"Caudal Duplication Syndrome in a Young Girl: A Complex Case of Gastrointestinal and Genitourinary Duplication with Spinal Dysraphism"

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No Conflict of Interest

Introduction

Caudal Duplication Syndrome is an extremely rare congenital anomaly involving duplication of distal gastrointestinal tract, genitourinary structures along with distal spinal & lower limb anomalies. Anatomical, functional & cosmetic reconstruction remains a major challenge.

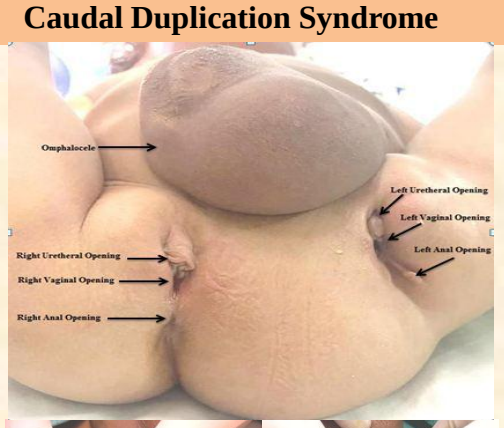
Patient: Medical Summary

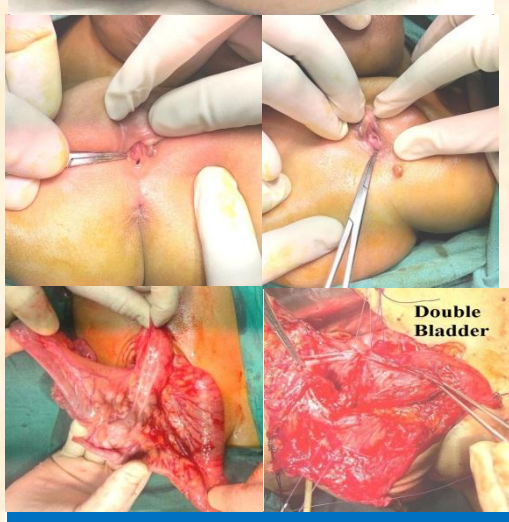
DOB: 5th July 2019 | Age: 6 years

Delivery: via C-Section. Born with double vaginal urethral & anal openings

Diagnosis:

Caudal Duplication Syndrome





Investigations

U/S, MRI/CT: Bladder & rectal duplication, abdominal wall defect 7x10 cm, spinal anomalies (thoraocic hemivertebrae, diastomatomylia & meningocele), ovaries and uterus not identified. Separate ureters draining into ipsilateral bladders.

Barium Study: Malrotation,

Barium Enema: Rectosigmoid and colonic duplication till cecum.

EUA & Cystoscopy: No sphincter response on nerve stimulation; trabeculated right bladder. Unable to cannulate on left side

CT Urogram: Ipsilateral ureters draining into separate urinary bladders.

Key Findings

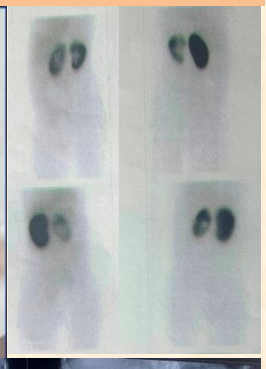
Two swellings at birth (umbilical & lower back). Two separate set of perineal openings including 2 separate urethral, vaginal and anal orifices (5 cm apart) Complete bladder & rectal duplication Omphalocele (10×10 cm). Malrotation on contrast study (DJ junction on right side), Hemi vertebrae (thoracic/ lumbar), tethered cord, myelomeningocele on MRI.

Major Procedures

05-02-2021 (1.5 yrs.):

Myelomeningocele Repair.

23-07-2022 (3 yrs.): Exploratory laparotomy. Left total colectomy, Urinary Bladders unification, Omphalocele repair, Urethral mobilization & left urethral intra vesical closure by Pursestring suture



Current Status

The child walks with support but has no bladder or bowel control. She is school going. On going urinary leakage from re-fistulization of left urethera. The patient has two vaginal openings and weighs 16 kg. On MCUG grade 5 VUR on left side. Renal Scan shows Right Kidney Function 70% & Left 30%. She remains under regular follow-up on CIC.

Discussion

Caudal Duplication Syndrome (CDS) is an exceptionally rare congenital anomaly, with fewer than 100 cases reported (Dominguez et al., 1993; J Pediatric Surgery, 2001). It involves varying degrees of duplication of the gastrointestinal, genitourinary, and spinal systems, posing significant diagnostic and surgical challenges. Due to complex anatomy, meticulous preoperative planning & staged surgical intervention is pivotal if needed. Management focuses on restoring function, improving cosmesis and minimizing risks to vital structures. A multidisciplinary team—including Pediatric surgeon, neurosurgeon, pediatric urologist, radiologist & afterward gynaecologist is crucial for achieving favourable long-term outcomes.

Conclusion

This rare case of Caudal Duplication Syndrome was managed through staged surgical interventions and multidisciplinary care. The child has shown good physical growth, improved mobility, and resolution of major GI issues. On going challenges include urinary leakage, lack of urinary & fecal continence along with preservation of renal function. Continued care aimed at maximizing functional independence and quality of life.

References

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Acknowledgement:
Muhammad Umer