SPLIT-APPENDIX TECHNIQUE: SURGICAL CHOICE FOR COMPLETE INCONTINENCE IN CAUDAL REGRESSION SYNDROME

E. Cerchia, A.L. Bulotta, M. Ruscelli, R. Angotti, G. Di Maggio, M. Messina

The Section of Pediatric Surgery, Department of Medical Sciences, Surgery and Neuroscience, University of Siena, Siena, Italy

Abstract. In the last decades the surgery for total continent reconstruction has been changed by the introduction of intermittent catheterized stoma with the purpose to preserve urinary tract function, urinary continence, elimination of fecal soiling, preservation of quality of life with complete self-sufficiency. We report a rare case of complete incontinence in caudal regression syndrome in whom the appendix was long enough to be divided for creation of both channels for the treatment of urinary and fecal incontinence. A 9-year-old male patient with complete incontinence in caudal regression syndrome was submitted to a Mitrofanoff appendicovesicostomy procedure (AV) in conjunction with appendicocecostomy procedure (ACE). The appendix was divided into two different parts preserving adequate perfusion. Fecal continence was achieved for a period of 18-20 hours after a colonic irrigation with 500 ml of saline solution one time daily, while urinary continence was obtained after suburethral endoscopic injection of dextranomer/hyaluronic acid (Deflux) and intermittent catheterization every 3 hours with an evident reduction of upper urinary tract dilatation. The combination of ACE and Mitrofanoff principle have revolutionized the management of urinary and fecal incontinence in patients who are unable to utilize their urethra to keep themselves dry.

Key words: Caudal regression syndrome, split-appendix technique, appendicovesicostomy, complete incontinence, appendicocecostomy

INTRODUCTION

In the last decades, the management of urinary and fecal incontinence has been revolutionized by the introduction of intermittent catheterized stoma with the purpose to preserve urinary tract function, urinary continence without external presides, elimination of fecal soiling, preservation of quality of life with complete self-sufficiency.

In some patients urethral catheterization is difficult for urethral abnormalities or skeletal deformities. There have been many surgical approaches for urinary reconstruction that usually consist of a catheterizable stoma for emptying a good capacity low-pressure reservoir.

In this kind of patients the Mitrofanoff technique is the best surgical choice creating a continent, catheterizable stoma by bringing the tip of the appendix into the bladder at the end of a submucosal tunnel with the other end hemmed to the skin [1].

Coexistence of fecal and urinary incontinence is common and accounts for a large percentage of children who required more complex diversion, because treatment of 1 system could be limited management of the other. An important surgical development for these complex cases has been the non-refluxing appendicocecostomy, which allows administration of an antegrade continent enema (ACE). In most of cases, the anatomy allows to use the appendix only for a single successful channel. We report a rare case of complete incontinence in caudal regression syndrome in whom the appendix was long enough to be divided for creation of both channels.

CASE REPORT

A 9-year-old male patient was referred for complete incontinence in caudal regression syndrome. His antenatal history was unremarkable and family history revealed a non-insulin diabetic mother. At the birth a loop colostomy was performed in the left upper quadrant for imperforate anus with recto-urethral fistula that was closed at the age of two months and completed by a posterior sagittal anorectoplasty. At the age of four years, he was submitted to surgical removal of bladder stone. We took care of him when he was 9 years old. At physical examination, he showed an hypoplastic and curved penis with anterior hypospadias.

He also had sacral and lower limb anomalies with popliteal webbing associated to equinovarus deformity

Correspondence to:
Dr. Elisa Cerchia
Division of Pediatric Surgery, Dept of Medical Sciences, Surgery and Neuroscience, University of Siena. Polyclinic "Le Scotte"
Viale Bracci 16, 53100 Siena, Italy
Tel.: +39.577.586502; Fax :+39.577.586174
E-mail: elicerc@virgilio.it

of the feet, and no craniofacial or upper limb anomalies were evident. His motor and sensory development was normal. Clinically, he was continually wet and dirty for urinary and fecal incontinence, but he referred sensation of a full bladder and to defecate. Abdominal ultrasound scan revealed a great stone in the bladder and a great bilateral renal pyelectasis. Abddominal and pelvic magnetic resonance showed a complete hipothrophic pelvic floor (Figure 1 a-b), a malrotated incomplete duplex kidney on right side, a bilateral hydroureteronephrosis with focal cortical thinning as the result of recurrent infections, an hyperthrophic bladder detrusor and a voluminous stone in the bladder itself (Figure 1 c-d). Computerized tomography of the pelvis confirmed the presence of bladder stone (5×2.5 cm), the absence of sacral vertebrae from S3 to coccyx, intervertebral osteochondrosis from L4 to L5 with consequent stenosis of the channel at this level and "hourglass" appearance of the pelvic foramen (Figure 2 a-b). Also an upper gastro-intestinal X-ray was performed for documented the intestinal morphology and the presence of the appendix.

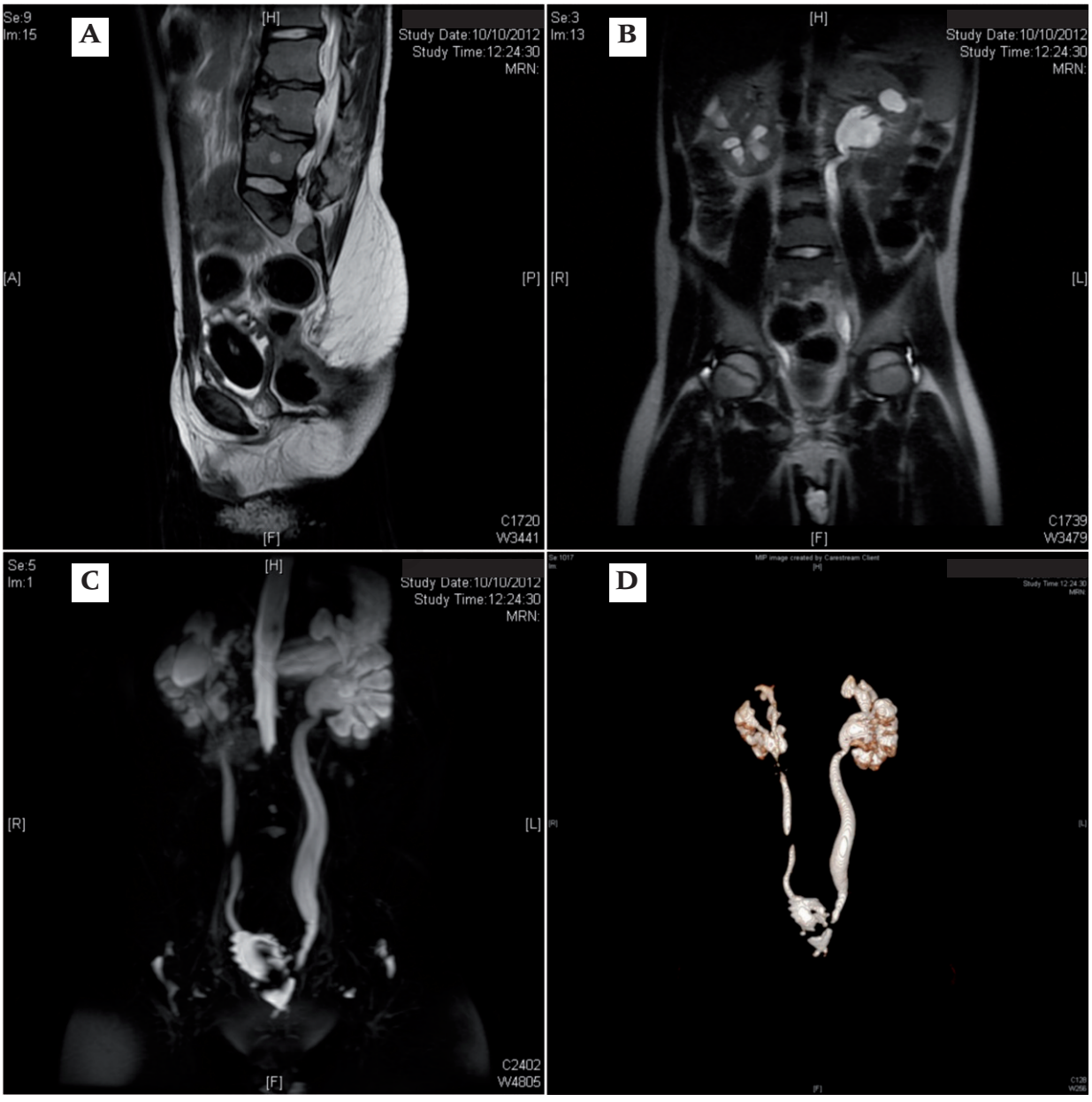

Figure 1 a-b-c-d. Preoperative abdominal-pelvic magnetic resonance showed a complete hipothrophic pelvic floor. Urodynamic and three dimensional magnetic resonance imaging evidenced a malrotated incomplete duplex kidney on right side, a bilateral hydroureteronephrosis with focal cortical thinning as the result of recurrent infections, an hyperthrophic bladder detrusor and a voluminous stone in the bladder itself.

A preoperative cystography showed a bladder of low capacity with irregular wall due to hyperthrophic detrusor and a left vescico-ureteral reflux of III grade.

At examination under general anaesthesia, the perineal electrostimulation showed absence of anal contraction, then it was performed a diagnostic cystoscopy that visualized a residual of the closed recto-urethral fistula, a bladder with trabeculated wall bladder and a great stone at the center, and a normopositioned ureteral meatus.

Surgical technique

The skin incision was made through Mc Burney's point, the cecum was identified and broght into the wound. The appendix was examined carefully to verify a good length and vascularization. An appendiceal length of approximately 8 cm well vascularized was isolated on its mesentery. Another access was via a Pfannestenstiel incision allowing to open the anterior bladder wall and to remove the great stone (4 cm in the major diameter). Then was carefully evaluated the distance between the ileo-cecal junction and the bladder wall and from this last and the abdominal wall, so it was decided to performe a Mitrofanoff appendicovesicostomy procedure (AV) in conjunction with appendicocecostomy procedure (ACE). The appendix was divided into two different parts preserving adequate perfusion (Figure 3).

The distal portion of the appendix (2/3 of the initial length) with its vascular pedicle was directed through an antirefluxing tunnel into the anterior bladder wall. The tunnel was of 3 cm in length (to maintain a 5:1 ratio), and the appendiceal opening was secured with an anchoring resorbable suture to the muscle and mucosa. The abdominal end of the conduit is brought through the abdominal wall, and a stoma was fashioned as rose-bud.

The proximal portion of the appendix (1/3 of the initial length) was kept in continuity with the cecum. A cecal myotomy was created along the anterior tenia for 2 cm and the residual appendix was tunneled with muscular and serosal layer of the cecum to provide a mechanism for fecal continence. An anastomosis of the skin, after a star-shaped incision was performed, to the apex of the spatulated appendix with creation an appendicocecostomy as rose bud (Figure 4). Also was performed the first stage of surgical correction for hypospadic penis.

Postoperatively, a suprapubic catheter was left in situ for six weeks, initially on free drainage and after four weeks alternatively opened every 4-5 hours. A 6 Ch catheter was inserted through each conduit, that it was positioned in appendicecostomy was removed after 3 weeks and then used only for colonic irrigation once daily with 500 ml of saline solution, the other positioned in appendicovesicostomy was removed after five weeks and the patient was trained to perform intermittent catheterization every 3 hours during the day. Fecal continence was achieved for a period of 18-20 hours, while urinary continence was obtained after suburethral endoscopic injection of dextranomer/hyaluronic acid (Deflux) and intermittent catheterization every 3 hours

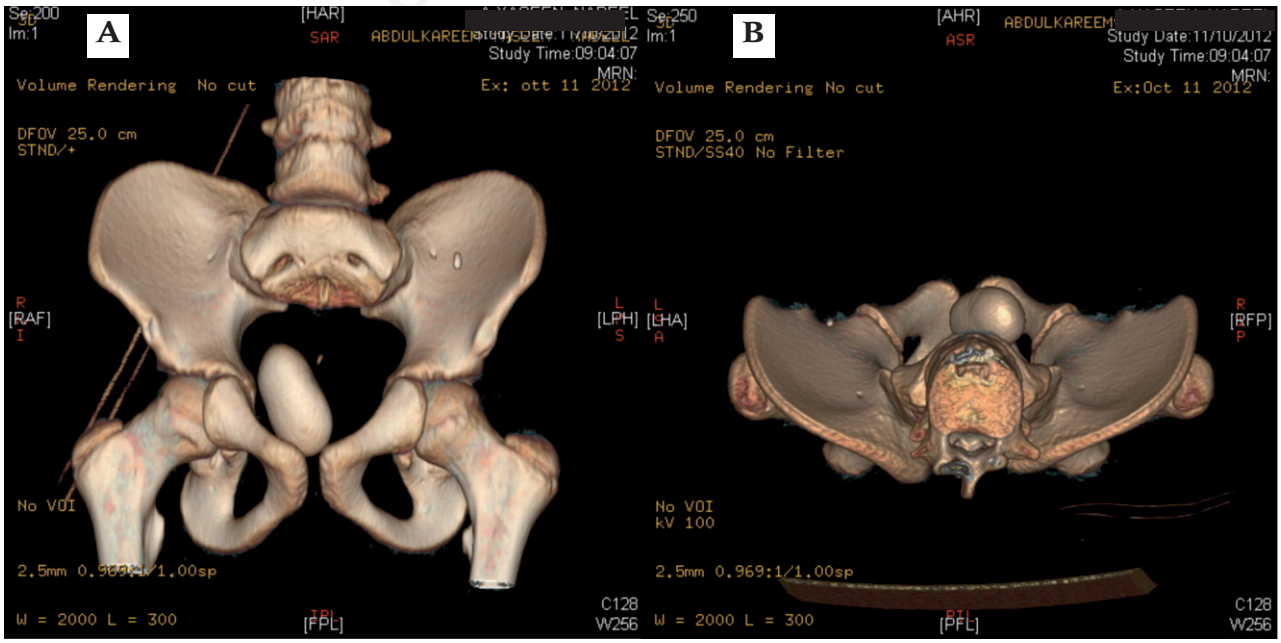

Figure 2 a-b. Three-dimensional computerized tomography of the pelvis showing the presence of bladder stone (5x2.5 cm), the absence of sacral vertebrae from S3 to coccyx, intervertebral osteochondrosis from L4 to L5 and "hourglass" appearance of the pelvic foramen.

with an evident reduction of upper urinary tract dilatation. No stomal stenosis or leakage were present. The patient was discharged from the hospital after 2 months.

DISCUSSION

Caudal regression syndrome is a rare congenital disorder with a reported incidence of 0.01 to 0.05 for 1000 live births, that can affect multiple organs with devastating effects [2-3]. Urologic problems are frequently and can have a great impact on the renal function and quality of life of these patients [4]. Neurogenic bladder is very common in these cases and is consistently present if more than one sacral segment is missing [5]. Also different disorders as anorectal and genital malformations can occur frequently. As a consequence of this functional deficiency of many system, there is often a complete incontinence.

In the last decades total continent reconstruction has been increasingly performed as experience grows with various surgical techniques. Continent catheterizable channels have become an important means to achieve urinary and fecal continence.

The concept of the bladder emptying through clean intermittent catheterization was introduced by Lapides in 1972 [6]. Then in 1980, Mitrofanoff described the transappendicular continent cystostomy as an alternative route for catheterizing the bladder when the urethra could not be used [1], including urethral strictures and injuries, bladder dysfunction associated with an intact urethral sensation (bladder or cloacal exstrophy and epispadias, prune-belly syndrome and idiopathic dysfunctional bladder). The basic principle was the persistence of a positive pressure gradient between the conduit lumen in the antireflux tunnel and the reservoir [7]. This concept was later adopted in the management of bowel incontinence, a procedure known as the antegrade continence enema trough a catheterizable appendicocecostomy and described by Malone in 1990 [8].

The appendix is the preferred conduit for a catheterizable continent stoma, in fact the main advantages of the appendix are good blood supply, satisfactory lumen and auto-lubrication. Several authors have described variations on the classical technique described by Mitrofanoff. Wedderburn [9] and Kajbafzadeh [10] reported the use of the divided appendix in children requiring simultaneous ACE and AV procedures. The split-appendix technique allows the surgeon to create two channels from a single luminal structure. An appendiceal length of approximately 9 to 15 cm was necessary for the surgeon to contemplate the appropriateness of the split-appendix technique. Additional factors involved in the decision-making process were cecal mobility and appendiceal vascular anatomy. Even if the successful results of this procedure, many problems of catheterization are common. Barqawi [11] analyzed the factors associated with problematic conduits in a study of 22 ACE and AV conduits. At 4-year follow up, 5% of their AV required revision. The most common complications were stenosis in 25%, leakage in 8%, and false passage in 2%. Following revision, 97% of their Mitrofanoff channels, and 99% of the ACE conduits, were continent. In another series of 43 patients submitted to split-appendix technique, VanderBrink [12] reported a stomal continence rate of 95% with the surgical revision rate of 19% for stomal stenosis, incontinence, difficulty

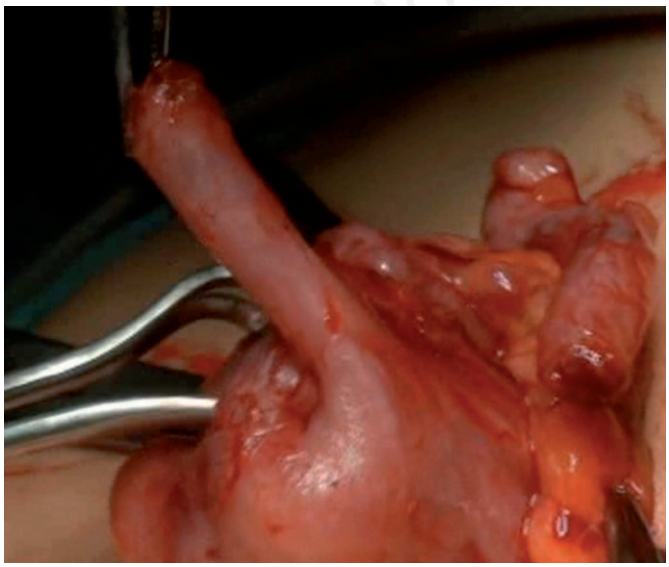

Figure 3. Intraoperative image of the splitting appendix.

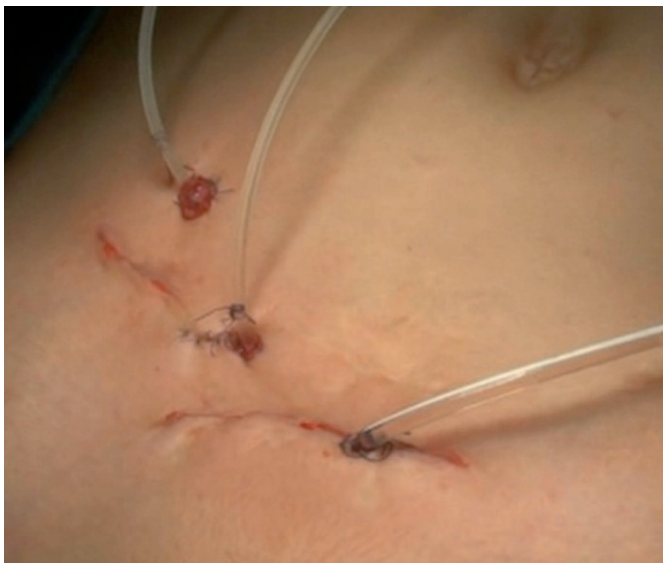

Figure 4. Postoperative result after the anastomosis of the skin to the apex of the splitted appendix with creation of an appendivescicostomy and an appendicocecostomy as rose bud.

Table 1.

Author	Year	N° of patients	Mean length of the appendix	Stomal continence	Revision rate	Follow up (months)
Kajbafzadeh and Chubak	2001	40	10.3 cm	100% AC 100% AV	2.5% AC 5% AV	22
Wedderburn *et al.*	2001	4	Not reported	96% AC	Overall 17%	44
Barqawi *et al.*	2004	22	Not reported	99% AC 97% AV	Overall 15%	48
VanderBrink *et al.*	2010	43	10 cm	100% AC 94% AV	18% AC 18% AV	40

catheterizing and peristomal abscess. Also in 9 of these patient, asymmetrical separation with stapled appendicocecal extension was used, mean length of the appendix for AV and AC was 6 and 4 cm, respectively.

The stomal continence and the surgical revision in our series using a split-appendix technique is similar to other studies. We observed that in literature there is a lack of experience about unequal division of the appendix as a practical option for total continent reconstruction when its length is borderline. In our case there was an appendix of length 8 cm for the creation of two channels and so we preferred an unequal division of itself for preservation of blood supply and performing a tension free anastomosis. Our preference was to use the longer portion of the appendix for creating a good catheterizable appendicovesicostomy.

These changes to the basic technique allowed to reach successful and reproducible outcomes using an appendix of reduced length compared with other studies (Table1).

In our complex case the presence of sacral hypoplasia with associated malformations had an impact on the outcome of the surgical intervention, in fact our patient has reached complete fecal continence, but he has presented dribbling of urine even if in smaller quantities which will probably require closure of the bladder neck and reconstruction of an augmented bladder of increased capacity in a second surgical stage.

The combination of ACE and Mitrofanoff principle have revolutionized the management of urinary and fecal incontinence in patients who are unable to utilize their urethra to keep themselves dry. Most of the complications are preventable by meticulous technique, and eventually multistaged surgical corrections can lead to a functionally and an aesthetically superior reconstruction with a potentially excellent prognosis.

REFERENCES

1. *Mitrofanoff P. Trans-appendicular continent cystostomy in the management of the neurogenic bladder. Chir Pediatric 1980;21:297-305.*
2. *Andrish J, Kalamchi A, MacEwen G. Sacral agenesis: a clinical evaluation of its management, heredity, and associated anomalies. Clin Orthop 1979;139:52.*
3. *Pang D. Sacral agenesis and caudal spinal cord malformations. Neurosurgery 1993;32:755.*
4. *Wilmshurst JM, Rosemary K, Borzyskowski M. Presentation and outcome of sacral agenesis: 20 years experience. Dev Med Child Neurol 1999;41:806.*
5. *Pang D. Caudal agenesis and associated caudal spinal cord malformations. In: Wilkins RH, Rengachary SS, eds. Neurosurgery. 2nd ed., vol.3. New York: McGraw-Hill; 1995. pp 3527-3552.*
6. *Lapides J, Diokno AC, Silber SJ, Lowe BS. Clean, intermittent self-catheterization in the treatment of urinary tract disease. J Urol 1972;107:458-61.*
7. *Hinman FJr. Functional classification of conduits for continent diversion. J Urol 1990;144:27-30.*
8. *Malone PS, Ransley PG, Kiely EM. Preliminary report: the antegrade continence enema. Lancet 1990;336:1217-8.*
9. *Wedderburn A, Lee RS, Denny A, et al. Synchronous bladder reconstruction and ACE procedure. J Urol 2001;163:2392-3.*
10. *Kajbafzadeh AM, Chubak N. Simultaneous Malone antegrade continent enema and Mitrofanoff principle using the divided appendix: report of a new technique for prevention of stoma complications. J Urol 2001;165:2404-9.*
11. *Barqawi A, De Valdenebro M, Furness III PD, et al. Lessons learned from stomal complications in children with cutaneous catheterizable continent stomas. BJU Int 2004;94:1344-7.*
12. *VanderBrink BA, Cain MP, Kaefer M, et al. Split-appendix technique for simultaneous appendicovescicostomy and appendicocecostomy. J Ped Surg 2011;46:259-62.*