

Adolescent gynaecology

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Abstract

Paediatric and adolescent gynaecology involves the care of young women and their special needs as their bodies change and grow into adulthood. Diagnosis and management of common problems in gynaecology in this age group requires knowledge regarding congenital reproductive anomalies, disorders of gender development, reproductive endocrinopathies, gender identity, and gynaecologic malignancies. Gynaecologic disease states in paediatric patients frequently present with vulval and vaginal manifestations, while those in the adolescent age range present as abdominal-pelvic pain and abnormal menstrual bleeding. A case-based learning of common childhood gynaecologic disorders and their treatment is presented.

Keywords dysfunctional uterine bleeding; immature malignant teratoma; imperforate hymen; ovarian mass; polycystic ovary syndrome; precocious puberty; primary amenorrhoea

Introduction

Paediatric and adolescent gynaecology requires knowledge of normal female embryological development, endocrine function around adolescent years, fertility, the impact of female genital system abnormalities on gender identity and mental health, and the medical and surgical management of these systems. Best care is administered with a multidisciplinary team (including adolescent gynaecologists, paediatricians, paediatric surgeons, endocrinologist, clinical geneticists and psychologists) to treat the underlying disorder, preserve fertility and future sexual function, and maintain a healthy mental state. This review presents different scenarios in children presenting with a diverse range of gynaecological diseases requiring knowledge of these systems.

Case 1: polycystic ovary syndrome and dysfunctional uterine bleeding

A 14-year-old girl presents with irregular menses since her menarche at the age of 11. Her menstrual flow usually lasts from 4 to 6 days, is heavy, and is associated with painful cramping. The intercycle interval varies between 40 and 60 days. Her mother's onset of menarche occurred at 12 years of age. The girl also complains of increased acne and facial hair over the last year accompanied by weight gain disproportionate to her growth in height.

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This adolescent has presented with clinical findings commonly associated with polycystic ovary syndrome (PCOS). As many as 5–15% of teens and women are affected by PCOS. Polycystic ovary syndrome most commonly begins during adolescence and presents with irregular menstrual cycles and hyperandrogenism. Difficulties with infertility and metabolic complications often develop over time. Polycystic ovary syndrome is diagnosed in adults using the Rotterdam consensus criteria. Two of the following criteria are required for a diagnosis of PCOS in adults:

- Oligo- or anovulation
- Clinical and/or biochemical signs of hyperandrogenism
- Polycystic ovaries

These criteria require special interpretation in adolescents. Polycystic ovaries contain 12 or more follicles measuring 2–9 mm in diameter in each ovary, or an ovarian volume greater than 10 mL on abdominal ultrasound in adult women. Similar criteria are sometimes used, but not well supported in adolescents. Ovaries generally remain stable in size, less than 2 cm³, in girls from 2 to 9 years of age, and progressively increase in size during adolescence. Follicle counts in adolescents are higher than in adults, and can lead to a misdiagnosis if the Rotterdam criteria are used. Best ovarian visualization is obtained with vaginal ultrasound in adults, but transabdominal ultrasound is used to visualize the ovaries in girls who are not sexually active. Girls experiencing regular menstruation should be scanned in the early follicular phase (cycle days 3–5), while those not yet experiencing menstruation can be evaluated at any time.

The 1990 Consensus Conference of the National Institute on Child Health and Human Development based the diagnosis of PCOS in adolescents on the presence of anovulation and hyperandrogenism, not explained by other endocrine disorders. Adolescents with polycystic ovary syndrome (PCOS) often present at menarche with irregular and long menstrual cycles. Adolescents with two years of irregular cycles following the onset of menarche should be evaluated for PCOS. Irregular cycles can occur from 45 days to a year apart and are generally heavy and associated with cramping pain. Adolescents with regular menstrual cycles and hyperandrogenism and/or polycystic ovaries should also be evaluated for PCOS. Many normal adolescents have irregular cycles regulated with oral contraceptives. Adolescents with undiagnosed PCOS treated with oral contraceptives to regulate their cycle can remain undiagnosed until they seek evaluation for infertility.

Dysfunctional uterine bleeding (DUB) is defined as irregular uterine bleeding with no pelvic or general medical disease, or pregnancy. Affected patients have a loss of the normal cyclic pattern of ovulatory luteinizing hormone (LH) surge in response to rising oestradiol levels during ovulation. Endometrium in these girls continues to proliferate under the influence of unopposed oestrogens, and does not undergo secretory change. Endometrial shedding in these girls is usually prolonged and heavy. DUB is observed in anovulatory and oligo-ovulatory women with PCOS. About 1–2% of women with anovulatory bleeding, who are not properly treated, would develop endometrial cancer.

Decreased insulin-mediated glucose utilisation, aka insulin resistance, is found in as many as half of affected patients. Girls with PCOS have a three to seven times increased risk of

developing type 2 diabetes. This risk is greatest in girls with anovulatory cycles, those with polycystic ovaries, obese girls, and girls with a family history of type 2 diabetes. The presence of altered lipid metabolism suggests these girls may be at increased risk for cardiac disease as well.

Clinical findings

Relevant history includes age at menarche, breast bud development, and appearance of pubic hair, duration of periods, amount of menstrual flow, presence of pain or cramping, time between periods, diet, and exercise patterns. Girls with PCOS most frequently have regular periods to start and then develop irregular periods. Growth charts of adolescents with PCOS can demonstrate a marked increase in weight following initiation of menses. A family history for PCOS and diabetes is also obtained.

Adolescents with PCOS and hyperandrogenism can demonstrate Hirsutism and virilisation. *Hirsutism* is characterized by excess facial and body hair. The frequency of Hirsutism is directly related to age, with adolescents being least commonly affected. Signs of *virilisation* include deep voice, frontal and temporal balding, increased muscle bulk, decreased breast size, and clitoral hypertrophy. The clinical finding of acne is generally normal during adolescence and can be confusing in virilised adolescents. *Acanthosis nigricans*, characterized by darkening of the skin on the back of the neck and other areas, is associated with moderate to severe insulin resistance.

At first clinic appointment, patient's height, weight, BMI, waist circumference, and blood pressure are evaluated. Affected adolescents generally have a central obesity pattern.

Laboratory testing

Patients are diagnosed with PCOS after excluding the diagnosis of congenital adrenal hyperplasia, androgen-secreting tumours, high-dose exogenous androgens, and Cushing's syndrome. A basal morning 17-hydroxyprogesterone level is determined in order to exclude 21-hydroxylase deficient non-classic adrenal hyperplasia in hyperandrogenic patients.

Serum follicle stimulating hormone (FSH) and 17 β -oestradiol levels are determined to exclude hypogonadotropic hypogonadism in young women presenting with oligo- or anovulation.

The free androgen index (total testosterone level divided by sex hormone-binding globulin level) and serum dehydroepiandrosteronesulphate (DHEAS) levels are used to identify biochemical hyperandrogenism. A dexamethasone suppression test is performed to exclude Cushing's syndrome.

At subsequent review appointments, consideration should be given to assess fasting serum glucose level, total, and low-density lipoprotein and high-density lipoprotein cholesterol levels to determine insulin resistance. Oral glucose tolerance testing is performed as indicated.

Luteinizing Hormone (LH) levels are significantly elevated in about 60% of girls with PCOS, due to the increased amplitude and frequency of LH pulses. The LH/FSH ratio is elevated in about 95% of affected patients. The clinical relevance of LH in PCOS and the potential effects of LH suppression with gonadotropin releasing hormone (GnRH) analogues are not known, so LH levels are not routinely determined.

Thyroid hormones are known to alter androgen metabolism and contribute to hyperandrogenism. However, the incidence of

hyperthyroidism in women with PCOS is not felt to be greater than that of the general population and the use of routine thyroid function determination is not recommended. Prolactin secretion can increase the production of androgens by the adrenal glands and prolactin levels are generally determined.

Treatment

Girls with PCOS require family and personal counselling to better understand the long-term impact of this diagnosis. Depression and anxiety can develop as the patient experiences the clinical impact of their disease. The treatment of PCOS is symptomatic. General recommendations include, diet, exercise, and weight reduction, which can improve serum androgen, insulin, LH and lipid abnormalities. Further treatment may be needed if the adolescent woman with treated PCOS desires pregnancy.

Medroxyprogesterone or Norethisterone administered for 10–14 days each month can be used to avoid abnormal endometrial proliferation and facilitate normal ovarian androgen production in adolescents. The low-dose oral contraceptives pills can also be used in girls with regular menses. Such treatment will provide contraception, prevent endometrial hyperplasia, decrease testosterone levels, and can improve hirsutism and acne. Adolescents with irregular menstrual cycles may also be started on oral contraceptives without evaluation for PCOS. Girls already taking oral contraceptives can be withdrawn for 3 months if biochemical testing is needed to diagnose hyperandrogenism or PCOS. Other contraceptive measures e.g. barrier contraception should be planned for during this testing period, when appropriate.

The use of metformin has been beneficial in increasing insulin sensitivity and decreasing serum LH and free testosterone levels. The use of metformin has been associated with the return of normal menstrual cycles in 68–95% of girls with prior normal cycles.

Antiandrogens such as spironolactone, flutamide, or cyproterone can be combined with oral contraceptive pills to decrease testosterone levels and treat hirsutism. GnRH analogues are generally reserved for girls that do not respond to these treatments or do not tolerate oral contraceptive pills. Long-term use of GnRH analogy can lead to bone demineralization, hot flushes, and atrophic vaginitis due to chronic low oestrogen levels, and may require monitoring as long as they are on this treatment.

Case 2: ovarian mass

A 15-year-old girl presents with acute peri-umbilical pain, palpable abdominal mass, nausea, vomiting, and radiation of pain to the groin. Pelvic ultrasound demonstrates a 17 cm complex pelvic mass. Plain computed tomography (CT) imaging demonstrates a large, heterogeneous mass with a prominent solid component, calcifications, and small foci of fat.

As many as 10% of women undergo evaluation for an ovarian mass during their life. Almost all ovarian masses in premenopausal females are benign. Pre-pubertal adolescents develop functional cysts due to the failure of involution of follicles. These cysts are caused by gonadotropin stimulation of the ovary by the immature hypothalamic–pituitary axis. Benign ovarian cysts/or immature follicular cysts, less than 1 cm in diameter, have been reported in 2–5% of pre-pubertal females undergoing

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