

Evaluation of dysfunction and malformations of the urinary tract in patients with meningomyelocele, by renal dynamic scintigraphy and direct radionuclide cystography.

An indian perspective

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Abstract

Meningomyelocele (MMC) is the most common form of neural crest closure defect and may be associated with dysfunction and malformations of multiple organs including urinary tract. It affects as many as 2-4 in 1000 live births, however there are no epidemiological studies available from India. Anatomical and neurogenic abnormalities of the urinary tract are known to occur in these patients. Scanty literature is available regarding the utility of radionuclide procedures in the evaluation of urinary tract in patients of MMC. *The aim* of this study was to evaluate urinary tract dysfunction and malformations in patients operated of MMC by diuretic renal dynamic scintigraphy (RDS) and direct radionuclide cystography (DRCG). In this retrospective analysis, 140 operated patients for MMC, mean age: 4.46, range: 1-18 years; 83 male and 57 female, referred to our department for evaluation of the upper renal tract, were studied. Neurogenic bladder (NB) was associated in 69 patients. The diagnosis of NB was based on clinical features like incontinence, expressibility of urine bladder and on urodynamic studies. RDS was performed after the intravenous administration of 10.36 MBq/kg of technetium-99m diethylene triamine penta-acetate or ^{99m}Tc-L, L-ethylenedicysteine. All patients with hydronephrosis (HDN) or hydroureteronephrosis (HDUN) on RDS underwent DRCG using 11.1-18.5 MBq of ^{99m}Tc-sulphur colloid. *Our results* showed that: out of the 140 patients, 78 (56%) including 33 with NB, had normal renal scintigraphy. Six patients (4%) had congenital renal anomalies. Twenty patients (14%) revealed non-obstructive HDN and had no vesico-ureteric reflux (VUR). Thirty-six patients (26%) with bilateral HDUN also had NB of which 19 (53%) had VUR. Fourteen (74%) of these patients with VUR had impaired renal function. *In discussion*, patients with MMC may present with disordered innervation of the detrussor muscle and external sphincter, which adversely affects the bladder function and potentially threatens the upper urinary tract. Patients of MMC with or without NB can be evaluated with RDS for congenital anomalies and initial assessment of renal function. Patients with NB and particularly those with HDUN can be evaluated with DRCG for the presence of VUR to prevent renal scarring and subsequent impairment of renal function. Long-term surveillance of the urinary tract is important in the management of these patients and this can be accomplished with RDS and DRCG. *In conclusion*, in our patients with MMC, the tests of RDS, and DRCG could differentiate those with normal or our abnormal renal anatomy and function while the major group of patients had bilateral HDUN and NB, and/or VUR or impaired renal function.

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Introduction

Meningomyelocele (MMC) is the most common congenital malformation of the central nervous system due to incomplete closure of the vertebral column during embryogenesis, resulting in traumatic exposure or in infections of the meninges and the spinal cord. It affects as many as 2-4 per 1000 live births, however there are no epidemiological studies available from India [1]. The lumbosacral region is involved in 75% of cases and may be associated with dysfunction of multiple organ systems, including the urinary tract. Neurogenic bladder (NB) is a major complication associated with MMC [2]. Detrusor sphincter dyskinesia occurs in almost 50% of these patients. Hydroureteronephrosis (HDUN) is seen in 72% of patients with dyssynergia while 80% of patients with dyssynergia develop vesico-ureteric reflux (VUR). Periodic urine cultures, renal function tests, intravenous pyelogram (IVP), ultrasonography (USG) and micturating cystourethrogram (MCU) are commonly employed to assess the morphological and functional status of the upper urinary

tract. Although the role of radiological procedures in the evaluation of the urinary system in cases of MMC can offer a complete functional diagnosis [3-6], scanty literature is available regarding the utility of radionuclide procedures in the patient of MMC [7, 8].

The purpose of this study was to assess the role of renal dynamic scintigraphy (RDS) and direct radionuclide cystography (DRCG) for the presence of normal or abnormal renal function and renal anatomy findings and also for VUR in MMC patients from India, operated for the treatment of MMC.

Patients and Methods

A total of 140 patients 83 male and 57 female mean age: 4.46 yr., range: 1-18yr operated for the treatment of MMC and referred to our department for the evaluation of the upper renal tract, were retrospectively analyzed. Sixty-nine per cent of these patients had NB. All patients underwent diuretic RDS and DRCG on two separate days within a week. Informed consent was obtained from all the patients.

Diuretic renal dynamic study was performed with the intravenous (iv) administration of technetium-99m diethyl-tri-amine-penta-acetic acid (^{99m}Tc -DTPA) (TCK-7, Board of Radiation and Isotope Technology, Bhabha Atomic Research Center, India) (10.36 MBq/kg) or ^{99m}Tc -ethylenedicycysteine (^{99m}Tc -EC) (TCK-43, Board of Radiation and Isotope Technology, Bhabha Atomic Research Center, India) (3.7 MBq/kg). Minimum dose was 74 MBq and maximum dose was 185 MBq for both radiopharmaceuticals. Furosemide in an iv dose of 0.5-1 mg/kg was given 15 min after the injection of the radiopharmaceutical. Flow images were acquired at 2 sec per frame for the first minute followed by 1-min images acquired over 20 min in a 64X64 acquisition matrix. Data were analyzed by vendor provided 'renal processing software' by marking the whole kidney as a region of interest (ROI) followed by background subtraction using a semi lunar ROI at the lower pole of both kidneys in each study. Differential renal function was estimated using the time activity curve generated by the integral method.

All patients with hydronephrosis (HDN) or HDUN on renal scintigraphy underwent DRCG on a separate day within a week of RDS. In continent and patients older than 5 yr who could void on command, supra-pubic direct radionuclide cystography was performed. ^{99m}Tc -sulphur colloid (TCK-5, Board of Radiation and Isotope Technology, Bhabha Atomic Research Center, India) in a dose of 0.1-0.2 ml (11.1-18.5MBq) was injected into a full urine bladder suprapubically with a small cranial tilt into the bladder by using a 1.5 inch needle (23 gauge) attached to a 2 ml disposable syringe containing the tracer. Before injection it was confirmed that the needle tip was in the bladder by withdrawing a small amount of urine into the syringe. In incontinent patients with NB, the catheter DRCG-method was performed [9] with the tracer injected into the catheter. Pre-void, during micturition and post-void images were acquired in the posterior projection with the kidney and bladder in the field of view.

Both the RDS and DRCG studies were acquired on a single head gamma camera (SPX4, Elscint, Haifa, Israel).

Results

Out of the 140 patients who underwent RDS, 78 (56%) including 33 with NB had normal renal scintigraphy with normal renal morphology, well-preserved function and non-obstructive outflow tract. Six patients (4%) revealed congenital anomalies (2 horse-shoe kidney, 3 ectopic kidney and one crossed fused ectopia). Twenty patients (14%) had non-obstructive HDN (11 unilateral and 9 bilateral HDN) of which two with bilateral HDN had impaired renal function on RDS. Thirty-six patients (26%) with bilateral HDUN were also associated with neurogenic bladder of which 14 (39%) had impaired renal function on RDS. Cases with bilateral VUR and also bilateral HDUN in a patient with NB are shown in Fig 1 and 2 respectively.

Among patients with HDN and HDUN who underwent DRCG, no patient with HDN alone had any VUR, while 19 of 36 patients with HDN (53%) showed evidence of VUR of which, 14 (74%) had impaired renal function. In all, out of the 69 patients with NB, 19 (27.5%) had VUR and 14 (20%) had impaired renal function, Table 1.

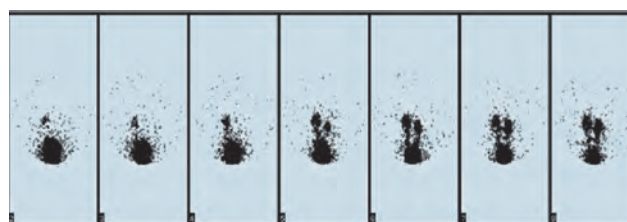


Figure 1. Direct radionuclide cystography revealing bilateral vesico-ureteric reflux.

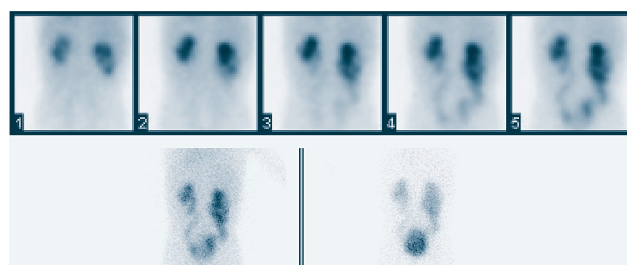


Figure 2. Renal scintigraphy revealing bilateral hydro-ureteronephrosis in patient with neurogenic bladder.

Table 1. Normal and abnormal findings in our patients.

	Normal renal function	Abnormal renal function
NB (n=69)	55 (80%)	14 (20%)
HDN (n=20)	18 (90%)	2 (10%)
NB + HDUN (n=36)	22 (61%)	14 (39%)
NB + HDUN + VUR (n=19)	5 (26%)	14 (74%)

NB: Neurogenic bladder, HDN: Hydronephrosis, HDUN: Hydroureteronephrosis, VUR: Vesico -ureteric reflux

Discussion

Urological problems are the major source of morbidity and mortality in patients with MMC. Congenital renal anomalies such as ectopia, agenesis and duplication are reported with increased frequency [10-12]. In our study we detected by RDS, congenital renal anomalies in 6 patients (4%).

In addition to anatomical abnormalities, neurogenic abnormalities of the urinary tract are known to occur in these patients [13, 14]. Children with detrusor sphincter dyskinesia can be categorized into high and low risk groups for secondary damage from NB based on intravesical pressure. Those with raised intravesical pressure have a high risk for HDUN and reflux [4]. Natural history of these patients at high risk shows that 72% of patients with dyssynergia develop HDUN and 80% develop VUR [3, 15]. Timely intervention with day and night clean intermittent catheterization as well as the administration of anticholinergic drugs, ensure regular and complete emptying of the urine bladder maintaining a low residual volume that prevents urinary tract infections and reflux, leading to pyelonephritis [16].

Sixty-nine patients (49%) in our study had associated NB. Of these patients 33 (48%) had on RDS normal renal morphology and function and 36 had also HDUN with normal/or impaired renal function. All having impaired renal function had also VUR on DRCG. Patients with HDN did not have VUR and seldomly had impaired renal function. In MMC patients, NB and HDUN associated with VUR are more likely to cause impairment of renal function (Table 1).

Although radiological procedures have been routinely used for the evaluation of patients with MMC [3-6], scanty literature is available regarding the utility of radionuclide procedures in patients with MMC [7, 8]. It has been reported that radionuclide hippuran nephrography and renal scintigraphy when compared with iv pyelography (IVP), are suitable investigation techniques for monitoring the kidneys and the upper urinary tract in children with MMC, especially because in these cases, renal function may decrease without clinical symptoms [7]. Others have shown that in neonates with MMC, radionuclide cystography provides comparable information to micturating cystourethrography (MCU) with a much smaller radiation dose [8]. Others have shown that DRCG gives less than 1/20 the radiation dose of the conventional contrast enhanced MCU [17].

Our study demonstrates the role of RDS and DRCG as a screening modality for the evaluation of urological complications associated with MMC in addition to the procedures that may be followed routinely. Patients of MMC with or without NB can be evaluated with RDS for congenital anomalies and initial assessment of renal function. Patients with NB and par-

ticularly those with HDUN can be evaluated with DRCG for the presence of VUR to prevent renal scarring and subsequent impairment of renal function. Long-term follow-up and patients at risk, can also be evaluated with RDS and DRCG. *In conclusion*, in our patients with MMC, the test of RDS, and DRCG could differentiate those with normal or abnormal renal anatomy and/or function, while the major group of our patients had bilateral HDUN and NB, and/or VUR or impaired renal function.

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