

Pediatric Vulvovaginal Disorders: A Diagnostic Approach and Review of the Literature

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Abstract

Vulvovaginal complaints in the prepubertal child are a common reason for referral to the health care provider. The Cochrane Library and Medline databases were searched for articles published in English from 1980 to December 2004 relating to vulvovaginal conditions in girls. The following search terms were used: vulvovaginitis, prepubertal, pediatric, lichen sclerosis, labial fusion, labial adhesion, genital ulcers, urethral prolapse, psoriasis, and straddle injuries. The objectives of this article are to review the normal vulvovaginal anatomy, describe how to perform an age-appropriate examination, and discuss common vulvovaginal disorders and their management in young girls.

Résumé

Les plaintes de nature vulvo-vaginale chez l'enfant prépubère sont couramment à l'origine d'une orientation vers les services d'un fournisseur de soins de santé. Des recherches ont été menées dans les bases de données Cochrane Library et Medline en vue d'en tirer les articles publiés en anglais, entre 1980 et décembre 2004, portant sur les pathologies vulvo-vaginales chez les filles. Les termes de recherche suivants ont été utilisés : *vulvovaginitis*, *prepubertal*, *pediatric*, *lichen sclerosis*, *labial fusion*, *labial adhesion*, *genital ulcers*, *urethral prolapse*, *psoriasis* et *straddle injuries*. Les objectifs de cet article sont de passer en revue l'anatomie vulvo-vaginale normale, de décrire la façon de mener un examen approprié en fonction de l'âge et de discuter des troubles vulvo-vaginaux courants et de leur prise en charge chez les jeunes filles.

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INTRODUCTION

Some of the most common problems presenting to a pediatric gynaecologist are those involving the vulvovagina. The pediatric vulvovagina is particularly susceptible to problems because of the anatomy of the external genitalia, lack of estrogenization, and frequent contact with irritants. We describe the normal anatomy, discuss how to perform a proper examination, and review basic hygiene measures to maintain vulvovaginal health in these young patients. We also review common disorders including vulvovaginitis, labial adhesions, lichen sclerosus, genital ulcers, urethral prolapse, and straddle injuries, and discuss appropriate management strategies for these conditions.

METHODS

The Cochrane Library and Medline databases were searched for English language articles published from 1980 to December 2004 relating to common vulvovaginal conditions in girls. The following search terms were used: vulvovaginitis, prepubertal, pediatric, lichen sclerosus, labial fusion, labial adhesion, genital ulcers, urethral prolapse, psoriasis, and straddle injuries. Additional publications were then identified using the bibliographies of these articles.

ANATOMY OF THE PEDIATRIC VULVA

Prior to the onset of puberty, the pediatric vulva will differ significantly from the adult in its anatomic features. The transient estrogen effect from maternal estrogens in the newborn gradually resolves within the first six months of

life. Compared with the adult vulva, the pediatric vulva is hairless and has very little subcutaneous fat under the lateral aspects of the mons pubis and labia majora. The labia minora lack pigmentation and have an atrophic appearance. The distance from the anus to the vestibule is comparatively short, leaving the pediatric vulva more prone to irritation and inflammation.^{1,2}

Histologically, the vulva is covered by keratinized, stratified squamous epithelium. The vestibule is composed of squamous epithelium resembling vaginal mucosa, which is not glycogenated in the pediatric patient and does not have estrogen effect until puberty. The labia majora in the adult will have sebaceous glands associated with hair follicles and can open directly on to the surface epithelium toward the medial aspect; the labia minora do not contain glandular elements but may have sebaceous glands near the interlabial sulcus. The apocrine glands of the labia majora, prepuce, posterior vestibule, and perineal body are not activated until puberty and are not often involved in pathology in the prepubertal child. However, the eccrine sweat glands do function prior to puberty. Miliaria related to obstruction of the eccrine sweat glands is common in neonates.¹

The linea vestibularis is a white streak in the midline of the posterior vestibule and may be seen in up to 25% of newborn females. It is important to be aware of this as it can be misinterpreted as a scar secondary to abuse.¹ The fossa navicularis (the shelf-like area in the vestibule leading up to the hymen) and the lateral vestibular sulci are densely vascular and in most children will have an erythematous appearance. The glans clitoridis may appear relatively more prominent in children nearing puberty than in adults due to the flat appearance of the labia majora and minora. The actual size of the clitoris (glans and shaft) in a newborn should measure < 0.9 cm when fully stretched.^{3,4}

The hymen is a fold of vascularized mucous membrane that lies within the vaginal orifice and separates the vagina from the vestibule (Figure 1). It can show great variation in thickness, size, and shape. Normal diameters of the opening can be up to 1 cm. There is no distensibility prior to puberty. The most common type of hymen is the crescentic hymen, which starts in the periurethral area at 1 o'clock and extends around to the 11 o'clock position; the circumferential (annular) hymen extends full circle. The hymen is originally

Figure 1. Normal anatomy



a solid membrane that opens during the fetal period. Abnormalities in this process can result in variations including imperforate hymen (no opening), microperforate hymen (a single small opening), cribriform hymen (multiple small openings), and septate hymen (a residual band, usually in the anteroposterior diameter).² The vagina in the child is proportionally smaller in length and diameter, reaching 6 cm in length prior to puberty. It also has very little distensibility. The cervix will be flush with the vaginal vault or protrude slightly. The columnar mucosa of the cervix in the child is not exposed to the vagina.²

It is important to know the normal genital anatomy of the prepubertal child and to be familiar with the variations that may exist. This will help to ensure that a normal variation is neither interpreted as pathological nor suspected as sexual abuse. One should also be aware that because of the anatomy and lack of estrogenization, the pediatric vulva is more susceptible to irritants and trauma.

THE GYNAECOLOGIC EXAMINATION OF THE CHILD

Successfully completing a gynaecologic examination in the pediatric patient can be challenging for all parties involved: the patient, her caregiver, and the clinician. Prior to beginning the examination, it is important to explain it carefully to both the child and her parent or caregiver. These young patients must be assured that the examination, although perhaps uncomfortable or embarrassing, will not be painful. Emphasize that if any instruments are used, they are specifically designed for little girls. Provide both parent and child the opportunity to ask questions. Educate the child in developmentally appropriate language and emphasize that only parents or caregivers and doctors or nurses can touch or examine her genital area.

ABBREVIATIONS

EBV	Epstein-Barr virus
LS	lichen sclerosus
HPV	human papillomavirus
HSV	herpes simplex virus
VZV	varicella zoster virus

In most situations, the parent will remain in the room to assist in the examination and to hold the child's hand. Do not rush the examination, as this will precipitate anxiety and resistance in the child. If the child is very resistant, consider deferring the examination until the next visit, or if the problem is urgent, consider performing the examination under anaesthesia. Always start the examination with a general pediatric assessment of the child's weight and height, head and neck, chest, abdomen, and inguinal areas. Starting with a general examination will often put the child at ease prior to proceeding with the genital examination.

The examination should include inspection of the external genitalia, visualization of the vagina and, rarely, a rectoabdominal examination. Some physicians use mirrors during the examination to show normal and abnormal anatomic details. Alternatively, the use of video colposcopy for the genital examination can be very useful since it reduces apprehension and is well-accepted by young patients. A permanent record can be obtained and used for further discussion and documentation.

The most common position used for genital examination is the frog-leg position. This can be attained by asking the child to lie on her back, spread her knees apart and have her feet touch together. Having the child in the lithotomy position with use of adjustable stirrups, or positioning the child in the frog-leg position on her mother's lap are alternatives. Lateral spread technique can be used to visualize the external genitalia, hymen, and distal vagina. The thumb and index finger grasp the posterior aspect of each of the labia majora and gentle outward and lateral traction is applied (Figure 2). The patient is asked to "Valsalva" or cough to aid in the visualization of the distal portion of the vagina. In girls over two years old, the knee-chest position provides a particularly good view of the vagina and cervix without instrumentation.⁵ The chest is placed on the table, resting the head to one side on folded arms, while the buttocks are raised in the air with knees bent (6–8 inches apart).⁵ An assistant helps to hold the buttocks apart, pressing laterally and slightly upward. When the child takes a deep breath, the anterior vagina wall will move down allowing for visualization of the vagina and cervix. Since the vagina of the prepubertal child is quite short, a foreign body or lesion can often be detected in this position.⁵ A supine position with the child's knees flexed on her abdomen is an alternative method for visualizing the hymen, vagina, and anus.⁶

Although not universally recommended, visualization of the vagina can be better accomplished using a small nasal speculum or veterinary otoscope (7 mm) with viscous lidocaine placed at the introitus.⁷ An otoscope can be used to provide magnification and light. If the vagina is not easily seen with good positioning, vaginoscopy can be performed

Figure 2. Separation and traction



to visualize the lower vagina and cervix. A step-by-step approach for inserting the vaginoscope in the office setting has been described by Capraro.⁸ Vaginoscopy requires a light source, irrigation, and a hysteroscope or pediatric cystoscope. It can be performed without a general anaesthetic, but relies on the child being extremely cooperative. If an ideal view of the lower vagina is needed (e.g., to rule out a foreign body), an examination under anaesthesia is recommended.

Finally, a gentle rectoabdominal examination can be performed if a mass is suspected or if the patient's complaint is primarily abdominal pain. This should be done last because it is the part of the evaluation that the child is most apt to dislike.⁵ With the child in frog-leg position, the examiner performs a bimanual examination, placing the index or little finger of one hand into the rectum and the other hand on the abdomen. The child should be reassured that a finger has a smaller diameter than a bowel movement and should not cause any discomfort.⁶ Since the ovaries are not palpable in the child and are located higher in the pelvis than in the adult, masses should alert the physician to the possibility of a cyst or tumour.⁶ At the end of the rectal examination, as the finger is removed from the rectum, the vagina can be gently "milked" to promote the passage of any discharge or extremely rare polypoid tumours.⁶

VULVAR HYGIENE

There are two key elements in the institution of proper vulvar hygiene: the removal of irritants and the institution of healthy vulvar hygiene practices. These practices will promote healthy lifelong hygiene habits and prevent non-specific vulvovaginitis.

All harmful, irritating, or offending agents must be removed. Nylon clothing, tight-fitting clothing (stockings,

ballet leotards), and prolonged exposure to wet bathing suits should be avoided.⁹ Loose-fitting cotton clothing is best. At night, a loose-fitting night garment with no underwear is recommended. Undergarments should be washed with a mild, unscented detergent, and fabric softeners and anti-static dryer sheets should not be used.^{9,10} If convenient, air-drying of the child's undergarments is preferred.

Proper vulvar hygiene begins with good bath practices. Girls who are overweight are particularly susceptible to non-specific vulvovaginitis. The child should sit in a tub of warm clear water for 10 to 15 minutes daily.¹⁰ The vulva should never be scrubbed. If smegma is present between the labial folds, this area may be cleansed with gentle front to back washing with a mild soap such as Dove, Aveeno, or Neutrogena.¹¹⁻¹³ The child should never sit in a tub with soap, shampoo, or bubble bath. The child can stand up to have her hair washed and rinsed so that she doesn't sit in the soapy water. The vulva should be dried with gentle patting of the vulvar skin or air-drying. When voiding, the child should urinate with her legs wide apart, and she should lean forward to minimize urine pooling in the lower vagina. Following urination or defecation, the child should always wipe in a front to back direction to minimize contamination of the vulva and vagina with enteric organisms.^{14,15}

These helpful tips should be emphasized to all parents and toilet-trained children to help avoid irritation and infections.

VULVOVAGINITIS

Vulvovaginitis continues to be the most common indication for referral to a pediatric gynaecologist. Most children with vulvovaginitis are managed by primary care physicians or pediatricians, with the more severe or recurrent cases referred for specialist care.¹⁶ One must be cautious when reviewing the medical literature regarding the etiology and the pathogens responsible for vulvovaginitis in children, because the majority of evidence is based on referral populations and not children in primary care settings.¹⁷

Vulvitis and vaginitis in the pediatric population may occur separately or in combination. Symptoms and signs of vulvitis alone include pruritus, tenderness, dysuria, and erythema of the vulva (Figure 3). The presence of discharge is more indicative of a vaginitis. The pathologic discharge associated with vaginitis should be distinguished from the physiologic clear, mucoid discharge that occurs during the newborn period and with the onset of pubertal development.¹⁸ The most common presenting clinical features in a recent review of vulvovaginitis in a referral population of prepubertal girls were vaginal discharge in 92%, pruritus in 45%, and dysuria in 30%. Bleeding and pain were reported much less frequently at 8% and 5% respectively. Redness of

Figure 3. Vulvovaginitis



the vulva was reported by 30% of caregivers.¹⁹ Soreness of the vulvar region is more frequently reported in a primary care setting (74%) and may be the predominant presenting symptom.²⁰

The increased susceptibility of children to vulvovaginitis is due to a combination of anatomic and behavioural factors. The anatomic factors include the close proximity of vagina to anus, the lack of labial fat pads and pubic hair, the thin atrophic non-estrogenized vaginal mucosa, the thin delicate vulvar skin, and an alkaline vaginal pH.^{19,21} Behavioural factors that may facilitate the development of vulvovaginitis include a tendency to poor hygiene, children's natural curiosity with exploration of their bodies including masturbation, and, in some, underlying chronic constipation.^{15,19,22-24}

The assessment of any child with vulvar irritation includes taking an appropriate history and conducting a physical examination. The history should include the child (if possible) and involves assessment of the duration of symptoms, presence and description of discharge, prior home or prescribed therapies, presence or absence of pubertal development, trauma, history of foreign body insertion, history of dermatitis or atopy, perineal hygiene, and concern regarding abuse. The physical examination should include a general physical examination, with particular attention to Tanner staging and general assessment of skin and mucosal surfaces for lesions. An inspection of the external genitalia will demonstrate perineal hygiene, vaginal discharge, hymenal anomalies, skin lesions, secondary excoriations, and evidence of trauma. A rectoabdominal examination may be of value in the presence of a foreign body and may express discharge not previously visualized.²⁵ Further assessments may include a vaginoscopy or vaginal cultures, depending on the differential diagnosis.

The differential diagnosis of vulvovaginitis includes vaginal foreign bodies, sexual abuse, sexually transmitted infections, pinworms, lichen sclerosus, psoriasis, eczema, contact dermatitis, scabies, lichen planus, ectopic ureter, congenital enteric fistula, and systemic disorders (e.g., Kawasaki disease, Crohn's disease, scarlet fever).^{16,21,25,26}

Non-specific vulvovaginitis is vulvovaginal irritation without an identifiable bacterial pathogen, and it accounts for 74% to 80% of all cases.^{19–21,24} It may also be referred to as irritant or atopic contact dermatitis.¹⁹ The term allergic contact dermatitis is used when a specific irritant can be identified (e.g., latex) but is uncommon in the pediatric age group.¹⁹ Non-specific vulvovaginitis often responds to a regimen of hygiene measures and avoidance of any identified irritants.²¹ With severe inflammation, topical estrogen or a topical steroid cream may facilitate healing.²¹ Referral to a specialist should occur in clinical situations when symptoms fail to resolve with appropriate treatment measures, when the diagnosis is unclear, or when parents require reassurance.

Positive cultures are more likely in the clinical setting of visible vaginal discharge with moderate to severe inflammation extending beyond the introitus.^{19,20,27} Cultures are obtained trans-hyemenally from the lower vagina with the child relaxed in a frog-leg or knee-chest position. A fine cotton swab moistened with saline (Calgiswab or urethral aluminum swab) may be passed easily into the vagina without touching the hymenal edges and causing discomfort. Another method of obtaining samples is by inserting the proximal end of an intravenous butterfly catheter into the end of a red rubber catheter.²⁸ With a syringe attached, this can be used to flush 1 mL of sterile normal saline into the vagina and then used to aspirate the sample for cytology, wet mount, and cultures. If abuse is suspected, Chlamydia and gonorrhea can be recovered from vaginal secretions in the prepubertal population.²⁷

Pathogens may be demonstrated in vaginal cultures of up to 26% of children with vulvovaginitis.^{19,20} Recognition of bacteria present in the normal flora will limit the over-treatment of non-pathogens. The non-sexually transmitted pathogens in vulvovaginitis are group A beta-hemolytic *Streptococcus*, *Haemophilus influenzae*, *Staphylococcus aureus*, *Moraxella catarrhalis*, *Streptococcus pneumoniae*, *Neisseria meningitidis*, *Shigella*, and *Yersinia enterocolitica*. *Escherichia coli* may be present in asymptomatic patient populations, and therefore coliforms are often not considered pathogens.²⁹ Group A beta-hemolytic *Streptococci* and *Haemophilus influenzae*, transferred from the upper respiratory tract, are the two most common causative agents, in both primary care settings and referral populations.^{17,19,29–31} Cyclical variations in the incidence may follow the trends of upper

respiratory tract infections with similar organisms; with streptococcal vulvovaginitis, concurrent throat swabs are positive in 5% to 92%.^{21,24, 29,31,32} *Candida* is rarely, if ever, a pathogen in the prepubertal child with vulvar irritation.^{23,24} Contrary, however, to evidence of pathogens in cultures, a survey of general practitioners in the United Kingdom revealed that 41% believed the most common pathogen associated with prepubertal vulvovaginitis was *Candida*. As a result of their expectation of causation, the most common treatments prescribed to this patient population were antifungals.¹⁶ Many families may hold a similar erroneous belief, hence it is important to ask about home remedies applied during the intake history. *Shigella* is an uncommon cause of prepubertal vulvovaginitis. Although it has been reported that 50% of patients with *Shigella* vulvovaginitis present with a bloody discharge, a more recent review from an endemic community found that bloody discharge and diarrhea are absent in the majority of cases.^{33,34} A quinolone or cefixime antibiotic should be used for empiric therapy.³³ Treatment of a specific pathogen includes the hygiene measures recommended for non-specific vulvovaginitis and appropriate antimicrobial therapy, usually by oral administration.

In the presence of a vaginal foreign body, the presenting symptoms are more likely to be recurrent or resistant to previous treatment modalities and accompanied by foul smelling discharge or, less commonly, vaginal bleeding. A vaginoscopy, which may be facilitated by sedation or general anaesthesia, will allow visualization of the foreign body. Toilet paper is the most common foreign body found. It may be flushed from the vagina using a pediatric feeding tube, Foley catheter, or red rubber catheter as previously described. Viscous lidocaine can be placed at the introitus, if necessary, for anaesthesia.

In the setting of predominant perianal pruritus, nocturnal symptoms, and a low-grade vulvovaginitis, consider the diagnosis of pinworms (*Enterobius vermicularis*). Secondary infection with fecal organisms such as *E. coli* is common with spread of the pinworms from the anus to the vagina. Demonstration of pinworms involves either inspection of the anal area at night with a flashlight or application of clear adhesive tape to the anal area in the morning to collect ova.²⁶ As both of these measures may prove awkward and difficult, empiric treatment with 100 mg mebendazole, repeated two weeks later, is an option.¹⁹

LABIAL ADHESIONS

Labial adhesions, also known as labial agglutination or labial fusion, constitute an acquired condition in which the labia are adherent in the midline (Figure 4). The estimated incidence of labial adhesions in prepubertal girls has typically

been reported as 0.6% to 3%.^{35–39} Recent studies have revealed that it may be present in as many as 38.9% of healthy girls.^{40,41} It is not present in newborn females.³⁹ The hypothesized etiology is that irritants denude the thin, non-estrogenized epithelium of the labia.⁴² Adhesions then form and re-epithelialization occurs, forming an avascular connection between the two labia. It has been suggested by some authors that labial adhesions should arouse suspicion of sexual abuse,^{43–45} but given how frequently labial adhesions are identified, this is likely an uncommon association.

Young girls typically present between six months and six years of age.^{37,45} The patient may mistakenly be referred for congenital absence of the vagina, ambiguous genitalia, or imperforate hymen.

There is controversy regarding the best treatment for prepubertal girls with labial agglutination.⁴⁶ The limited information available in the literature is all retrospective. Only one study looking at the natural history of the condition has been published; this reported that all agglutination resolved without treatment within 18 months ($N = 10$).⁴⁷ The hypothesis is that once the young girls begin endogenous estrogen production, the adhesions will resolve spontaneously.

Many authors suggest using hygiene measures with or without some form of bland cream (e.g., petroleum jelly or vitamin A + D ointment).⁴⁸ Hygiene measures consist of removal of all potential vulvovaginal irritants (soaps, bubble baths, restrictive clothing) and daily sitz baths. Bland creams protect the labial epithelium from irritants, thereby discouraging adhesions. One study with data on the treatment with bland cream ($N = 5$) showed no change after one month.⁴⁹ The authors did not mention use of additional hygiene measures (i.e., sitz baths).

When patients are symptomatic or when significant agglutination is present, estrogen cream can be used. Symptoms can include vulvovaginal irritation, urinary dribbling, and urinary tract infections. Urinary retention is uncommon.⁵⁰ Conjugated estrogen cream is approved for use in postmenopausal women for short-term management of urogenital symptoms, but is not approved for use in children, despite its common use in adults. The efficacy of estrogen cream for the treatment of prepubertal labial agglutination ranges from 50% to 91% in the literature, while treatment duration varies, with application once or twice daily for two to six weeks.^{49,51,52} Estrogen cream should be applied with a gentle amount of pressure to the line of fusion only, either with a finger or cotton swab. The pressure itself may aid in resolving the adhesions. Side effects can include local irritation (erythema or burning), vulvar pigmentation, or pubertal changes (breast buds).³⁷ Most studies ($N = 25–50$) do not report any pubertal

Figure 4. Labial adhesions



changes.^{34,49} One study reported breast budding in 8.7% (2/23), but the length of treatment was not specified.⁵³ The breasts regressed following cessation of treatments in both cases. The exact incidence of all these various side effects and their relation to estrogen dose is not known. Following successful treatment with estrogen, the risk of recurrence may be minimized by daily application of a barrier cream (e.g., petroleum jelly); we suggest that a duration of six months should be sufficient.

Manual separation should be reserved for patients with acute urinary retention (complete agglutination) or failed medical treatment. This may be accomplished in an office setting using topical anaesthesia (e.g., EMLA cream or xylocaine)^{53,54} or awake sedation (e.g., midazolam),⁵⁵ with minimal discomfort and high success. General anaesthesia in the operating room is also an option, particularly if the child is very anxious or the adhesions are thick. One should note, as with estrogen treatment, there is a significant recurrence rate, which may be as high as 40%.⁵³

LICHEN SCLEROSUS

Lichen sclerosus is a chronic skin disorder of unknown etiology. Familial cases have been reported, suggesting a genetic tendency.^{56–60} There is an established association between LS and autoimmune diseases such as vitiligo, thyroid disease, alopecia areata, rheumatoid arthritis, and diabetes mellitus.^{56,60,61} There also appears to be an immunogenetic association with HLA class II DQ7.^{56,60,62,63} There may also be a hormonal component, as patients with LS are often premenarchal or postmenopausal.^{57,58,60,63,64} One study reported a 75% improvement of symptoms at menarche, with only 30% concomitant improvement in physical signs.⁶³ It has also been postulated that genital carriage of human papillomavirus in prepubertal girls may be a

trigger for the development of LS. At the present time, there is no evidence to suggest HPV-typing in patients with lichen sclerosis.⁶⁵

Lichen sclerosis is seen most frequently in adult women, but 10% to 15% of cases arise during childhood.^{56,63,66–68} Its prevalence has been estimated at less than 1 in 900 in premenarchal girls.⁵⁷ The mean age at diagnosis in children is 5.5 to 6.7 years.^{56–59,68} Clinically, these young girls may present with pruritus, soreness, erosions and fissures, papules and lumps, bleeding, dysuria, constipation, or pain with defecation.^{57,63,68} Because of the variable presentation, these patients may present to pediatricians, urologists, gastroenterologists, dermatologists, or gynaecologists. Important points to elicit on history include age at presentation, symptoms, vulvar hygiene practices, history of trauma or sexual abuse, personal or family history of skin diseases, atopy or autoimmune diseases, and previous treatments.

LS typically appears as a sharply demarcated white figure-of-eight encircling the vulva and anus (Figure 5).^{58,63,64,66,68,69} Erythema, purpura, fissuring, erosions, and scarring are often present. Over time, altered architecture with fusion and resorption of the labia minora and introital narrowing may occur. Papules and lichenification involve the labia and anal area without affecting the hymenal structures. Extragenital manifestations are uncommon, but may involve the arms, trunk, shoulders, and head.^{57,66,70} There have been reports of lichen sclerosis leading to mistaken accusations of abuse.^{57,58,63,66,70} The diagnosis is usually made by clinical examination; a biopsy is rarely required.⁶⁸

Lichen sclerosis is best treated with a potent topical corticosteroid. The use of a short course of high potency topical corticosteroid appears to be effective, safe, and well-tolerated, with minimal side effects. The pediatric literature includes a few small prospective case series (N = 10–15).^{66–69} Various formulations have been used, including 0.05% betamethasone dipropionate and 0.05% clobetasol propionate ointments.^{66–68} The frequency and duration of treatment ranges from two to three times daily application for up to 6 to 12 weeks, until clinical resolution.^{66–68} Some authors report continuing 1% hydrocortisone ointment daily or 0.1% triamcinolone following resolution for an additional three months.⁶⁷ Clinical improvement has been reported in 93% to 100% of pediatric patients.^{66–68} Recurrences are common and reported to be as high as 60%,⁶⁶ or an average of two flare-ups per year.⁶⁸ Side effects may include steroid-induced atrophy, telangiectasias (0–27%),^{66,67} or erythema (18%).⁶⁷ Although not yet well studied in the treatment of LS, tacrolimus, a novel immunomodulating agent, may prove useful in the future.^{69,71}

Figure 5. Lichen sclerosis



PSORIASIS

Psoriasis affects 0.5% to 3.0% of adults.⁷² In one study, 31.5% of patients with psoriasis could recall that the onset of symptoms was prior to age 16.⁷³ In another review, 2% of patients were diagnosed with psoriasis as infants, 8% as children, and 25% as adolescents.⁷² In an audit of prepubertal girls seen by dermatologists, psoriasis was diagnosed in 17% of those referred with vulvar abnormalities.⁷⁴ Nearly one half of patients may have a first degree relative with psoriasis.⁷³

Psoriasis most frequently affects the scalp, extensor surfaces of the limbs (elbows and knees), and the sacral region.⁷⁵ In the genital area, the vulva, perineum, and anus may be involved, sparing the labia minora and vagina.^{74,76,77} While the classic appearance of psoriasis is discrete erythematous papules and plaques covered with silvery scales, the lesions of the genital region are not scaly.⁷⁷ The lesions may have a glazed appearance with or without superficial erosions and fissuring deep in folds (flexural psoriasis).⁷⁸ Nail pitting or the presence of scalp or postauricular erythema and scaling may be subtle signs to help confirm the diagnosis.⁷⁴ In rare instances, the vulva may be the only site of disease.⁷⁵ Stress, cutaneous injury, and upper respiratory streptococcal infections may trigger exacerbations.⁷² The appearance of a psoriatic lesion at the site of skin injury is referred to as Koebner's phenomenon, appearing 3 to 30 days post injury.^{72,75} As psoriasis is primarily a process that involves the epidermis, scarring is rare.⁷⁸ An earlier onset of disease does not indicate a more aggressive form of disease.⁷⁸

The treatment of psoriasis in children does not differ from that in adults, although some of the treatments applied to psoriasis elsewhere on the body are too irritating for vulvar application.^{76,77} Avoidance of skin injury and good vulvar

hygiene should be emphasized. Emollients and moisturizers may be helpful.⁷² Investigate for and treat concurrent infections with antibiotics and antifungals as indicated. Streptococcal vulvovaginitis may be seen in conjunction with psoriasis.⁷⁴ Moderate to high potency topical steroids are commonly used for initial control; fluorinated ointments are recommended to be used two to three times daily.^{72,76} Low potency steroids can be used for maintenance with or without weak tar preparations. Systemic treatments, such as methotrexate or retinoids, should be considered second line agents for vulvar psoriasis and some authors suggest avoiding their use in the pediatric population.^{72,77}

GENITAL ULCERS

In developed countries, most sexually active patients with genital ulcers are diagnosed as having herpes (30%), but up to 55% will have a non-specific ulcer of unknown etiology.^{79,80} This is in contrast to Africa and Asia, where the most common diagnoses are chancroid (45%) and syphilis (25%).⁸⁰ Genital ulcers are relatively uncommon in girls and young women without a history of sexual activity, and published reports are scarce. The largest case series reported findings in nine such girls; eight of nine were premenarchal. Six patients had viral systemic symptoms but no etiology could be determined, and three patients had recurrent ulcers and a possible diagnosis of Behçet's syndrome.⁸¹

Genital erosions and ulcers in children and adolescents can present a complex diagnostic dilemma and therefore a good history is crucial. Enquire about sexual activity, because this is pivotal to the differential diagnosis. Be aware that inoculation of infectious agents can occur through digital- and oral-genital contact. Explore the possibility of sexual abuse if there is any suspicion. Enquire if the ulcer is painful (it would be painless with syphilis), recurrent (as in herpes or Behçet's), or present elsewhere (suggesting dermatitis or vasculitis).⁸²⁻⁸⁴ Ask if there are associated symptoms such as fever or malaise (suggesting a viral etiology, such as mononucleosis or herpes) or gastrointestinal symptoms (as in Crohn's disease).^{84,85} A history of travel should alert the clinician to infections rarely seen in North America (e.g., amebiasis or leishmaniasis).^{80,84} Enquire about medications (to rule out fixed drug reaction).^{79,84} A thorough past medical history is important, as the presentation and natural history of genital ulcers can be influenced by many factors including immune status, secondary infection, systemic disorders, and dermatologic disease.

Carefully examine the skin and oral mucosa and assess for signs of systemic illness. Examine the vulva and perineum. A blister is a fluid-filled vesicle that can rupture to leave an erosion, involving only the epidermis; an ulcer affects both

Figure 6. Vulvar ulcers



the dermis and epidermis⁷⁹ (Figure 6). The differential diagnosis of solitary ulcers should include syphilis (*Treponema pallidum*), chancroid (*Hemophilus ducreyi*), Crohn's disease, pyoderma gangrenosum, ulcerative vulvitis (bacterial pathogen or trauma), and cancer (basal or squamous cell carcinoma).^{79,84,86} Multiple ulcers are more typical with herpes, chicken pox or shingles (VZV), mononucleosis (EBV), secondary syphilis, candidiasis, scabies, Behçet's syndrome, aphthous ulcers, and fixed drug reactions.^{79,84,86}

Acute ulcers should be cultured for bacterial, viral, and fungal infections.^{79,80,84} A biopsy may be performed, particularly if the ulcers are unexplained and chronic or recurrent.^{79,84} The biopsy should be done at the edge of the lesion, not at the base, and should include normal skin. Serology may be helpful (HSV, VDRL, EBV).^{79,80,82,84} Depending on the level of suspicion, consultation with a specialist in dermatology, gastroenterology, ophthalmology, or rheumatology may be necessary to exclude a specific diagnosis. The following provides a brief description of some of the more common diagnoses to consider in young girls (Table).

1. Herpes simplex virus infection presents with painful, multiple, vesicular, or ulcerative lesions. The incubation period is two to seven days. During the first episode, 50% to 75% have systemic symptoms including fever, malaise, headache, myalgias, and bilateral tender inguinal lymphadenopathy.^{79,80} The majority of genital lesions are caused by HSV-2.^{79,80,84} Auto-inoculation from HSV-1 can occur, and it often has less frequent recurrences.^{79,80} The virus is usually shed from active lesions, but asymptomatic shedding can occur.^{79,80} The inactive virus resides in the dorsal root sacral ganglia. Virus isolation by tissue culture is the most accurate means of diagnosis.^{80,83,87} Treatment

Differential diagnosis of genital ulcers in young girls

Infectious	Non-infectious
Venereal Herpes (HSV) ^{15,76,79-91,93} Syphilis (<i>T. pallidum</i>) ^{79,81,82,86-88,90} Chancroid (<i>H. ducreyi</i>) ^{79,80} Lymphogranuloma venereum (<i>C. trachomatis</i> L1,2,3) ^{79,80} Granuloma inguinale (<i>C. granulomatis</i>) ^{79,80} Scabies ^{79,80,83,84}	Dermatologic Aphthous ulcers ^{77,79,82} Dermatitis: lichen sclerosus, lichen planus, psoriasis, eczema ^{79,80,82,84,85,87} Vesiculobullous skin disease: Bartholinitis, hidradenitis suppurativa, pemphigus ^{74,76,79,82,84,86} Vasculitis Behçet's syndrome ^{77,79-83,86-88,91} Systemic lupus erythematosus ^{79,84,86}
Non-venereal Viral: chicken pox/shingles (VZV) ^{79,82} Mononucleosis (EBV) ^{81,82,84,86,87} HIV, other Fungal: Candida, Actinomycosis ^{79,80,82,84} Bacterial: pseudomonas aeruginosa, diphtheria, (para) typhoid ^{74,79,82,84} Other: amebiasis, brucellosis ^{79,80}	Other Crohn's disease ⁸⁶ Insect bites ^{79,84} Trauma: foreign body, sexual injury ^{79,84,90} Adverse drug reaction ^{76,77,79,80,86,87} Neoplasm: Paget's, Bowen's, basal or squamous cell carcinoma, lymphoma, leukemia ^{79,82,84,86}

is with analgesics, oral antiviral medication (such as acyclovir), and antibiotics if secondary infection is present. Intravenous therapy may be required for severe disease.

- Epstein Barr virus is the infectious agent causing mononucleosis.⁸⁶ Patients typically present with flu-like symptoms and a sore throat. Various case reports have confirmed acute infection with EBV in the presence of vulvar ulcers.^{81,86} The ulcers are typically described as painful, punched-out lesions with irregular borders.⁷⁹ They may last a few weeks but are usually self-limited. The monospot heterophile antibody test may not be reliable and therefore EBV titres are needed for a definitive diagnosis. Treatment is supportive and may include sitz baths and topical or oral analgesics.
- Aphthous ulcers, known as canker sores when oral, can also be seen in the genital area. They are painful, shallow ulcers involving mucosal surfaces. They are non-infectious and possibly immune-mediated. Treatment is symptomatic.
- Behçet's syndrome is a chronic, relapsing systemic vasculitis of unknown etiology.^{79,81,82,87,88} It is more prevalent in individuals of Japanese or Mediterranean origin. Patients typically present with recurrent, painful oral or genital ulcers (or both) that may be single or multiple, shallow or deep, round to oval in shape, and have a yellowish necrotic base.^{79,83,84,85} There are no specific diagnostic tests. The International Study Group for Behçet's suggests that the diagnosis includes

the presence of oral aphthous ulcers recurring at least three times a year, plus at least two of the following: recurrent genital ulcers, uveitis or retinal vasculitis, skin involvement such as erythema nodosum, or a positive pathergy test.^{79,84,86,87} Treatment is primarily symptomatic; corticosteroids and other drugs (e.g., colchicine, methotrexate) have been used.^{83,84,85}

- Crohn's disease, an inflammatory bowel disease, may have vulvar manifestations including linear ulcers with prominent edema, fissures, or fistulas,^{74,76,79,81,83,84,89-91} Occasionally vulvar ulcers may precede gastrointestinal symptoms.⁹²
- Fixed drug eruptions can involve the genitalia.^{77,79,82,84,87} Ulcers are recurrent, occurring in the same location each time the drug is ingested. Possible implicated medications include NSAIDs, acetaminophen, metronidazole, sulfonamides, tetracycline, phenytoin, oral contraceptives, or barbiturates.^{77,84,87}

In conclusion, the differential diagnosis of genital ulcers is extensive. The history should ascertain if the person is sexually active and whether this is a first episode or recurrent. If vesicles are present, HSV is the most common diagnosis. The ulcer(s) should be cultured and a biopsy may be considered. In young girls, HSV or syphilis should raise the suspicion of sexual abuse. Even after thorough investigations, a specific cause for vulvar ulcers is often difficult to establish, especially in non-sexually active children.

Figure 7. Urethral prolapse

URETHRAL PROLAPSE

Urethral prolapse occurs in prepubertal and menopausal females. It is more common in black female children than in white.⁹³ The hypo-estrogenic state predisposes the child to urethral prolapse, but often the precipitant is a history of repeated Valsalva, such as would occur with chronic constipation, chronic cough, or a urinary tract infection with the constant sensation of needing to strain. The urethral mucosa protrudes beyond the urethral meatus, forming a beefy, red, friable, congested mass (Figure 7). The patient may be referred with urinary tract symptoms (hematuria, dysuria), blood staining in the diaper or undergarments, a vaginal mass, or concerns about abuse.

Opinions differ regarding the ideal and most effective management strategy. Some authors advocate surgery as first line therapy,^{93,94} while others believe that conservative approaches are successful in the vast majority of cases.^{95,96} Unfortunately the case series are often small and always retrospective chart reviews.^{97,98} Rubin et al. in 1997 reported the largest series, involving 58 white prepubertal girls. Of those children, 38 failed conservative medical therapy either initially or because of recurrence.⁹⁹ Nonetheless, local care and medical therapy is minimally invasive and does not require an anaesthetic. Most authors agree that with the first presentation, these basic measures should be tried before opting for mucosal excision.⁹⁵⁻⁹⁹ Conservative treatment includes topical estrogen (at least nightly), soothing tub soaks, and analgesics.⁹⁵ Some authors report using hexachlorophene soap and topical proviodone in addition to estrogen cream and sitz baths.⁹⁶ The child may need to void in a tub bath to reduce the discomfort. Treating the Valsalva-related precipitant is paramount. When treatment is successful, the urethral mucosa will regress over the course of a few days to a few weeks. Alternatively, when

Figure 8. Straddle injury

conservative measures are unsuccessful or when the problem is recurrent, surgical excision is warranted. Essentially, the prolapsed distal mucosa is excised with re-anastomosis of the proximal urethral mucosa to the vestibule using a fine absorbable suture.⁹³⁻⁹⁹ There is little consensus on technique or success rates.⁹³⁻⁹⁹ Rubin et al. reported surgical complications that included bleeding, urethral stenosis, and recurrence.⁹⁹ Another study of 23 patients concluded that ligation over a Foley catheter should be discouraged.⁹⁸

STRADDLE INJURY

Straddle injury usually refers to a genital injury resulting from inadvertent trauma to the perineum. As the term implies, the mechanism of injury often results in separation of the child's legs, allowing direct force on the vulva and perineum. Occasionally the mechanism can involve something sharp, but usually the force is blunt. Examples would include falling on to the cross-bar of a bicycle, slipping off a diving board and making contact with the edge upon descent, or mishaps related to monkey bars or jungle gyms.¹⁰⁰

The history is paramount in the context of genital trauma. There must be a clear and plausible explanation, preferably provided by both the child and an adult witness independently.¹⁰¹ Most injuries are minor lacerations or abrasions of the labia minora and posterior fourchette, accompanied by bruising of the labia majora and mons (Figure 8). With very rare exception, the labial fat pads of the vulva protect the hymen and lower vagina, preventing tearing or compromise.^{102,103} Often the perineal body and posterior fourchette are splayed apart, resulting in a shallow separation from the shear force. The labia minora can be severed, and this often results in persistent bleeding. External bruising, which may not be noted until the following day, is a common finding on physical examination. Fortunately, significant injuries to

the urethra and bladder are uncommon in females with a straddle injury except when there has been a great force and a concurrent straddle-related pelvic fracture. The management of trauma-related injuries to the urinary tract is outlined in the urologic literature.^{104–109} Indications for examination under anaesthesia include an inability to void or concern about urethral integrity, ongoing bleeding requiring suture or hemostasis, a large or expanding hematoma that needs to be evacuated, suspected anal sphincter injury, and penetrating injury necessitating inspection of the upper vagina. When explanations or reports are inconsistent, deliberate injury should be suspected.^{102,103} Hymenal, vaginal, or perianal lacerations suggest a penetrating genital injury and are suspicious for sexual assault or abuse. Signs of other injuries elsewhere on the body may raise suspicion of abuse. An assessment for abuse by an experienced and skilled care provider should be undertaken.¹⁰¹ The collection of forensic evidence may be necessary.

Conservative management consists of analgesics, soothing soaks, and intermittent ice packs to reduce swelling. The child may need to void in water to avoid stinging. If this is unsuccessful and/or if the child develops urinary retention, an indwelling urethral catheter may be required in the short term. This may be placed during examination under anaesthesia or with some sedation to avoid an additional painful experience. Education must be provided, and follow-up should be arranged within one week.

CONCLUSION

Caregivers should be aware of the normal vulvovaginal anatomy in young girls. The examination should be conducted by a physician who is familiar with manoeuvres to maximize patient compliance and visibility, while minimizing discomfort for the child.

Conditions such as vulvovaginitis and labial adhesions will often resolve with the institution of proper vulvar hygiene and the elimination of irritants. Vaginal swabs for culture should be considered if vaginal discharge is present. A diagnosis of lichen sclerosus is typically made by clinical examination alone and is treated with potent topical corticosteroids. Vulvar ulcers in this age group, without a history of sexual activity, can be difficult to diagnose; despite thorough investigations, the etiology often remains unclear. Urethral prolapse can usually be treated with topical estrogen therapy and rarely requires surgery. Straddle injuries should be managed conservatively with sitz baths unless a penetrating injury is suspected or there is overt bleeding, in which case an examination under anaesthesia should be performed. If a child presents with an injury to the vulva or perineum without a reasonable history to explain the injury, abuse should always be considered.

Referral to a specialist should be considered for any vulvovaginal condition if the symptoms persist despite suggested treatment regimens.

REFERENCES

1. Kurman RJ. Blaustein's Pathology of the female genital tract. 5th ed. Springer-Verlag: New York; 2002.
2. Sanfillipo J, Muram D, Lee P, Dewhurst JC. Pediatric and adolescent gynecology. WB Saunders: Philadelphia; 2001:187–9.
3. Ogilvy-Stuart A, Brain C. Early assessment of ambiguous genitalia. *Arch Dis Child* 2004;89(5):401–7.
4. Low Y, Hutson JM. Rules for clinical diagnosis in babies with ambiguous genitalia. *J Pediatr Child Health* 2003;39:406–13.
5. Emans SJ, Goldstein DP. The gynecologic examination of the prepubertal child with vulvovaginitis: use of the knee-chest position. *Pediatrics* 1980;65(4):758–60.
6. Emans SJ. Office evaluation of the child and adolescent. In: Emans SJ, Laufer MK, Goldstein DP, eds. *Pediatric and adolescent gynecology*. 4th ed. Lippincott-Raven: Baltimore; 1998:1–48.
7. Billmire MD, Farrell MK, Dine MS. A simplified procedure for pediatric vaginal examination: use of veterinary otoscope specula. *Pediatrics* 1980;65(4):823–5.
8. Capraro VJ. Gynecologic examination in children and adolescents. *Pediatr Clin North Am* 1972;19(3):511–28.
9. Wilson MD. Vaginal discharge and bleeding. In: Carpenter SEK, Rock JA, eds. *Pediatric and Adolescent Gynecology*. 2nd ed. Lippincott Williams and Wilkins: Philadelphia; 2000:151.
10. Emans SJ. Vulvovaginal problems in the prepubertal child. In: Emans SJ, Laufer MK, Goldstein DP, eds. *Pediatric and adolescent gynecology* 4th ed. Lippincott-Raven: Baltimore; 1998: 75–107.
11. Paek SC, Merritt DF, Mallory SB. Pruritus vulvae in prepubertal children. *J Am Acad Dermatol* 2001;44(5):795–802.
12. Morelli JG, Weston WL. Soaps and shampoos in pediatric practice. *Pediatrics* 1987;80(5):634–7.
13. Baranda L, González-Amaro R, Torres-Alvarez B, Alvarez C, Ramírez V. Correlation between pH and irritant effect of cleansers marketed for dry skin. *Int J Dermatol* 2002;41:494–9.
14. Mroueh J, Muram D. Common problems in pediatric gynecology: new developments. *Curr Opin Obstet Gynecol* 1999;11(5):463–6.
15. Farrington PF. Pediatric vulvo-vaginitis. *Clin Obstet Gynecol* 1997;40(1):135–40.
16. Cox RA. *Haemophilus influenzae*: an underrated cause of vulvovaginitis in young girls. *J Clin Pathol* 1997;50:765–8.
17. Jones R. Childhood vulvovaginitis and vaginal discharge in general practice. *Fam Pract* 1996;13(4):369–72.
18. Vandeven AM, Emans SJ. Vulvovaginitis in the child and adolescent. *Pediatr Rev* 1993;14(4):141–7.
19. Stricker T, Navratil F, Sennhauser FH. Vulvovaginitis in prepubertal girls. *Arch Dis Child* 2003;88:324–6.
20. Jaquiere A, Stylianopoulos A, Hogg G, Grover S. Vulvovaginitis: clinical features, etiology, and microbiology of the genital tract. *Arch Dis Child* 1999;81:64–7.
21. Barron SA. Index of suspicion. Case 3. Diagnosis: vulvovaginitis. *Pediatr Rev* 1998;19(2):51–4.
22. van Neer PAFA, Korver CRW. Constipation presenting as recurrent vulvovaginitis in prepubertal children. *J Am Acad Dermatol* 2000;43:718–9.
23. Pierce AM, Hart CA. Vulvovaginitis: causes and management. *Arch Dis Child* 1992;67:509–12.