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Y – TYPE FEMALE URETHRAL DUPLICATION WITH URINARY INCONTINENCE– A CASE REPORT

Synopsis of Video

Aim

An unusual presentation of Female urethral duplication with urinary incontinence is presented for its rarity and the problems that are encountered during surgical management of this case are discussed.

A 16-year-old girl presented with dribbling of urine since childhood. At birth she was evaluated for ambiguous genitalia and diagnosed as a genetic female. At 6 months of age underwent bladder neck, urethral, vaginal reconstruction and cohens reimplantation of both ureters. During the reconstruction, gonadal biopsy was taken which was reported as ovary. Incontinence continued. At 8 years of age she underwent an unsuccessful repair of urethrovaginal fistula and the phallic urethra was left undisturbed. She has attained menarche at the age of 13 and started menstruating regularly.

She continued to have urinary incontinence. She voided mostly per orthotopic urethra but also voided per clitoral [phallic] urethral opening. After voiding she continue to dribble urine from vaginal pool which gets collected through the urethrovaginal fistula

On clinical evaluation she had a prominent clitoris with phallic urethra opening in it, absent labia minora, hypospadiac orthotopic urethra with urethrovaginal fistula. Anus was situated close to vaginal introitus due to poorly formed perineal body. Urodynamics study showed evidence of stress leak, which is due to the surgical procedure in childhood, which has resulted in a short orthotopic urethra with poor urethral supports.

Static cystography showed good capacity bladder with no vesicouretric reflux and competent bladder neck. During micturation phase phallic urethra was visualized joining orthotopic urethra with vaginal pooling of contrast. Intravenous urography showed malrotated left kidney. Computer tomographic cystography showed uterus didelphys with single cervix. Endoscopic evaluation showed roomy vagina with single cervix, short hypospadiac orthotopic urethra with urethrovaginal fistula at 1 cm from the external meatus and phallic urethra joining the orthotopic urethra 0.5 cm proximal to it. The childhood reconstruction of urogenita lsinus into distal urethra has resulted in urethrovaginal fistula.

Prominent clitoral hood may be due to ventral development of urethra with lack of caudal curvature. Duplication of urethra might be due to delayed union of mullerian duct with urogenital sinus.

Surgery was contemplated with the main aim of achieving continence. Plan was to make the patient void urine through a single opening and to address the stress incontinence at the same sitting as the patient was not willing for another surgical intervention. Aesthetically acceptable appearance of external genteelism was also planned.

In exaggerated lithotomy position the vaginal introitus was stretched. The vagina was so roomy that no other manipulation was needed to improve the access. After mobilizing the orthotopic and phallic urethra, phallic urethra was found tobe robust and longer. Orthotopic urethra was flimsy, short and poorly developed. It was laid open on its posterior wall and closed with vicryl sutures. The tissue in between the two external urethral openings were mobilized and kept as tongue shaped flap over the urethral closure site as a live tissue interposition. Spatulated phallic urethra was brought down caudally and neo external urethral meatus was created .TVT placed loosely over the flap. Vaginal reconstruction was done. Clitoral prominence was addressed with simple plication sutures. Postoperative period was uneventful. Urethral catheter was removed on the 14th postoperative day and suprapubic catheter was removed after two days after checking for post void residue. Relocation of phallic urethra facilitated caudal migration of clitoris and simple plication has given the acceptable cosmetic appearance. The patient is on regular follow-up and she is voiding per single urethra and free of urinary leakage.

Female urethral duplication is extremely rare and literature reports about obstructive features with duplication. This case presented with urinary incontinence, probably of iatrogenic aetiology. Preserving of the phallic urethra, which is an accepted step during reconstruction of

duplication, has proven to be right and has formed the ideal substitute for the poorly developed orthotopic urethra.