

# Vulval disease in children

## Part 1: presentation

When a prepubertal girl presents with a complaint of an itchy or sore vulval rash, a common assumption is that she has 'thrush' or a urinary tract infection. She may be considered to have poor hygiene, or even to have been sexually abused. In fact, none of these diagnoses is likely to be true.

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Vulval disease in children is less common than in adults, and although many of the vulval diseases that affect adults also affect children, there are some important differences between the two groups. In both adults and children, dermatitis, psoriasis and lichen sclerosus are the three most common causes of a chronic vulval rash. Candidiasis is another common adult vulval disease, but it is not seen in the non-oestrogenised vulva and vagina of the child. Birthmarks of the vulva area are an important issue in children but not in adults (where they have long ago resolved or been diagnosed). Fusion of the labia is a self-limiting condition sometimes seen in young children whereas it is seen in adults only in the setting of lichen sclerosus or severe lichen planus. Group A  $\beta$ -haemolytic streptococcal vulvovaginitis is a disease that affects only children, but apart from this, infective vaginitis is rare in children. Sexual abuse is always an issue to be considered in any genital presentation in children, but in fact is rarely a cause of vulval disease. Causes of vulval symptoms in children are listed in the Table.

There has been little published work on paediatric vulval disease. Most of what exists focuses on infective conditions, anatomical abnormalities and tumours. The latter two of these conditions are very rare in everyday practice and even infection is unusual.

There is often an assertion in articles on paediatric vulval disease that the skin of the prepubertal vulva is fragile and sensitive because it is poorly oestrogenised. There is, however, no evidence to back this up. It is physiological for a child's vulva to be low in oestrogen and the fact that children have much less trouble with vulval rashes than adults do does not support the assumption that the vulval skin of a child is prone to disease. The presence of oestrogen predisposes to vaginitis, particularly candidiasis. Furthermore, oestrogen creams are often very irritating when applied to children. Obviously oestrogen is not meant to be there in this age group.

Another common assertion is that vulval disease in children is due to 'poor hygiene' and 'faecal contamination'. This is a poorly evidenced

### IN SUMMARY

- Most children with an itchy vulval rash have dermatitis or psoriasis.
- Vulvovaginal candidiasis does not occur in healthy prepubertal children.
- The commonest cause of acute infective vulvovaginitis in children is group A  $\beta$ -haemolytic *Streptococcus*.
- Not all children with genital warts have acquired them sexually.
- A greenish discharge on the underwear in an asymptomatic child with negative swabs is a normal variant.
- Beware of the child who complains of persistent symptoms despite normal examination and negative bacteriology – she may be attention-seeking.

statement that trivialises the problem and stigmatises the child. In fact, mothers of small girls, particularly those with a vulval problem, are usually highly conscientious about genital hygiene and are more likely to be doing more washing than is necessary, rather than too little.

This article will consider the presentations of the various vulval diseases in children. Part 2, to be published in the next issue of *Medicine Today*, will consider their management and the issue of child sexual abuse.

### Itchy rashes of the vulva

The common causes of vulval rashes are the usual dermatoses of the prepubertal age group, i.e. dermatitis and psoriasis, and the much rarer condition lichen sclerosus, which is primarily a genital rash in children.

#### Dermatitis

Although it is a common assumption that candidiasis is the usual cause of itchy vulval rashes, dermatitis is most often the culprit in both adults and children. Furthermore, in both age groups, most patients who suffer from vulval dermatitis are atopic, and atopic individuals have the sort of skin that reacts to common environmental irritants.

The onset of vulval dermatitis is often delayed until the child is out of nappies because the well-hydrated skin under the nappy rarely shows signs of atopic dermatitis.

The skin in the nappy area of babies is remarkably resistant to disease, considering it is continuously covered by nappies. This resistance appears to be lost in the older child, where the ongoing wearing of nappies, such as overnight in those with enuresis, starts to cause problems with irritant dermatitis similar to that seen in incontinent adults.

Irritant contact dermatitis may occur after contact with faeces, such as in children with diarrhoea or chronic constipation with soiling. Children who shower

rather than bath may not wash the vulval area effectively; in my experience this is how the alleged 'poor hygiene' most often occurs. However, the most common causes of irritant contact dermatitis in children are overuse of soap and bubble bath, using shampoo in the bath and swimming in chlorinated swimming pools.

Irritation from overuse of medications and perfumed products is very common in adults but less so in children, mainly because children are not exposed to so many of these products. However, it is not uncommon for parents to treat their child's itchy vulva with antifungal creams: these are not appropriate in this age group and may cause irritation.

Vulval dermatitis presents with vulval itching and a fluctuating rash that is often precipitated by contact with irritants, and worsened by excessive washing and use of antifungal creams. The child's scratching behaviour is often embarrassing for her parents and attracts unwelcome attention at school. It is common for children with vulval itching to wake in a distressed state at night with night terrors.

Examination is often fairly unremarkable. Close inspection will reveal some erythema, scale and slight rugosity of the labia majora, and increased erythema and desquamation of the minora. The desquamation may stain the child's underwear and be misinterpreted as a vaginal discharge. If the rash is severe it may extend to the inguinal areas and buttocks (Figure 1). Superinfection with *Staphylococcus aureus* may occur on the skin, but there is no vaginitis and vaginal swabs and urine culture are invariably negative.

#### Psoriasis

The onset of psoriasis may be at any age, although it more commonly starts in adult life than in childhood. Nevertheless, if children with vulval disease are taken as a group, it is a relatively common cause, and should always be considered in the differential diagnosis of

persistent genital rashes in both sexes.

In babies, psoriasis may present for the first time as a persistent nappy rash. The features at this age include a well demarcated edge and involvement of the inguinal folds, but the typical scale of

### Table. Causes of vulval disease in children

#### Red itchy rash

Dermatitis  
Psoriasis  
Tinea  
Vaginal foreign body with persistent discharge

#### White rash

Lichen sclerosus

#### Blisters, erosions or ulcers

Impetigo  
Folliculitis  
Herpes simplex virus infection  
Herpes zoster virus infection  
Varicella virus infection  
Aphthous ulcers  
Bullous pemphigoid

#### Acute vulvovaginitis

Group A  $\beta$ -haemolytic streptococci  
Fixed drug eruption  
Erythema multiforme

#### Lesions

Molluscum contagiosum  
Human papilloma virus infection  
Pyramidal perineal protrusion

#### Normal appearing vulva

Subacute vulvitis  
Pinworm infestation  
Attention-seeking behaviour

#### Abnormal appearing vulva

Fusion of the labia  
Imperforate hymen  
Ambiguous genitalia

continued

psoriasis is lacking under the nappy. Although psoriasis may respond to standard nappy rash treatment with 1% hydrocortisone topically, it is not unusual for the rash to be resistant to this treatment.



Figure 1. Acute dermatitis with erosions.



Figure 2. Psoriasis, showing typical colour and well defined edge.



Figure 3. Lichen sclerosus. White eruption with ulceration and telangiectasia.

In older children the morphology of the rash is much the same, with itchy, red, well-demarcated, symmetrical plaques (Figure 2). Again there is no scale. The vulva, perineum and perianal areas, and often also the natal cleft, may all be involved. In older children, 1% hydrocortisone is rarely effective.

If psoriasis is confined to the vulva, it is difficult to make a definite diagnosis unless other diagnostic clues are present, such as a history of cradle cap or difficult nappy rashes as a baby, nail pitting, post auricular or scalp rashes, and a family history.

### Lichen sclerosus

Lichen sclerosus is a rare skin disease that presents as well demarcated white plaques anywhere on the skin but mainly in the genital area. It is much more common in females than males, and is mainly seen in adults but may start at any age. The tendency to develop it is probably genetic and there is an association with autoimmune disease. Although it is often asymptomatic outside the genital area, when it occurs on the vulva it tends to be intensely itchy.

Although unusual, lichen sclerosus is familiar to doctors who deal frequently with vulval disease because of its tendency to involve the vulva. If children with vulval disease are examined as a group, about 10% of them will have lichen sclerosus.

Girls with lichen sclerosus tend to present with more complex symptoms than adults, who usually complain mostly of itch. Soreness, dysuria, bleeding and chronic constipation may also occur in children. Not surprisingly, affected children are often investigated for bowel and urinary tract abnormalities.

The clinical appearance is of a well demarcated white plaque with a wrinkled surface and scattered telangiectasia that may bleed from time to time (Figure 3). The typical distribution is a figure of eight plaque surrounding the vagina and anus, but any pattern on the vulva, perineum

or perianal area may be seen. The vagina is not involved and extragenital involvement is very rare in children. Unless the labia are parted and a careful inspection made, the condition can easily be missed.

The characteristic atrophy and scarring of the vulva that occurs in adults with lichen sclerosus is rare in children in the early stages of the condition. However, in longstanding cases, particularly those that have been neglected, there can be loss of the labia minora and clitoris (both become buried under scar tissue).

A concern with this condition in children is that it may be suspected to represent child sexual abuse. This relates to the unusual and unfamiliar appearance of the rash, and the fact that there may be recurrent bleeding. It is important for the GP to realise that this is a skin condition. Its presence does not rule out abuse, but the disease itself is not a cause to suspect it.

Whether lichen sclerosus remits at puberty has been controversial. It is now accepted, however, that although this does appear to happen sometimes, a permanent remission cannot be assumed in every case. Symptoms may settle but there may be silent progression of scarring and atrophy, and symptom activity may recur in adult life. There is also a worrying association (of about 2 to 6%) with squamous cell carcinoma of the vulva in adult life. This has been reported in relatively young women who have had lichen sclerosus since childhood. This condition should, therefore, be actively managed and referral to a dermatologist is recommended.

### Birthmarks on the vulva

Birthmarks may occur on the vulva, as on any other part of the skin, but the importance of lesions in this location is that they may be mistaken for more sinister conditions. For example, pigmented naevi on the vulva often raise queries of melanoma, where they might be ignored elsewhere, and epidermal naevi, which are rare and therefore not

familiar to GPs, are often mistaken for warts or recalcitrant dermatitis.

### Haemangiomas

In infancy, capillary haemangiomas or strawberry naevi are common, being found in 10 to 20% of babies, especially girls (Figure 4). They are very prone to ulceration when located on the vulva, and sometimes may ulcerate so extensively that it is difficult to tell that there was ever a birthmark. Not surprisingly, when ulceration occurs, sexual abuse may be suspected. It is most important not to jump to such conclusions but to seek the opinion of a dermatologist or paediatrician.

Fortunately, like haemangiomas elsewhere, these lesions resolve, and significant sequelae are rare unless they were very large.

### Pigmented naevi

Pigmented naevi may occur on the vulva either as a congenital lesion or one that appears at any stage of childhood. The congenital lesions may be larger than late onset ones. It is normal for children to acquire naevi at any age on any part of the skin, and the vulva and perianal area is no exception (Figure 5). However melanoma in children is rare, and there have been very few reports of childhood vulval melanoma.

Pigmented naevi of the vulva do not have significant malignant potential. If a lesion has benign features, such as symmetry, evenness of colour, and growth stability, it can be safely observed.

### Epidermal naevi

Epidermal naevi are quite rare, and are not always present at birth. They often have a warty surface and may be arranged in whorls or streaks. A lesion that involves the vulva can be part of a larger one that extends to the leg and buttock (Figure 6). They are sometimes quite itchy and they have a tendency to enlarge with time, possibly then interfering with function

(such as wiping after toileting).

Such naevi can be mistaken for warts, which may raise suspicions of sexual abuse. If they are itchy, they can be mistaken for treatment-resistant dermatitis or nappy rash. Epidermal naevi of the genital region do not have a significant malignant potential but it is not uncommon for them to cause enough trouble to require at least partial excision. However, if they are not causing problems, it is best to leave them alone.

### Vulvovaginal infections Streptococcal vulvovaginitis

Most GPs are aware that group A  $\beta$ -haemolytic streptococci (*Streptococcus pyogenes*) can cause a low grade perianal rash (streptococcal perianal dermatitis). It is less well known, however, that the same organism can cause vulvovaginitis and balanitis, and is, in fact, the most common cause of acute infective vulvovaginitis in prepubertal children in Australia. Adults are not prone to group A streptococcal vulvovaginitis, although very occasionally a group B *Streptococcus* may cause a true vaginitis in an adult.

Clinically, streptococcal vulvovaginitis presents with sudden onset of an erythematous, swollen, painful vulva and vagina, with a thin mucoid discharge (Figure 7). There may have been a preceding throat infection with the same organism, or preceding perianal dermatitis. Sometimes the infection can be low grade (similar to the perianal disease), presenting as a subacute vulvitis.

The infection is easily diagnosed by introital and perianal swabs, and it is not necessary to insert the swab right into the vagina, which children usually find traumatic, particularly when the area is tender. Although a differential diagnosis of acute candidiasis would be reasonable in an adult, this is not appropriate in children. Also to be considered in the differential diagnosis are psoriasis, but only where the vulvitis is subacute, and a fixed drug eruption, a much rarer cause of



Figure 4. Small haemangioma.

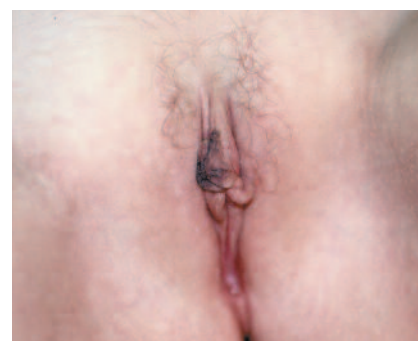


Figure 5. Pigmented naevus of the labia minora.

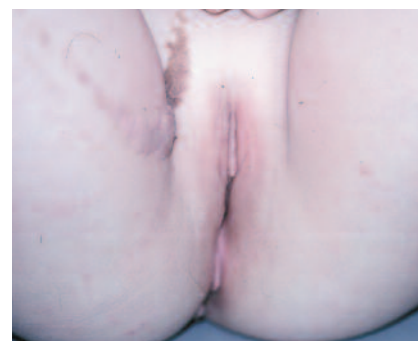


Figure 6. Systematised epidermal naevus with vulval and perianal involvement.

acute vulvitis (Figure 8). Erythema multiforme may also affect the vulva, but is usually part of a generalised reaction in children (Figure 9).

Recurrent streptococcal infections should raise the possibility of an intra-vaginal foreign body.

Any case of acute vulvitis in a child should suggest *S. pyogenes* infection, and

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swabs should be taken and antibiotic treatment started (see part 2 of this article in next month's *Medicine Today*).

### Staphylococcal folliculitis and impetigo

Staphylococcal folliculitis is common on the buttocks of children, particularly those with dermatitis and those who are still in nappies at night (Figure 10). Sometimes it spreads to the vulva or it may be found there primarily. Impetigo sometimes occurs on the vulva and perianal area.

The presentation is with pustules and crusted lesions, which are often more itchy and irritating than painful. The diagnosis is made with a skin swab, *Staphylococcus aureus* usually being isolated. Impetigo usually responds quickly to antibiotics but folliculitis can be very persistent and is often better treated with topical agents.

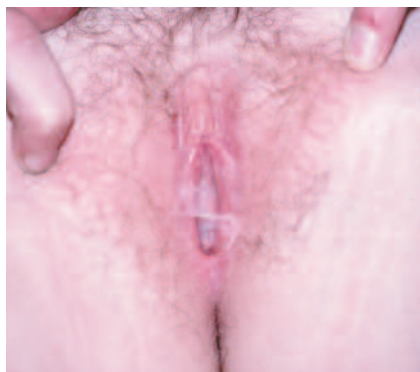


Figure 7. Streptococcal vulvovaginitis with erythema and mucoid discharge.



Figure 9. Erythema multiforme.

### Pinworm infestation

The symptoms of pinworm infestation are perianal and vulval itching, particularly at night, and sometimes a dermatitic rash. Many cases of pinworm infestation are, however, asymptomatic. Pinworm is well known as a cause of genital itching in children, and many children with vulval disease will already have been treated with mebendazole (Combantrin-1 with Mebendazole, Vermox) by their parents or their pharmacist before they see a doctor.

### Molluscum contagiosum

The lesions of molluscum contagiosum are very common in children. The virus spreads in water, hence the predilection for the lower body where the child sits in the bath. It is not uncommon for mollusca to be found on the vulva, often as



Figure 8. Fixed drug eruption.



Figure 10. Staphylococcal folliculitis.

part of a more generalised eruption.

Sometimes vulval mollusca can be difficult to differentiate from genital warts: close examination under magnification will show the typical central core of mollusca (Figure 11). The differentiation is important because mollusca, unlike genital warts, are generally not considered to be sexually transmitted in children.

In most cases, it is not necessary to treat vulval mollusca as spontaneous resolution invariably occurs.

### Genital warts

Lesions of genital human papilloma virus are very uncommon in children. However, genital warts should always raise the question of sexual abuse. The difficulty is that sometimes warts may be transmitted nonsexually from other family members, autoinoculated from fingers or even acquired at birth from a mother with genital warts. It is, therefore, best to refer the decision about sexual abuse to, for instance, a child protection unit.

Genital warts typically have a filiform appearance and may involve the vulva, vaginal opening and perianal area (Figure 12). They are usually self-limiting, but can occasionally become large enough to interfere with toilet routines and may then need treatment.

### Genital herpes

Herpetic lesions of the vulval area are very uncommon in children. In infancy, herpes simplex virus can be acquired at birth from a mother with genital herpes, or inoculated from an adult with a herpetic lesion on the mouth or finger. Children with severe atopic dermatitis are also prone to herpetic infections in unusual locations, including the genital area.

The presentation of genital herpes in children is the same as in adults, i.e. painful vesicles that rapidly erode and ulcerate, and lymphadenopathy. There may also be oedema of the labia. The finding of a primary attack of genital herpes in a child should be confirmed by

culture, and should raise the possibility of sexual abuse if herpes simplex virus type 2 is isolated.

There are two very uncommon conditions in the differential diagnosis of herpes: vulval aphthous ulcers and vulval bullous pemphigoid. Both present with painful ulcerating lesions, and it is not surprising that they are often mistaken for herpes. Viral culture, however, is negative. Such cases are often queried as sexual abuse.

Both varicella and herpes zoster in children may involve the genital area but are usually part of a more widespread eruption (Figure 13).

### Fungal infections

Candidiasis does not occur in children who are no longer wearing nappies. In adults with chronic vulval symptoms, about 15% have candidiasis, but this oestrogen-dependent condition is not seen after infancy in children with normal immune systems. This is an important point because it is common for children with skin diseases such as dermatitis and psoriasis to be diagnosed as having 'thrush' and to be treated with antifungal creams, which may cause irritation, particularly if dermatitis is present.

Tinea is rarely found on the vulva in young girls (Figure 14), although it sometimes causes vulval rashes in women and is a common cause of groin rashes in men. When it does occur on the vulva of children, it hardly ever has typical features, often because of previous treatment with topical corticosteroids. It may be more common than we think because the imidazole cream used to treat the many cases of vulval eruptions that are 'assumed' to be candidiasis would, in most childhood cases, also treat tinea.

In cases where no antifungal agent has been used and the rash has been treated as dermatitis, tinea of the vulva or tinea under the nappy in a baby presents as a dermatitic rash that does not respond to

treatment. The diagnosis requires a high index of suspicion but once thought of is easily confirmed by a skin scraping.

### Anatomical abnormalities

#### Fusion of the labia

Fusion of the labia is sometimes seen in children aged 3 years and under. It is not a malformation but is acquired and may appear very early – it has even been seen at birth. The labia minora or majora are agglutinated to a variable degree from the tip of the clitoris to the posterior fourchette (Figure 15). The vulva may look abnormal with no apparent vaginal opening, or relatively normal but with what appears to be a membrane across the vagina when the labia majora are parted.

It is not clear why the labia join in some children, but many of the affected children do have an underlying dermatitis. Some affected children are asymptomatic but others have soreness or itching. Urine can pool behind the fusion causing irritating maceration. Urinary tract infections, however, are rarely a complicating factor.

The condition tends to resolve spontaneously, although treatment with oestrogen cream may be required in the meantime.

#### Pyramidal perineal protrusion

Although pyramidal perineal protrusion has only recently been labelled as an entity in the medical literature, it is probably not rare. It is noticed in infancy as an asymptomatic soft protrusion of the median raphe in girls. The overlying skin is normal. The aetiology of this acquired condition is unknown, and it resolves spontaneously.

#### Ambiguous genitalia and imperforate hymen

The anatomical malformations ambiguous genitalia and imperforate hymen can be confused with fusion of the labia as an opening to the vagina may not be apparent. A specialist opinion should be sought if there is any doubt.



Figure 11. Molluscum contagiosum.



Figure 12. Human papillomavirus infection.

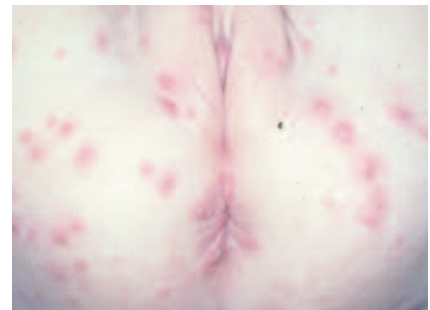


Figure 13. Varicella.

### Foreign bodies

Although intravaginal foreign bodies are often mentioned as a cause of vulval disease in the medical literature, they are not a common event. Usually the foreign material is a fragment of toilet paper or fluff. Larger objects, such as small toys, are less common.

The child presents with a persistent purulent discharge heavy enough to cause maceration of the vulval skin (Figure 16). Swabs show recurrent bacterial

continued

infection that responds to antibiotics but rapidly recurs. Referral is recommended for examination under anaesthesia and saline lavage.

### Psychological aspects of vulval problems

In adults with vulval complaints, there is a small but significant group who presents with symptoms (usually burning pain) but with no apparent abnormality. Although some of this group are malingering or somaticising, there are many more who have a genuine complaint of neuropathy or referred pain.

When a child presents in this way, with symptoms but nothing to see, even after close examination when symptoms are maximal, it is unlikely that there is any physical complaint. A common scenario is the child who is presented because of a greenish discharge noticed as staining of the underwear but who has no other symptoms and normal swabs and urine culture on investigation. This situation is a normal variant.

A somewhat less innocent situation is where a child constantly complains of vulval discomfort in the absence of findings and without any observable signs of being in pain. Children rapidly realise that complaining of genital pain, particularly at school or in public, attracts much adult attention and is a source of embarrassment for their parents. They may even find that they are sent home from school by teachers worried about sexual abuse allegations, and distraught parents are summonsed to explain the behaviour. In other words, havoc can be created. Children who capitalise on this have no idea how much adult distress they are causing, but they do know that it is an effective attention-seeking device.

Children who masturbate and whose parents are shocked by this behaviour often learn to explain their actions to their parents by saying they are in pain. The parents may need to come to terms with the normality of their child's actions.

These cases can catch out the unwary. Nonintervention, reassurance and not giving in to attention-seeking behaviour are usually the best treatments, although psychiatric help may occasionally be required.



Figure 14. Tinea. The rash is nonspecific but does not respond to treatment.



Figure 15. Agglutination of the labia.



Figure 16. Foreign body, causing maceration of vulval skin.

### Conclusion

Most conditions of the vulva in children are dermatological rather than gynaecological, and can be diagnosed on clinical appearance and history alone, the only investigation needed being a skin swab for bacterial infection. The range of vulval conditions seen in children is smaller than that in adults, and the prevalence is also less. However, with the exception of candidiasis, the common diseases seen in both groups are similar. In Australia, virtually all cases of acute infective vulvitis are due to *S. pyogenes*. Birthmarks may occur on the vulva, but malignancy is exceptionally rare. Anatomical abnormalities are uncommon, although the acquired condition fusion of the labia is seen sometimes in young girls and may be confused with anatomical malformations.

Vulval symptoms in the absence of signs are unusual in children. It should be kept in mind that some children might make an issue of vulval symptoms as an attention-seeking device. **MT**

Next month, part 2 of this article will discuss the diagnosis and management of vulval disease in children and also the issue of vulval conditions being misdiagnosed as child sexual abuse.

### Further reading

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3. Pokorny SF. Prepubertal vulvovaginopathies. *Obstet Gynecol Clin North Am* 1992; 19: 39-58.