Pub: 22/2008

CASE REPORT

Double uteri with cervicovaginal agenesis

Gurcharan Singh, M.Ch.,[a] *and Venigalla Sunita, M.D.*[b]

[a] Department of Plastic Surgery, and [b] Department of Obstetrics and Gynecology, Sri Devraj Urs University, Kolar, India

Objective: To report results of managing complex malformation of the genital tract and to validate a newer classification (vaginal, cervix, uterus, adnexa-associated malformation [VCUAM]) for describing such complex anomalies.
Design: Case report.
Setting: University hospital.
Patient(s): A patient with complex genital malformation.
Intervention(s): Laparotomy, double hysterectomy, and vaginal reconstruction with use of bilateral neurovascular pudendal thigh flaps. Use of VCUAM classification for description of the anatomy of malformation.
Main Outcome Measure(s): Creation of a neovagina and accurate and complete description of the anomaly.
Result(s): Relief of pelvic pain and creation of patent and sensate vagina retaining the innervations of the erogenous zones. The VCUAM classification could accurately depict complex anatomic variations.
Conclusion(s): Vaginal reconstruction with use of bilateral neurovascular pudendal thigh flaps yields good postoperative results. The VCUAM classification reflects anatomic variations in external and internal organs precisely. (Fertil Steril® 2008;90:2016.e13–e15. ©2008 by American Society for Reproductive Medicine.)

Key Words: Müllerian duct anomalies, cervicovaginal agenesis, double uteri

Maldevelopment of müllerian ducts occurs in a variety of forms, and each anomaly is distinctive. Anomalies in the formation and fusion of müllerian ducts can result in a variety of anomalies of the uterus and vagina.

Multiple etiologic factors are suggested for these syndromes. Variable expression of a genetic defect possibly precipitated by teratogenic exposure between 37 and 41 days of gestation, the time during which the vagina is formed, is suggested in the etiology of vaginal agenesis. Genital malformations have an incidence of up to 5% in the general female population.

This case of double uterus with functioning endometrium and single fallopian tube with cervicovaginal aplasia is reported for its rarity. A 16-year-old girl with cryptomenorrhea with complex genital malformation was the subject of our study. She required vaginal reconstruction and correction of internal genital malformation to relieve her of the pain ensuing from collected menstrual blood. A laparotomy with a plan for either hysterectomy or canalization of uteri and connecting them to a reconstructed vagina was planned along with vaginal reconstruction. An Institutional Review Board clearance was obtained regarding the plan of management.

Received February 29, 2008; revised June 17, 2008; accepted June 26, 2008.
G.S. has nothing to disclose. V.S. has nothing to disclose.
Reprint requests: Venigalla Sunita, M.D., Professor, Department of Obstetrics and Gynecology, Sri Devraj Urs University, No. 560 Bangarpet Circle, Kolar, Karnataka 563101, India (E-mail: venigallasunita@yahoo.co.in).

The method of vaginal reconstruction chosen was by posteriorly based neurovascular pudendal thigh flaps as described by Joseph and Wee (1). This method can reconstruct the vagina reliably in both congenital and acquired conditions. The reconstructed vagina has a natural angle and is sensate. Donor site scarring in the groin is inconspicuous. The main vascularity is from anastomosis of deep external pudendal and posterior labial arteries. The posterior part of the pudendal thigh flap retains its innervation from posterior labial branches from the pudendal nerve and posterior rami of the posterior cutaneous nerve of the thigh. The anterior part, which is innervated by the genitofemoral nerve and ilioinguinal nerves, may be denervated at the time of flap elevation. Sensation would be retained in the lower part of the reconstructed vagina.

Surgery was done with the patient under general anesthesia in the lithotomy position, which allowed access to both abdomen and perineum simultaneously. Initially vaginal reconstruction was done followed by laparotomy. A rectovaginal space was created by dissecting between bladder and rectum.

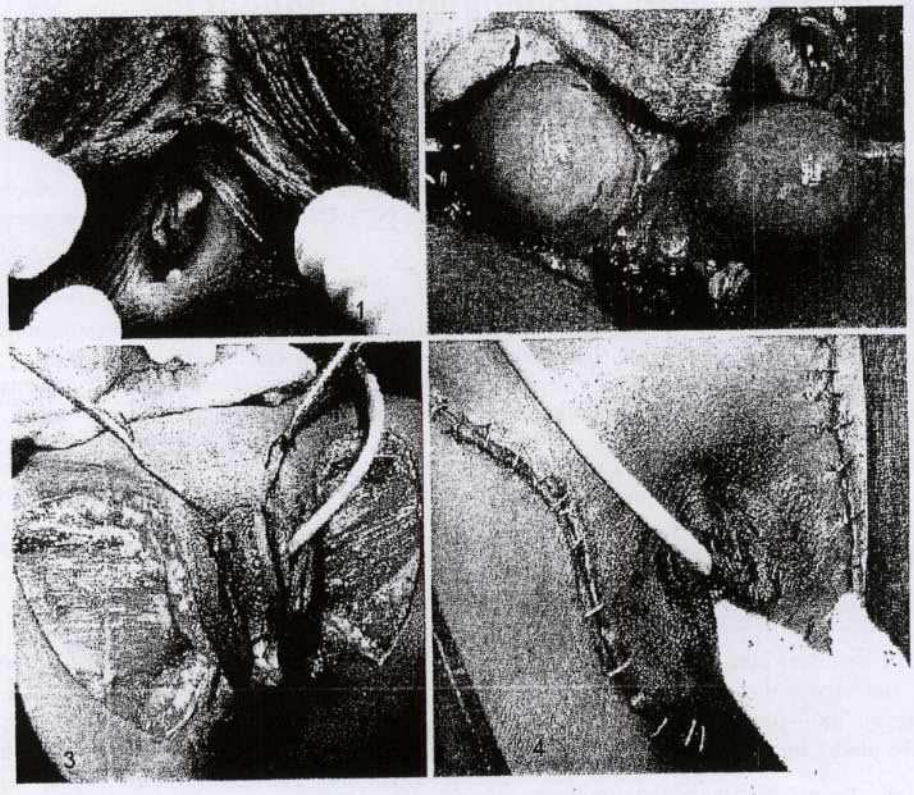
A 15 × 6-cm flap with posterior skin flap at the level of the posterior end of introitus was marked out. The flap was horn-shaped and was planned lateral to the hair-bearing area of the labia majora, centered on the crease of the groin. The incision was deepened up to deep fascia. The subfascial plane was developed raising the epimysium of the adductor muscles with deep fascia. The posterior margin was incised through dermis to subcutaneous tissue to a depth of 1 to 1.5 cm and then undermined in a plane parallel to the skin for a distance of 4 cm

0015-0282/08/$34.00

doi:10.1016/j.fertnstert.2008.06.048

FIGURE 1

(1) Preoperative photograph showing vaginal dimple. (2) Intraoperative photograph showing double uteri with single fallopian tube. (3) Raised and tunneled and sutured flaps. (4) Reconstructed vagina admitting two fingers.



Singh. Double uteri with cervicovaginal agenesis. Fertil Steril 2008.

posteriorly. This enabled the flap to be transferred through 70 to 90 degrees to meet its counterpart in the midline and posterior skin margin to be sutured to labia minora. The flaps were tunneled under the labia and sutured, creating a tubular structure that was interposed into the rectovaginal space. The tip of the vagina was anchored to the rectum. The opening of the vagina was sutured to the mucocutaneous edge of the labia minora. The vagina was packed with povidone-iodine (Betadine)–soaked gauze. Drains were inserted into the donor sites, which were closed by approximating the skin edges. Laparotomy showed two hypoplastic uteri with one fallopian tube attached to left uterus and two ovaries. Upon opening both uteri showed collected menstrual blood with hematosalpinx. As both uteri were hypoplastic and not feasible to canalise to vagina, double hysterectomy was done.

A patent and sensate vagina admitting two fingers easily was obtained, which remained patent at 1-year followup (Fig. 1). There was complete relief of cyclical abdominal pain, which was present before surgery.

In this case there was an unusual combination of dysgenesis of the caudal portion of the müllerian ducts and failure of fusion of two müllerian ducts. There previously have been few case reports of similar cases. In two of the cases reported earlier, precise diagnosis of the malformation was delayed. Only magnetic resonance imaging (2) and explorative laparotomy (3) could depict the complete extent of the malformation. However, in our patient, both transabdominal and transrectal ultrasound scans have been helpful, especially transrectal in suspecting absence of cervical tissue. This comprehensive understanding of the anatomy involved allowed us to plan the operative procedure so that laparotomy and vaginal reconstruction were undertaken at the same sitting. The patient and relatives were counseled before surgery regarding future reproductive potential.

Another important difference from the earlier cases is that we found a single fallopian tube. A third difference in this case is the technique of vaginal reconstruction. A patent and sensate vagina of 7 to 8 cm retaining the same innervations of the erogenous zones ensures adequate sexual life for the patient.

This case presented with a combination of cervicovaginal aplasia and double uterus with functioning endometrium; therefore it cannot be categorized according to AFS classification (4) or under Mayer-Rokitansky-Küster-Hauser (MRKH)

syndrome. According to the vaginal, cervix, uterus, adnexa-associated malformation (VCUAM) classification of Oppelt et al. (5), this case can be classified as V5b, C2b, U4b, A1a, M0. This classification, unlike the AFS classification, depicts each genital organ individually, similar to the TNM classification of tumors. The classification is simple and easily reproducible and gives a comprehensive description of the anomaly including adnexal and associated malformations, which was lacking in the AFS classification. Accompanying malformations of vagina or cervix such as hypoplasia and duplication could not be depicted in the AFS classification. Therefore the VCUAM classification appears a better way to classify the complex anomalies and provides precise orientation toward anatomic variations in external and internal organs.

A precise and comprehensive description of the anatomy is the key to the proper management. Joseph and Wee's technique of vaginal reconstruction (1) using bilateral posteriorly based neurovascular pudendal thigh flaps in this case yielded excellent postoperative results.

Acknowledgments: The authors gratefully acknowledge the assistance given by Dr. G. K. Narayana in preparing and submitting this manuscript.

REFERENCES

1. Joseph VT, Wee JTK. New technique of vaginal reconstruction using neurovascular pudendal-thigh flaps. In: Strauch B, Vasconez LO, Hall-Findlay EJ, eds. Grabbs encyclopedia of flaps. Vol. 3. New York: Lippincott-Raven, 1998:1461–70.
2. Bakri YN, Al-Sugair A, Hugosson C. Bicornuate nonfused rudimentary uterine horns with functioning endometria and complete cervical-vaginal agenesis: magnetic resonance diagnosis. Fertil Steril 1992;58:620–1.
3. Goluda M, St Gabrys M, Ujec M, Jedryka M, Goluda C. Bicornuate rudimentary uterine horns with functioning endometrium and complete cervical-vaginal agenesis coexisting with ovarian endometriosis: a case report. Fertil Steril 2006;86:462.e9–11.
4. The American Fertility Society. The American Fertility Society classification of adnexal adhesions, distal tubal occlusion secondary to tubal ligation, tubal pregnancies, Müllerian anomalies and intrauterine adhesions. Fertil Steril 1988;49:944–55.
5. Oppelt P, Renner SP, Brucker S, Strissel PL, Strick R, Oppelt PG, et al. The VCUAM (Vagina Cervix Uterus Adnexa-associated Malformation) Classification: a new classification for genital malformations. Fertil Steril 2005;84:1493–7.