
The Clinical Management of Maldevelopment of the Uterine Cervix

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Objectives: To outline the anatomical variations of malformations of the uterine cavity and to discuss the clinical management of cervical agenesis and dysgenesis.

Design: Patients who were treated for cervical malformations during a 55-year interval are reviewed and followed prospectively. The diagnosis of cervical agenesis or dysgenesis is assigned at the time of exploratory laparotomy.

Setting: Hospital based tertiary care reproductive endocrine-infertility units.

Main Outcome Measures: All surgical findings were carefully reviewed to determine the anatomical characteristics of the malformed cervix. When cervical reconstruction was performed, the patient was followed to determine the need for reoperation or if the patient achieved a pregnancy.

Results: Patients were found to lack a cervix (cervical agenesis) or to have one of three variants of cervical dysgenesis. Patients with cervical dysgenesis were characterized as having 1) an intact cervical body with obstruction of the cervical os, 2) a cervical body consisting of a fibrous band, or 3) cervical fragmentation. One patient conceived after reconstruction. Reoperation was less likely to occur among women who had an obstructed endocervical canal but an otherwise normal cervical body.

Conclusions: There are two basic types of anatomic congenital cervical malformations, the second of which may be considered as three distinct variations. The weight of the international clinical experience suggests that women with cervical agenesis or fragmentation should receive hysterectomy, whereas women with cervical dysgenesis may benefit from a reconstruction operation.

Congenital cervical abnormalities are rare with less than 50 reported cases in the world literature. To select reconstructive procedures for cervical atresia, surgeons

have emphasized the importance of the integrity of structures superior and inferior to the atretic cervix and the presence of associated abnormalities of the Wolffian duct. There are few articles that focus on the anatomical variants that may result from maldevelopment of the cervix. This study seeks to carefully describe the cervical abnormalities in 21 women with cervical maldevelopment. Subsequent pregnancy success or the need for

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Table 1. Surgical Approach to Patients With Abnormalities of Cervical Development (January 1, 1940–December 31, 1994)

Anomaly Observed	No. of Patients	Primary Hysterectomy	Cervicovaginal Reconstruction	Pregnancy Success	Reoperation and Hysterectomy
Cervical Agenesis	10	8	2	0	2
Cervical Dysgenesis					
Fragmentation	4	2	2	0	2
Fibrous cord	3	1	2	0	1
Cervical obstruction	4	1	3	1	0
Total	21	12	9	1	5

reoperation among women receiving cervical reconstruction is addressed.

Materials and Methods

Twenty-one women were examined and treated for cervical malformation at one of four hospitals (The Johns Hopkins Hospital, Union Memorial Hospital, Emory University Hospital, and Grady Memorial Hospital) between January 1, 1940 and December 31, 1994. All patients were between the ages of 13 and 18 years of age. Delay in diagnosis ranged from 1 to 4 years. The surgical approach to the management of these patients has been fully described elsewhere.¹⁻⁴

Briefly, the differential diagnosis in a patient with such an outlet obstruction is between a high transverse vaginal septum and cervical atresia. Initially, an attempt is made to identify the upper vagina by opening a neovaginal space. If the mass is high and cannot be reached from below, the abdomen should be entered to carefully delineate the anatomical findings from above. If a cervix is present, a probe may be introduced through the uterine fundus into the cervix to tent the upper vagina so that it may be entered safely from below. If there is cervical atresia, then a careful description of the findings should be recorded.

If cervical stroma is present, an attempt to create a cervicouterine fistula through the fibrous cord can be made. The surgeon may pass a probe through the cervical stroma or obstructed segment of endocervix into the vagina and pass a rubber catheter through this tract to maintain patency. If a large segment of vagina is undeveloped, then a vaginoplasty using a modification of the McIndoe technique may be performed. The uterine catheter may be left in place over the next 6 months allowing cyclic menstruation to occur and a permanent fistulous tract (epithelialized endocervical canal) to form. A full thickness skin graft (FTSG) 2 cm × 4 cm may be sewn around the uterine catheter to speed the process of epithelialized endocervical canal and to shorten the time required for the catheter, thus reducing the

chances of endometritis. The one patient who conceived had the FTSG rather than the split-thickness skin graft (STSG) that is usually preferred for reducing constriction of the cervical os.

Results

A review of the anatomic configurations at the time of exploratory laparotomy revealed that patients either had no cervix (cervical agenesis, $n = 10$), or an incompletely formed cervix (cervical dysgenesis, $n = 11$). Cervical dysgenesis presented as one of three groups. In the first group, there is a well formed cervix, but a portion of the endocervical lumen is obliterated. The length of the obliterated portion of the endocervical canal may vary ($n = 4$). In the second group, a cervical cord of variable length and diameter is noted with a completely obliterated endocervical canal. Endocervical glands were sometimes noted on pathologic examination ($n = 3$). In the third group, when fragmentation of the cervix was noted, a portion(s) of the cervix could be palpated below the fundus. In some instances several fragments of the cervix were noted ($n = 3$), or a single fragment was noted to be separate from the uterine fundus ($n = 1$) (Table 1) (Figs. 1, 2A–D).

Associated anomalies were noted in nine patients. These anomalies were ovarian malposition ($n = 4$), tubal abnormalities ($n = 3$), endometrial hypoplasia ($n = 5$), and solitary kidney ($n = 2$).

Absence or fragmentation of the uterine cervix ($n = 14$) was the most frequently observed malformation. Although cervical reconstruction was completed in four patients, all ultimately received hysterectomy.

Two of three patients with a cervix consisting of a fibrous cord received cervicovaginal reconstruction. After surgery, one patient began menstruating, whereas the other subsequently underwent hysterectomy.

Finally, three of four patients with cervical obstruction underwent a reconstructive procedure. One patient conceived 5 months after the procedure and delivered a viable premature infant by cesarean section at 34 weeks.

Table 2. Pregnancy After Cervical Reconstruction

Author	Year	Vaginal Status	Surgical Procedure	Follow-up
Zarou	1973	Atretic cervix, normal vagina	Cervicovaginal fistula formed after placement of two polyethylene drainage tubes	Onset of cyclic menses. Six years postoperatively delivered term infant by cesarean section
Welker	1988	Cervical obstruction partial vaginal agenesis	T drain placed in uterine cavity which drained through cervix and vagina	Cyclic menstruation established. Intrauterine pregnancy achieved; patient delivered viable fetus at 31 weeks gestation by cesarean section
Fraser	1989	Cervix fragmented with atretic midportion; normal vagina	Excised equal sized portions of lower uterus and upper cervix. Infant feeding tube sutured to the vaginal aspect of cervix and removed in three weeks	Cyclic menstruation established. Delivered a viable fetus at 37 weeks gestation by cesarean section
Hampton	1990	Atretic cervix dilated and cystic; vaginal agenesis	McIndoe vaginoplasty cervical graft with pediatric endotracheal tube	Onset of cyclic menses. Intrauterine pregnancy achieved. Abdominal cerclage performed. Delivery at 38 weeks gestation by cesarean section
Thijssen et al	1990	Atretic cervix normal vagina	Rudimentary cervix was sutured to the top of the vaginal vault. A polyethylene T drain was placed in the uterine cavity and removed 3 months later	Onset of cyclic menses on oral contraceptives. Pregnancy achieved with zygote intrafallopian transfer. Delivered at 38 weeks gestation
Present Study	1995	Atretic cervix, normal vagina	Cervicovaginal fistula formed after placement of polyethylene catheter with skin graft	Onset of cyclic menses. Intrauterine pregnancy achieved twice with delivery by cesarean section.

The patient has since delivered a second child. All patients are now menstruating, although one patient has severe dysmenorrhea thought to be secondary to extensive endometriosis.

Discussion

The uterine cervix provides a conduit for menstrual flow, maintenance of an intrauterine pregnancy, mucus for sperm transfer, and a barrier to infection from vaginal microflora. Atresia (dysgenesis) of the cervix may result from local segmental atrophy.⁵

Buttram has suggested a classification for "müllerian" agenesis or hypoplasia.⁶ Class I defects consist of a variety of malformations. Cervical anomalies are designated IB. Our findings suggest that this classification should be broadened to include patients with the various forms of cervical dysgenesis. The presence or absence of a normal vagina should also be taken into consideration. These modifications of the current system would allow a more precise definition of cervical malformations.

There is a consensus in the international literature that hysterectomy is the procedure of choice in a patient with cervical agenesis.^{1,7} Alternatively, when cervical dysgenesis is noted, reconstruction of the cervix may be warranted.

The goals of reconstructive surgery for cervical malformations are to provide a conduit for menstruation to relieve pain and preserve reproductive potential. The

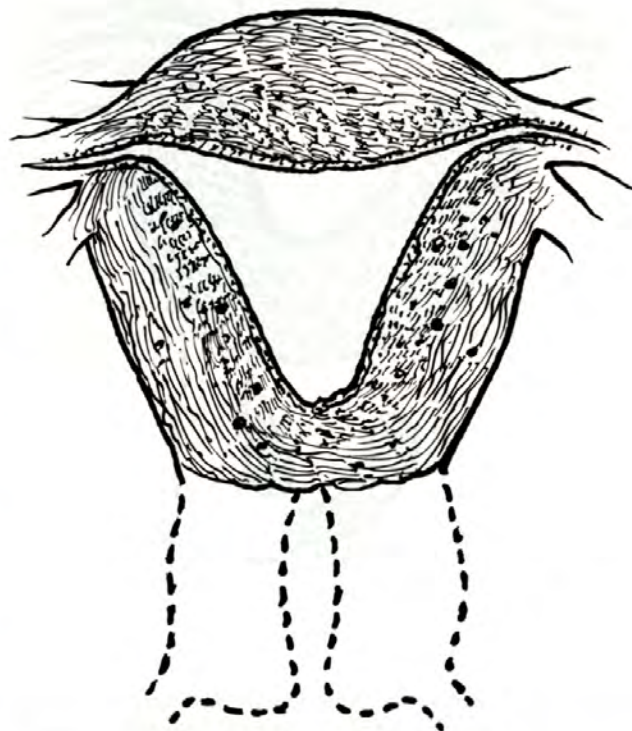


Figure 1. Congenital cervical agenesis. The fundus of the uterus is noted without a cervix.

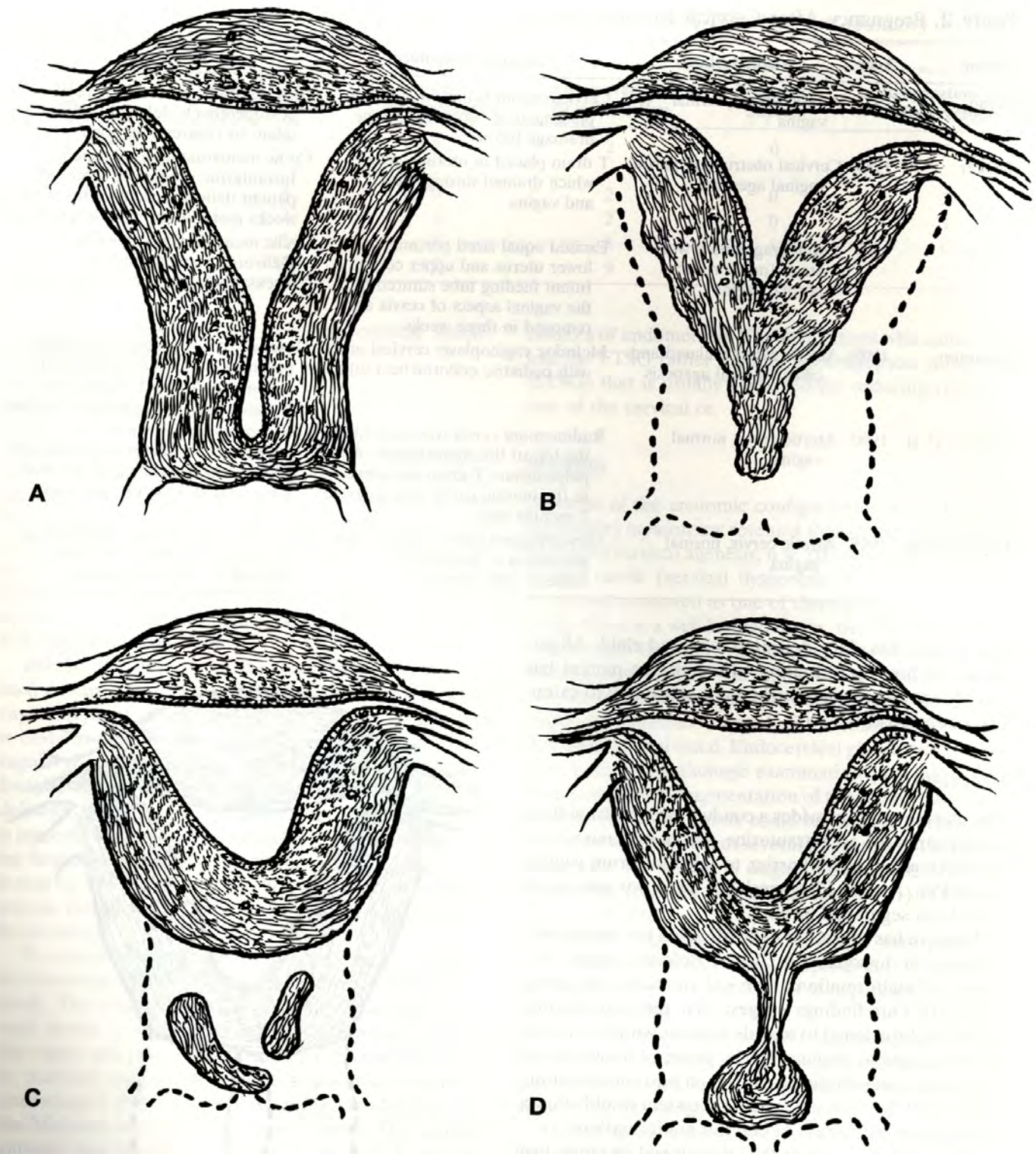


Figure 2. Congenital cervical dysgenesis. The cervical body is intact (A) with obstruction of the cervical os. Variable portions of the cervical lumen are obliterated. The cervical body (B) consists of a fibrous band of variable diameter that may contain endocervical glands. Fragmented portions of the cervix are noted (C) with no connection to the uterine body. Hypoplasia of the uterine cavity may be associated with cervical cords or fragmentation. The midportion of the cervix (D) is hypoplastic with a bulbous tip. No cervical lumen is identified.

goals are usually achieved when there is substance to the cervix. Moreover, pregnancy has been documented after cervical reconstruction when cervical stroma is substantial.⁸⁻¹² Cervical stroma was also present in our patient who ultimately conceived (Table 2).

Two groups of surgeons have emphasized the need for fabricating an epithelial cervical stent to graft the cervical lumen to maximize the chances for maintaining patency and preventing ascending infection.^{10,13} The need for such grafting may depend on the length of the atretic portion of the endocervical canal.

Patients with atresia or cervical fragmentation are not usually candidates for canalization. Patients with either cervical obstruction or a fibrous cord may reasonably be considered for reconstruction. The results of our study indicate that patients with cervical dysgenesis may have one of three anatomical variations. Patients with cervical fragmentation are usually not candidates for canalization. Alternatively, women with cervical obstruction or a fibrous cord may be candidates for reconstruction. All of the women with either clinical agenesis or fragmentation on whom canalization was attempted failed, requiring reoperative and hysterectomy within 6 months. One patient with a fibrous cord received hysterectomy because of persistent infection, although cyclic menstruation had been established.

This study and the international experience suggest canalization procedures may be performed on carefully selected patients, although a fragmented cervix usually warrants hysterectomy. Consideration should be directed to the presence of adequate cervical stroma to allow a cervicovaginal anastomosis to be achieved. If a long segment of cervix consists of a fibrous cord, a cervical grafting technique may be required. Cervical grafting may speed epithelialization of the newly created cervical canal. If a fragmented cervix is noted, hysterectomy is usually warranted. Nevertheless, as demonstrated by Fraser, anastomosis of the cervix to the uterine body may be accomplished with success, provided the distal cervical segment is well formed.

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