

Categorization of Bladder Dynamics and Treatment after Fetal Myelomeningocele Repair: First 50 Cases Prospectively Assessed

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Abbreviations and Acronyms

CIC = clean intermittent catheterization

MMC = myelomeningocele

MOMS = Management of Myelomeningocele Study

UE = urodynamic evaluation

US = urinary tract ultrasound

UTI = urinary tract infection

VCUG = voiding cystourethrography

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Purpose: We categorized bladder patterns and principles of treatment applied to patients who underwent myelomeningocele repair during gestation in a prospective urological assessment.

Materials and Methods: We performed urinary tract ultrasound, voiding cystourethrography and urodynamic evaluation. We then categorized patients into 4 patterns, including normal, high risk (overactive bladder with detrusor leak point pressure greater than 40 cm H₂O and high filling pressure also greater than 40 cm H₂O), incontinent and underactive bladder.

Results: A total of 51 patients were enrolled in study at the first medical appointment. Urodynamic evaluation was done in 48 of these patients as the initial investigation, enabling attribution of a bladder pattern. The high risk pattern was found in 27 patients (56.2%), 18 were incontinent and 1 had an underactive bladder. Only 2 patients (4.2%) in this series had a normal bladder.

Conclusions: Of patients who underwent myelomeningocele closure during gestation 93.7% had significant lower urinary tract dysfunction consisting of high bladder pressure or incontinence. These data reinforce the absolute need to follow these patients closely. The potential benefits of fetal surgery in the urinary tract remain to be proved.

Key Words: urinary bladder, neurogenic; meningomyelocele; fetal therapies; urodynamics; risk

FETAL closure of the MMC abnormality has been gaining popularity after the 2011 publication of MOMS.¹ That randomized trial compared infants operated on prenatally and postnatally. In the fetal surgery group lower rates of ventriculoperitoneal shunt placement and improvement in the composite score for mental development and motor function were confirmed. Groups at several tertiary centers throughout the world started performing MMC

surgery during gestation. Other possible goals and benefits of this procedure are under investigation, including its urological status.

Current urological literature in this regard is sparse and mainly retrospective, and includes only case series. We previously presented our preliminary data on prospective urological assessment at first presentation in 19 cases.²

We have an ongoing protocol of continuous surveillance in this

population and currently more than 50 patients are enrolled in this study. The aim of this series is to categorize bladder patterns and principles of treatment applied to this whole population and review the clinical outcome in patients who underwent at least 2 UEs.

MATERIALS AND METHODS

Since November 2011, all patients with MMC referred to our facility have undergone a protocol consisting of detailed medical history with special attention to the previous febrile UTI history. US is performed to identify hydronephrosis. We use the SFU (Society for Fetal Urology) classification to score hydronephrosis as mild (grade I-II), moderate (grade III) or severe (grade IV). Bladder wall thickness is recorded as normal or as hypertrophic when the thickness of any considerably filled bladder is greater than 3 mm.³ Patients undergo VCUg to rule out vesicoureteral reflux and study bladder morphology. Detrusor-sphincter dyssynergia is defined as persistent proximal urethra dilatation without micturition documented on VCUg that correlates with UE.

Our UE protocol consists of bladder filling with body temperature saline solution at a rate of 5% to 10% of expected bladder capacity per minute through a 4Fr double lumen catheter. This is continued until expected bladder capacity by age is achieved, according to ICCS (International Children's Continence Society) protocol and terminology.^{4,5} Rectal catheters are routinely used as part of the urodynamic assessment and no electromyogram is done. Post-void residual urine volume is considered significant at more than 15% of expected bladder capacity for age. We estimate bladder capacity according to the Holmdahl formula, bladder capacity in ml = 38×2.5 age in months.⁶

We perform US, VCUg and UE, and categorize patients into 4 patterns, including normal—stable bladder cystometry without leakage, high risk—overactive bladder with detrusor leak point pressure higher than 40 cm H₂O and high filling pressures also above 40 cm H₂O, incontinent—overactive bladder with detrusor leak point pressure lower than 40cm H₂O or stable but leaking below 40 cm H₂O and underactivity—underactive bladder with post-void residual urine. Clinical history, physical examination, analysis of radiological examinations and UE are performed by a single urologist (AM). According to these findings we recommended 1 of 3 options as the initial approach, including 1) anticholinergics (0.2 mg/kg) 2 or 3 times daily in association with CIC, 2) clinical surveillance only or 2) CIC only. A urotherapist nurse makes a 50-minute appointment to teach parents how to catheterize the bladder of the child.

All patients in the high risk subgroup are treated with anticholinergics and CIC, those with under contractility undergo CIC only and those who are incontinent and normal remain under surveillance. Treatment can change according to new findings on subsequent clinical and radiological evaluations.

Parents are instructed that urine examinations and cultures are needed only in case of febrile events and to

check with us about the need to treat the event as a UTI. US is repeated at 6-month intervals, and VCUg and UE are repeated every 12 months unless the clinical presentation is not favorable (recurrent UTIs). We define UTI as a febrile event with abnormal urinalysis and positive cultures yielding more than 100,000 colonies per ml. Surgery is never considered an option unless initial treatment fails and the patient presents with a breakthrough febrile UTI.

RESULTS

Currently 51 patients are enrolled in the study after the first medical appointment and UE was completed in 48, enabling us to attribute a bladder pattern and suggest a corresponding treatment. VCUg and US were available in 42 and 46 patients, respectively. The initial patient was born on May 21, 2011. The 51 patients were referred from certain regions of Brazil, including 32 (62.7%) from the southeast, 4 (7.8%) from the south, 8 (15.7%) from the central west, 1 (2%) from the north and 6 (11.8%) from the northeast, composing a nationwide experience. Mean age at diagnosis was 20.7 weeks (range 16 to 25). Mean age at fetal surgery and at birth was 25.6 (range 23 to 27) and 33 weeks of gestation (range 26 to 38), respectively. The maternal complications were pulmonary edema in 1 mother with lupus, and premature membrane rupture in 8 of 51 (15.7%), uterine scar dehiscence in 7 (13.7%) and uterine wound thinning in 4 (7.8%).

A ventriculoperitoneal shunt was placed in 3 of the 51 cases (5.8%). Orthopedic evaluation was available in 47 patients. Motor status was defined as good in 37 cases (78.7%), regular in 6 (12.8%) and poor in 4 (8.5%).

Mean age at first urological evaluation was 4.8 months. One, 2 and 3 or more UTI episodes were documented in 15 (29.4%), 10 and 5 patients (9.5%), respectively. Seven patients (13.7%) were treated with antibiotics as a protocol in hospital.

US revealed hydronephrosis in 12 cases (26.1%) and a thick walled bladder in 7 (15.2%). VCUg, which was available in 42 patients, showed a trabeculated bladder in 12 (28.6%), a dilated urethra in 14 (33.3%) and vesicoureteral reflux in 11 (26.2%).

UE was done in 48 patients at a mean age of 5.4 months (range 1 to 14). The bladder was overactive in 38 cases (79.2%) and stable in 10 (20.8%). Compliance was normal in 6 patients (12.5%), decreased in 21 (43.7%) and could not be determined in another 21, mostly due to leakage. Detrusor-sphincter dyssynergia was found in 24 patients (50%). Bladder capacity was reduced in 31 patients (64.6%), could not be determined in 5 (10.4%) and was normal in 12 (25%). Urinary loss was seen during cystometry in 36 of 48 cases (75%).

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