**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. **General surgical principles**

- 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/prevpucial 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
Hairly urethra
Disordered psychological adjustment

Congenital megaprepucex
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 prepucplasty 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 puberty 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 propionate (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, prepuciplasty

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

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):

<table>
<thead>
<tr>
<th>Condition</th>
<th>NNT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal urinary tract</td>
<td>111</td>
</tr>
<tr>
<td>Recurrent febrile UTI</td>
<td>11</td>
</tr>
<tr>
<td>High-grade VUR and UTI</td>
<td>4</td>
</tr>
</tbody>
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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 tension 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
   Division 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
   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

Management of impalpable testis
20% UDT impalpable of which:
40% intra-abdominal testis (usually just above inguinal ring)
30% absent intra-abdominal (blind-ending vessels)
20% absent intracanalicular (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 pediatric surgery or pediatric 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%

Varicocele
See notes on adult varicocele
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 volume in children. Prader orchidometer unreliable – more valuable to assess size using USS
Varicocele 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