Newborn with complete double penis and two separate scrotums: A case report

Ahmed Maher *, Tarek Abdelazeem Sabra, Hussein Ibrahim, Mahmoud Mostafa

Pediatric Surgery Unit at Assiut University Children Hospital, Assiut, Egypt

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ABSTRACT

INTRODUCTION: We present a newborn with double penis and double scrotum as a part of a caudal duplication syndrome (CDS) which is a condition includes duplication of the distal organs of the body. It is crucial to have knowledge about it to be able to be identified.

PRESENTATION: A male newborn presented with double penis, double scrotum double urethra, double colon, and double imperforate anus. After work up a low descending colostomy was done (4 stomas of duplicated colon) and started feeding with normal passage from colostomy.

DISCUSSION: The cause of CDS is unknown many theories have tried to explain that, but the most accepted theory is failure of monochorial twins to separate completely. CDS may be associated with other congenital anomalies as imperforate anus, renal anomalies, and omphalocele.

CONCLUSION: CDS is a very rare condition which needs multidisciplinary team to manage and needs staged repair. Most pediatricians and pediatric surgeons are unable to diagnose it, we add a case of CDS to the literature.

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1. Introduction

Caudal duplication syndrome (CDS) is an extremely rare condition that includes duplications of the caudal part of the body including genitourinary and gastrointestinal systems along with the vertebral column. CDS is first described by Dominguez [1]. We present a neonate with CDS with a double high imperforate anus and double scrotum. This case report has been reported in line with the SCARE criteria [2].

2. Presentation

A male newborn aged 6 h weighted 2.6 kg with no history of prenatal exposure and could not be diagnosed prenatally, was admitted to the NICU (neonatal intensive care unit) in our institution. He was born of a non-consanguineous marriage. On physical examination, the patient has a double penis (diphalus) with double pinpoint urethral opening, through both the patient micturates normally, double scrotum containing one palpable testis for each Fig. 1. He has two separated natal clefts with no anal orifice. Initial diagnosis of anorectal malformation with CDS is established. The Invertogram showed a high variety of anorectal malformation, the US revealed a hydrenephrotic left kidney, his echocardiography showed no abnormalities. Subsequently, he underwent a low descending colostomy that revealed a duplicated colon(4 stomas) Fig. 2. The patient started oral feeding and tolerating and pass regularly from the colostomy (The two proximal ends). The Subsequent distal Loopgram (from the distal two openings showed duplicated colon down to the rectum with no fistulae with the urinary tracts Fig. 3. The MRI (Magnetic Resonance Imaging) showed left moderate hydrenephrosis due to left congenital PUJ (pelvi-ureteric junction) stenosis and duplicated urinary bladder. Also, there is a tethered cord with no duplication of the vertebral column Fig. 4. The patient discharged 3 days post-operatively for follow up and subsequent planning for a staged repair.

3. Discussion

Caudal duplication syndrome was presented in the literature by Dominguez [1] Nevertheless, the exact etiology still obscured. However, its embryological etiology is claimed to be conjoined twins with incomplete fusion of some areas of the body giving duplication of these parts or monochorial twins with incomplete separation lastly, fibrous band dividing the caudal embryo into two [3,4]. An event in early embryological development is suggested by Alfadhel et al., relying on formation of the pathology by the derivatives of the three germ layers [5].

In our case, we found two natal clefts with an imperforate anus (IA) which is commonly seen in CDS. Variant forms of imperforate anus were mentioned in cases of CDS, two anal openings on...
Fig. 1. Shows our patient with a double penis, a double scrotum, and two natal clefts with absent anal orifice.

Fig. 2. Shows a low descending colostomy that demonstrates four stomas POD3: the two white arrows indicate the duplicated proximal colon and the two black arrows indicate the duplicated distal one.
both sides, normal opening at one side, imperforate one at the other side and imperforate anus at both sides. Timing of occurrence of CDS as suggested by Bajpai et al. and Bannykh et al. [3] at 25th which falls in a time interval of cloacal membrane formation and its rupture (19th day and 9th weeks of gestation) [6] that can explain the increased frequency of IA with cases of CDS.

Double penis with two urethra and septated urinary bladder found in our case represents the caudal duplication of the genitourinary system in addition to a tethered cord and congenital left PUJ obstruction. Associations of these anomalies (caudal tubal duplication) in our case may favor the caudal twining theory as the mechanism behind CDS. Table 1 provides an overview of various cases of CDS reported in literature.

4. Conclusion

CDS is a rare condition that needs a multidisciplinary team for its management that includes staged repair of duplications. In our case, we are planning to start staged repair at the age of 16 months.

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.
| Author(year)         | Age    | Gender | Anal region                        | External genitalia         | Internal genitalia   | Urinary system | GIS  | Neurologic/vertebral system                        |
|---------------------|--------|--------|-----------------------------------|----------------------------|----------------------|-----------------|------|----------------------------------------------------|
| Suppiger[1876]      | NB     | F      | DIA and duplex rectovulvar fistulas | DEG                        | DUT and DV           | DUB and DU     | DTC  | Vertebral duplication (L3 and caudal parts)         |
| Piccoli[1892]       | NB     | F      | Duplex anal orifices              | Duplex vaginal orifices, one external genitalia | DUT and DV |                  |      |                                                    |
| Volpe[1903]         | NB     | M      | DIA, right rectovesiceral and left rectourethral fistulas | DEG                        | DUB and DU           | Horseshoe kidney | DC distal to cecum | Spina bifida, meningocele |
| Bar et al.[1908]    | 14 days|M      | Blind anal dimple on each side of anal dimple | Bifid scrotum, one penis, two urethral orifices | DUB and DU           | DC distal to cecum |      |                                                    |
| Aitken[1912]        | NB     | F      | One normal anus and vulvar anus   | Uterus unicornuate,       | DTC                  | DTC + TI        |      | Vertebral duplication (L3 and caudal parts)         |
| Lesbre[1927]        | ?      | F      | Duplex anal orifices              | DEG                        | DUB and DU           | DTC             |      |                                                    |
| Ombredanne[1936]    | 2 years| F      | Right normal anus, left rectourethral fistula | DEG                        | DU                   | DTC             |      |                                                    |
| Aitken[1950]        | NB     | F      | DIA and duplex rectovaginal fistula | DEG                        | Bicornate uterus and DV | DTC + TI        |      |                                                    |
| Ravitch[1953]       | 4.5 years|M     |                                   |                             | DUB and DU           | DC distal to cecum | DTC + TI | Meningomyelocele                                    |
| Beach et al.[1969]  | ?      | F      | DIA                               | DEG                        | DUT and DV           | DUB and DU      |      |                                                    |
| Dutta et al.[1974]  | ?      | M      | Duplication of penis, scrotum     | DUB and DU                 |                      | Duplication of L5, split sacrum, hemivertebra |
| Veeraraghavan et al.[1983] | ? | M | DIA | Bifid scrotum | DUB and DU | DTC | Lateral deviation of sacrum, hemivertebra |
| Zamir et al.[1984]  | ?      | F      | DIA                               | DEG                        | DU and DV            | DUB             | DTC  |                                                    |
| Magalhaes et al.[1999] | NB | F      | DIA, left rectovulvar, right rectoperineal fistulas | DEG                        | DV                   | DUB and DU      | DTC  |                                                    |
Table 1 (Continued)

| Author/year                  | Age     | Gender | Anal region                                      | External genitalia                                                                 | Internal genitalia                      | Urinary system | GIS       | Neurologic/vertebral system. |
|------------------------------|---------|--------|--------------------------------------------------|-----------------------------------------------------------------------------------|------------------------------------------|----------------|----------|-----------------------------|
| Bannykh et al. (2001) [22]   | NB      | M      | Right normal anal orifice, left imperforate anus with rectourethral fistula | DEG, left hypoplastic penis with hypospadias, right scrotal testis, left bilobed intraabdominal testis | DU                        | DTC + TI       | Meningomyelocele, scoliosis, hypospasia of left side of the lumbar spine, lumbar vertebral duplication, Sacral duplication |
| Kros et al. (2001)-1 [23]    | NB      | F      | Ectopic anus at right fold, A rudimentary, non-functional anus to the two vaginal openings | Normal urethral orifice, duplex vagina orifice                                   | Duplication of cervix and DV (uterus unknown) | Pelvic right kidney, duplication of left ureter opening, urethra caudal to bladder neck, normal urethral opening | DTC       | Complete duplication of the spine from L4 downwards, abnormalities in vertebral segmentation, abnormally shaped vertebrae and sacrum, myelocele |
| Kros et al. (2001)-2 [23]    | NB      | F      | DIA, rectoperineal and rectovaginal fistulas       | Duplication of anus (high anorectal malformation)                                | DU and DV?                     | DUB and DU, dilated pelvis of the right kidney and DU, right VUR | DTC + TI   | Hemivertebra (T6, T10), abnormal curvature of sacrum Lumbosacral lipomingomyelocele, lumbosacral spinal dysraphism, spina bifida |
| Bajpai et al. (2004) [3]     | NB      | M      | Imperforate anus (high anorectal malformation)      | Bifid scrotum, double phallus                                                | DU and DV?                     | DUB and DU, dilated pelvis of the right kidney and DU, right VUR | DTC       | Duplication of sacrum, coccygeal vertebrae, terminal spine |
| Vijayaraghavan et al. (2004) [24]| 19-wk fetus | F | Duplication of anus Imperforate anus | ?                                      | DU and DV | ? DUB and DU | ? | Normal |
| Siebert et al. (2005) [25]   |        |       |                                                  | Vaginal atresia                                                                  | DU and DV                      | DUB and DU | DTC | |
| Jianhong et al. (2005) [26]  | F       |        | Right normal anal orifice, left low-level anal atresia | DEG                                                | DV?                        | Bilateral complete duplication of kidney and ureter, DUB and DU | DTC | Hemicorpus vertebral fusion of T11–T12, vertebral corpus subfissure of L5-S1 |
| Liu et al. (2009) [27]       | 13 years | M | Two anuses beside perineal raphe, right anus was normal, left was closed as a small anal dimple | Diphallia (1 penile shaft, 2 glans) | DV?                        | Bilateral complete duplication of kidney and ureter, DUB and DU | DTC | Hemicorpus vertebral fusion of T11–T12, vertebral corpus subfissure of L5-S1 |
| Author/year | Age | Gender | Anal region | External genitalia | Internal genitalia | Urinary system | GIS | Neurologic/vertebral system |
|-------------|-----|--------|-------------|--------------------|--------------------|---------------|-----|----------------------------|
| Taneja et al. (2009) [28] | NB | F | One anus, one urogenital sinus | DEG (urine output by the right orifice and feces by the left) | ? | Pelvic left kidney, two urethral orifices, bladder diverticulum, left VUR | ? | Complex malformations of the thoracic and lumbosacral spine, coccgeal vertebrae were absent, spinal cord duplication from T1, lipoma from L1 | Left-sided lipoma at the tip of the conus medullaris |
| Alfadhel et al. (2009) [5] | NB | M | Right rectum opening to anus and left rectourethral fistula | Normal | DUB | DTC + TI | | |
| Bansal et al. (2011) [29] | 2.5 years | F | Left normal anus, right anovestibular fistula | DEG | DUB and DV | DC with a single cecum | | Scoliosis, lipomyelomeningocele, tethered cord and hydrosyinx, Absence of right hemisacrum, Vertebral dysplasias and bifid L5 and sacrum |
| Acer al et al (2013) [4] | NB | F | DIA and duplex rectovulvar fistulas | DEG | DUB and DV | DU | Duplication of appendix, cecum, sigmoid colon and rectum | DC distal to caecum | DTC + TI | Spinal lipoma |
| Swaika et al. (2013)-1 [30] | NB | M | Duplex stenotic anal orifices DIA (right is low, and left is high with recto-bladder neck fistula) | Two hemiphalus | DUB and DU | DTC + TI | Spinal lipoma | Duplication of lower 3 ribs, double pelvis, and tetrapagus. |
| Al Alayet YF (2014) [31] | NB | M | Duplex anal orifices | Double perineum with DEG | DUB, Duplicated left pelvicalyceal system | DC with single caecum | hemivertebrae |
| Chaussy et al. (2015)-1 [32] | NB | F | Normal | DEG | DUB and DU | DTC + TI | Spinal lipoma | Duplication of lower 3 ribs, double pelvis, and tetrapagus. |
| Samuk et al. (2016) [33] | NB | M | Duplex anal orifices | Single penis with two Urethra and bifid scrotum | DUB and DU | DC to caecum | Tethered cord |
| Our case | NB | M | DIA | DEG Double penis and double urethral orifices | DUB, DU, left congenital PUJ stenosis and ipsilateral hydrenephrosis | DC to caecum | Tethered cord |

NB: Newborn. M: Male. F: Female. GIS: Gastrointestinal system. DIA: Duplex imperforate anus. DEG: Duplication of external genitalia. DUB: Duplication of uterus, DV: Duplication of vagina. DUB: Duplication of urinary bladder. DU: Duplication of urethra. VUR: Vesicoureteral reflux. PUJ: pelvi-ureteric junction. DC: Duplication of colon. DTC: Duplication of total colon. DTC + TI: Duplication of total colon and terminal ileum.
Fig. 4. Shows the MRI demonstrating a duplicated urinary bladder (the white arrows) and left hydronephrosis (the black arrow).

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

The case report is exempt from ethical approval in my institution.

Consent

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Ahmed Maher: main author of the paper and wrote the manuscript.

Hussein Ibrahim: literature review and revised the manuscript. Mahmoud Mostafa: supervising and editing. Tarek Abdelazeem Sabra: supervising and editing.

Registration of research studies

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