A 15-YEAR-OLD PREMENARCHAL GIRL PRESENTED WITH A 4-DAY HISTORY of increasing abdominal pain. Her medical history was unremarkable, and she reported that she had never been sexually active. Physical examination revealed a palpable, nontender, nonpulsatile midline mass extending from the pelvis to the umbilicus. The patient declined a vaginal examination. Abdominal ultrasonography revealed a large pelvic mass of uncertain origin. The uterus could not be identified ultrasonographically. Computed tomographic images of the pelvis showed a hyperdense, nonenhancing, fluid-filled pelvic mass measuring 25 cm by 11 cm by 12 cm (Panel A, axial image, white arrows; Panel B, sagittal image, white arrows) that was causing substantial compression of the bladder (Panels A and B, black arrows). A tubular structure at the superior pole of the mass was identified as the uterus (Panel B, arrowhead). The patient was given a presumptive diagnosis of hematocolpometra (accumulation of menstrual blood in the vagina and uterus). Subsequent vaginal examination revealed a thickened imperforate hymen. After hymenotomy, two liters of coagulated blood drained from the vagina and uterus. The patient was discharged after 3 days without pain or vaginal bleeding.

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Imperforate Hymen with Hematocolpometra

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Case Report

A Rare Presentation of Imperforate Hymen: A Case Report

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Introduction. Acute urinary retention in a child is rare. Haematocolpos can cause a mechanical obstruction, resulting in acute urinary retention. Case Report. A 12-year-old girl presented to the surgical department with a one-day history of acute urinary retention and suprapubic tenderness. She had not started menses but had described period-like pains every month for the past six months. On examination, she had a palpable bladder with over 500 ml of residual urine and a bluish-grey bulge posterior to her urethral meatus. An US scan showed a large mass posterior to her bladder resembling a haematocolpos, and this was confirmed with an MRI scan. She was catheterised and eventually underwent a hymenectomy using a cruciate incision. She made a good recovery postoperatively. Conclusion. In the case of a peripubertal female presenting with acute urinary retention, haematocolpos should be considered as a diagnosis.

1. Introduction

This is a case report of acute urinary retention as a result of an imperforate hymen causing haematocolpos. The incidence of imperforate hymen is 1 in 2000 girls, and approximately half of these will present with urinary retention [1]. Haematocolpos is a rare condition, where the vagina is filled with menstrual blood, caused by uterovaginal pathologies such as an imperforate hymen [2]. Most cases of imperforate hymen are sporadic in nature; however there have been reports of familial cases, where both recessive inheritance and dominant inheritance have been shown [3].

2. Case

In September 2012, a 12-year-old girl presented to the accident and emergency department with a one-day history of acute urinary retention associated with suprapubic pain and dysuria. There was no history of vomiting or a change in bowel habit. She reported cyclical abdominal cramping pains in the preceding six months but denied having started menses. Her birth history and developmental history were unremarkable.

On examination, her abdomen was soft with mild tenderness suprapubically and in the left iliac fossa. Her bladder was palpable and she was noted to have a nontender bluish-grey bulge posterior to the urethra on examination of her external genitalia. Neurological examination was normal.

Urine dipstick was normal, and a urinary pregnancy test was negative. A bladder scan revealed over 500 ml of residual urine; therefore, a 10 Ch urinary catheter was inserted, which relieved her suprapubic pain. On repeat examination, the bladder was no longer palpable and a PR examination was normal with no palpable masses. Initial blood tests showed a mildly raised WCC at 11.7 and raised neutrophils at 10.15; all other blood results were unremarkable.

An ultrasound scan of the kidneys showed an 11 × 7.8 × 8 cm fluid-filled mass lying posterior to the bladder, inseparable from and lying immediately inferior to the uterus (Figure 1). The mass had a fluid level, and findings were consistent with a hydrometra. The right kidney showed mild hydronephrosis. No other abnormal findings were detected.

Following the ultrasound findings, she was referred to the gynaecology department and underwent an MRI scan. This showed an 11 × 7.8 × 8 cm mass lying within the midline of the pelvis, which had several fluid layers indicating that it consisted of blood products (Figures 2(a) and 2(b)). Superiorly, the fluid was in continuation with a single uterine cavity, and inferiorly, it extended down to the perineum. Appearances were consistent with a hugely distended uterus filled with menstrual products.
Subsequently, she underwent a hymenotomy (using a cruciate incision) with drainage of her hydrocolpos. Postoperatively, she made a good recovery with a successful removal of the urinary catheter. Since returning home, she has started experiencing normal menses and has had no further urinary problems.

3. Discussion

Acute urinary retention is not a common presentation in children and is more common in males [4]. When young females present, the causes can include mechanical obstructions (urinary tract stones, urethral strictures, trauma to external genitalia, and imperforate hymen), neurological disorders, and urinary tract infection [4].

Imperforate hymen is a rare genital tract anomaly which has an incidence of about 1 in 2000 [1]. Acute urinary retention can subsequently occur due to the pressure effect imposed on the bladder and urethra [5].

This case serves to illustrate that in peripubertal females with amenorrhoea and acute urinary retention, even though uncommon, a diagnosis of haematocolpos should be considered and excluded.

Conflict of interests

The authors declare that they have no conflict of interests.

References


CASE REPORT

A uterovaginal septum and imperforate hymen with a double pyocolpos

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ABSTRACT: The presence of both a uterovaginal septum and imperforate hymen is described in a young patient presenting with ongoing chronic pelvic pain and a double pyocolpos. Ultrasound and magnetic resonance imaging scans were performed. The patient underwent laparoscopic adesiolysis, hymenotomy with drainage of 200 mL of pus, and excision of a complete longitudinal vaginal septum. Over the past 5 years of regular follow-up examinations, the patient has always reported regular menstrual cycles and an absence of pelvic pain.

Key words: Müllerian / uterovaginal septum / septate uterus / vaginal septum / imperforated hymen

Introduction

A septate uterus is the most common uterine malformation and it originates from the incomplete resorption of the adjacent walls of the two fused Müllerian ducts. This septum may extend to the cervical canal and can be associated with a vaginal septum which can be partial or can reach the introitus. An imperforate hymen is a congenital resorptive defect which, on the other hand, does not apparently derive from the Müllerian ducts. The rare finding of both a uterovaginal septum and coexisting imperforate hymen (Fig. 1) is described in this case of a young patient presenting with ongoing chronic pelvic pain and a double pyocolpos.

Case Report

A premenarchal 13-year-old girl presented with severe chronic pelvic pain which had started 1 year earlier. The patient reported that her gynaecologist had justified her symptoms as being caused by cystitis, although she had not undergone a uterine culture or other uterine tract examinations. She was ultimately seen by a gynaecologist who performed a hymeneal incision for a pyocolpos due to an imperforate hymen. Despite this, symptoms persisted, and the patient was sent 1 year later to our tertiary referral centre for female genital malformations. At gynaecologic examination, the vaginal dimple ended blindly with a slight bulge. A pelvic ultrasound showed a septate uterus and an endo-vaginal hypoechoic cystic mass of 21 cm. Magnetic resonance imaging (MRI) scans noted a uterovaginal septum with a right hematocolpos of 20 × 6 cm (Fig. 2). Both kidneys were documented to be normal.

The patient was brought to the operating room, where she underwent an open laparoscopy followed by a vaginal procedure. Laparoscopy showed severe adhesions involving the omentum, anterior abdominal wall, bowel, uterus and adnexae. The uterus was laterodeviated to the right, had a mid-sagittal indentation of the external fundus and a large isthmic bulge. The pelvic adhesions were freed and no endometriosis nodules were seen. The tubes and ovaries appeared to be normal. The hymen showed fibrotic scarring resulting from the prior left hymeneal incision and drainage. We performed hystectomy on both left and right hemimembranes, and drained ~200 mL of pus from both hemivaginas. The longitudinal vaginal septum was identified and septoplasty was performed with straight Haney clamps placed along the anterior and posterior aspects of the vaginal septum. The 6-cm septum was excised along its length and the edges of the vaginal mucosa were reapproximated, both anteriorly and posteriorly, with a series of interrupted 3-0 Vicryl stitches. One month after surgery, the patient had an adequate vaginal diameter without evidence of mucosal scarring or narrowing. The patient has undergone regular annual check-ups since then, and after 5 years of follow-up, reports regular menstrual cycles and no recurrence of symptoms since surgery.

Discussion

The presence of a uterovaginal septum and concomitant imperforate hymen is likely to be coincidental, since these portions of the female genitals derive from two distinct embryological structures, and although both are resorptive defects, they occur at different stages of embryological development. Although the septate uterus is the most
frequently encountered Müllerian anomaly, this specific anomaly of a coexisting longitudinal vaginal septum and imperforate hymen has been previously described only in one case (Oakes et al., 2010) in the scientific literature. However, a case of a concurrent imperforate hymen with a transverse vaginal septum in a unicornuate uterus has been described by Creatsas et al. (Deligeoroglou et al., 2007), while both a longitudinal and a transverse vaginal septum were encountered by (Moawad et al., 2009). All authors indeed conclude that an adequate anatomical evaluation of each patient is paramount in tailoring the appropriate conservative and surgical treatment.

While a septate uterus is commonly diagnosed during a work-up for infertility, the diagnosis of imperforate hymen is made in adolescence, when the retained secretions consist of menstrual products, and the resulting mass effect in the vagina and uterus are referred to as hematocolpos and hematometrocolpos, respectively. A pyocolpos may result from an infection that is ascending through microperforations in the membrane, which may also explain why our patient did not firstly present with a muco/hydrometra. However, the pyocolpos with which the patient presented was most likely iatrogenic due to the previous hymeneal incision.

The combination of a uterovaginal septum with an imperforate hymen does not seem to fit into the existing classification systems and is inconsistent with the theory of linear caudal to cephalad Müllerian fusion as described by Crosby and Hill (1962). According to this theory, uterine development results from the fusion of the Müllerian ducts, which starts at the caudal-most aspect known as the Müllerian tubercle, and proceeding in a cranial direction. Septal resorption is thought to follow shortly thereafter, beginning at any point of fusion, and moving in either or both directions. The case reported here is not supported by this theory because a dual vagina/cervix complex suggests failure of caudal fusion, whereas a septate uterus indicates normal cephalad fusion with failure of septal resorption. The present case does in fact fit better into the alternative hypothesis suggested by Musset et al. (1967), which proposed a three-stage process in which the medial aspects of the Müllerian ducts begin to fuse in the middle and proceed in both the cephalad and caudad directions simultaneously. This is then followed by rapid cellular proliferation between the ducts which result in the development of the uterine body and cervix, and septal resorption, all of which occur simultaneously in both directions. The dual vagina/cervix complex in this case could therefore be due to failed fusion of the Müllerian ducts starting at the uterine isthmus towards the caudal direction. The septate uterus and vagina could then be explained by completely failed septal resorption after normal fusion.

The hymen is formed from the endoderm of the urogenital sinus epithelium and represents the junction of the sinovaginal bulbs with the urogenital sinus. An imperforate hymen is the result of failure of canalization of the vaginal plate. Since the lower third of the vagina is thought to derive from the urogenital sinus, this described association of a uterovaginal septum and imperforate hymen might shed new light onto the possibly intertwined embryological derivation of these caudal structures. An alternative view is presented in a recent study (Kimberley et al., 2012) which described 7 patients out of 31 with Rokitansky syndrome also presenting with hymenal variations. The authors suggest that those women without a hymen are also more likely to have renal tract anomalies, and postulate that in these patients the primary problem is with the underlying Wolffian duct defect, rather than being primarily a Müllerian duct problem. Cases like these and the one described in this report and the one described by Oakes et al. give some insight on how there is still much to be learned regarding the complex embryological networks that are engaged in organ formation.

The importance of understanding the embryological background of rare malformations allows the clinician to adequately treat each case despite the absence of a classification system for following the current treatment guidelines. The authors of the only other case of uterovaginal septum and imperforate hymen reported in the literature (Oakes et al., 2010) performed hymenotomy with a subsequent vaginal septoplasty and did not perform laparoscopy. Our decision to perform laparoscopy in this patient was based on the possibility
of treating possible pelvic adhesions and the presence of endometrio-
osis that may occur secondarily to chronic menstrual reflux in ob-
structive malformations such as in the present case.

Authors’ roles
L.F.: author and main surgeon, G.F.: co-author and assistant surgeon,

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Case Report

Abdominal Tumor in a 14-Year-Old Adolescent: Imperforate Hymen, Resulting in Hematocolpos—A Case Report and Review of the Literature

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1. Introduction

An emerging abdominal tumor in young females is a rare situation and requires a specific clinical and ultrasonographic approach. Common causes of a newly diagnosed abdominal mass in young females include cysts and solid tumors of various origins. In this context, hematocolpos is a rare entity that can cause such symptoms: it comprises the blood collection in the distal closed vagina and is usually diagnosed in young adolescents with no menstruation and cyclic abdominal pain. Its incidence is approximately one every 2000 young adolescents [1] and in 90% of cases is caused by an imperforate hymen.

Usual clinical signs include cyclical low abdominal pain, urinary retention, back pain, primary amenorrhea, and/or a quickly enlarging pelvic tumor. It may also affect neonatal age and can be manifested as fetal ascites or renal failure [2, 3], sometimes leading to variable degrees of hydroureter and/or hydronephrosis [4]. For its diagnosis, 2D sonography is the usually indicated imaging method of choice. 3D sonography and MRI are rarely suggested and used, although they both provide a better visualization and differentiation of the tissues and a safer distinction among other causes of hematocolpos, such as vaginal septum or partial agenesis. In addition, an endocrine profile of the patient is usually necessary [5, 6]. Surgical management is the treatment of choice, through incision or excision of the hymen, using cold knife, scissors, electrocoagulation, or laser. The recurrence rate remains low, occurring more often during minor surgical approaches, such as after a cruciate incision. Notably, a spontaneous rupture of an imperforate hymen is likely to precede any decision for surgical management [7]. Finally, further issues have to be
weighted, such as the bleeding and the subsequent emotional stress of the young female after the procedure, along with the completion and keeping of legal documentation.

2. Case

A 14-year-old girl was admitted to the Pediatric Emergency Department of the Department of Obstetrics and Gynecology, Johannes Gutenberg University of Mainz, Mainz, Germany, with primary amenorrhea, an expanding abdominal mass, and mild abdominal pain. There was no history of severe abdominal pain during the last year and the patient complained of polyuria during the last month; there were no signs of defecation. Her parents sought for medical assistance because of a growing tumor in her abdomen. At clinical examination, secondary sexual characteristics were present and within normal ranges. The clinical presentation was quite impressive: a thin girl with a BMI of 22 with a painless, nontender, soft, and homogenous mass, distorting her abdominal wall and expanding up to 5 cm over the umbilicus (Figure 1).

The patient’s vital signs were normal; laboratory tests revealed a hemoglobin concentration of 13 g/dL and white blood cell count of 11/nL, while CRP and tumor markers’ concentrations were within normal ranges. In addition, her endocrine hormonal profile was indicating a girl with a mature hypothalamic-pituitary axis. Urinalysis was normal. Clinical examination of the abdomen did not reveal any pain or signs of peritonism. Clinical gynecological examination after retracting the labia minor revealed an imperforate hymen, which was bulging forwards. Rectal digital examination revealed a large bulky mass positioned anteriorly. A structure of 34 cm length, 11 cm width, and 11 cm height was evidenced at 2D transabdominal ultrasound (Figure 2).

On the cranial, frontal end of the structure, cranial from the umbilicus and adapted to the front abdominal wall, we observed a uterus of normal size (no hematometra) (Figure 3), while both ovaries were present with a normal appearance. Both kidneys were present, with no anomalies or dilatation of the ureters.

3D ultrasound displayed the clarity of the wall of this structure: it appeared straight, with no adherence to the neighboring organs, homogenous, with a fluid-like content in it.

As the diagnosis was clear, surgical management was decided after providing the written informed consent of both parents and scheduled for the following day. A hymenotomy was performed under general anesthesia: at first, laser was used, followed by electrocoagulation, and an oval shaped piece of hymen was excised. A total sum of 2400 mL dark red, tarry blood was drained from the vagina. Of note, the maximum quantity reported in the literature is 3000 mL [8, 9]; spontaneous drainage was continued the following day too. No suturing of the remnant hymen was performed. Antibiotics were given prophylactically for the next 4 days.

3D imaging during the first postoperative day revealed a waveform vagina, with a length of approximately 21 cm, whereas the size of uterus regressed for 10 cm under the umbilicus but did not enter the minor pelvis. The patient was discharged from the hospital after two days and a weekly follow-up with 3D ultrasound was scheduled. Menstruation occurred 20 days postoperatively and vaginal length was normalized 3 days after. During a scheduled follow-up appointment, 2 months postoperatively, a small amount of blood was detected in the vagina through 3D imaging; recurrence of the hematocolpos was confirmed after genital inspection. Reoperation was booked immediately, leading to a wider triangular tissue excision.

The patient is not sexually active yet and during the last 12 months she has normal menstrual cycle and vaginal length, measured at ultrasound.
3. Summary and Conclusion

The approach of a young patient presenting with a newly diagnosed abdominal tumor is always a demanding process. It causes fear to the child, emotions of guilt to the parents, and additional responsibility to the clinician. Apart from the pediatrician, other medical specialties can assist towards diagnosis and management, such as general surgeon, gynecologist, endocrinologist, and radiologist.

Although hematocolpos consists of a rare clinical feature, it should be always considered a possible diagnosis in young females with primary amenorrhea and abdominal mass. Both diagnosis and treatment of hematocolpos are relative easy, but due to the sensitive nature of the disease, the approach of the patients presenting with that disease is demanding. Since the most serious complication is recurrence, in our experience, we recommend wide tissue excision as an initial approach, through a triangular or oval shape, instead of a cross or “X” shape incision. 2D ultrasound is the diagnostic tool of choice, but 3D ultrasound can reveal more details, such as the exact relationship of the feature with the neighboring organs and structures, since it provides better tissue differentiation and can assist in the vaginal length surveillance.

Conflict of Interests

The authors declare no potential conflict of interests.

References

Hematometrocolpos Secondary to Imperforate Hymen

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14 year old girl referred to our department with lower abdominal pain with abdominal distension.on examination large tender midline cystic mass noted extending from pelvis to epigatrium.

Ultrasound showing Huge midline oval hypoechoiec mass with scattered internal echoes.uterus on top of the mass.bladder not made out seperately rest normal.

Ultrasound showing over distended bladder catheterisation.Catheter is anterior to the lesion,little urine drained. No vaginal opening bulging membrane with bluish hue seen at the introitus.secondary sexual characters well developed.patient not attained menarche.

C.T and MRI confirmed the diagnostic findings of ultrasound.

DIAGNOSIS: Hematometrocolpos secondary to imperforate hymen.

Hydrocolpos is characterised by an expanded fluid filled vaginal cavity, when associated with distention of the uterine cavity as well ,the term hydrometrocolpos is used . When the fluid is mixed with menstrual products, it is termed as hematocolpos and hematometrocolpos .

Causes: Imperforate hymen (most common); Complete vaginal stenosis; Segmental vaginal atresia; Transverse vaginal septum Associations: Imperforate anus; uterus didelphys; Renal agenesis.

IMPERFORATE HYMEN

Imperforate hymen is the most common and most distal form of vaginal outflow obstruction. Persistence of the intact hymenal membrane results in the condition of imperforate hymen. The imperforate hymen is a solid membrane interposed between the proximal uterovaginal tract and introitus.

PATHOPHYSIOLOGY: Any obstruction of the vaginal tract during the prenatal, perinatal, or adolescent period results in the entrapment of vaginal and uterine secretions.

In patients with imperforate hymen, this obstruction is at the level of interoitus and becomes evident when the distensible membrane bulges between the labia.

Various terms such as mucocolpos, hematocolpos, and pyocolpos, are used to describe this condition depending on the retained contents.

When the diagnosis is made in adolescence, the retained secretions consist of menstrual products, and the resulting mass effect in the vagina and uterus are referred to as hematocolpos and hematometrocolpos, respectively.

MANAGEMENT: This patient underwent cruciate excision of the hymen under GA. More than 2 litres of blood coloured fluid drained after two days of hospital stay, the girl is relieved not only from the hospital-even from her pain, distress, girl went home with ooh and ALL IZ WELL.

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Case report

Imperforate Hymen - a rare cause of acute abdominal pain and tenesmus: case report and review of the literature

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Key words: Imperforate hymen, amenorrhea, pubertal girls, urine retention

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Abstract

Imperforate hymen is a rare condition that presents with amenorrhea, cyclical abdominal pains and urine retention among pubertal girls. A 14 year old girl with imperforate hymen underwent hymenotomy for hematocolpometra, having presented with abdominal pains and tenesmus.


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Introduction

Imperforate hymen, despite being the commonest female genital tract malformation [1], is a rare occurrence with a prevalence of 0.014-0.1% [1-3]. It mostly presents during puberty [1, 4] although diagnoses in utero [3, 5, 6] and during the new-born period and childhood [3, 7] are also documented.

There are few cases of Imperforate hymen reported in Africa. A case of unique presentation with tenesmus besides other documented symptoms was managed at a rural Kenyan hospital. There is no recorded case of Imperforate hymen presenting with tenesmus according to literature search. In this article, a review of the literature concerning the symptomatology of Imperforate hymen among pubertal girls is also presented.

Patient and observation

14 year old Kenyan girl of African descent presented to hospital with a weeklong complaint of lower abdominal pains associated with tenesmus. She had reduced appetite and poor intake of food due to the colicky pains. There was no abdominal distension but she had observed some suprapubic fullness. She did not have constipation, diarrhoea, vomiting or fevers. Her urinary habits were normal. She had never had her menstrual periods but she had developed secondary sexual characteristics. On examination, she was in severe pain, walking stooped over and had moderately tender suprapubic mass corresponding to a uterus at 16 weeks. Rectal examination revealed an anterior mass. Perineal examination revealed a bulging imperforate hymen exaggerated on valsalva manoeuvre. Pelvic ultrasound done revealed distended uterus and vagina all filled up with homogenous thick fluid (Figure 1, Figure 2). A diagnosis of hematocolpometra was made.

In theatre, an X-shaped incision of the hymen was made under anaesthesia and approximately 600mls of thick chocolate coloured blood evacuated. The edges of the hymen were everted and anchored by Vicryl 2/0 sutures. Analgesic cream and prophylactic oral antibiotics were prescribed. She made uneventful recovery and was doing well at 1 month. She was however lost to follow-up after that.

Discussion

Imperforate hymen is a layer of connective tissue that forms a transverse septum and obstructs the vaginal opening at the level of the introitus [5]. Usually, the hymen is a membrane that embryologically develops through the fusion of the caudal end of the paramesonephric ducts and the urogenital sinus [4, 5, 7, 8]. The central portion of this membrane perforates through the degeneration of its epithelial cells [5]. Failure of the degeneration of the epithelial cells and subsequent perforation leads to a hymen that is termed imperforate [5].

The function of the hymen is not clear but is thought to include innate immunity as it provides a physical barrier to infections during the pre-pubertal period when the vaginal immunity is not fully developed [3].

Imperforate hymen is rarely associated with other female genital tract malformations [1, 4] although some authors [2, 9] have emphasized the need to rule out associated mullerian malformations. It occurs sporadically but few familial cases have been reported [8].

Imperforate hymen can present during three main stages in life;

1. In utero: This is the rarest and occurs due to maternal estrogenic stimulation that leads to uterovaginal secretions filling up the blind vagina and presenting as hydrocolpos diagnosed through obstetric ultrasound [6]. The diagnosis should be confirmed post natally.

2. New-born-infanthood-childhood: In new-born period this may occur due to maternal estrogenic stimulation that leads to uterovaginal secretions filling up the blind vagina and presenting with hydrocolpos [1, 3, 7, 8, 10].

3. At puberty: This is the commonest. It occurs when a girl starts menstruating and the menstrual blood accumulates in the vagina [3, 10]. The age of presentation (mean, range) is 13.2 and 11-16 years respectively according to Liang et al [5] or 12 and 10-15 years respectively according to Lui et al [9]. Liang and colleagues did a ten year retrospective analysis of 15 women treated for Imperforate hymen through telephone based researcher administered questionnaire and a subsequent physical and sonographic examination. In their study, Lui et al did a ten year retrospective analysis of the data of 15 patients treated for Imperforate hymen but did not do any follow up patient interview or examination. Kurgodu and colleagues argue that the age of presentation is 2.5-4 years after thelarche [12].

Among the pubertal girls, Imperforate hymen will present in the following ways.

Amenorrhea

I. Primary amenorrhea

a. This is because the girl has started menstruating but does not experience any menstrual flow as the blood accumulates in the vagina, then in the uterus and occasionally, eventually into the fallopian tubes [3, 4, 7].

II. Secondary amenorrhea

a. This can occur following spontaneous closure of previously perforate hymen [8]. This can happen with a micro perforate or stenosed hymen. In such initial light periods will be experienced but continuous stenosis leads to complete obstruction and amenorrhea [8].

b. It can also occur as a result of stenosis of the hymenal opening following surgical or sexual trauma [8].

c. Lastly, it can occur as failure of hymenotomy [10]. In the months following hymenotomy the patient experiences her menstrual flow but the margins of the hymenotomy incision adhere and eventually occlude the vaginal outflow leading to amenorrhea.

III. Cryptomenorrhea

Pain

Recurrent cyclical lower abdominal/pelvic pains (up to 60%) [2, 4, 8, 9, 11, 12]. This is due to continued distension of the vagina and uterus by accumulating menstrual blood.
Low back pain (38-40%) [4, 13, 14]. Occurs as referred pain following irritation of the sacral plexus and nerve roots by the distended vagina and uterus.

**Obstruction**

**I. Urinary outflow obstruction and its complications (58%) [9]**

a. Acute urine retention (3-60%) [7, 9, 10, 13, 15]. This occurs by a number of mechanisms:
   i. Pressure on the bladder by the distended uterus causing angulation at the bladder neck and kinking of the urethra [10].
   ii. Direct pressure on the urethra causing urethral tamponade [10].
   iii. The bulging hymen distends the vagina and may cause cephalad angulation at the urethral meatus further stretching the urethra and worsening tamponade [15].

b. Complications of prolonged or recurrent urinary retention/obstruction
   i. Hydroureters [2]
   ii. Hydronephrosis [2]
   iii. Renal failure [1]
   iv. Acute bacterial nephritis [16]

**II. Vaginal outflow obstruction- Cryptomenorrhea**

**III. Intestinal obstruction**

a. Constipation (20-27%) [9, 13]

b. Tenesmus

**IV. Lymphovenous obstruction**

Compression of the pelvic veins and lymphatics can impair lymphovenous return from the lower limbs leading to oedema [1].

**Mass**

I. Distended uterus felt as pelvic mass on abdominal examination (20%) [9]

II. The distended vagina is felt as a pelvic mass on digital rectal examination

III. A bluish bulging hymen is observed beneath the labia (60%) [9]

IV. A cystic retroperitoneal mass is revealed on ultrasound or MRI [9]

With above in mind and a high index of suspicion, it is easy to make a diagnosis of imperforate hymen. Late presentation may be accompanied with complications such as ruptured hematosalpinx [9, 11], endometriosis [4, 15] and infection (pyocolpos and nephritis) [5, 16]. A clinical diagnosis negates the need for extensive laboratory and radiological investigations [10] and reduces the delay of intervention and length of hospital stay [9].

The management is aimed at re-establishing vaginal outflow and mainly consists of surgical hymenotomy under local or general anaesthesia [7]. Simple vertical, T-shaped, cruciform, X-shaped and cyclical incisions may be used [4, 7, 8]. X-shaped incision has the advantage of reduced risk of injury to the urethra-which should be stented during the procedure [7]. Pressure on the uterus in order to expel more blood is discouraged as it can lead to retrograde flow through the tubes causing endometriosis and tubal adhesions [15]. Hymenectomy and hymenotomy with a two week indwelling catheter have also been reported [8]. The outcome is good and the recurrences are rare [5].

**Conclusion**

Imperforate hymen is a rare condition but should be easy to diagnose when it presents. It should be suspected in pubertal girls who presented with acute abdominal pain.

**Competing interests**

The author declares no competing interest.

**Tables and figures**

Figure 1: Distended uterus

Figure 2: Bladder compression by the distended uterus

**References**


10. Abu-Ghanem S et al. Recurrent urinary retention due to imperforate hymen after hymenotomy failure: a rare case


Figure 1: Distended uterus

Figure 2: Bladder compression by the distended uterus
Case Report

Imperforate hymen and its complications: report of two cases and review of literature

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ABSTRACT

Imperforate hymen is a rare female genital tract malformation, arises as a result of complete failure of the inferior end of the vaginal plate to canalize. We report two cases of imperforate hymen with different presentation. Our first case was a 14 year old girl presented with lower abdominal pain and acute retention of urine with history of cyclical lower abdominal pain for last 6 months. Examination revealed mass of 14 weeks gravid uterus with bulged imperforate hymen. The second case was a 16 year old girl presents with primary amenorrhea with mass per abdomen. There was history of difficulty in micturition and constipation for last 4 months. On examination, a mass of size corresponding to 22 weeks gravid uterus with bulged bluish colour imperforate hymen was found. Both the cases were managed with incision and passive drainage of collected menstrual blood. On follow up both the cases resumed menstruation and doing well.

Keywords: Imperforate hymen, Primary amenorrhea, Acute urinary retention

INTRODUCTION

Imperforate hymen is the commonest female genital tract malformation and is noted in approximately 1 in 2000 females.1 Although the most common age of presentation is around puberty2,3 diagnosis in utero3,4 and during the new born period and childhood4,7 are also reported. We report two cases of imperforated hymen with different symptoms of presentation.

CASE REPORT

Case 1

A 14 year old girl presented with colicky lower abdominal pain and acute retention of urine for one day. She had not attended her menarche but had developed secondary sexual characters. There was history of cyclical colicky lower abdominal pain with backache for which she took medication from local hospital. For last one month there was increased frequency and difficulty in micturition. Although she had reduced appetite and poor intake of food due to colicky pain, there was no history of constipation, diarrhoea, vomiting or fevers.

She was catheterised with an indwelling Foley’s catheter and one litre of straw coloured urine was drained. Abdominal examination revealed, pain and tenderness in lower abdomen with a suprapubic mass corresponding to a uterus at 14 weeks. Examination of external genitalia revealed a bulging bluish colour imperforate hymen (Figure 1) which was exaggerated on Valsalva manoeuvres. The mass was found to be anterior to rectum in per rectal examination. Pelvic ultrasonography revealed distended uterus and vagina all filled up with homogenous thick fluid with internal echoes (Figure 2). She was diagnosed as a case of hematometra and hematocolpos with imperforate hymen and planned for
incision and drainage. In operation theatre, a cruciate incision was given and 800 ml of thick tarry blood was drained. The quadrants of the hymens are excised; the mucosal margins were everted and anchored by fine delayed absorbable suture (Vicryl 2/0). Local analgesic cream and prophylactic oral antibiotics were prescribed. Postoperative period was uneventful and she was discharged on 2nd postoperative day after removal of catheter. Follow up after one month revealed patent outflow tract and consummation of normal menstrual cycle.

Figure 1: 14 year old girl with bluish bulged imperforate hymen with associated vulvar distension.

Figure 2: Ultrasound scan showing hematometra and hematocolpos.

Case 2

We report a case of 16 year old girl who presented with primary amenorrhoea and mass per abdomen. She had history of colicky cyclical lower abdominal pain 4 months back for which she was treated in a local peripheral hospital after which pain subsided. She gave history of difficulty in micturition, constipation and occasional pain in back for last 4 months.

Examination revealed well developed secondary sexual characters with tanner stage IV breast, pubic and axillary hair. On per abdominal examination, the size of mass corresponds to 22 weeks gravid uterus and it was well defined, mobile, tender with non-palpable lower border.

Examination of genitalia revealed bulged blush colour imperforated hymen (Figure 3) and per rectal examination revealed that the mass was anterior to rectum. Abdominal and pelvic ultrasonography revealed large fluid collection in dilated and enlarged vagina, uterus and tubes with fine internal echo’s suggestive of hematocolpos, hematometra with hematosalpinx (Figure 4 & 5). Bilateral mild hydroureteronephrosis due to mass effect of the lesion with no free fluid in peritoneal cavity was reported in ultrasound. CT scan revealed no associated Mullerian duct and skeletal anomalies. She was planned for incision and drainage. In theatre, X shaped or cruciate incisions was made through the hymenal membrane at the 2-, 4-, 8-, and 10-o’clock positions after putting an indwelling Foleys catheter and 1000 ml of collected chocolate coloured blood was drained passively (Figure 6). The individual quadrants were excised along the lateral wall of the vagina, avoiding excision of the vagina. Margins of vaginal mucosa were approximated with fine delayed-absorbable suture (Figure 7). Local analgesic cream and oral antibiotic were given. She had an uneventful postoperative period and was discharged on 2nd postoperative day after removal of the catheter. She had menstruation after one month and is doing well in follow up examination.

Figure 3: 16 year old girl with imperforate hymen presented as a case of primary amenorrhoea and mass abdomen.

Figure 4: Ultrasound scans showing hematometra and hematocolpos.
DISCUSSION

The hymen is the membranous vestige of the junction between the sinovaginal bulbs and the urogenital sinus. It generally becomes perforate or patent during fetal life to establish a connection between the vaginal lumen and the perineum. Imperforate hymen is due to complete failure of the inferior end of the vaginal plate to canalize. Although most cases occur sporadically, cases of imperforate hymen involving multiple family members have been reported. The function of the hymen is not clear but is thought to include innate immunity as it provides a physical barrier to infections during the prepubertal period when the vaginal immunity is not fully developed.

Imperforate hymen is rarely associated with other female genital tract malformations although some authors have emphasized the need to rule out associated Mullerian malformations. If the hymen is imperforate, mucus and blood from endometrial sloughing accumulate in the vagina which can present during three main stages in life. As congenital hydrometrocolpos in intrauterine period which is the rarest and occurs due to maternal estrogenic stimulation that leads to uterovaginal secretions filling up the blind vagina and diagnosed through obstetric ultrasound. The diagnosis should be confirmed postnatally. In new-born and childhood period this may occur due to maternal estrogenic stimulation that leads to uterovaginal secretions filling up the blind vagina and presenting with hydrometrocolpos. However, more commonly adolescent girls present after menarche when menstrual blood trapped in the vagina behind the imperforate hymen which is known as hydrometrocolpos creating a bluish bulge at the introitus. With cyclic menstruation, the vaginal canal becomes greatly distended, and the cervix may begin to dilate and allow formation of a hematometra and hematosalpinx.

The age of presentation (mean, range) is 12 and 10-15 years respectively according to Lui et al. and 13.2 and 11-16 years respectively according to Liang et al. The common mode of presentation of imperforate hymen includes-

I. Amenorrhea, which may be primary due to accumulation of blood behind the imperforate hymen or secondary which can occur following spontaneous closure of previously perforate hymen. The later mainly occurs in micro perforate or stenosed hymen following surgical or sexual trauma where initial light periods will be experienced but continuous stenosis leads to complete obstruction and amenorrhea.

II. Recurrent cyclical lower abdominal/pelvic pains (up to 60%) due to continued distension of the vagina and uterus by accumulating menstrual blood and low back pain (38-40%) which is a referred pain following irritation of the sacral plexus and nerve roots by the distended vagina and uterus.

III. Obstruction

1. Urinary outflow obstruction and its complications (58%).

A. Acute retention of urine which is due to pressure on the bladder by the distended uterus causing angulation at the bladder neck and kinking of the urethra and direct pressure on the urethra causing urethral tamponade.

B. Chronic or prolonged urinary retention leading to hydrourereteronephrosis, acute bacterial nephritis and renal failure.

2. Vaginal outflow obstruction which is seen as a bluish bulge at the introitus.
3. In chronic cases intestinal obstruction leading to constipation (20-27%)\textsuperscript{10} and tenesmus\textsuperscript{13} also seen.

4. Lymphovenous obstruction due to compression of the pelvic veins and lymphatics can lead to edema of limbs.\textsuperscript{2}

IV. Mass per abdomen due to distended uterus and vagina with accumulated menstrual blood.\textsuperscript{10}

V. Retrograde menstruation may lead to the development of endometriosis and laparoscopy can be performed at the time of excision of an imperforate hymen to detect this.\textsuperscript{5}

 Differential diagnosis of imperforate hymen includes other obstructive reproductive tract anomalies like lower transverse vaginal septum. The associated vulvar distension, however, uniquely suggests imperforate hymen.\textsuperscript{1} Imperforate hymen is usually a clinical diagnosis which can be confirmed by ultrasonography.

The treatment includes surgical hymenotomy under anaesthesia following catheterization with an indwelling Foleys catheter to re-establish vaginal outflow. An X-shaped incision at 2-, 4-, 8-, and 10-o’clock positions is used which has the advantage of decrease risk of injury to the urethra. The quadrants of the hymen are then excised, and the mucosal margins are approximated with fine delayed-absorbable suture.\textsuperscript{15} Pressure on the uterus in order to expel more blood is discouraged as it can lead to retrograde flow through the tubes causing endometriosis and tubal adhesions.\textsuperscript{15} Needle aspiration of mucocolpos or hematocolpos should be avoided as it can lead to infection and pyocolpos formation.\textsuperscript{1} The outcome of surgical hymenotomy is good and the recurrences are rare.\textsuperscript{5}

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Imperforate hymen- Can it be treated by a simple vertical incision – A case report.

KEYWORDS

imperforate hymen, hymenotomy, cyclic pelvic pain

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ABSTRACT

Imperforate hymen is a rare congenital anomaly, with an incidence of 0.05%-0.1%, although it is the most common obstructive congenital anomaly of the female genital tract. It is thought that cause of this anomaly may be congenital defect in the degeneration of central cells of hymen. Generally it presents as an isolated anomaly, but some times genitourinary anomalies may be seen with it.

Patients are generally asymptomatic until menarche. Patients may present with primary amenorrhea, cyclic abdominal pain, urinary retention, suprapubic lump and tenesmus. Diagnosis is made with history and pelvic examination. Surgical treatment for this anomaly is hymenectomy after a cruciate or X shaped hymenotomy incision. Here we are reporting a case of imperforate hymen treated with a more conservative surgery which involved a simple vertical incision (hymenotomy). Post operative follow up was uneventful and patient attained normal menses. Hence it could be an alternative option as it is a hymen sparing procedure which involves preservation of hymen and so maintains integrity of female genitalia. This procedure is more acceptable by the patient and their families.

Introduction

Imperforate hymen is a rare congenital anomaly of female genital tract and is observed in 0.01-0.5% of new born. It is a developmental defect in which there is defective degeneration of central cells of hymen. It is generally regarded as an isolated congenital anomaly, sometimes genitourinary anomalies may be seen with it. These patients remain asymptomatic until puberty when they present with the complaints of primary amenorrhea, cyclic abdominal pain, suprapubic bulge, urinary retention, tenesmus and constipation.

Diagnosis is made easily by simple genital examination in which bluish bulging hymenal membrane is found. It usually manifests as a sporadic case, however familial inheritance has also been reported.

Approximately 58% of patients may complain of pain while urinating and retention of urine because of collection of menstrual blood in the vagina (hematocolpos). Untreated or late treatment may result in complications like infertility, endometriosis, and adhesions.

Standard treatment of this condition is hymenectomy after a cruciate incision or X shaped hymenotomy incision. An intact hymen is important in some cultures and religions. Patients and families have fears about losing virginity after surgical interventions.

We report a case admitted to our emergency ward with retention of urine, was diagnosed as a case of imperforate hymen, and was treated with a conservative hymen sparing surgery with single vertical hymenotomy incision. The procedure is less invasive than other methods described in the literature and more comfortable for the patients.

The aim of this case report is to show simple virginity preserving socially acceptable procedure to provide an intact hymen.

Case report

A 16 year old female came to our surgery department with the complaints of severe abdominal pain and retention of urine for 2 days. She suffered twice with the same episode of acute retention of urine with abdominal pain for the past three months. For this she was catheterised in some other hospital. She also had not attained menarche. No other significant history was found in personal and family history. On her physical examination secondary sexual characters were normally present. On local pelvic examination it was noted that hymenal opening was not there, instead a bluish bulge was there. An USG scan showed a large mass posterior to her bladder suggesting haematocolpos. Bilateral ovaries were normal. She was catheterised and underwent hymenotomy by simple vertical incision under general anaesthesia because of patient’s preference.

Approx 600 ml dark colored tarry blood was suctioned. Foley catheter no 16 F was inserted for one week. Post operative period was uneventful. She was discharged and followed up after one and a half months. She started having normal menses and was doing well. There was no stenosis or infection.

Discussion

The incidence of imperforate hymen in newborn has been found to be a rare congenital anomaly, with the reported incidence of 0.05%-0.1%. It is thought to be a developmental defect in which due to defective degeneration of central cells of hymen, it remains imperforated. This condition causes collection of menstrual blood behind imperforated hymen at menarche. Patients are usually not asymptomatic until puberty. At puberty they typically present with cyclic abdominal pain and primary amenorrhea. Other complaints are urinary retention, back pain and constipation.

These patients are easily missed when they present with retention of urine in emergency department. Proper history and pelvic examination is not done like it was seen in our patient in which patient had two previous episodes of urinary retention in which catheterisation was done but con-
It is interesting that patient was catheterised twice before coming to us but the nurse who catheterised did not notice the abnormal bluish bulge that was enough for diagnosis, neither the doctor suspected the possibility of imperforate hymen.

Acar et al reviewed 65 cases and found average age of 14 years at the time of diagnosis and hematocolpos was inconsistently present.[4]

Standard surgical procedure for this condition is hystero-nectomy using cruciate incision, but in our case we performed hymen sparing surgery in which single vertical incision was used and hymen was not cut.

Hymen sparing surgery was done primarily by M Basaran who treated cases with simple vertical incision. They used few oblique suture to prevent refusion.[4] In our case we did not use sutures instead Foley’s catheter was inserted for one week.

Temizkan et al also did virginity sparing surgery for imperforate hymen in two patients. They did simple central excision of the hymen leaving an intact annular hymen, but they did not use Foley’s catheter. No restenosis was seen in their patients.[5]

Acar et al did central oval incision with insertion of 16 f Foley’s catheter for two weeks on 65 patients and found equally good results.[5] Cheli et al conducted a study in which radial incision technique was applied on 3 out of 5 cases. In rest of two cases hystero-natomy procedure with simple excision was performed and Foley’s catheter was inserted,[8], as was done in our case.

Ali A et al did simple central excision of the hymen making an annular intact hymen using a Foley’s catheter for two weeks in order to prevent restenosis.[5]

In our case we gave simple vertical incision after drainage of blood from the vagina, Foley’s catheter 16F was put for one week and then she was discharged. There was no stenosis and infection in the follow up and patient resumed normal menses.

Conclusion-
As standard surgical procedure for imperforate hymen is cruciate incision over the bulging hymenal membrane. Another method could be single vertical incision, as was done in this case. This hymen sparing procedure seems to be more acceptable by most of the patient as it involves preservation of hymenal tissue. The result of one cases is not sufficient to provide a conclusion so studies with large number of cases are required to show the efficacy of procedure, so that it will have world wide acceptance.

REFERENCE
Case Report

Imperforate hymen: cause of lower abdominal pain in teenage girls


ABSTRACT

Imperforate hymen is a relatively rare congenital anomaly. However, it is not an uncommon cause of lower abdominal pain presenting in teenage girls. Without careful history taking and thorough examination, the condition can be missed easily. We report an imperforate hymen presenting as abdominal pain in three teenage girls aged 12, 12 and 13 years, respectively, within a six-month period. The presentation was reviewed and the various types of hymenotomy were discussed.

Keywords: adolescents, abdominal pain, haematocolpos, hymenotomy, imperforate hymen

INTRODUCTION

Imperforate hymen is a relatively rare congenital anomaly, in which the hymenal membrane occludes the vaginal orifice, resulting in haematocolpos, which often leads to abdominal pain in adolescent girls. It was reported that imperforate hymen occurs in one in 1,000 to one in 10,000.1 We report three cases of imperforate hymen, presented over a period of six months, that were initially missed. The importance of detailed history taking and thorough examination is highlighted.

CASE REPORTS

Case 1

In April 2006, a 12-year-old premenarchal girl was admitted with a two-month history of cyclic lower abdominal pain. Before admission, she had been treated by several general practitioners with analgesics. On admission, there was an obvious huge lower abdominal mass, and on careful perineal examination, a bluish bulging imperforate hymen was identified (Fig. 1). A hymenotomy and drainage of around 600 ml of old blood products were performed (Fig. 2). She was discharged on postoperative Day 2 with an uneventful recovery. On follow-up, she had remained well and had normal menses.

Fig. 1 Photograph shows the bulged hymen with a collection of menstrual blood.

Fig. 2 Operative photograph shows the appearance of the introitus after the hymenotomy.

Case 2

In July 2006, a 12-year-old premenarchal girl presented with a five-month history of lower abdominal pain associated with tenesmus and dysuria. On the day of admission, she could not pass urine for more than 12 hours. Despite the drainage of 300 ml of urine through a Foley catheter, there was still the presence of a large lower abdominal mass. Further detailed examination of the perineum revealed a bluish bulging imperforate hymen. The diagnosis was made clinically, and a hymenotomy and drainage of 500 ml of blood clot were performed. She could pass urine after the removal of the Foley catheter on postoperative Day 1 and was discharged on Day 2 with no more tenesmus or urinary symptoms. During follow-up, she had had normal menses since the operation.
Case 3
In September 2006, a 13-year-old premenarchal girl was admitted with a ten-month history of on-and-off lower abdominal pain. She had attended the accident and emergency department on several occasions for the same symptom. On physical examination, the abdomen was soft with no palpable mass. Per rectal examination revealed a large pelvic mass, but due to labial adhesion, the hymen was not seen. Ultrasonography showed a big heterogeneous collection, measuring 11 cm × 6.6 cm × 7.8 cm, in the vagina. Differential diagnoses included a simple imperforate hymen with haematocolpos or transverse vaginal septum. Examination under anaesthesia revealed an imperforate hymen above the labial adhesion. A mini-hymenotomy was performed and 500 ml of old menstrual blood was drained. She was then discharged on postoperative Day 3. At the six-month follow-up, she was found to have no menses after surgery and examination revealed a completely intact hymen with no opening. She then underwent a re-do hymenotomy and had regular menses afterwards.

DISCUSSION
Imperforate hymen is an anomaly, which when presenting during the adolescent period, can usually be diagnosed by thorough history taking and a physical examination. Adolescents typically present with primary amenorrhoea, a cyclic pattern of lower abdominal/pelvic pain, with or without associated symptoms like back pain (38%–40%), urine retention (37%–60%) or constipation (27%).(2-4) On physical examination, a lower abdominal mass may be palpable, or a pelvic mass may be detected on bimanual rectal examination. The diagnosis of imperforate hymen can often be established readily during the perineal examination when a bluish bulging imperforate hymen is found at the introitus. However, the condition can be easily missed if a careful history taking and detailed examination are not carried out, as illustrated by our cases. This highlights the importance of pursuing the basic principles in medicine, viz. thorough history taking and physical examination. In girls presenting with abdominal pain, a careful examination of the introitus, apart from per rectal examination, is mandatory. Imaging or laboratory studies are usually not indicated for a classical presentation of imperforate hymen. However, if the diagnosis is uncertain, as illustrated in the third case, imaging studies would be needed for better surgical planning.

The treatment for imperforate hymen is hymenotomy. Before the procedure, the urethra has to be stented to avoid possible damage during the procedure. Two techniques of hymenotomy are commonly advocated: a simple incision or a small excision of the membrane. In Case 3, a simple cruciate incision of the hymen, without the excision of any part of the membrane, was made in the first operation in an attempt to preserve the traditional Chinese concept of the importance of first-coitus bleed. Acar et al advocated the use of “mini-hymenotomy” (0.5-cm incision) together with keeping a Foley catheter in situ for two weeks.10 However, it is not the universally-accepted method due to the high recurrence rate. It is generally advocated that at least part of the membrane be excised. In addition, the incidence of recurrence might further be reduced by plicating the edge of the incised membrane.

Though the outcome after adequate hymenotomy for imperforate hymen is usually excellent, follow-up is still necessary to ensure that there is no recurrence of the problem. With adequate surgery, symptoms of dyspareunia, abnormal menstruation or persistent problems of micturition/defaecation seldom recur.10 In conclusion, imperforate hymen is not an uncommon cause of abdominal pain and abnormal menstruation in adolescent girls. Without a careful history taking and physical examination, the diagnosis can be missed easily, resulting in a delay in diagnosis and treatment. An adequately-performed hymenotomy usually leads to an excellent outcome.

REFERENCES
Case Report

Imperforate Hymen Causing Bilateral Hydroureteronephrosis in an Infant with Bicornuate Uterus

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A rare case of imperforate hymen associated with bicornuate uterus in an infant is presented as a cause of bilateral hydroureteronephrosis and pelvic mass in infancy. The importance of postoperative radiologic evaluation for diagnosis of accompanying uterine abnormalities is introduced. A 8-month-old girl with restlessness and intermittent fever was brought to the daily outpatient clinic by her parents. Ultrasound exam showed bilateral grade 4 hydroureteronephrosis and a large cystic pelvic mass. Magnetic resonance scan of the pelvis revealed marked hematocolpos. A cruciate incision was made over the hymen under general anesthesia. During a 6-month followup gradual resolution of bilateral hydroureteronephrosis was documented. Although the details of the uterine anomaly were obscured in preoperative imaging, postoperative US and MR demonstrated bicornuate uterus. Postoperative pelvic radiologic examination is highly recommended to verify the resolution of hematocolpos and to screen for any concomitant anomalies that can have long-term clinical significance.

1. Introduction

Imperforate hymen which has an incidence of 0.014%–0.1% is usually asymptomatic until menstruation starts [1–3]. However, under endogenous maternal estrogen stimulation, secretions produced by the fetal uterovaginal mucosa can accumulate in the vagina and uterus resulting in hydrocolpos before puberty. This may cause a mechanical effect on the urethra and bladder and lead to obstructive urinary symptoms. When so, manifestation as a pelvic mass which severely compresses the bladder, and the ureters causing hydroureteronephrosis is rare in infancy [4–6].

Since imperforate hymen is generally considered not to be associated with other Müllerian abnormalities, further investigation of these patients for concomitant urogenital abnormalities has been thought to be unnecessary until today [7].

The aim of this report is to increase the awareness about the possibility of this condition as a cause of bilateral hydroureteronephrosis and pelvic mass in infancy and to introduce postoperative radiologic evaluation for diagnosis of accompanying uterine abnormalities. Informed consent was taken from the patient’s parents, and the case was presented as a poster in 31st National Radiology Congress, Antalya, Turkey in November 2010.

2. Case

A 8-month-old girl presenting with restlessness and intermittent fever of unknown etiology was brought to the daily clinic by her parents. The parents did not complain about any problems related to urination, and there was some amount of daily urine output.

She had been born term after an uneventful pregnancy via normal vaginal delivery. Far to the parents’ knowledge the newborn examination was normal. On physical examination, she had normal vital signs. She was found to have a midline
abdominal mass. The rest of the examination was normal. Initial laboratory values were unremarkable, except for a mild leukocytosis and plenty of erythrocytes in the urine. The urine culture did not reveal any pathological findings. Blood urea levels and creatinine levels were normal.

Abdominopelvic sonography showed bilateral grade 4 hydronephrosis and a large well-circumscribed midline cystic mass including internal echoes. The cyst reached up to the umbilical level (Figure 1(a)). No bladder could be identified on control pelvic ultrasonography (US) exams until the exam was repeated after the insertion of a Foley urethral catheter (Figure 1(b)). Magnetic resonance (MR) scan of the abdomen and pelvis was obtained; it revealed hematocolpos that was causing marked distention of the uterus and cervix. The urinary bladder was significantly compressed (Figure 2). The presumptive diagnosis of hydrometrocolpos secondary to an obstructing lesion was made.

The patient underwent voiding cystourethrogram (VCUG) which demonstrated no reflux but a compressed urinary bladder with a diminished urine volume of approximately 25cc (Figure 3). The diagnosis of imperforate hymen was made under sedation during the instrumentation for the procedure by the inspection of a protuberant mass on retraction of the labia. The family had not noted any perineal abnormalities prior to presentation to the clinic.

The patient was taken into the operating room, and a simple cruciate incision was made over the hymen under general anesthesia which resulted in drainage of approximately 500 mL cloudy, yellowish, nonbloody mucosal secretions from the vagina. No acute or subacute complications occurred.

During a period of 6-month followup, repeated ultrasound exams documented the gradual resolution of bilateral hydroureteronephrosis. The suspicion of bicornuate uterus raised by pelvic control ultrasound was verified by a postoperative MR exam (Figures 4(a), 4(b), and 4(c)).

3. Discussion

Imperforate hymen is an uncommon congenital disorder of the female genital tract [1, 2]. The hymen is an embryological remnant of mesodermal tissue which is supposed to

![Figure 1: Pelvic US. (a) Transverse view shows a large well-defined cystic mass with internal echoes, which could easily be misinterpreted as overly distended urinary bladder. Note the bilateral ureteral dilatation. (b) Insertion of a Foley catheter makes it clear that the cystic mass is separate from the urinary bladder which is severely compressed and therefore hard to detect on ultrasound.](image1)

![Figure 2: Sagittal-T2-weighted MR image. Marked distention of the uterus and cervix is demonstrated. Note the compressed urinary bladder with little urine in it.](image2)

![Figure 3: Preoperative VCUG. Image reveals no reflux but a urinary bladder which could not receive appropriate amount of contrast material due to severe compression secondary to hematocolpos.](image3)
Figure 4: T1-weighed postoperative MR images. (a) and (b): two consecutive pelvic coronal images clearly show the two cavities (arrows) of the uterus separated by an incomplete longitudinal septum which was difficult to depict earlier. (c) Axial view through corpus shows bicornuate uterus.

perforate during the later stages of embryonic development [8]. The usual clinical presentation of imperforate hymen is as an expanding abdominal mass and cyclic lower abdominal or back pain in an adolescent girl with primary amenorrhea. It is rarely diagnosed in the neonatal period or infancy. Hydrocolpos or mucocolpos triggered by endogenous maternal estrogen stimulation rarely presents as bilateral severe hydroureteronephrosis in infancy [4–6].

This particular case did not present as acute urinary retention which would be far more alarming. Less obvious changes in urination can be missed by the family in a child of this age. Imperforate hymen can be hidden under a very nonspecific set of complaints with a broad differential diagnosis, like fever of unknown etiology and restlessness.

In this case ultrasound examination revealed bilateral grade 4 hydronephrosis but was unable to demonstrate the normal pelvic anatomy. It revealed a giant pelvic cystic mass without any change in appearance on more than one ultrasound and which could be easily misinterpreted as a distended urinary bladder since the bladder could not be visualized on either exam. Insertion of a Foley catheter might be helpful to distinguish between a real pelvic mass or urinary bladder overdistension in such cases.

Incorporation of the external genitalia into the newborn nursery exam and well baby examination is highly recommended so that genital anomalies can be diagnosed early. When the diagnosis of imperforate hymen is made in a newborn or an infant, assuming that there are no urinary signs or obstruction, observation throughout childhood and a planned hymenotomy after the onset of puberty and before menarche is optimal. Surgery in the presence of adequate estrogenization avoids scarring and needs to repeat surgery. When there are signs of urinary obstruction or an abdominal mass as in this case, immediate surgery is needed.

Whereas imperforate hymen is a problem that could be easily solved by a minor operation without sequel [9], even though rare [7], any accompanying uterine anomaly like bicornuate uterus as in this case could potentially have a long lasting impact on fertility [10].

Early diagnosis of accompanying genital anomalies would not affect the immediate management but would save time and money on the long range. Postoperative imaging is also recommended for the followup of resolution of findings in response to surgery.

Although preoperative US and MR examination both revealed a very distended uterus in the form of a large cystic mass and the details of the uterine anomaly were obscured, postoperative radiologic imaging was diagnostic. Not every case receives pre- and/or postoperative MR exams. The diagnosis is usually based on clinical examination and preoperative ultrasound. No further information regarding the pelvic anatomy may be obtained.

We suggest postoperative radiologic examination, preferably by pelvic ultrasound since it is more accessible and cheaper, both to verify the resolution of hematocolpos and to screen for any concomitant anomalies that can have long-term clinical significance.

References


CASE REPORT

Imperforate hymen causing congenital hydrometrocolpos

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Abstract A 3-day-old girl in good health was referred to our department for the evaluation of an abdominal mass detected at birth. Prenatal ultrasound (US) examinations had shown no anomaly. US examination revealed the presence of a hypoechoic and corpusculated cystic formation of about 8 × 5 × 4 cm located in the mid region of the abdomen. The uterus was not visible and the kidneys were normal with no sign of hydronephrosis. The ovaries were normal. Physical examination confirmed US findings revealing the presence of a curved membrane which covered the vaginal opening. Based on these findings, the patient was diagnosed to have hydrometrocolpos. Hymenectomy was performed and about 100 ml of milky fluid was subsequently removed by aspiration. The patient presented no other congenital anomalies and US follow-up showed a normal structure of the uterus.

Keywords Imperforate hymen · Congenital hydrometrocolpos · Pelvic mass · Ultrasound

Introduction

Hydrometrocolpos is an accumulation of uterine and vaginal secretions as well as menstrual blood in the uterus and vagina. Usually this condition manifests at puberty caused by an obstruction of the female genital tract. The most frequent cause of hydrometrocolpos is the presence of imperforate hymen due to failure of partial resorption of this membrane during the embryonic development; the incidence is 0.0014–0.001 % in full-term newborns [1, 2]. Congenital hydrometrocolpos is a rare event with an incidence of about 0.006 % [3]. We present the US features of hydrometrocolpos in a newborn girl before and after surgical treatment.

Description of the case

A 3-day-old girl born by Cesarean section at 38 weeks of gestation was referred to our department for the evaluation of an abdominal mass detected at US examination performed on the day of birth. The patient was otherwise in good condition and weighed 3.330 kg. Prenatal US examinations showed no anomaly.
US examination was carried out on GE Logiq 9 using a micro-convex 5–9 MHz probe. The image showed the presence of an inhomogeneous hypoechoic and corpusculated cystic formation of about $8 \times 9 \times 5$ cm with well-demarcated margins; it was located in the mid region of the abdomen extending from the upper middle to the lowest region (Figs. 1, 2). The bladder was empty and compressed, the kidneys and ovaries were normal. The uterus was not visible.

Hydrometrocolpos was suspected and subsequently confirmed at physical examination of the external genitalia, which revealed the presence of a soft oval mass with an imperforate hymen at the vaginal opening. After incision of the hymenal membrane, approximately 100 ml of milky fluid was aspirated. Subsequent US imaging showed normal appearance of the uterus and vagina with no signs of other congenital anomalies (Fig. 3).

Discussion

Hydrometrocolpos is an unusual finding in newborn infants. It occurs when a genital tract obstruction is associated with accumulation of cervical and endometrial gland secretions.

This condition may be caused by congenital malformations of the genital tract such as vaginal atresia, transverse vaginal septum and imperforate hymen. It may also be associated with the McKusick–Kaufman syndrome, an autosomal recessive disorder characterized by vaginal atresia with hydrometrocolpos, polydactyly, congenital heart defects and non-immune mediated hydrops fetalis [4]. In the present case, imperforate hymen was the cause of hydrometrocolpos.

Imperforate hymen is a result of the hymen failing to rupture during the eighth week of gestation; it may be an isolated abnormality or associated with other malformations, such as imperforate anus, bifid clitoris, polycystic kidney.

As described in other cases [5], hydrometrocolpos is usually diagnosed prenatally as the cause of abdominal cystic mass. However, in the present case prenatal examinations were all negative.

Differential diagnosis of a perinatally identified abdominal mass should include ovarian cysts, intra-abdominal sacrococcygeal teratoma (type IV), neuroblastoma, mesoblastic nephroma, bowel duplication, genital-urinary anomalies and anterior sacral meningocele.

Hydrometrocolpos may cause urinary stasis and acute renal failure due to obstructive uropathy [6, 7]. In the present case, physical and US examinations showed that

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**Fig. 1** Longitudinal scan reveals a large hypoechoic mass in the mid region of the abdomen extending from the upper middle to the lowest region

**Fig. 2** Axial US scan shows the mass and the right ovary which is intact

**Fig. 3** Longitudinal pelvic US scan obtained after drainage
both kidneys were normal with no sign of hydronephrosis and that there were no other congenital anomalies.

In hydrometrocolpos caused by imperforate hymen, hymenectomy has proved to be an adequate, conservative treatment [8]. A peculiar phenomenon was in the present case the amount of fluid removed (about 100 ml).

In conclusion, although hydrometrocolpos is a rare event, this disorder should be kept in mind if pre- and/or postnatal examinations reveal the presence of a pelvic mass.

Conflict of interest The authors have no conflict of interest to declare.

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Imperforate Hymen

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Background

Imperforate hymen is at the extreme of a spectrum of variations in hymenal configuration. Variations in the embryologic development of the hymen are common and result in fenestrations, septa, bands, microperforations, anterior displacement, and differences in rigidity and/or elasticity of the hymenal tissue. Inspection of the external genitalia and anus are important components of the physical examination of the female neonate and child.[1]

While this examination can and should be accomplished by the pediatrician, the observant delivering obstetrician can learn much about the normal variations in genital configuration by examining the female neonate in the delivery room, keeping in mind the influence and structural changes induced by maternal estrogens. Under this influence, the labia majora are plump, the hymen is elastic and often fimbriated, and the mucosal surfaces (ie, introitus, fossa navicularis, vaginal vestibule) are pale pink.

Problem

Imperforate hymen has been diagnosed with prenatal ultrasound documentation of bladder outlet obstruction due to hydrocolpos or mucocolpos. However, in spite of the recommendations for inspection of the external genitalia during the neonatal and early childhood period, variations in hymenal anatomy commonly escape diagnosis until the time of menarche. See the image below.

Imperforate hymen, classic appearance of bulging, blue-domed, translucent membrane.

Different normal variants in hymenal configuration are described, varying from the common annular, to crescentic, to navicular ("boatlike" with an anteriorly displaced hymenal orifice). Hymenal variations are rarely clinically significant before menarche. In the case of a navicular configuration, urinary complaints (eg, dribbling, retention, urinary tract infections) may occasionally result. Sometimes, a cribiform (fenestrated), septate, or navicular configuration to the hymen can be associated with retention of vaginal secretions and prolongation of the common condition of a mixed bacterial vulvovaginitis.

Occasionally, a hymenal tag will protrude from the vaginal vestibule, leading to concerns about a tumor or other significant pathology. These hymenal tags are of no clinical significance, and they do not require therapy if hymenal origin can be excluded based on findings from a careful examination.

Imperforate hymen in infancy or childhood

On occasion, an infant or young child may be thought to have an imperforate hymen. However, after the neonatal period, when maternal estrogen levels have declined, examination of the area may be challenging owing to the small area involved. Careful examination with pressure applied to the fourchette may reveal microperforations, sometimes with an anteriorly displaced opening just beneath the urethra. Capraro described a surgical technique similar to a perineotomy to correct such a defect; however, in asymptomatic patients, waiting until puberty is generally recommended before deciding whether such a technique is necessary.

The hymenal changes that result from estrogenization (increased elasticity and fimbriation) may reveal the hymen to be open and obviate the need for surgery. With estrogen stimulation, the hymen could be described as having the appearance of an annular "scrunchie" (ie, a fabric-covered elastic hair tie). In addition, surgical procedures to
the vagina and hymen during childhood, when endogenous estrogen levels are low, may result in scarring and the need for subsequent surgical revision. Thus, surgery during this time should generally be avoided if possible. If the hymen is suspected to be imperforate during childhood, re-examination should be performed after the onset of estrogen production, as signaled by breast development. If required, surgery can be performed at this time when healing is optimal and prior to the accumulation of a hematocolpos.

In a review of 23 cases of imperforate hymen, Posner et al emphasizes the ease of making a diagnosis of imperforate hymen by routine genital examinations in childhood.[2] The authors compared the significant delays and difficulties in making the diagnosis after the onset of puberty, primarily because the diagnosis was not considered, with the simplicity of making the diagnosis in asymptomatic prepubertal children by a simple genital examination.

Sexual abuse

Accurate description of the morphology and integrity of the hymen is critical in the diagnosis of female sexual abuse. Imperforate hymen has been described as occurring as a result of scarring from penetration and abuse, thus emphasizing the importance of an early examination to document the congenital, rather than acquired, etiology.[3] Concerns about hymenal disruption and lacerations associated with sexual abuse with digital or penile penetration have led to discussions of the normal hymenal diameter. However, this concept has now largely been abandoned.[4]

Experts in sexual abuse assessment have used unaided visual examination and colposcopy to examine the integrity of the hymenal ring. A normal examination or nonspecific findings are commonly found in cases of alleged sexual abuse unless the abuse is quite recent.[5]

Lacerations through the hymen into the fossa navicularis and introitus suggest a penetrating injury. Frequently, sexual abuse evaluations are conducted at some time remote from the immediate injury; thus, normal findings or healed or healing lacerations may be noted.

Muram concluded that the use of the colposcope by an experienced examiner adds little to an evaluation by an experienced examiner with expertise in abuse.[6] In addition, Muram proposed a scale that the examiner can use to evaluate physical findings as normal, abnormal and nonspecific, abnormal and suggestive of abuse, and definitive for abuse.[6] That last category includes only the situation in which sperm are found during the examination. Additional aids to the examination of the hymen have been described, including the procedure of inserting a Foley catheter into the vagina and inflating the balloon behind the hymen to stretch the hymenal margin and allow for a better examination.[7]

Anatomic anomalies

The classic image of an imperforate hymen is noted at the time of typical diagnosis: after the onset of menses, when a hematometocolpos is present (see the image below).

Consider anatomic anomalies that can be confused with imperforate hymen in the differential diagnosis. These anomalies include the following:

- Acquired labial adhesions (see image below)
Extensive labial adhesion. Not to be confused with imperforate hymen.

- Obstructing or partially obstructing vaginal septa (longitudinal or transverse)
- Vaginal cyst
- Vaginal agenesis (Mayer-Rokitansky-Kuster-Hauser syndrome) with or without the presence of a uterus or functional endometrium (see image below)

Vaginal agenesis. Not to be confused with imperforate hymen.

- Complete androgen insensitivity syndrome (previously termed testicular feminization)

Epidemiology

Frequency

Imperforate hymen is likely the most frequent obstructive anomaly of the female genital tract, but estimates of its frequency vary from 1 case per 1000 population to 1 case per 10,000 population. A population-based study estimated the frequency at 0.5 case per 1000 women (95% confidence interval, 0.3-0.7).\(^8\)

Heger et al examined 147 premenarchal girls with a mean age of 63 months to collect normative data on genital anatomy; an imperforate hymen was found in only one patient (< 1%) and hymenal septa were found in 3 (2%).\(^9\)

Imperforate hymen usually occurs sporadically, but a handful of cases have been reported to be familial.\(^{10, 11}\)
Examination of first-degree relatives/female siblings of affected individuals has been recommended.

Etiology

Imperforate hymen and related genital tract anomalies result from abnormal or incomplete embryologic development.

Pathophysiology

The genital tract develops during embryogenesis, from 3 weeks' gestation to the second trimester. The initial development of both the male and female genital tracts is identical and is referred to as the indifferent stage of

Prior to birth, the müllerian ducts are paired and a solid evagination from the distal aspects of the müllerian tubercle forms the sinovaginal bulbs (of urogenital sinus origin) or vaginal plate. The initial or cephalad portion of the müllerian ducts forms the fimbria and fallopian tubes; the more distal segment forms the uterus and upper vagina. The canalization of the sinovaginal bulbs and/or upper vagina joins with the vaginal plate, which canalizes beginning caudally and creates the lower vagina. By the fifth month of gestation, the canalization of the vagina is complete. The hymen itself is formed from the proliferation of the sinovaginal bulbs, becoming perforate before or shortly after birth. An imperforate hymen results when this "sheet" of tissue fails to completely canalize. Varying degrees of perforation result in findings such as a cribriform or septate hymen.

Gonadal development

The development of the gonads occurs from the migration of primordial germ cells to the genital ridge, while the genital tract itself develops from the müllerian ducts (paramesonephric ducts), urogenital sinus, and vaginal plate. Thus, anomalies of the vagina, hymen, and uterus are not accompanied by abnormalities of ovarian development. In girls with hymenal anomalies, hormonal and endocrinologic function is normal, leading to expected pubertal breast and pubic hair development. In cases of urorectal agenesis, imaging may fail to detect ovaries in the normal location (they may be located high and/or lateral in the pelvis), leading to unnecessary concern that the ovaries may be absent. Patients and families can be easily reassured that given both embryologic development and normal hormonal function (evidenced by the presence of normal breast development), the ovaries are present and functioning appropriately.

Because the mesodermal layer contributes to the development of the kidneys, gonads, and ductal structures, defects or insults in embryologic development may result in congenital defects of the kidneys or ureters that accompany abnormalities of the vagina and uterus. These anomalies should be considered with vaginal and uterine anomalies. However, given the embryologic origins of hymenal anomalies, urologic abnormalities are not associated.

The lining of the urethra and urinary bladder derives from endoderm, and the urogenital sinus forms the urethra and vestibule in females. The ectoderm fuses with the endoderm to contribute to the patency and canalization of the genital tract. Defects in this process lead to fusion failures and imperforate and obstruction defects.

Familial occurrence

Familial occurrence, although rare, is reported and screening by history or examination of family members is warranted. Dominant transmission (either sex-linked or autosomal) and sibships suggesting a recessive mode of inheritance are described. The inheritance of müllerian defects likely is polygenic or multifactorial, although some syndromes of heritable disorders are described with associated genital and nongenital anomalies.

Anomalies of the female reproductive tract

Anomalies of the female reproductive tract can result from agenesis or hypoplasia, vertical fusion and/or canalization defects, lateral fusion and/or duplication abnormalities, or failure of resorption, resulting in septa. Recent reports have noted the concurrent presence of lateral fusion defects with imperforate hymen.

Presentation

Prenatal diagnosis

Rarely, diagnosis of imperforate hymen in the fetus has been made with obstetric ultrasonography. In such cases, the anomaly is visible on the imaging study because of hydrocolpos, hydrometrocolpos, or mucocolpos.

Diagnosis in infancy or childhood

The diagnosis is infrequently made during infancy in the neonatal nursery. The infant may have a bulging, yellow-gray mass at or beyond the introitus. Several case reports describe the presence of an abdominal mass in association with urinary obstruction.

Ultrasonography is an essential first step in diagnosis, precluding unwise and unplanned surgical intervention with resultant injury to the urethra or other pelvic structures, and excluding other more complicated anomalies.

Routine examination of the female genitalia by primary care clinicians during childhood is strongly recommended so that genital abnormalities can be diagnosed early. Observation throughout childhood, with a planned hymenotomy after the onset of puberty is a reasonable course of action in most cases diagnosed in infancy or childhood, assuming no urinary symptoms or obstruction is present. Surgery in the presence of adequate estrogenization avoids scarring and the potential need for a repeat surgery that can occur to correct scarring when surgery is performed on the unestrogenized hymen and vagina.
If the diagnosis is equivocal (i.e., imperforate hymen vs labial adhesions vs late-onset congenital adrenal hyperplasia), referral to a pediatric gynecologist may be warranted. Typically, a mucocele is not present even if the condition is noted at birth. If a patient is diagnosed with an asymptomatic imperforate hymen in infancy or childhood beyond the neonatal period, the optimal time for surgical repair is after the onset of puberty and prior to menarche.

**Diagnosis and surgical repair in adolescence**

Diagnosis of imperforate hymen depends on an awareness of the condition as a possible anomaly and surveillance with well-child care. The typical presenting complaint is primary amenorrhea, but this is a late presentation of a condition that should have been diagnosed at an earlier time. Textbooks frequently state that amenorrhea is not pathologic until age 16 years. Statistically, this statement is not evidence-based, as age 15 years represents the 98th percentile for girls in the United States and other developed countries.[16]

Additionally, failure to menstruate beyond 2-3 years from the onset of breast development, thelarche, is also statistically uncommon, and should be investigated to determine a cause. Imperforate hymen is one uncommon, but important, anatomic cause of primary amenorrhea.

When the condition presents as abdominal pain or an abdominal mass (see image below), diagnostic testing is often extensive because the condition is not considered. An abdominal mass may prompt the consideration of an ovarian tumor and tumor markers may be obtained. While a false-positive elevation of CA-125 in premenopausal women has numerous causes, and testing has thus been discouraged, elevated CA-125 and 19.9 have been described with imperforate hymen, and may delay the diagnosis.[17, 18]

![Abdominal mass with imperforate hymen.](image)

Surgical repair after the onset of puberty but before menarche is optimal. The most common scenario is that in which a young woman presents with increasingly severe intermittent abdominal and pelvic pain due to a large hematocolpos and hematometra. This situation is preventable, as routine examinations of the genitalia can detect this obstruction and allow correction before menarche.

Walsh and Shih present a case of a 14-year-old elite athlete who presented to the emergency department and her pediatrician on multiple occasions over the course of several months with symptoms of cyclic abdominal pain, urinary retention, and constipation due to hematocolpos and hematometra.[19] This is an all too common presentation. In this reported case, even after placement of a Foley catheter for urinary retention on 2 separate occasions, the diagnosis of imperforate hymen was missed.

While these young adolescents typically present to an emergency department with relatively acute pain, this condition should generally not be managed emergently until the definitive diagnosis is made. Defining the anatomy with appropriate imaging techniques and arranging for the most skilled and experienced gynecologist to perform surgery on a scheduled rather than emergent basis is essential. If necessary, menstrual suppression with gonadotropin-releasing hormone (GnRH) analogs can minimize pain pending appropriate imaging and clarification of anatomy. This is more likely to be necessary with complex genital anomalies than with a straightforward imperforate hymen.

Urinary pressure and even retention, with hydrouretere and/or hydronephrosis, may occur due to the mass effect and resultant obstruction. Vaginal and rectal pressure is typically present. Severe constipation and low-back pain are described as presenting symptoms. The laborlike menstrual cramps may be severe and cyclic, although the cyclic nature of the symptoms may not be easily or immediately appreciated by the young woman or her family.

Unfortunately, the typical findings at diagnosis may include a large collection of blood within the uterus (hematometra) and an even larger collection of blood within the more distensible vagina (hematocolpos). Additional findings may include blood-filled fallopian tubes (hematosalpinges) and signs of retrograde menses, occasionally to the point of the development of intra-abdominal endometriosis and severe pelvic adhesions. The classic teaching is that endometriosis associated with obstructive anomalies resolves spontaneously and does not cause problems with subsequent pain and infertility compared with endometriosis arising spontaneously; however, this assertion is anecdotal rather than evidence-based.

Clinically, families are often concerned about whether the ovaries are normal when vaginal or hymenal anomalies are present; the course of separate embryologic development allows assurance of normal hormonal function without any need for hormonal testing or ovarian imaging. The exception to this is the diagnosis of androgen insensitivity syndrome with XY chromosomal complement in which the gonads require removal to prevent malignant transformation.

**Differential diagnosis**
The differential diagnosis of an imperforate hymen includes many conditions, some rare and others relatively common. Absolute confirmation of the diagnosis of an imperforate hymen is imperative prior to any attempted surgical repair in order to prevent vaginal scarring that can occur if a thick vaginal septum is inadvertently confused with a thin imperforate hymen.

Labial adhesions

The presence of acquired labial adhesions in a prepubertal girl is a common situation that is often confused with absence of the vagina. Labial adhesions, sometimes incorrectly termed vaginal adhesions, are not congenital and result from acquired labial agglutination most commonly due to inflammation. Small areas of labial adhesions can be managed expectantly. Extensive labial adhesions or those associated with such symptoms as recurrent urinary tract infections, urinary dribbling, or recurrent vulvovaginitis can be managed easily using the topical application of estrogen cream for 2-6 weeks. Such treatment results in marked thinning of the adhesions, often with spontaneous resolution.

Separation of thick adhesions is possible in an office setting with a child who can be restrained; however, this procedure ultimately is counterproductive because the examination frequently is difficult and traumatic, resulting in the subsequent inability to adequately examine the genital area due to the child's refusal because of memories of pain. Such traumatic lysis should be avoided. General anesthesia in an operative setting may thus be required.

Management of labial adhesions can be problematic as recurrence is common. Parents or caretakers must be instructed on how to ensure the child maintains excellent perineal hygiene and avoids vulvovaginitis. Families are often incorrectly encouraged to avoid baths in favor of showers; while bubble-baths may occasionally contribute to vulvar inflammation, a plain water bath with soaking and cleansing of the interlabial folds using a washcloth without soap is preferable to a shower, which makes interlabial cleansing more difficult. The daily application of a topical emollient (such as A&D ointment) helps reduce the risk of recurrence until endogenous pubertal estrogen stimulation alleviates the risk. Thus, the application of a topical emollient should be continued until the child shows signs of estrogen-stimulated breast development.

Rarely, an adopted child will be found to have what appears to be labial adhesions, and these may be suggestive of female genital mutilation that occurred at a young age. The thick adhesions that result from this trauma may require surgical separation and management by a gynecologist with experience in managing female genital mutilation.

Labial adhesions may be confused with posterior labial fusion encountered in persons with congenital adrenal hyperplasia and may be differentiated by careful physical examination with attention to the presence or absence of clitoromegaly. This abnormality is noted at birth, rather than acquired.

The differential diagnosis for a cystic mass at the hymen includes ectopic ureter, hymenal cyst, hymenal skin tag, periurethral cyst, and vaginal cyst.[20]

Incomplete hymenal obstruction

In the case of incomplete hymenal obstruction due to a cribiform hymen or hymenal band, the typical presenting symptom is difficulty inserting a tampon or even the inability to achieve vaginal intercourse in an adolescent. Anatomic variations must be distinguished from involuntary vaginismus or contraction of the perineal and pelvic musculature or levator ani muscles, which can be associated with the learning process of tampon insertion, becoming a vicious cycle when persistent insertion is attempted without success and causes pain.

Hymenotomy occasionally may be indicated in the case of a rigid inelastic hymen, particularly for young female athletes (eg, swimmers, divers, gymnasts, cheerleaders) who may be hypoestrogenic, leading to the rigid hymenal configuration. As athletes, these girls are often eager to use tampons. A reasonable alternative to surgical correction involves the use of progressive dilation in a motivated young woman, along with topical estrogen. In these athletes with a rigid hymen, an evaluation for hypoestrogenism associated with overly vigorous physical activity should be considered; if present, estrogen replacement improves the hymenal characteristics and increases hymenal elasticity.
Hymenal bands

This condition is typically amenable to division using a local anesthetic in the office; however, the young woman's age and tolerance of such an office procedure must be predicted and judged. Her degree of motivation for tampon use or intercourse impacts the timing at which she requests such a procedure. A typical presenting history of an individual with a hymenal band is the ability to insert a tampon but extreme difficulty removing it. The author has encountered a patient in whom the tampon string became wrapped around the hymenal band, leading to marked edema and pain when removal was attempted.

Obstructing longitudinal or transverse septa

These conditions require careful preoperative evaluation to define the anatomy prior to any attempted surgical reconstruction. The repair of such complicated anomalies should usually be referred to a gynecologist at a tertiary care center where these cases are not a rarity. MRI is the optimal imaging modality for defining complicated female reproductive anatomy.[21]

Vaginal agenesis or androgen insensitivity

The evaluation and management of vaginal agenesis or androgen insensitivity syndrome is beyond the scope of this article, but these conditions should be considered in the differential diagnosis. Like imperforate hymen, primary amenorrhea is typically the presenting complaint.

Androgen insensitivity is diagnosed based on findings of a blind vaginal pouch, with an XY chromosomal complement. Mayer-Rokitansky-Kuster-Hauser syndrome (uterovaginal agenesis) may include uterine remnants, some containing endometrium as well as myometrium. These patients should be referred to a gynecologist who specializes in adolescents and who has experience in managing these conditions.

Others

The presentation of an abdominal mass must be differentiated from urinary obstruction or tumors such as sacrococcygeal teratoma with abdominal extension, ovarian tumor, or other masses like mesenteric cysts or anterior meningoceles.[20]

Neovagina options

The options for creation of a neovagina are nonoperative (preferred approach) or operative, such as a McIndoe, Davydov, Vecchietti, or Williams procedures.[22]

Nonoperative management, using progressively larger Lucite dilators, is generally thought to be the first-line approach to management. Nonsurgical management at a time when the young woman is motivated to use vaginal dilators minimizes the potential for scarring and has high rates of success.[22]

Coital dilation has also been described as a successful management strategy.

Indications

An imperforate hymen at the time of puberty must be corrected surgically. The surgical decision-making process should focus on appropriate diagnosis and timing of surgical repair. While the patient may present with acute pain, the repair should not be performed emergently before carefully defining the anatomy. The surgery should be performed by a gynecologist who is skilled and experienced in the care of adolescents with genital anomalies.

Relevant Anatomy

An imperforate hymen presenting after the onset of menstrual shedding is visible upon examination as a translucent thin membrane just inferior to the urethral meatus that bulges with the Valsava maneuver. This bluish discoloration is due to the presence of a hematocolpos visible behind the translucent hymenal membrane. Vaginal septa do not typically appear translucent (see the image below).
Depending on the size and volume of the hematometra, hematocolpos, or hematosalpinges, a pelvic or abdominal mass may be palpable during abdominal or rectal examination. See the image below.

Radiographic documentation must demonstrate that the true diagnosis is not an obstructing transverse vaginal septum or other anomaly. Pelvic ultrasonography via the transabdominal, transperineal, or transrectal route is indicated as the initial diagnostic test, followed by MRI if any questions remain about the anatomy. Transperineal ultrasonography can be helpful in measuring the thickness of the septum. Because renal and urologic abnormalities are associated with müllerian abnormalities, imaging of the upper urinary tract can help diagnose ipsilateral renal agenesis, duplex collecting systems, and other complex renal anomalies if there are uterovaginal anomalies.

The prevalence of renal agenesis is estimated at 1 case per 600-1200 persons in patients with müllerian anomalies on the basis of autopsy studies. As many as 25-90% of women with renal anomalies are suggested to have concurrent genital anomalies; thus, abdominal and pelvic imaging of these patients is also warranted for these patients.

Contraindications

The contraindications for a surgical repair of an imperforate hymen relate to the surgeon's inexperience with this condition, failure to adequately consider the alternative diagnoses, or failure to carefully define the anatomy.

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Introduction

Imperforate hymen, as its name implies, is a condition where the hymen, a thin membrane in the shape of a half moon, covers the entire opening of the Vagina [1]. This condition, although rare, is the most common female genital tract malformation with a prevalence of up to 0.1% [2,3]. Diagnosis can go undetected until a patient starts experience one, or many, of a variety of symptoms.

Symptoms can range from mild abdominal pain and tenesmus to urinary retention and hematocolpos, a mass that forms due to the accumulation of menstrual blood that cannot leave the vaginal cavity. In this case, we present a young female with cyclical abdominal pain and difficulty urinating, to urinary retention and tenesmus. Ultrasound is the study of choice for further evaluation and definite treatment is via hymenotomy.

The Case

Case of an 11 year-old pre-menarchal female with lower abdominal and pelvic pain for four days after moving some boxes in her bedroom. Associated to amenorrhea, increased urinary frequency and palpable abdominal non-pulsatile mass that extended above the umbilicus (Figure 1). She denied any poor oral intake, nausea, vomiting or problem with bowel movements. She had never been evaluated by a gynecologist. Upon further history, her sister had menarche at 10 years old and mother also had menarche at 10 years old.

Abstract

Introduction: Imperforate Hymen is the most common female genital tract malformation with prevalence of up to 0.1% and can present with wide variety of symptoms, ranging from abdominal pain to urinary retention.

Case: Case of 11 y/o female with cyclical abdominal pain and palpable mass up to umbilicus. Patient diagnosed with imperforate hymen and taken to operating room (OR) by Gynecology team for hymenotomy; 2,500 mL of blood was evacuated.

Discussion: Imperforate Hymen is an uncommon cause of abdominal pain. Presentation varies from abdominal pain and difficulty urinating, to urinary retention and tenesmus. Ultrasound is the study of choice for further evaluation and definite treatment is via hymenotomy.

Conclusion: Imperforate hymen is an easily missed diagnosis in the Emergency Department (ED). It has to be included in the differential diagnosis for abdominal pain in pre-menarchal females.

Figure 1: Protuberant abdomen due to enlarged mass.

Figure 2: Sagittal view from abdomino-pelvic CT scan showing huge abdomino-pelvic mass.
Discussion

Imperforate hymen is a rare congenital anomaly reported at an approximate rate of 0.1% and occurs due to the incomplete canalization of the Mullerian system and the urogenital system [1]. In the embryological period, the lateral portion of the hymen originates from a fold of urogenital sinus at the union of the Mullerian ducts, whereas in its posterior part, it originates from the cells of the urogenital sinus externally and from Mullerian ducts internally. Usually in the eighth week of gestation, it partially ruptures in the inferior part of the Mullerian ducts, remaining as a fold of mucous membrane around the entrance of the vagina. Failure to partially rupture results in a persistence of the septum, which can be diagnosed as imperforate hymen clinically [5].

Imperforate hymen is an isolated abnormality, where diagnosis should ideally be done at birth by careful examination of the external genitalia of all newborn females [2-17]. During the neonatal period, it may present with fetal ascites or acute renal failure. The hematocolpos or hydrocolpos may lead to variable degrees of hydrorrhea, and hydronephrosis. If the diagnosis is not made in the newborn period and the hymen remains imperforate, the mucus will be reabsorbed and the child usually remains asymptomatic until menarche [10]. At menarche, usually between 9 to 13 years of age, the child starts getting cyclic abdominal pains associated with primary amenorrhea. Retained blood in the vagina, uterus, and fallopian tubes can result in hematocolpos, hematometra and hematosalpinx. Hematocolpos gets worse with each menstrual period [17].

Over thirty cases of hematocolpometra due to imperforate hymen have been reported [2-17]. Most of these cases present with cyclic abdominal pain, but presentation can vary widely, from low back pain [15] to acute urinary retention [3,5,9,11,12,16] and tenesmus [2,12]. Of the 20 cases being reviewed [2-17], 55% experienced urinary retention as a result of mass effect. Diagnosis is done through a thorough history and physical exam, which needs to include genitalia; something that is not always performed. Most of these reported cases presented in adolescence upon menarche. The youngest patient was a 3-month-old girl who had suffered from repeated urinary tract infections because of urinary retention related to pyocolpos [10]. Menarche typically occurs within 2 to 3 years after the larche (breast budding), at Tanner stage IV breast development, and is rare before Tanner stage III development [18-20]. Also, upon evaluation of the vagina, a bluish membrane will be seen protruding the vaginal introitus.

If patient or parents refuse genital exam evaluation, imaging studies should ideally be done at birth by careful examination of the external genitalia of all newborn females [2-17]. During the neonatal period, it may present with fetal ascites or acute renal failure. The hematocolpos or hydrocolpos may lead to variable degrees of hydrorrhea, and hydronephrosis. If the diagnosis is not made in the newborn period and the hymen remains imperforate, the mucus will be reabsorbed and the child usually remains asymptomatic until menarche [10]. At menarche, usually between 9 to 13 years of age, the child starts getting cyclic abdominal pains associated with primary amenorrhea. Retained blood in the vagina, uterus, and fallopian tubes can result in hematocolpos, hematometra and hematosalpinx. Hematocolpos gets worse with each menstrual period [17].

References


Imperforate hymen: a cause of abdominal pain in female adolescents

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Abstract

A 16-year-old girl presented with primary amenorrhea and had had cyclical abdominal pain for almost a year. At examination we observed a painful mass in the lower abdomen and normal secondary sex characteristics. Perineal examination showed a bluish bulging hymen. Transabdominal ultrasonography revealed a dense mass in the pelvis measuring about 12×11 cm. We diagnosed an imperforate hymen with haematocolpos and haematometra. The hymen was opened surgically and a large quantity of menstrual blood was drained from the vagina and uterus. Postoperative recovery was normal without any pain. The patient now menstruates regularly. An imperforate hymen occurs in 0.05% of women. It is important to be aware of this while examining a female adolescent presenting with cyclical abdominal pain and primary amenorrhea. Late discovery of an imperforate hymen may lead to pain, infections, hydronephrosis and endometriosis with subfertility as a possible consequence.

BACKGROUND

Imperforate hymen is a rare cause of abdominal pain in female adolescents. It is seen in approximately 1 in 2000 females, although information on the true incidence is difficult to obtain. The lack of menses in an adolescent girl of (post-)menarchal age with cyclical abdominal pain, urinary retention, constipation and/or a (symptomatic) lower abdominal mass suggests this condition should be considered. This case illustrates that there is quite often a considerable delay before a proper diagnosis is reached. The aim of this communication is to increase awareness of imperforate hymen among clinicians examining adolescent girls with lower abdominal pain.

CASE PRESENTATION

A 16-year-old girl was admitted with a history of lower abdominal pain. She was asymptomatic until a year previously. She then started developing cyclical crampy pain in the lower abdomen, which lasted for 7 days every month. The pain had become more severe during the previous 2 months and the size of her abdomen had increased over the past few months. There was no history of nausea, vomiting, fever, altered bowel habits or problems with urinating. She was 13 years old and had not yet had a menstrual period, but did have pubic hair and breast buds, confirming the onset of puberty. The patient denied any vaginal discharge and there was no history of sexual activity. No other members of her family had similar or other physical complaints. The medical history was unremarkable.

INVESTIGATIONS

On physical examination, the secondary sex characteristics, such as pubic hair and breast buds, were well developed. A mobile, non-tender mass, arising from the pelvis to the belly button, was felt in the abdomen (fig.1,
Gynaecologic examination revealed a bulging bluish hymen (fig 2). No other gross external abnormalities of the external genitalia were observed. A pelvic ultrasound showed a homogenous, hypoechoic mass in the vagina and uterus measuring about 12×11 cm. Both the ovaries were normal.

DIFFERENTIAL DIAGNOSIS

We diagnosed an imperforate hymen with consequent accumulation of blood in the vagina and uterus (haematocolpos and haematometra, respectively).

TREATMENT

Two days later the patient underwent a hymenectomy with a cruciate incision. Approximately 500 ml of viscous, chocolate coloured blood and clots were drained (fig 3).

OUTCOME AND FOLLOW-UP

Immediately after the operation the size of the abdomen had regained normal proportions (fig 1, lower image).

During a 2-month follow-up, the patient was asymptomatic and started to have regular menstrual cycles.

DISCUSSION

Imperforate hymen is a rare though serious cause of abdominal pain in female adolescents. It is seen in approximately 1 in 2000 females.\(^1\) The hymen is an embryological remnant of mesodermal tissue that normally perforates during the later stages of embryo development. The hymen is called imperforate if there is no perforation of this membrane.

The reason for non-perforation of this membrane is unknown. Imperforate hymen occurs mostly in a sporadic manner, although some familial occurrences have been reported. Both the recessive and dominant modes of transmission have been suggested, but no genetic markers or mutations have been proven as aetiological factors.\(^5\) An imperforate hymen in younger children (<10 years) is discovered by chance in 90% of cases, while 100% of affected adolescents first present with symptoms.\(^2\)

The most common symptoms of an imperforate hymen are cyclical abdominal pain and urinary retention, usually presenting between the ages of 13 and 15 years (when menarche occurs).\(^7\) There is primary amenorrhea but secondary sex characteristics are well developed. Because the vaginal outflow is obstructed by the non-perforated hymen, menstrual blood accumulates in the vagina (haematocolpos) and the uterus (haematometra). This may lead to mechanical effects on the urethra, bladder, intestines or pelvic blood vessels which can result in urinary retention, obstipation or oedema of the legs.\(^2\) Irritation of the sacral plexus or nerve roots can cause lower back pain.\(^8\) Problems with intercourse are rarely mentioned, probably because most of the patients are still sexually inactive.\(^15\)

Some symptoms of appendicitis are similar to those of an imperforate hymen, and there are cases where groundless appendectomies have been performed.\(^16\) In addition, there is often an initial diagnosis of infection of the bladder, nephrolithiasis or abdominal tumour, which leads to unnecessary examinations and treatment. The history and physical examination are frequently incomplete.\(^2\) One should always consider an imperforate hymen if there is a discrepancy between the Tanner stage and menarcheal status.\(^2\)

Gynaecological examination reveals a bluish bulging hymen and generally an abdominal mass. The diagnosis can be established with an abdominal ultrasound showing the pelvic cystic mass. Imperforate hymen should be differentiated from a low transverse vaginal septum using the Valsalva manoeuvre: an imperforate hymen should bulge and a transverse vaginal septum should not.\(^4\) Imperforate hymen is usually not associated with any other Müllerian abnormalities and extensive investigation for urogenital anomalies is unnecessary.\(^4\)
Early diagnosis of an imperforate hymen is important, since it can lead to serious complications such as infections, hydronephrosis, kidney failure, endometriosis and subfertility. A retrospective study showed that eight of nine patients with haematocolpos or haematometra due to an outflow obstruction had endometriosis at the time of intra-abdominal operation. Treatment of haematocolpos or haematometra due to an imperforate hymen is to make cruciate incision in the hymen which allows the accumulated blood to drain away. This has to be done aseptically as a closed vagina lacks protective Doederlein’s bacilli and the pH is alkaline or weakly acidic; there is a poor natural resistance to bacteria entering from the lower genital tract and the blood and debris provide a good culture medium. The complications of a hymenectomy are bleeding, scarring and stenosis of the vaginal opening. Less invasive treatments for an imperforate hymen include the use of CO₂ lasers or a Foley catheter.

In a retrospective study of the long term results of surgical correction of imperforate hymen, nine of 15 patients had an irregular menstrual cycle and six of 15 patients had dysmenorrhea (over a follow-up of 8.5 years). Most patients had no sexual dysfunction. Pre-operative complaints of cryptomenorrhea (n=15), abdominal pain (n=11), palpable mass in the lower abdomen (n=9), urinary retention (n=6), dysuria (n=3) and problems defecating (n=4) almost all disappeared after surgery. Eight patients worried about their future fertility; two of them were attempting pregnancy and were successful. Another study showed that 86% of patients who attempted pregnancy succeeded after surgical correction of imperforate hymen.

**LEARNING POINTS**

- In adolescent girls with cyclical pain, it is very important to take a complete gynaecological history and perform a gynaecological examination as serious complications may follow an unnecessary delay in diagnosis.
- Consider an imperforate hymen when there is a lack of menses in an adolescent girl with cyclical abdominal pain.
- Consider an imperforate hymen if there is a discrepancy between the Tanner stage and menarcheal status.

**Footnotes**

Competing interests: none.

Patient consent: Patient/guardian consent was obtained for publication.

**REFERENCES**


Figures and Tables

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3029536/?report=printable
Swollen abdomen with haematometra due to an imperforate hymen and the abdomen after hymenectomy.

**Figure 2**
Imperforate hymen.

Figure 3

Surgical treatment of an imperforate hymen.

Articles from BMJ Case Reports are provided here courtesy of BMJ Group
The treatment of 65 women with imperforate hymen by a central incision and application of Foley catheter

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Objective To determine the surgical outcome of 65 women with imperforate hymen treated with a central surgical incision and insertion of a Foley catheter.

Design A prospective study.

Setting The study was carried out at Department of Obstetrics and Gynecology, Faculty of Meram Medicine, Selcuk University, between 1 January 1996 and 30 June 2006.

Population A total of 65 women diagnosed as imperforate hymen.

Methods A central oval incision was performed to imperforate hymenal membrane, then 16F Foley catheter was protruded and the balloon was insufflated. Catheter was removed after 2 weeks duration. Estrogen cream was prescribed to all women for application onto hymenal structure for 2 weeks.

Main outcome measures Efficacy of procedure in treatment of imperforate hymen, preserving hymenal structural integrity that is accepted as important for virginity in some societies.

Results After the procedure, hymenal orifice created remained open and intact in all women except two women. Closure of artificially created hymenal orifice in these two women was believed to be related to inappropriate administration of estrogen cream. Subsequent treatment with local estrogen treatment results in the hymenal orifice remaining opened in these two women.

Conclusions We have previously reported the technique in 2002, but now we are able to demonstrate results of our technique in an expanded number of women. This technique is less invasive than other methods and prevents many social problems related to virginity by preventing destruction of the integrity of the hymenal structure and providing an annular-intact hymenal ring.

Keywords Foley catheter, imperforate hymen, intact hymenal ring.

Introduction

The imperforate hymen is a common genital disorder. The incidence of hymen imperforatus is approximately 1 among 2000 girls. Some cases are recognised because of muocolpos at birth, but the diagnosis is usually not detected before puberty. The symptoms that appear after the onset of puberty are due to accumulation of menstrual blood. Menstrual blood usually accumulates in the uterus and upper vagina resulting in abdominal pain, distension of the lower abdomen and often acute urinary retention. The management is easy, but many women visit several doctors before the exact diagnosis is achieved. Haematocolpos due to imperforate hymen is treated by simple star-shaped incision or by other surgical interventions protecting the Bartholin gland orifices to separate the hymen and the meatus.

The aim of this study was to determine the outcome in 65 women with imperforate hymen whereby the integrity of hymenal structure is preserved with a central oval incision and the insertion of a Foley catheter and local treatment with estrogen cream for 2 weeks.

Methods

Sixty-five women with cyclical pelvic pain and primary amenorrhoea were included into the study at the Department
of Obstetrics and Gynecology, Faculty of Meram Medicine, Selcuk University, between 1 January 1996 and 30 June 2006. All these women were diagnosed with imperforate hymen by visual inspection of totally closed hymenal structure, bulging of hymen out of the vagina and ultrasonographic detection of haematocolpos. First of all, women were informed about classical surgical methods, but they usually opted for an alternative procedure to attempt to preserve their hymenal ring as much as possible.

Women were positioned in the lithotomy (Figure 1). The hymen was perforated by a sterile injection from the middle of the distended and imperforate hymenal membrane, and then the perforation site was enlarged to 0.5 cm in diameter to allow for the insertion of a Foley catheter (Figure 2). After drainage of the blood in the vagina using irrigation with a saline solution, a 16F Foley catheter was then inserted and the balloon of the catheter is insufflated by 10 ml of water (Figure 3). The distal end of the Foley catheter is fixed to medial part of the patient’s thigh by using a plaster to prevent any traction on it, and the presence of the Foley catheter allowed further drainage of blood in the uterus and vagina. Estrogen cream (Premarin vaginal cream; Wyeth, Istanbul, Turkey) was applied to the hymenal opening for the next 14 days to augment vaginal epithelisation. A single dose intravenous prophylactic antibiotic was administered to all women (1 g Ceftriaxon, Rocephin; Roche, Istanbul, Turkey).

Women were observed in hospital for 6 hours after the procedure and were warned about the risks of infection before they were discharged home. We advised women to keep vulva clean with iodine solution. We reviewed women for pelvic infection 1 week after the procedure. The Foley catheter was removed after 2 weeks of operation (Figure 4), and women were followed up monthly for the initial 6 months and then every 6 months thereafter.

Results

The mean age of women was 13.9 ± 2.1 years. The main presenting complaints of all these women were primary amenorrhoea and pelvic pain. Some of these women also complained of low back pain. Haematocolpos was detected in all the 65 women (100%).

After the procedure, the hymenal orifices created remained open and hymenal structures were found to be annular and intact except in two women. In two women, we observed reclosure of hymenal orifice that we created. Failure in these two women was believed to be related to the inappropriate administration of the estrogen cream to the hymen. Subsequent repeat surgical incision and drainage followed by a sufficient amount of local estrogen treatment resulted in a successful outcome for these two women. Complications such as bleeding and infection were not detected in any
women. The mean follow-up period for these women was 56.5 ± 16.2 months.

The mean age at marriage among these women was 19 ± 4 years. Twenty-eight women got married in the follow-up period, and they were all found to bleed following the first coitus. None of the women complained of severe dyspareunia at the first coitus. Nineteen of the married women fell pregnant in the follow-up period and 15 of them delivered vaginally, whereas the remaining 4 women underwent caesarean section.

Discussion and conclusion

In the classical surgical treatment of imperforate hymen, stellate or cruciate incisions are made through the hymenal membrane. The individual quadrants are excised along the lateral wall of the vagina. Inset margins of vaginal mucosa are approximated using with fine delayed-absorbable sutures. These procedures produce good surgical outcome, but the hymenal structure is usually damaged.4–6 Because the hymen is accepted as a sign of virginity, damage to the hymen may be undesirable in some families and societies. The technique described in this manuscript was originally used on a girl who insisted on preservation of her hymen as an indication of her virginity.7 We have already reported the technique in 2002 in a limited number of women, but we are now able to evaluate our results with an expanded number of women to confirm our previous results and also to report two failed cases, which we believe were related to the inappropriate usage of local topical estrogen cream.

In the literature, there are reports on labial adhesions being successfully treated with topical estrogen application.8 The closure of the central hymenal incisional defect in our series is also prevented by the administration of topical estrogen cream for 2 weeks.7

In a study performed by Liang et al., women who underwent surgical correction for imperforate hymen was found to have irregular menstrual cycles in the majority of cases. Most of these women were also worried about their future fertility and six women suffered from dysmenorrhoea. Out of the 11 women who began having intercourse, 2 women fell pregnant with none of them complained of sexual dysfunction. After hymenectomy, it was also reported that the women with imperforate hymen were markedly relieved of cryptomenorrhoea, and there was improvements in micturition and defecation.9

There is no known complication of this method, and no study similar to this was reported in the literature. This technique is less invasive than other methods and can potentially preserve the integrity of the hymenal ring.

References

Acar et al. have described a novel technique for the treatment of imperforate hymen. The authors state that the operation is performed to maintain the hymenal structures as a sign of virginity in some cultures. I submit that until a woman has intercourse, she remains a virgin, regardless of whether or not she has had a corrective surgical procedure performed upon her hymen. In my opinion, the idea that a woman’s virginity is solely determined by her anatomy is detrimental to the social status of women worldwide.

As a physician, it is my duty to serve my patients with the best care medicine can provide and to act as an advocate on their behalf to advance their standing in society regardless of race, religion or sexual orientation. The idea that the ‘defloration’ of a woman’s hymen upon marriage to a man perpetuates the idea that women are no more than prizes to be vied over and eventually won, possessed and dominated.

In this case, as all potential women for the treatment would be minors, the patient herself will be unlikely to make decisions regarding the treatment plan; rather, her parents or guardians will do so. As such, it calls into question the role of the physician treating a minor patient. Are we to be the enforcers of religious beliefs upon a patient, simply because she has not yet reached an age suited for consent? Truly, this dilemma plagued my thoughts from the moment I first read the article and shall continue to do so.

Optimally, a patient should be able to choose a treatment plan which suits her understanding of her own body, the society in which she lives and her role in that society. This decision-making process should be a dialogue between the patient and her physician, free of coercion and bias. Given the intimate nature of the clinical situations we face as obstetrician-gynecologists, this mind-set should be ever present during our encounters with patients.

Academic debate of opposing viewpoints in controversial matters such as this is an essential component of the advancement of knowledge: medical, social and ethical. We must be sure that our practices and values are in line with one another. In our global society, will we act to ensure the equality of those among us who have been subjugated in the past? or will we continue to perpetuate the submission of one individual to another? I submit these questions to challenge the minds of BJOGs readership with the hopes that they will analyse and identify where their biases lay and how they may reconcile them with their duties as a healthcare provider.

CM Estes

Reference

Case Report

Tuboovarian Abscess as Primary Presentation for Imperforate Hymen

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Objective. Imperforate hymen represents the extreme in the spectrum of hymenal embryological variations. The archetypal presentation in the adolescent patient is that of cyclical abdominopelvic pain in the presence of amenorrhoea. We reported a rare event of imperforate hymen presenting as a cause of tuboovarian abscess (TOA).

Case Study. A 14-year-old girl presented to the emergency department complaining of severe left iliac fossa pain. It was her first episode of heavy bleeding per vagina, and she had a history of cyclical pelvic pain. She was clinically unwell, and an external genital examination demonstrated a partially perforated hymen. A transabdominal ultrasound showed grossly dilated serpiginous fallopian tubes. The upper part of the vagina was filled with homogeneous echogenic substance. Magnetic resonance imaging (MRI) demonstrated complex right adnexa mass with bilateral pyo-haemato-salpinges, haematometra, and haematocolpos. In theatre, the imperforate hymen was opened via cruciate incision and blood was drained from the vagina. At laparoscopy, dense purulent material was evacuated prior to an incision and drainage of the persistent right TOA.

Conclusion. Ideally identification of imperforate hymen should occur during neonatal examination to prevent symptomatic presentation. Our case highlights the risks of late recognition resulting in the development of sepsis and TOA.

1. Introduction

Imperforate hymen represents the extreme in the spectrum of embryological variations in hymenal configuration, with reported incidence ranging from 0.014% to 0.1% [1]. Antenatal diagnosis of imperforate hymen is challenging; thus, neonatal diagnosis is optimal to prevent symptoms and complications seen when primary presentation occurs in adolescence.

The archetypal presentation in the adolescent patient is that of cyclical abdominopelvic pain in the presence of amenorrhoea [2]. The accumulated blood behind an intact hymen may compress the adjacent pelvic organs or vessels, resulting in some of the less common presentations such as urinary retention, back pain, or constipation. We report a rare case of spontaneous partial rupture of an imperforate hymen resulting in tuboovarian abscess (TOA) and sepsis.

2. Case Study

A 14-year-old girl presented to the emergency department of the Gold Coast University Hospital complaining of severe left iliac fossa pain with her first episode of heavy bleeding per vagina since the morning of presentation. Her background history was unremarkable with no significant medical or surgical history. She had no previous sexual encounters (virgo intacta). Further questioning elucidated that she had not gone through menarche and had cyclical pelvic pain for the previous three linebreak months.

Examination revealed a clinically unwell patient. She presented as febrile to 39 degrees Celsius, with tachycardia of 118 bpm. Her blood pressure was 120/60, showing a postural drop to 105/40. Her abdomen was soft; however there was marked lower abdominal tenderness on palpation, rebound tenderness and abdominal guarding. External genital examination was limited by copious amounts of both old and fresh
blood. However, it did reveal a partially perforated hymen with the rest of the hymen still intact. Intravenous antibiotics were empirically commenced.

Biochemical workup indicated signs of infection, with a c-reactive protein of 315, elevated white cell count of 12, and a neutrophil count of 8.9. Blood cultures were obtained, and ultimately a coagulase negative staphylococcus (probable skin contaminant) was grown. The urine specimen was grossly contaminated with blood and grew no organism. The beta-human chorionic gonadotropin was negative.

The transabdominal ultrasound images showed grossly dilated, serpiginous fallopian tubes with low level internal echoes and peripheral vascularity. There was echogenic substance within the endometrial cavity; the upper part of vagina was dilated and filled with homogenous echogenic substance (Figure 1). A right complex mass was noted on ultrasound (Figure 2). The lower vagina was not well visualized. The renal tract was assessed as normal. Magnetic resonance imaging (MRI) further elucidated the complex right adnexal mass with bilateral pyo-haemato-salpinges, haematometra, and haematocolpos (Figures 3 and 4). The pictures were in keeping with a partial imperforate hymen rather than vaginal septum.

An examination under anaesthesia with diagnostic laparoscopy was undertaken. The imperforate hymen was opened via a cruciate incision and blood drained from distended and oedematous upper vagina. Purulent material in the abdominal cavity was drained, and division of omental adhesions to uterus and fallopian tubes, caecum, and appendix was necessary to adequately view pelvic organs. A right TOA was identified, incised, and drained (Figure 5). A thorough washout was performed and a pelvic drain was inserted.

She had an uncomplicated recovery and was discharged six days following the procedure with a course of oral antibiotics. At outpatient review six weeks following surgery, she reported a resolution of her dysmenorrhoea with the menstrual cycle experienced postoperatively.

3. Discussion

A literature search was conducted using Medline, Pubmed, and Cochrane with the terms [mullerian anomaly] OR [mullerian tract] OR [imperforate hymen] OR [vaginal septum] AND [pelvic abscess] OR [pelvic infection]. This yielded several case reports of common and unusual presentations for imperforate hymen.

On occasion, antenatal ultrasound can detect an imperforate hymen due to the presence of hydrocolpos in the fetus in response to maternal oestrogens [11]. Imperforate hymen can also be identified on examination of the newborn, and if it is asymptomatic at this time, the recommendation is to delay surgical management until puberty, when the Oestrogenization improves elasticity and healing. The diagnosis is infrequently made in infancy, if the patient presents with a mucocele, in which case hymenectomy may be indicated.
A retrospective cohort study demonstrated a bimodal distribution of age at diagnosis. 43% of patients were diagnosed at less than 4 years of age, and the remaining 57% were diagnosed over 10 years of age. In the group of patients diagnosed over 10 years, 100% of patients were asymptomatic, compared with just 10% of girls diagnosed under the age of 4 years [2].

In spite of recommendations for early inspection of the external genitalia, variations in hymenal anatomy frequently escape diagnosis until menarche. Although the most common adolescent presentation of an imperforate hymen is haematocolpos manifesting as abdominal pain [2], there are several case reports of more unusual or severe presentation of imperforate hymen, and these are presented in Table 1.

Of interest are the two case reports of pelvic infection arising from an imperforate hymen, which demonstrates the breakdown of the barrier from a sterile haematocolpos, either iatrogenically [6], or embryologically [7]. This paralleled our case presentation in that our patient did not become septic until the spontaneous rupture of the hymen which thus facilitated ascending infection of her reproductive tract.

Only one paper reviewed the long term outcome of late diagnoses of imperforate hymen. It reported the persistence of menstrual dysfunction even after hymenectomy, with 60% demonstrating abnormal menstruation and 40% with ongoing dysmenorrhea [11]. Furthermore, the data assessing fertility in these patients is sparse but encouraging. 86% of women who attempted pregnancy conceived after surgical correction of their imperforate hymen [12].

### 4. Conclusion

Ideally identification of the imperforate hymen should occur during the neonatal examination. Delay in diagnosis results in symptomatic presentations, with the potential for long-term menstrual and reproductive ramifications. Our case highlights the risks of late recognition of imperforate hymen, through the development of sepsis and TOA.

### Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

### References


