Original Article

Diagnosis and Management of Congenital Anomalies of Vagina

Sarwat Ara, Sumera Tahir

Abstract

Objective: To create awareness by presenting case series of congenital anomalies of the vagina, their diagnosis and management. **Design:** Interventional case series. **Setting:** Allied Hospital PMC Faisalabad. **Materials and Methods**: 18 patients with vaginal anomalies were reviewed between March 2008 and February 2011. Patients were evaluated by symptoms, physical examination and investigations. After management they were followed up to a period of minimum 3 months to a maximum of 2 years. **Main Outcome Measures:** Improvement in symptoms, creation of vagina with at least a 3-5cm in width leading to perfection of sexual life and fertility. **Interventions:** were done in 16 (88.88%) cases of transverse and longitudinal vaginal septum, imperforate hymen and absent vagina. Transverse vaginal septum and longitudinal septum was removed, imperforate hymen incised and no postoperative contractures occurred. Mayer-von Rokitansky-Küster-Hauser's Syndrome (MRKH) was managed by non surgical method. While absent vagina and cervix with functioning uterus was treated with abdominal hysterectomy followed by vaginoplasty and absent vagina with rudimentary/hypo plastic uterus treated with vaginoplasty. **Results:** 7(38.88%) cases of vaginal septum, 4(22.22%) cases of imperforate hymen, 1 (5.55%) case of vaginal atresia with absent cervix and functioning uterus,1(5.55%) hypo plastic uterus with absent lower vagina were operated for creation of vagina. Functioning vagina with a minimum of 4-5cm width in the former area of the septum and atretic vagina was created. 3(16.66%) longitudinal vaginal septum were operated during labor, a satisfactory sexual life was reported by all except in one of the group. We achieved 88.23 %(15/17) successful results. While 1(5.55%) cases of absent uterus and vagina in non Inter- vented group did not follow back. **Conclusion:** Congenital anomalies of the vagina have long term impact on patient sexual and reproductive life. New born girls must be examined for vaginal defects initially so that investigations could be arranged for further abnormalities such as upper mullerian duct and renal tracts. **Key Words:** vaginal anomalies, transverse vaginal septum, vaginoplasty, genital malformations, Haematocolopos, Haematometra, Imperforate hymen, Mayer-von Rokitansky-Küster-Hauser's Syndrome (MRKH)

INTRODUCTION

Female genital malformations occur in 5% of the general population[1]. Different Vaginal abnormalities include Müllerian aplasia[2] (MRKH), vertical fusion defects resulting from incomplete fusion of mullerian ducts to the urogenital sinus. It includes transverse vaginal septum and cervical agenesis. Lateral fusion defects of the two Müllerian ducts resulting in longitudinal vaginal septum with or without uterine abnormalities[2]. MRKH is a syndrome characterized by congenital aplasia of the uterus and the upper part (2/3) of the vagina in women with or without renal, skeletal and hearing problems[3,4] occurs 1:4,000-5,000 female births[5], 30-36% of such genital malformations are associated with anomalies of the kidneys and skeleton[6]. A vaginal septum is a congenital separation within the vagina. Longitudinal vaginal septum develops during embryogenesis due to an incomplete fusion of the lower parts of the two mullerian ducts. As a result there is a double vagina[7]. A transverse septum appears between the junction of urogenital sinus and the sinovaginal bulb. It generally occurs between the upper one-third and lower two-thirds of the vaginal canal[8].

The reported incidence of all transverse vaginal septum is 1:2,100 - 1:72,000 [9,10]. Imperforate hymen is a gynecological abnormality which is usually not detected until the onset of menstruation. The exact incidence and prevalence remained uncertain. However it occurs round about 1:1000-10,000 population[11], mainly in teenage girls with the symptoms of primary amenorrhea, recurrent lower abdominal pain and urinary retention[12]. Imperforate hymen, transverse septa, and distal vaginal agenesis and Müllerian aplasia may appear similar on symptoms and assessment[13].

History, examination Sonocolpography[14] is significant in diagnosing the complete transverse vaginal septum and other related conditions. MR imaging is opted for distinctive delineation of congenital anomalies of vagina when results of physical examination and ultrasound are inconclusive[15].

MATERIALS AND METHODS

Patients with vaginal anomalies were reviewed between March 2008 and February 2011. Patients were admitted through OPD and emergency. They were evaluated by history examination and relevant investigation like ultrasonography, MRI and reviewed by their relevant interventions like hymenectomy, removal of vaginal septum, vaginoplasty and abdominal hysterectomy. All patients were followed for a minimum of 3 months to a maximum of 2 years.

RESULTS

During mentioned period a total of 18 cases with four types of congenital vaginal malformations were picked out. These were transverse and longitudinal vaginal septum, imperforate hymen, absent vagina with or with out functioning uterus (MRHK syndrome). Out of 18 patients, 7 (38.91%) were selected for removal of transverse vaginal septum. All were having partial vaginal septum with a fenestre at variable sites. All these 7 patients had history of dyspareunia and prolonged duration of menstruation. After surgery 5 of 7 patients with transverse vaginal septum were relieved of dyspareunia and conceived during follow up period. All 4 (22.22%) patients with imperforate hymen came with retention of urine, primary amenorrhea and cyclical lower abdominal pain. Imperforate hymen, transverse septa, and distal vaginal agenesis differentiated by history, examination and Sonography. For Imperforate hymen cruciate incision in the hymen was given and haematocolopos was drained. They remained well during their follow up. Longitudinal vaginal septums were detected in 3 (16.66 %) patients. All were discovered during labor and evacuation and curettage; septum was removed during delivery and at the time of evacuation. During their follow up vagina remained normal and patients relieved of dyspareunia. The aim in absence of uterus and vagina was to create a blind-ending vaginal pouch by surgical or conservative approach. The surgical technique (Abbe-McIndoe) involves dissection between bladder and rectum followed by placement of a mold covered with an amnion graft into the space, and conservative approach (Franck's dilator method) by forceful dilation of a shallow rudimentary vaginal pit with the serial application of progressively wider and longer dilators. There were 2 (11.11%) patients belonging to MRHK syndrome and had absent uterus and vagina with unilateral renal agenesis and presented with primary amenorrhea. In these patients non-surgical method of neovagina was opted. They were counseled, explained about vaginal dilation. They adopted willingly conservative approach. 1 (5.55%) patient remarried; relieved of symptoms while 1 (5.55%) did not come for follow up. 1 (5.55%) patient had absent vagina but functioning uterus and presented with primary amenorrhea with cyclical abdominal pain. In this patient, vagina and cervix were absent but uterus was functional so abdominal hysterectomy with removal of endometrioma was done. Later on vaginoplasty with amnion grafting was performed as second step procedure. Fortunately the above patient did well and got remarried. Remaining 1 (5.55%) patient with absent lower vagina having hypo plastic uterus underwent vaginoplasty with amnion grafting; it was followed by vaginal stenosis.

DISCUSSION

A wide variety of Müllerian anomalies has been described in the literature. Various combinations of anomalies may coexist in a single subject[16]. Symptoms of vaginal obstruction occur at the time of menarche; depending upon complete or partial obstruction. If remain uncovered may lead to dyspareunia or obstruction in labor. It was suggested that whenever a patient presented with amenorrhea, dyspareunia, apareunia, prolonged menstruation and infertility; an evaluation for vaginal obstruction must always be performed[17].

In this study 44.44% patients presented with primary amenorrhea in contrast to 59.2% in one study[18]. In the present study 100% patients with imperforate hymen presented with acute retention of urine along with primary amenorrhea and abdominal pain in contrast to a ten year study[19] which revealed that 53% presented with acute retention of urine while 47% with other symptoms like lower abdominal pain, lower abdominal mass, and protruding Introital mass.The current case series explored 38.88% patients with transverse vaginal septum, 16.66% longitudinal vaginal septae, 22.22% imperforate hymens, 11.11% absent uterus and vagina, 11.11 % absent vagina with uterus. Comparing our results with one Chinese study[18] we found dissimilar results of 12.3% transverse vaginal septum, 17.2% longitudinal vaginal septae, 9.8 imperforate hymen and 19.7% absent vagina. It is also contrary to study[20] where we found transverse vaginal septum, longitudinal vaginal septae, and imperforate hymen 11.5%, 27%, and 50% respectively. Patients having primary amenorrhea along with cyclical pain need to be differentiated between transverse vaginal septum and imperforate hymen. They are differentiated by bluish bulging membrane of imperforate hymen from pinkish thickened membrane of transverse vaginal septum. Imperforate hymen can also be differentiated from a low transverse vaginal septum by Valsalva maneuver. Former bulges outward with Valsalva whilst there is no outward bulge in patients with transverse vaginal septum[21].

Transverse partial septum with pinpoint openings may remain hidden during gynecologic examination as it occurred in our cases that were revealed during period of menstruation. In present study patients with transverse septum were treated with by incising and removing the partial septum. Patient did very well after surgery. Then reproductive outcome was good in 71 .4 %(5/7) patients who became pregnant. It is not in accordance to study[20] where pregnancy outcome was 94% in the transverse septum The reason for less success rate might be that in present study 28.5 %(2/7) were divorced however it is more than 47% found in another study[22] .

In this study all longitudinal vaginal septae (3/18) were received during labor and evacuation. Patients with longitudinal vaginal septum may or may not have any symptoms. It may rupture during coitus or could persist and be recognized by obstetrician during labor. In the present study 66% patients with longitudinal vaginal septum were asymptomatic while it was 56.4% in another study[23]. Obstetric complications are mainly linked to a uterine malformation as occurred in 33% cases contrary to 87.8% uterine malformations documented in the previous study [23].However we removed longitudinal vaginal septum due to predisposition to dystocia[23]. Handling imperforate hymen, longitudinal septum, and low thin transverse septum are fairly easy and simple, but surgeries involving vaginal agenesis in association with functional uterus are complicated. Absent uterus and vagina is less problematic as compared to absent vagina and functioning uterus without cervix along with endometriosis as occurred in 1/18 cases. It requires skill and experties[24] Creation of a cervix is associated with a very high rate of infection and mortality[25].

The best management in during the study our opinion was abdominal hysterectomy and removal of endometrioma followed by second step Vaginoplasty. In mullerian aplasia or absence of uterus and vagina, the creation of a vaginal pouch remains the objective so Franck's dilator conservative approach and Abbe-McIndoe surgical technique were tried. We adopted non surgical methods were adopted for neovagina in11 % (2/18) came out successful in 50% (1/2). It seems dissimilar to success rate of vaginal dilation of 92-100% in different studies[26,27]. The decrease in success rate might be that our 50% (1/2)loss of patients during follows up. In mentioned study 11 % (2/18) underwent surgical Vaginoplasty with amnion grafting, out of which 50% (1/2) came out to be successful. It is in contrast to100% clinical outcome in another study[18].Mizia K et al[27] concluded in their study that dilators are effective in creating a functionally useful vagina because surgery mandates well-trained experts and definitive primary operation. Regarding renal and other system anomalies, 16.66 % had absent one sided kidney while no patient had skeletal abnormality in current study. Associated congenital anomalies of the upper urinary tract are reported to occur in 30-40% of all cases of MRKH syndrome[28]. Unilateral renal agenesis were present in 21% patients with major uterine malformations in one study[29] while Urinary symptoms and associated urinary abnormalities were detected in more than 50% of cases in another study.[17]

Table-1
Distribution of vaginal anomalies in relation to age, marital status, parity and other associated anomalies

Type of vaginal anomalies	Number	Mean age	Marital status	Parity	Associated anomalies
Transverse vaginal septum	7	23.8	Married-5 divorced-2	zero	nil
Imperforate hymen	4	13.3	Unmarried 4		nil
Longitudinal vaginal septum	3	22.3	Married - 3	Primigravid-2 Abortion-1	Septate uterus with absent kidney
Absent uterus and vaginal	2	24	Married and divorced 2	zero	Both had one absent kidney
Absent cervix vaginal and functioning uterus	1	19	Unmarried	----	nil
Hypo plastic uterus, Lower vagina absent	1	18	Unmarried	----	nil
Total	18		18		3/18

Table-2
Types of vaginal anomalies with clinical presentation

Type of vaginal anomalies	No.	Percentage (%)	Presentation
Transverse vaginal septum	7	38.88	Dyspareunia/ Apareunia/ prolonged menstruation
Imperforate hymen	4	22.22	Primary amenorrhea/ cyclical pain/ retention of urine
Longitudinal vaginal septum	3	16.66	h/o dyspareunia All presented during pregnancy/ labor
Absent uterus and vaginal	2	11.11	Desire for remarriage & primary amenorrhea
Absent cervix vaginal & functioning uterus	1	5.55	Primary amenorrhea/ cyclical pain
Hypo plastic uterus Lower vagina absent	1	5.55	Primary amenorrhea
Total	18	100	

Table-3
Different Anomalies and Management

Anomalies	Surgical Procedure	No.	Conservative	No.
Transverse vaginal septum	Picking, incising and cutting of septum	7		
Longitudinal vaginal septum	Removed during labor	3		
Imperforate hymen	Hymenectomy &drainage of haematocolopos	4		
Absent uterus & vaginal	----------		Creation of neovagina by vaginal dilators	2
Absent cervix vaginal & functioning uterus	Total abdominal hysterectomy with removal of endometrioma & Vaginoplasty	1		
Hypo plastic uterus Lower vagina absent	Vaginoplasty	1		
Total		16		2

Table-4
Effect of Intervention /non Intervention on outcome

Anomalies	Improved symptoms regarding fertility, Menstrual and sexual problems.	Remarks or symptoms not improved	Total Patients
Transverse vaginal septum	5	2 were divorced	7
Imperforate hymen	4 having normal menstruation	Unmarried	4
Longitudinal vaginal septum	No dyspareunia	Discovered during labor and pregnancy Fertility was not problem	3
Absent uterus & vaginal	1 got married and improved apareunia dyspareunia	1 Lost during follow up	2
Absent cervix vaginal and functioning uterus	Improved symptoms of endometriosis got married with normal sexual function		1
Hypo plastic uterus Lower vagina absent	-	symptoms not Improved Vaginoplasty which was followed byVaginal stenosis remained dyspareunia	1
Total			18

CONCLUSION

Examination of newborn female babies should always include an inspection of external genitalia. Delayed detection is not infrequent and can result in major risk to the patient because of associated urinary tract anomalies and pelvic endometriosis. Vaginal septum can be removed by simple minimal surgery. Diagnosis of Imperforate hymen should be kept in mind when dealing premenarcheal girls with lower abdominal pain and retention of urine.

REFERENCES

1. Oppelt P, von Have M, Paulsen M, et al. Female genital malformations and their associated abnormalities. Fertil Steril 2007; 87:335-42.
2. Caloia DV, Rahmani MR. Congenital transverse vaginal septum: vaginal hydrosonographic diagnosis. J Ultrasound Med 1998; 17:261-264.
3. Tolhurst DE, van der Helm TW. The treatment of vaginal atresia. Surg Gynecol Obstet 1991; 172: 407-14.
4. Bryon A, Nigro J, Counsellor V. One hundred cases of congenital absence of the vagina. Surg Gynec Obstet 1949; 88:79-85.
5. Griffin JE I, Konje JC: Mullerian agenesis: etiology, diagnosis, and management. Obstet Gynecol Surv 2000; 55:644-649.
6. Chandran L, Outflow obstructions differential diagnosis 2011; 03:25 Available from:http://emedicine.medscape.com/article/ 953015-differential.
7. Varras M, Akrivis C, Demou A, Kitsiou E, Antoniou N. Double vagina and cervix communicating bilaterally with a single uterine cavity: report of a case with an unusual congenital uterine malformation. J Reprod Med 2007; 52: 238–40.
8. Üstün Y, Üstün YE, Zeteroğlu Ş, Şahin G, Kamacı MA Case of Transverse Vaginal Septum Diagnosed During Labor. Erciyes Medical Journal 2005; 27:136–138.
9. Kil KC, Yoon JH, Chung JE, You SH, Park MS, Kwon DJ, Yoo YO. A case of transverse vaginal septum with modified Garcia technique. Korean J Obstet Gynecol 2007; 50:1563-1568.
10. Banerjee R, Laufer MR. Reproductive disorders associated with pelvic pain. Semin Pediatr Surg 1998; 7:52-61.
11. Paula J, Hillard A. Imperforate Hymen: 2011;09:19. Available from http://emedicine.medscape.com/article/269050-overview#a0199
12. Saks EK, Vakili B, Steinberg AC. Primary amenorrhea with an abdominal mass at the umbilicus. J Pediatr Adolesc Gynecol 2009; 22:1-3.
13. Miller RJ, Breech L: Correction of Vaginal anomalies. Clinical Obstetrics & Gynecology 2008; 51:223-23.
14. Thabet SMA. Role of new sono-imaging technique 'Sonocolpography' in the diagnosis and treatment of the complete transverse vaginal septum and other allied conditions. J Obstet Gynecol Res Japan 2002; 28:80-85.
15. Hricak H, Chang YC, Thurnher S. Vagina evaluation with MR imaging. Part I. Normal anatomy and congenital anomalies. 1988; 169:169-174.
16. Moawad NS, Mahajan ST, Moawad SA, Greenfield M. Uterus didelphys and longitudinal vaginal septum coincident with an obstructive transverse vaginal septum . Journal of paediatric and adolescent gynecology J Pediatric Adolese Gyneacol 2009; 22:163-165.
17. Nazir Z, Rizvi RM, Qureshi RN. Congenital vaginal obstructions: varied presentation and outcome. Pediatr Surg Int. 2006; 22:749-53.
18. Huang L, Ye M, Wang YB, Ji B, Tang JL. Analysis of 81 cases of congenital anomalies of the vagina. Nan Fang Yi Ke Da Xue Xue Bao. 2009; 29:1468-70.
19. Lui CT, Chan TWT, Fung HT, Tang SYH. A retrospective study on imperforate hymen and haematometrocolpos in a regional hospital. Hong Kong J Emerg. Med. 2010; 17:435-440.
20. Joki-Erkkilä MM, Heinonen PK. Presenting and long-term clinical implications and fecundity in females with obstructing vaginal malformations. J Pediatr Adolesc Gynecol. 2003; 16:307- 12.
21. Gorkem SB. Haematocolopos secondary to imperforate hymen. Journal of Clinical Imaging Science 2009; 06: 12.
22. Rock JA, Zacur HA, Dlugi AM, et al: Pregnancy success following surgical correction of

imperforate hymen and complete transverse vaginal septum. Obstet Gynecol 1982; 59:448-51.
23. Haddada B, Sylvestrea CL, Poitoutb P, Paniela BJ. Longitudinal vaginal septum: a retrospective study of 202 cases. Eur J Obstet gyneacol. 1997; 74: 197-199.
24. Quint EH, McCarthy JD, Smith YR.Vaginal surgery for congenital anomalies. Clin Obstet Gynecol. 2010; 53:115-24.
25. Amulya K Saxena. Vaginal Atresia. 2010; 03:03. Available from http://emedicine.medscape.com/article/954110-overview#a03].
26. Dharamsi N, Sheldon C. Management quandary. Case 2005: management of vaginal agenesis. J Pediatr Adolesc Gynecol. 2005; 18:359-62.
27. Mizia K, Bennett MJ, Dudley J, Mullerian MJ. Dysgenesis: a review of recent outcomes at Royal Hospital for Women.The Aust NZJ of Obstet Gynaecol 2006; 46:29-31.
28. Basile C, De Michele V.Renal abnormalities in Mayer-Rokitansky-Küster-Hauser's syndrome. J Nephrol. 2001; 14:316-8.
29. Acién P, Acién M.Unilateral renal agenesis and female genital tract pathologies. Acta Obstet Gynecol Scand 2010; 89:1424-31.

AUTHORS

- **Dr. Sarwat Ara**
  Assistant Professor Gynecology & Obstetrics
  PMC/Allied Hospital, Faisalabad
- **Dr. Sumera Tahir**
  Associate Professor Gynecology & Obstetrics
  PMC/Allied Hospital, Faisalabad
  Email: razatahir@hotmail.com