PAEDIATRIC RENAL & UROLOGY

URINARY SYSTEM EMBRYOLOGY

- Urogenital system derived from **intermediate mesoderm** → forms a urogenital ridge on either side of the developing aorta.
- Kidneys develop through 3 successive sets of tubular nephric structures:

(1) **Pronephros → (2) Mesonephros → (3) Metanephros (gonads)**

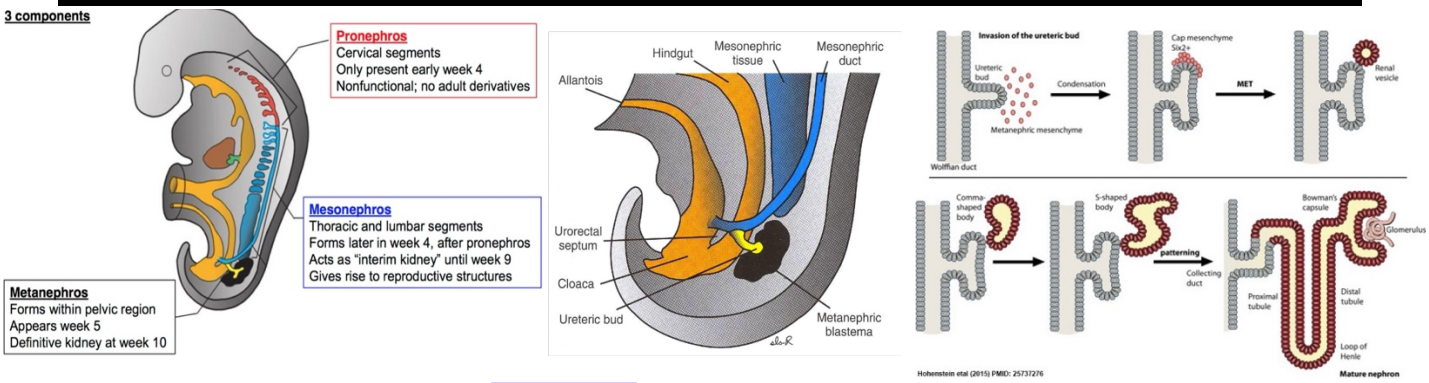
Urine production in utero

Metanephros– **primordia of permanent kidneys** begin to produce urine early in week 9 → mixed with the amniotic fluid → continues throughout foetal life +

- A mature foetus swallows several hundred millilitres of amniotic fluid each day → then **absorbed** by intestine.
- Waste products eliminated by placental circulation into maternal blood for **elimination** by the kidneys.

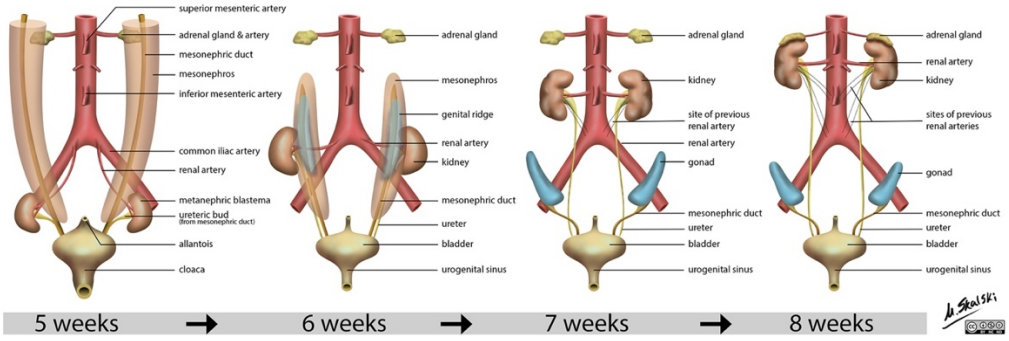
The metanephros develops from an outgrowth of the:

Outgrowth	Forms the	Function	Adult derivatives of
Condensation of intermediate mesoderm	**Metanephric blastema**	Secretes growth factors that induce growth of the ureteric bud	• Podocytes and bowman's capsule • Proximal convoluted tubules • Loop of Henle • Distal convoluted tubule
Caudal mesonephric duct	**Ureteric bud**	**Ureteric bud** responds and secretes growth factors to induces growth and differentiation of **metanephric blastema**	• Collecting tubules and ducts • Minor and major calyces • Ureters



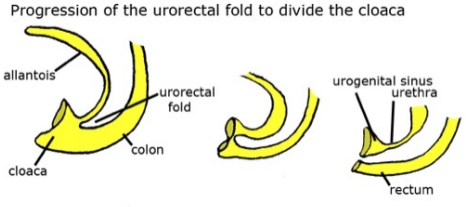
Positional changes of kidneys [ascending → inferior → superior inc. renal arteries]

- During ascent of the embryonic kidneys (from pelvis → upper posterior abdominal wall
- New blood vessels arise from the aorta and supply the kidney, while the vessels at the lower level disappear.

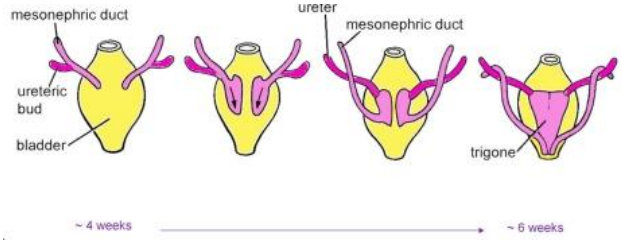


Development of the urinary bladder and urethra

- **Cloaca** = terminal hindgut lined with endoderm + receives the ***allantois*** (finger-like diverticulum) on its ventral side.
- **urorectal septum** (mesenchyme) divides cloaca into:
  - Ventral (urogenital sinus)
  - Dorsal parts (rectum and anal canal)



- *Bladder (lined by **mesoderm**)* ← **vesical part of the urogenital sinus**,
- *BUT Trigone region* ← **caudal ends of mesonephric ducts.**
  - Due to *traction* with kidney ascent + *ureters* needing to *enter obliquely* through the base of the bladder.
- **Orifices of mesonephric ducts** move close together and enter prostatic part of the urethra to become the **ejaculatory ducts**.

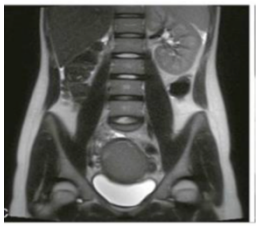


Types of embryological renal anomalies

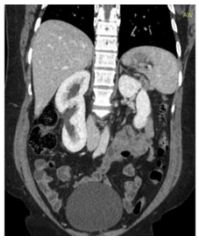
Anomalies of number

- **Unilateral renal agenesis** (absence of one kidney)
- **Supernumerary right kidney (***separate or partially fused extra kidney***)**

Unilateral renal agenesis

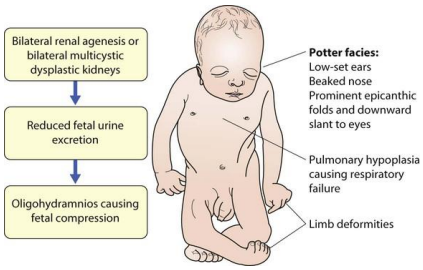


Supernumerary right kidney



What embryological urinary tract abnormality will cause oligohydramnios during pregnancy?

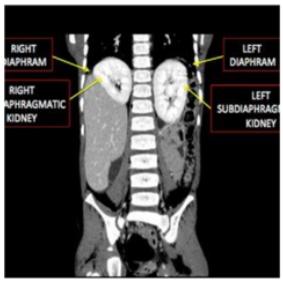
- If foetus kidneys are not producing urine (e.g. bilateral renal agenesis)
- **decrease in amount** of the amniotic fluid in pregnancy (*oligohydramnios*) since urine produced is usually mixed with amniotic fluid
- No fluid to cushion umbilical cord from uterine compression
- baby cannot survive



Anomalies of ascent

- Renal ectopia is a congenital renal anomaly characterized by the **abnormal location of one or both kidney**s
- e.g. diaphragmatic kidney, pelvic kidney, cephalad renal ectopia, thoracic kidney

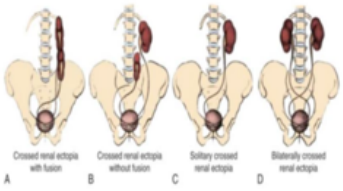
PELVIC KIDNEY

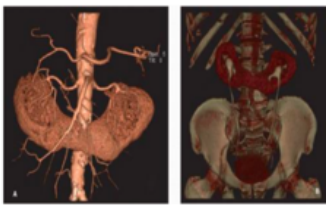


DIAPHRAGMATIC KIDNEY

Anomalies of form and fusion

- Crossed renal ectopia with or without fusion
- **Horseshoe kidney** (arrested by IMA)





Anomalies of rotation

- **Malrotation**

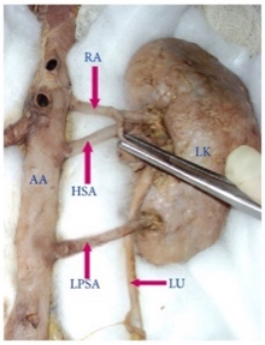
Normal

Malrotation

Anomalies of renal vasculature

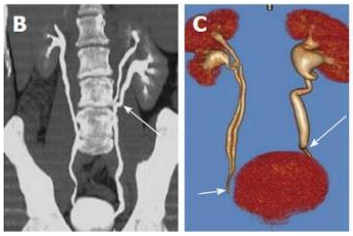
Accessory renal vessels

- Failure of lower vessels to degenerate during embryonic kidney ascending = persistent accessory renal arteries (**end arteries**)
- Consequently, if damaged or ligated the part of the kidney supplied by accessory artery is likely to become ischemic ➔ may need transplant



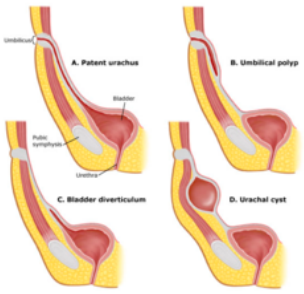
Anomalies of collecting system

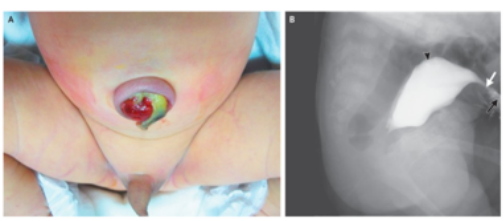
- **Duplicated collecting system**



Patent Urachus

- Opening between the bladder and the umbilicus → closes before birth
- An open ***urachus*** typically occurs in infants ➔ can lead to cysts or umbilical polyps





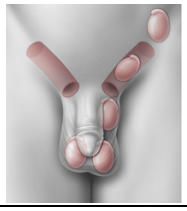
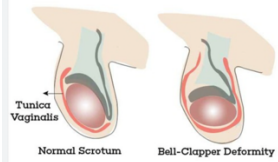
PAEDIATRIC RENAL & UROLOGY

	UTI	Vulvovaginitis	Nephrotic	Nephritis
PP	➢ Urethritis ➢ Cystitis ➢ Pyelonephritis	Inflammation and irritation of vagina and vulva	When basement membrane in glomerulus becomes permeable for protein leakage	Inflammation within nephrons of kidneys causing: ➢ Reduced kidney function ➢ Gross Haematuria ➢ Proteinuria
RF	**Females (8%) > males (2%)** • Posterior urethral valve • vesico-ureteric reflux – higher risk of Upper UTI • Poor hygiene • Uncircumcised males (urine in prepuce) • Hx of constipation • IDC	• Girls 3-10yo • Wet nappies • Tight clothing • Poor toilet hygiene • Constipation • Chemicals and soaps cleansing area	**Minimal change disease** (ages 1-10) ➢ no HTN ➢ normal C3/C4 ➢ normal EUC *Intrinsic causes of minimal change* ➢ FSGS ➢ membranoproliferative *Systemic cause of minimal change* ➢ *HSP* ➢ Diabetes ➢ Infection (HIV, hepatitis, malaria) *Other causes of nephrotic* ➢ **Membranous nephropathy** – Ab-Ag-complex on membrane causes protein leakage → **spikes on silver stain**	**Post-strep GN** (ages 5-12 yo) ➢ 1-3 weeks after GAS pharyngitis/ tonsillitis or ➢ 3-6 wks after skin infection (impetigo) ***IgA nephropathy (Berger's disease)*** ➢ Recurrent macroscopic haematuria ➢ Post-viral GITI or URTI (days after) ➢ Progressively crescenteric GN *OTHER CAUSES:* • *Deaf + FHx* → Alport • *Autoimmune or BBV* → SLE – MPGN, • *Young + rash, arthritis, bleeding* → vasculitis • *XS exercise + recent URTI* → BFH
Sx	**In babies:** • Fever • Lethargy • Poor feeding • Frequency **In children** • Dysuria • Suprapubic pain • N/V • Incontinence • Frequency	• Vulva soreness • Itchy • Redness around labia • Vaginal d/c • Dysuria • Constipation	*Pathogenesis* and signs: 1. **Inflammation** = damaged foot processes and glomeruli 2. **Proteinuria** – frothy urine (>3-3.5g/day) 3. **Hypoalbuminemia (Serum)** → peripheral oedema, ascites, pleural effusion (SOB), leuconychia 4. **Hypoimmunoglobulinemia** → infection risk and hypercoagulable state (due to loss of anticoagulants esp. antithrombin III) ➢ **Hyperlipidemia** → stroke, VTE, xanthalasma	*Pathogenesis* and signs: ➢ **Inflammation** = damaged capillary wall ➢ *Mild Proteinuria – frothy urine* ➢ **↓GFR and ↑RAAS activation →** ➢ Na and water retention → ↑ plasma volume and vasoconstriction ➢ Oliguria + azotemia ➢ **↑RAAS activation → Volume overload** → **Acute HTN + oedema** ➢ **Macroscopic haematuria** (coca-cola coloured urine)
Comp.	➢ Pyelonephritis (> 38 deg + loin-pain tenderness) **Recurrent UTI definition**: ➢ **2+ UTI w/ pyelonephritis, upper UTI** ➢ **1+ UTI w/ pyelonephritis and 1+ cystitis/lower UTI** ➢ **3+ UTI w/ cystitis/lower UTI**	UTI, thrush	➢ **Hypovolaemia** (fluid shift into interstitial space – oedema) ➢ **VTE** (XS lipids and low albumin not binding pro-thromb proteins) ➢ **Infections** (due to excretion of Ig) ➢ **Relapse** ➢ AKI or CKD	CKD – Kidney scarring ➢ Immune complexes created by strep antigens, antibodies and complement get stuck in glomerulus causing inflammation
Ix	➢ **FBC, EUC, LFT, CRP** ➢ **Catheter urine or clean catch** (non-toilet trained) ➢ **Mid-stream Urine** (toilet trained) ➢ **Urine dipstick** → Urine M/C/S o *Nitrites, leucocytes (pyuria)* ➢ **KUB USS** (if vesicoureteral reflux)	➢ **Urine dipstick +ve leucocytes** (BUT no nitrites)	➢ **Vitals (HTN)** ➢ **FBC, EUC, LFT (albumin < 25g/L), lipids (VLDL, LDL)** ➢ **Urine dipstick –** proteinuria ➢ **Urinalysis *ACR > 200*** ➢ **ASOT, anti-DNaseB, C3/C4** ➢ **Renal USS + biopsy** (no abnormality for minimal change) ➢ **EM** (podocyte effacement)	➢ **FBC, EUC, LFT (albumin), lipids** ➢ **Urine dipstick + analysis** (*RBC casts) + ACR* ➢ **POST-STREP → ASOT + Low complement** ➢ **Throat + skin swabs** (GAS) ➢ **ANCA** (vasculitis) ➢ **Anti-GBM** (goodpasture) ➢ **Renal USS** + biopsy
Mx	*Prophylaxis/ prevention* ➢ **Oral Empirical Abx** (Keflex, Bactrim, Augmentin). Esp. o 3-7 days for cystitis o 7-10 days for pyelonephritis o IV Gentamicin + ampicillin (if atypical UTI and not tolerating oral intake and unwell) ➢ **Targeted ABx** (after M/C/S results) ➢ **circumcision** esp. if have PUV or VUR/VUJ. ***If child <3yo + fever*** ➢ Empirical IV ABx ➢ Septic screen *If recurrent UTI* ➢ surgery = VUR/VUJ ➢ intermittent IDC → bladder dysfunction. ➢ **USS scan** within 6 weeks ➢ **DMSA scan (post 4-6/12 the UTI)** assess damage from recurrent or atypical UTI (damaged areas do NOT take up contrast) *When to discharge (if not → admit child)* 1. typical UTI 2. tolerating oral intakes & relatively well 3. FU with GP in 48 hrs or ED if deteriorating **If vesico-ureteric reflux** ➢ AVOID constipation ➢ Avoid XS full bladder ➢ Prophylactic ABx ➢ Urology input	➢ Avoid soap and chemical washes ➢ Good toilet hygiene ➢ Keep groin area dry ➢ Loose clothing ➢ Rx constipation or threadworms present ➢ Emollients (e.g. sudacrem can help)	*When to biopsy?* 1. < 1yo or >12 yo 2. Gross hematuria 3. Low C3, persistent HTN 4. Unresponsive to steroids ***Treatment for minimal change*** 1. **Lifestyle:** *1) low salt diet* *2) fluid restrictions* *3) regular urinalysis* 2. ***Oedema*** → *diuretics (if severe)* 3. ***4 wks of High-dose* steroids THEN weaned over next 8 weeks (to prevent relapses and tolerance)** (*prednisolone 1mg/kg – for remission and reduce relapse*) → curative intent) [80% will be fully cured] 4. ***Albumin IV*** *-if severe hypoAlb* *If unresponsive to steroid Rx:* ➢ *ACEi* ➢ *Immunosuppressants (cyclosporin, tacrolimus, rituximab)*	***Acute? → DR ABCDE*** ➢ Salt and water restrict ➢ Loop diuretics (1st line: furosemide) ➢ anti-HTN **Post-strep GN:** Mainly supportive - 80% make full recovery ➢ penicillin ➢ diuretics - for oedema ➢ anti-HTN – for HTN **IgA nephropathy:** ➢ *Supportive treatment* ➢ *1st line* =corticosteroids to slow progression of disease ➢ alternative immunosuppressive o

PAEDIATRIC RENAL & UROLOGY: Structural Issues

	STRUCTURAL ISSUES			HUS	Nocturnal Enuresis
	ARPKD	**Wilm's tumour**	**Posterior Urethral valve**		
PP	➢ Children more commonly have **AUTOSOMAL RECESSIVE PKD** unlike adults	Nephroblastoma	Proximal tissue in urethra (closest to bladder) causes obstruction and **hydronephrosis**	***Most common kidney*** ➢ IgA vasculitis ➢ Thrombosis within the small BVs	**Incontinence during sleep in children > 5 yo** 1) Poor sleep 2) Small bladder capacity 3) Nocturnal polyuria **Types** 1) **Primary Enuresis** = *incontinence since birth* → **nocturnal detrusor (overactive bladder),** *nighttime fluid intake, failure to wake (underdeveloped bladder signals), psychological distress at home/school* 2) **Secondary** enuresis = wetting after period of dryness > 6/12 (**e.g. UTI, T1DM, psychosocial issues, maltreatment, constipation, CP**) 3) **Diurnal enuresis (daytime incontinence) e.g. urge or stress incontinence recurrent UTI, constipation, psychosocial issues**
RF	• Neonates • Mutated PKHD1 gene on Chr 6 → improper development of tubules and healthy epithelial tissue in kidneys, liver and pancreas	• < 5 year olds • WAGR syndrome (deleted WT1 gene)	➢ Newborn boys	➢ Viral URTI ➢ Shiga toxin producing E. coli or POST-DIARRHOEA or post-pneumococcal	
Sx	**Ante-natal period:** • Oligohydramnios • Pulmonary hypoplasia • Potter syndrome • Congenital liver fibrosis • Cystic enlargement of renal collecting ducts	**Prominent unilateral abdo mass** ➢ **Aniridia (complete absence of iris)** May present with • Abdo pain • Haematuria • Lethargy • Fever • HTN • UWL	➢ **Palpable enlarged abdo mass** May present with ➢ Oliguria ➢ Abdo pain ➢ Altered mental state ➢ Chronic urinary retention ➢ Difficulty urinating ➢ Weak stream ➢ **Recurrent UTI**	*Symptoms* ➢ Abdo pain ➢ Lethargy / irritable *Signs* ➢ Reduced urine output ➢ Haematuria or dark brown urine ➢ HTN ➢ Oedema ➢ Bruising ➢ Confusion	
Comp.	**Liver fibrosis** ➢ Liver failure → cirrhosis ➢ Portal HTN → variceal bleeding **Renal dysfunction** ➢ ESKD ➢ HTN **Pulmonary hypoplasia** ➢ Chronic lung disease	Death	**Ante-natal** ➢ Bilateral or unilateral hydronephrosis ➢ Oligohydramnios + pulmonary hypoplasia **Post-natal** ➢ Recurrent UTI ➢ Respiratory failure (2nd to pulm. Hypoplasia)	➢ ISS ➢ Testicular swelling ➢ ttp	**Red flags:** ➢ **SC issue (cauda equina –** sphincter dysfn w/ lower limb weakness ➢ **Day and night time polyuria** (e.g. DI, renal abnormality) ➢ **Recurrent UTI (**renal abnormality, dysuria, poor stream**)** ➢ **PMHx (**diabetes, OSA, ADHD**)** ➢ **FHx** and **psychological stresses**
Ix	➢ **Ante-natal scans** ➢ **Genetic testing** **Genetic dysmorphic features of potter's:** ➢ Low set ears ➢ Flat nasal bridge ➢ Abnormal skeleton ➢ Underdeveloped ear cartilage	➢ USS KUB - visualise mass ➢ CT/MRI – stage tumour ➢ **Biopsy and histology** for definitive diagnosis	➢ **KUB USS -** hydronephrosis or enlarged thickened bladder ➢ **Micturating cystourethrogram (MCUG) -**visualise urine reflux ➢ **Cystoscopy** – ablate or remove extra tissue	➢ **FBC** 1) -Microangiopathic Anaemia (haemolysis) 2) Thrombocytopenia ➢ **EUC** = Ureamia ➢ **Urine dipstick** = Haematuria + proteinuria ➢ **Blood film** = schistocytes (haemolysis)	**Establish underlying cause** ➢ **2 week diary –** toileting, fluid intake and bedwetting episodes ➢ **Ht, wt, BP (***growth + HTN)* ➢ **Lower limb exam (**cauda equina) ➢ **Tufts of hair on back (**spina bifida**)** ➢ **FBC, EUC,** ➢ **UA, Urine M/C/S → ?UTI**
Mx	**Poor prognosis** ➢ 1/3rd die in neonate period ➢ 1/3rd survive into adulthood **Most need:** ➢ Specialist peadiatrician care ➢ Renal dialysis ➢ Steroids to promote long maturation during ante-natal period	**Prognosis** ➢ **Good** = early stage (90% survival) ➢ **Poor** = **m**ets present **Surgical excision (curative intent)** ➢ Adjuvant RT OR ➢ Adjuvant chemo	**If asymptomatic:** ➢ Conservative approach (watch and wait) **Symptomatic**: ➢ Temporary IDC ➢ Surgical ablation using cystoscopy (definitive Mx)	MEDICAL EMERGENCY (10% MORTALITY RATE) ➢ Refer to paediatrician and renal specialist → possible referral for renal dialysis ➢ Usu. self-limiting and supportive Mx best ➢ Anti-HTN – as required ➢ Careful maintenance of fluid balance 70-80% make full recovery	**Lifestyle:** ➢ *Bladder retraining* (alarm training – but requires compliance and adherence) ➢ *Minimise* fluid intake before bed ➢ *Avoid* blame and shame ➢ *Minimise* constipation (to avoid reducing bladder capacity) ➢ *Enuresis alarms?* **Medical:** ➢ **Minirim (desmopressin)** = reduce nocturia **→ best for short-term** *(e.g. upcoming camp or school trip)* ➢ **Anti-cholinergic for overactive bladder in urge incontinence** (*e.g. oxybutynin, tolteridone)* ➢ **TCA (e.g. imipramine) → 2nd line ? relax bladder to lighten sleep**

PAEDIATRIC RENAL & UROLOGY: Male children and their issues

	Undescended testes (cryptorchidism)	TESTICULAR TORSION	Varicocele (L > R)	Hydrocele
PP	Failure for testes to migrate down inguinal canal into scrotum ➢ Congenital ➢ Usu. found in inguinal canal (80%) ➢ **DDx: retractile testicles (normal)** – when child exposed to cold – cremasteric reflex activated	➢ **Intravaginal "bell-clapper"** = testes not fixed to tunica vaginalis "testes + cord twists" ➢ **Extravaginal** = spermatic cord not fixed within inguinal canal "vaginalis + cord twists"	Dilated PPV draining testes due to distal obstruction ➢ Large tumour compression L renal vein ➢ Renal vein thrombosis ➢ Accelerated growth	Collection of fluid within **tunica vaginalis which surrounds the testes** ➢ Simple = fluid trapped in tunica vaginalis ➢ Communicating = tunica vaginalis connected to peritoneal cavity via processus vaginalis
RF	➢ FHx- inheritance ➢ LBW ➢ SGA ➢ Pre-term ➢ Maternal smoking (pregnancy)	➢ Young male ➢ **bell-clapper deformity -** *abnormally high fixation between tunica vaginalis and spermatic cord*		➢ Newborn males (esp. simple hydrocele)
Sx	➢ **Asymptomatic**	➢ **Sudden onset Severe suprapubic groin pain (***usu. no preceding trauma* **)** ➢ **Nausea and vomiting**	➢ **BAG OF Worm appearance and feeling** ➢ **Non-tender** ➢ **Standing or valsalvre**	➢ **Irreducible soft smooth non-tender swelling** ➢ **Trans-illuminable**
Comp.	➢ **subfertility,** ➢ **torsion** ➢ **testicular cancer** (if not descended by 6/12)	**Swollen testes and impaired gait** ➢ **-ve prehn's sign** ➢ **-ve cremasteric** ➢ **high-riding** testes (red/blue) ➢ **Blue dot sign** (upper scrotal edge) = testicular appendage torsion	➢ **subfertility**	DDx: of scrotal mass ➢ Testicular torsion ➢ Hydrocele ➢ Haematoma ➢ Inguinal hernia ➢ Crypto-orchidism ➢ Testicular tumour (Rare)
Ix	Newborn exam	Clinical ➢ Diagnostic USS – reduced testicular blood flow		Scrotal USS – confirm Dx and exc. other causes
Mx	➢ Conservative = Usu. resolve by 1 y ➢ Orchidopexy (if undescended after 6/12)	➢ Urgent surgical consult (risk of infarction of 8-12 hours) ➢ NBM, clear fluids + analgesia	Supportive care ➢ Surgery – risk of bowel perforation ➢ Radiological emboilsation	• **Simple hydrocele** = 90% spontaneously resolve by 3 yo • **Communicating hydrocele** – surgical ligation required
		 *Testes and cord twist for intravaginal torsion		

	Hypospadias	Balanitis	Paraphimosis	Inguinal hernia
PP	Congenital penis abnormality where urethral meatus is in abnormal position **ventral surface towards scrotum** ➢ Glandular (90%) ➢ Medial ➢ Penoscrotal	➢ Inflammation of glans of penis	➢ XS unretracted foreskin causes distal oedema → stricture of external urethral meatus	***DIRECT*** – weakness of AAW allows bowel to protrude into inguinal canal ➢ ***INDIRECT*** – follows course of spermatic cord
RF	➢ Congenital	➢ Trauma, infection ➢ Irritation (soap residue) ➢ Poor hygiene	➢	➢
Sx	➢ **Incomplete foreskin on ventral surface** ➢ **Urine leakage** *DDx:* ➢ *Epispadias (on dorsal side)*	➢ **Red painful glans** ➢ **Possible rash** ➢ **+/- penile discharge**	➢ Poor flow ➢ Suprapubic pain (full bladder) ➢ Will see **constricted penile ring**	➢ **Reducible mass** ➢ **Irreducible (if strangulated)**
Comp.	➢ **Curvature (chordee)**	➢ Paraphimiosis	➢	➢ obstructed hernia → bowel ischaemia → bowel necrosis ➢ incarcerated hernia
Ix	Newborn check			
Mx	*Paediatric specialist urologist referral* • if mild = watch and wait • **(avoid circumcision before surgery)** • Surgical ED for > 3/12 old – correct meatus position and straighten penis *Complications of surgery* ➢ Urination difficulty ➢ Sexual dysfn ➢ Aesthetically unappealing and psych concerns	• Warm salt water wash • 1% hydrocortisone topical • Antifungal topical if candida?	• Emergency -→ manually reduced w/ analgesia OR surgery	***Semi-urgent surgery*** ➢ within days for newborn ➢ within wks for infants ➢ within mths for children