Urinary Tract Abnormalities

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Pictures from “The 18 to 23 weeks scan”
ISUOG Educational series

Embryology:

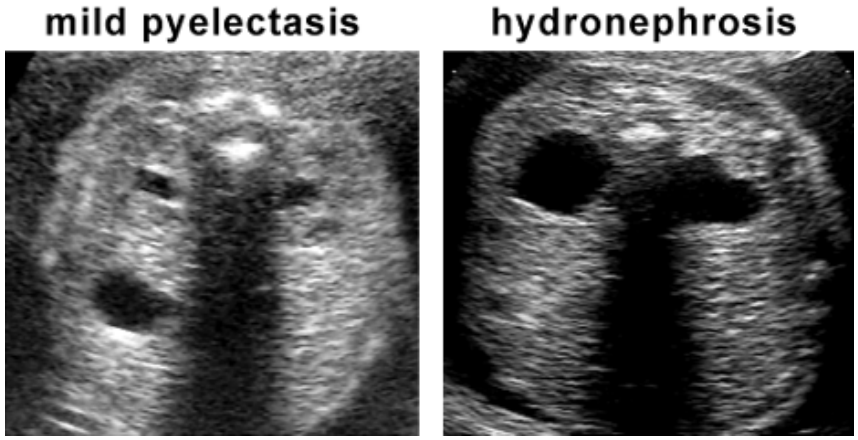
- Intermediate mesoderma: Pronephros, Mesonephros and finally Metanephros
- Mesonephros:
  - Longitudinal swelling minimal urine production 6-10 weeks
  - Mesonephric duct connecting cloaca to kidney
- Metanephros
  - Mesonephric buds
  - Connection of ureteral bud with metanephric blastema induces nephron formation
  - Functional by 10 weeks

Antenatally detected hydronephrosis

- 0,5% out of 12 000 antenatal scans revealed fetal malformations
- 0,25% had genitourinary tract abnormalities

Approach to hydronephrosis:

- Wide differential diagnosis



Differential diagnosis

- UPJ obstruction
- VUR
- Primary nonrefluxing megaureter
- Ureterocele
- Uterovesical junction obstruction
- Ectopic ureter
- Posterior urethral valves
- Megacystitis megaureter
- Physiological dilatation
- Multicystic dysplastic kidney
- Autosomal recessive polycystic kidney disease
- Extrophy
- Prune belly syndrome

Approach to hydronephrosis:

- Important factors
  - Fetal well being
  - Gestational age
  - Unilateral vs bilateral
  - Amniotic fluid volume

Diagnosis

- Different diagnostic criteria:
  - Siemens and colleagues:
  - > 6mm at < 20 weeks
  - > 8mm at 20 – 30 weeks
  - > 10mm at > 30 weeks

  - Stocks and co-workers:
  - > 4mm before 22 weeks
  - > 7mm after 33 weeks

Grading hydronephrosis

- Grade 1: APD 1cm and no caliectasis
- Grade 2: APD 1-1,5cm and no caliectasis
- Grade 3:>1,5cm and slight caliectasis
- Grade 4:> 1,5cm and moderate caliectasis
- Grade 5:> 1,5cm, severe caliectasis and cortical atrophy less than 2mm

Prognostic tests:

- Glick and co-workers:
  - Normal hypotonic urine
  - Normal to moderately decreased amniotic fluid
  - Normal to echogenic appearance of the kidney

Fetal intervention for hydronephrosis

- Controversial
- 1st was in 1980
  - Open hysterotomy and urinary diversion
- Indication:
  - Oligohydramnios and bladder outlet obstruction
  - Normal kariotype
  - Singleton

Fetal intervention for hydronephrosis

- Types of interventions:
  - Vesico-amniotic shunts
  - Fetal cystoscopy and endoscopic valve ablation

Post natal evaluation:

- Day 1: Cases with oligohydramnios, urethral obstruction, multicystic renal dyplasia, bilateral moderate-to-severe hydronephrosis or uncertainty of diagnosis

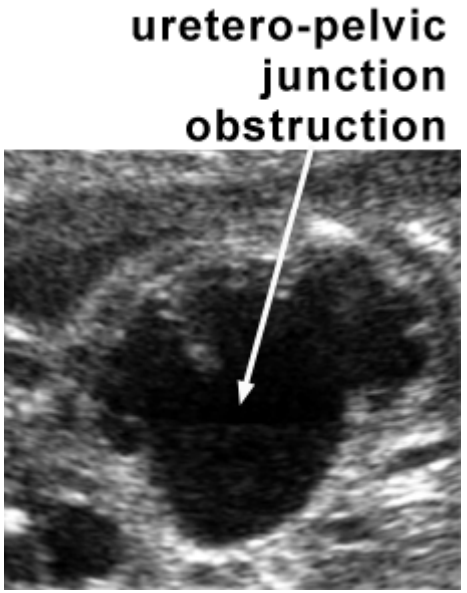
- Days 7-10: For mild or unilateral hydronephrosis

Post natal evaluation

- Voiding cystourethrography
  - Not indicated if normal sonogram post natal
  - Value if still post natal hydronephrosis

Uteropelvic junction obstruction:

- 44-65% the cause of hydronephrosis
- 90% unilateral
- Cause:
  - Intrinsic narrowing
  - at the junction
  - Extrinsic pressure from
  - accessory lower pole artery



Uteropelvic junction obstruction:

- Sonographic features:
  - Dilated renal pelvis
  - Caliectasis
  - Enlargement of the kidney
  - Distension ends abruptly
- 25% Contra lateral renal abnormalities:
  - Renal agenesis
  - Renal cystic dysplasia
  - Vesicoureteric reflux
- 10% extrarenal abnormalities

Uteropelvic junction obstruction:

- Amniotic fluid
  - Normal
  - 30% polyhydramnios, impaired renal functions

Uteropelvic junction obstruction:

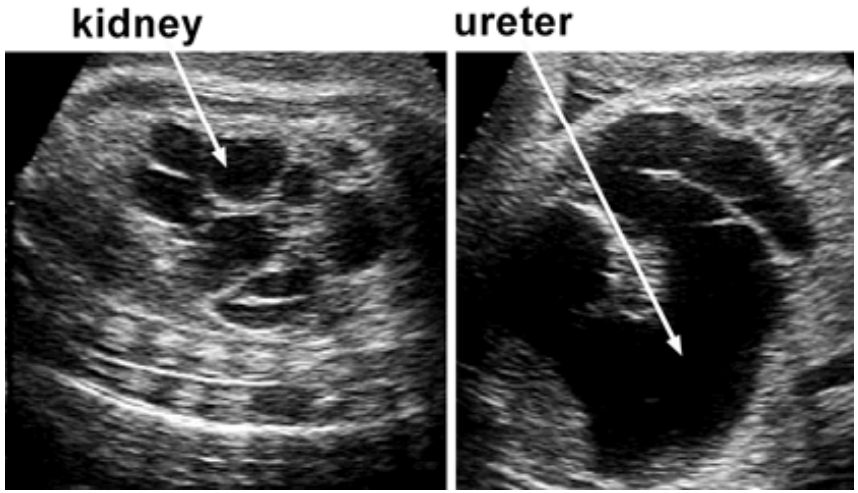
- Follow up 4 – 6 weeks
  - Evaluate for obstructive cystic dysplasia

Uteropelvic junction obstruction:

- Management:
  - Controversial between operative and observation
  - Ulman and colleagues evaluated 104 cases
  - 22% underwent pyeloplasty and all had improvement
  - 69% of non operatively managed patients resolved and 31% improved renal function

Vesicoureteral reflux

- 10-20% of hydronephrosis
- Variable degree of hydronephrosis
- No specific prenatal sonar findings



Vesicoureteral reflux

- Mostly in males

- Management
  - Observation with antibiotic cover
  - Endoscopic treatment
  - Ureteroneocystostomy

Primary nonrefluxing megaureter

- Cause
  - Aperistaltic segment of the distal ureter causing abnormal propulsion of the urine
- Ultrasound
  - Dilated ureter and renal pelvis
  - Variable atrophy of the renal parenchyma

Primary nonrefluxing megaureter

- Management
  - Surgery
  - Follow-up if differential function between 35 – 40%.
- Resolution rate depend on the grade of initial presentation with 12 months for grade 1 and 48 months for grade 5.

Primary nonrefluxing megaureter

- Indications for surgery:
  - With grade 4 or 5 hydronephrosis
  - A retrovesical ureter diameter > 1cm

Ureterocele

- Cystic dilatation of distal ureter
- Associated with renal duplication
- Classified based on position:
  - Ectopic: Extending trough the bladder neck
  - Intravesical: Remaining in the bladder

Ureterocele

- Incidence 1:9000 live births
- Gynaecological malformations in 50% of females
- Contra lateral duplication in 20%

Ureterocele

- Prenatal diagnosis:
  - Hydroureteronephrosis
  - A cystic structure in the bladder
  - Oligohydramnios
  - Obstructive cystic dyplasia of the upper pole
- If hydronephrosis always search for signs of duplication

Ureterocele

- Management
  - Antenatal decompression only when bladder outlet obstruction or oligohydramnios
  - Endoscopic decompression
  - Ureteral re-implantation surgery
  - Heminephroureterectomy

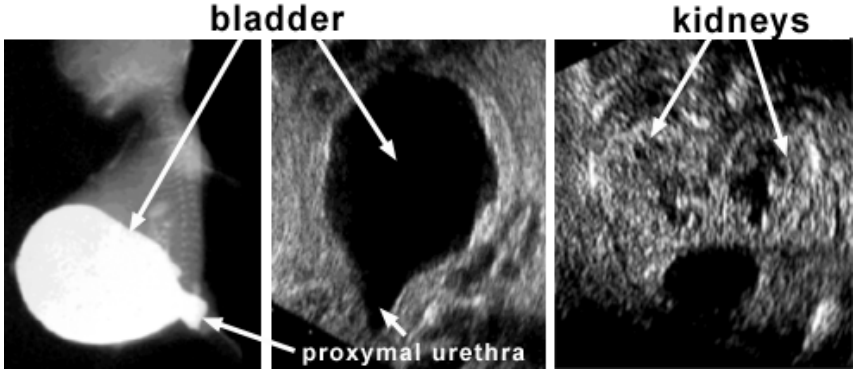
Posterior urethral valves

- Incidence: 1 in 5000 to 1 in 8000

- 3 types of valves
  - Type 1 leaflets extending distally to the level of the urogenital diaphragm
  - Type 2 extend to the level internal sphincter or bladder neck
  - Type 3 Diaphragm with central perforation

Posterior urethral valves

- Sonographic findings:
  - Keyhole sign



Posterior urethral valves

- Sonographic findings:
  - Keyhole sign
  - Ureterectasis
  - Caliectasis
  - Hydronephrosis
  - Renal dysplasia
  - Cortical cysts
  - Bladder distension
  - Thick-walled bladder

Posterior urethral valves

- Sonographic findings:
  - Renal cortical cysts 100% predictive of renal dyplasia
  - Oligohydramnios 80% fatality rate
- 43% associated malformations
  - Cardiac
  - VACTERL

Posterior urethral valves

- 43% associated malformations
  - Cardiac
  - VACTERL
    - Vertebral defects
    - Anal atresia
    - Cardiac anomalies
    - Trageoesophageal fistula
    - Esophageal atresia
    - Renal abnormalities
    - Limb abnormalities

Posterior urethral valves

- Poor prognostic signs:
  - Echogenic kidneys
  - Worsening hydronephrosis
  - Oligohydramnios
  - First detection in the second trimester

Posterior urethral valves

- Prognosis:
  - Overall mortality 25-50%
  - >90% with olygohydramnios
  - Renal insufficiency develop in 45% of survivors

Posterior urethral valves

- Management:
  - Kariotyping
  - Perform serial bladder drainage every 3-4 days
  - Use sample of 3rd drainage
  - Isotonic urine indicate poor function

Posterior urethral valves

- Good prognostic biochemical markers:
  - Na < 100meq/L
  - Cl < 90meq/L
  - Osmolarity <210mOsm/L
  - B2 microglobulin < 4mg/L
  - Ca < 8mg/dl
- Indication for vesico amniotic shunts

Prune Belly Syndrome

- Features:
  - Dramatic dilatation of the collecting system
  - Deficiency of the abdominal musculature
  - Cryptorchidism

Prune Belly Syndrome

- Sonographic Features:
  - Large thin walled bladder
  - Bilateral hydroureter
  - Bilateral hydronephrosis
  - Entire ureter dilated

Prune Belly Syndrome

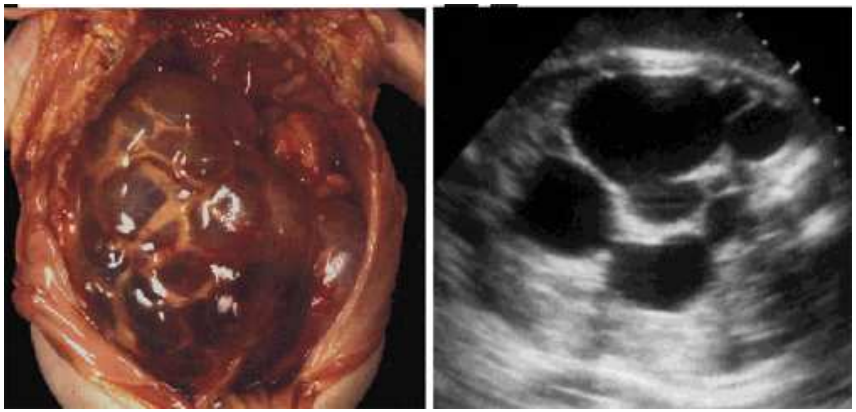
- Outcome
  - Depends on olygohydramnios
  - Renal failure a common problem

Prune Belly Syndrome

- Management
  - Follow up during pregnancy
  - Vesico amniotic shunting
- Neonatal management
  - May require renal transplant

Multicystic dysplastic kidney

- Sonographic findings:
  - Multiple variable sized non-communicating cysts
  - No central large cysts
  - Minimal to no renal parenchyma
  - Kidney enlarged
  - Unilateral in 80% of cases



Multicystic dysplastic kidney

- Common associations
  - Meckel-Gruber syndrome
    - Encephalocele
    - Postaxial polydactyly
    - Renal cystic dysplasia
  - Trisomy 13
  - Trisomy 18

Multicystic dysplastic kidney

- Gender issues
  - M:F 2:1
  - Female fetus worse prognosis
  - 2x more likely to have bilateral disease
  - 4x more likely to have aneuploidy

Multicystic dysplastic kidney

- Outcome
  - Unilateral has a good prognosis
  - Involution over time
    - 50% over 5 years
  - Bilateral disease is fatal
  - If contra lateral renal disease the diagnosis of that kidney will predict the prognosis

Multicystic dysplastic kidney

- Management
  - Termination if bilateral
  - Neonatal work up
  - Surgical excision reserved for
    - Recurrent infection
    - Hypertension
    - Wilms tumor

Conclusion

- Evaluate fetus carefully for other structural abnormalities
- Use colour Doppler to distinguish bladder from other cysts
- Evaluate the amniotic fluid volume to predict prognosis
- Consider if shunting is an option
- Careful neonatal evaluation