Rehabilitation of Urinary Incontinence Following Delayed Bladder Exstrophy Repair in a Female Child

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ABSTRACT

Bladder exstrophy is relatively rare and is even more rarely referred for rehabilitation. A nine-year-old girl with congenital bladder exstrophy from Samar presented with an abnormally low umbilicus, an abdominal opening below the umbilicus, and an anteriorly located anus. The patient has had no prior medical consult. She underwent primary repair of the exstrophy, anterior innominate bone osteotomy, application of external fixators, and traction. Postoperatively, the patient was referred for multidisciplinary rehabilitation management. The rehabilitation bladder program consisted of bladder training with clean intermittent catheterization, use of voiding diaries, fluid restriction, timed voiding, electrical stimulation, and pelvic floor muscle exercises. Bed sore care, pulmophysiotherapy, and strengthening exercises for lower extremities were also implemented. At present there are no validated guidelines in the management of urinary incontinence among female children with pelvic surgery, specifically for bladder exstrophy. This case report aims to present the rehabilitation management of a female pediatric patient with delayed bladder exstrophy repair, resulting in improved outcome.

Key Words: bladder exstrophy, urinary incontinence, delayed bladder exstrophy repair, rehabilitation

Introduction

Bladder exstrophy is a rare developmental defect that is seen in one per 30,000 live births.[1] Records in the University of the Philippines—Philippine General Hospital (UP-PGH) include only 23 cases from 1993 to 2002.

Bladder exstrophy presents in two forms, namely, classic exstrophy and cloacal exstrophy. Classic exstrophy is characterized by wide pubic separation and an exposed bladder. Cloacal exstrophy has the features of classic exstrophy with the addition of intestinal prolapse.

Considered the most severe bladder anomaly, bladder exstrophy results from the persistence of the cloacal membrane, preventing cephalad migration of mesoderm to the midline during development. When the cloacal membrane degenerates, this leaves behind a midline defect of the urogenital sinus. The anterior wall of the bladder, and overlying muscle and skin are not developed.

Clinically, the patient will present with an open pelvic ring, urine draining from an opening on the abdomen, and a waddling gait. Bladder salvage, the closure of the bladder with or without osteotomies in neonates, has been the management of choice.[2]

When closure is delayed, as in the case of the nine-year-old child with classic bladder exstrophy in this report, there is often a need for approximation of the pubic symphyses via anterior innominate bone osteotomy to facilitate the midline repair. Immediately after bladder operation, rehabilitation is commenced with the goal of achieving urinary continence in addition to other problems secondary to the surgical procedure.

The objective of this case report is to present the postoperative rehabilitation management of urinary incontinence after delayed repair of bladder exstrophy in a child.

Case Report

The patient was born full term to a 29-year-old G4P3 (3003) mother via normal spontaneous vaginal delivery at home assisted by a traditional birth attendant. At birth, she had good cry, good suck and no feto-maternal complications. However, an unusually low umbilicus and a separate abdominal opening below it were noted. No other congenital anomalies were observed. The mother was

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advised to bring the child to a clinic for consultation but she did not comply.

The family noted that the patient grew up apparently well, with normal developmental milestones except for a waddling gait. A liquid discharge was noted to drain from the abdominal opening below the umbilicus when the patient cried, when the patient was in a prone position or when her abdomen was pressed. The patient was unable to retain urine, was wet after less than an hour, and wore diapers when going to school. Family history was unremarkable. The patient was doing well in school with honors as a Grade 3 pupil. She could walk independently around their bungalow house and in the single-storey building at school. Since the patient had no complaints and the parents had financial difficulties, medical consult was postponed until the child was older.

At eight years old, the patient started experiencing intermittent low grade fever and abdominal pain. She was brought to a local physician who then referred the patient to a tertiary government hospital in April 2004 for further management. She was initially seen by the Pediatrics Service. The impression was urinary tract infection. Pelvic radiograph revealed an open pelvis with widened symphysis pubis (15 cm gap), intact cortical outlines, with no lytic or sclerotic changes, and with rounded density at the midline of the symphysis pubis (Figure 1). She was diagnosed with classic bladder exstrophy and was referred to the Pediatric Surgery Service for surgical repair of the exstrophy. Anterior innominate bone osteotomy and application of external fixator on the pelvis were performed with primary bladder repair two months after that consult.

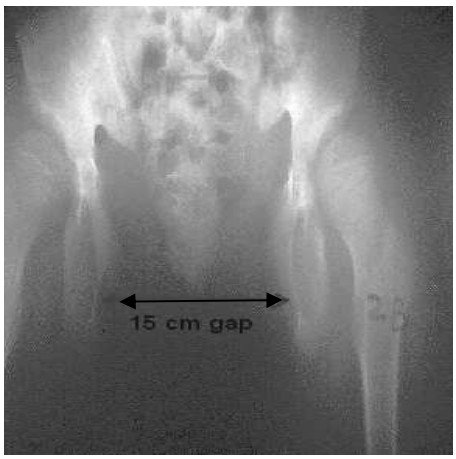

Figure 1. Radiograph (antero-posterior view) of pelvis showing diastasis of symphysis pubis.

The patient was confined at the Surgery Ward for 10 weeks and was managed by Pediatric, Orthopedic, Urology and Rehabilitation Medicine Services. For seven weeks, the patient was bed-bound, kept in a supine position with hips immobilized in an abducted and slightly flexed position by external fixators, and traction on both lower extremities.

A suprapubic cystostomy was inserted with draining catheter via the abdomen and another catheter per urethra to maintain patency while healing. A postoperative cystometrogram to assess bladder function was done by the Urology Service. The cystometrogram showed no extravasation or leakage of dye. After the surgical repair, the patient was observed to have spontaneous voiding but was only partially continent. Oxybutynin HCl 5 mg/tablet 1 tablet two times a day was also given to relieve urinary incontinence.

The patient was referred to the Rehabilitation Medicine Service on the fifth week post-surgery. She was communicative, interactive and cooperative when examined. Both her upper extremities were unremarkable. The lower extremities, however, were atrophied, with both hips immobilized by an external fixator and traction. Bilateral knee joint range of motion (ROM) was full but the knees were in flexion since the patient was immobilized in a supine position with hips in flexion. Ankle dorsiflexors were graded 1/5. Sensory findings were normal. There was a grade 2 decubitus ulcer at the sacrum measuring 2 cm at widest diameter with minimal purulent discharge.

Nerve conduction velocity and electromyography studies showed prolonged left femoral nerve (across the inguinal ligament) compound motor action potential (CMAP) latencies. However, the latencies were within normal limits when taken at the Hunter's canal. The CMAP amplitude of both femoral nerves was also decreased. Bilateral femoral palsy probably secondary to compression was considered.

Initial rehabilitation goals were establishing urinary continence, improving strength, and preventing complications from prolonged immobilization, such as decubitus ulcers, contractures, muscle atrophy, and deconditioning. The following were started while the patient was immobilized at the Surgery Ward: bed sore care; pulmophysiotherapy; range of motion exercises; electrical stimulation of pelvic floor muscles, quadriceps, and tibialis anterior; isometric exercises including abdominal strengthening exercises; and pelvic floor muscle exercises (PFME). The patient developed sacral decubitus ulcer and nosocomial pneumonia for which appropriate treatments were provided.

After seven weeks on pelvic medialization with external fixators, she underwent inguinal ligament release and removal of the external fixator. Both suprapubic cystostomy and foley catheter were removed (Figure 2). The patient was partially continent after catheter removal. Wet periods were noted at intervals of less than 2 hours especially with movement. A bladder training program was initiated. Together with the mother, she was instructed on clean intermittent catheterization (CIC) every four hours, voiding

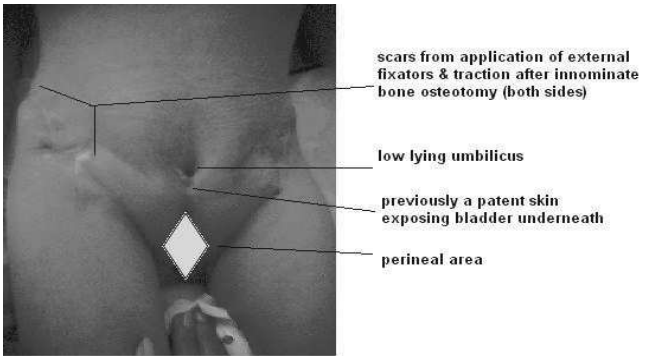

Figure 2. Patient after surgical repair.

diaries, fluid restriction, and continuation of PFME which the patient performed for three weeks. The patient was discharged from the Surgery Ward on the 10th week post-surgery, wheelchair ambulant with weak lower extremities graded 2/5 with partial weight-bearing precautions advised by the Orthopedic Service. She achieved at least two to three hours "dry period," improved from an almost hourly interval of wetness before surgery. She was advised to continue CIC and taper it to six-hour intervals.

The rehabilitation intervention was continued on an out-patient basis with the goal of achieving complete continence, independent ambulation, return to performance of age-appropriate activities of daily living and return to schooling. Timed voiding, voiding diaries, CIC every six hours were continued as a home program. Although her out-patient physical therapy sessions at the tertiary hospital were focused on ambulation and strengthening of the lower extremities, PFME and electrical stimulation were continued. She attended only six out-patient sessions for two weeks since she returned to Samar. Discharge rehabilitation evaluation showed a patient who was partially continent with dry intervals of two to three hours, with some stress incontinence and occasional wetting at night. She became ambulatory without support, with improved lower extremity strength graded 4/5.

Discussion

Various surgical techniques with variable success have been employed to repair bladder exstrophy in the hope of achieving complete dryness, full control over delivery of urine, freedom from catheters and a protected upper urinary tract. The International Continence Society (ICS) with International Consultation on Urological Diseases have published consensus reports on the management of urinary incontinence.[3] The rehabilitation management of urinary incontinence also includes behavioral strategies, pelvic muscle rehabilitation, and patient education with a pharmacologic approach.[4,5,6] This case report highlights the importance of CIC, PFME, and behavioral interventions in the rehabilitation management of bladder incontinence in a child after delayed repair of bladder exstrophy.

The clean intermittent catheterization technique aims to improve bladder capacity and function. It is believed to promote better bladder cycling, maintenance of patent urethra, habituation of parents to performing CIC, and kidney preservation by the prevention of urinary tract infections caused by post-void residual urine.[7,8,9]

Pelvic floor muscle exercise (PFME) has been recognized as beneficial for urinary incontinence.[10,11,12] In this pediatric patient, she was started on PFME immediately after surgery. Instructions for the patient included 10-minute contractions done at two repetitions per session and performed three times a day for three months.[13] Both mother and patient were given instructions to ensure accurate execution of the exercise.

Aside from exercises, the patient was instructed on behavioral interventions such as bladder training, timed voiding, voiding diaries, and fluid limitation. Bladder training enables a patient to accommodate increasingly greater volumes of urine in the bladder and gradually to extend the interval between voiding. Training on timed voiding, use of voiding diaries and fluid limitation increase a patient's awareness of bladder function and strengthens compliance to exercises that promote continence. Behavior therapies were reported to be effective in relieving symptoms and improving psychosocial well-being related to urinary incontinence.[14,15,16,]

Figure 3 is an algorithm for the rehabilitation management of post-bladder exstrophy repair.

This patient showed improvement in continence three months after surgical repair of bladder exstrophy. Immediately after operation, the patient showed spontaneous voiding with partial continence (with dry periods of one to two hours), an improvement from her preoperative incontinent status. Further improvement in continence was observed based on the reduced amount of urine leakage and interval of spillages per day. Three months after surgical repair, the patient was discharged from the outpatient clinic of the tertiary hospital socially continent and was observed to be "dry" for two to three hours with infrequent night time enuresis.

Conclusion

Rehabilitation plays an essential role in the multidisciplinary management of children after delayed repair of bladder exstrophy. Interventions for bladder incontinence in adults such as clean intermittent catheterization, pelvic floor muscle exercises and behavioral interventions can also be instituted in children as presented in this case report. There is evidence to suggest that these programs can improve urinary continence in a child who has undergone delayed surgical repair for bladder exstrophy, although complete continence was yet to be achieved in this

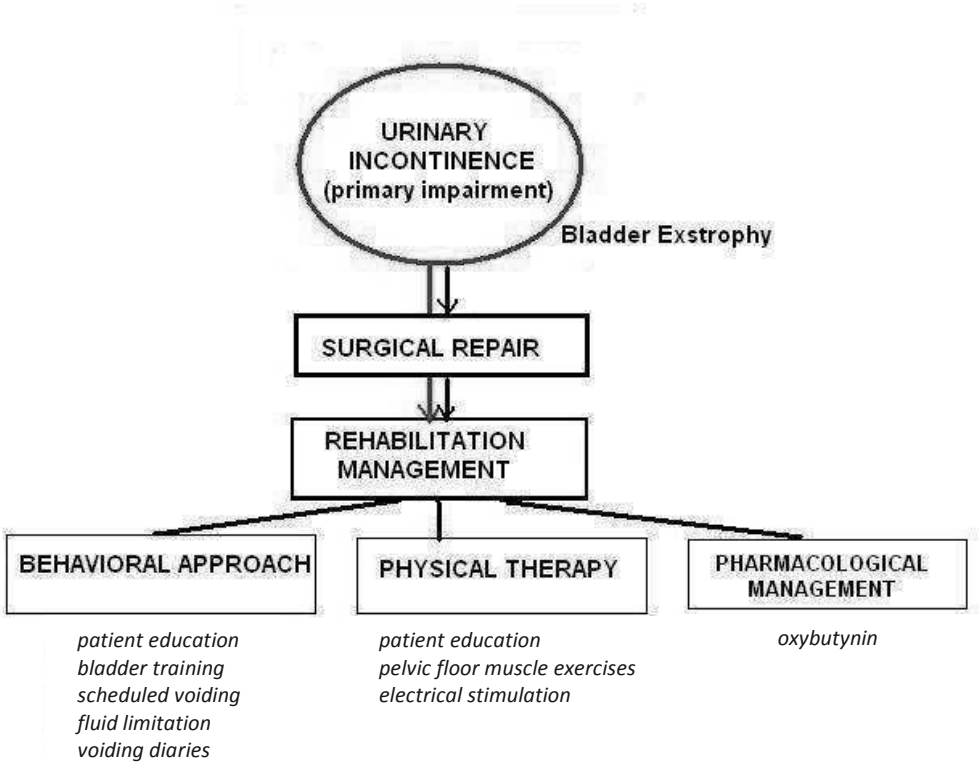

Figure 3. Algorithm for rehabilitation management of urinary incontinence in bladder exstrophy.

case. The assessment of functional outcomes, including long-term social continence through surgical and rehabilitation management, will need well-designed studies involving a large cohort and longer follow-up period. Studies comparing the rehabilitation management of urinary incontinence in early versus late repair of bladder exstrophy are also recommended.

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