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Caudal 'duplication' or 'split' syndrome: Is there a misnomer?

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ABSTRACT

'Caudal duplication syndrome' was coined to describe the apparent duplication of organs derived from the hindgut, the neural tube and the adjacent mesoderm. Review of the anatomy suggests that the word 'duplication' may be a misnomer. This paper describes the management of 2 girls with caudal duplication syndrome who underwent multistage reconstructive surgery. Both had a large omphalocele and a severe diastasis of the pubic symphysis. The first patient also had an apparent duplication of the vulva, the perineum and the anus to either side of a wide midline. Each vulva contained a urethra, a hemi-clitoris with ipsilateral labium minor, and a hemi-vagina with hemi-uterus. The second child had an infrapubic sequestrated appendico-cecal duplication lying between two hemi-bladders each with ipsilateral ureter and urethra. The everted duplication split the single vulva longitudinally in the midline as far as the fourchette. To each side were a hemi-clitoris, and a hemi-vagina with hemi-uterus and ipsilateral fallopian tube. Analysis of our patients' anatomy and a literature review indicates for the most part 'hemi' organs on either side and suggests that the term 'duplication' is a misnomer such that caudal 'split' syndrome may be a more appropriate title.

Caudal duplication syndrome was coined to describe the apparent duplication of the organs derived from the hindgut, neural tube and adjacent mesoderm resulting from 'insults' at different stages of embryogenesis [1]. The condition is rare and only few reported cases describe the combination of anomalies of the colorectum, the lower urogenital tract, the abdominal wall and spinal dysraphism.

We report our experience with two girls, born 20 years apart and in different countries. We recommend that early management should be conservative and not compromise fecal or urinary continence. Complex multistage surgery should be patient tailored toward an anatomical, functional and esthetic reconstruction [2,3], and should be delayed until after functional and dynamic evaluation of all anatomical parts. A study of our patients' anatomy and a literature review suggest that the term 'duplication' in the title is, for the most part, a misnomer and caudal 'split' syndrome may be more appropriate.

1. Case reports

1.1. Patient 1

Born in 1980, the child was referred after a normal pregnancy and term delivery, because of a large omphalocele, severe pubic diastasis, and ano-urogenital malformations. To either side of a wide fatty midline there was a separate smaller vulva. Each contained a hemi-clitoris and ipsilateral labium minor, a complete vaginal orifice and a continent urethral orifice that passed urine. Posterior to each vulva was a perineum and an anus, with the left demonstrating an intact and continent sphincter and the right anus being open anteriorly. Both ani passed meconium. Routine pelvic radiology did not show any lumbosacral anomalies. Intravenous urography, examination under anesthesia, and cystoscopy through each urethra revealed two separate hemi-bladders lying side by side with each draining its single ipsilateral normal ureter and kidney. Bilateral vaginoscopy demonstrated a full-length hemi-vagina with cervix. At neonatal repair of the omphalocele, a laparotomy confirmed two separate hemi-bladders each with an ipsilateral single ureter, two hemi-uteri each with ipsilateral fallopian tube, and a normal ovary bilaterally (Fig. 1a). The hemi-bladders were opened longitudinally on their medial aspects and the right bladder neck was detached from its urethra and closed.

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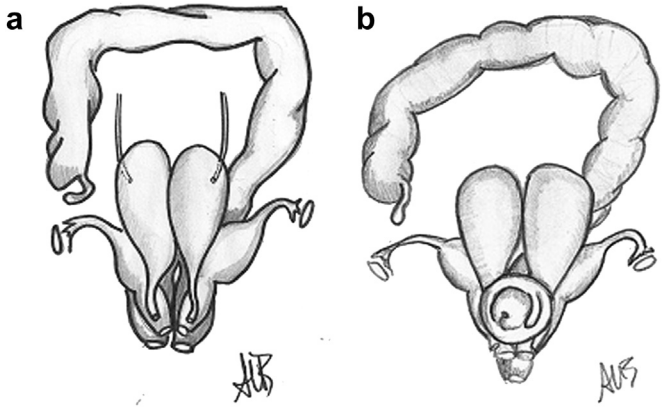

Fig. 1. (a) Patient 1 had two separated hemi-bladders lying side by side, and separate Mullerian structures. The two hemi-recti pass to separate ipsilateral anal orifices. (b) Patient 2 had a sequestrated cecum and appendix lying in the midline between two separate hemi-bladders and separate Mullerian structures.

The hemi-bladders were combined to form a single larger volume bladder that received both ureters and that voided through a competent left bladder neck and urethra. The right hemi-vagina with hemi-uterus and fallopian tube was detached from the right vulva and excised. The cecum, appendix and colon were single and normally sited, but the rectum was split longitudinally with each hemi-rectum passing to its ipsilateral anus. The right hemi-rectum was resected leaving the colon to evacuate through the left hemi-rectum that was controlled by a continent intact left anus and anal sphincter complex. The abnormal right anus was left in situ. No pelvic surgery was undertaken and the pubic diastasis was left open. Healing was relatively uneventful and the child was followed up as an outpatient in another institution where the residual right vulva, perineum and split anus were eventually resected. She developed normal fecal and urinary continence through the left structures. Serial radiographic and ultrasound monitoring outlined a stable large capacity bladder with no upper tract changes. Her renal function was normal and there were no urinary infections.

She returned to our institution at 25 years of age, for evaluation toward possible sexual relationships and esthetic adjustments. The pelvic diastasis was still present (Fig. 2) and had led to difficulties with appropriate clothing, but her gait was normal. She was fully continent of urine and feces through the left structures, and had gone through normal puberty establishing regular menstrual cycles. There had not been any urinary infections and evaluation revealed a large bladder with normal upper tracts and normally functioning unscarred kidneys. Examination under anesthesia revealed a smaller vulva to the left side of a wide fatty midline. The left labium major and labium minor were normally formed but were separated from the contralateral matching right structures by a wide fatty midline. There was a left hemi-clitoris with prepuce, and a full length left vagina leading up to a left cervix. The large capacity bladder without any trabeculation was accessible through a longer angled urethra. A single ureteric orifice with hemi-trigone opened separately to either side of the previous bladder suture line. The esthetic appearance of the genitalia and perineum was poor (Fig. 3) and she was very self-conscious with strong psychological concerns relating to her lower body appearance. There had not been any sexual relationships. She was otherwise a well-adjusted, extrovert, intelligent and educated young lady.

1.2. Patient 2

The second patient presented as a healthy 10 year-old 46XX female with an unremarkable antenatal history and scars suggestive of surgery for a large omphalocele. The anus was normal and she was fully continent for feces, however she was continually wet because of urinary incontinence. Her serum electrolytes, renal function, and lumbosacral spine were normal. She had a severe pubic diastasis beneath which was a large midline mucosal structure that protruded through the longitudinally split vulva anterior to the intact fourchette, perineum and anus (Fig. 1b). Abdominal ultrasound scan and a barium enema revealed a cecum and appendix in the right iliac fossa in continuity with a normal single colon and rectum that did not communicate with the abnormal midline mucosal structure. Abdominal and pelvic magnetic resonance showed a single normal kidney and ureter bilaterally, each draining into a separate ipsilateral hemi-bladder (Fig. 4a and b). There was a well-formed hemi-uterus on each side. At examination

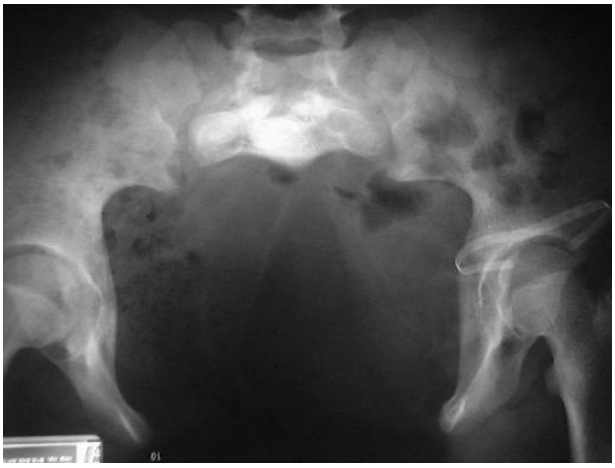

Fig. 2. Patient 1 – preoperative pelvic x-ray showing a severe pubic diastasis. Similar for both patients.

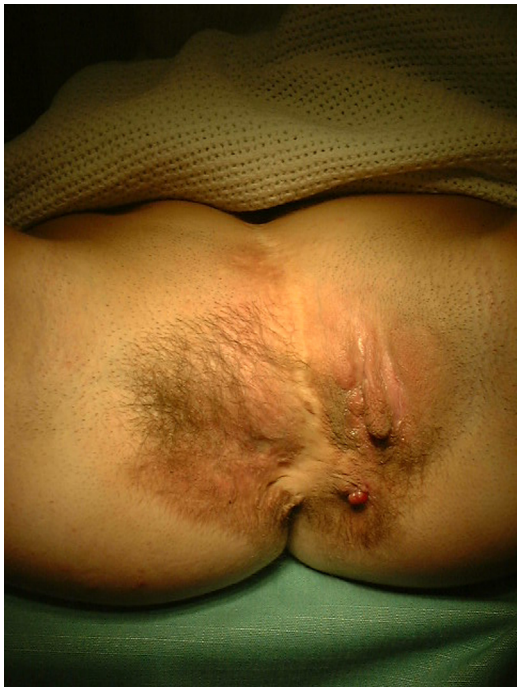

Fig. 3. Patient 1 – lower abdomen and perineum. Very poor esthetic result.