Hypospadias

Common

1:200 boys

Very rare but occasionally seen in girls

? incidence increasing (possibly foetal exposure to oestrogens)

Poorly characterised genetic predisposition

- More common in monozygotic twins and in offspring of fathers with hypospadias (6-8%)
- Risk of affected second child:
  - 1 in 8 if no family history
  - 1 in 5 if one other male relative affected
  - 1 in 4 if two other male relatives affected

Characterised by location of external urethral meatus*

Glanular	}	80-85%
Subcoronal	}	
Penile		10-15%
Penoscrotal	}	5-10%
Perineal	}	

* technically the level of hypospadias can only be defined after correction of chordee, although for pragmatic purposes the above classification is widely used

Features (3)

- Abnormal ventral opening of urethra
- Ventral chordee
- Hooded foreskin (deficient ventrally)

Associated abnormalities (4)

- Hernia
- Patent processus vaginalis/hydrocoele
- Persistent prostatic utricle
- Undescended testis
  - Palpable unilateral/bilateral UDT and hypospadias = 15% chance finding underlying DSD
  - Impalpable unilateral/bilateral UDT and hypospadias = 50% chance of underlying DSD
- NB. Incidence of upper tract abnormalities is identical to normal population (2%). No requirement for upper tract imaging

Management

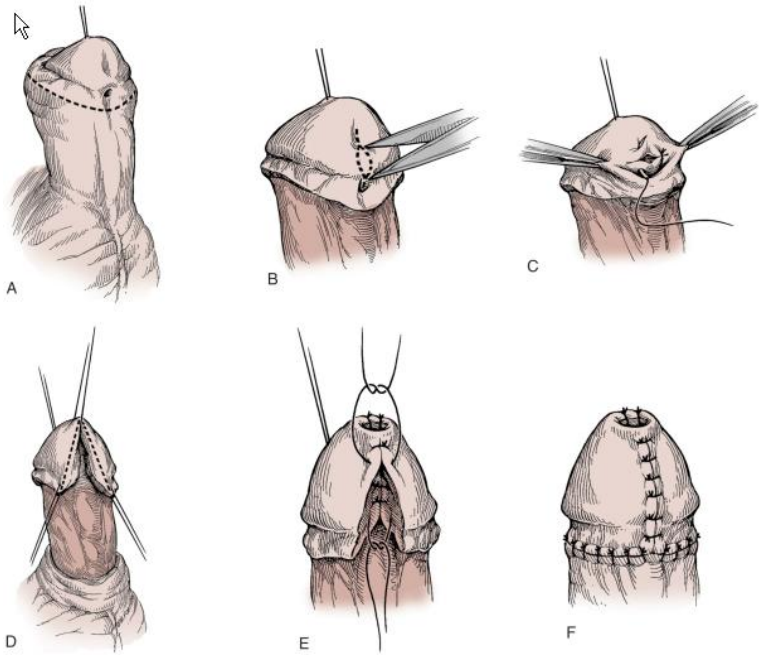
- Adult patients who void and inseminate without difficulty require no specific treatment. There is however an appreciable psychological impact associated with hypospadias and therefore it is recommended that children are undergo surgery. American Society of Paediatrics recommends surgery between 6-12 months, certainly before 2 years.
- A. <u>General surgical principles</u>
  - Careful tissue handling
  - Tension-free anastomosis
  - Use of well vascularised tissues
  - Meticulous haemostasis
  - Fine suture material (6/0 or 7/0 absorbable)
  - Non-overlapping suture lines
  - Adequate urinary diversion

B. Specific surgical principles

(i) Correction of chordee depends on underlying problem
tethering of ventral penile skin – deglove penis
atretic c. spongiosum – excision of atretic segments
tethering of urethral plate to c. spongiosum - release
fibrous ventral c.cavernosum - plication

(ii) Urethroplasty

(a) MAGPI (Duckett, Arap 1984)
Distal only
Meatal advancement and glanuloplasty
Heineke-Mikulicz type advancement of meatus with lateral glanular incisions and wraparound
Re-operation rates < 5%
MAGPI – see below



(b) Tubularisation (Duplay, Snodgrass, Hayes)
Use of urethral plate to form tube
Wide plate = Duplay
Narrow plate = Snodgrass (vertical relaxing incision aka tubularised incised plate (TIP)) – **most commonly performed procedure worldwide**
Hayes (v. incision and onlay graft)
Vascularised, skin-less pedicle on top
Glanuloplasty and skin coverage
Duplay ~ 7-10% re-operation rate; approx 5% for Snodgrass. NB. Dartos flap for waterproofing has dramatically reduced fistula rate.

(c) Pedicled flaps
Meatal flap (Matthieu)
Prepucial flap (Bracka)

(d) Two-stage free flaps
Incision and dorsal onlay of windowed graft

Graft may be buccal mucosa or post-auricular skin
Tubularisation after 6 months.
Windowed grafts allow imbibition and inosculation

(iii) Adequate skin coverage
Glanuloplasty
Rotation of excess doral/prepucial skin to ventral side

C. Complications

Early
Bleeding
Infection
Rarely dehiscence

Late
Fistula
Commonest late complication
From cath removal to many years post-op
Initial Mx – catheter replacement
Persistent – exclude stricture or diverticulum, then refer to specialist centre for re-do surgery (up to 50% recurrence after re-do). Two-stage repair favoured for complex re-do surgery
Meatal stenosis
Urethral stricture
Urethral diverticulum
Persistent chordee
Hairy urethra
Disordered psychological adjustment

Congenital megaprepuce

Massively enlarged foreskin
Probably acquired ? secondary to phimosis
Important to identify prior to circumcision, as may lead to buried penis
Initial Rx = dorsal slit or prepucioplasty and evacuation of retained urine/inspissated secretions, followed by referral for plastic surgical correction

Micropenis

Normal neonatal stretched penile length = 3-5cm
Definition of micropenis = stretched penile length < 2cm
Commonest cause is hypogonadotrophic hypogonadism [Kallman's syndrome, Prader-Willi, Lawrence-Moon-Biedel, CHARGE syndrome]
Associated with midline brain defects
Rx is testosterone replacement (penis will still enlarge at pubity with T replacement)

Phimosis

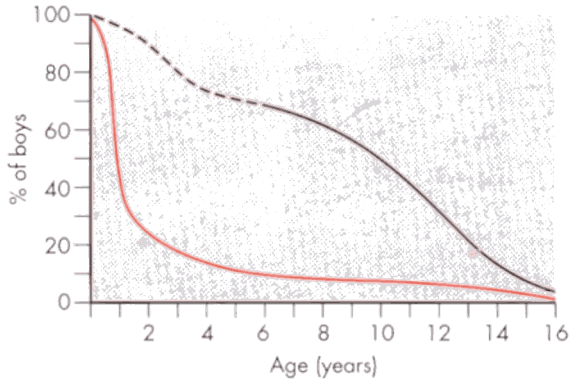
Male circumcision commonest surgical procedure in world

Incidence varies around world due to religious and cultural differences

UK	3.3% falling
Scandinavia	2% static
Australia	13% falling
US	60% rising

Prepuce develops 8-16 weeks gestation, contiguous with glans – childhood phimosis therefore physiological

At birth < 5% foreskins retractile; ~5% non-retractile by 16 yrs (Oster 1968, see below; red = non-retractile foreskin, black = prepucial adhesions)



Prepucial adhesions separate by proximal desquamation, possibly due to infection of retained smegma.

Separation of prepucial adhesions may be accelerated by topical steroid therapy. Largest study to date in 1185 boys showed resolution rate of over 90% in boys receiving 8 weeks of potent topical steroid fluticasone proprionate (Cutivate) 0.05% (Zavras 2009) without any reported complications. Similar results reported for Betnovate (0.025% -0.1%). AUA recommends application tds for 6 weeks.

Current indications for circumcision

BXO phimosis	0.6% incidence
Recurrent severe balanoposthitis	~1% incidence
Recurrent febrile UTI and abnormal UT	rare

BXO phimosis rare before the age of 5 yrs; a/w meatal involvement in 20% - anterior urethral involvement very rare in children

Recurrent balanoposthitis presents with erythema +/- discharge. Incidence approximately 3%; in one-third recurrent.

Alternatives – counselling, topical high-potency steroids, prepucioplasty

Complications:

Oozing		36%
Bleeding req. re-operation		1.5%
Infection		8%
Discomfort > 1 week		26%
Meatal stenosis	}	
Glans amputation	}	rare
Buried penis	}	

Routine neonatal circumcision (Jews, Muslims, traditional Catholics, Aborigines (subincision))

'Arguments' for routine circumcision (very limited evidence)

Reduced infection rate
Reduced penile cancer (HPV)
Reduced partner cervical cancer (HPV)
Reduced HIV infection

(i) Cx reduces infection rate
Multiple studies (Wiswell, Schoen, To) have shown that UTI frequency in young males is higher in uncircumcised males vs. circumcised males (OR = 0.13; ~8x lower). May be that keratinised squamous epithelium prevents p-fimbriated bacterial adherence. However incidence of phimosis ~1% and there is an appreciable complication rate. Thus (from Singh-Grewal meta-analysis 2005):

normal urinary tract	NNT =111
recurrent febrile UTI	NNT = 11
high-grade VUR and UTI	NNT = 4

only boys at high risk of UTI should therefore be offered Cx.
(ii Cx and penile cancer
Most penile carcinoma associated with phimosis. Furthermore HPV infection rates 70% lower in Cx males than in unCx (Castellsague 2002) However incidence of penile carcinoma very rare and although conceivable, there is no evidence that routine circumcision influences development of penile carcinoma (or cervical carcinoma)
(iii) Cx for HIV
No evidence that routine neonatal circumcision reduces the risk of contraction of HIV. Some have suggested that foreskin with its high concentration of Langerhan's cells easily traumatised and removal may reduce risk of transmission.ANRS trial (Auvert 2005) performed in South African males showed 60% protective effect for Cx even after adjustment for sexual and health seeking behaviour. Trial controversial however (paid inducements, bias etc.)

Undescended Testis
Common
Right > left
Bilateral in 25%
Affects 4% of term boys and 22% of premature boys; 1% of boys at one year
Spontaneous descent among premature boys common (especially infants > 2kg birthweight); In normal term infants may be expected in first 3 months of life, associated with neonatal LH surge (4% reduced to ~1%). Very few testes spontaneously descend thereafter (0.8% at 12 months).
Bilateral UDT a/w:
Neural tube defects
Prune-belly syndrome
Androgen insensitivity
Karyotype abnormalities
Denys-Drash Syndrome
UDT develop abnormal histological findings after ~24 months
Leydig cells hypoplasia (#1 finding)

Defective transformation of gonocytes to spermatogonia (and spermatocytes/spermatids)
Reduced total numbers of germ cells

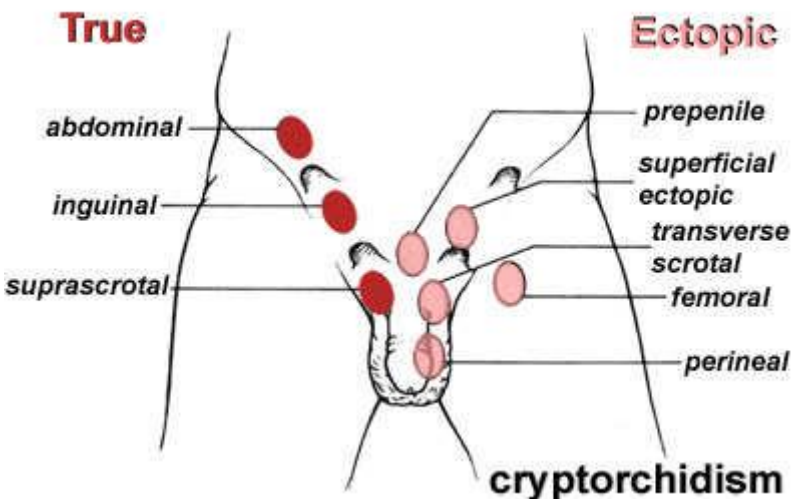
Although impact upon rates of fertility and malignancy controversial, recommended that orchidopexy performed before 2 yrs of age. Probably due to temperature – difference between core temperature and scrotum = 2 degrees

UDT always a/w PPV; ectopic testis usually not

Cremasteric reflex absent in first 3 months of life

Investigation

If UDT diagnosed at birth – re-examine at 3 months of age
Crucial to distinguish between UDT and ectopic testis, and retractile or ascending testis
Ectopic testes may be superficial inguinal pouch (aka Dennis-Browne pouch), prepenile, transverse scrotal, femoral or perineal (commonest).
Retractile testes can be brought to the floor of scrotum where they stay without tension: high retractile testes are always under tesion and should be treated as UDT.
Ascending testis is a definite phenomenon; aetiology unknown, spontaneous re-descent in 80% but orchidopexy recommended.
Examination considerations
Warm room
Inspection – scrotal hypoplasia
Palpation – soap test



Management of palpable testis (80%)

No evidence for the use of BHCG in treating congenitally UDT – may have a role in improving likelihood of success in difficult cases

Orchidopexy

1.8% of boys undergo orchidopexy by 2 yrs of age
Slightly higher incidence cf. incidence of UDT (1%) – presumably due to surgery on retractile testis
Inguinal UDT
Inguinal incision
Separate cord structures from PPV
Divide all cremasteric fibres
Dartos pouch (absorbable suture)

- High/intra-abdominal UDT
  - (i) Jones pre-peritoneal approach (Steve Jones – Australia)
    - High transverse incision
    - Retroperitoneal mobilisation
    - Divsion of inferior epigastrics
    - Medial placement
  - (ii) Fowler-Stephens procedure
    - Ligation of testicular vessels and preservation on artery to vas
    - Typically performed as two-stage procedure to allow collateral supply to develop
    - First stage usually laparoscopic; second stage (6 months) open or lap-assisted
  - (iii) Microvascular orchidopexy
    - ? still performed
    - Transection and anastomosis to inferior epigastrics
  - (iv) Lap-assisted
    - Usually mobilisation of testicular vessels at time of diagnostic lap for impalpable testis. If testis can be obilised with enough length to reach contralateral deep inguinal ring, then will reach scrotum.
  - Complications
    - Failure to bring testis down
      - > EIR 8%
      - Canalicular 13%
      - Intra-abdominal 26%
    - Testis atrophy 5%
    - Injury to vas 1%
  - Outcomes
    - Fertility
      - Overall
        - Unilateral UDT 80-90% paternity
        - Bilateral UDT 50-60% paternity
      - Timing of orchidopexy
        - < 2 years 90% fertile
        - 3-4 years 60% fertile
        - 9-12 years 30% fertile
    - Malignancy
      - UDT a/w increased risk of testis cancer (RR~4)
        - Normal population 1:500
        - Unilateral UDT 1:125
        - Bilateral UDT 1:45
      - CIS found in ~25% of adults with retained intra-abdominal testes and 1.7% of non-abdominal testes
      - Commonest tumour seminoma
      - Evidence regarding the timing of orchidopexy controversial and currently undefined
        - Chilvers 1994 = no increased risk provided orchidopexy performed before age of 10

(from UK Testicular Cancer Study Group). Similar results from Danish study (Moller 1996). However:
Preber 1996 and Swerdlow 1997 = increased risk irrespective of timing
Recent excellent study using Scandinavian database suggests and overall increased risk, which is exacerbated by late orchidopexy (Pettersson NEJM 2007)

< 13 years RR 2.2 vs. gen. popn.
>= 13 yrs RR 5.4 vs. gen. popn.

Management of palpable ectopic testis

Orchidopexy if possible, orchidectomy if not

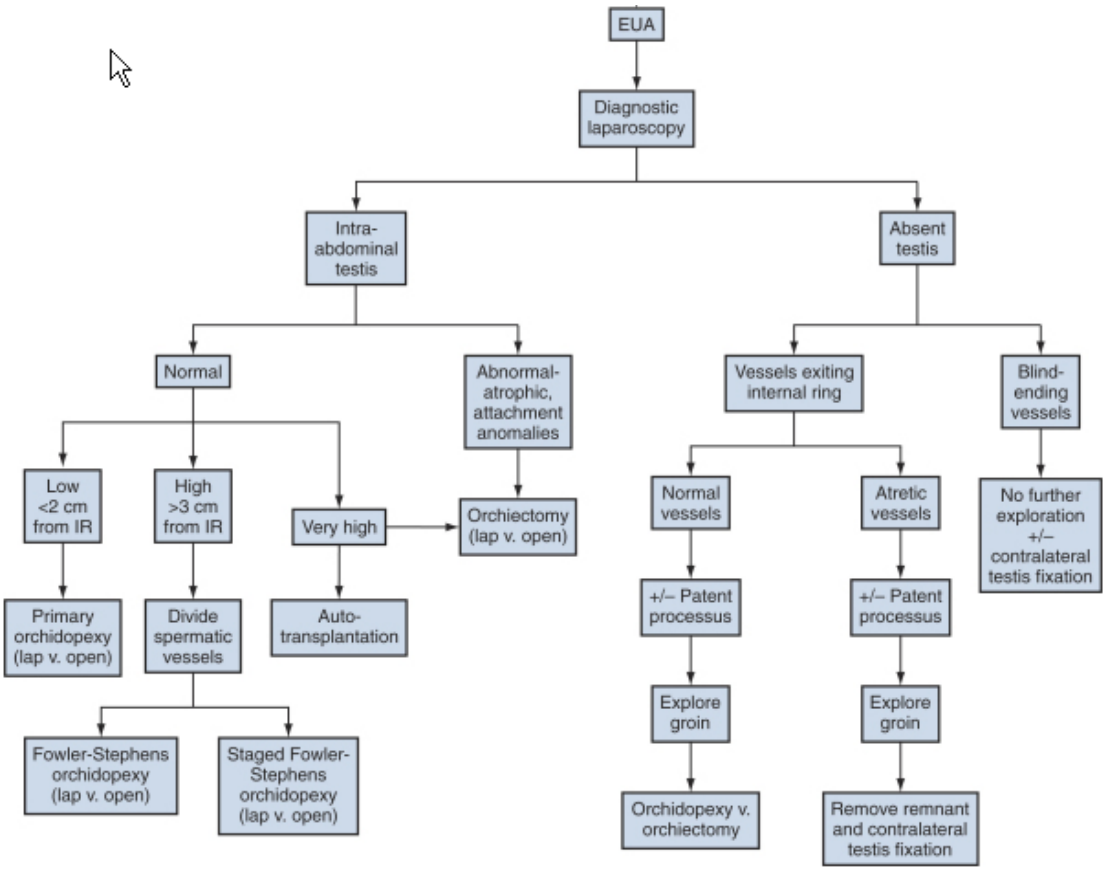
<u>Management of impalpable testis</u>

20% UDT impalpable of which:

40% intra-abdominal testis (usually just above inguinal ring)
30% absent intra-abdominal (blind-ending vessels)
20% absent intracananalicular (nubbin)
10% inguinal testis

Thus ~ 50% have salvageable testis

Assuming negative outpatient examination by experienced practitioner, EUA +/- laparoscopy investigation of choice. USS of inguinal canal may be useful in settings without padiatric surgery or paediatric urology facilities
Campbell's algorithm for impalpable testis below:



IR = internal ring
Fixation of contralateral testis has been recommended due to higher incidence of bell-clapper abnormality in contralateral testis. However fixation a/w risk of testis atrophy.
Initial laparoscopic approach has been criticised by some. In 30% of cases vessels exit via IR and inguinal exploration required, thereby avoiding laparoscopy. As intra-intrabdominal testis likely to require an inguinal incision at some stage, laparoscopy only avoids inguinal incision in 30%. However also allows management of intra-abdominal testis and prevents re-operation through groin incision when staged procedure performed.

The acute scrotum

Causes of acute scrotum

- Torsion of testis*
- Torsion of appendix testis
- Torsion of appendix epididymis
- Epididymo-orchitis
- Idiopathic scrotal oedema
- Strangulated inguinal hernia
- Henoch-schonlein purpura

* Torsion accounts for ~90% of causes of acute scrotum in adolescents. Second to torted appendages in younger children

Testicular Torsion

Common
Incidence 1:4000
Accounts for 25%-33% of all cases of acute scrotum
Viable testis can only be salvaged if blood supply restored < 4-6 hours after onset. Even if <4 hours, 50% have abnormal sperm parameters (Bartsch 1980 - ? anti-sperm antibodies)
Neonatal or intrauterine torsion extravaginal; all other ages intravaginal due to high insertion of tunical vaginalis (bell-clapper deformity)
UDT = 10x RR, but history of UDT in only 5% of cases
Diagnosis

- Bottom-line = No reliable diagnostic test
- (i) History
  - Commonly unreliable, especially in younger boys
- (ii) Examination
  - High-riding swollen testis suggestive
  - Prehn's sign unreliable
  - **Absent cremasteric reflex highly sensitive** (100% of 145 boys over 7 yr period (Rabinowitz 1984), but also associated with other causes of acute scrotum (poor specificity). Normal cremasteric reflex suggests diagnosis unlikely, although there are a number of case reports of torsion in the presence of a normal cremasteric reflex.
- (iii) Doppler USS

Demonstration of normal blood supply to the testis reassuring but not infallible and highly operator dependent. False negative rates of 11-25% remain unacceptable. High-resolution US (detects twist in spermatic cord) reportedly highly sensitive (96%) and specific (99%)(Kalfa 2007), but further training necessary and uptake low

(iv) Radionucleotide scan

Largely historical. Invasive and limited availability with PPV of only 75%

Management

Manual detorsion – largely unfeasable

Surgical detorsion – midline raphe, detorsion, dartos pouch with non-absorbable sutures, fixation of other side

Outcome

Semenalysis usually impaired, including 50% of those repaired within 4 hours. Few studies relating Hx of torsion with susequent paternity

Neonatal torsion

Usually referred from post-natal ward with swollen, indurated scrotum. No prospect of salvage and no additional risk to the other side – therefore conservative management and serial USS (ensure involution and not congenital testis tumour). However controversial. Some advocate surgical intervention for the following reasons:

1. Makes diagnosis – exclude intravaginal torsion
2. Limits symptoms
3. There have been case reports of bilateral metachronous extravaginal torsion which is catastrophic

Torsion of appendage

Peak age 10-12; blue dot sign in only 20%. If history sufficiently suggestive, USS for flow and conserve Mx. Otherwise Sx

Epididymo-orchitis

Relatively uncommon – wastebasket diagnosis however
Classically due to retrograde reflux of urine into epidiymidis, often with underlying anatomical anomaly or voiding dysfunction
Cultures often negative – when diagnostic GNB most common
Cremasteric reflex usually present.
Analgaesia and Abx for obvious cases; surgery if doubt re. Torsion

Idiopathic scrotal oedema

Peak incidence 5-6 yrs
Usually painless
Diagnosis clinically or USS to exclude hydrocoele
? lymphangitis (often extends towards anus)
Spontaneous resolution – no treatment required

Hydrocoele

Patent PPV very common – 2-5% of boys
Painless swelling, often fluctuates in size, typically worse at end of day

90% resolve by one year, vast majority by 2 years.
Surgery indicated if persists > 2 years or de-novo cases in later childhood
Surgery – identify and isolate PPV in inguinal canal; exclude co-existent hernia; ligate and divide at IR; aspirate and incised distal portion
Recurrence ~1%

Varicocoele

See notes on adult varicocoele
6% 10 yr olds; 15% 13 yr olds
Hudson classification applies to children

Subclinical	Doppler USS only
Grade 1	palpable on valsalva
Grade 2	palpable but not visible
Grade 3	visible

Important to document testicular voulume in children. Prader orchidometer unreliable – more valuable to assess size using USS
Varicocoele repair generally indicated in children for a) symptoms; or b) catch up growth of affected testis. Surgery should be considered if > 20% difference in volume between the 2 testes on ultrasound scanning.
Similar operative options and outcomes for children as in adults