

## CASE REPORT

# Complete Tubular Duplication of Colon associated Genito-Urinary Duplication in Female Baby (Caudal Duplication Syndrome): A Rare Case

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### ABSTRACT

Caudal Duplication Syndrome is a rare case, congenital anomalies, involved the alimentary tract duplications and urogenital tracts duplication. The incidence is 1:100.000 births. We present a case of two months old female baby with abnormal genitalia and imperforate anus related to caudal duplication syndrome. On physical assessment we found duplication of vagina with single uterus and urethra with anorectal malformation (anovestibular and rectovaginal type). Intraoperative findings showed double-duplication of ascending colon, transverse, descending and half of sigmoid with duplication of the rest of sigmoid and rectum; left-sided rectum was adjacent to left vestibule and right-sided rectum adjacent to the right vagina; duplication of bladder and urethral duplication. On fourth, the reconstruction surgery was performed to correct digestive abnormality with Posterior Sagittal Anorectoplasty (PSARP) and separating duplication segment using stapler. Stapler was enough to separate duplication. Further reconstructive surgery were needed to achieve better gastrointestinal and genitourinary function outcome.

**Keywords:** Caudal duplication syndrome, Colonic Duplication, Genitourinary duplication, Congenital

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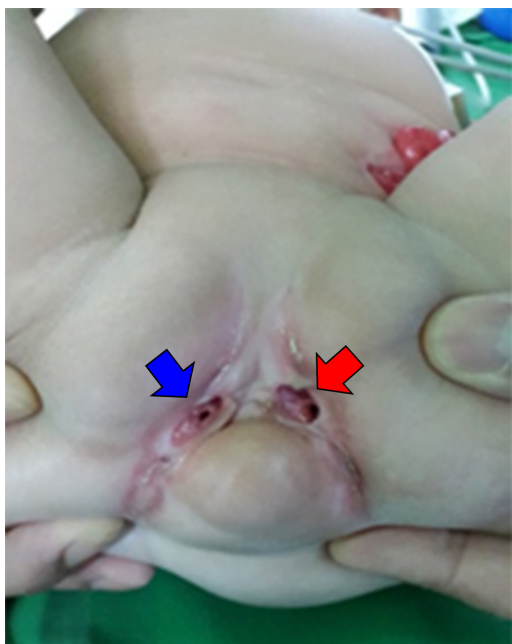
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### INTRODUCTION

Caudal Duplication Syndrome (CDS) is congenital anomalies, involving complete or incomplete duplication of the alimentary tract, spinal cord and urogenital tracts (1). Caudal duplication syndrome is very rare type of alimentary tract duplication found in clinical practice. its incidence about 1:100.000 births (1). Caudal duplication derived from the embryonic cloaca and notochord duplicated in various degrees (1,2). It was defined as cystic or tubular structure, often accompanied by sharing common muscular wall and vascular in 15% cases (3). Therefore, a well setting surgical and postoperative approach with individual-based is mandatory to achieve gastrointestinal and genitourinary function. We present an extremely rare case of female baby with CDS, surgical approach and management in our institution.

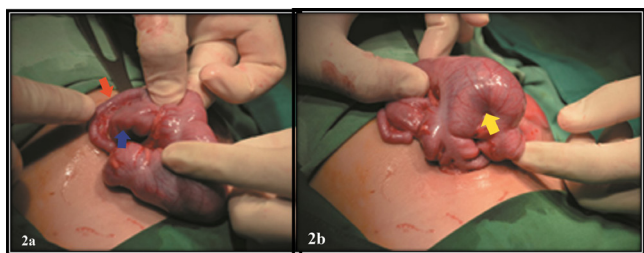
### CASE REPORT

A two-months old female baby was referred to our hospital with urogenital duplication and imperforate anus. On physical examination, we found duplication of vagina (with single uterus) and urethra duplication with double anorectal malformation krickenbeck rare other variant (anovestibular and rectovaginal-type) (Figure 1). The baby passed stool from two respective genitalia openings. The pregnancy and delivery history was normal. The baby was aterm, normal birth weight, and blood analysis within normal ranges. There was no abnormality of spine and patient didn't have any neurological deficit. Sacral ratio of this patient was 0.80. After admission, she underwent divided colostomy procedure for diversion (duplication colostomy divided). Complete colostomy duplication was done and four stoma divided were made (one pair for each colon) on the left side of abdomen. No exploration laparotomy performed. The patient showed good condition with viable stoma post surgery. Four months later, diagnostic laparotomy combined with contrast imaging were done for assessing anomaly of both digestive and

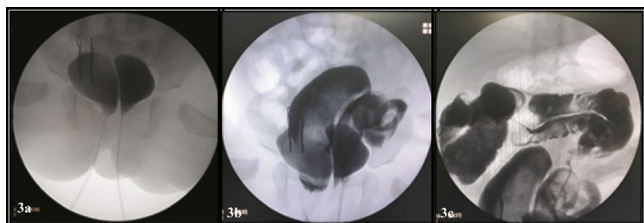


**Figure 1: Genitalia Examination showed left-sided vagina with vestibular fistula (red arrow) and right-sided vagina with vaginal fistula (blue arrow)**

genitourinary tract structure anatomy. During surgery, quadruplication (double duplication) of ascending, transverse, descending and half of sigmoid colon with duplication of the rest of sigmoid and rectum were found; left-sided rectum was adjacent to left vestibule (rectovestibular-type) and right-sided rectum was adjacent to the right vagina (rectovaginal-type) (Figure 2). Intraoperative retrograde urethrography was done and we found duplication of bladder, urethra, and single uterine (Figure 3). No abnormality and duplication of ureter and kidney on examination.

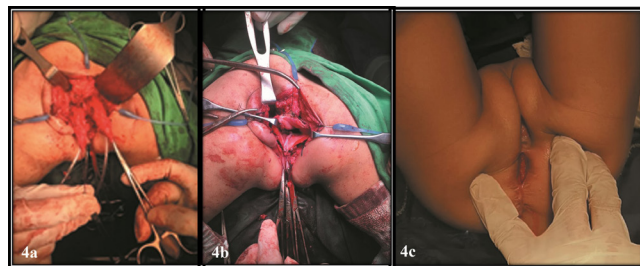


**Figure 2: Identification of colonic duplication; 2a) Appendix (red arrow), terminal ileum (blue arrow); 2b) Ascending colon (black arrow), bifurcation of colon (yellow arrow)**



**Figure 3: Imaging Study; 3a) Urethral and Bladder Duplication; 3b) Contrast filling from fistula to colon; 3c) Contrast filling from stoma**

The reconstruction surgery was performed to treat digestive abnormality with posterior sagittal anorectoplasty (PSARP) from anovestibular and rectovaginal fistula. Then, rectal duplication was separated by using two-75 cm-length stapler (Figure 4). Colostomy closure was done by stapling method. The genitourinary abnormalities will be evaluated further after the digestive abnormality correction. No complication after surgery and patient was home from hospital with good condition. On follow up, the outcome was good. There were no voluntary bowel movement, soiling, and constipation.



**Figure 4: PSARP procedure of rectovagina and anovestibular duplication; 3a) before separation; (3b) after separation; 4c) new anal**

## DISCUSSION

Caudal duplication syndrome is an uncommon malformations, a very rare case, with incidence 1:100.000. In a complex cases, a multidisciplinary team approach is mandatory. They are defined as a cystic or tubular form, accompanied by sharing common muscular wall and vascular supply (1-3). The first reported case was published by Galder in 1733 (1-3). Ladd was the first introducing the term “duplication of the alimentary tract” in 1937 (3). The rarest case of tubular duplications was affecting several segments of colon or entire colon (4). Total colon or complete colon duplication was a term applied to the duplications involving four or more segments of colon without involvement of small intestine segment. Duplication can be found in any part of alimentary tract. Only 4%-18% are in colonic region (4).

The aetiology of these anomalies is still unclear. Jellali et al. reported CDS were found at neonatal period because of their association with anal atresia (5). Female have a high prevalence of rectogenital fistulas and related duplications of internal or external genital structures (5). Preoperative diagnosis of alimentary tract duplications is often difficult. Ultrasonography was helpful establishing preoperative diagnosis and screening for 10-20% of multiple lesions (4,5). Once the diagnosis was made, a surgical resection, performed to avoid complications, was reserved for symptomatic cases. Indication for surgical intervention depends on acute setting, symptoms, types of duplications and associated anomalies.

There was many operating technique used for colonic duplication reported. Riedel, Brunschwig, and Sarpel

et al. had tried multiple technique for treating colonic duplication such as dividing the septum between rectum, anal orifice, and perineum reconstruction (1,2). Mortality rate was 4%-8% (2). Total tubular colonic duplication treated before the onset of complications generally have a favourable prognosis, hence the importance of early diagnosis is mandatory (5).

The limitation of this report was the complexity of abnormality. There was multiple procedure and multidisciplinary approaches involved in treating this patient.

## CONCLUSION

Caudal Duplication Syndrome, a rare congenital anomaly, need careful investigation and well setting surgical procedure due to its nature syndrome and deformities complexity. Further surgical correction should be planned and multidisciplinary approaches were needed to achieve better gastrointestinal and genitourinary function outcome.

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