Aims

- To discuss important specific diagnoses relevant to renal physicians
- To describe presentations and management of these conditions
- To outline surgical interventions available both minimally invasive and open surgery
Paediatric urological conditions

- Pelviureteric junction obstruction
- Vesicoureteric reflux
- Posterior urethral valves
- Neuropathic bladder
Pelviureteric junction obstruction

- Most common cause of paediatric hydronephrosis
- Majority detected on prenatal ultrasound
- Prenatally detected cases usually completely asymptomatic
- Older patients present with pain, UTI, haematuria, stone formation, following trauma or with hypertension
Antenatal detection

- 1% of fetal kidneys show mild dilatation
- 1 in 600 pregnancies fetal uropathy
- 35% PUJO anomaly
- PUJO occurs in 1 per 1000-2000 newborns

Aetiology

- **Extrinsic abnormalities**
  - Aberrant vessels
  - Kinks
  - Bands
  - High ureteric insertion
- **Intraluminal**
  - Stones
  - PUJ fibroepithelial polyps
  - Fungal balls

Associated with?

- Horseshoe kidneys
- Pelvic kidneys
- Duplex collecting systems
  - Usually involves the lower pole moiety
Current indications for surgery

- Renal preserving surgery if
  - Function initially or subsequently < 40%
  - Increasing hydronephrosis
  - Definitely if initial AP > 50mm
  - Symptoms - pain, UTI, haematuria, abdominal mass
  - Persisting obstruction after age 3

- More likely to intervene early in cases of
  - Severe bilateral PUJO
  - Single kidneys

- Differential function < 10% nephrectomy
Surgical management of PUJO

- **Endourological procedures**
  - Balloon dilatation
  - Endopyelotomy
  - Endopyeloplasty

- **Open procedures**
  - Anderson-Hynes dismembered pyeloplasty (1949)
  - Culp/De Weerd pyeloplasty (1951)
  - Y-V Foley advancement (1936)

- **Laparoscopic pyeloplasty**
  - Conventional
  - Robotic surgery

- **Laparoscopic transposition lower pole vessels**

*Figure 1 - Anderson-Hynes pyeloplasty.*

Gold Standard
Open pyeloplasty

- Incision
  - Anterior - muscle split
  - Loin - muscle cut
  - Posterior lumbotomy
  - Minimally invasive open pyeloplasty (< 1 year old incisions 1-1.5cm)

Laparoscopic pyeloplasty

- Retroperitoneal
- Transperitoneal
- Robotic-assisted
- Reported decreased hospital stay, decreased narcotic use
- Operation times longer than open pyeloplasty ~ 3 hours

Vesicoureteric reflux

- 1% general population
- Boys predominate in prenatally detected cases and with UTI in 1st year of life (usually high grade reflux)
- VUR and UTI later in childhood mainly girls (usually low grade reflux) - dysfunctional elimination syndrome
Primary VUR

- Short submucosal tunnel
- Laterally sited ureteric orifice
- Congenital – ureteric bud anomaly
Secondary VUR

- Abnormal bladder function
- Raised intravesical pressure
- eg Neuropathic bladder, PUV
Reflux nephropathy

- Congenital
- Acquired
  - Infective
  - Intrarenal reflux
  - Compound papillae
  - Ransley’s ‘big bang’ hypothesis
  - Greatest risk <4 years old
Methods of urine collection
Management of VUR

- Conservative - spontaneous resolution rates
- Role of uroprophylaxis
- Treat any underlying bladder dysfunction
- Treat constipation
- “Surgical management of VUR is not routinely recommended” NICE CG54 2007
Surgery: vesicoureteric reflux

- Controversy on when or if ever required
- Failure of medical management – breakthrough infections
- Problems with compliance/follow-up
- Poor renal function
- Development of new renal scars
- Solitary kidneys
- Pain

VUR - ureteric reimplantation
VUR - alternatives to reimplantation

- STING/HIT technique
- Nephroureterectomy
- Vesicostomy
- CIC
Posterior urethral valves

- First described by Hugh Hampton Young 1919
- Bladder outflow obstruction in boys
- 1 in 5,000 live male births
- Common cause of renal failure in childhood
- 25-30% ESRD by puberty
Clinical presentation

- Prenatally detected
- Infection
- Abdominal mass
- Voiding symptoms
- Respiratory insufficiency, pulmonary hypoplasia
Prenatal detection

- male fetus
- thick-walled bladder, infrequent/incomplete voiding
- upper tract dilatation
- echogenic kidneys
- oligohydramnios in severe cases
Initial Investigations

- renal ultrasound
- micturating cystourethrogram
- functional imaging - DMSA
Surgical management

- transurethral valve ablation - neonatal resectoscope, neodymium:YAG laser, fluroscopically guided Whitaker hook, balloon disruption
- antegrade valve disruption
- urinary diversion-nephrostomy, pyelostomy, ureterostomy or vesicostomy
Management of the ‘valves bladder’

- uroflow rates/post void residuals/urodynamics
- voiding regimes
- anticholinergics, α blockers
- Intermittent, overnight catheterisation
- bladder augmentation (ureterocystoplasty, enterocystoplasty)
- Mitrofanoff stoma
Renal transplantation

- 15-20% of PUV patients
- More UTI’s post transplant than other groups
- Graft survival lower when transplanting into ‘valves bladder’
- Careful preoperative evaluation—augmentation and Mitrofanoff procedure
Neuropathic bladder

Aetiology

- Congenital e.g. myelomeningocele, lumbosacral lipoma, sacral agenesis, occult
- Acquired e.g. cord ischaemia, tumour, dermal sinus (intradural abscess), trauma
Urinary incontinence

- Organic causes rare - 1% of referrals from GP’s
- Continuous daytime wetting always structural or neurological
- Abdomen, genitalia, spine, neurology – palpable bladder, phimosis, meatal stenosis, epispadias, U/G sinus, hairy patches, cutaneous haemangiomata, sacral agenesis, muscle wasting, foot deformities, sacral segment innervation
- If you don’t look you won’t find
- Haemangiomata can fade in time
- Don’t forget VUR can be secondary
- Ultrasound useful screening tool
Neuropathic bladder

Surgery

- Cutaneous vesicostomy - closure age 3-4
- Bladder augmentation
- Creation of continent catheterizable channel e.g. Mitrofanoff
- Surgery for sphincter incompetence
- Continent diversion
Bladder augmentation

- Enterocystoplasty
- Gastrocystoplasty
- Autoaugmentation
- Ureterocystoplasty
The Mitrofanoff procedure

- Used when urethral access difficult or during planned bladder neck closure
- First described by Paul Mitrofanoff 1980
- Appendix most commonly used channel
- Other conduits - distal ureter, gastric tube, tapered ileal segment, prepuce, bladder flap, fallopian tube
- Monti procedure - described 1997, ‘appendiceal tube’ reconfigured from ileum or colonic segments
Appendicovesicostomy

- Conduit of choice
- But may be atretic, short, previously removed or used for in-situ ACE procedure
- Stoma site – umbilicus, right lower quadrant
- Recessed skin flaps decrease rate of stomal stenosis and improve cosmesis
- VQZ stoma
Appendicovesicostomy
Yang-Monti catheterizable channel

Surgery for sphincter incompetence

- Endoscopic bulking agents e.g. teflon, collagen, Macroplastique, Deflux 22%-48% cure rate (Bomolaski 1996, Lottman 2006)
- Bladder neck sling procedures - 93% success rate (Gosalbez 1998)
- Urethral lengthening procedures e.g. Kropp and Pippi-Salle procedures 78% success rate (Belman 1989)
- Artificial urinary sphincter - success rate 86% (Herndon 2003)
- Periurethral constrictor – bladder neck or bulbar urethra
Future technologies

- Tissue engineering
- Stem cell research

An organ transplant without rejection? This spring Kaitlyne McNamara.............

Atala et al. Tissue-engineered autologous bladders for patients needing cystoplasty. Lancet 2006 – 7 patients follow up 22-61 months, mean compliance increase 2.79 fold.