Objective: Despite worldwide availability of prenatal ultrasound, many patients are diagnosed in adult life with congenital anomalies such as ureteropelvic junction obstruction (UPJO), undescended testicle (UDT), ureterocele, hypospadias, vesicoureteral reflux (VUR) and primary obstructing megaureter (POM). The aim of this review was to describe these clinical conditions and their suggested management based on the available medical literature.

Review: Adult UPJO is not a rare condition; symptomatic patients should be treated rather than observed. Treatment options are nephrectomy for non-functioning kidneys and reconstructive surgery for functioning renal units. The adult UDT has low fertility potential and increased cancer risk; hence most of the data in the literature indicate performing an orchietomy. Adult ureteroceles are usually related to single systems and they are intravesical and less obstructive. For symptomatic patients endoscopic incision showed high efficacy for symptom elimination with minimal side effects. Primary hypospadias correction in the adult patient is feasible, but success rates are low compared to the pediatric age group. Secondary correction, whether primary correction was performed in childhood or adulthood, is a challenging task with a high complication rate. Treatment decisions regarding adult patients with VUR are difficult to make as the available data are inconsistent; there is no strict evidence that reflux in an adult is directly related to renal growth impairment, ascending pyelonephritis, and/or embryo loss in a pregnant woman. In contrast to the pediatric age group, adult POM is usually a symptomatic condition and related to a high complication rate including infections, stone formation and renal failure. Spontaneous resolution is rare and hence active surgical management is advocated.
Introduction

The urology system is easily demonstrated in prenatal sonography [1]. Prenatal identification of congenital urological anomalies has tremendous impact on the postnatal baby’s life. Prenatal parental consultation enables explanation of the predicted anomalies and planning a postnatal follow-up regimen. Preparations for a high-risk baby with urologically related anomalies may reduce antenatal mortality and morbidity [2]. Congenital anomalies diagnosed in adulthood may require a different perspective than in children regarding management and surgical correction. Here we present descriptions of some of the anomalies for which surgical management is an option in children. We propose management options according to the current medical literature.

Adult Ureteropelvic Junction Obstruction (UPJO)

The incidence of UPJO in the pediatric age group is well defined, affecting around 60% of all newborns with hydronephrosis. The exact incidence of primary congenital UPJO in adults is unknown, although estimated as not uncommon [3]. In contrast to prenatal screening, adult UPJO may be discovered in several ways: (a) evaluation following symptoms such as chronic back pain, acute renal colic, especially after fluid overload, hematuria, UTI and pyelonephritis; and (b) incidentally during diagnostic imaging of the abdomen or spinal area for other problems such as abdominal or back pain. Diagnosis of adult UPJO does not differ from that of the pediatric age group, using nuclear medicine static and dynamic renal scans, intravenous pyelography, or CT urography. Unfortunately, there is no consensus regarding which is the optimal imaging modality to determine obstruction. Moreover, there are no uniform standards for how to perform the above-mentioned diagnostic tests, and various protocols exist. Management decisions in the adult are quite different. In children, the main goal is to eliminate the obstruction in order to restore maximal renal function and to allow maximal growth of the kidney. In adults, the ability to restore renal function or to allow compensatory renal growth following surgery is limited, as chronic obstruction may have caused irreversible changes in the renal parenchyma and vasculature, while maximal growth potential has already been achieved. Factors that should guide the physician regarding the management of adult UPJO are: symptoms, function, age and co-morbidities.

Symptomatic patients with UPJO should be treated when symptoms endanger the patient: recurrent UTI, pyelonephritis or hypertension can be life threatening, especially in elderly patients with other co-morbidities. Non-life-threatening symptoms such as chronic pain that interfere with a patient’s quality of life may also be an indication for definitive treatment. Definitive treatment includes reconstructive surgery to eliminate the obstruction or nephrectomy. In order to choose the most suitable arm of treatment for the patient, several factors should be taken into consideration: the function of the obstructed kidney, the function of the other kidney, the patient’s age and co-morbidities. A non-functioning symptomatic kidney in the presence of a normal functioning contralateral kidney should be removed. In patients with a single kidney or those with overall poor renal function, reconstructive surgery should be offered.

Nephrectomy in adults is a well established operation. The decision between open and laparoscopic routes should be made by the patient and the urologist based on individual circumstances. Both can achieve the surgical goal successfully, with the advantage of a shorter recovery period with less analgesic consumption in patients who undergo laparoscopic nephrectomy [4].

Reconstructive surgery in adults can be performed by a wider range of surgical techniques than in children. The open surgical approach — open pyeloplasty — is still considered the gold standard for UPJO in adults, with a 91% success rate overall and 100% success rate for grade 1–3/4 hydronephrosis [5]. Laparoscopic and laparoscopic robotic pyeloplasty already show similar success rates to open surgery. Bauer et al. [6] compared 42 laparoscopic pyeloplasties to 35 patients who underwent open surgery, and the overall success rates were comparable (98% and 94%, respectively). Robot-assisted laparoscopic pyeloplasty also showed comparable success rates with minimal complications and short recovery time [7,8]. In contrast to the pediatric age group, adult patient may choose to undergo endoscopic correction of the UPJO. Endoscopic endopyelotomy has the advantage of being a minimally invasive procedure that may be performed as ambulatory or day-care surgery, with short recovery time and rapid (within 24 h) return to normal activity. Due to its minimally invasive nature and the ability to perform the procedure under regional anesthesia, endoscopic endopyelotomy is more suitable to patients with co-morbidities. The idea was originally described by Wickham and Kellet [9] who, using an endoscopic urethrotome via an antegrade endoscopic approach, performed a full-thickness incision of the upper ureter and PUJ. Among the three initially treated patients, two achieved improved drainage. Danuser et al. assessed the results of 212 consecutive antegrade endopyelotomies over a period of 8 years and showed overall 85% success. With the introduction of smaller endoscopes with better visual imaging and the use of laser energy antegrade and later on retrograde, endopyelotomies became even easier to perform. Today, endopyelotomy for UPJO in adults is considered as first-line treatment in various urological centers [10]; however, more recent reviews with longer-term follow up show that success rates are worse than previously reported. Dimarco et al. [11] assessed the long-term results...
of antegrade endopyelotomy (182 patients) and open pyelo-
plasty (175 patients). The estimated 3-, 5-, and 10-year
recurrence-free survival rates for the endopyelotomy group
were only 63%, 55% and 41%, respectively, compared to
85%, 80% and 75% for the pyeloplasty group (p < 0.001).
Additional crucial information arising from this study is the
fact that failures continue to appear after 5 and 10 years,
and patients should be followed accordingly. Rassweiler
et al. [12] showed similar results comparing the success rates
of laser endopyelotomy (113 patients) and laparoscopic pye-
lopasty (143 patients). The laparoscopic procedure showed
a superior overall success rate of 94.4% versus 72.6%.

In summary, for the adult patient with UPJO various
optional treatment modalities are available: nephrectomy
or a reconstructive procedure. Although minimally invasive
endoscopic techniques enable almost any patient to have
definitive treatment, recent reports show disappointing re-
results and late appearance failure. Patients should be followed
beyond 10 years, and given advice regarding the higher suc-
cess rate of ‘formal’ open and laparoscopic pyeloplasty.

**Adult cryptorchidism (undescended testis, UDT)**

In the pediatric age group, orchidopexy at an early age is
guided by the need to preserve functions being damaged by
the inappropriate anatomic position of the UDT, i.e. sperm
production and testosterone secretion. Unfortunately, sper-
matogenesis in the UDT decays with time and after the age of
2 years the rate of germ-cell aplasia irreversibly accelerates.
Rogers et al. [13] analyzed the histology of 52 resected UDTs.
Patients’ mean age at surgery was 26 years, and among the 52
specimens only one (1.9%) testis showed normal spermato-
genesis. In the remaining testicles the histology slices
showed Sertoli cells only in 30 (58%) patients, maturation
arrest in 15 (28.5%), and testicular agenesis in six (11.5%).
The authors concluded that the majority of UDTs cannot con-
tribute to fertility. Grasso et al. [14] produced similar results
by performing testicular biopsies in 22 patients who had post-
pubertal orchidopexy for cryptorchidism. More than 83% of
the biopsies showed azoospermia and severe oligospermia.
Although Leydig cells are less vulnerable to damage, endo-
crine function is also impaired in the adult UDT [15].

Concerning malignancy it has already been shown that
orchidopexy does not prevent or change the rate of
testicular cancer [16]; hence orchidopexy may only improve
the ability to palpate the testicle. Due to its abnormal
location, UDT may cause discomfort and undergo torsion.
Zilberman et al. [17] showed that the rate of salvaging UDT
torsion is lower than for normally positioned testes. Among
11 patients with torsion of UDT only two testicles (18%) re-
mained viable; five had massive necrosis during surgery and
had to be resected and four vanished post orchidopexy.

In summary, the majority of adult UDTs have very low
fertility potential, impairment of endocrine function and
increased risk of testicular cancer. Hence, in patients with
a normal contralateral testicle, UDT orchietomy should be
offered. In patients with a single testis or bilateral UDT,
preservative management may be considered mandating
careful follow up and awareness of the patient and
physician of the possible complications.

**Adult ureteroceles**

In the pediatric age group ureteroceles may be related to
a wide variety of complex anomalies, such as duplex kidney,
ectopic ureter, bladder outlet obstruction, incontinence and
reflux. In adults most ureteroceles are related to a single
system. They are intravesically located and the degree of
obstruction is less severe. Presentation in an adult could be
either by flank/back pain and recurrent UTI, or asymptom-
atic hydronephrosis detected incidentally. Stasis at the
ureteral meatus due to the partial obstruction of the
ureteroceles may induce stone formation inside the ureter-
oceles, causing renal colic and/or UTI. A single case report
in the English medical literature described a patient who
deteriorated to renal failure due to bilateral ureteroceles
[18]. Diagnosis can be established with ultrasound or intrave-
sous pyelography in patients with normal renal function.
Ultrasound demonstrates a cystic mass within the bladder;
Doppler mode can demonstrate a urine jet at the meatus of
the ureteroceles with volume changes during urine expul-
sion from the meatus. With evaluation of renal parenchyma,
degree of hydronephrosis and jet sign with the advantage of
no radiation energy exposure, ultrasound may be a sufficient
tool to diagnose and follow up adult patients with uretero-
cele [19]. Intravenous pyelography may demonstrate the
classic picture of the ‘cobra head’ intravesically. In addition,
renal function, excretion delay and degree of hydronephro-
sis can be assessed. Management should be based on symp-
toms and renal function. Symptomatic patients should be
treated. In the case of no or poorly functioning kidney with
normal contralateral kidney and normal renal function ne-
phrectomy should be offered. For symptomatic functioning
kidney endoscopic incision of the ureteroceles with stone
fragmentation will lead to symptom elimination with mini-
mal morbidity. Chtourou et al. [20] described 20 patients
with ureteroceles diagnosed due to chronic back pain; the
mean age was 48 years, 16 had a single system and four du-
pex. All had an endoscopic incision and, in the presence of
stones, fragmentation. Elimination of pain was successfully
achieved in all patients. In a single patient the procedure
was complicated with sepsis and one patient developed tran-
sient VUR. Vasu et al. [18] described a rare case of bilateral
ureteroceles causing progressive renal failure that reversed
following bilateral incision. Although these are single case
reports, such complications should be born in mind.

In summary, adult ureteroceles are usually intravesical
and only mildly obstructing. Although it seems logical to
treat symptomatic patients with endoscopic incision, phy-
sicians should be aware of the lack of data in the medical
literature; hence our recommendations for adults are based
on scanty reports.

**Adult hypospadias**

Distal hypospadias in itself poses only a cosmetic problem,
and sometimes may cause deviation of the urinary stream.
Erectile function is fully preserved and is not related to the
anomaly. Ejaculation and fertility are not impaired. There
are very little data regarding hypospadias correction in
adults. Adayener and Akyol [21] reviewed the results of pri-
mary distal hypospadias repair in 80 adults and secondary
repair in additional 17. The location of the meatus was glan-
nular in six, coronal in 35 and subcoronal in 56. Operative
technique used was meatal advancement in 42, Mathieu
in 41 and tubularized incised plate in 14. Overall success
rate was 91.3%. However, the meatal position-related suc-
cess rate was slightly different with 91% success for glanular
and coronal hypospasias and only 85% success rate for sub-
coronal hypospasias. This rate is low compared to the pedi-
astric age group. Senkul et al. [22] achieved a 89.9% success
rate operating on 59 adults with a mean age of 22 years,
among whom 48 had distal, nine mid-shaft and two prox-
imal hypospasias. Operating on secondary hypospasias
yields much lower success rates, with no differences in
complications and failure rates noted whether the failed
primary correction was performed in childhood or in adult-
hood. Barbagli et al. [23] assessed 60 adults with complica-
tions following pediatric hypospasias surgery; 36% of the
patients had one complication and 64% had two or more.
Complications included stricture 34, residual hypospasias
26, fistula 18, meatal stenosis 11, penile curvature nine,
hair four, diverticula two and stone in one. Twenty-nine pa-
tients had one-stage repair with buccal or skin grafts or di-
rect repair, and 31 underwent multistage repairs with buccal
or skin grafts. Forty-five (75%) patients had a final
successful outcome, 15 (25%) failed. One-stage repair pro-
vided 24 (82.7%) successes and five (17.3%) failures. Multi-
stage repair provided 21 (67.7%) successes and 10 (32.3%)
failures. The authors concluded that adults with complica-
tions following childhood hypospasias repair are still a diffi-
cult population to treat with a high failure rate for re-
operative surgery. Senkul et al. showed a 27% complication
rate for second attempt hypospasias correction in patients
operated in adulthood.

For comparison we review results from the experience
in the pediatric age group. Snidgrass and Yucel [24] reported
about 30 pediatric patients who had tubularized incised
plate repair for mid-shaft hypospasias. Fistulae were not-
ticed in 3/30 (10%) and an additional two patients (6%)
had one stricture and one glans dehiscence. Cheng et al.
[25] reported about 514 pediatric patients operated for
distal (414) and mid-shaft (100) hypospasias. For the distal
repair no fistulae were reported and only a single case (0.2%)
of stenosis, in the mid-shaft group three (3%) fistulae were
noticed and one (1%) case of urethral stenosis. Combining
both groups overall complications were less than 1%. Ham-
mouda et al. [26] with at least 6 months’ follow up showed
that 4% (2/48) of patients who had tubularized incised plate
repair developed fistulae and 10% (5/48) meatal stenosis.
Duncan and Snidgrass [27] reviewing 26 articles comprising
around 2035 pediatric patients operated for distal hypospa-
dias revealed an average complication rate of only 9%.

In summary, primary and secondary hypospasias cor-
rection in adults is feasible. Surgical success rates are
lower and there is a high complication rate compared to
the pediatric age group. Patients should be informed
regarding these data.

**Adult reflux**

The treatment of pediatric reflux underwent tremendous
changes in the past decade. Open surgery was replaced
with conservative medical treatment and minimally in-
vasive endoscopic procedures. Although a high percentage
of patients with reflux will resolve spontaneously, in
10–40% reflux will persist (depending mainly on grade and
patient age). Symptomatic reflux, causing new renal scars,
and deterioration of renal function should be treated;
however, the proper management for asymptomatic per-
sistent reflux in the adult is yet to be determined.

Consideration of increased risk in the adult patient with
reflux may arise for several reasons. In females, adulthood
may be related to an increased rate of bacterial UTI during
sexual activity and at menopause. Pregnancy is related to
overall urinary tract dysfunction, bacteriuria and UTI.
Above all the chance of spontaneous resolution of the
reflux in adulthood is low. On the other hand, the adult
kidney is more resistant to infection, is less susceptible to
developing new scars following infection, and the kidney
growth potential has already been achieved. Based on the
available medical literature it is not possible yet to
determine the exact effect of asymptomatic reflux on the
adult patient. Choi et al. [28] assessed 86 adult females
with pyelonephritis for the significance of VUR. All patients
underwent VCUG, 31 of them at the 3rd treatment day and
55 at the 7th day. Only two patients (2.3%) had low-grade
reflux. Hence the authors concluded that, unlike in
children, VUR is not a significant factor causing ascending
infection leading to the development of acute pyelonephri-
ts in adult women. el-Khatib et al. [29] assessed the influ-
ence of reflux on the course of renal failure in adult
patients with reflux nephropathy. Among several factors
studied the presence of persistent reflux had no indepen-
dent influence on the course of renal failure; the most im-
portant factor that was associated with renal function
deterioration was proteinuria.

Olbing et al. [30] showed in a 10-year follow-up study
that even in the presence of high-grade reflux the appear-
ce of new scars is rare after the age of 5; moreover
there was no difference in new scar formation between
patients treated conservatively and patients who were
operated surgically. Olbing et al. [31] in a different study
assessed renal growth in patients with severe reflux. The
study compared renal growth in two groups of patients
with high-grade reflux, one treated conservatively and
the other surgically. There was no significant difference
in renal growth during 10 years between surgical and med-
tical treatment in patients with severe reflux. The question
whether reflux can impair pregnancy outcome was studied
in several works: el-Khatib et al. [32] studied the outcome
of 345 pregnancies in 137 women with reflux nephropathy.
Overall fetal loss was 48 (14%) of which only six (2%) were
therapeutic abortions. Fifty-two pregnancies took place in
women with plasma creatinine >0.11 mmol/l prior to con-
ception. Fetal loss after 12 weeks’ gestation (excluding
therapeutic abortions) was 18% compared to 8% in the
104 pregnancies where maternal plasma creatinine was
lower than 0.11 mmol/l (p < 0.05). Maternal complications
were also more common in the impaired renal function
group (p < 0.001). Comparison of pregnancies in women
with unilateral versus bilateral renal scarring revealed no
significant difference in fetal loss but an increased inci-
dence of maternal complications in the bilateral renal
scar group (p < 0.01). Persistent VUR was not associated
with increased fetal loss or maternal risk; however, impaired renal function prior to conception is associated with increased fetal loss and maternal complications in pregnancy.

In summary, currently there is no firm evidence that reflux in an adult is directly related to renal growth impairment, ascending pyelonephritis and/or embryo loss in pregnant woman with reflux.

**Adult primary obstructing megaureter (POM)**

In the pediatric age group a high percentage of patients with megaureters may be manage conservatively as long as renal function is good. Although adult POM is uncommon patients are usually symptomatic and need intervention. Hemal et al. [33] described a series of 55 adult patients treated over a period of 12 years: 52/55 (94%) were symptomatic and the mean follow-up time was 7 years. In this series five (12%) patients with bilateral POM suffered already from renal failure. Tatlisen et al. [34] described five patients with POM who all suffered from flank pain. Dorairajan et al. [35] reported a series of 37 adult patients with POM: 26 (70%) had flank pain, 15 (40%) UTI, 17 (46%) urinary calculus and five (13.5%) presented with azotemia.

Diagnostic imaging included ultrasound, intravenous pyelography, diuretic renogram and VCUG as in the pediatric age group. Gherin et al. [36] used antegrade injection of contrast material and multi-detector CT to diagnose urinary tract obstruction including megaureters without the need for intravenous contrast material injection, so that patients with impaired renal function could also be evaluated.

In contrast to the pediatric age group, uniformly, the medical literature recommends active treatment in adults with POM [33,35,37,38]. Surgical management for functioning renal units includes ureteral neocystostomy (open or laparoscopic) and endoscopic endoureterotomy (electrocoagulation or laser), and nephrectomy for non-functioning units. Hemal et al. [33] operated 41 patients for POM: 38 patients underwent reimplantation (21 with tailoring and 17 without) and three endoscopic ureteral meatotomy. Forty three of the 47 (%) showed improvement in collecting system dilatation, and the mean follow-up time was 7 years. In this series five (12%) patients with bilateral POM suffered already from renal failure on diagnosis, despite surgical treatment only one patient improved, and two died of renal failure. Tatlisen and Ekmeckioglu [34] performed direct nipple ureteroneocystostomy in five adult patients in order to avoid ureteral tailoring. Ansari et al. [39] successfully performed three laparoscopic ureteroneocystostomies for POM; ureteral tailoring was done extracorporeally followed by Lich-Gregoir reimplantation. Biyani and Powell [40] performed four endoscopic laser ureterotomies for adult POM; a full-thickness 2.5 cm long incision was performed on the intramural and juxtavesical part of the ureter followed by internal stent diversion. In a 24-month follow-up period all patients improved; a single female patient developed reflux. Bapat et al. [37] used a cutting current to treat six POM. Follow up of 1–4 years showed reduction of hydronephrosis and elimination of symptoms.

In summary, adult POM in contrast to children is usually symptomatic. Due to symptoms complications such as infections, stones and reduced renal function, and low spontaneous resolution active management is advocated. For functioning units, reconstructive surgery is advised; for poorly functioning units nephrectomy would be a better option. Ureteral reimplantation with or without tailoring is an established procedure. Endoscopic endoureterotomy showed good results, but should be evaluated for a longer term with higher numbers of patients.

**Conclusions**

Diagnosis of a congenital urological anomaly in an adult is not a rare event. Management considerations in the adult patient are sometimes different compared to the pediatric age group. In adults, more emphasis is given to symptoms, patient age and additional co-morbidities. The adult age group may benefit from a wider choice of surgical techniques that cannot be applied in children such as endoscopic procedures. Despite utilizing similar operational skills and techniques, surgical outcome in adults may be different from that in children, as is clear in hypospadias cases. In general, there is a relatively large arsenal of available therapeutic solutions to offer to adult patients with congenital urological anomalies.

**References**


