Vulval disease in children

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I have no conflict of interests
<table>
<thead>
<tr>
<th>Inflammatory dermatoses</th>
<th>Infections</th>
<th>Drug reactions</th>
<th>Blisters and ulcers</th>
<th>Naevi</th>
</tr>
</thead>
<tbody>
<tr>
<td>Psoriasis</td>
<td><em>S. Pyogenes</em></td>
<td>Allergic contact dermatitis</td>
<td>NSAGU</td>
<td>Haemangioma of infancy</td>
</tr>
<tr>
<td></td>
<td>Vulvovaginitis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dermatitis</td>
<td>HPV</td>
<td>Fixed Drug Eruption</td>
<td>Bullous pemphigoid</td>
<td>Epidermal naevi</td>
</tr>
<tr>
<td>Lichen Sclerosus</td>
<td>Molluscum contagiosum</td>
<td></td>
<td>Erythema multiforme</td>
<td>Pigmented naevi</td>
</tr>
<tr>
<td>Contact Dermatitis</td>
<td><em>S. aureus</em></td>
<td></td>
<td></td>
<td>Other hamartoma</td>
</tr>
<tr>
<td>Labial fusion</td>
<td>Tinea</td>
<td></td>
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</tbody>
</table>
Common parental experiences

- Child abuse fears
- My child is abnormal
- Issues at school
- Embarassment
- Frustration with diagnoses such as thrush, worms, UTI
- Fear regarding use of topical corticosteroids
Genital Dermatitis

- Common cause of genital itch and irritation
- Signs can be subtle
- Atopic children
- Exposed to irritants
Dermatitis Treatment

- 1% hydrocortisone ointment
- Emollient
- Rule out and treat superinfection: usually *S. aureus*
- Reassurance and explanation
Irritants and practical advice for parents

› Night nappies
› Swimming lessons
› Bubble bath and shampoo
› Ballet lessons
› Cycling and horse-riding
› Fecal incontinence and dysfunctional voiding
› Stop using antifungals and wet wipes (MI)

› It is VERY unusual to have a hygiene problem
Vulval Psoriasis in children

› Less common than dermatitis generally
› More common as a cause of genital rashes
### Causes of Non-Specific Vulvitis
(Fischer G Australas J Dermatol 2010; 51:118)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Pre-pubertal n=38</th>
<th>Post-menarchal n=68</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dermatitis</td>
<td>9 (24%)</td>
<td>14 (21%)</td>
</tr>
<tr>
<td>Psoriasis</td>
<td>27 (71%)</td>
<td>8 (12%)</td>
</tr>
<tr>
<td>Chronic vulvovaginal candidiasis</td>
<td>0</td>
<td>42 (62%) p&lt;0.001</td>
</tr>
</tbody>
</table>
Genital psoriasis: Treatment

- Same environmental modification as for dermatitis
- Stronger topical corticosteroids
- Steroid sparer: emollient, tar ointment
Lichen Sclerosus

- Mean age at onset 5-7 years
- Commonest presenting symptoms: itch and soreness
- Other symptoms at presentation
  - Dysuria
  - Constipation
- Referrals to urology and gastro-enterology common
- May be mistaken for sexual abuse
Management of LS in Girls

- No different to adult women
- Disease suppression with potent topical corticosteroid: well validated
- Little data on long-term control

- Fischer G Rogers M Treatment of vulvar lichen sclerosus with potent topical corticosteroid Pediatr Dermatol 1997; 14:235-8
Prognosis

- Vulvar lichen sclerosus does not resolve at puberty
- Early onset malignancy in adult life
- Inadequately treated childhood disease may result in scarring


Long-term management

› Single centre, retrospective study, 2014
› 46 with pre-pubertal onset VLS
› Comparision of
  - Compliant patients (72%)
  - Partially compliant patients (28%)
Why are patients non-compliant?

- Fears around topical corticosteroids
- Denial
- Forgetful
- Not enough tough love
Treatment used to achieve remission

Strength of topical corticosteroid used for initial treatment
Treatment used to maintain remission

Strength of topical corticosteroid used for long-term treatment:

- Super potent: 0%
- Moderate: 20%
- Moderate + Mild: 40%
- Mild: 60%
## Results

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Adherent</th>
<th>Non-adherent</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time to initial remission</td>
<td>4.2 months</td>
<td>5.2 months</td>
<td></td>
</tr>
<tr>
<td>Complete control on maintenance treatment</td>
<td>91%</td>
<td>8%</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Progression of disease during treatment</td>
<td>0</td>
<td>69%</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Progression of scarring</td>
<td>0</td>
<td>16%</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Side effects (erythema, irritation)</td>
<td>4 (12%)</td>
<td>2 (15%)</td>
<td>&gt;0.99</td>
</tr>
</tbody>
</table>
Implications for practice

› Maintenance treatment can prevent progression of disease
› Opportunity for early intervention
› Potentially reduce future cancer risk
› Regular preventative rather than “as needed” treatment
Streptococcal Genital Infection

› Presentation: acute, sore, red
› May be associated perianal infection
› Source: oropharynx
› May recur due to pharyngeal carrier state
› Swabs: Group A Beta-Hemolytic Strep
› May precipitate psoriasis
› Treatment: appropriate antibiotic 10 days
Does thrush happen to pre-pubertal children?

- Studies that EXCLUDE teenagers in the cohort have found that *C. albicans* is rarely found in genital isolates
- *C. albicans* clinical infection requires estrogenisation

Unless
- Recent oral antibiotics
- Diabetes
- Immunosuppression
<table>
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<tr>
<th>Pathogen</th>
<th>Pre-pubertal n=38</th>
<th>Post-menarchal n=68</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>C. albicans</em>(causal)</td>
<td>0</td>
<td>35 (50%) p=&lt;0.001</td>
</tr>
<tr>
<td><em>C. Albicans</em> (colonization)</td>
<td>2 (5%)</td>
<td>3 (4%)</td>
</tr>
<tr>
<td>Gp A β-haem strep</td>
<td>4 (10%)</td>
<td>0</td>
</tr>
<tr>
<td><em>H. Influenzae</em></td>
<td>3 (7.5%)</td>
<td>0</td>
</tr>
<tr>
<td><em>S. Aureus</em></td>
<td>1 (2.5%)</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>Gardnerella</td>
<td>0</td>
<td>1 (1.5%)</td>
</tr>
<tr>
<td>No pathogen isolated</td>
<td>28 (74%)</td>
<td>28 (41%)</td>
</tr>
</tbody>
</table>
Non-sexually acquired genital ulceration

- Benign, non-infective acute and/or recurrent ulceration
- Typical morphology of aphthous ulcer
- Involves mucosal and adjacent surfaces only
- Painful
- Recurrent
Acute NSAGU

- Early adolescent to teenage girls
- Ulceration of sudden onset
- Flu-like prodrome in most
- Ulcers often large (>10mm)
- May be obscured by intense swelling of labia
- Incapacitating
- Patients often unable to walk or urinate
All forms of NSAGU are frequently misdiagnosed:
- HSV
- Other forms of STI
- Trauma

For young girls this can present a major source of iatrogenic trauma
Etiology of NSAGU

- Acute form may be reactive, virally induced
- EBV has been implicated but not always found
- Recurrent form: many theories, essentially unknown
- Helicobacter, other micro-organisms
- Oral aphthae associated with Coeliac Disease
- Aphthous-like ulcers seen in Crohn’s and Behçet’s
› No evidence-based treatment guidelines exist
› The literature is vague on management
› There are recommendations for treating oral aphthae which can be extrapolated
Investigations should be minimal: Viral swab
Serology interesting but does not aid management
Severe: oral prednisone 0.5-1mg/kg daily
Mild: potent TCS
Analgesia
Wait….. reassure
If recurs, prophylaxis: Doxycycline 50-100mg/day
Non-invasive approach to young girls

Vaginal foreign bodies: a common problem?

› No
› Worth considering if persistent discharge
› Said to be toilet paper most often
› In practice, rare
› Treat with saline lavage
Recurrent toxin-mediated perineal erythema

- Recurrent asymptomatic erythema and desquamation of vulva and perineaum
- Follows a Streptococcal pharyngitis
- Superantigen phenomenon
- May be desquamation of hands, feet
- Associated with strawberry tongue
- DD: Kawasaki Syndrome
Staphylococcal Folliculitis

- Often also folliculitis on buttocks
- Itchy rather than sore, pustules
- *S. Aureus* is cultured
- Consider perineal staph carriage
- May superimpose on any skin condition
Not always the result of sexual transmission but this needs consideration in all cases.

Remains controversial

We don’t know outcome re later cervical infection

If maternal source: screen mother

Treatment: imiquimod, podophyllin

Observation only
Molluscum Contagiosum

› Common viral infection in children
› Spread in water
› Uncommon as isolated vulval presentation
› Not usually considered an STD in children.
› 4 genotypes: 2 are sexually acquired
› May be giant
› May be extensive in children with HIV
Tinea of the vulva in children

- Very rare
- Often incognito due to treatment
- Diagnosis: scraping
- Rx: Topical antifungal
Naevi and other lesions

- Haemangiomas
- Pigmented naevi
- Epidermal naevi
- Hamartomas
Birthmarks are commonly seen on the vulva in children.

Any birthmark that occurs on skin generally may be found on the vulva.

They are often confused with dermatoses and genital warts.

Haemangionas may be confused with sexual abuse.
Hidradenitis Suppurativa in Children

› HS has been reported in children as young as 5
› More often a family history
› Look for androgenisation
› Most common is premature adrenarche
› Treatment: spironolactone, antibiotic combinations
Vulval bullous pemphigoid
Linear IgA disease
Erythema Multiforme
Fixed Drug Eruption
Vesicular nappy rash
Allergic contact dermatitis

Erosive lichen planus is very rare in children (more likely to be BP)
Childhood vulval bullous pemphigoid

- Erosive or blistering disease
- Painful
- May be cicatricial
- May be associated with ocular disease
- Biopsy with IF diagnostic
- Responds to potent topical TCS
Vulval fixed drug eruption

- Occurs in children
- Acute erosive vulvitis
- Ibuprofen
- Paracetamol
- Sulfa antibiotics
- Often missed
- Can scar
Labial Fusion

- Seen only in young children often with an underlying vulvitis
- Not seen in adults
- Abnormal vulval appearance
- Confused with ambiguous genitalia
- Asymptomatic, but pooling of urine may result in vulvitis or urethritis
- Often recurs after treatment
- Treated with topical oestrogen cream and potent topical corticosteroids
Neuropathic vulval pain has been described

Look for scoliosis

Attention getting behavior
Key point: Genital dermatoses

- Chronic genital disease in children is uncommon relative to adults
- Almost all with genital itch and redness have psoriasis or dermatitis
- Children with genital dermatoses require TCS, not antifungals
Key Points: Lichen Sclerosus

› LS in children should be treated aggressively with potent TCS
› LS does not resolve at puberty
› LS may result in scarring if inadequately treated
› Follow up is essential to ensure adherence to long-term treatment
Key Points: Genital Infections

- The commonest genital infective condition in children is due to Gp A Streptococcus, *which is not seen in adults*.
- Children do not suffer from recurrent or chronic candidiasis.
Key Points: Genital ulceration

› Acute genital ulceration in adolescent girls is usually not infective
› Minimize investigations: HPV only
› Acute treatment with prednisone and analgesia
› Rarely recurs
› Recurrent disease: doxycycline
 › The commonest chronic genital dermatosis is psoriasis
 › Lichen sclerosus requires long-term treatment and follow up
 › Genital candidiasis does not occur in healthy pre-pubertal children
 › Non-sexually acquired acute genital ulceration is an important differential from STI and child sexual abuse