Upper urinary tract obstruction

Pelvi-ureteric junction obstruction
Prior to routine antenatal testing, most children with presented with pain, UTI or incidentally. Now majority of patients identified on antenatal screening. PUJO accounts for 30–50% of patients with antenatally detected hydronephrosis
Incidence PUJO based on second trimester screening 1:750 – 1:1000
[However should be appreciated that this is a broad estimate, as some cases arise de novo in childhood and a significant proportion of prenatally detected cases resolve spontaneously]
Equal sex ratio
Left kidney twice as commonly affected
Bilateral in 10–40% of cases

Aetiology
Intrinsic
short stenotic segment
Crossing vessel
~ 5% infants (cf. 30% adults)
Ureteric folds
may explain spontaneous resolution as ‘ironed out’ with growth
Secondary causes
Horseshoe kidney
Retrocaval ureter (right-sided)
Gross VUR with kinking of PUJ

Natural history of antenatally diagnosed PUJO
Controversial
Long-term observational studies (Ransley & Dhillon, Great Ormond Street 1990) in patients with good renal function showed:
17% pyeloplasty for deteriorating renal function
56% stable renal function
27% spontaneous resolution
Spontaneous resolution has been related to renal pelvis AP diameter USS imaging (Desai & Dhillon 1999, Progress in Paediatric Urology; updated in 2006)
< 20 mm 3% require surgery
20 – 29 mm 20% surgery
30 – 39 mm 50% surgery
40 – 49 mm 80% surgery
> 50 mm 100% surgery
Overall if > 30mm approximately 90% of patients end up with surgery
Above raises question of follow-up. Most centres use a combination of MAG-3 renogram and USS to monitor progress. A split function of < 40% on affected side reported as the threshold for intervention in the form of pyeloplasty.

Investigation
Antenatal USS
Normal third trimester neonatal renal pelvis AP diameter 6mm
Abnormal fetal dilatation
4mm in second trimester
7mm in third trimester

Abnormal scan in second trimester should trigger further scan in third trimester
Any degree of ureteric dilatation pathological on prenatal USS

Postnatal USS
Neonatal urine output reduced immediately following delivery
Repeat USS should be deferred at least 48 hours when normal diuresis established. Often performed at one week post-natally
If significant (>15mm AP diameter) dilatation exists without ureteric dilatation, then MAG-3 renogram required ideally after 6 weeks.
If bilateral hydronephrosis identified then MCU probably first line as VUR more common.
If fetal hydronephrosis has resolved or mild (<15mm) traditionally MCU was performed to exclude VUR. However recent studies have shown that 2 normal postnatal US studies effectively rules out clinically significant VUR (Ismaili 2002) – therefore a repeat USS performed by some.

MAG-3 diuretic renography
Next step in the absence of ureteric dilatation
Filtration and secretion reduced background counts and enhances excretion (and thus time on the scanner for kids)
Generally good for determining obstruction*
Can provide an assessment of split function, but in the presence of severely impaired renal function, DMSA more accurate
* NB. Caution should be used in interpreting MAG-3 curves in infants, as impaired response to diuretic. Also children < 2yrs have renal pelvis expansion in response to fluid loading, which can affect washout curves. Important to standardise renogram technique

Management
(i) Conservative management
For normally functioning kidneys with AP renal pelvis diameter 20-30mm (opinion varies)
Lower limit of normal 40% in affected kidney – widely used but empirically determined. Some evidence that isotopic function poorly correlates with histological features of obstructive nephropathy. Some authors advocate using Single Kidney GFR (split function on MAG3 x $^{51}$Cr-EDTA clearance) to more accurately document a decline in function, but experimental and expensive
NB. Renography and SKGFR are unhelpful in kids with bilateral PUJO (does deterioration on one side represent improvement on the other?) – AP pelvic diameter alone used to determine need for Rx in this situation
As in adults, resistive index has been shown to correlate with obstruction (Gilbert 1993) but subject to observer error and not widely performed

(ii) Surgery
Indications for pyeloplasty
Symptoms (pain/UTI)
- Split function < 40% on the affected side
- AP diameter > 50mm
- Decline in function of > 10% of affected side

Operative considerations
- Culp and Foley give worse long-term outcomes cf. Anderson-Hynes (a/w 3-5% re-operation rate)
- No difference in outcome for different drainage techniques. Nephrostomy alone not recommended.
- Repeat renogram 6-12 months

Alternatives to pyeloplasty:
- Endopyelotomy
- PCN
  - originally performed to identify improved renal function. Largely abandoned 2’ minimal improvement in function and more difficult surgery
- Nephrectomy
  - 10-15% function (DMSA advised)
- Ureterocalicostomy
  - May be useful for redo-operations
  - ? horseshoe kidneys

Algorithm for prenatally diagnosed hydronephrosis
Algorithm for incidentally diagnosed hydronephrosis in older children

**Obstructed megaureter**
Should be differentiated from refluxing megaureter, and non-refluxing, non-obstructed megaureter
Usually associated with a stenotic segment of distal ureter (Analogous to Hirshprung’s disease)
Incidence based on prenatal USS 1:1500 – 1:2000
Accounts for 10% of prenatally detected hydronephrosis
Boys > girls
Left kidney > right
Usually sporadic; occasionally heritable in AD/variable penetrance fashion
Typically picked up prenatally; occasionally with pain, UTI, mass, deteriorating renal function.

**Imaging**
USS first line
Ureteric dilatation < 1cm usually a/w normal function
MCUG
Next step after USS to exclude reflux
MAG:3
Next step once reflux excluded
Important to look at hard copies vs. curves as an unobstructed unit may simply represent the renal pelvis emptying into a dilated ureter: regions of interest have not been too successful in eradicating this

**Management**
(i) Conservative
Similar to PUJO
Provided renal function >= 40% on affected side observation only
UTI does not necessarily mandate surgery but prophylaxis should be initiated
Follow-up for minimum of 5 yrs prior to considering discharge

(ii) Surgical intervention
Indicated for deteriorating renal function, pain or mass
Surgery avoided in first year of life as re-implantation technically very difficult with poorer outcome, especially neurological damage – therefore temporise with JJ stent [cannot be inserted endoscopically in infants; requires open cystotomy, ureteric dilatation and i/o JJ stent]
Surgery for children > 1 yr
Ureteric diameter <= 1cm
   Excision of stenotic segment and Cohen re-implant
Ureteric diameter > 1cm
   Excision of stenotic segment, Starr plication and Leadbetter-Politano re-implant with psoas hitch
   Bilateral disease = unilateral operation as above
   and TUU (cannot perform bilateral psoas hitches)
Follow up at 6-12 months with USS & renogram