Review Article

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Obstructive lesions of the Genital Tract in Adolescent Female

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Abstract

Adolescents have suffered from disorders of the genital tract since human race began.Malformations may affect the genital Tract alone or in association with urinary tract or intestinal tract Anomaly. Formation of haematocolpus and or haematometra due to the obstruction to the outflow tract is also known as cryptomenorrhoea. In presence of normal uterus,this could occur mainly due to imperforate hymen,transverse vaginal septum,absent vagina.Obstructive lesions of the genital tract in adolescent femalemay present as acute abdomen. The issues of congenital malformations of genital tract are complexpscycologically and gynaecologically.Patient should be treated in multidisciplinary , holistic manners. Knowledge of the clinical presentation and Management of these cases is exlremely important.

Keywords: Obstructive Lesions; Imperforate Hymen; Transeverse Vaginal Septum; Acute Abdomen.

Introduction

The subject of Adolescent Gynaecology is not new. Adolescent having suffered from disorders of the genital tract since the human race began [1]. adolescents, mode of presentation of surgery and associated problems may vary at times, and also certain types of surgical disease are known to have a predilection in adolescents [2].

Ever since man evolved on this planet and medical science emerged for giving solace to his various disorders, Acute abdomen pain has always persisted as a diagnostic and therapeutic dilemma despite resolution of many dark alleys, there still remains many to be solved.

Abdomen is a Pandora's box. It has a wide variety of organs within and they all have overlapping of symptoms and signs which give rise to difficulty in their interpretation so as to formulate a firm diagnosis.The common gynaecological diseases known to present as acute abdomen are pelvic inflammatory disease, ectopic pregnancy and its complications, tubo-ovarian cysts, torsion of ovary, hemorrhage, haematometra, haematocolpos and mittleschmerz. Mullerian anomalies should always be considered in case of acute abdomen in Adolescents [2].

The malformation which become evident at birth afterwards may affect the genital tract alone or may affect it in association with urinary tract or intestinal tract anomaly. Congenital malformations are often multiple, and it should be remembered that although malformation of the genital tract seems to be an isolated lesion, there may be renal tract abnormalities too which may not be recognizable unless IVP undertaken [1].

Diagnosis of associated malformation of mullerian ducts and urologic system are of atmost importance for successful results and minimizing complications. An Adolescent girl with genital tract anomaly presents with primary Amenorrhoea and / or acute lower abdominal pain in majority of cases. In an emergency without making an attempt for correct diagnosis, girls are subjected to unnecessary and incomplete surgery [3].

Acute Abdominal pain in Children presents a diagnostic dilemma. Although many cases of acute

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abdominal pain are benign,some require rapid diagnosis and treatment to minimize morbidity. The pain may herald a surgical or medical emergency. The most difficult challenge is making a timely diagnosis so that treatment can be initiated and morbidity prevented[4]. High degree of suspicion is the key to diagnosis. In acute presentation, the decision as to which test to order or which therapies to initiate must be made almost totally on presenting symptoms and signs [5].

No. of Principles can be Recommended

1. Adolescentshave different presentations than do younger children.
2. Adolescents have different presentations than do adults.
3. Be aware of entities related to lifestyle changes and exposure to environmental precipitants.
4. History taking requires excellent communication to overcome shyness relating to patient's self-awareness of sexuality or outright denial of symptom's significance.
5. In acute pain presentation, clinical judgment requires consideration of potential negative consequences of missed "surgical" abdomen, and surgical / gynaecological consultation is often indicated [5].

Genital tract outflow is important for the expulsion of normal secretions from the cervix and vagina. Outflow is also critical for menstrual efflux. Outflow obstruction may occur at different levels with resultant variations in clinical presentation. With appropriate treatment, the prognosis is excellent, and full recovery is typical.

Embryologically, the lower two thirds of the vagina develops from the urogenital sinus. The upper vagina, cervix, uterus, fallopian tubes, and ovaries form from the müllerian duct system. Failure of vertical fusion or canalization of the two systems in utero may result in bicornuate uterus, uterus didelphus, cervical stenosis or atresia, vaginal atresia, or longitudinal or transverse vaginal septa. In addition, hymenal tissue may be imperforate.

The exact etiology of such genital tract anomalies is unclear. Any defect in the normal organogenesis involving the urogenital sinus or the müllerian duct can result in genital tract anomalies. Interest has been focused on abnormalities in the structure; expression or function of the mammalian *HOX* genes may be a possible etiology for such malformations [6].

Epidemiology

Female genital malformations occur in 5-7% of the general population. The incidence of müllerian agenesis is about one in 4500 women; 30-36% of such malformations are associated with other anomalies as well, particularly anomalies of the kidneys and skeleton [6].

Formation of haematocolpus and / or Haematometra due to obstruction to the outflow tract is also known as cryptomenorrhoea. In the presence of normal uterus, this could occur due to:-

Imperforate Hymen

Transverse Vaginal Septum

Absent Vagina

[American Society of Reproductive Medicine ASRM Class-I]

Imperforate hymen is the most common genital outflow tract anomaly. Since development of Kidneys and urinary tract is closely linked to development of mullerian ducts, anomalies of kidneys and urinary tract are common in these girls [7]. A trans- verse vaginal septum / imperforate hymen represents only 3% - 5% of cases of Primary amenorrhoea .Lack of adequate reproductive organ development requires spe-cific surgical, medical, psychological and long-term fol-low-up [8].

Imperforate hymen

The hymen is a thin membrane that occurs at the junction of the sinovaginal bulb with the urogenital sinus and is usually perforated during fetal life. Failure of this perforation leads to the membrane remaining intact and as puberty begins, menstrual blood collects behind the membrane and the vagina begins to distend [9]. Variations of an imperforate hymen include microperforate, septate, stenotic, and cribriform hymen. If menstruation occurs and hematocolpos develops, the hymen may bulge and exhibit a bluish discoloration [6]. This is often painless initially and only when the vagina becomes sufficiently distended does a haematocolpos result and cause discomfort. Occasionally, if the mass is sufficiently large, it may affect micturition and defaecation, and may be palpable abdominally. Inspection of the vulva reveals a membrane that is blue in appearance with the darkened blood transilluminating through the thin membrane [9]. Atresia of the cervix has similar results but only the uterus and tubes are affected [10]. Retrograde menstruation can result in endometriosis and in infertility due to adhesion of the fimbrial ends of the

fallopian tubes. A similar condition may affect the newborn owing to the accumulation of cervical secretions behind the membrane [11].

A differential diagnosis of a transverse vaginal septum must always be considered but the appearances here are totally different with the septum being pink, although bulging, because the septum is so much thicker. Surgical treatment involves a cruciate incision to relieve the obstruction and the remaining quadrants of the hymen may be left in situ or may be excised. Following surgery, the haematocolpos will completely drain within 3–5 days and usually with no sequelae to this condition whatsoever especially in cases presenting early [9].

However, this apparently minor operation demands great care over asepsis, as do the more complicated ones mentioned above. The vagina has been closed throughout life so it is without the protecting lactobacilli, its epithelium is poorly formed, and its reaction is alkaline or weakly acid. There is therefore little natural resistance to bacteria entering from below; indeed, the degenerated blood and debris offer a favourable medium for their growth. Postoperative salpingitis and peritonitis are therefore real hazards and, in the past, were sometimes fatal. Bilateral hydrosalpinges occur as late sequelae.

The fluid which escapes after removal of the obstruction resembles liquid chocolate. It is devoid of fibrinogen and prothrombin and contains mucins (from the cervix), lactic acid (from the blood), calcium and altered blood pigments. Its amount varies from 200 mL to 2 litres or more, and it may take several days to run away. Its escape should not be hurried by the insertion of a drainage tube or by intermittent pressure over the lower abdomen, because such procedures encourage the entry of organisms. For the same reason vaginal examination should be avoided and the state of the uterus and tubes left in doubt for 1–2 months. Meanwhilethe vulva is kept covered with a sterile pad and the patient is discouraged from sitting in a bath.

At the end of the prescribed time, vaginal examination is carried out to see if there is any remaining evidence of haematometra or haematosalpinx. Fortunately, the uterus and tubes show enormous powers of recovery so that radical treatment such as salpingectomy and hysterectomy is rarely necessary. A high percentage of girls treated for haematocolpos later prove to be fertile [10]. Transverse vaginal septum The incidence of this is unclear but probably not greater than 1:30 000–50 000. The septae may occur anywhere along the length of the vagina although they are classified as upper, mid and lower, with the upper septae accounting for 46%, 30–40% in the mid vagina, and 15–20% in the lower. The presenting symptoms are usually increasing cyclical abdominal pain and the absence of menstruation. It is not uncommon for the diagnosis to be missed for several months. Only when a clinically palpable mass is discovered does the possibility of an obstructed outflow tract disorder arise and ultrasound imaging will confirm the presence of a haematocolpos and occasionally a small haematometra. If there has been sufficient time lapse between the onset of menarche and the diagnosis, a haematosalpinx can also occur. The surgical management of this condition requires the excision of the septal defect in its entirety and subsequently an end-to-end anastomosis of the upper and lower vagina. It is imperative that dissection occurs laterally to excise all of the septal tissue or the risk of stenosis will occur. A firm vaginal mould should be inserted through the site of the anastomosis for a minimum of 10 days and thereafter the patient should be instructed in the use of vaginal dilators for 2–3 months to ensure that stenosis does not occur and that a functional result will ensue. Excellent results are normally obtained for lower and middle septal defects but for higher septal defects, the results are less encouraging. When the upper vaginal portion is short, there is a risk of damage to the bladder or the rectum and great care has to be taken during the procedure. When these higher septal defects are resected, a mould may need to remain in situ for 3–6 months to obtain the best results.

However, one of the difficulties with these patients is the age of presentation and often they can be as young as 12 or 13 years when the cooperation with the use of either moulds or dilators is difficult to ensure. This means that in some cases, patients may return some years later and although they have been able to obtain menstrual drainage, and use tampons, the ability to have sexual intercourse may be difficult due to stenosis. At this stage, a further procedure can be carried out in order to remove the stenotic area and attempt re-anastomosis but this should only be done if the patient is capable of maintaining a regimen of using vaginal dilators. Pregnancy success in patients with this type of disorder vary with the level of obstruction and Rock et al reported a pregnancy rate of 100% in patients with lower third obstruction, 40% in middle third and only 20% in the upper third and the likely explanation for this is the incidence of endometriosis at the time of the obstructive problem which may cause architectural damage to the pelvis. However, IVF is appropriate in this patient group but it should be emphasised that other than those patients with a lower third

anastomosis, delivery should be by caesarean section. Patients with lower third problems can be allowed to have a vaginal delivery but care must be taken in ensuring the use of an episiotomy to prevent lateral damage during head descent.

Longitudinal vaginal septum

Longitudinal fusion defects may occur in the presence of two hemi-uteri and two hemi-cervices. Embryologically, each cervix fuses with the urogenital sinus to develop into two hemi-vaginas.

If one of the hemi-vaginas fails to completely canalise, a blind vaginal cavity results and at the time of puberty when menstruation begins, menses from the unobstructed vagina are found to flow normally, whereas the obstructed hemi-vagina accumulates menstrual fluid. This can create a confusing clinical situation which often leads to late diagnosis. However, eventually a clinically palpable mass should lead the clinician to suspect outflow tract disorder. Dysmenorrhoea can be quite severe and patients are sometimes admitted as emergencies. The surgical management of these conditions involves careful excision of the vaginal septum in its entirety but care has to be taken because this type of septum can be very thick. It is ill-advised to drain a hemi-vagina as a temporary means as this may result in ascending infection, septicaemia and a life-threatening situation. It is imperative therefore that these procedures are carried out by surgeons whose skills allow them to do this surgery effectively and that the operation performed is curative. The results of surgery in these circumstances are excellent. It is important to inform the patients that their uterus didelphus remains and therefore obstetric complications need to be explained.

Uterine anomalies

From an outflow tract perspective, the only uterine anomaly that may cause a problem is the presence of a rudimentary horn. In this circumstance, the caudal ends of the Müllerian duct fail to fuse and a unicornuate uterus with an adjacent or attached uterine horn may result. This horn is functional and therefore at the time of the onset of menses, the endometrium within the horn will shed and therefore create a haematometra, with retrograde menstruation and severe dysmenorrhoea. In girls with dysmenorrhoea that is unresolved through normal medication, an ultrasound scan should be performed to identify whether or not there is a rudimentary horn present. When these are non-communicating, the horn needs to be removed surgically and the uterus reconstructed. If this is carried out meticulously, reproductive performance is the same as with a unicornuate uterus. (9)

Cervical Atresia

Obstruction at the level of the cervix is least often encountered , but is the most controversial. Congenital atresia of the cervix of an otherwise normal uterus or of a bicornuate uterus is rare.When it does occur, a reasonably normal vagina is invariably present. It is more common to encounter apperent cervical atresia in association with absence of Lower vagina.

The conservative approach is of uterovaginoplasty, while the radical method is to perform a hysterectomy. The decision is based on the clinical features, psychological status, approach and experience of the surgeon and, not the least, the wishes of the patient and her family.

When the vagina is well developed and the uterus well formed and functional, uterovaginoplasty can be attempted by a combined abdominovaginal approach. This is a technically difficult operation in which the uterus and vagina are opened, the collected menstrual blood drained from the uterus and the posterior wall of the vagina sutured to that of the uterus. A Foley catheter is inserted through the vagina into the uterus and anchored there. The uterine body is then reconstructed and sutured to the anterior vaginal wall. The Foley catheter remains as a stent for 8–12 weeks to permit

epithelialisation of the new tract. In patients with cervical atresia, restenosis may occur in 50% and hysterectomy is then required. In some of these patients, severe sepsis may resultand even the occasional fatality has been reported. All these patients have severe morbidity from the incapacitating pain. These features prompt many gynaecologists to recommend a hysterectomy to patients with cervical atresia. However, since these are young women, the psychological sequelae of such a radical step are tremendous, hence most surgeons nowadays attempt reconstructive surgery at least once. Pregnancy has occurred in some cases after creation of a neocervix and this fact further encourages surgeons towards the conservative approach. When the outflow of menstrual blood is prevented by a thick vaginal membrane at any level, incision of the latter alone is inadequate. The raw area in the vagina always sealsover rapidly within days if not hours. It is therefore essential tocover any deficiency with vaginal epithelium; this is best done by dissecting free the upper vaginal walls and “advancing” them downwards to be sutured to the

margins of the vagina around the lower limit of the obstruction. Pregnancy following such procedures is not very rare and is often best terminated by elective caesarean section. (10)

Vaginal Atresia

This occurs in various degrees and forms. The whole vagina may be represented by a solid strand of tissue which is difficult if not impossible to recognise. Whether or not this is present the condition is labelled congenital absence of the vagina. Next, the upper (46%), middle (40%) or lower (14%) zones of the vagina may be imperforate over an area 0.5–6.0 cm in depth. More frequently, the vagina is obstructed by a thinner membrane situated low in the vagina, just above the hymen. It represents a failure in breakdown of the partition between the Müllerian and sinovaginal bulb contributions to the vagina. This condition of imperforate vagina is frequently misdiagnosed as the much less common imperforate hymen. However, the distinction between the two is largely academic because In patients with müllerian agenesis, vaginal reconstruction is performed. However, repeat operations are often required, and complications (eg, stenosis and fistula formation) are not uncommon. Vaginal strictures may follow, necessitating repeated dilatation.

Laboratory Studies

The history and physical examination usually suffice to establish the diagnosis. In rare cases of transverse vaginal septum with microperforation, ascending infection may lead to pyohematometra. Fluid culture is helpful in determining appropriate antibiotic therapy. If the patient has a blind vaginal pouch and no pubic or axillary hair, karyotyping or serum testosterone levels are helpful in establishing the diagnosis of androgen insensitivity syndrome.

Imaging Studies

Three-dimensional (3D) ultrasonography of the pelvis can reveal additional associated congenital malformations of the uterus or cervix. Ultrasonography also reveals hematocolpos or hematometra. Magnetic resonance imaging (MRI) can delineate other associated abnormalities, such as those of the urinary tract. For patients undergoing surgery, an accurate and full assessment of the underlying anatomy is important and is performed via MRI before the operation is performed.(6)

Conclusion

Congenital abnormalities of the genital tract are uncommon and the general gynaecologist will only encounter these on rare occasions during their career. However, knowledge of the clinical presentation and management of these cases is extremely important in differentiating those patients whose care can be provided locally and those who need referral to a tertiary centre where appropriate management can be determined. Congenital abnormalities of the genital tract can be isolated, as in the case of an imperforate hymen, or more complex, involving the cervix, uterus and fallopian tubes and thus potentially affecting fertility and in some cases pregnancy may be impossible. Occasionally, congenital abnormalities of the genital tract may be associated with more complex syndromes and the importance of these relationships needs to be understood by those dealing with complex problems. Some patients who have these anomalies will find it extremely difficult to cope psychologically and may need a multidisciplinary team approach to manage their care. This is particularly pertinent as the majority of these patients are teenagers.

Congenital malformations of the genital tract in teenage and adult women generate understandable anxiety with regard to sexual function and reproduction. The issues are complex psychologically as well as gynaecologically. Superior outcome is achieved for patients treated in a multidisciplinary, holistic manner. Centres that try to manage these patients without an experienced team of health professionals are not giving their patients the best care, and as such, pose an ethical question about the appropriateness of their approach. Gynaecologists, like all doctors, must have the patient's best interests at the forefront of their decisions with regard to best practice.

Once the diagnosis of obstructive lesions of the Genital tract has been established, consultation with a pediatric gynecologist or urologist who is familiar with these conditions is recommended. In considering the appropriate management of the patient, the age of the patient as well as the psychological implications for the patient due to the condition must be taken into account.An accurate surgical intervention. Simple obstructive outflow tract disorders, such as an imperforate hymen, can be treated very effectively by the the approach of tertiary surgeons to obtain optimum results.

Obstructive lesions of the genital tract are rare but obstetricians and gynaecologists must be aware of them as they can affect obstetric outcome and fertility if the diagnosis is delayed.

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