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Original Research Article

Neurovesical dysfunction in anorectal malformation

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ABSTRACT

Background: Anorectal malformations are congenital anomalies characterized by the absence of a normally formed anus. Obstructive uropathy, vesicoureteral reflux neurovesical dysfunction are the leading causes of mortality and morbidity in ARM. Early diagnosis and treatment of NVD prevents renal parenchymal damage. Surgical interventions for the correction of anorectal malformation may also lead to the development of NVD. The incidence of urologic and spinal anomalies associated with ARM and the relationship between the anorectal malformations, spinal abnormalities and voiding dysfunction was studied by means of MRI spine and urodynamic studies**.**
Methods: A prospective, observational study was done over a period of 2 years. All patients with ARM underwent MRI of the spine, ultrasound of abdomen, micturating cystourethrogram and urodynamic study.
Results: Forty two patients were included in this study. Out of the 42 patients 17 (40.5%) were female and 25 (59.5%) were male. The ages of the patients ranged from 4 months to 14 years with an average of 32.9 months. 5 patients (12%) had complaints pertaining to neurovesical dysfunction. MRI of the spine revealed abnormalities in 10 patients (23.8%). Urodynamic study was abnormal in 8 patients (19%). Therefore a total of 8 patients (19%) had neurovesical dysfunction in the present study. This included 3 female patients with cloaca and 5 male patients with high anorectal malformations.
Conclusions: Neurovesical dysfunction is frequently seen in patients with anorectal malformations. NVD may be due to the spinal abnormalities or due to iatrogenic injuries to the bladder innervations during surgical reconstruction.

Keywords: Anorectal malformations, Neurovesical dysfunction, Magnetic resonance imaging, Urodynamic study, Voiding dysfunction

INTRODUCTION

Anorectal malformations are congenital anomalies characterized by the absence of a normally formed anus.[1] Cardiovascular, spinal and urogenital abnormalities are the most common abnormalities seen in association with ARM. Obstructive uropathy, VUR and NVD are the leading causes of mortality and morbidity in ARM. Early diagnosis and treatment of NVD prevents renal parenchymal damage.[2] Surgical interventions for the correction of ARM may also lead to the development of NVD.

The objective of current study was to identify patients with voiding dysfunction among the cases of anorectal malformations and to calculate the incidence of voiding dysfunction in ARM; to evaluate those patients with MRI spine and pelvis and urodynamic study; to calculate incidence of spinal abnormalities in patients with anorectal malformations and their correlation with voiding dysfunction; and to manage patients with voiding dysfunction in anorectal malformations.

METHODS

Current study was a prospective, observational study done over a period of 2 years from January 2015 to December 2016.

Inclusion criteria for current study were patients with anorectal malformation who presented to our department and underwent a full workup with an ultrasound abdomen, MCUG, urodynamic study, MRI of the spine. Patients who did not undergo the full workup were excluded from the study.

The previous records of the patients were reviewed and a detailed history was obtained and physical examination was done. Ultrasound of the abdomen, MCUG was done for all patients. All the included patients underwent whole spine axial and sagittal T1 and T2 weighted MRI with a 1.5 tesla surface coil. Ultrasound of the abdomen and micturating cystourethrogram was done for all patients. Urodynamic study (UDS) was done in sitting position using two 5 Fr urethral catheters, a rectal balloon catheter and surface EMG electrodes. Urethral and rectal catheters are used to obtain intravesical and intra-abdominal pressure recordings. Detrusor pressure is calculated by continuous subtraction of the intra-abdominal from the intravesical pressure to correct for changes that might occur during laughing, coughing, movement or talking during the study. Four parameters were used in the evaluation of UDS. Analysis of bladder volume, compliance and detrusor activity allowed assessment of the filling phase, and analysis of vesicosphincteric synergy was used to assess the voiding phase. Bladder filling was performed at a rate of 5% to 10% of the expected bladder capacity per minute with saline at room temperature. Maximum bladder volume was measured when the child started to urinate or showed any signs of discomfort. Compliance was measured by dividing the increment of bladder volume by the concomitant increment of intravesical pressure (V/P) during the filling phase. Detrusor activity was analyzed during the filling phase to detect uninhibited bladder contractions. Management of the anorectal malformations (ARM) and neurovesical dysfunction (NVD) was done as per standard protocols. Statistical analysis were done using SPSS version 16.0.

RESULTS

Forty two patients were included in current study. Out of the 42 patients 17 (40.5%) were female and 25 (59.5%) were male. The ages of the patients ranged from 4 months to 14 years with an average of 32.9 months. Twelve patients (28.5%) had high anorectal malformation, 10 (23.8%) had intermediate and 17 (40.5%) had low anorectal malformations. Three patients (7.2%) had cloacal malformation.

All patients with intermediate and high anorectal malformations and cloaca underwent colostomy in the newborn period. Female patients with anovestibular fistula underwent cutback anoplasty in the newborn period. The definitive procedures done for the patients included posterior sagittal anorectoplasty (PSARP) in 10 patients, Rhode's abdominoperineal pull through in 12 patients, anal transposition in 10 patients, anoplasty in 7 patients and posterior sagittal ano vagino rectoplasty (PSAVURP) in 3 patients. PSARP was done in 7 male patients. Abdominoperineal pull through was done in 11 male patients. Anoplasty was done in 7 male patients. PSARP was done in 3 female patients. Abdominoperineal pullthrough was done in 1 female patient. PSAVURP was done for the 3 female patients with cloaca. Anal transposition was done in 10 female patients.

Out of the 42 patients included in the study, 5 patients (12%) had complaints pertaining to neurovesical dysfunction. Three male and two female patients were symptomatic. The various complaints included nocturnal eneuresis in one patient, dribbling of urine in one patient, frequency of urination in one patient, incontinence of urine and faeces in one patient and incontinence of urine in one patient. Two male patients were evaluated prior to the definitive procedure. One of these cases had an intermediate anomaly and the other had a high anorectal malformation. These patients did not have any voiding problems at the time of evaluation. Both these patients had normal MRI spine and urodynamic studies. Five female patients were evaluated prior to the definitive procedure. Two of these cases had rectovestibular fistula and 3 had anovestibular fistula. These patients did not have any voiding problems at the time of evaluation. One patient with an anovestibular fistula and a solitary right kidney. All 5 female patients who were evaluated preoperatively had normal MRI spine and urodynamic studies.

Ultrasound abdomen was done in all the 42 cases included in this study. Three (7.1%) cases of unilateral renal agenesis were found. 4 cases (9.5%) had unilateral hydroureteronephrosis and one (2.3%) case showed a left sided crossed fused ectopia of kidney. Associated anomalies were seen in 6 cases (14.3%). 2 patients (4.7%) had hypospadias. 4 cases (9.5%) had cardiac anomalies. Ventricular septal defect was seen in one case, patent ductus arteriosus was seen in one case, patent foramen ovale was seen in 2 cases. Micturating cystourethrogram was done in all the cases. Left sided grade 5 vesicoureteral reflux was identified in 2 cases (4.7%) and features suggestive of neurogenic bladder were found in 6 cases (14.3%). Magnetic resonance imaging (MRI) of the spine revealed abnormalities in 10 patients (23.8%). These included sacral agenesis in 5 cases (11.9%), caudal regression syndrome in 3 cases (7.1%) and spinal dysraphism in 2 cases (4.7%) out of which one was associated with meningocele. One case with caudal regression syndrome was asymptomatic. Two sacral pieces were missing in the 2 patients who had sacral agenesis without voiding dysfunction.

Table 1: Details of the patients with neurovesical dysfunction.

Age	Sex	Type of anomaly	Definitive procedure done	Complaints	MCUG	MRI spine	UDS	Treatment
4 y	M	Recto-prostatic urethral fistula	Abdomin-operineal pull through	-	S/o neurogenic bladder	Segmentation anomaly of lumbosacral spine with spinal dysraphism of sacral spine	Severe detrusor overactivity	CIC, anti cho-linergics
11 y	M	Recto-prostatic urethral fistula	Abdomin-operineal pull through	Dribbling of urine, fecal incontinence	No VUR, bladder appears normal	Nonvisuali-zation of S4-S5, moderately filled urinary bladder	Severe detrusor over-activity, incontinence bladder can't be filled completely	CIC, oxy-butinin
3 y	M	Recto-prostatic urethral fistula	PSARP	Incontinence of urine and stool	Oblong bladder, diverticulae left grade 5 VUR	Conus abruptly terminates at lower border of D12 level, s/o caudal regression syndrome	Low pressure detrusor overactivity, compliance and capacity reduced	CIC, oxy-butinin
11 m	M	Recto-prostatic urethral fistula	Abdomin-operineal pull through	Incontinence of urine	S/o neurogenic bladder	Sacral agenesis	Low pressure detrusor overactivity, compliance and capacity reduced	CIC, oxy-butinin
7 m	M	Recto-prostatic urethral fistula	Abdomin-operineal pull through		S/o neurogenic bladder	Sacral agenesis	Low pressure detrusor overactivity, compliance and capacity reduced	CIC, oxy-butinin
12 y	F	Cloaca, long channel	PSAVURP	Nocturnal eneuresis	Neurogenic bladder	Sacral agenesis	Low pressure detrusor activity, reduced capacity	CIC, oxy-butinin
4 y	F	Cloaca, long channel	PSAVURP		No VUR, bladder appears normal	Caudal regression syndrome group, absent S 2-5	Acontractile detrusor, reduced compliance	CIC, oxy-butinin
14 y	F	Cloaca, long channel	PSAVURP	Frequency of urine, fecal incontinence	S/o neurogenic bladder	Sacral agenesis	Low pressure detrusor activity, poor compliance, reduced capacity	CIC, oxy-butinin

y; years, m; months, M; male, F; female.

Urodynamic study was abnormal in 8 patients (19%). The urodynamic studies showed detrusor overactivity in 2 cases, low pressure detrusor activity in 5 cases and an acontractile detrusor in one case. Among those with neurovesical dysfunction, the 5 male patients had high ARM and the 3 female patients had cloaca. All the eight patients with abnormal urodynamic studies were treated

with clean intermittent catheterization (CIC) andanticholinergics. The details of the patients with neurovesical dysfunction are shown in (Table 1).

DISCUSSION

The etiology of urinary incontinence and neurovesical dysfunction (NVD) in ARM patients is multifactorial. NVD involves an impaired innervation to the lower urinary tract, which affects both the filling and emptying functions. During the filling phase detrusor pressure may be increased and the detrusor may be overactive, together with sphincter disturbance and detrusor sphincter dyssynergy (DSD). This may lead to incomplete bladder emptying which, in turn, may cause urinary tract infections (UTI) and renal damage.

A significant proportion of these patients also have associated urologic abnormalities, which include vesicoureteral reflux (VUR), hydronephrosis, or renal agenesis. In anorectal malformations, and especially in high level variants, the late diagnosis and treatment of lower urinary tract dysfunction caused by NVD will lead to the development of permanent upper urinary tract damage. Renal parenchymal injury is more extensive, especially in cases of DSD caused by high intravesical voiding pressures.[3]

Malformations of the lower spinal cord in ARM may lead to NVD and these patients need further urologic workup. The overall incidence of spinal cord abnormalities in patients born with ARM has been reported in different series to be 18% to 50%.[4] The type of lumbosacral pathology had no predictive value with respect to the type of NVD.

De Gennaro et al compared the low incidence of NVD in a group of young children to the older ones and confirmed that spinal dysraphism, although present, may be clinically silent during the first years of life.[5] They suggested that early and repeated urodynamic evaluation is mandatory to detect as earliest as possible the onset of deterioration before irreversible neurological damage has occurred.

Sacral agenesis accompanying imperforate anus in children might indicate the presence of a neurogenic voiding dysfunction recent data have suggested that this dysfunction may exist in the absence of any vertebral anomalies.[6]

Bladder innervations as well as nerve erigenti in cases of high ARM seem to run closer to the midline than normal, becoming more susceptible to surgical damage when violating the basic principle of staying in the midline in PSARP operations. Any injury to those nerves may lead to the development of NVD in the postoperative period. Damage to the external vesical sphincter during ligation of rectourethral fistulas, iatrogenic injury to the sacral nerve fibres during operations in the pelvis minor may play an important role in postoperative urinary incontinence and NVD.

All types of anorectal malformations were included in the present study. There was a wide spectrum of cases and symptoms. Majority of the cases, that is 37 of the 42 included cases (88%), did not have any complaints regarding voiding. 5 patients (12%) had complaints pertaining to neurovesical dysfunction out of which 3 were boys and 2 were girls. The various complaints included nocturnal eneuresis in one patient, dribbling of urine in one patients, frequency of urination in one patient, incontinence of urine and faeces in one patient and incontinence of urine in one patient.

Out of the 37 asymptomatic patients 3 patients had abnormal urodynamic studies. One patient had severe detrusor overactivity. One patient had an acontractile detrusor and reduced compliance. One patient had low pressure detrusor overactivity and reduced compliance and capacity. Therefore a total of 8 patients (19%) had neurovesical dysfunction in the present study. This included 3 female patients with cloaca and 5 male patients with high anorectal malformations. All these patients had sacral agenesis. MRI spine revealed segmentation anomaly of lumbosacral spine with spinal dysraphism of sacral spine in one case, nonvisualization of S4-5 in one case and conus abruptly terminating at the lower border of D12 level which was suggestive of caudal regression syndrome in one case. Caudal regression syndrome with absent S 2-5 vertebrae was seen in one case. Sacral agenesis was seen in 4 other cases.

In study done by Sheldon et al the neurovesical dysfunction was evaluated in patients with anorectal malformations.[7] 90 patients aged 1 to 11 years were evaluated. Patients presented with high imperforate anus, low imperforate anus, anal stenosis, cloacas and cloacal exstrophy.[1-3,8] 10 (11%) patients had clinically proven neurovesical dysfunction based on urodynamic criteria. Two patients had NVD presumed clinically on the basis of urinary retention compounded by urosepsis in the neonatal period. Six patients were documented to have tethered spinal cords and one patient had lumbar stenosis. 16 patients were managed surgically with ureteral reimplantation, vesicostomy, Mitrofanoff procedure.[1-3] They opined that all patients with imperforate anus should have a screening renal ultrasound and a radiographic VCUG. Children who present at an older age with evidence for a sacral mass, significant cutaneous dimpling, a sacral sinus, sacral pigmentation, or sacral hypertrichosis warrant careful neurological evaluation, which in our hands usually includes either MRI or computed tomographic myelography. Similarly, any patient with imperforate anus who presents with urinary retention, urinary incontinence, or deterioration of the upper tracts must be suspected as having NVD and evaluated accordingly.

De Filippo et al studied neurogenic bladder in infants born with anorectal malformation.[4] 26 patients were examined. All these patients underwent PSARP. Urodynamic study, radiologic evaluation of spinal cord with either ultrasound scan or MRI and radiologic imaging of the genitourinary tract with a renal ultrasound and voiding cystourethrogram (VCUG) was done. 21 of the 26 patients had elevated leak point pressures. 6 patients had spinal abnormalities (prominent filum in three, blunted cords in two patients with myelomeningocele, and a low lying cord in one). Ten of the 21 patients had bony abnormalities of the spine or sacrum. Two of the five patients with normal LPPs had spinal cord abnormalities and four had bony abnormalities. This shows that spinal cord and bony spinal column abnormalities are common in patients with and without elevated LPP. The spinal evaluation did not predict the urodynamic result. 12 of the 21 children with elevated LPPs had hydronephrosis or VUR on imaging studies even though six of these patients had normal spinal imaging. VUR was present in 11 of the 26 patients out of which 9 had elevated LPPs. Their results were similar to those found by Kilic et al that PSARP has been found to have little or no effect on bladder dynamics.[3]

De Gennaro et al conducted MRI in 50 patients with anorectal malformations.[5] 25 patients (50%) (1 male and 12 female) had abnormal findings. The abnormalities were not significantly related to the various types of ARM. The pathological MRI findings of the 25 patients with spinal cord anomalies included fibrolipoma with thickened filum in 19 patients, syringomyelia in 4, and tethered cord in 2 and meningocele in 1 patient. 24 patients underwent urodynamic studies out of which 11 had pathological urodynamic findings. 4 infants had detrusor hyperreflexia whereas older children had detrusor hypocontractility.

In 1991 Ralph and others suggested that severe urological problems present in a population of adolescents previously operated by a pull-through procedure were presumably secondary to spinal dysraphism which had led to progressive bladder dysfunction.[8] A neuropathic bladder was found in more than half of the patients in their series and the authors concluded that it is secondary to the spinal abnormalities which were present in 92% of the cases, and not iatrogenic because the lesions were mainly upper motor neuron (UMN) whereas a rectal resection is likely to produce a lower motor neuron (LMN) lesion.

Borg studied the impact of spinal cord malformation on bladder function in children with anorectal malformations.[9] 45 patients with anorectal malformation were studied. 21 were boys out of which 16 (8 prostatic, 5 bulbar, and 3 no fistula) were considered to have normal urodynamics both before and after surgical reconstruction. Seven of these boys were found to have mild detrusor overactivity (4 before and 6 after surgery), which was considered to be of nonneurogenic origin. 5 boys (1 bladder neck, 4 prostatic) were considered to have NVD before and after reconstructive surgery. The urodynamic pattern was characterized by neurogenic detrusor overactivity during filling phase with maximal pressure median of 45 cm H_2O (range, 29-65 cm H_2O). Ten of the 13 girls with vestibular fistulas had normal urodynamics both before and after surgical reconstruction. Three girls had NVD both before and after reconstructive surgery. None of the 3 patients with cloaca had NVD before or after reconstructive surgery.

Thus 25% of the children with anorectal malformation were found to have neurogenic bladder dysfunction. Urinary incontinence is normally accepted in children below 5 years of age. This may lead to delay in the diagnosis of NVD and result in renal damage. The urinary incontinence may also be hidden by concomitant anal incontinence and by the patients wearing diapers.

Parents of newborn babies with ARM are subjected to huge amounts of information. Much of this concerns future surgical procedures and the chance of achieving a normal or acceptable anal function. It is also desirable to inform the parents early about the risk of bladder dysfunction. This information increases awareness of the babies' micturition. It also facilitates parental acceptance of investigations regarding bladder function and treatment for NVD.

NVD generally is not considered to be a consequence of a PSARP, but rather a pre-existing condition. However, during surgery for high cloacal malformations, rectoprostatic fistulae and rectovesical fistulae, the bladder innervation is close to the surgical field and postoperative micturition disturbances may occur. NVD may lead to urinary tract infection (UTI), which may cause impairment in renal function. Loss of voluntary control of micturition frequently leads to urinary incontinence, which is a severe social handicap for the children, who may already suffer from impaired anal continence. CIC is now a standard treatment for NVD. It can prevent UTI and renal damage. It is, either alone or together with anticholinergic medication and surgical procedures, a cornerstone in the treatment of urinary incontinence. An early diagnosis of NVD may result in the early introduction of CIC, which is easier to accept for the patients and reduces the risk of renal damage.

For the early diagnosis of NVD, a 4 hour micturition observation should be done preoperatively in all babies born with ARM during the first months of life. If this is pathological the patient might suffer from NVD and should be investigated further. As NVD is more common in patients with sacral anomalies and in patients with cloacal malformations, a cystometric investigation may be considered in these children. After surgery for high cloacal malformations, rectoprostatic fistulae and rectovesical fistulae, micturition should also be observed.
Once the child is born, a thorough perineal inspection gives the most important clues about the patient's type of

malformation and to some extent even the functional outcome. The newborn boy with a flat perineum, poor developed muscles, hardly visible anal dimple, poor midline, and a palpable defective sacrum will be at high risk of having a spinal cord pathologic condition. The spinal cord malformation is crucial to determining the need for future management of the bladder and bowel. In case of a spinal cord regression with a high location of the conus that ends abruptly, as opposed to a tethered cord, we can tell the parents that the neurologic symptoms will probably not progress. With this information, it is often possible to predict the need for therapeutic interventions to achieve fecal and urinary continence. On the other hand, in case of radiologically detected tethering, there is a risk that the child might develop the tethered cord syndrome. Therefore, there is a need for repeat follow-up of lower limb neurology and urodynamics, for early detection of deterioration of both motor and bladder function.

On the basis of results, author recommends screening for neurovesical dysfunction in patients with ARM. Early detection and institution of treatment for neurovesical dysfunction helps to prevent renal damage.

CONCLUSION

Neurovesical dysfunction is frequently seen in patients with anorectal malformations. NVD may be due to the spinal abnormalities or due to iatrogenic injuries to the bladder innervation during surgical reconstruction. Therefore, screening for neurovesical dysfunction in patients with ARM is very important. All cases of anorectal malformation need to be evaluated for spinal defects. Magnetic resonance imaging of the spinal cord should be done to detect spinal and vertebral anomalies. Urodynamic studies are useful to determine the type of bladder dysfunction as well as detect occult neurovesical dysfunction in patients with anorectal malformations.

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