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Congenital malformations of the genital tract and their management

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While congenital malformations of the genital tract are not common, the sequelae of their presence can be serious. The practising gynaecologist must be aware of the range of congenital abnormalities that may occur and the symptoms that may result from them. Failure to manage these patients correctly may have long-term sequelae for their psychological, sexual and reproductive health. The involvement of a multi-disciplinary team in dealing with these patients is imperative, and preparation for surgery—particularly in congenital malformations of the vulva and the vagina—is imperative if the long-term sexual function in these patients is to be fulfilled. Surgical correction of vulval abnormalities in adolescence is related solely to sexual function as most of the reconstructive surgery is done in childhood. For the management of Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome, the recommendation is now that passive dilatation by Franks' technique is the treatment of first choice and only if that fails should surgical approaches be embarked upon. The results of the surgery are similar in all techniques and the particular surgical centre will have its own preference of which technique it adopts. Congenital absence of the cervix is a complex surgical problem and should be dealt with solely in centres with expertise.

Key words: vulval abnormalities; vaginal anomalies; vaginal obstruction; congenital absence of the vagina; cervical atresia; haematocolpos; haematometra.

A number of congenital malformations may not manifest themselves until adolescence and some congenital malformations which have been present at birth or in childhood may remain problems throughout the adolescent years, particularly following the onset of puberty. The genetics and aetiology of these disorders have been referred to extensively in the previous chapter, and the management of these disorders in adolescence is a complex, multi-disciplinary problem. Not only does the team caring for the adolescent need to address the anatomical difficulties but there are functional, reproductive and psychological issues which also need attention—and very often by health professionals with particular expertise in these areas, especially that related to the psychological health of these young women.

In addressing the various malformations and abnormalities, the conditions have been divided into various sections for ease of discussion.

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CLINICAL ASSESSMENT

In considering the malformations, some universal principles of clinical management need to be employed. It is imperative that the correct diagnosis of the underlying conditions be made and also that patients are investigated fully—detailing not only their genital anatomy and its variance from normal, but also carrying out appropriate endocrine and genetic investigations. A large number of these congenital malformations have associated disorders of other organ systems, which also need to be delineated and defined. This may involve a number of forms of imaging, depending on the most appropriate to determine the abnormality. Finally, an extremely careful assessment of the psychological status of the patient is imperative. The risks of not involving the psychologists in the management of these patients may cause long-term irreparable damage. Adolescents are very difficult to assess psychologically and intellectually because intellectual development varies from patient to patient. There is a complete discrepancy between physical maturity and intellectual maturity, and while physical maturity may be achieved by the age of 15 or 16, intellectual maturity is not reached until 18–20. This discordance presents a major problem for management as a number of the malformations that we will be discussing have physical symptoms, which demand surgical attention at a younger age than would be ideally desirable in terms of being able to cope with the difficulties that these young women encounter. The risks of ignoring the psychological aspects of these disorders are long-term sexual dysfunction and the inability to integrate into normal society. So profound are these issues that feelings of heterosexuality may mean that these patients have difficulty sustaining relationships due to the psychological harm that has been ignored in early adolescent years. However, the psychological difficulties of adolescence make assessment much more complex than in childhood or adult life and therefore specially trained psychological counsellors are needed for all units carrying out this type of reproductive surgery.

DISORDERS OF THE VULVA

In adolescence, there are three situations which arise with the vulva which may require the attention of the reconstructive gynaecologist/urologist. These are (1) patients with a congenital adrenal hyperplasia (CAH) who have had surgery performed in childhood and who, having progressed through puberty, have ongoing physical problems, (2) patients with late-onset CAH in whom clitoromegaly may result, and (3) patients with androgen-secreting virulizing tumours of the ovary who may also get some degree of clitoromegaly.

Congenital adrenal hyperplasia

The management of CAH currently involves the majority of surgery being performed in infancy. The objective of reconstructive procedures is to delay the surgery until such time as the anomalous structures have reached a size that permits effective surgery, and yet to have completed the procedure before the abnormalities present become embarrassing and alarming to the patient or her family. The philosophy that proceeding as early as possible has psychological advantages has recently been challenged.[1] Most of the vulvo-vaginal surgery is carried out as a one-step procedure in an attempt to create an anatomically acceptable and functional vulva and vagina, which will grow with

the child through adolescence into adulthood. However, the results of the revision of external genitalia in childhood when followed-up in adolescence and adulthood indicate that the surgery is far from successful in the majority of patients. In the study of Azziz et al, 79% of patients required further vaginal reconstructive surgery as an adolescent or adult to allow comfortable intercourse and 25% had undergone more than one procedure before puberty. Of all patients having repeat vaginal reconstructive surgery, 72% were successful with their second attempt and these authors recommended that exteriorization of the vagina be postponed until puberty or thereafter when the young woman has reached a sufficient level of maturity to comply with the use of post-operative dilators. Krege et al[2] reported similar findings in 27 patients who underwent surgery for CAH. In 20 patients the procedure was a one-stage procedure and in five it was a two-stage procedure. Some 36% of patients developed intravaginal stenosis and all of these were in the single-stage procedure group. However, 14 out of 16 patients who were followed-up in the long term had overall problems with their body image, vaginal stenosis, anxiety about sexual intercourse and problems with orgasm. This group also concluded that this type of surgery should be delayed until the beginning of puberty or subsequently for the same reasons as Azziz et al. In a retrospective study by Creighton et al[3], cosmetic results were judged to be poor in 41% of 44 patients who were studied, and 98% needed further treatment to the genitalia for cosmesis, tampon use or intercourse. Some 89% of one-stage procedures required further surgery and the larger studies available so far have brought similar results. The timing of genital surgery is obviously a difficult and controversial issue but these results highlight the need for appropriate debate.

In adolescence, secondary surgery may well be required on the vaginal outlet. The stenosis usually results from a failure of the original surgery to carry the midline incision far enough posteriorly and a second procedure in adolescence may be required to complete this. Enlargement of the vaginal orifice may require several approaches which include a Fenton's procedure or the use of skin flaps which may be swung from the labia—or there may still be perineal skin available in some cases to allow this to be used as a flap. Labial tissue expanders have been used in these circumstances to try to reduce the risk of re-stenosis.[4]

Late-onset congenital adrenal hyperplasia

This uncommon endocrine condition occurs during puberty when, following adrenarche, excessive androgen is produced by the adrenal. This impacts on clitoral growth and therefore may lead to a presentation of clitoromegaly in adolescence. These patients are universally extremely embarrassed by the size of the clitoris, and its prominence when wearing swimming or sporting clothing leads to extreme dissatisfaction with their physical appearance. However, in early adolescence, little understanding of the functional part of the clitoris in terms of sexual satisfaction is evident to the patient and therefore great care and counselling has to occur before any decision is made to correct the clitoromegaly.

The clitoromegaly associated with CAH is usually dealt with in infancy and the original techniques involved amputation of the clitoris, leaving a non-functioning clitoral stump. By the middle 1970s, questions were asked about the sense of this approach and the development of the reduction clitoroplasty, which involves the partial resection of the corpora and the glands of the clitoris re-anastomosed to the stump with retention of the neurovascular bundle, has become very popular. The theory behind this approach was that when the children reached adolescence and beyond, there would be

retention of the sensory part for sexual enjoyment. Long-term follow-up of 42 women at a mean of 24 years post-repair showed excellent functional success and, although there were limited data available, sexual function was satisfactory in 61% of cases.[1]

However, it is difficult to believe that any surgical procedure which leads to complete or partial resection of the clitoris can be successful in leaving this individual with normal clitoral function, and while there may be some sensation in the glands, this is difficult to define. Retention of the clitoris with its glands must be the ideal. Surgical reconstruction with recession of the entire clitoris beneath the symphysis pubis has been achieved by Randolph and Hung[5] and a long-term follow-up study published by Newman et al[6] gave highly satisfactory anatomical results; all but one of the nine women who were followed-up in the long term achieved regular, painless orgasm, which was equivalent to normal clitoral orgasm in women without clitoromegaly. This approach of clitoral recession with retention of the clitoris in its entirety should be considered in all women with late-onset CAH, and it is an approach that really ought to be considered in the paediatric patient also. The results reported so far are extremely encouraging and perhaps this should be considered the approach of choice.

In all patients presenting with clitoromegaly in adolescence, consideration of androgen-secreting tumours of the ovary must be borne in mind and appropriate endocrine screening should be carried out.

DISORDERS OF THE VAGINA

Congenital malformations of the vagina fall into three categories: remnant cyst, obstructive outflow tract disorders and congenital absence of the vagina.

Cysts of the vagina

The majority of vaginal cysts are located on the lateral or posterior walls of the vagina. Most patients complain of swelling in the vagina, and in adolescence they may also complain that they cannot insert a tampon at period time or, in those teenagers who are sexually active, they may have dyspareunia. The origin of these vaginal cysts histologically indicates that the majority are Müllerian remnant cysts or epidermal inclusion cysts, while cysts of Gartner's duct are much less common than generally believed (Table 1).

Management of vaginal cysts in general should be by excision. However, high, large Müllerian duct cysts which contain clear mucous are best treated by marsupialization. Attempts to excise these deep cysts totally may lead to unexpectedly profuse bleeding or damage to the ureter which may occur as a result of the placement of deep sutures to arrest the bleeding.

Table 1. Vulval cysts.

Author	Müllerian remnant cyst (%)	Epidermal inclusion cyst (%)	Cyst of Gartner's duct (%)	Unknown origin (%)
Deppisch[64]	33	45	12	10
Pradhan and Toban[65]	56	20	15	9

Obstructive outflow tract disorders of the vagina

The embryological development of the vagina may lead to a number of conditions which result in obstruction to the flow of menstrual blood once menarche occurs. All of these conditions present with primary amenorrhoea in association with increasing dysmenorrhoea as subsequent cycles unfold. Menstrual loss does not occur but the cyclicity of the pain is diagnostic. Management of these cases is dependent on the level of obstruction, and the anatomical and functional success becomes increasingly poor as the obstruction becomes increasingly high.

Imperforate hymen

The hymen is a thin, mucous membrane which occurs at the junction of the sinovaginal bulbs with the urogenital sinus and is usually perforated during fetal life—although the mechanism involved in achieving this remains unknown. Failure to develop a perforation leads to the membrane remaining intact, resulting in the imperforate hymen. As puberty unfolds, menstrual blood collects behind the membrane and the vagina begins to distend. This is often painless for some months and eventually the vagina becomes greatly distended and a haematocolpos results. When the mass becomes sufficiently large, it may affect micturition and defaecation and even overflow incontinence may occur. Clinically, a mass arising from the pelvis is palpable abdominally and inspection of the vulva by separation of the labia will reveal a membrane through which menstrual blood may be seen and which appears as a dark blue mass. This appearance is quite unlike the appearance of a transverse vaginal septum (see later) in which the bulging membrane remains entirely pink.

The surgical management of this condition involves a cruciate incision in the membrane from 2 o'clock to 8 o'clock and 10 o'clock to 4 o'clock. The remaining quadrants of hymen may be excised or they may be left in situ. No attempt to evacuate the vagina at the time of surgery should be made, as there is a risk of perforation of the vagina with the introduction of instruments and the risk of ascending infection through the introduction of swabs into the haematocolpos. In general, drainage is complete within 3–5 days and thereafter there should be no further problems.

There are generally no sequelae following imperforate hymen and its treatment; reproductive performance subsequently compares equally with that of the normal population.[7]

Transverse vaginal septa

The incidence of this phenomenon is unclear but it is much less common than congenital absence of the vagina and uterus. The developmental defect in this situation is one that results from incomplete fusion between the Müllerian duct component of the vagina and the urogenital sinus component. This incomplete vertical fusion leaves a transverse vaginal septum that varies in both the level and thickness. It would seem that these septa can be classified into those that occur in the upper vagina, the mid-vagina and the lower vagina with incidences of 46% occurring in the upper, 35–40% in the mid and 15–20% in the lower.[8] The observations on the degree of absence of the vagina show that greater absence occurs the higher the septal defect. It may be associated with other congenital malformations of the urological tract or rectum, and anus and uterine

abnormalities, for example, bicornuate and septate uteri, are also described in association with this.

Presenting symptoms are similar to those of imperforate hymen, with increasing cyclical abdominal pain in the absence of menstruation at the time of puberty. It is often the case that the adolescent's diagnosis may be missed for several months. Only when the adolescent is admitted as an emergency with acute abdominal pain and/or urinary retention is the diagnosis made and clinically at this time there is a pelvic mass arising from the pelvis, which may extend into the abdomen. The mass is tender to palpation, and ultrasound imaging will confirm the presence of a haematocolpos and a haematometra.

Occasionally, the presence of a haematosalpinx may be seen or, more rarely, the presence of an endometrioma.

Surgical management

The principle of the management of transverse vaginal septae is to excise the septal defect and perform a vaginal end-to-end anastomosis between the upper portion of the vagina and the lower. A transverse incision is made in the centre of the vault of the lower short vagina, and a passage is made which will eventually allow incision into the upper vagina where the haematocolpos will be apparent. The dissection laterally then allows excision of the septal tissue and this has to be completed in its entirety. The upper vaginal wall may need to be mobilized in order to bring it into apposition to the lower vaginal portion and an end-to-end anastomosis is usually possible in these circumstances, leaving a vagina that is normal in calibre; it is the author's preference to place a firm vaginal mould at the site of the anastomosis for 10 days post-operatively in order to try to prevent stenosis occurring subsequently. The use of vaginal dilators for 2–3 months subsequent to the removal of the mould will result in excellent functional results, particularly for the lower and middle septal defects. The approach for the higher septal defects, which have a greater absence of the vagina, may require an abdominal approach as well as a vaginal one. The upper vaginal portion may be quite short and there is the risk of damage to the bladder or the rectum in trying to enter this through the vagina, rather than exploring abdominally and placing a probe into the upper vagina to act as a guide for the vaginal surgeon to find the appropriate point of entry. Again, if possible, an end-to-end anastomosis should be attempted but there are a significant number of cases where the length of absent vagina precludes this approach. Here, a mould needs to be placed with a central drainage passage to allow menstrual blood to escape and the mould should be left in situ for 3–6 months to allow epithelialization to occur between the two portions of vagina. This process may be promoted by the use of amnion which may be placed around the mould and which encourages epithelialization. Subsequent to the removal of the mould, again, the use of vaginal dilators is imperative in order to ensure that the vagina remains functionally patent. In some cases of high vaginal septum, when drainage of the upper haematocolpos has been performed prior to referral for reconstructive surgery, the use of vaginal dilators to extend the lower vagina as much as possible can be a useful approach. Here, suppression of menstruation will allow time for the use of the dilators and the expansion of the lower vagina. This may then allow primary anastomosis to occur at the time of surgery, thereby offering a greater chance of success and reducing the incidence of stenosis. It is the author's belief that this approach is the best one to try to achieve the results that we seek. The difficulty with which we are often faced with these cases is the high vaginal septum in the girl who is only 12 or 13 years of age. The

use of dilators in these circumstances is often unsuccessful and therefore suppression of menstruation may be carried out for some 2–3 years in order that puberty can progress, and that the patient becomes more mature and is able to cope with the idea of the use of vaginal dilators prior to the surgery being performed. Suppression in these circumstances is usually with the oral contraceptive pill continually, and only if this is unsuccessful does surgery need to be attempted earlier.

In the author's own experience of 27 cases of lower third obstruction, sexual function in the follow-up of those patients has been 100% successful. In 12 cases of a middle third obstruction, eight patients have been found to have a normal sexual function and four patients have had some degree of partial stenosis which has required revision; one patient remains with some degree of dyspareunia in spite of attempts at revision. In 11 cases of high vaginal obstruction, seven patients have satisfactory sexual function. Of interest, however, is that the incidence of high stenosis remains high with eight patients having an anatomical stenosis, although only three of the eight found this to be a cause of dyspareunia. Similar results are found in numerous case reports in the literature.

In the only follow-up study of pregnancy after vaginal obstruction, Rock et al[8] reported a 100% pregnancy success rate in patients with a lower third obstruction, 40% success with a middle third obstruction and only 20% success with an upper third obstruction. The likely explanation for this is the incidence of endometriosis, which is much higher with the higher obstructive disorders. Here, prolonged retrograde menstruation may lead to architectural damage of the pelvis, which may result in anatomical infertility. In vitro fertilization (IVF) in these circumstances may be very successful but there are no data specifically related to these patients.

Longitudinal vaginal septum

If the two lateral Müllerian ducts fail to fuse at their lower border, uterine and vaginal anomalies may result. The failure of fusion results in a uterus didelphus with two hemi-uteri and two hemi-cervices. Each cervix creates a hemi-vagina and, in most cases, these two vaginas fuse with the urogenital sinus to give the clinical result of a double vagina. The septum that divides these vaginas may be partial or complete, and patients usually present either with difficulty with inserting tampons or the insertion of a tampon fails to stem the menstrual flow as only one hemi-vagina is protected. However, it is not uncommon for patients to fail to notice the hemi-vagina effect and to present with either dyspareunia or, when they are already pregnant, with an incidental finding during the pregnancy. Some patients will find that one of the hemi-vaginas is larger than the other, usually the right versus the left, and they are able to have intercourse in the right hemi-vagina without difficulty. However, this may be associated with infertility because sperm are then able to get into only one hemi-uterus. Excision of the vaginal septum is advisable in order to improve the chances of pregnancy in those women who are trying to conceive and also to avoid difficulties which may arise during vaginal childbirth. Excision is straightforward, but caution should be exercised and care taken to ensure that the septum is removed in its entirety, not leaving a band of vaginal tissue between the two cervices, which may, in itself, cause dyspareunia. The traditional method for removing a longitudinal vaginal septum is by excision and ligation—although the use of laser may in time bring better anatomical and functional results.

In some cases, one of the hemi-vaginas may fail to cannulate and reach the urogenital sinus. Here, a blind vaginal cavity exists and at the time of puberty, when menstruation begins, menses from the unobstructed vagina are found to flow

normally whereas the obstructed hemi-vagina results in an accumulation of menstrual fluid. This very confusing clinical situation is often late diagnosed, as the presence of menses does not alert the clinician to the possibility of an obstructed hemi-vagina. Only when the dysmenorrhoea resulting from this becomes quite severe, and the patient is admitted as an emergency, is the diagnosis made on imaging; this is clearly possible to delineate with real-time ultrasound. It is imperative that screening for renal agenesis is undertaken in these patients as they often have ipsilateral renal agenesis. Surgical management of these conditions involves careful excision of the vaginal septum in its entirety, and this is carried out per vagina, but care has to be taken in excising this type of septum because it may be very thick. It is simple to find post-operative haemorrhage as a complication if care is not taken. Also, if excision is not carried out—simply an attempt made to create an ostium through which the menses can flow—bacteria from that hemi-vagina which is functioning normally will enter the previously blind hemi-vagina and an abscess may result. This is an extremely serious situation, which may result in ascending infection, septicaemia and a life-threatening episode for the girl. It is imperative, therefore, that these procedures be carried out by surgeons whose skills allow them to do this surgery effectively and that the operation performed should be the first operation—which should be curative rather than a sequence of operations, which is ill-advised. The results of the surgery are excellent and retraction of the septal pedicles is almost complete. Dyspareunia is rarely a problem and reproductive performance in this group of patients is comparative to patients with solely a uterus didelphus.

CONGENITAL ABSENCE OF THE UTERUS [MAYER–ROKITANSKY–KÜSTER–HAUSER SYNDROME (MRKH)]

Complete vaginal agenesis in association with uterine agenesis or hypoplasia is uncommon and is known as the MRKH. The incidence of this syndrome is 1 in 5000.[9] These patients present as adolescents with primary amenorrhoea in the presence of normal secondary sexual characteristics other than menarche. Their ovarian function is normal and their karyotype is 46,XX.

Genetics of MRKH

MRKH is not usually found in a familial pattern but it is found quite commonly in conjunction with other malformation syndromes.[10] Although affected siblings have been documented with vaginal agenesis, discordant monozygotic twins have also been reported.[11] This indicates that a single autosomal recessive gene cannot be the explanation for this condition. Although Shokeir[12] proposed an autosomal dominant inheritance from a study of 16 families in Saskatchewan, subsequent studies have failed to confirm this.[13,14] It would therefore seem that the most logical explanation is a polygenic multi-factorial inheritance and these inherited abnormalities normally have a recurrence risk of between 1 and 2%. In the offspring reported by Petrozza et al.,[14] none of the female offspring have exhibited Müllerian aplasia.

Molecular genetic studies have so far failed to attract any candidate genes to explain this phenomenon. While a number of studies have looked at the possibility of defects in the homeobox series, as yet no abnormality has been found.

Diagnosis

As alluded to above, patients with MRKH present with primary amenorrhoea, and investigation by imaging reveals that there are two forms of the MRKH syndrome. In the first group, the only abnormality is congenital absence of the vagina and uterus, while in the second group, congenital absence of the vagina and uterus is also associated with renal, ear and skeletal abnormalities.[15] With regard to skeletal abnormalities, it is well known that these occur in some 10–12% of patients.[16,17] The incidence of hearing difficulties in MRKH patients has been reported by Strubbe et al[18], and a hearing loss of some degree was attributed to a congenital origin in 10% of MRKH patients. These findings illustrate the need to evaluate MRKH patients a little more thoroughly for associated abnormalities than perhaps has been previously practised.

Management

The management of these patients falls into two distinct areas: (i) the management of their congenital anatomical abnormality with the need to be sexually active, and (ii) the psychological impact of the knowledge that these individuals have no vagina or uterus.

Psychological aspects of Müllerian aplasia

It is difficult to quantify the emotional trauma which is associated with the knowledge that the patient has Müllerian aplasia. There are at least three parties involved in this process: the patient herself, who may be a young adolescent, aged 14 or 15, and her parents. The shock is considerable to all, and the first reference to the need for psychotherapy is cited in 1968 by Kaplan on the basis of nine patients with MRKH syndrome.[19] He describes the importance of the impact of a physician's lack of appreciation of the difficulties that may ensue and how this inability to communicate may make the psychological effects much worse. He further develops his hypothesis of the need for psychological input in 1970 when he describes a series of cases outlining the great difficulties that these patients have.[20] Poland and Evans[21] studied 54 patients and described the emotional reactions to the diagnosis and treatment—which varied with the age of the patient and her relationship both with her parents and with a heterosexual partner. Many patients were initially depressed, questioning themselves about their gender and their ability to fulfil the female role in the future as an adult. However, rather than the expected difficulty with sexual intercourse, it was infertility that was the most difficult part for these young women to accept. In having instituted a policy of prolonged counselling, they showed that emotional reaction and reinforcement were vital parts of the medical management of these patients but the input of psychological services was fundamental in improving the outcome of therapy. In the last 10 years there has been an evolution in the multi-disciplinary approach to the management of MRKH syndrome, and the importance of psychological support has been emphasized to an even greater extent.[22] The evaluation of group therapy, which has been ongoing in a number of centres for some years, was first reported by Weijenborg and ter Kuile[23] and demonstrated the invaluable use of group programmes for helping MRKH syndrome patients to deal with their psychological stress. It is impossible to emphasize too greatly the importance of psychological input into the preparation of patients, not only for the adaptation to their congenital abnormality and the ability to deal with this currently and

in later life, but also in the preparation of the patient for whichever therapy is chosen to help her to achieve a functional vagina. Without adequate psychological assessment, and the involvement of the psychological or the clinical nurse specialist in the timing of treatment, the chances of success are vastly reduced.

Non-surgical management of Müllerian agenesis

While as yet it is not possible to create a new uterus for these individuals, a number of techniques have been described for the creation of a vagina. A recent policy statement by the American College of Obstetricians and Gynecologists emphasizes the primary role of the non-surgical approach—with vaginal dilators being the treatment of first choice.[24] The use of vaginal dilators was first reported by Amussat in 1835.[25] Amussat's technique was to use strong digital pressure on the vaginal dimple over a series of sessions, but in 1938 Frank modified this method by using pyrex tubes.[26] Frank described six cases in his paper, only one of which was not successful but, interestingly, for almost 40 years this technique was only infrequently used. Rock et al. reported success with the use of dilators in only 40% of patients[8] and this report further reinforced the lack of enthusiasm for the use of this technique. However, organization of the therapy at that time was poor and had little psychological input; moreover, the patients were given almost no ongoing support from clinical nurse specialists with expertise in these areas. Once this had been introduced into normal practice, a number of reports supported the idea of a non-surgical approach.

The technique involved requires passive dilatation of the vaginal dimple using graduated dilators. The dilators that are used come in a number of formats and also are made of a number of different materials. No particular design is better than the others, and the results are significant in that a well-motivated, well-supported patient can achieve a vaginal length which is totally satisfactory for intercourse in 85% of cases[22] (Table 2). Importantly, however, whichever technique is used to try to create a vagina, a proper assessment must be made of the results of therapy. This has, in fact, been extremely poorly addressed over many years and—apart from sporadic reports of small numbers—there has been no comparative study until recently. In a study carried out by Nadarajah et al (personal communication), we have shown that in 60 patients followed-up for up to 5 years, over 90% of the girls had a totally satisfactory sexual experience. Some 25% of the patients complained that they had either poor lubrication or dyspareunia but this did not interfere with their enjoyment of sexual intercourse and this was extremely gratifying. We believe that this technique has now been shown to be highly successful and, as stated by the American College of Obstetrics and Gynecology, it is the procedure of first choice.

Table 2. Results of dilator therapy.

Author	Patient numbers	Success
Rock et al[8]	21	18 (66%)
Broadbent et al[66]	20	19 (95%)
Roberts et al[67]	51	46 (91%)
Edmonds[68]	242	205 (85%)
Total	334	288 (86%)

In an attempt to try to improve patient co-operation with this type of passive dilatation, Ingram developed a method whereby a dilator was placed through a bicycle seat stool so that the patient could lower herself onto the dilator and use her body weight to create the pressure.[27] Ingram reported the use of his technique on 24 women, of whom 50% had primary vaginal agenesis. Twenty out of the 24 patients had successful treatment, and the only four who failed to have a successful result had had previous neovaginal surgery. This technique is identical to Frank's procedure in terms of its principle, and the outcomes are equally gratifying.

Surgical management of Müllerian agenesis

In those patients who fail to achieve a functional vagina with passive dilatation, a surgical approach may be required. It is extremely important to realize that, whatever techniques are used, these patients will need to use vaginal dilatation post-operatively in almost all cases. As a result of this, in just the same way as it is important to prepare the girls for passive dilatation primary treatment, the fact that they have failed primary treatment at least means that they are familiar with the vaginal dilatation technique and it is more likely that they will persevere post-operatively and have a good result. However, failure of the use of dilators sometimes brings a negative approach from the patient who hopes that the surgery will be a one-off event, which will then mean that they do not need to use vaginal dilatation at all. It is important, therefore, that an appropriate psychological assessment be carried out to ensure that the patients are ready for this type of procedure and that they also get appropriate support post-operatively.

There are numerous descriptions of procedures which attempt to create a functional vagina; these procedures fall into various categories (Table 3).

Surgical creation of the neovaginal space

For all techniques in this group, the initial approach is identical in that a transverse incision is made at the apex of the dimple and a digital dissection of the space between

Table 3. Procedures which aim to create a functional vagina.

Surgical creation of a neovaginal space	Split-thickness skin graft Full-thickness skin graft Perineal skin graft Amnion Peritoneum Absorbal adhesion barriers Tissue expansion Flap vaginoplasty
Vulvo-vaginoplasty	Williams' operation
Bowel vaginoplasty	Small bowel Sigmoid colon Caecum
Vecchietti's operation	Laparotomy Laparoscopy

the urethra and bladder anteriorly and the rectum posteriorly is carried out. It is essential that the depth of the neovaginal space reach the peritoneum which lines the pelvic cavity as failure to do this results in excessive contracture post-operatively. A mould is then placed in the space and this mould may be lined with a number of materials. In the United Kingdom, the most widely used material is amnion but in the United States, it is more common to use the McIndoe Reed technique with the use of split-thickness skin grafts taken from buttock. The complications of this type of surgical approach involve intra-operative damage to either the rectum, urethra or bladder with subsequent fistula formation at rates ranging from 0 to 7.6%.[8,28] A number of authors have used different types of mould—soft, semi-rigid or rigid—and there seems to be good evidence that the use of soft moulds reduces the risk of fistula formation, presumably from lack of avascular necrosis from pressure. An alternative to the use of these moulds is the use of the inflatable soft stent which, it is claimed, reduces the risk of haematoma formation without compromising healing and without the risk of fistula formation.[28] In the largest reported series by Alessandrescu et al[29], involving the treatment of 201 patients, the surgeons used a rigid mould throughout their experience and had a fistula rate of $<1\%$; therefore, it may well be that it is not the type of mould that is important but the skill and experience of the surgeon. The functional outcome of the McIndoe type of procedure is summarized in Table 4. Although the functional outcome is reported anecdotally, with success rates ranging between 80 and 100%,

Table 4. Results of McIndoe skin grafting.

Author (reference)	Patient numbers	Serious complication	Sexual satisfaction (%)
LeDuc et al[69]	15	0	8 (47%)
Garcia and Jones[70]	54	N/A	53 (99%)
Farber and Mitchell[71]	12	2	10 (80%)
Alessandrescu et al[29]	201	N/A	188 (94%)
Wiser and Bates[72]	92	2	89 (96%)
Buss and Lee[73]	47	5	40 (85%)
Rock et al[8]	79	5	79 (100%)
Hojsgaard and Villadsen[28]	23	3	18 (78%)
Counseller[74]	150	N/A	142 (95%)
Varner et al[75]	29	3	29 (100%)
Khanna and Khanna[76]	17	1	15 (88%)
Harkins et al[77]	21	3	18 (88%)
Bryans[78]	15	0	12 (80%)
Karjalainen et al[79]	33	1	32 (97%)
Salvatore and Lodovicci[80]	90	9	81 (90%)
Kunz et al[81]	24	0	22 (92%)
Feroze et al[82]	28	7	21 (75%)
Page and Owsley[83]	21	N/A	17 (81%)
Cali and Pratt[84]	93	N/A	84 (90%)
Jackson[85]	128	4	109 (85%)
McIndoe[86]	96	N/A	91 (95%)
Roberts et al[67]	14	0	14 (100%)
Ozek et al[87]	29	N/A	29 (100%)
Total	1311		1201 (92%)

there has not been a specific study surveying sexual satisfaction to ensure that these results are indeed true.

The use of amnion has been described by Ashworth et al[30] and subsequent experience with this technique has shown that it has success rates very similar to those of the McIndoe procedure. It has the advantage that no graft site is required, thereby leaving no external scars for the patient to have to tolerate. However, it is important that the use of this material is properly governed and that the donors are suitably screened for HIV and CJD.

The use of adhesion barriers has been reported, although only in small numbers of patients, but in the report of Jackson and Rosenblatt[31] the four patients treated had 100% success in creating a neovagina after 6 months.

The use of peritoneum to line the neovaginal space has been popularized in Russia by Davydov and Zhvitiashvili.[32]

Bowel vaginoplasty

Segments of intestine have been used as a vagina ever since intestine was introduced for this purpose in 1892 by Sneguireff.[33] He used the rectum as a substitute for a vagina and created a colostomy but this did not become popular. Ileum was used for the first time in 1907 when Baldwin reported its use in a patient;[34] the procedure therefore became known as Baldwin's procedure. The advantage of using ileum is that the calibre of the bowel remains constant and there is good lubrication; however, there are disadvantages as the mucosa of the small intestine is very easily traumatized by intercourse so that bleeding often occurs. Also, there is chronic secretion of mucus from the loop and, in the end, this technique has not remained popular.[35] The use of an isolated segment of sigmoid colon was first reported in 1914 by Ruge.[36] He used lower sigmoid colon and, although initially a number of these procedures ended with bowel necrosis and fistulae, over the ensuing 80 years the technique has gradually become modified and improved and is now known as a colocolpopoiesis.[37] The most recent results of the use of bowel are summarized in Table 5 in which it can be seen that, although the success rate overall is between 77 and 90%, the complication rates are not inconsiderable. This technique of using the bowel is therefore generally reserved for more difficult cases where simple vaginoplasties have failed and the post-surgical situation is one that results in excessive scarring. In these circumstances, in order to create a neovagina of some function, the whole neovaginal area needs to be excised and replaced by a loop of bowel; it is recommended that sigmoid colon is the best segment to use.

Syed et al[38] reported the outcome of 18 children who had undergone colovaginoplasty when aged between 1.5 and 8 years. They used sigmoid colon in the hope that this might abolish the problems of the emotional difficulty of reconstructive surgery in adolescence by creating a functional vagina in childhood. Although there were no major complications in the early follow-up period, three patients developed severe vaginal discharge problems within 2–7 years and the histology confirmed diversion colitis. This is a reasonably serious complication and one which has made these authors suggest that childhood bowel vaginoplasty should not be continued.

Finally, the long-term sequelae of this procedure are unknown but Hiroi et al[39] recently reported a mucinous adenocarcinoma arising in a neovagina using the sigmoid colon.

Table 5. Results of bowel vaginoplasty.

Author (reference)	Patients	Success	Complications	Comment
Novak et al[88]	63	59	2 necrosed; 2 prolapse	Sigmoid
Burger et al[89]	9	8	1 stenosis	Sigmoid/ileum
Ghosh and Kwawukume[90]	15	15	?	?
Franz[91]	13	12/13	1 stenosis	Sigmoid only
Hensle and Reiley[92]	31	20/26 (77%)	4 vaginal d/c; 3 stenosis; 1 dyspareunia; 3 patients required repeat procedure	Sigmoid better than ileum
Parsons et al[93]	28 (ages 6–21)	14/16 adults	4 stenosis; 4 prolapse; 2 partial bowel obstruction	
Syed and Malone[38]	18 (all children)	(80% satisfied)	3 d/c; 3 cases of diversion colitis	? Not use bowel as primary procedure
Freundtet al[37]			3 cases of prolapse of sigmoid	
Hiroi et al[39]			Adenocarcinoma in sigmoid 30 years later	

Neovaginoplasty using peritoneum

Although it had been described earlier, the use of peritoneum to line the neovagina was first reported in a series by Davydov and Zhvitiashvili in 1974[32] and has since been known as the Davydov operation. In his procedure, Davydov performs a laparotomy, having created a neovagina in the normal way; then, by mobilizing the peritoneum from the peritoneal cavity—including the rectum—he uses this to line the neovaginal cavity. The advantages of this technique as claimed by Davydov and co-workers are the lack of granulation and scar formation. However, as can be seen in Table 6, the success rate is not universally 100%—although the results are remarkably good in terms of success rates for vaginal intercourse.

Table 6. Results of Davydov operation.

Author (reference)	*N*	Success (%)	Complications
Davydov[94]	200	100	0
Mobus et al[95]	44	82	0
Tamaya and Imai[96]	24	24	0

Recently, a laparoscopic approach for the use of peritoneum has been reported by Soong et al[40] In this series the authors claim a 100% success rate and suggest that it is unnecessary for this procedure now to be performed through a laparotomy. Similar experience has been reported by other authors.[41–43]

Vecchietti's operation

This procedure involves the creation of a neovagina using dilatation of the vaginal dimple with a traction device attached to the abdomen. In the conventional operation, a laparotomy is performed and a suture is passed through the perineal membrane from above. This suture is threaded through a plastic olive and the suture is then passed back through the vault and up through the abdomen. The abdomen is then closed, and the ligature is then attached to a traction device, which is strapped to the patient's abdomen. Traction on the suture is then increased on a daily basis to pull the olive into the neovagina and to stretch the vaginal skin in order to create a vagina. This mimics the technique of Frank but does not rely on the woman herself to use the dilators. After 7–9 days, Vecchietti claims to have produced a vagina of some 10–12 cm in length and, at that stage, he introduces the use of vaginal dilators to ensure that the skin remains stretched. In Vecchietti's personal series of 522 procedures, he claims a 100% success rate and only nine complications—which included one rectal and one bladder fistula. Table 7 outlines the conventional Vecchietti technique and its results.

Recently, a number of series have reported a laparoscopic approach to this procedure, thereby avoiding the laparotomy to insert the suture. The rest of the procedure is as in the conventional technique and the results are seen in Table 8.

Vaginoplasty using skin flaps

Graves first reported the use of skin flaps from the labia minora in 1921[44], and in 1927 Frank and Geist suggested that a tube graft from the inner aspect of the thigh could be used for vaginoplasty.[45] The principle behind using skin flaps is the advantage of full-thickness grafting. The disadvantage of the McIndoe technique, which uses a split-thickness graft technique, is the problem of contraction of the vagina post-operatively and in the full-thickness grafts this ought to be avoided. Over the ensuing years, a number of procedures have been suggested using gracilis myocutaneous flaps[46] and rectus abdominis myocutaneous flaps.[47] In 1989, Wee and Joseph described the technique of pudendal thigh flap vaginoplasty,[48] and in the follow-up study of 12 patients[49] they found that this technique was extremely useful—particularly in patients who had vulval anomalies secondary to CAH. A further report of the use of pudendal thigh flaps in eight patients with MRKH syndrome showed that this was successful in

Table 7. Results of Vecchietti's conventional technique.

Author (reference)	Number of patients	Anatomical success	Functional success	Complications
Borruto[97]	522	522	522	9
Brun et al[98]	17	15	17	1 (re-operated with skin graft)
Janisch et al[99]	9	9	9	0

Table 8. Results of laparoscopic Vecchietti technique.

Author (reference)	Number of patients	Anatomical success	Functional success	Complications
Keckstein et al[100]	9	9	N/A	0
Fedele et al[101]	52	52	51	0
Khater and Fatthy[102]	6	5	5	0
Giacalone et al[103]	7	7	6	0

100% of cases in achieving a functional vagina.[50] However, the disadvantage of this technique—and of any technique that uses hair-bearing skin as the donor site—inevitably means that there is some hair growth in the created vagina and this can be a problem, both in terms of dyspareunia and discharge.

The use of a free flap graft from the scapula was first reported by Johnson et al in 1991.[51] This technique was carried out on three women and although they achieved a good functional length vagina at the end of the procedure, the surgical undertaking was enormous and this technique has not subsequently become popular. In 1996, Giraldo et al reported the use of a vulvoperineal fasciocutaneous flap (the Malaga flap) in the treatment of MRKH in six patients.[52] The results are very encouraging.

Finally, the use of tissue expanders to create excessive vulval skin to act as the donor site was first reported by Lilford et al in 1988[53]; these authors subsequently reported their experience of 17 cases.[54] They reported that vaginal length was satisfactory in 16 of the 17 women—although, again, they had two patients who complained of vaginal discharge and a number of patients who subsequently complained of hair in the vagina causing dyspareunia. In 1993, Serra et al[55] reported their experience of six patients with 100% success, only one patient complaining of dyspareunia. In 1997, Belloli et al[56] reported their successful experience with two patients. While this constitutes only 25 patients altogether, the results in creating a vagina seem to be very good. However, the complications of hair-bearing skin makes this more difficult, although Belloli's use of labial skin may have some promise.

Williams' vulvoplasty

In 1964, Williams described a vulvo-vaginoplasty which involved the creation of a vulval pouch using the labia majora and minora[57]; however, as this procedure creates an abnormal angle for intercourse—and may in fact create dyspareunia in the long term—it does not have a role in routine vaginoplasty. In those patients in whom a neovagina cannot be created, this may provide some form of pouch into which intercourse can occur and can be considered in these rare circumstances.

MRKH AND SURROGACY

In MRKH syndrome, the ovarian function is normal. Therefore, the possibility of IVF surrogacy is one that has come to fruition in terms of success. In studies initially looking at the ovarian response to gonadotrophins, the studies showed that there was an absolutely normal response rate in these patients in IVF programmes.[58,59] In 1997, Petrozza et al[14] reported on 162 IVF cycles in MRKH syndrome patients. Thirty-four

live children were born. The fact that no congenital abnormalities were found in these offspring was very encouraging and strongly suggests that the congenital absence of the vagina is not inherited in a dominant fashion. Similarly, Beski et al reported their experience with similar outcome figures and no congenital abnormalities.[60] The experience of Goldfarb et al[61] in their surrogacy programme gave equally encouraging results. Clinicians should therefore be encouraged to consider IVF surrogacy as an option for MRKH patients who wish to attempt pregnancy.

UTERINE ANOMALIES

The only uterine anomaly in the menstruating teenager that may be a problem is the presence of rudimentary horns. These cause increasingly severe dysmenorrhoea, and in all teenagers who fail to respond to the normal medication for primary dysmenorrhoea an ultrasound scan should be performed to identify the presence of a rudimentary horn. Occasionally, these may be communicating—in which case no haematometra will be seen—but if they are non-communicating a haematometra is apparent and retrograde menstruation may well lead to a haematosalpinx and/or an endometrioma. Treatment of this problem is excision of the rudimentary horn and reconstruction of the uterus; reproductive performance in these women is normal if the horn is non-communicating. If patients have a communicating horn, care must be taken at the time of surgery to reconstruct the uterine cavity to give it sufficient strength to withstand the physiological changes of pregnancy, and a decision should be made as to whether or not delivery should be by elective Caesarean section.

Occasionally, in the MRKH syndrome the uterine anlage may have functional endometrium and here excision of the anlage resolves the problem.

CONGENITAL ABNORMALITIES OF THE FALLOPIAN TUBES

These abnormalities are extremely rare and include duplication of the tube or absence of the fallopian tubes. In one fascinating case, a woman was reported to have bilateral absence of both fallopian tubes and ovaries.[62] These abnormalities are curiosities and have no clinical significance in teenage years. However, they may have implications for reproduction in later life.

CONGENITAL ABSENCE OF THE CERVIX

This is an extremely rare condition: the literature contains no more than 60 recorded cases. The clinical presentation is extremely similar to obstructive outflow tract disorder, with cyclical abdominal pain presenting in the presence of amenorrhoea and normal secondary sexual characteristics. Recurrent retrograde menstruation occurs due to failure of outflow so that haematosalpinx and endometriosis are extremely common associations. Diagnosis is usually made on ultrasound when it is quite possible to demonstrate the absence of the cervix and vagina and the presence of a haematometra. Management is controversial. Most experts would advocate hysterectomy in these circumstances owing to the risk of sepsis, with attempts to perform an utero-vaginal anastomosis. However, the author's own series of 15 patients has given

a 50% success rate with ongoing menstruation beyond 1 year—although seven patients have failed their surgery and have therefore been subjected to a hysterectomy.[63] It is also possible to retain the functional uterus using either continuous oral contraceptive pills or GnRH analogues with add-back therapy if an attempt is to be made to retain the uterus for subsequent assisted reproduction. What is clear here is that this type of congenital abnormality is rare, the problems of reconstruction are complex and these cases should be managed only in centres with existing expertise in the management of this condition.

SUMMARY

Congenital malformations of the genital tract are not common. However, the impact that they have on young women's lives can be devastating. The correct management of these cases is imperative and this requires a multi-disciplinary team of healthcare workers in order to ensure that the best outcomes—physically, mentally and socially—are achieved. As we look into the future, it is perfectly likely that new techniques will bring improved anatomical results from surgery, particularly in the creation of the new vagina. There are a number of tissue-engineering projects, aimed at growing new skin, which give great hope for full-thickness grafts in the future. However, tissue engineering

Practice points

- the best results in vulval surgery are achieved at the first attempt
- reduction cliteroplasty has less success in sensory outcome than previously believed
- congenital absence of the vagina requires a multi-disciplinary approach to achieve the best results
- surgery for outflow tract obstruction is best performed in centres with extensive expertise to avoid subsequent stenosis
- the use of a psychologist in the management of these cases is imperative
- congenital absence of the cervix requires complex reconstruction that should be carried out only in centres with expertise
- persisting rudimentary horns leading to increasing primary dysmenorrhoea should be excised

Research agenda

- identification of the genes involved in the control of the development of the lower genital tract
- the development of tissue-engineering techniques to create a neo-uterus and neovagina for transplantation
- the development of databases internationally to record a worldwide experience and database on the condition
- further psychosexual research to improve the outcome of the management of these conditions

will not stop at the creation of skin and we will be looking to the creation of new organs by tissue-engineering techniques which will bring us hope for girls with abnormalities to achieve physical normality in the longer term.

In the quest to understand these disorders, the hope of molecular genetics will be the discovery of the gene sequences that are responsible for development of the genital tract and an understanding of whether or not this knowledge could be used to influence development at a subsequent point in time.

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