Adolescent gynaecology

Holly Vickers

Swati Jha

Abstract

Paediatric and adolescent gynaecology is an area of special interest in gynaecology and encompasses a spectrum of conditions affecting the gynaecological health of girls from birth through to adulthood. Management of conditions during pubescent development can be challenging and require sensitive multidisciplinary input. It requires an in-depth understanding of anatomy, embryology, pubertal development as well as an understanding of the differences between presentations in childhood, adolescence and adulthood and the effects of those conditions on patients' adult lives and reproductive potential. Another important aspect to consider is gender identity and the psychological impact that those conditions can have on patients at a young age. This review contains a summary of four diverse presentations seen in adolescent gynaecology clinic with a discussion of their management.

Keywords amenorrhoea; dermoid cyst; disorders of sex development; Mullerian aplasia; polycystic ovary syndrome

Introduction

Paediatric and adolescent gynaecology is a complex and highly specialised area requiring an in-depth understanding of embryology and adolescent development. The gold standard of care typically requires a multi-disciplinary team of professionals including adolescent gynaecologists, specialist nurses, endocrinologists, paediatricians, paediatric surgeons, clinical geneticists, infertility specialists and psychologists to treat the condition, in addition to the psychological and reproductive implications associated with that condition. This review contains four case scenarios with common presentations seen in adolescent gynaecology, with aetiology, incidence, presentation, investigations and management discussed.

Case 1: pelvic mass and cyclical pelvic pain

A 15-year-old girl presents with cyclical lower abdominal pain and a pelvic mass. She developed secondary sexual characteristics at the age of 12 and has menstruated regularly since then. She underwent further imaging which demonstrated a unicornuate uterus with a communication to the vagina and a separate non-communicating horn.

Holly Vickers *MBChB MRCOG Specialty Trainee 4 in Obstetrics and Gynaecology, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK. Conflicts of interest: none.*

Swati Jha *MD FRCOG Consultant Gynaecologist, Sheffield Teaching Hospitals NHSFT and Honorary Senior Clinical Lecturer at University of Sheffield, Sheffield, UK. Conflicts of interest: none.*

Genital outflow tract abnormalities

Congenital abnormalities of the genital tract are abnormalities related to the development of the uterus, fallopian tubes, ovaries, cervix, vagina and vulva, and can affect the normal reproductive and sexual function of women. The incidence of congenital abnormalities of the female genital tract is 2–4%. There are three main types of congenital genital outflow tract abnormalities:

1) **Isolated** abnormalities (e.g. imperforate hymen or vaginal septum);
2) **Complex** abnormalities of the genital outflow tract involving the cervix, uterus and fallopian tubes (e.g. unicornuate uterus or uterus didelphus); or
3) **Multi-organ** abnormalities. These abnormalities maybe related to syndromes or adjacent organs may be involved. (e.g. Mayer-Rokitansky-Kuster-Hauser syndrome).

When managing patients with these abnormalities it is important to have an in-depth understanding of the embryological development of the genital tract. This enables clinicians to understand the developmental malformations and adverse effects which are associated with these conditions.

Uterine abnormalities usually occur as a result of failure of the caudal ends of the Mullerian ducts to fuse. This can result in a variety of abnormalities ranging from a bicornuate uterus to uterus didelphus (Figure 1). A unicornuate uterus with a rudimentary horn can develop as a result of this fusion failure. Rudimentary horns can either be communicating or non-communicating with the main body of the uterus. Communicating rudimentary horns will have few symptoms however can be dangerous if a pregnancy is implanted in it. Non-communicating horns can be functional and have active endometrial tissue which will be shed on a cyclical basis. Hence a lack of communication from the rudimentary horn to the uterus will result in haematometra and retrograde menstruation. Where the rudimentary horn is non-functional i.e. has no endometrial stripe, the patient is likely to be asymptomatic and this is usually an incidental finding on ultrasound scan.

Clinical findings

Most girls with a unicornuate uterus and non-communicating functional rudimentary horns present in adolescence with severe dysmenorrhoea due to the increasing volume of haematometra.

A thorough history and examination is important to exclude other genital outflow tract anomalies and also to rule out conditions affecting other organ systems. History of menarche, pubertal development, menstrual cycle, sexual history and function, contraception, and previous medical problems should be taken.

Abdominal examination should be undertaken to exclude a pelvic mass which may be an enlarged uterus as a result of haematocolpos. Vulvo-vaginal examination can be performed to exclude an imperforate hymen or vaginal septum in the presence of primary amenorrhoea – however care must be taken to determine whether this is appropriate in adolescent girls and in some cases would require a general anaesthetic to do so. The presence of secondary sexual characteristics should be noted as this is a sign of normal pubertal development.

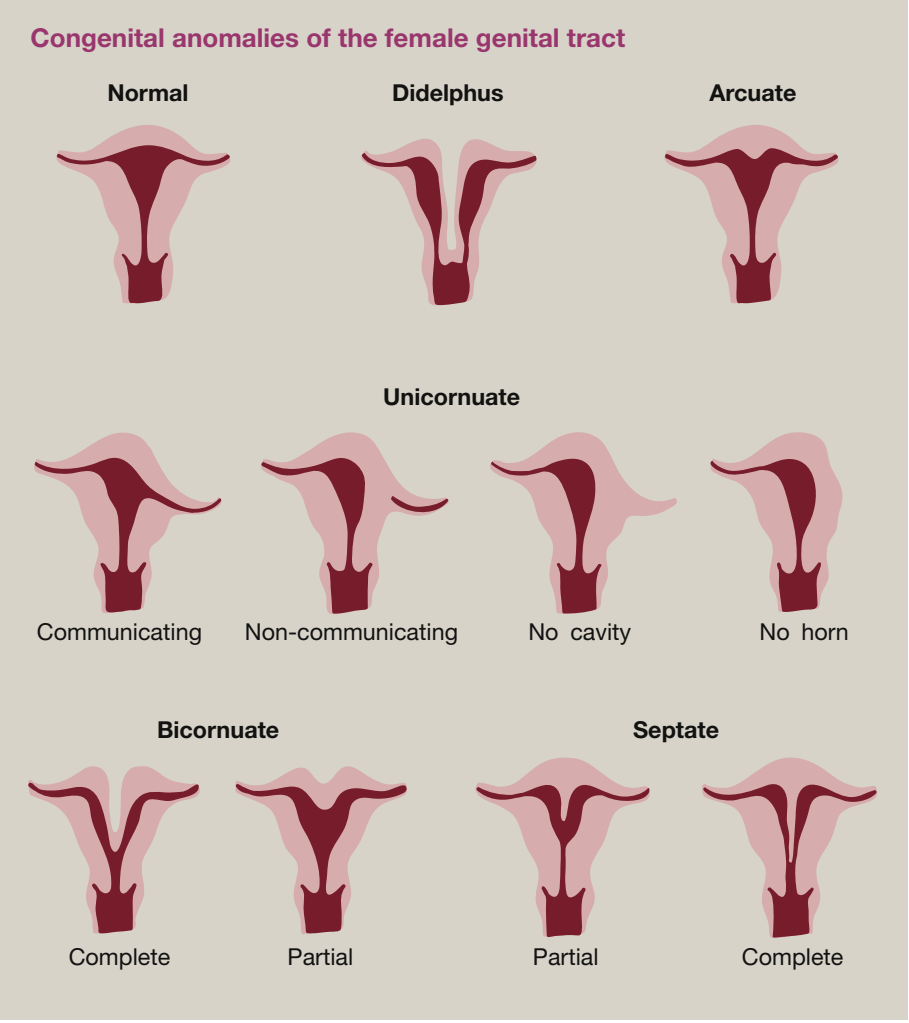

Figure 1

Laboratory testing

Biochemical laboratory testing in patients with unicornuate uterus and rudimentary horn is usually normal as this should not affect the normal ovulatory and menstrual cycle.

The primary imaging modality used is a pelvic ultrasound scan. Most sonographers who are experienced in adolescent gynaecology are able to identify the structural abnormality present through pelvic ultrasound. If there is doubt regarding the presence of these structural abnormalities then Magnetic Resonance Imaging (MRI) can be used. Hysterosalpingogram (HSG) or hysterosonography can be used to assess uterine abnormalities among sexually active women mostly those attending for infertility investigations.

Treatment

Patients with congenital genital outflow tract abnormalities are usually managed in a tertiary centre by a clinician with a special interest in adolescent gynaecology. Non-communicating uterine horns are removed surgically during adolescence and the unicornuate uterus is reconstructed. This will now almost exclusively be performed laparoscopically. This does not commonly affect fertility if the surgery is performed by a specialist with experience in this field. A subsequent pregnancy carries minimal risk of uterine rupture and the mode of delivery should be carefully discussed with the patient with a low threshold of proceeding to a caesarean section.

A communicating uterine horn that is detected prior to pregnancy should be removed electively due to the risk of uterine rupture during pregnancy; however if this abnormality is an incidental finding during pregnancy, patients are managed conservatively.

During pregnancy, patients with a unicornuate uterus should be managed by an obstetrician with experience in uterine abnormalities. The clinician must be aware that it is a high risk pregnancy and complications of pregnancy such as miscarriage, fetal demise, malpresentation and preterm delivery are more common.

One of the main factors of consideration with all genital outflow tract abnormalities is the psychological effect of the abnormality on the patient. Appropriate psychological support and counselling is central to their management.

Case 2: primary amenorrhoea

A 16-year-old girl presents with primary amenorrhoea. She has normal development of secondary sexual characteristics. Her

mother had menarche at the age of 13. Imaging revealed absence of a uterus on ultrasound scan.

Mayer–Rokitansky–Küster–Hauser syndrome

Mayer–Rokitansky–Küster–Hauser syndrome (MRKH), also known as Mullerian agenesis, is a syndrome characterized by agenesis or hypoplasia of the uterus and vagina. The aetiology is unknown – however it is thought to have autosomal dominant, polygenomic, multifactorial inheritance with variable penetrance. Familial clustering of the disease has been noted suggesting it is inherited.

This condition is associated with a rudimentary or absent uterus, and absent upper 2/3 of the vagina and a short blind ending vaginal dimple. Girls with MRKH present with primary amenorrhoea, however they will have normal pubertal development and secondary sexual characteristics prior to this. The incidence is approximately 1:5000 females. A large proportion of cases are isolated uterine and vaginal agenesis however 40% have associated renal tract abnormalities, and 12% having an absent kidney.

There are three main types of Mullerian agenesis:

Type 1: **Typical**

- isolated symmetrical uterovaginal aplasia/hypoplasia

Type 2: **Atypical**

- asymmetrical uterovaginal aplasia/hypoplasia
- the absence of one or both fallopian tubes
- malformation of the ovaries and/or renal system

Type 3: **MURCs (Mullerian duct aplasia, renal dysplasia, cervical somite anomalies)**

- uterovaginal aplasia
- Abnormalities of heart, skeletal system, muscular weakness &renal anomalies

The frequency of typical MRKH is 54.9%, atypical MRKH is 26% and MURCS 13.4%. MRKH syndrome is caused by partial or complete failure of the Mullerian system development.

Clinical findings

A thorough history and examination is pertinent to investigating these patients. It is important to take a thorough medical history including pubertal development, secondary sexual characteristics, menstrual history, sexual history, past medical history and family history.

Sensitive examination is highly important, particularly in adolescents who may be virgo-intacta. Examination in patients with MRKH will reveal normal secondary sexual characteristics and an absent or blind ending vagina. Clinicians must exclude imperforate hymen or transverse vaginal septum on examination as these can have a similar presentation. Abdominal palpation is important to exclude abdominal masses such as an enlarged uterus due to haematocolpos.

Another differential diagnosis of MRKH includes Complete Androgen Insensitivity Syndrome (CAIS) which presents similarly with a short, blind ending vagina, and primary amenorrhoea, normal external genitalia but absent axillary and pubic hair. Diagnosis of CAIS is confirmed by karyotyping as the patient will be phenotypically female with 46XY chromosomes (MRKH female has normal 46 XX chromosomes). In addition, pelvic imaging will show an absent uterus, and short vagina with rudimentary gonads. Diagnosis of CAIS can be excluded at examination if axillary and pubic hair is present.

Laboratory testing

The initial investigation of choice for MRKH is pelvic ultrasound. This will identify the majority of cases of uterine agenesis where the uterus is absent. Ultrasound is also beneficial in detecting renal abnormalities which may co-exist. Where a rudimentary uterus is present diagnosis may be more difficult. In this instance magnetic resonance imaging (MRI) may be more appropriate as this is the gold standard imaging modality for MRKH and can determine the degree of structural defect.

Additional testing should include endocrine profile and karyotyping. Patients with MRKH have a normal hormonal profile due to the presence of normal ovarian tissue and ovulatory function. If abnormalities are suspected of other organ systems these should be investigated and intravenous urography, spine x-rays, limb x-rays and echocardiogram may be required.

Treatment

Patients with MRKH are usually managed in tertiary centres by a gynaecologist with a specialist interest in adolescent gynaecology. There are many facets to the management of patients who are diagnosed with MRKH syndrome. Sensitive disclosure of the diagnosis is paramount as this can have a lasting impact on the patient. A diagnosis such as this will have an enormous psychological impact with respect to their psychological, social, reproductive and sexual function. It is therefore highly important that these patients receive extensive psychological support and counselling.

Most patients with MRKH have an absent or blind ending vagina, and as a result sexual function can be an issue. Most cases are managed non-surgically using graduated vaginal dilators for 20–30 minutes twice or three times daily. This involves passive dilatation of the vaginal dimple and patients can use local anaesthetic gel or simple oral analgesia. Treatment can be individualized and is usually undertaken when the patient is psychologically prepared and considering undertaking sexual intercourse. There is an 81–88% success with this mode of treatment – however evidence suggests that psychological preparation is vital and can vastly affect outcomes.

Surgery is an option if non-surgical methods fail or for patient choice, however to be successful still requires the patient to be able to perform vaginal dilatation. The most commonly practiced surgical options currently are the Vecchietti and Davydov procedures. The Vecchietti procedure is a laparoscopic procedure creating internal traction and dilatation on the vaginal dimple, which stretches and elongates a neo-vagina within 7–10 days. The Davydov procedure creates a neovagina laparoscopically from the patient's abdominal peritoneum.

Although sexual function can be restored and the patient has functioning ovaries, it has not been possible to create a uterus capable of pregnancy. In-vitro fertilisation (IVF) and surrogacy are a possibility – however transvaginal egg retrieval may not be possible due to the lack of vaginal length. In many cases egg retrieval is carried out trans-abdominally or occasionally laparoscopically. Alternatively patients could consider adoption.

Uterine transplantation has been attempted and successful pregnancy now reported. This is however still in its infancy and before this can be introduced more widely, several more carefully monitored pregnancies are required to evaluate major obstetrical risks, including miscarriage, pre-eclampsia, preterm birth, and fetal growth restriction.

Case 3: secondary amenorrhoea

A 14-year-old girl presents with secondary amenorrhoea. Her periods are irregular and have been since menarche aged 12 with a cycle interval of 45–60 days. Her BMI is 35 and has significant problems with hirsutism and acne. An ultrasound demonstrates appearances consistent with polycystic ovarian syndrome (PCOS).

PCOS

Polycystic ovarian syndrome (PCOS) is one of the most common endocrine disorders in women. It is categorised by a triad of menstrual cycle disturbances, hyperandrogenism and insulin resistance, causing a significant impact on many women of childbearing age. The prevalence of PCOS is thought to be around 10–15 % – however it is suspected that under-diagnosis is a significant issue, with much higher incidences when population screening has been undertaken.

PCOS causes many problems for women. Firstly as multiple immature ovarian follicles are produced, patients often do not ovulate regularly. This can result in dysfunctional uterine bleeding (DUB), most commonly oligomenorrhoea or amenorrhoea, as well as anovulatory infertility.

Insulin resistance and hyperinsulinaemia are a predominant feature in PCOS and can be seen in almost 50% of patients. Insulin resistance reduces the amount of sexhormone binding globulin, which usually circulates bound to testosterone. This will result in an increased level of free testosterone and as a result increase its peripheral action, resulting in hyperandrogenism. Therefore acne and hirsutism, which is characterized by male pattern distribution of terminal hair growth, are commonly experienced. Although the symptoms of PCOS are highly heterogenic, acne is a more common presenting complaint experienced by adolescents.

In addition patients with PCOS are at increased risk of obesity, Type 2 Diabetes Mellitus and cardiovascular disease due to altered lipid metabolism.

Clinical findings

It is important to take a detailed history to establish the onset of pubertal development, secondary sexual characteristics, menarche, characteristics of the menstrual cycle (including regularity, duration and interval), features of hirsutism, acne, virilisation and family history including a history of diabetes and PCOS.

Four of the common clinical features of PCOS are acne, hirsutism, virilisation and acanthosis nigricans. Acne is a common feature of adolescence and not specific to PCOS. Hirsutism is characterized by excess facial and body hair. Virilisation relates to 'masculinization' of their features and includes; increased muscle bulk, cliteromegaly, male pattern balding, decreased breast volume and deepening of the voice. These features are due to hyperandrogenism. Acanthosis nigricans refers to hyperpigmentation of skin in body folds caused by severe insulin resistance.

Laboratory testing

Diagnosis of PCOS presents many challenges within the adolescent population. The Rotterdam Criteria is currently largely accepted as the criteria for diagnosis of PCOS in the adult population -however as some of the signs and symptoms seen in PCOS are more common in adolescents, they can overlap with normal range of development, which poses a diagnostic challenge.

To fulfil the Rotterdam Criteria for diagnosis of PCOS in adults, patients must have two out of the three of the following:

1) Clinical or biochemical evidence of hyperandrogenism
2) Oligo-ovulation/Anovulation
3) Ultrasound evidence of polycystic ovaries (12 or more follicles 2–9 mm or an ovarian volume >10 ml)

Clinical evidence of hyperandrogenism would include acne, hirsutism and virilisation. Acne is one of the more common presenting complaints for PCOS within the adolescent population. Unfortunately acne is generally more common during adolescence, and so it may not be a good marker for hyperandrogenism. Hirsutism and virilisation are less common in adolescence and therefore may be a better marker for hyperandrogenism experienced in PCOS.

In addition, 65% of adolescents within the normal population experience menstrual cycle disturbances. Anovulatory cycles in this group are very common, particularly within the first two years following menarche and therefore amenorrhoea and oligomenorrhoea may also be a less sensitive markers for PCOS in adolescents.

There has been no agreement on how PCOS should be diagnosed in adolescents – however it has been proposed that Rotterdam Criteria could be used if all three criteria are fulfilled, and in addition it has been suggested that diagnosis should not be made until 2 years after menarche to allow for regular, ovulatory cycles to resume where possible. Where diagnosis is made careful follow-up is required.

It is important that clinicians exclude other conditions which may present in similar ways to PCOS. Non-classical, late onset Congenital Adrenal Hyperplasia (CAH) can be excluded by taking a basal morning 17-hydroxyprogesterone blood test to exclude 21-hydroxylase deficient non-classical adrenal hyperplasia. It is important to exclude Cushing's by taking a dexamethasone suppression test and exogenous androgens to check the free androgen index and serum levels of DHEAs. In addition patients with a testosterone >5 nmol/l will require further investigation to exclude adrenal tumours.

Treatment

NICE have issued guidance on the management of PCOS in adolescents. Adolescents should be screened for impaired glucose tolerance and type 2 diabetes as well as cardiovascular risk factors. Their emotional wellbeing should be assessed. Obesity is a common problem among patients diagnosed with PCOS, and many of the signs and symptoms of PCOS including infertility, menstrual cycle disturbances and adverse effects such as type 2 diabetes mellitus improve with weight loss and lifestyle changes. Diet, exercise and lifestyle advice are therefore the first line management of PCOS.

Metformin can be used to treat insulin resistance, resulting in increased insulin sensitivity as well as decreased LH and testosterone levels. In the long term this may help to restore fertility, correct menstrual cycles, reduce hirsutism and supplement weight loss. Around 70% of patients who had previously normal menstrual cycles prior to diagnosis with PCOS resumed normal menstrual cycles following treatment with metformin.

Hirsutism can be managed in a variety of ways: there are many options for hair removal including electrolysis, laser hair removal, waxing and bleaching, as well as antiandrogens such as Dianette (cyproterone acetate) or Spironolactone. Dianette has a higher risk of thromboembolism than other contraceptives hence duration of use should be limited, and should be stopped 3 months after an improvement in symptoms have been noted.

Patients suffering from menstrual cycle problems may benefit from cyclical hormone therapy such as the combined oral contraceptive pill (COCP) or progesterone only pill (POP) to regulate their menstrual pattern. Many adolescents are already taking oral contraceptive pills and this may be another reason that PCOS is underdiagnosed.

Oligomenorrhoea and amenorrhoea increases the risk of endometrial cancer due to unopposed oestrogen stimulation on the endometrium. It is important that all patients who are of child bearing age menstruate every 3 months to protect their endometrium. For those who are oligomenorrhoeic or amenorrhoeic it is important to induce a withdrawal bleed by commencing the oral contraceptive pill or with medroxyprogesterone 10 mg twice a day for 7 days every 3 months, in order to reduce the risk of endometrial hyperplasia and subsequent malignancy.

Case 4: adnexal masses

A 15-year-old girl present with mild abdominal pain. An ultrasound scan demonstrates bilateral complex ovarian cysts of greater than 8 cm on both ovaries. Tumour markers are normal and a subsequent CT scan demonstrates heterogeneous adnexal masses with prominent solid components, calcification and foci of fat on both ovaries. There is no evidence of torsion.

Adnexal masses

Ovarian cysts are a relatively common finding, particularly among women of reproductive age. The majority of these cysts are simple, functional cysts which resolve over the forthcoming 2–3 menstrual cycles. The type of cyst seen in patients is generally different depending upon their age at presentation. The most common cysts experienced during reproductive years are simple cysts related to ovulation of an oocyte from an ovarian follicle. In childhood it is more common to see germ cell tumours of the ovary. The incidence of adnexal masses in the under 18 year olds has been estimated at 2.6 per 100,000 with 10% of these being non ovarian in origin. About 10% of ovarian masses in this age group are malignant. Overall ovarian malignancy is particularly uncommon prior to the menopause, with 0.4–8.9/100,000 risk of malignancy in the reproductive age group.

Dermoid cysts, also known as mature teratomas are usually benign cysts that originate from totipotent germ cells (primordial follicles) found in the ovary. They are the most common benign tumour in girls less than 20 years of age. These cells have the potential to become any type of mature tissue from throughout the body. As a result dermoid cysts contain many different types of tissue commonly including, hair, skin, nails, bone, teeth, sebum and thyroid tissue. 15% of dermoid cysts are bilateral and the average age at detection is 30, which may be because many of these are identified incidentally during pregnancy. Dermoid cysts are almost always benign, however occasionally malignancy occurs, most commonly due to an endodermal sinus tumour in childhood or a squamous cell carcinoma in adults. It is important to exclude malignancy in these patients – however regardless of this the cysts usually require removal due to their size or the potential for them to cause ovarian cyst accidents, in particular ovarian torsion.

Malignant immature teratomas comprise less than 1% of ovarian teratomas and 10–20% of malignant tumours in this age range. Germ cell tumours can be associated with elevations in tumour markers including serum a-fetoprotein (a-FP) (33–65% of patients with malignant immature teratoma) and b-human chorionic gonadotropin (b-hCG) levels. About 26% of affected patients may have associated ipsilateral mature cystic teratoma and 10% contralateral immature teratoma. Malignant immature teratomas are diagnosed most frequently between the ages of 10 and 20 years, with a median age of diagnosis at 17 years.

Due to the contents of dermoid cysts, they are heavy. As a result they are prone to twisting on the ovarian pedicle, which is known as torsion. This can compromise the blood supply to the ovary and risk the integrity of the ovarian function. This, like testicular torsion, is a medical emergency which require surgical correction, as a delay in diagnosis can result in loss of the ovary. Torsion is particularly common in dermoid cysts and cysts over 5 cm diameter.

Clinical findings

Dermoid cysts are often identified incidentally on a pelvic ultrasound. In the absence of an ovarian cyst accident, these cysts are commonly asymptomatic and many are detected in pregnancy.

A detailed history is important to management. It is important with any cysts to identify any risk factors such as family history of breast and ovarian cancer, history of abdominal distension, appetite suppression, abdominal pain and urinary symptoms such as frequency and urgency.

Examination, both abdominal and vaginal may reveal any palpable masses, tenderness and local lymphadenopathy. Identification of pain could suggest a diagnosis of ovarian cyst accident including torsion, rupture or haemorrhage.

The presentation of ovarian torsion can be difficult to predict. The most common presentation is intermittent, waxing and waning pain along with nausea and vomiting. 85% of patients with ovarian torsion present with a combination of these symptoms. Other common clinical findings are a low grade pyrexia and sinus tachycardia.

Laboratory testing

Pelvic ultrasound scan (USS) is the most superior tool for identifying ovarian cysts. Since the advent of pelvic USS, many more ovarian cysts are being identified. Dermoid cysts usually appear to have solid components on USS and have hyper and hypoechoic areas.

Ovarian torsion may be difficult to identify on USS – features such as unilateral ovarian enlargement or ovarian oedema with peripherally arranged follicles can be seen. Occasionally a solid mass with hyperechoic and hypoechoic areas are identified suggesting haemorrhage and necrosis. The twisted pedicle in torsion may appear like a whirlpool and there could be abnormal doppler flow due to the reduction or complete obstruction of the flow of blood into the ovary.

Magnetic resonance imaging (MRI) is not routinely used in ovarian cyst management — however it can be used to evaluate complex masses for features suggestive of malignancy.

In young patients under 40 years of age presenting with complex ovarian cysts, various tumour markers are required to exclude a germ cell tumour of the ovary. These include lactate dehydrogenase (LDH), bHCG and alpha-fetoprotein (αFP). Ca125 should also be taken in complex cysts, however if the cyst appears to be simple it is of little benefit. It is particularly unreliable due to the number of conditions which cause false positive results including; fibroids, endometriosis, adenomyosis, and pelvic infections therefore it must be interpreted cautiously.

Risk of malignancy index (RMI) or IOTA rules can be used to identify those cysts that are at increased risk of being malignant. Patients with a high RMI must be referred to gynaecological oncology MDT for discussion. Unfortunately, RMI does not take into account tumour markers other than Ca125 and therefore may be less applicable to patients in a younger cohort.

Treatment

Dermoid cysts are usually managed surgically if they are over 5 cm in size due to the risk of ovarian cyst accident. This may be via laparoscopic ovarian cystectomy or via laparotomy. Most benign cysts are able to be removed laparoscopically and this is the treatment of choice due to the faster recovery, reduced postoperative pain and shorter hospital admission. Unfortunately, due to the solid nature of dermoid cysts, this often is not possible as the remaining cyst contents are too large (even when aspirated) to be removed through a laparoscopic port site. Spillage of dermoid cyst contents is associated with 0.2% risk of chemical peritonitis. It is therefore important to thoroughly lavage the peritoneal cavity with large amounts of warmed fluid. Warm fluid avoids the patient becoming hypothermic and it helps to dissolve the fat rich contents of the cyst which may solidify in colder environments making removal challenging.

Given the risk of these benign tumours being bilateral it is important to conserve ovarian tissue where possible in adolescents to preserve fertility. It is not usually possible to harvest eggs prior to removal of the dermoids as this carries the risk of spillage of contents and makes it almost impossible to access normal ovarian tissue. Surgery carries the risk of reduced ovarian reserve and patients should be warned of this preoperatively and before removal of the bilateral dermoids.

Simple ovarian cysts <5 cm in premenopausal women can be managed conservatively with no follow-up required. Between 5 and 7 cm these cysts should be scanned in 1 year to ensure they are not growing. Simple cysts larger than 7 cm may require an MRI or surgical intervention.

In patients presenting with torsion, management is usually via urgent surgical de-torsion and aspiration of the cyst contents. Research has shown this to be the most superior form of management because although ovaries may appear necrotic at first inspection, in the majority of cases a proportion of ovarian function will resume. Previously it has been known for clinicians to perform a partial or total oophorectomy for these cases — however this practice is becoming less popular and is to be avoided due to the impact on fertility. Laparoscopic drainage may increase the risk of recurrence, and as a result it may be pertinent to follow patients up to determine if laparoscopic cystectomy is required at a later date.

Summary

Development and function of the reproductive system can be affected in many ways. The cases summarised characterize the diversity of presentations seen by adolescent gynaecologists and touch on the in-depth understanding of embryology required to manage such patients. Diagnosis is complex and multifaceted and these conditions may have far reaching implications for future lives. It is important for obstetricians and gynaecologists to have an understanding of these patients due to the implications they may have on their reproductive function, pregnancy and gynaecological care. Multidisciplinary care is vital and psychological input is paramount to managing these conditions successfully. ◆

FURTHER READING

Balen A. Polycystic ovary syndrome (PCOS). *Obstet Gynaecol* 2017; **19:** 119–29.

Damigos E, Johns J, Ross J. An update on the diagnosis and management of ovarian torsion. *Obstet Gynaecol* 2012; **14:** 229–36.

Edmonds DK, Rose GL. Outflow tract disorders of the female genital tract. *Obstet Gynaecol* 2013; **15:** 11–7.

Hickey M, Lester S. The management of menstrual disorders in early reproductive life. *Obstet Gynaecol* 2003; **5:** 136–41.

Valappil S, Chetan U, Wood N, Garden A. Mayer–Rokitansky–Kuster–Hauser syndrome: diagnosis and management. *Obstetr Gynaecol* 2012; **14:** 93–8.

Practice points

- Paediatric and adolescent gynaecology is complex and often requires input from a clinician with a specialist interest in the paediatric and adolescent gynaecology.
- Mullerian agenesis presents with primary amenorrhoea and normal secondary sexual characteristics. It is usually diagnosed on pelvic ultrasound.
- Polycystic ovarian syndrome is very common in adolescence and adulthood however it is often under-diagnosed.
- Dermoid cysts are the most common benign tumours found in girls under 20 years of age. Large dermoid cysts are at higher risk of ovarian torsion.