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Clinical features and surgical procedures of congenital vaginal atresia—A retrospective study of 67 patients

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ABSTRACT

Objective: To explore the characteristics of congenital vaginal atresia, further improve its classification, and therefore help the clinical diagnosis and treatment of congenital vaginal atresia.
Methods: This was a retrospective study of 67 patients with congenital vaginal atresia (from March 1984 to March 2015). Clinical and surgical characteristics were analyzed.
Results: For lower vaginal atresia, 25 patients successfully underwent vaginoplasty at the lower portion of the vagina. For complete vagina atresia, 25 patients with type i cervical atresia were treated with artificial vaginoplasty + tracheloplasty, and all showed no dysmenorrhea within six months after surgery. Four patients with type ii cervical atresia and two patients with type iii cervical atresia successfully underwent hysterectomy + artificialvaginoplasty. Two patients with type iv cervical atresia underwent combined abdominoperineal artificial vaginoplasty + tracheloplasty. One patient with upper vaginal atresia successfully underwent hysterectomy via the narrow segment of the cervix. Three patients with top vaginal atresia had no dysmenorrhea after transvaginaltracheloplasty.
Conclusion: This study suggests two new categories of vaginal atresia (upper vaginal atresia and top vaginal atresia), which could be used as a reference for treatment of this condition. Appropriate treatments were performed using a personalized approach and satisfactory results were achieved.

Introduction

In 1976, Simpson [1] suggested that congenital vaginal atresia (developmental defects of the urogenital sinus) differs from the congenital absence of the vagina (vaginal agenesis; results from Müllerian duct dysplasia), but the American Fertility Society (AFS) did not recognize Simpson's hypothesis until 1998. Based on Buttram's classification (1979) and the AFS classification (1988), a modified AFS classification of uterovaginalabnormalities was developed according to the embryology theory [2]. The modified AFS classification divides uterovaginalabnormalities into two main types: type I [Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome: lack of reproductive potential due to Müllerian duct dysplasia]; and type II (transverse vaginal septum resulting from abnormal vertical fusion of Müllerian ducts and vaginal abnormalities associated with cervical dysplasia). Recently, an embryological-clinical classification system has been suggested for the general classification of the malformations of the female genital tract [3].

In 2002, Leng et al. further divided congenital vaginal atresia in two types: type I (lower vaginal atresia, showing normal development of the upper portion of the vagina and the uterus); and type II (complete vaginal atresia associated with cervical atresia, showing normal development or deformity of the uterine body and with a possible normal secretory function of the endometrium) [4]. In 2012, Ruggeri et al. suggested three types of vaginal atresia with subtypes based on the six types of vaginal malformations they describe [5]. In 2013, the European Society of Human Reproduction and Embryology (ESHRE) and the European Society of GynaelogicalEndopscopy (ESGE) presented the novel ESHRE/ESGE classification system for female congenital genital malformations, which was based on anatomy. Vaginal dysplasia could be classified into four subgroups, and vaginal atresia could be classified as subtype V4, only generally described as partial or complete vaginal dysplasia in all cases [6]. In addition, congenital vaginal atresia is not further classified into different subgroups in this classification system.

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Nevertheless, some patients were found to be with vaginal atresia but could not be classified using the above classification [4–8]. The aim of the present study was to explore the characteristics of congenital vaginal atresia, further improve its classification, and therefore help the clinical diagnosis and treatment of congenital vaginal atresia.

Materials and methods

Patients

From March 1984 to March 2015, 67 patients with congenital vaginal atresia were admitted to the Department of Gynecology, Fuyang Clinical College of Anhui Medical University, China. The patients were aged 13 to 35 years. Congenital transverse vaginal septum and hymenal atresia were excluded by physical examination at admission. The patients' data were analyzed retrospectively. All patients consented to the surgical treatments. The study was approved by the ethical committee of Fuyang Clinical College of Anhui Medical University, China. The need for individual consent was waived by the committee because of the retrospective nature of the study.

Classification of vaginal atresia

The patients were classified according to physical examination, hysteroscopy, ultrasound, and/or computed tomography. Classification was based on previous systems [4–8]. Patients who could not be classified using these previous systems were examined in order to create new categories for the 67 patients.

1) Lower vaginal atresia (25 patients). Most patients had imperforated and bulging hymen with upper vaginal hematocele. Ultrasound examination showed normal development of the uterus and cervix, with occasional hematocele in the uterine cavity.
2) Complete vaginal atresia (36 patients). All patients showed cervical atresia. Ultrasound examination showed hematocele in the uterine cavity and the cervix, with a relatively long distance between the lower segment of the uterus and the hymen, varying between 3 and 10 cm
3) Upper vaginal atresia (one patient). The patient also showed cervical atresia. Gynecological examination showed normal width but shallow vaginal depth. Ultrasound examination showed little hematocele in the uterine cavity, and a 2-cm of loose solid atresia between the lower segment of the uterus and the top of the vagina.
4) Top vaginal atresia (five patients). Patients had normal depth and width of the vagina, but had uterine abnormalities. They had a blind vaginal ending and the uterine orifice could not be seen. Case 1 was admitted to the hospital because of severe abdominal pain and left lower quadrant abdominal mass for four months after her marriage. She was diagnosed with uterusdidelphys associated with left type i cervical atresia (Fig. 1). Case 2 was admitted to the hospital because of a lack of menarche, periodic hypogastralgia for half a year, and pelvic mass for a month. She was diagnosed with complete uterusbicornis and type i cervical atresia (Fig. 2). The third patient (20 years old) was hospitalized because of abdominal pain, purulent vaginal discharge, and lower quadrant abdominal mass for three months. She was diagnosed with uterus didelphysand right type i cervical atresia with infection. This patient had underwent ostomy at the top of the rightvagina and cecum of the uterine isthmus 5 years before for severe dysmenorrhea to release accumulated menstrual blood, but incomplete atresia of the stoma, and subsequent hematocele andinner cyst infection at the uterine isthmus were found (Fig. 3). The remaining two patients were diagnosed with primordial uterus associated with top vaginal atresia.

Classification of cervical atresia

Cervical atresia may be classified into four types: type i (normal uterine isthmus; atresia at the histological internal os, with a normal isthmus and normal anatomical internal os); type ii (short and solid cervix with a round end; atresia at the whole isthmus, including anatomical and histological internal os); type iii (complete cervical atresia); and type iv (absence of uterine isthmus, the lower uterus body is directly connecting with a blind cervical canal, uterine orifice could not be seen, and uterus isthmus is missing) [9]. The classification of cervical atresia according to vaginal atresia of the 67 patients is presented in Table 1.

Results

Among the 65 patients who underwent surgery, most were treated with the appropriate therapeutic options and surgical procedures according to their specific anatomical structures, achieving satisfactory results, except one patient with complete vaginal atresia and type i cervical atresia that underwent hysterectomy + vaginoplasty, and one with complete vaginal atresia and type ii cervical atresia that was treated by combined abdominoperineal artificial vaginoplasty + tracheloplasty (the patients had cervical re-atresia after surgery, and needed reoperation).

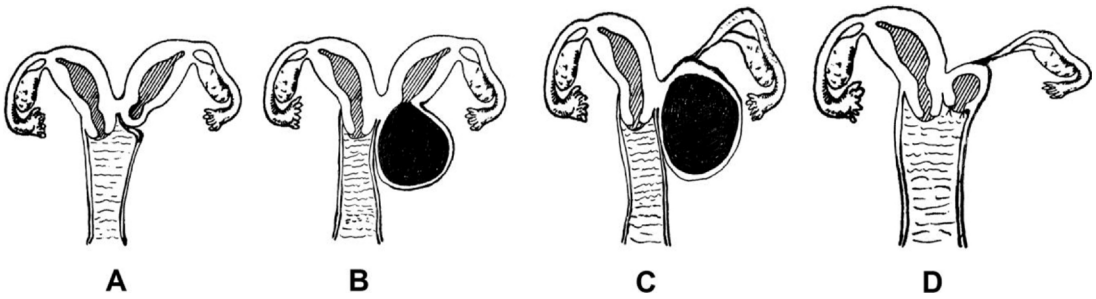

Fig. 1. Schematic diagram of diagnosis and classification of congenital vaginal atresia. (A) Type I: lower vaginal atresia (uterus is well developed). (B) Type II: complete vaginal atresia (accompanied with cervical atresia). (C) Type III: upper vaginal atresia (accompanied with cervical atresia). (D) Type IV: atresia at the vaginal top (accompanied with primordial uterus or cervical atresia).