Mitrofanoff urinary diversion in a patient with cerebral palsy

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ABSTRACT

Cerebral palsy is the most common motor disability in childhood which result in huge socioeconomic costs. This children have a significant incidence of lower urinary tract symptoms. Clean intermittent self-catheterization is needed to avoid deterioration of renal function. But significant spasticity and resulting contractures of the adductors can interfere with the caretakers’ ability to provide perineal hygiene. Surgery in cerebral palsy affected child is challenging due to multiple associated co-morbidities. The aim of this report is to describe quality of life and renal function for a mentally retarded child with cerebral palsy before and after Mitrofanoff diversion without bladder augmentation.

Key Words: Cerebral palsy; urinary diversion; life quality; morbidity.

Introduction

Developmental disability is a diverse group of chronic conditions that are due to mental or physical impairments and cause individuals living with them many difficulties in certain areas of life, as in the examples in "language, mobility, learning, self-help, and independent living. One of the leading cause being cerebral palsy which result in huge direct and indirect lifetime economic costs. Cerebral palsy describe a group of chronic, non-progressive motor disorders characterized by impaired voluntary movement resulting from prenatal developmental abnormalities or perinatal or postnatal central nervous system damage [1]. Children with cerebral palsy have a significant incidence of lower urinary tract symptoms including urinary incontinence, urgency, frequency, hesitancy, urinary retention and urinary tract infection [2]. Urinary assessment is neglected issue as incontinence is widely attributed to diminish cognition. As a result, urinary tract care is often reactionary rather than proactive. For such patients, continent urinary diversion (CUD) offers a useful alternative for bladder emptying. The
cutaneous appendicovesicostomy firstly described by Mitrofanoff in 1980 [3] remains the most commonly performed CUD. CUDs allow the restoration of urinary continence, preserve the upper urinary tract and improve the patient’s quality of life [4]. Careful assessment of the upper renal tracts and renal function is indicated and such assessment should be ongoing. Spasticity and resulting contractures of the adductors can interfere with the caretakers’ ability to provide perineal hygiene. The aim of this report is to describe quality of life and renal function for a mentally retarded child with cerebral palsy before and after Mitrofanoff diversion without bladder augmentation.

**Case report**

A 9 year child with cerebral palsy with spastic quadriplegia and mental retardation presented to us with history of recurrent urinary tract infection (UTI). He was not able to void and had incontinence of urine. Difficulty for clean intermittent catheterization (CIC) was due to diminished cognition and bilaterally adducted spastic lower limbs. His mother was doing CIC every 3 hourly since patient was of 6 years age, with normal 24 hour urine output. He used to have 3-4 episodes of febrile urinary tract infection per year. Patient was investigated. Urine routine and microscopic examination had plenty of pus cells. Urine culture reports were persistently showing E. coli growth sensitive to higher antibiotics like vancomycin, imipenem. Blood investigations including serum creatinine was normal. Ultrasonography of KUB revealed mild dilatation of pelvicalyceal system on right side with significant post–void residue. On micturating cysto-urethrogram, urinary bladder capacity and contour was normal with grade 2 vesico-ureteric reflux on right side. DMSA scan had bilateral moderate reduced function of cortex with cortical defects at the both poles. On urodynamic evaluation detrusor overactivity was excluded. Antibiotics according to sensitivity report were given for 1 year but in spite of that there were recurrent episodes of UTI. Before surgery we gave parenteral antibiotics according to urine culture and sensitivity till there was no growth on culture. We used mitrofanoff principle and appendico-vesicostomy was done. A lower midline was performed to allow access to the bladder and ileo-cecal junction and appendix. The appendix identified and disconnected from the cecum whilst preserving its mesentery. After passing a 14 Fr catheter down the isolated appendix to check its patency, the end going into the bladder is tunneled submucosally for 3-4 cm into the bladder to achieve an anti-reflux effect. The appendix then secured with absorbable sutures to the bladder muscle and mucosa [Fig. 1A-C].
Fig. 1A-C. Intraoperative photograph. Vertical arrow-bladder, horizontal white arrow-caecum, horizontal black arrow-appendix.

Patient’s per urethral catheter removed on post-operative day 7, drain removed on post-operative day 3 [Fig. 2].

Fig. 2. Patient’s post-operative photograph.

CIC through Mitrofanoff was started after 3 weeks. Then after patient didn’t present with urinary tract infection within 1 year follow up period. Also now it is easy for his primary caretaker to do CIC through an easily accessible stoma. Post-operative ultrasound scan revealed no dilatation of pelvicalyceal system.

Discussion
The prevalence of cerebral palsy is approximately 2-3 per 1000 live births [5]. India however, being a developing country, the expected actual figure may be much higher. Bowel and bladder problems in children with cerebral palsy known as urinary incontinence or enuresis, fecal incontinence or encopresis, and constipation. Additionally, age of achieving bladder and bowel control in the children with cerebral palsy are higher than in their healthy individuals [6,7]. Furthermore, the frequency of urinary tract infections may be higher in these children [8,9]. Hence, it is important to think about the possibility of a bladder problem in any child with cerebral palsy who would be expected to be dry [10]. Vesicoureteral reflux and incomplete bladder emptying resulting from detrusor hyper-reflexia and detrusor sphincter dys-synergia are possible reasons for the propensity to urinary tract infections [2]. In addition, the impaired cognition, mobility and the inability to communicate bladder fullness and the need to void may also explain the tendency to urinary retention and risk of urinary tract infections [2,11]. The prevalence of urinary tract infections in patients with cerebral palsy has been reported as 2.2–32.5% by authors from developed countries [2,12,13].

The management of bladder dysfunction can be complex. Long-term urethral catheterization is associated with problems such as catheter blockage, peri-catheter leak, urethral trauma/strictures, and infection. The trans-appendicular continent cystostomy was first described by Mitrofanoff in 1980, using the appendix as a conduit between the bladder and skin [3]. The indications for this procedure include refractory neurogenic bladder (with or without myelomeningocele), refractory idiopathic bladder dysfunction, as an adjunct to reconstruction in congenital urogenital abnormalities (cloacal extrophy, posterior urethral valves, epispadias and prune belly syndrome).
Pre-operative assessment of co-morbidities, spine evaluation and respiratory evaluation was done in our patient. Cognitive and communication problems in patients with cerebral palsy may make preoperative assessment more difficult. Parents and caretakers are best to be involved during this period not only to gain information but also allay the fears of the child. Children with cerebral palsy are prone to constipation. So pre-operatively soft diet and laxatives were given. Supine positioning of patient and intravenous access during surgery was difficult due to contractures. Also intra-operative hypothermia was a problem due to hypothalamic dysfunction and the malnourished child. Recurrent pneumonias, pulmonary aspiration are aggravated by the inability to cough and poor nutritional state which decreases immune response. Also patient had kypho-scoliosis which may compound the problem with a restrictive lung pattern. So post-operative pain management and chest physiotherapy was discussed with parents beforehand. Thorough pre-operative evaluation with multidisciplinary team is essential in cerebral palsy patient by taking into consideration all risk factors.

Mitrofanoff diversion and CIC can improve quality of life, also reduce socioeconomic burden by reducing number of hospital admissions for such unusual patients.

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References
