



# Vaginal reconstruction for distal vaginal atresia without anorectal malformation: is the approach different?

Andrea Bischoff<sup>1</sup> · Veronica I. Alaniz<sup>2</sup> · Andrew Trecartin<sup>1</sup> · Alberto Peña<sup>1</sup>

Accepted: 20 June 2019 / Published online: 29 June 2019  
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## Abstract

**Introduction** Distal vaginal atresia is a rare condition and treatment approaches are varied, usually driven by symptoms.

**Methods** A retrospective review was performed to identify patients with distal vaginal atresia without anorectal malformation. Data collected included age and symptoms at presentation, type and number of operations, and associated anomalies.

**Results** Eight patients were identified. Four presented at birth with a hydrocolpos and four presented with hematometocolpos after 12 years of age. Number of operations per patient ranged from one to seven with an average of three. The vaginal reconstruction was achieved by perineal vaginal mobilization in four patients and abdomino-perineal approach in four patients. One patient, with a proximal vagina approximately 7 cm from the perineum, required partial vaginal replacement with colon. In addition, she had hematometocolpos with an acute inflammation at the time of reconstruction despite menstrual suppression and drainage which may have contributed to the difficulty in mobilizing the vagina. In five patients, distal vaginal atresia was an isolated anomaly. In the other three cases, associated anomalies included: mild hydronephrosis that improved after hydrocolpos decompression (2), cardiac anomaly (2), and vertebral anomaly (1).

**Conclusion** In this series, a distended upper vagina/uterus was a common presentation and the time of reconstruction was driven by the presence of symptoms. Drainage of the hydrocolpos/hydrometrocolpos with menstrual suppression in post-pubertal patients, followed by further work-up, and planned reconstruction is a good surgical strategy.

**Keywords** Distal vaginal atresia · Agenesis of the lower vagina · Hydrocolpos · Hydrometrocolpos · Imperforate hymen · Transverse vaginal septum

## Introduction

Congenital agenesis of the lower vagina is a rare condition. Diagnosis can be made with hydrocolpos on antenatal or post-natal ultrasound, though more common diagnosis is made during adolescence with symptoms of primary amenorrhea, cyclic abdominal pain, and sometimes an abdominal mass. Clinically, distal vaginal atresia presents similarly

to an imperforate hymen and a transverse vaginal septum, however, differentiation is imperative for proper surgical planning [1–5].

The hymen is the distal most part of the vagina and various hymenal anomalies have been described. When completely imperforate, a thin and sometimes bulging membrane can be identified. Hymenectomy is performed for definitive treatment and post-operative dilation is not required [3].

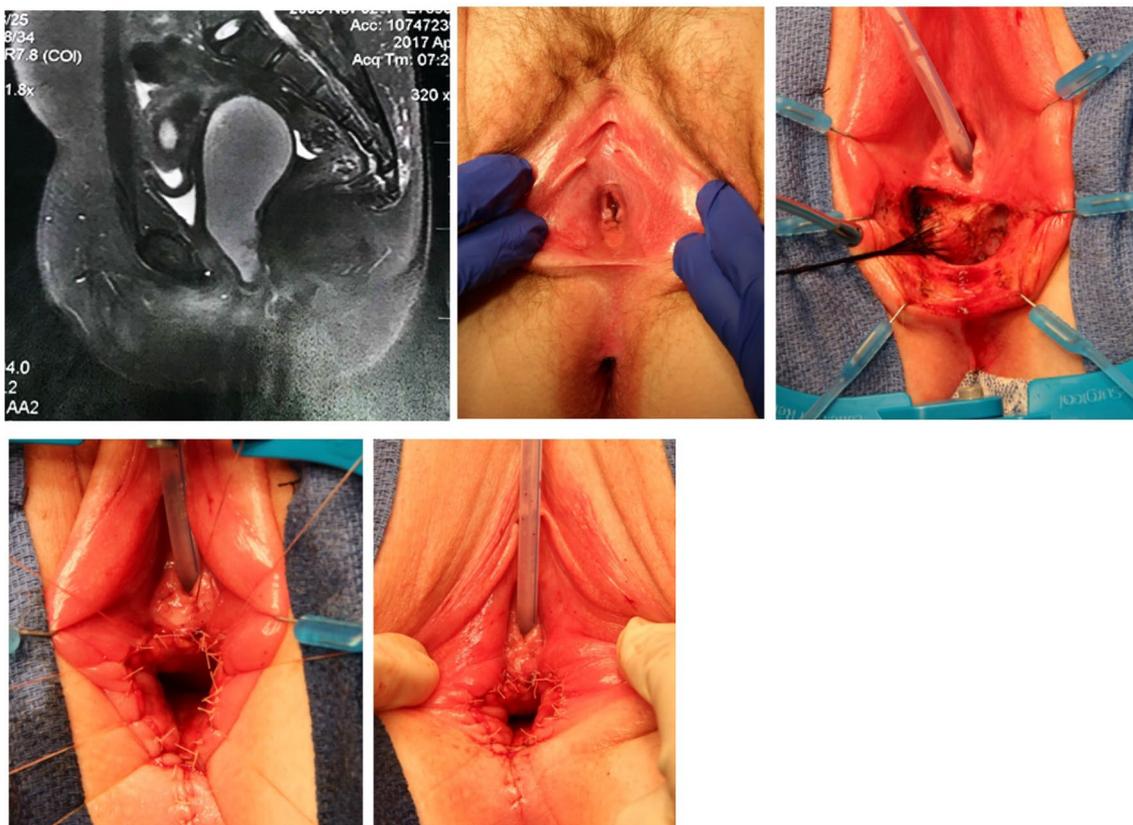
A complete transverse vaginal septum is an obstructive vaginal anomaly that can be located in the proximal, mid, or distal vagina. The septum can be thick or thin and there are typically normal vaginal walls on both sides of the septum. For surgical treatment, the septum is excised and the proximal and distal vaginas are anastomosed. In some cases, vaginal mobilization is required. Post-operative dilation is recommended to assure patency and adequacy of the anastomosis [5].

Patients with distal vaginal atresia have an absent vaginal opening and a proximal vagina that is located at different

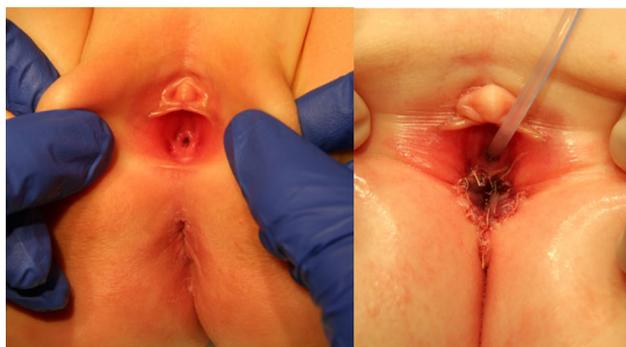
✉ Andrea Bischoff  
andrea.bischoff@childrenscolorado.org

<sup>1</sup> Department of Pediatric Surgery, Children's Hospital Colorado, International Center for Colorectal and Urogenital Care, University of Colorado, 13123 East 16th Avenue Box 323, Anschutz Medical Campus, Aurora, CO 80045, USA

<sup>2</sup> Division of Pediatric and Adolescent Gynecology, International Center for Colorectal and Urogenital Care, Children's Hospital Colorado, Aurora, CO, USA



**Fig. 1** Patient with distal vagina atresia, at a short distance from the perineum, repaired through perineal mobilization only



**Fig. 2** Patient with distal vaginal atresia that required abdomino-perineal mobilization of the vagina

lengths from the perineum. Depending on the distance from the perineum, vaginal mobilization through the perineum can be achieved or abdomino-perineal approach may be needed. The surgeon should also be ready for a partial vaginal replacement if needed [1].

Our aim was to review our cases of distal vaginal atresia without anorectal malformation and provide insights on how to manage these patients (Figs. 1, 2, 3).

**Fig. 3** Patient with distal vaginal atresia and hematometrocolpos that required partial vaginal replacement with colon due to inability to mobilize a very inflamed vagina



## Methods

A retrospective review was performed to identify patients with distal vaginal atresia without anorectal malformation. The study period was from October 1998 until December 2017. Data collected included: age and symptoms at presentation, type and number of operations, and associated anomalies. Institutional IRB was obtained for this study (19-0388).

## Results

Eight patients were identified. Four patients presented prior to puberty (ranging 1 day to 6 months) with hydrocolpos. Of these four, one presented with sepsis shortly after birth, and another one with frequent urinary tract infections. The four patients who presented with hematometra had abdominal pain after 12 years of age.

Associated anomalies were present in only three patients: mild hydronephrosis that improved after hydrocolpos decompression (2), cardiac anomaly (2), vertebral anomaly (1), and Chiari malformation (1).

Number of operations per patient ranged from one to seven with an average of three. Operations were: permanent drainage of the vagina with tube or vaginostomy (9), single aspiration (1) that resulted in re-accumulation of blood, despite menstrual suppression.

Seven out of eight patients had drainage of the hydrocolpos or hematocolpos prior to definitive surgery. In four patients, vaginal reconstruction was achieved by vaginal mobilization through the perineum. The other four patients required abdomino-perineal approach. One patient, with a proximal vagina approximately 7 cm from the perineum, required partial vaginal replacement with colon. In addition, she had hematometrocolpos with acute inflammation at the time of reconstruction despite menstrual suppression and drainage (re-accumulation after single aspiration) which may have contributed to the difficulty in mobilizing the vagina.

The only complication was neo-vagina prolapse that required an ambulatory neo-vagina prolapse repair.

## Discussion

It is imperative that obstructive vaginal anomalies are correctly diagnosed prior to surgical treatment [6, 7]. In our series patients that underwent multiple surgeries resulted from an error in the initial management. The most common error was diagnostic, mistaking vaginal agenesis for an imperforate hymen and failing primary attempted reconstruction.

Re-accumulation of hematometrocolpos occurred when vaginal decompression was performed without placement of a vaginostomy tube and was another cause of multiple surgeries.

In the treatment of distal vaginal agenesis, drainage of the vagina with a vaginostomy tube prior to surgical reconstruction should be considered. Without drainage, obstructed menses and/or fluid causes a severe inflammatory reaction in the vagina, thickening the vaginal walls and making mobilization more difficult. When appropriate and if needed to delay surgery, hormonal suppression of menses can be used to prevent recurrent hematometrocolpos. Hormonal contraception (contraceptive pills, contraceptive patch, or depo provera), high dose progestin-only pills, or complete suppression of the hypothalamic pituitary ovarian axis with Depot Lupron can be utilized.

A distended vagina facilitates localization of the proximal vagina during surgery and thus there is concern about pre-operative drainage. This can be mitigated with the use of the vaginostomy tube which allows injection of the saline solution intra-operatively to re-distend the proximal vagina.

Our recommendation for a pre-operative image includes an abdominal ultrasound for diagnosis followed by an abdominopelvic MRI (that includes sagittal views), and injection of contrast material through the vaginostomy tube. The lateral view is also important during the contrast study, in the event that the surgeon decides to proceed with a trans-anorectal approach [8].

Based on the pre-operative studies, the surgeon can decide if the vagina can be reached through the perineum or not. We recommend bowel preparation pre-operatively for a possible transanorectal approach which can improve chances for perineal mobilization.

Vaginal dilation is not required when the repair is done at an early age. Depending on the age and maturity of the patient, post-operative vaginal dilation might be recommended to assure patency. Vaginal examination under anesthesia should be done prior to intercourse, as introitoplasty may be needed [9, 10].

## Conclusion

In this series, a distended upper vagina/uterus was the common presentation and the time of reconstruction was driven by the presence of symptoms. Immediate drainage of the hydrocolpos/hydrometrocolpos, followed by further work-up, and planned reconstruction is a good surgical strategy.

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