Adolescent and Adult Outcomes in Women Following Childhood Vaginal Reconstruction for Cloacal Anomaly

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Purpose: We examined outcomes in female adolescents and women who underwent vaginoplasty in childhood during genitourinary reconstruction for cloacal anomalies.

Materials and Methods: We retrospectively reviewed the medical notes on girls and women attending an adult specialist center for genitourinary anomalies. Data were collected on vaginal reconstruction, menstruation, sexual and reproductive function, and urological and gastroenterological outcomes.

Results: We identified 19 patients with a mean age of 22 years (range 13 to 35), of whom 16 (84%) underwent vaginoplasty in the first year of life. Nine of these 16 patients (56%) had required 1 (7) or 2 (2) further vaginal reconstructions to facilitate menstruation or sexual activity. The remaining 7 patients (44%) required no further vaginal reconstruction. Nine of the 19 patients (47%) had associated müllerian anomalies, obstructed menstruation developed in 5 (26%) and 1 required hemihysterectomy. Eight patients were sexually active, of whom 1 experienced difficult penetration. Three patients attempted to conceive, including 1 with a complex preterm delivery and 2 undergoing fertility treatment. Of the patients 74% underwent further reconstruction of the renal tract and 36% had an enteric stoma.

Conclusions: This study confirms the complexity of vaginal reconstruction in this group with a notable vaginoplasty revision rate. Müllerian anomalies were identified in almost half of the patients, a higher incidence than previously reported, and in a quarter obstructed menstruation developed in puberty. A specialist team with gynecologic input should treat patients with cloacal anomalies. Outcome data are sparse. There remains a need for well planned, prospective cohort studies that include assessments of psychological, sexual and reproductive outcomes.

Key Words: urogenital abnormalities, vagina, cloaca, reconstructive surgical procedures, treatment outcome

Abbreviations and Acronyms

PSARVUP = posterior sagittal anorectovaginoplasty

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Persistent cloaca, defined as confluence of the rectum, the vagina and the urethra into a single common channel, has an estimated incidence of 1/50,000 live births.[1] It is a subtype of anorectal malformation. A wide spectrum of anomalies can occur that can have immediate and lifelong implications. Short-term outcome data on surgical intervention are well reported but long-term outcome data and quality of life data are less well represented in the current literature.

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After presentation at birth immediate management focuses on patient stabilization and decompression of the hydrometrocolpos, a consequence of urinary reflux into the uterus. Improved pediatric management strategies have increased patient survival into adulthood. However, little literature exists on the outcomes of early vs late intervention in this population.

We reviewed adolescent and adult outcomes after vaginal reconstruction in childhood for cloacal anomalies. In this retrospective review we examined adolescent and adult outcomes in patients who underwent vaginoplasty in childhood during genitourinary reconstruction for cloacal anomalies.

MATERIALS AND METHODS

After receiving local approval we identified patients with cloacal anomalies who presented for review at the adolescent and adult urogynecology clinic at our tertiary institution through a period of years. Study inclusion criteria included a diagnosis of cloacal anomaly with primary vaginal reconstruction in childhood. We excluded from study patients with disorders of sexual differentiation and cloacal extrophy.

After identifying the study population we reviewed the medical notes. Collected data included patient characteristics, initial reconstructive procedure, operation complexity, reoperation rate, intervention for obstructed menstruation or sexual activity, renal anomalies and renal function impairment, and müllerian anomalies. Final urinary and fecal continence was assessed.

RESULTS

We identified 19 patients with a mean age of 22 years (range 13 to 35). Anomalies at birth were recorded (see table). Of the patients 37% had an associated anomaly, which involved the renal tract in 4 of 7 (57%). Müllerian anomalies were noted in 10 of 19 patients (52%).

The primary procedure was performed in the first year of life in 16 of 19 patients (84%) (see Appendix). Five patients (26%) underwent PSARVUP while 8 (42%) underwent vaginoplasty. The difficulty in assessing the primary procedure when the patient has moved from a pediatric hospital to adolescent/adult care was clear in this cohort as we could not identify the nature of previous vaginoplasties from the referral letters. In 3 of 19 patients (16%) the primary procedure was not identified and 1 did not undergo a reconstructive procedure.

Patient characteristics at birth

Pt No.	Associated Anomalies*
2	Tracheal-esophageal atresia, dysplastic kidneys
3	Solitary kidney
4	Solitary kidney
5	Vertebral anomaly
9	Solitary kidney, vertebral anomaly
14	Single pelvic kidney

* None in patients 1, 6 to 8, 10 to 13 and 15 to 19.

Of the 16 patients (84%) who underwent a primary reconstructive procedure in childhood 9 (56%) required a further vaginal reconstructive procedure and 2 (12.5%) required 2 further vaginal reconstructions each to facilitate menstruation and achieve sexual activity. The Appendix shows the procedures.

Obstructed menstruation was documented in 5 of 19 patients (26%) and 1 (5%) required hemihysterectomy. Eight of 19 patients (42%) were reported to be sexually active, of whom 1 identified difficult penetration. Three patients in our cohort were actively attempting to conceive with assisted conception methods and 1 had a complex preterm delivery.

Further reconstructive procedures on the renal tract were required in 14 of 19 patients (74%), including 1 and 2 further procedures in 4 (28.5%) and 1 of 14 (7%), respectively. Spontaneous voiding was reported in 9 of 19 patients (47%) and 10 (52%) required catheterization via the urethra or a Mitrofanoff channel for successful bladder emptying. Three of these patients had a neobladder with a Mitrofanoff channel and 4 had an ileocystoplasty, of whom 2 each catheterized via the urethra and via a Mitrofanoff channel. Two patients had a Mitrofanoff channel to the native bladder and 1 had an end ureterostomy. Renal function was impaired in 4 of 19 patients (21%) with renal transplantation in 1.

Seven patients had a colostomy/ileostomy for bowel evacuation with an antegrade continence enema channel in 1. One patient had a perineal sinus from a rectal stump after end colostomy formation. All other patients were continent of feces.

DISCUSSION

In our series of 19 patients the mean age was 22 years, making this one of the longest reported series of followup for cloacal anomalies. Our study was a retrospective review of the medical notes with the associated limitations.

There is a well documented correlation between cloacal anomalies and müllerian anomalies with a reported rate of 30%.[2,3] In our cohort the rate was 47%. This could be explained by the difficulty of predicting/identifying müllerian structures in infancy due to the poor sensitivity and specificity of radiological investigations.[4] Thus, our report could incorporate the structures that are only identified after puberty. Postpubertal studies echo this effect, identifying an obstructed menstruation rate of 36% after reconstruction in infancy.[5] Our obstructed