The paediatrician and the management of common gynaecological conditions

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ABSTRACT

Paediatric gynaecology is an emerging discipline. Since 2000, there has been an advanced training programme in paediatric gynaecology available for obstetric and gynaecology trainees; additionally, a set of clinical standards[1] for the care of paediatric and adolescent patients has been developed by The British Society of Paediatric and Adolescent Gynaecology (BritSPAG). BritSPAG is a multidisciplinary group of professionals including gynaecologists, paediatricians, paediatric urologists and endocrinologists.

Girls with gynaecological conditions are often seen in general paediatric services; it is important that those assessing them are confident in identifying patients who require more specialist care. Despite this, gynaecology does not appear in the Royal College of Paediatrics and Child Health curriculum. This article aims to increase the knowledge base and confidence of paediatricians in dealing with common paediatric and adolescent gynaecological conditions.

VULVAL DISORDERS

Differential diagnoses of vaginal discharge (table 1)

Vulvovaginitis is the most common[2 3] gynaecological problem seen in girls 2–8 years of age (table 1).[4]

Many factors predispose to vulvovaginitis, including the prepubertal anatomy of underdeveloped labia, lack of fat pads, proximity of the anus to the vagina and absence of pubic hair.[3 5 6] The vaginal introitus is exposed and there is no physical protection against infection, particularly when a small girl sits or squats.[3] In prepubertal girls, in whom oestrogen concentrations are low, the vulvovaginal mucosa is thin, delicate and highly susceptible to invasion by pathogens.[2 3] The pH of the prepubertal vagina is less acidic than in older females, and resistance to infection is lower.[7]

Most cases of vulvovaginitis in this age group are non-specific in nature and are caused by the normal vaginal flora of mixed aerobes and anaerobes. Candida vulvovaginitis is rarely seen in childhood.

The most common presenting complaints are vaginal discharge, soreness, irritation, burning and redness of the region. Symptoms can vary in severity from minor itching to distressing pain waking the child up from sleep. Examination findings can range from minimal redness to severe inflammation with excoriation and lichenification. It is important that all carers are sensitively asked about the risk of sexual abuse but reassured that this is a rare cause of vaginal discharge.

Investigation

No investigations are required routinely in vulvovaginitis. In cases of severe symptoms or failure to respond to conservative treatment, a swab may be taken from the area of the posterior fourchette/introitus.[8] The swab can be taken either with the child lying on their parent's lap or in the frog leg position.[3 7 8] It is important not to undertake a true vaginal swab as performed in adults as this will cause unnecessary pain and distress to the child. Any further examination would require a general anaesthetic and should be reserved only for severe or refractory cases or when a foreign body or sexual abuse is suspected.

Management

Reassurance of the child and family about the benign, recurrent nature of the condition and natural resolution of symptoms by the onset of puberty[6] is important. The mainstay of management is avoidance of irritants such as bubble-baths, synthetic underwear, tight clothing, education about perineal hygiene including wiping front-to-back and wearing cotton underwear.[6 8] Hypoallergenic emollient shower gels can be used as a replacement to bubble-baths and soaps; a salt or vinegar bath could be considered, and some children may find having showers rather than baths helpful. Application of bland barrier creams to inflamed areas can help to relieve soreness. There is no evidence to support the routine use of topical antibiotics, antifungals or oestrogen creams.

Antibiotics should only be used in severe cases if a pure or predominant pathogen is identified from a swab.[2] If a sexually transmitted infection is identified, local child protection services should be contacted and further investigations as mandated in the local hospital safeguarding guidance will be required.

Foreign body

Foreign bodies should be considered if vaginal discharge does not respond to treatment or if there is a bloody or offensive discharge.[5 7] If a foreign body is suspected, this is best removed under general anaesthetic with vaginoscopy done with a paediatric cystoscope or a hysteroscope.

Differential diagnoses of vulval irritation (table 2)

Labial adhesions

Fused labia or labial adhesions are usually asymptomatic and are discovered during nappy changing or bathing. Infrequently it can cause urinary

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Table 1 Differential diagnoses of vaginal discharge

Vulvovaginitis	Most common cause
Foreign body	*Diagnosis not to miss*
Child sexual abuse	*Diagnosis not to miss* A thorough social history is mandatory and safeguarding procedures should be initiated where concerns arise

dribbling, vulval irritation and soreness. On examination, the labia can be seen to be fused from the level of the posterior fourchette up towards the urethra.[6] Often a fine line is seen in the midline between the two fused labia with a pinpoint hole through which urine is passed (table 2).

Management

No treatment is usually required, and the parents can be reassured that the adhesions will spontaneously resolve at the onset of puberty. In symptomatic cases, those with difficulty in passing urine, treatment is usually with topical oestrogen cream. A small portion of topical oestrogen cream is applied daily to the midline generally up to a maximum of 6 weeks.[6] Recurrence is common and a repeat course of treatment might be necessary.

Surgical separation may rarely be required in symptomatic cases which do not respond to oestrogen cream.

Threadworm/pinworm infestation

Threadworms are a common cause of anal irritation, but they can also cause vulval itching typically worse at night. Treatment is with a single dose of mebendazole 100 mg. Treatment of all family members is recommended.

Lichen sclerosus

Lichen sclerosus is a chronic skin disorder of presumed autoimmune origin, usually associated with postmenopausal women, but also seen in prepubertal girls. It usually presents with severe vulval irritation, pain and itching. On examination, the appearance is of white plaques over the vulva and perianal area, in more severe cases bleeding, erosions and ulcerations[3] can occur.

Vulval biopsies are rarely warranted in children and examination alone is sufficient.[5]

Management

If symptoms are mild, only an emollient is needed. In more severe cases, a topical corticoid steroid such as clobetasol propionate 0.05%, applied twice daily for up to 2 weeks can be used.[9]

Parents can be reassured that, unlike in postmenopausal women, there is no association with progression to vulva squamous cell carcinoma.[3 6–8] Lichen sclerosus is believed to resolve with the onset of puberty, although recent evidence suggests this may not always be the case.[10]

Elongated labia

Both the labia majora and the labia minora can vary in their size, colour and shape, with a wide range in physiological appearance.

Table 2 Differential diagnoses of vulval irritation

Labial adhesions	
Threadworm/pinworm Infestation	
Lichen sclerosus	
Elongated labia	
Child sexual abuse	*Diagnosis not to miss*

Table 3 Differential diagnoses of amenorrhoea

Physiological: constitutional, anorexia, excessive exercise	Most common cause
Polycystic ovarian syndrome	Can be a cause of both primary and secondary amenorrhoea
Premature ovarian failure ,for example, Turner's syndrome	Can be unexplained
Complete androgen insensitivity syndrome	*Diagnosis not to miss* Management must be within a multidisciplinary disorders of sexual development team, so that expert psychological care can be accessed
Mayer-Rokitansky-Kuster-Hauser syndrome	*Diagnosis not to miss* Management must be within a multidisciplinary disorders of sexual development team, so that expert psychological care can be accessed
Outflow tract disorder: imperforate hymen, transverse septum and uterus didelphys	*Diagnosis not to miss*

Girls should be reassured about their labial appearances. Simple measures to relieve labial discomfort should be suggested, such as wearing comfortable underwear, not shaving pubic hair and the use of emollients.[11]

Labial reduction is a contentious issue; a recent Royal College of Obstetrics and Gyanecology statement confirms that it should not be undertaken in those aged under 18.[12]

MENSTRUAL DYSFUNCTION

Differential diagnoses of amenorrhoea (table 3)

Definition: Primary amenorrhoea is defined as the absence of menarche by the age of 15 years or by 3 years after the onset of breast development (table 3).

The first step in the evaluation of girls with amenorrhoea is to determine if puberty has commenced. Girls with normal breast development have functioning gonads, directing the clinician to consider anatomical abnormalities of the reproductive tract, or in the obese patient with clinical or biochemical features of androgen excess, polycystic ovarian syndrome (PCOS). In girls with no evidence of secondary sexual characteristics by the age of 13 years, puberty is considered to be delayed,[13] and consideration should be given to endocrine disorders of the hypothalamic–pituitary–ovarian axis.

Measurement of follicle stimualting hormone (FSH) is a helpful screening test (table 4). If FSH is not elevated and there is a family history of delayed puberty, constitutional delay is likely. However, any chronic medical condition can result in delayed puberty, as can sudden weight loss, excessive exercise and anorexia. Rarely, delayed or arrested puberty in the presence of a normal or low FSH concentrations can be the first presentation of a lesion of the hypothalamic–pituitary pathway, such

Table 4 Follicle stimulating hormone (FSH) as a screening test

Low FSH	High FSH
Structural lesion of the hypothalamic pituitary pathway Constitutional Excessive exercise Anorexia Chronic disease	Turner's syndrome Swyer's syndrome Premature ovarian failure

as craniopharyngioma or pituitary adenomas, including prolactinoma. Thyroid dysfunction must always be excluded.

Elevated FSH concentrations point to a diagnosis of primary ovarian failure, and blood should be sent to exclude Turner's syndrome and Swyer's syndrome (46 XY).

In the presence of normal breast development, a transabdominal pelvic ultrasound should be performed to determine whether the uterus is present and to examine for evidence of outflow tract disorder. If there is suspicion of developmental anomaly or a suboptimal ultrasound, MRI scan should be performed.

Complete androgen insensitivity syndrome

Affected girls have a short vagina, and the uterus and fallopian tubes are absent. During puberty, breast development may progress normally as testosterone is aromatised to oestrogen; however, axillary and pubic hair do not develop. Endocrine investigations show that FSH is appropriate for the stage of breast development and testosterone is high. Genetic testing shows a 46XY karyotype. Most commonly, the diagnosis is made at puberty, when girls present with primary amenorrhoea. Gonadectomy may be considered as the testes are usually intra-abdominal and therefore at increased risk of malignant change, after which oestrogen replacement will be necessary.[6]

Turner's syndrome and other causes of premature ovarian failure

Patients with Turner's syndrome should be managed by a group of experienced professionals fully conversant with the complex needs of these patients. International consensus guidelines are available,[14] and professionals from a wide range of disciplines are required to ensure that all their healthcare needs are addressed.

Approximately 25% of girls with Turner's syndrome enter puberty spontaneously, and 16% are reported to achieve menarche with regular menstrual cycles.[15] In girls in whom FSH is clearly elevated, and there are no signs of breast development, pubertal induction can be started from the age of 10,[13] with the aim of developing secondary sexual characteristics, attaining a normal height in conjunction with growth hormone, and possibly oxandrolone therapy for normal peak bone mass. Pubertal induction is started with oestrogen replacement.[13] After 2–3 years, progesterone is added alongside oestogen[13] for endometrial protection.

Structural abnormalities of the uterus and vagina

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome

Patients with MRKH have normal secondary sexual characteristics, a normal endocrine profile and a 46XX karyotype. The uterus is absent, and there may also be some shortening of the vagina. Renal tract abnormalities occur in 30% of patients.[16] Vaginal dilatation is often required for normal sexual function.

Outflow tract disorders

Like patients with MRKH syndrome, breast development and endocrine tests are normal.

An outflow tract disorder will usually be diagnosed on examination or with imaging that will reveal a haematocolpos, the accumulation of blood in the vagina. The girl may be asymptomatic and present only with amenorrhoea or more commonly with cyclical abdominal pain. Other presentations include urinary retention, back pain, vomiting and diarrhoea or an abdominal mass.[3]

Table 5 Differential diagnoses in heavy menstrual bleeding

Dysfunctional uterine bleeding	Anovulation is the most common cause
Hypothyroidism	Thyroid function tests should be performed if there is any suspicion of thyroid dysfunction
Polycystic ovarian syndrome	Often with irregular cycles
Bleeding disorders: Von Willebrand disease, platelet function defects	***Diagnosis not to miss***

The most common cause of outflow tract disorder is an imperforate hymen. On examination, an imperforate hymen typically has the appearance of a bulging, bluish membrane which is visualised at the vaginal introitus.[6] A hymenectomy is a surgically straightforward procedure to relieve the obstruction.

A transverse vaginal septum, being thicker, will not create a bluish membrane as blood will not be visible behind it and instead will have a pinkish appearance. It may present in the same way as an imperforate hymen with delayed menstruation and cyclical pain, but more extensive surgery is required to correct and preoperative imaging is important if the classical appearance of an imperforate hymen is not evident.

Uterus didelphys is a rare genital abnormality which results from failure of the two Müllerian ducts to fuse together and is commonly associated with a vaginal longitudinal septum creating a double vagina. A small number of patients may present with a unilateral obstructed hemivagina and haematocolpos. The diagnosis can be difficult and is likely to be delayed as these patients will often already be menstruating from the non-obstructed hemivagina.

Differential diagnoses in heavy menstrual bleeding (table 5)

A thorough history should be taken including detailed menstrual history, personal history of excessive bleeding (eg, after tooth extraction or prolonged epistaxis) and family history of bleeding disorders (table 5).

Investigations

A full blood count and ferritin should be measured. The coagulation screen should be interpreted in collaboration with a haematologist in cases of severe menorrhagia (causing anaemia and in patients unresponsive to standard treatments).[17]

Management

Non-hormonal

If there are no contraindications, non-steroidal anti-inflammatory drugs (mefenamic acid or ibuprofen) are the first-line treatment to reduce blood flow with or without tranexamic acid (1 g four times a day) to be given during the days of heavy flow up to a maximum of 4 days every cycle.

Hormonal

1. *Progesterone*: Norethisterone 5 mg or medroxyprogesterone acetate 10 mg two or three times a day from day 5–25 every cycle for 6 months is an option when there is contraindication or reluctance to use the combined pill.

2. *Combined contraceptive methods: combined contraceptive pill/oral contraceptives (COCP)*: These methods should produce a regular, painless, light, predictable cycle. They can be taken with a monthly break or taken consecutively for 3 months at a time. For those who cannot take COCP, the alternative routes of administration are transdermal patch or vaginal ring.

Box 1 Investigation of adolescents presenting with PCOS-like symptoms[19]

Follicle stimualting hormone, lutenizing hormone, oestradiol (in amenorrhoeic adolescents)

Thyroid stimulating hormone

Prolactin: Mildly elevated in patients with PCOS. If concentrations exceed 1000 mU/L, imaging of the pituitary should be performed to exclude a prolactinoma.[20]

Total and free testosterone: The free androgen index (total testosterone divided by sex hormone binding globulin (SHBG) × 100).[21]

Dehydroepiandrosterone

17-OH progesterone

Other causes of hyperandrogenism, including atypical congenital adrenal hyperplasia, Cushing syndrome and tumours of the adrenal gland, should be excluded if the testosterone level is >5 nmol/L,[22] or 17 hydroxyprogesterone or dehydroepiandrosterone sulfate are above the upper limit of the adult reference range.[18]

Ultrasound of ovaries

Once PCOS has been confirmed

Fasting and 2-hour GTT increased risk of metabolic syndrome and type 2 diabetes.

Lipid panel

Fasting insulin

PCOS, polycystic ovarian syndrome.

Polycystic ovarian syndrome

PCOS is a cause of heavy menstrual bleeding and primary as well as secondary amenorrhoea. Diagnosis is usually made with the Rotterdam criteria (all three must be present):

1. Polycystic ovaries (either 12 or more follicles or increased ovarian volume (>10 cm^3)) on ultrasound scan.
2. oOligo-ovulation or anovulation.
3. Clinical and/or biochemical signs of hyperandrogenism.

However, diagnosis can be difficult in the adolescent. First, polycystic appearances on ultrasound can be normal. Transvaginal scans are not performed routinely in patients of this age and abdominal scans may be less accurate, particularly in patients who are significantly overweight or obese.[18] Additionally, oligo and anovulation are also normal in the first 18 months following menarche and clinical features of androgen excess are part of normal puberty. This confusion has led to recommendations that different diagnostic criteria for PCOS should be used in the adolescent including otherwise unexplained, persistent hyperandrogenic anovulation, using age-appropriate and stage-appropriate standards.[18]

Investigation of adolescents presenting with PCOS-like symptoms

See box 1.

Management

For those with PCOS who have less than four periods a year, it is recommended to have a withdrawal bleed with cyclical progestogens for at least 7–12 days, every 3–4 months.[20]

For those with menstrual dysfunction as well as androgenic symptoms, combined pill containing cyproterone acetate can be considered for up to 2 years. The long-term management is weight loss and symptomatic treatment of presenting symptoms.

SUMMARY

Girls present commonly with gynaecological complaints, most of which are due to benign conditions that can be treated safely by a paediatrician. However, they may be the first presentation of serious or complex pathology, or child sex abuse, and the prompt recognition of patients requiring specialist care is essential. Close collaboration between specialists from a range of disciplines may be required to manage some of the most complex cases, and the development of multidisciplinary services within the paediatric setting has the potential to facilitate the sharing of knowledge and skills in this new and evolving specialty.

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