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 $^{Cr\text{-}}$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

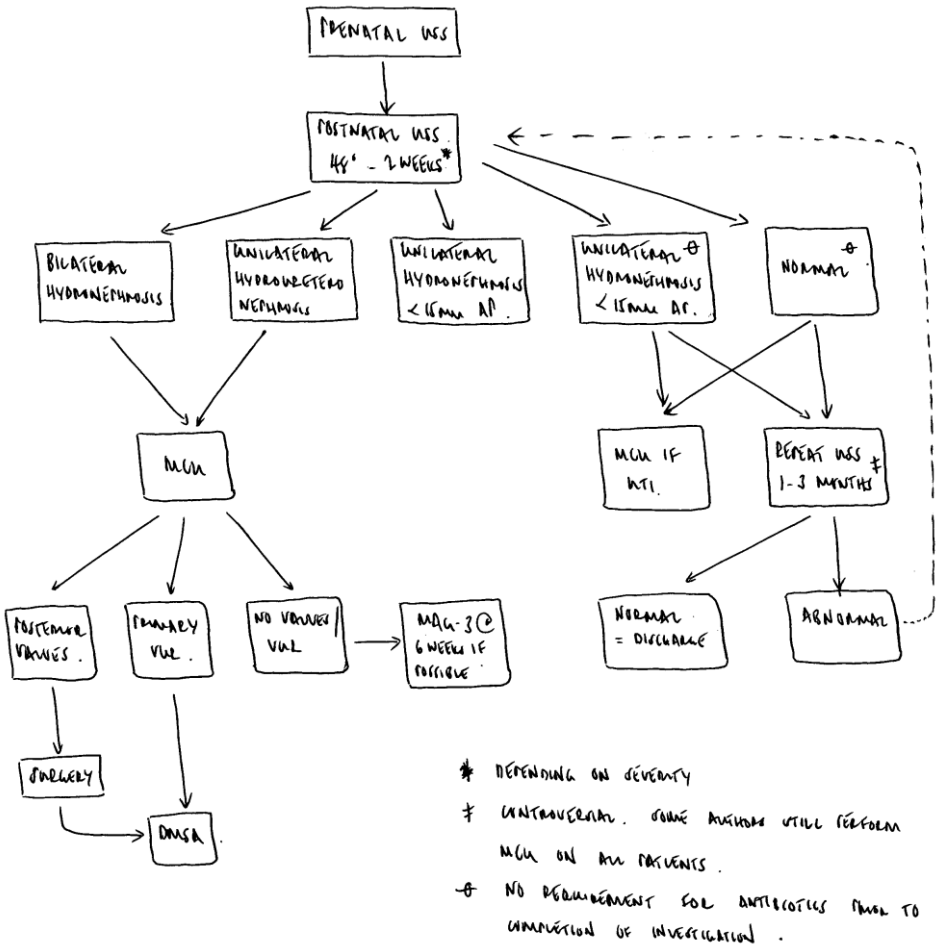
Indications for pyeloplasty

Symptoms (pain/UTI)
Split funcion < 40% on the affected side
AP diameter > 50mm
Decline in function of > 10% of affected side

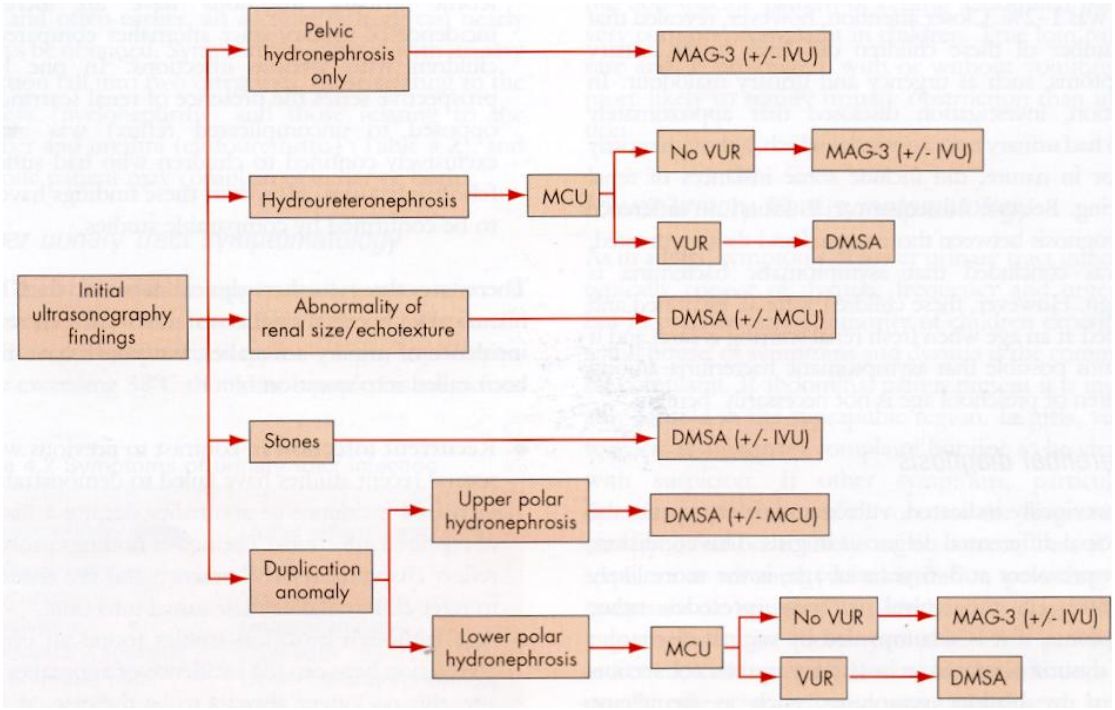
Operative considerations
Culp and Foley give worse long-term outcomes cf. Anderson-Hynes (a/w 3-5% re-operation rate)
No difference in outcome for different drainage techniques. Nephrostomy alone not recommended.
Repeat renogram 6-12 months

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



<u>Obstructed megaureter</u>

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 prohylaxis 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 diasease = unilateral operation as above and TUU (cannot perform beliateral psoas hitches)

Follow up at 6-12 months with USS & renogram