $p<0.001$ (Pohl et al. 2012).

## Renal Ultrasound Parameters

Retrospective analysis reported by Pohl et al. (2012) mentioned above evaluated renal ultrasound parameters from the first postnatal study in 42 PUV patients diagnosed at median age <3 months and compared them to functional outcomes of normal calculated GFR versus CRF (<90 mL/min/1.73 $m^2$). Total renal volume less than the third percentile was significantly more common in those with CRF (OR 17.42 [95 % CI 3.28–92.61]), with 79 % of those with CRF having hypoplastic kidneys. Similarly, hyperechogenicity greater than the liver or spleen (OR 21.33 [95 % CI 2.37–192.03]) and loss of corticomedullary differentiation (OR 28 [95 % CI 3.11–252.48]) correlated with CRF.

The retrospective review by Sarhan et al. (2011) referenced above including 120 males with PUV diagnosed at mean age 2 years (1 day–15 years) with follow-up a median 3.6 years also considered first renal ultrasound appearance versus calculated GFR. However, in this study, CRF was defined as ≤59 mL/min/1.73 $m^2$. Univariable analysis found significant differences in hyperechogenicity occurring in 12/76 (16 %) with normal GFR versus 16/44 (36 %) with CRF, $p=0.014$.

## VUR

VUR was not predictive for CRF (<90 mL/min/1.73 $m^2$) in the retrospective analysis reported by Pohl et al. (2012) mentioned earlier. VUR was initially diagnosed in 21/40 (52.5 %)

(no information was available in 2), which was unilateral in 12 and bilateral in 9 patients for a total of 30 renal units. VUR was grades 1–2 in 9 and 3–5 in 21 renal units. No difference in VUR was noted in patients with normal versus reduced renal function (OR 2.89 [95 % CI 0.79–10.57]).

Similarly, the retrospective analysis by Sarhan et al. (2011) discussed above that defined CRF as calculated GFR ≤59 mL/min/1.73 $m^2$ found that VUR was not a predictive factor on univariable analysis that compared no, unilateral, and bilateral VUR but did not report grade differences, if any.

Another retrospective review identified 142 patients with PUV treated between 1975 and 2005, of which 119 had sufficient data for analysis at mean follow-up of 7 years (3–24). Fifteen (13 %) progressed to ESRD. Although VUR grades III–V occurred in 93 % with ESRD versus 48 % in patients without ESRD, multivariable logistic regression analysis found differences were not significant (OR 2.0 [95 % CI 0.2–24]) (DeFoor et al. 2008).

In contrast, one retrospective analysis did find bilateral VUR a risk factor for ESRD in multivariable logistic regression. There were 116 males with PUV presenting at mean age of 13 months (0–120) who underwent primary valve ablation, followed by diversion in 32 (28 %) by Sober ureterostomies (31) or vesicostomy (1) for "severe VUR and HN" (22) or poor renal function (9). Subsequent follow-up was a mean of 10 years (18 months–22 years), during which time 49 (42 %) developed ESRD. VUR was found in a total of 52/116 (45 %) patients, including 18/49 (37 %) with ESRD, unilateral in 10 and bilateral in 8. Bilateral VUR, found in 16 % of those with ESRD, was reported a significant risk factor in multivariable logistic regression, but the OR was not stated. However, neither high-grade VUR (3–5) nor unilateral VUR was found to be a risk factor (Ghanem et al. 2004).

The retrospective analysis by Ylinen et al. (2004) of 46 boys reported VUR initially in 32 (70 %), unilateral in 17 and bilateral in 15 (grades not stated). Both unilateral and bilateral VUR increased risk for CRF (<60 mL/min/1.73 $m^2$)/ESRD versus no VUR. Among patients with VUR, there was no difference in renal outcomes for grades 1–2 versus 3–5.

## UTI

Recurrent fUTI was another risk factor for ESRD considered by DeFoor et al. (2008), occurring in 58/119 (49 %) patients despite antibiotic prophylaxis for VUR, bladder dysfunction requiring CIC, and/or "severe" HN. These infections were equally likely in patients with preserved renal function versus those with ESRD.

The analysis by Ansari et al. (2010) discussed above also reported that recurrent fUTI (not further defined) did not correlate with progression to ESRD in 227 males presenting at mean age of 30 months. Seventy seven (34 %) had fUTI during mean follow-up of 7 years, with no difference in those with versus without ESRD.

One study that specifically defined fUTI similarly found it was not a significant risk factor for renal function > than or <90 mL/min/1.73 $m^2$ calculated GFR, occurring in a total of 33/42 (79 %) patients (OR 3.72 [95 % CI 0.67–20.63]). However, >3 fUTIs (occurring in 50 %) did correlate with reduced renal function (OR 6.40 [95 % CI 1.65–24.77]) (Pohl et al. 2012).

However, Ylinen et al. (2004) reported that UTIs occurred in 30/46 (65 %) boys with PUV, with a mean of 1.9 episodes of fUTI in 22 of them, and that these patients were more likely to have CRF (<64 mL/min/1.73 $m^2$)/ESRD than those without UTI.

## Bladder Dysfunction "Valve Bladder"

Ansari et al. (2010) defined bladder dysfunction for their retrospective multivariable analysis as the presence of one or more factors: end filling pressure >40 cm $H_2O$, post-void residual urine >30 % of the maximum cystometric bladder capacity, myogenic failure, and/or need for CIC. By these criteria, 21/27 (78 %) with ESRD had bladder dysfunction versus 38/200 (19 %) patients without ESRD (OR 5.67 [95 % CI 1.90–16.93]).

A retrospective analysis included urodynamic findings in 116 boys with PUV presenting at mean age 13 months (0–120) and then undergoing primary valve ablation, with 31 also having Sober ureterostomies and 1 a vesicostomy. A total of 52 (45 %) developed ESRD. Age and number of UDs (assuming some had more than one study) were not stated. Studies were done transurethrally with a 7-Fr catheter at a filling rate of 5–10 cc/min. Expected bladder capacity was determined by formula (age + 2) × 30 cc; "loss" of bladder compliance was not defined, although used as a factor in logistic regression. Eighty percent of all patients had abnormal bladder parameters. Univariable analysis found bladder compliance and/or detrusor overactivity to be significant predictive factors for ESRD, specific results of multivariable analysis for these factors were not stated (Ghanem et al. 2004).

Bladder dysfunction leading to CIC was found on retrospective chart review in 37/119 (31 %) patients with PUV, generally due to low capacity and filling pressures >40 cm $H_2O$ in younger patients or myogenic failure with retention in older patients ("low capacity, younger, older, retention" were not defined). These patients were found on multivariable logistic regression to have increased risk for ESRD (OR 8.9 [95 % CI 1.1–73]) (DeFoor et al. 2008).

In contrast, Sarhan et al. (2011) reported that bladder dysfunction occurring in 35 % of toilet-trained boys did not correlate with ESRD. However, definition of "bladder dysfunction," means to diagnose it, and time to diagnosis after valve ablation that was done at mean age 2 years, were not stated.

Ylinen et al. (2004) simply defined bladder function as total urinary continence (day and night) and compared achieving this threshold at age <5 years versus >5 years. By this definition, bladder function did not correlate with CRF/ESRD (OR 0.45 [95 % CI 0.13–1.65]).

## VURD/Urinary Extravasation

The analysis by Ylinen (2004) of 46 PUV boys included VURD, defined as unilateral VUR into a nonfunctioning kidney, which was found in nine patients, with three developing ESRD. Urinary extravasation was noted in four patients, one progressing subsequently to ESRD. Neither "pop-off" mechanism was found protective of renal function.

"Pop-off" mechanisms, including VURD, large bladder diverticulum, urinoma, and/or ascites, were considered a single factor in the retrospective analysis of 120 PUV cases by Sarhan et al. (2011). Presence of these did not predict future renal function.

Another retrospective study involved 73 consecutive patients with PUV presenting at mean age of 1.6 years (0–11). Of these 35 had VUR, with VURD (unilateral VUR with renal dysplasia) in 21. Follow-up was a mean of 5.6 years (1–17). Logistic regression analysis was not done, but chi square analysis found no differences in patients with versus without VUR, or in those with VURD versus others having VUR without VURD (Hassan et al. 2003).

A retrospective review divided 89 consecutive patients with PUV diagnosed as neonates and treated between 1989 and 2009 into the 9 (10 %) with urinomas versus those without to determine impact, if any, on renal function. These urinomas were bilateral in one and unilateral in the remainder, ranging in size from 4 × 3 × 4 to 9 × 9 cm. Median initial nadir creatinine after valve ablation or vesicostomy was significantly lower in those with urinomas (0.35 mg/dL [0.2–0.5] vs. 0.5 mg/dL [0.2–6.5], $p < 0.01$). Ipsilateral renal function was not reported. Initial management of the urinomas was conservative, although four underwent percutaneous drainage due to respiratory or feeding impairment; neither the time of drainage nor the time for resolution of those observed was stated (Wells et al. 2010).

## Diagnosis by VCUG

**Our review found no report on the sensitivity and specificity of VCUG in diagnosis of PUV. Two studies agreed that the catheter in the urethra during voiding imaging does not impair diagnosis of PUV.**

Three radiologists reviewed preoperative VCUGs in 48 patients with cystoscopically confirmed PUV to determine if the urethral catheter used in the test impacted visualization of the valve. Studies were done leaving the catheter in the urethra in 28, without in 17, and both in 3. Valves were diagnosed in 25/28 (89 %) with the catheter versus 15/17 (88 %) without (Ditchfield et al. 1995).

In another study, 123 males had VCUG using a 6- or 8-Fr catheter at median age 2.6 months during investigations of prenatal HN or UTI. For inclusion, four voiding phase images were required, two with and two without the catheter; only 80 studies met this criteria. Of these, three showed PUV, seen in views with and without the catheter (Chaumoitre et al. 2004).

## Initial Management

**No RCT compares outcomes for treatment in newborns by primary valve ablation versus diversion by vesicostomy or supravesical diversion.**

**Our review found only one study comparing primary valve ablation versus vesicostomy for newborn management of PUV, with selection determined by whether or not the urethra admitted a 9-Fr resectoscope. Preoperative mean serum creatinine and mean serum creatinine at 1 year were reported as similar in the two groups.**

**Another review of patients <1 year at surgery found no difference in renal insufficiency or ESRD based on initial valve ablation versus supravesical diversion, although reasons for different interventions were not stated.**

**Within the limitations of retrospective chart reviews and the potential for selection bias in deciding valve ablation versus diversion, our review found no report indicating diversion superior to ablation.**

**Valve ablation can be done by a variety of methods, including fulguration, laser, or cold knife incision. Our review found few descriptions of technique and outcomes.**

**"Residual valves" have been reported by several authors, described by one as leaflets that could be engaged with a cold knife hook but not defined by others. Two studies using objective urethral ratios to diagnose persistent posterior urethral dilation reported residual valves in those with ratios above the medians of controls (1.73, 2.6) during follow-up VCUG 6–12 weeks later. Re-ablation resulted in normalization of ratios.**

**Urethral strictures occurring in up to 50 % of newborns and infants when 10-Fr or larger resectoscopes were used prompted vesicostomy or ablation using either hooked electrodes without direct visualization or bugbee electrodes introduced through smaller cystoscopes. Today 9-Fr resectoscopes potentially allow more primary ablations under direct vision in newborns with less concern for urethral trauma. Our review found only one contemporary study reporting inability to use a 9-Fr resectoscope in newborns, occurring in 47 %, and none concerning stricture presence or absence after ablation.**

## Ablation Technique

Thirteen neonates at mean age 9 days underwent transurethral YAG laser valve ablation at maximum power of 25 W via an 8-Fr cystoscope. Vaporization began centrally and moved peripherally, leaving a small rim circumferentially. VCUG was repeated at 4 weeks, with subsequent follow-up to a mean of 2.3 years (6 month–5 years). No strictures occurred, and no delayed repeat ablations were reported. Four infants who had ureteral reimplantation, apparently within 6 months of ablation, also had cystoscopy stated to demonstrate no residual valves (Biewald and Schier 1992).

## Adequacy of Valve Ablation

The retrospective report from Sarhan et al. (2011) with 120 PUV patients treated at mean age 2 years stated that all underwent primary valve ablation, done in 65 (54 %) with a cold knife versus fulguration using a hot loop or bugbee hook electrode (43 [36 %] and 12 [10 %]). Repeat VCUG was obtained 1–3 months later, with "adequate valve ablation" in 85 %. The remaining 18 (15 %)

underwent a second ablation. Specific findings to determine adequate ablation on VCUG were not defined, nor were findings at residual fulguration described or correlated with initial surgical methods.

Retrospective analysis was done in 31 consecutive patients with post-ablation cystoscopy during a 2-year period from 2006 to 2008. Ablation was done using an 11-FR cold knife ($n=21$) or 3-Fr point electrocautery ($n=9$) and was unknown in one. Further details (location and number of incisions) were not stated. Follow-up cystoscopy and VCUG were done in all at median 5 months (3–12). VCUG reported resolution in 10, improvement in 8, and persistence of posterior urethral dilation in 12 (39 %). Cystoscopy found valves sufficient for further ablation in 16 (52 %). Cold knife ablation was done in all but one, and these incisions were described: at 5, 7, 12 O'clock ($n=5$); at 5, 7 O'clock ($n=8$); at 7 or 12 O'clock only in one each. Potential clinical relevance for these valve remnants was assumed by ability to engage them with the hook of the cold knife. Based on secondary ablation, the positive/negative predictive value of visualized valves plus persistent posterior urethral dilation on VCUG was 83 %/75 %, but either valve or posterior urethral dilation alone was only 40 %/50 %. (Smeulders et al. 2011).

In another retrospective analysis, 20 patients underwent valve ablation at median age 1.5 months (1–12) and then had urethral ratios (transverse midpoint diameter of posterior urethra/transverse diameter at widest point of bulbar urethra during voiding without catheter) determined by repeat VCUG 6–8 weeks later. Ablation was performed using an 11-Fr resectoscope with Collins knife and cutting diathermy current or a 9-Fr cystoscope with 2.4-Fr electrode, with location and number of incisions not stated. Controls were similarly aged boys undergoing VCUG "for suspected urinary tract pathology." Median urethral ratio in 13 patients with available studies before valve ablation was 8.6 (4–14.7) versus post-ablation median in 20 patients of 3.4 (1.9–15) and controls of 2.6 (1.3–5.5). Repeat cystoscopy was selective, in five patients with persistent dilation a mean of 8 (5–15.5), who all had secondary ablation and resultant decrease in urethral ratio to 3.1 (2.9–6.4) (Bani Hani et al. 2006).

VCUG and repeat cystoscopy was done in 30 patients 8–12 weeks after valve ablation at a median age of 13 months (1 day–11 years). Ablation was performed using either a 7- or 10-Fr cystoscope with bugbee electrode, but otherwise technical details (location and number of fulgurations) were not described. There were 30 controls for urethral ratio undergoing VCUG for "suspected urinary tract pathology." Mean urethral ratio in controls was $1.73 \pm 0.57$, versus pre-ablation mean in patients of $4.94 \pm 2.97$, $p < 0.001$ (21/30 had ratios >2.3). Mean post-ablation ratio was $2.13 \pm 1.19$, which was not different from controls. However, two patients had persistently abnormal post-ablation ratios (6.5) and both had residual valves not seen in others (Gupta et al. 2010).

## Urethral Stricture After Ablation

A review of 28 patients with transurethral valve ablation noted that strictures, diagnosed by VCUG, all occurred in newborns and infants. Ablation was done using a loop electrode via a 10-Fr ($n=2$) or 12-Fr ($n=12$) resectoscope, and strictures developed in 7/14 (50 %), located in the membranous ($n=2$), bulbar ($n=4$) or anterior ($n=1$) urethra (Myers and Walker 1981).

Urethral stricture at the site of prior ablation occurred in 3/82 (4 %) patients in one retrospective series in which all newborns and infants underwent vesicostomy with subsequent closure and valve ablation at age 9–12 months. Fulguration was done using a 9-Fr resectoscope with a loop electrode applied at 5, 7, and 12 O'clock. VCUG was done 6 months after ablation. Of the three strictures, one occurred following urethral trauma with a false passage and "profuse bleeding" done elsewhere, with re-fulguration subsequently done for residual valves. The other two had preliminary diversion. Means of diagnosis, time after ablation to stricture, and any associated symptoms were not stated, nor were treatment and results for the strictures described (Lal et al. 1998). Considering the subgroup of patients

<12 months old at presentation (comprising those with diversion before ablation) strictures occurred in 2/38 (5 %).

## TUR Valves Versus Urinary Diversion

A review of 45 newborns with PUV managed between 1997 and 2002 found that 24 underwent primary valve ablation, while 21 had vesicostomy because the urethra would not admit a 9-Fr scope. Mean preoperative serum creatinine was similar in the two groups (1.6 ± 1.5 mg/dL vs. 1.7 ± 1.5). Nine were lost to follow-up, and six died before 12 months (four valve ablation, two vesicostomy). Apparently, diverted patients still had vesicostomy at 12 months. Mean postoperative serum creatinine at 1 year was 0.7 ± 0.2 mg/dL in 12 patients after ablation versus 0.9 ± 0.7 mg/dL in 9 with vesicostomy, which was said to be similar by chi square analysis (Narasimhan et al. 2004).

Another review included 46 patients, 23 prenatally detected and 23 diagnosed within 1 year of life. In each group approximately 50 % underwent primary valve ablation or supravesical diversion, but decision making was not described. During a mean observation period of approximately 12 years, six (13 %) developed renal insufficiency (<59 mL/min/1.73 $m^2$) and eight (17 %) ESRD, with no differences in outcomes based on initial surgical management (Ylinen et al. 2004).

A retrospective review included 100 consecutive patients with PUV born before 1985 and reported in 1996. Forty-two presented at less than 1 month of age, and 56 at less than 1 year (mean, median, range not reported). Initial management included valve ablation (74), vesicostomy (13), or supravesical diversion (9); after valve ablation, three also had supravesical diversion without further improvement in serum creatinine, and four others had vesicostomies, with one then having decreased creatinine. Median follow-up was 11 years, but follow-up for each group was not stated. Data were presented as Kaplan Meier curves, showing no statistical differences in risk for ESRD based on initial therapy (Smith et al. 1996). Selection of patients for primary valve ablation versus urinary diversion was not described, nor was analysis presented to demonstrate if patient groups had similar initial renal function.

Another retrospective review included 67 patients treated between 1985 and 2000, 38 undergoing primary valve ablation, 25 vesicostomy for a small urethra, and 4 ureterostomy for "gross pyuria and sepsis with dilated and tortuous ureters, or failure of serum creatinine to diminish with catheter drainage." Patients undergoing vesicostomy were significantly younger at a mean of 16.3 ± 29.1 months versus 37.2 ± 36.1 and 40.5 ± 17.9 months for valve ablation or ureterostomy. Initial renal function for each of the three groups was not stated or compared, nor was timing for post-surgical analysis of renal function described—the authors only stating that improvement in serum creatinine levels was not significantly different in diverted patients versus those with valve ablation (Puri et al. 2002).

## Bladder Function

**Although bladder dysfunction is a potential risk factor for ESRD in boys with PUV, we found no published systematic protocol to diagnose and manage patients. While functional characteristics of the bladder may evolve over time after valve ablation, our review found no studies describing longitudinal UD evaluations.**

**Two studies reported bladder capacity and compliance better after primary valve ablation versus diversion by vesicostomy or supravesical diversion, although selection bias for diversion could account for this observation.**

**One study with UD performed in infants within 2 weeks of valve ablation found no difference in median maximum detrusor pressures during voiding compared to age-matched controls evaluated after UTI.**

**Bladder dysfunction is also suspected from urinary incontinence in boys after the usual age of toilet training, reported in 42–80 %, but few reviews described ages at which continence was achieved, and none was found that used objective assessments to define normal versus altered**

**bladder function. Potential contributing factors, such as detrusor overactivity, poor compliance, and/or polyuria, have not been systematically assessed when continence is delayed.**

**Few studies report medical therapy with anticholinergics, alpha-blockers, and/or CIC in patients after valve ablation. Indications, age at initiation of therapy, duration of treatment, and results have not been systematically reported.**

**One study found imipramine abolished detrusor overactivity in nearly 90 % and improved compliance in 43 % of patients treated, with symptomatic improvement reported in 59 %.**

**Two studies reported alpha-blocker therapy using terazosin for PVR >10 % predicted bladder capacity reduced post-void residuals from a mean of approximately 15 cc to a mean of 2 cc.**

**One matched cohort study of adult men using a validated questionnaire found symptoms of voiding dysfunction reported by 32 % of former valve patients, who also had significantly greater urge and/or stress incontinence compared to age-matched controls.**

## Urodynamics

A retrospective study created two groups: 11 patients treated from 1970 to 1983 who underwent supravesical diversion versus 8 undergoing primary valve ablation from 1980 to 1983. Indications for diversion were small urethra, "severe urinary tract infections and failure to stabilize renal function or improve a poor clinical condition despite bladder drainage via a urethral catheter and medical treatment." Mean patient age at surgery was 10.2 ± 8.1 months (diversion) versus 11.8 ± 11.2 months (valve ablation), and mean duration of diversion was 57 ± 39 months. All had "rapid fill" UD testing at mean ages of 13.6 ± 2.2 years (diversion) versus 11 ± 4.1 years. Mean maximum bladder capacity adjusted for age by the formula age + 2 × 30 was significantly greater in those ablated (1.37 ± 0.19 %) versus diverted (0.9 ± 0.3 %), and end filling pressure at this capacity was significantly less in ablated (8 ± 2.6 cm $H_2O$) versus diverted (19.3 ± 4 cm $H_2O$) bladders (Podesta et al. 2002).

The retrospective analysis of 67 patients treated initially by primary valve ablation ($n = 38$) versus vesicostomy ($n = 25$) or supravesical diversion ($n = 4$) mentioned above (Puri et al. 2002) included UD testing in all patients. Studies were performed using an 8-Fr transurethral catheter at a filling rate of 10 % expected bladder capacity for age. A significant difference was found in maximum bladder capacity between ablated versus vesicostomy patients (mean % 96 ± 33 vs. 62 ± 28). Compliance of <10, 10–20, and >20 cc/cm $H_2O$ was also significantly better in ablated versus vesicostomy patients. Age at which UD was performed, time interval from valve treatment, and number of UDs performed per patient were not described.

UD was obtained in 116 patients who underwent valve ablation at mean age 14 months (6 days–164 months), followed by supravesical diversion in 32 (28 %) for "severe VUR and HN" in 22 and "poor renal function" in 9 for an unreported duration. Time after ablation for UD testing, the number of tests obtained per patient or, if more than one study was done, which was selected for analysis, were not stated. Filling was done via a 7-Fr transurethral catheter at 5–10 mL/min. Abnormal bladder parameters were found in 93 (80 %). Cystometric capacity was greater than expected by formula for age in 34 (29 %) and less in 33. Compliance loss was reported for 30 (26 %) and detrusor overactivity in 44 (38 %). Patch EMG electrodes indicated impaired pelvic floor relaxation in 31 (27 %), while retention >10 % voided volume was noted in 47 (41 %). Although 80 % of patients were considered to have abnormal bladder function, when the authors categorized them further, 86 (74 %) had normal compliance (61 without and 25 with detrusor overactivity), while 30 (26 %) had decreased compliance (13 without and 17 with detrusor overactivity). Definitions of increases/decreases in capacity, decreased compliance, or abnormal pelvic EMG were not stated. Differences, if any, between those with ablation versus ablation plus diversion were not discussed. Multivariable regression analysis found only bilateral VUR, and not bladder dysfunction, to independently predict ESRD (Ghanem et al. 2004).

Another retrospective analysis identified 25 males evaluated from 1994 to 2007 at a median age of 0.5 month (0–10.2). UD was performed in six boys 2–6 days before valve ablation, and in the other 19 at median 2 days (0–15) after ablation. Detrusor pressures were monitored using 4-Fr catheters, presumably transurethrally, with infusion done at 2–5 cc/min using a second 4-Fr tube placed suprapubically in 8. A control group of males without PUV was created from infants having UD after UTI at median age 3.3 months (1.5–9.6). Median maximum detrusor pressure during voiding was similar in PUV patients (112 cm $H_2O$, 40–331) and controls (91 cm $H_2O$, 48–191, $p=0.39$). Repeat UD was obtained at 12 months after valve ablation in 17 patients, finding a significant increase in median bladder capacity and decrease in median maximal voiding pressures to 100 cm $H_2O$, 60–193, $p=0.01$ (Taskinen et al. 2009).

## Urinary Continence

Continence was assessed in 100 boys with PUV, excluding an unspecified number who died or had ESRD before age 5 years. Total continence (dry day and night) occurred in 19 % of boys by age 5, 46 % by age 10, and in 99 % by age 20, with no significant differences based on initial therapy by valve ablation versus urinary diversion (Smith et al. 1996). Additional therapies (anticholinergics, CIC), if any, were not described.

In another study of 46 patients diagnosed either prenatally or within the first year of life and then undergoing either valve ablation or supravesical diversion, total urinary continence was achieved in 17 (37 %) by age 5 years, and in 22/27 (81 %) with follow-up to 10 years. Differences, if any, based on initial surgery were not described (Ylinen et al. 2004).

In another retrospective series, 63/65 cases of PUV diagnosed prenatally underwent primary valve ablation. During follow-up a median of 6.8 years, 55 patients were toilet-trained, of which 32 (58 %) were dry at mean age 3 years. Of the remaining 23, 7 were dry using anticholinergics ± alpha-blockers, 3 were dry on CIC, 3 had nocturnal enuresis (presumably dry during the day), and 10 were wet day and night (Sarhan et al. 2008).

Of 70 boys presenting at mean age 7.5 years (2–14), 33 (47 %) had diurnal incontinence. Following valve ablation, incontinence resolved in 24 at an unspecified interval and persisted in 9 (27 %). In four of nine, symptoms were controlled with medication (desmopressin, anticholinergics), but use in others was not stated. Incontinence resolved in six of nine patients during a mean of 11 months (4–18) and persisted in two receiving medical therapy; one was lost to follow-up (Schober et al. 2004).

Bother from bladder dysfunction can extend into adulthood. A study from Finland identified 106 patients with PUV treated after 1953, and received a response from 68 (64 %) to a validated LTUS questionnaire. These data were compared to age-matched controls at a 4:1 ratio (272 controls: 68 PUV patients) at median age of 37.5 years (18–57). Of PUV patients, 32 % reported at least one moderate or severe symptom, versus 16 % of controls ($p=0.002$). Overall, patients had 2× increase in LTUS (hesitancy, weak stream, incomplete emptying, straining) over controls. Urge incontinence was reported by 15 % PUV patients versus 5 % of controls ($p=0.014$), while stress incontinence occurred in 12 % versus 3 % ($p=0.005$) (Tikkinen et al. 2011).

## Medical Bladder Therapy (Anticholinergics, Alpha-Blockers, CIC)

A longitudinal study involved 30 PUV patients, all ≥5 years of age (5–20, mean or median not stated), who had UD testing and were treated with imipramine. Indications for investigation of these patients versus others of similar age, if any, were not described. Although all were considered toilet-trained, 27/30 had symptomatic voiding dysfunction described as diurnal incontinence in 22 and only nocturnal enuresis in 5. UD used an 8-Fr transurethral catheter with filling rates at 10 % expected capacity for age by formula, and needle electrodes for sphincter

EMG. All were considered to have abnormal UD, with detrusor overactivity in 18 (60 %) and compliance <10 mL/cm $H_2O$ in 21 (70 %). Imipramine was used at 1.5–2 mg/kg/24 h in a single or divided doses. “Significant” symptomatic improvement occurred in 16/27 (59 %). Repeat UD was reported at 1 and 2 years. Detrusor overactivity resolved on medication in 16/18 (89 %), while compliance improved in 9/21 (43 %) (Puri et al. 2005).

As mentioned above, of 23 toilet-trained boys with prenatally diagnosed PUV and primary valve ablation, 7 (30 %) were dry using anticholinergics ± alpha-blockers, 3 were dry on CIC, 3 had nocturnal enuresis (presumably dry during the day), and 10 were wet day and night. Specific indications for these therapies were not described (Sarhan et al. 2008).

Another retrospective analysis of 119 patients undergoing valve ablation reported 37 (31 %) were started on CIC for “hostile bladder dynamics” (DeFoor et al. 2008). The authors generally described these patients in two categories: “younger” ones who had low capacity and high-end fill pressures that also received anticholinergics, and “older” ones with myogenic failure and retention who did not need anticholinergics. The number of older versus younger patients, age at initiation of medical therapy, and response to treatment were not described.

Forty two patients were diagnosed with post-void residual volumes by ultrasound >10 % of expected bladder capacity for age 1 week after valve ablation. Age ranged from neonates to >5 years, with 69 % less than 1 year. Terazosin, 0.02–0.4 mg/kg, was given daily, with 25 % increases in dose every 2 weeks until residual volumes were <10 % capacity. After 6 months of successful therapy, medication was weaned. Mean pretreatment residual volume was 16 cc (34 % expected capacity) versus mean residual 2 cc (1.5 % expected capacity) on therapy. Symptomatic hypotension occurred in one, and another two boys did not respond. Four had therapy successfully ended after 14 months, while others apparently continued medication at mean follow-up 17 months (2 months–6 years) (Abraham et al. 2009).

In another retrospective series of 65 consecutive patients undergoing valve ablation at median age 1.5 months (1 day–13 years), post-void residuals were measured by ultrasound during outpatient follow-up, with those having PVR >10 % expected bladder capacity started on terazosin 0.04–0.4 mg/kg/day. Patient age, interval after ablation before alpha-blocker treatment, duration of therapy, and number of patients treated were not stated. Mean PVR pretreatment was 15 and 2.5 cc with terazosin (Sudarsanan et al. 2009).

## Radiologic Changes After Valve Ablation

**Posterior urethral dilation decreases within 12 weeks, with persistent dilation suggesting inadequate valve ablation (reviewed above).**

**VUR occurs in approximately 50 % of patients with PUV and is reported to resolve in 25 % to 80 % of renal units, often within a year of valve ablation. Resolution is more likely and occurs more rapidly in grades 1–3 versus 4–5 VUR.**

**Two studies reported decreased HN in 70 % of renal units within 6–12 months of valve ablation.**

### Posterior Urethral Dilation

Thirty patients underwent repeat VCUG 12 weeks following valve ablation, with urethral ratio (transverse midpoint posterior urethral diameter/transverse widest anterior urethral diameter) measured and compared to controls without PUV undergoing VUCG after UTI. Median age of patients at ablation was 13 months (1 day–11 years) and 12 months (2 days–16 years) in controls. Mean pre-ablation urethral ratio was 4.94 ± 2.97, mean post-ablation was 2.13 ± 1.19 ($p$ < 0.001), and was 1.73 ± 0.58 in controls ($p$ < 0.001 vs. pre-ablation and $p$ = 0.1 vs. post-ablation). Two patients with persistent dilation (ratios 5,6) after ablation were both found to have residual valves (Gupta et al. 2010).

### VUR

A prospective observational study included 20 patients undergoing valve ablation at median

15 months (12 days–5.5 years), of which 12 had VUR into19 renal units (grade 1 in 2, grade 3 in 4, grade 4 in 6, and grade 5 in 7). Follow-up VUCG was obtained at 3 and 6 months. By 6 months VUR had resolved in 15 (79 %) renal units, leaving one case each with grade 1–4 and no grade 5 (Priti et al. 2004).

A retrospective review concerned VUR in 127 patients and 200 renal units treated from 1953 to 2003. Data were available for 141 refluxing ureters after valve ablation. Resolution occurred at median 1.28 years (0.04-15.16) in 88 (62 %) renal units, while another 24 (17 %) were removed for poor function, and 29 (21 %) were reimplanted. Resolution was significantly faster for unilateral VUR (median 0.7 vs. 1.36 years), and for grades 1–3 (median 0.68 years) versus grades 4 and 5 (median 1.47 years), and was not influenced by ipsilateral renal function (<10 %, found in 22/73 with scintigraphy). Likelihood for specific grades to resolve was not stated (Heikkila et al. 2009).

Of 73 consecutive patients with PUV, 35 (48 %) had VUR, unilateral in 18. There was mean follow-up of 5.6 years (1–17) in this retrospective analysis, during which resolution occurred in 11/44 (25 %) units at a mean of 12 months. There was no difference in unilateral versus bilateral reflux resolution, nor in resolution according to ipsilateral function <20 % versus greater, but grades 1–3 were more likely than grades 4–5 to resolve (6/9 [67 %] versus 5/35 [14 %], $p = 0.0038$) (Hassan et al. 2003).

Another retrospective series included 65 consecutive boys with PUV treated from 2001 to 2007 with primary valve ablation at median age 1.5 months (1 day–13 years). VUR occurred in 26 (40 %), unilateral in 18. During median follow-up of 24 months (6–75), 11/32 (34 %) ureters resolved (grades not stated). (Sudarsanan et al. 2009).

## Hydronephrosis

The prospective study by Priti et al. (2004) also reported renal ultrasonography findings at ablation and repeated 3 and 6 months following ablation, reported as grades 0–3. All initially had HN, which was bilateral in 90 %. During follow-up HN resolved in one patient, but diminished significantly in the remainder; of 17 renal units with moderate to severe dilation, 5 (29 %) had persistent moderate to severe HN at 6 months after ablation.

Fifty consecutive patients, mean, median age 2 years (1–12), undergoing valve ablation during a 3-year period from 2004 to 2007 had follow-up a mean of 30 months and minimum of 1 year. Pre-ablation HN was described as 0 in 8 (16 %), grade 1 in 10 (20 %), grade 2 in 12 (24 %), grade 3 in 14 (28 %), and grade 4 in 6 (12 %) (grading system not defined). At 1 year after ablation, 31 (62 %) had grade 0, 4 (8 %) had grade 3, and none had grade 4 HN (Gupta et al. 2009).

## Ureteral Reimplantation or Endoscopic Injection

**As discussed above, VUR often resolves within a year of valve ablation, and it is unclear if VUR represents a modifiable factor for renal function.**

**One study found less postoperative VUR after excisional tapering with versus without psoas hitch reimplantation.**

VUR was present in 38/54 (70 %) boys in a retrospective study. Spontaneous resolution occurred in 9/38 (24 %) after ablation, and nephrectomy was done in 7 patients for nonfunction. Twenty patients had surgical reflux resolution, by reimplant in 5 and by injection with polytetrafluoroethylene in 15 patients and 24 ureters (grade 4 in 6, grade 5 in 18). Indications for intervention were not described. Age at injection was a mean of 2.2 years (8 months–6 years), and all but one (with UTI) had injection ≥12 months after valve ablation. A single injection resolved VUR in 17/24 (71 %) ureters, and with up to three injections all were reported successful. The number and results of any subsequent VCUGs, if done, were not discussed (Puri and Kumar 1996).

A retrospective review of 106 boys with PUV found 20 patients undergoing ureteral surgery, by reimplantation in 25 renal units and transureteroureterostomy in 7 renal units. Of these, 5 patients and 12 ureters had VUR, while the remaining 15 patients and 20 ureters had UVJ obstruction (discussed below). Mean interval from valve ablation to reimplant was 1.8 years (6 months–4.5 years) for the entire group, and grade of VUR was not clearly stated, but was grade 4 or 5 "in most." Excisional tapering to 10–12-Fr was done in all 25 reimplanted ureters, with a psoas hitch in 18 and not in the other 7, whose technique was not further described. Four out of five refluxing patients had persistent VUR; grades were not stated. Of the entire group undergoing surgery for either obstruction or VUR, psoas hitch reimplantation had significantly less postoperative VUR than did repairs without hitch (2/18 ureters with hitch vs. 7/7 ureters without hitch, $p<0.004$) (El-Sherbiny et al. 2002).

## Ureterovesical Junction Obstruction

**Impaired ureteral drainage across the ureterovesical junction generally has been attributed to valve bladder and/or intrinsic ureteral dysfunction rather than UVJ obstruction. Our review found only one article specifically reporting UVJ obstruction in 12 % of PUV patients.**

El-Sherbiny et al. (2002) diagnosed UVJ obstruction in13/106 (12 %) boys and 20 renal units following valve ablation at a mean of 1.8 years (6 months–4.5 years). All had bilateral "grade 3 or 4" HN (grading scale not defined). Diuretic renography demonstrated T1/2 >20 min in 7 patients and 11 renal units, while Whitaker test found renal pressures >22 cm $H_2O$ over bladder pressures in 6 patients and 9 renal units with supravesical diversion before repair.

## Nephrectomy to Reduce Urine Output

**One study reported that unilateral nephrectomy reduced polyuria by 40 % before renal transplant.**

A retrospective study evaluated consecutive patients undergoing unilateral or bilateral native nephrectomy from a total of 126 consecutive pediatric kidney transplants. These nephrectomies were done a median of 1.9 months (0–41) before transplant. The indication was polyuria, defined as sustained urine output >2.5 cc/kg/h, in 22 children, of which 10 had congenital urinary tract anomalies that were not further described. Of these 22 children, 15 had unilateral surgery (sometimes the first of staged removal) and median urine output decreased from 3.9 cc/kg/h to 2.4 cc/kg/h, a change of −40 % from 2.1 to 1.4 L per day (Ghane Sharbaf et al. 2012).

## Renal Transplantation

**Several cohort studies indicate that graft survival is similar in patients with renal failure due to PUV versus those with medical renal disease.**

A retrospective cohort study compared graft survival after living unrelated donor transplantation in 15 PUV patients, mean age 12.5 ± 2.8 years, to 45 matched controls of similar age also receiving living unrelated grafts and all managed with a similar immunosuppressive protocol. Preoperative urologic assessment included VCUG, cystoscopy, and UD; of these 15 patients, 4 took anticholinergics, 2 used CIC, and 9 had prior ileocystoplasty. There were no differences in acute or chronic rejection episodes between patients and controls, and mean graft survival was 7 years in patients and 6.2 in controls, $p=0.9$ (Otukesh et al. 2008).

Another retrospective study compared renal graft outcomes in 18 PUV patients transplanted at mean age 9.1 years (1–18) 1:1 with a selected control group with medical renal disease of similar age, donor type, and immunosuppressive therapies. No pre-transplant UD or bladder reconstruction was done in any patient, but no mention was made of other medical therapies for bladder management, if any, and the authors admitted there were neither UD data nor voiding histories for any PUV patients before or during 10 years of post-transplant follow-up. Nevertheless, 10-year graft survival was not significantly different between patients and controls, 54 % versus 41 % $p=0.35$ (Indudhara et al. 1998).

A third retrospective matched cohort study compared cadaveric transplantation in 19 PUV patients at mean age of 10 years (1.3–17) to 62 controls with glomerulonephropathies and 42 controls with renal dysplasia—neither control group including patients with known bladder dysfunction. There were no differences in PUV patients versus controls for graft survival at 1, 5, or 10 years. In this series, all PUV patients underwent pre-transplant UD, which diagnosed "severe" bladder dysfunction in 8 (42 %) described as detrusor overactivity, poor compliance, reduced bladder capacity, and bladder sphincter "incoordination" (none specifically defined). Pre-transplant augmentation was performed in five of these. All transplant reimplantations were antirefluxing done into the native bladder. No patient had pre-transplant bilateral nephrectomies, but eight had unilateral nephrectomy before or during the transplant, and six had unilateral nephrectomy afterwards, resulting in three with bilateral removal of native kidneys. Another patient had bilateral nephrectomy after transplant, so that altogether 4/19 (21 %) received bilateral nephrectomies. Indications for nephrectomy were not clearly stated (Mendizabal et al. 2006).

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