Case Report

Access this article online
Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.115053
Quick Response Code:

Early abdomino-perineal pull-through vaginoplasty

Ilhan Ciftci, Ayhan Tastekin[1], Ali Annagur[1], Mustafa Koplay[2]

ABSTRACT

Vaginal agenesis is known as one of the Müllerian anomalies. Müllerian anomalies occur during gonadal development and differentiation, and may lead to complex outcomes. McKusick–Kaufman syndrome (MKS) is an autosomal recessive multiple malformation syndrome characterized by Müllerian anomalies with hydrometrocolpos (HMC) and postaxial polydactyly (PAP). We report a case of a female newborn that developed hydronephrosis and respiratory distress due to compression of the cystic mass and underwent surgery on postpartum day 3. Pull-through vaginoplasty was performed with an abdomino-perineal approach. We report the unique treatment approach in this case.

Key words: Congenital, hydrometrocolpos, McKusick–Kaufman syndrome, neonate, polydactyly, pull-through, vaginoplasty

INTRODUCTION

Vaginal agenesis is known as one of the Müllerian anomalies. Müllerian anomalies occur during gonadal development and differentiation, and may lead to complex outcomes. Possible reasons for these anomalies include cessation or regression of Müllerian duct development and deficiency of estrogen and gestagen receptors.[1] We report a case of a female newborn with McKusick–Kaufman syndrome (MKS). MKS is an autosomal recessive multiple malformation syndrome characterized by Müllerian anomalies with hydrometrocolpos (HMC) and postaxial polydactyly (PAP).[1,2]

Departments of Pediatric Surgery, [1]Neonatalogy, [2]Radiology, Selcuk University, Medical Faculty, Konya, Turkey

Address for correspondence:
Dr. Ilhan Ciftci,
Department of Pediatric Surgery,
Selcuk Universitesi Tip Fakultesi,
Cocuk Cerrahisi A. D. Konya, Turkey.
E-mail: driciftci@yahoo.com

The patient developed hydronephrosis and respiratory distress due to compression of the cystic mass, and underwent surgery on postpartum day 3. Pull-through vaginoplasty was performed with an abdomino-perineal approach. We report the unique treatment approach in this case.

CASE REPORT

This 3 kg, female newborn infant was born by Cesarean section at 37 weeks of gestation to a 36-year-old gravida 7 para 7 mother at a local obstetric clinic. Her prenatal sonograms showed no abnormalities at 35 weeks of gestation.

After birth, an abdominal examination showed a swelling in the pelvic region. Palpation revealed a fixed mass with a smooth surface. Pelvic examination exhibited normal labia majora. Urethral opening was normal. Vaginal opening was not present [Figure 1].

Abdominal sonography showed a 10 × 7 cm cystic mass at the suprapubic area, and revealed HMC, bilateral grade 2 hydronephroureteroses, and anterior displacement of the compressed bladder.

The patient underwent abdominal magnetic resonance imaging (MRI), which showed HMC [Figure 2]. Physical examination of the hands and feet showed polydactyly and dysplastic phalanx.

Operative technique

A gynecologic examination showed normal labia majora, while there was only a single orifice similar to the urinary meatus. The patient was placed in the supine position and surgical preparations were carried out. The abdominal wall was incised with an infraumbilical transverse incision. During exploration, there was an approximately 12 × 10 × 7 mm cystic mass extending from the pelvic region in the retroperitoneum to the back of the stomach. The mass proved to be the vagina. The vaginal mass was evacuated by making an

incision. Proximal to the uterus, fallopian tubes and ovaries were present. An incision was made between the labias to create a vaginal opening by pushing the lower pole of the vagina between the urinary bladder and rectum.

The operation was continued by blunt and sharp dissection through the vertical plane, following the posterior of urethra from the distal portion of the urinary meatus. The proximal portion of the urethra was observed to be considerably wide, and there were adhesions to the adjacent tissues. No vaginal tissue was determined to exist between the urethra and rectum. Moreover, we determined an area of large vagina, extending from beneath the uterus to the pelvic floor. The vaginal tissue that we detected was liberated from the distal aspect [Figure 3a]. The neovagina was brought from between the urethra and rectum and anastomosed with 5-0 polyglactin sutures to the area that would host the opening. The opening was fixed by placing a 10F silicon tube within neovagina [Figure 3b]. A schematic view of the operative technique is given in Figure 4. After the anastomosis was created, the abdominal incision was closed.

The patient was included in a dilatation program. No problem occurred for 3 months. Urinary findings were improved.

DISCUSSION

Congenital vaginal atresia is a rare obstructive anomaly of the female genital tract. This generally occurs sporadically, with a reported incidence at term of 0.014–1%, and gives rise to hydrometrocolpos in less than 1/16,000 female births.[3,4] The management of patients with genital anomalies is a complex problem requiring individual surgical approaches, depending



Figure 1: Vaginal opening was not present in neonate

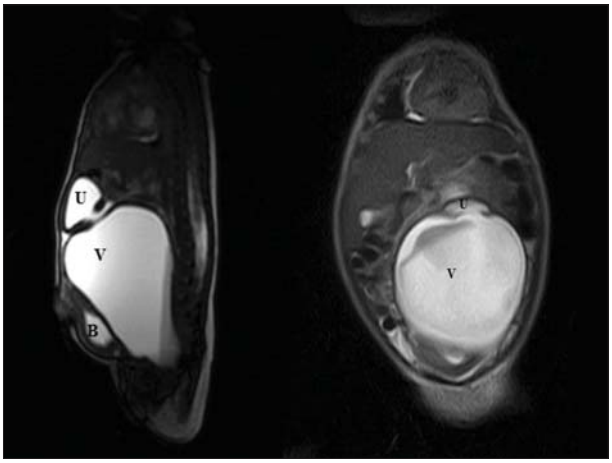


Figure 2: Magnetic resonance imaging for patient which showed hidrometrocolpos (V: upper vagina, U: uterus, B: bladder)

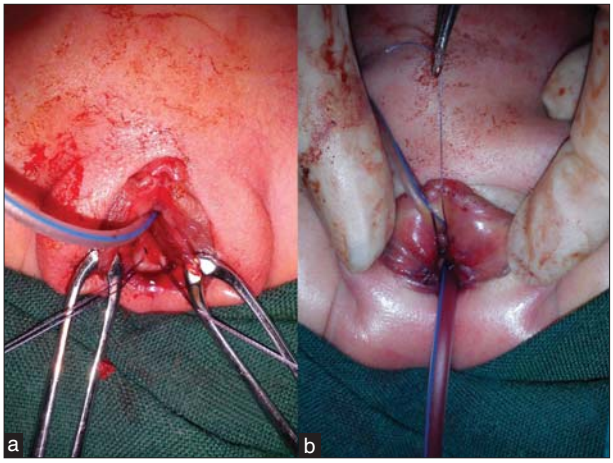


Figure 3: (a) Upper vagina between labias (V: Upper vagyna). (b) New vaginal opening

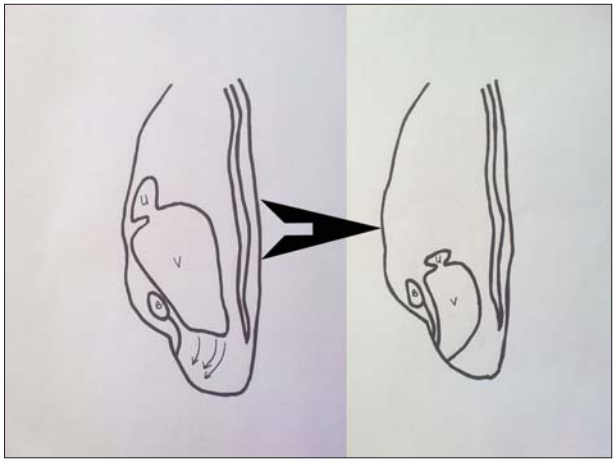


Figure 4: A schematic view of the operative technique (V: upper vagina, U: uterus, B: ladder)

on the anatomical conditions. The goals include the correction of visible anatomical anomalies, the creation of an appearance that corresponds to the patient's gender, and the restoration of function that will enable the individual to lead a normal life, including sexual function and, if possible, reproduction. Indeed, this implies that patient satisfaction should be among the goals. Many controversies about the management of genital anomalies center on this problem.[5]

There are various vaginoplasty techniques. The fundamental aim of these procedures is to form functional and comfortable vaginal tissue of adequate length and secretory character, which requires less dilatation.

The surgical method should be chosen based on the patient and the type of anomaly. Various methods can be used, including the McIndoe technique, the spontaneous epithelialization technique (Dupuytren), local muscle and skin flaps, free flaps, and ileal and colovaginoplasties.[6,7] Although numerous methods have been described since the first surgical intervention for the correction of vaginal agenesis, the Abbé–McIndoe technique is still the most popular and preferred method. Using a combination of blunt digital and scissor dissection, a cavity is formed digitally to admit two fingers.

A new technique developed by Vecchietti combines surgical and conservative methods, and involves epithelialization from the outer skin layer.[8] This method may have some disadvantages, including the need for prolonged vaginal dilatation sessions, which may cause psychological embarrassment for patients. Those dilatations may also lead to the formation of fistulas extending from the vagina to other organs, and epidermoid carcinoma has also been described.[9,10] Moreover, although stenosis is rarely observed in bowel vaginoplasties performed using grafts of the colon and ileum, the risks of continuous discharge prolapse the need for operative procedure(s) requiring prolonged recovery periods, bleeding, and infection are elevated,[11,12] while the risk of intestinal complications, including ileus due to fibrous adhesions (bridle ileus) and anastomotic leaks, is also increased. In addition, colonic vaginoplasty elevates the risk of neoplasia in the vagina.[13]

Our patient had an isolated vaginal agenesis and HMC. While the ovaries, uterus, and the proximal third of the vagina were developed, the distal two-thirds of the vagina were not developed. An emergency operation was planned due to the signs of ileus caused by severe abdominal distension, respiratory distress, hydronephrosis, and intestinal compression. We decided to create a vaginal opening, considering the risk of recurrence of another cyst formation because of possible inadequate emptying. By doing this, we were able to drain the reactionary fluid. We used the same vaginal tissue to create the new vaginal opening. Using this technique, we have provided a vaginal opening and prevented the formation of a cyst mass.

The ideal replacement material for an organ is the patient's own tissue, which is capable of normal functionality. Therefore, if possible, the replacement organ should be fabricated from autologous tissue. Thus, we freed up the dilated vaginal tissue and extended it up to the introitus.[5] No problems occurred for an 18-month period.

In this case, dilatation of the proximal vagina by approximately 3–4 cm facilitated our procedure. We encountered no problems, except for the tightness of the anastomosis.

Laparoscopy is a very useful method for visualization of intra-abdominal organs, and it can also provide assistance during dissection. Because of the severely distended abdomen and the significantly enlarged mass in this case, laparoscopy could not be performed. Exploration and dissection were completed via laparotomy.

In conclusion, early abdomino-perineal pull-through vaginoplasty can be effective in certain cases. It is particularly of great benefit in cases of vaginal agenesis.

REFERENCES

1. Stone DL, Agarwala R, Schaffer AA, Weber JL, Vaske D, Oda T, *et al*. Genetic and physical mapping of the McKusick-Kaufman syndrome. Hum Mol Genet 1998;7:475-81.
2. Son SH, Kim YJ, Kim ES, Kim EK, Kim HS, Kim BI, *et al*. A case of McKusick-Kaufman syndrome. Korean J Pediatr 2011;54:219-23.
3. Mor N, Merlob P, Reisner SH. Types of hymen in the newborn infant. Eur J Obstet Gynecol Reprod Biol 1986;22:225-8.
4. Benson CB, Doubilet PM. The fetal genitourinary system. In: Fleischer AC, editor. Sonography in Obstetrics and Gynecology. London: Prentice-Hall International; 1996. p. 444-50.
5. Ciftci I. Laparoscopic-assisted perineal pull-through vaginoplasty. J Pediatr Surg 2012;47:e13-5.
6. Wesley JR, Coran AG. Intestinal vaginoplasty for congenital absence of the vagina. J Pediatr Surg 1992;27:885-9.
7. Lima M, Ruggeri G, Randi B, Domini M, Gargano T, La Pergola E, *et al*. Vaginal replacement in the pediatric age group: A 34-year experience of intestinal vaginoplasty in children and young girls. J Pediatr Surg 2010;45:2087-91.
8. Borruto F, Chasen ST, Chervenak FA, Fedele L. The Vechietti procedure for surgical treatment of vaginal agenesis: Comparison of laparoscopy and laparotomy. Int J Gynaecol Obstet

1999;64:153-8.

9. Ingram JM. The bicycle seat stool in the treatment of vaginal agenesis and stenosis: A preliminary report. Am J Obstet Gynecol 1981;140:867-73.
10. Schouten van der Velden AP, de Hingh IH, Schijf CP, Bonenkamp HJ, Wobbes T. Metachronous colorectal malignancies: "Don't forget the neo vagina". A case report. Gynecol Oncol 2005;97: 279-81.
11. Freundt I, Toolenaar TA, Jeekel H, Drogendijk AC, Huikeshoven FJ. Prolapse of the sigmoid neovagina: Report of three cases. Obstet Gynecol 1994;83(5 Pt 2):876-9.
12. Toolenaar TA, Freundt I, Huikeshoven FJ, Drogendijk AC, Jeekel H, Chadha-Ajwani S: The occurrence of diversion colitis in patients with a sigmoid neovagina. Hum Pathol 1993;24:846-9.
13. Hiroi H, Yasugi T, Matsumoto K, Fujii T, Watanabe T, Yoshikawa H, *et al*. Mucinous adenocarcinoma arising in a neovagina using the sigmoid colon thirty years after operation: A case report. J Surg Oncol 2001;77:61-4.

Cite this article as: Ciftci I, Tastekin A, Annagur A, Koplay M. Early abdomino-perineal pull-through vaginoplasty. Afr J Paediatr Surg 2013;10:188-91.

Source of Support: Nil, **Conflict of Interest:** None declared.