INITIAL MANAGEMENT

In a prior article,1 the authors reviewed controversies regarding the urologic care of infants diagnosed with neurogenic bladder, primarily caused by spina bifida. Briefly summarized, initial management options include (1) universal institution of anticholinergics and intermittent catheterization (CIC), (2) observation using periodic radiologic imaging, reserving anticholinergics and CIC for infants developing bladder trabeculation, reflux, and/or hydronephrosis, or (3) urodynamic-based management using preemptive anticholinergics and CIC for infants with adverse findings. Proposed advantages of urodynamic-based management include avoidance of therapy in infants with low intravesical pressures, and prevention of bladder and upper tract damage in those with high pressures. Risk assessment according to bladder pressures is based on the observation by McGuire and colleagues2 that 80% of patients with intravesical pressures greater than 40 cm H2O at urinary leakage (detrusor leak point pressure [DLPP]) had reflux and/or hydronephrosis. Accordingly, infants with DLPP greater than 40 cm H2O, and/or detrusor sphincter dyssynergia believed to be an additional risk factor,3 have been prescribed anticholinergics and CIC prophylactically rather than at observation of secondary radiologic deterioration.

Although McGuire found a DLPP greater than 40 cm H2O predictive for upper tract risk, others subsequently suggested that sustained intravesical pressures as low as 20 cm H2O increase likelihood for bladder damage.4 Furthermore, how many infants with initially low intravesical pressures will subsequently manifest increased pressures with risk for bladder and upper tract deterioration is unclear, although two reports suggest it occurs in up to 40% of patients,5,6 usually within the first 6 months of life. Together, these observations have led to early treatment of more infants.

However, determining DLPP in infants has potential pitfalls, because intravesical pressures vary according to infusion rate7 and catheter size.8 Similarly, ability to diagnose detrusor sphincter dyssynergia with commonly used perineal patch electrodes rather than concentric needles has been questioned.6

CURRENT TEXAS SCOTTISH RITE HOSPITAL PROTOCOL

The authors perform initial evaluation using renal ultrasound and fluoroscopic urodynamic evaluation at approximately 6 weeks of age, when spinal shock from newborn back closure appears resolved. A 5-French transurethral urodynamic catheter and a rectal balloon are used to measure detrusor and intra-abdominal pressures, respectively. Contrast medium is infused through a pump with a filling rate based on 10% of estimated bladder capacity (approximately 5 mL/min). Infants with areflexic detrusor pressures exceeding 40 cm H2O start oxybutynin (0.2 mg/kg twice daily) and CIC (every 3 hours), with repeat
testing within 12 weeks to confirm lower pressures.

The authors do not treat uninhibited contractions (UBC), which can be seen in normal infants, unless baseline pressures before contraction exceed 40 cm H20, nor do they diagnose detrusor sphincter dyssynergia. All others with pressures less than 40 cm H2O are observed without active bladder management.

Urodynamic evaluation is repeated at 12 months and approximately 36 months of age. Renal ultrasonography provides surveillance for hydronephrosis every 6 months. Development of hydronephrosis, or new onset of febrile urinary tracts infections, prompts additional fluoroscopic urodynamic evaluation. Data from this protocol are being collected prospectively to document the number of infants diagnosed with high pressures through initial urodynamics at 6 weeks, and the number of patients initially believed to be at low risk who develop high pressures, reflux, or hydronephrosis over time.

Assuming fewer than 10% of children with congenital neurogenic bladder will develop satisfactory bladder control without need for CIC, all parents are initially counseled and reminded at periodic follow-ups to expect this intervention by the age of toilet training if urodynamic evaluation does not indicate earlier management.

**MANAGEMENT AT TOILET TRAINING AGE**

Classifying the neurogenic bladder into high and low pressures also helps predict continence at toilet training (Fig. 1). Those with baseline pressures greater than 40 cm H2O often become dry with anticholinergics (oxybutynin 0.2 mg/kg three to four times daily) and CIC (every 3 hours) alone. Most patients with high pressures have already been diagnosed and started on this medical regimen as a result of earlier urodynamic study.

Those with persistent incontinence undergo repeat testing to determine adequacy of oxybutynin for reducing pressures to less than 40 cm H2O and abolishing UBC. Persistent high pressure or UBC is treated by increasing oral oxybutynin to tolerance and/or adding intravesical instillation of 5 mg twice to three times daily as needed. In unusual cases with continued UBC, Botox injection is recommended. The indication for augmentation is persistent high pressure or UBC despite these medical treatments.

**Sphincteric Insufficiency**

Children who are incontinent on CIC and/or have a history of stress urinary incontinence and found to have detrusor areflexia and pressures less than 40 cm H2O are diagnosed with sphincteric insufficiency. Treatment requires surgical enhancement of outlet resistance using one of several bladder neck (BN) options: injection, reconstruction, sling, artificial sphincter, or closure.

In a series of patients with mixed causes of incontinence and mean follow-up of 28 months, BN injection using dextranomer/hyaluronic acid polymer resulted in a dry interval of 4 hours in 48% of 27 children with neuropathic bladders, 4 of whom underwent failed treatment with slings. Similarly, polydimethylsiloxane injection ended pad use in 34% of 44 children with neurogenic bladders, 24 of whom underwent prior bladder neck procedures, at median follow-up of 28 months. No difference in outcomes was seen between those who underwent prior interventions and those who did not. Both series used a mean volume per injection of approximately 3.5 mL, found that more than 2 additional injections were unlikely to succeed, and noted that the number of patients considered continent declined during the first 12 months and then seemed to stabilize.

Fig. 1. Classification of the neurogenic bladder: (A) low pressure; (B) high pressure.
BN procedures designed to achieve continence through urethral lengthening for neuropathic outlet incompetency include those described by Kropp and Angwafo and Salle. Each uses the anterior bladder wall to extend the urethra beyond the bladder neck into the bladder, through tubularization versus an onlay-type flap, respectively. Reported outcomes for these reconstructions and various modifications are summarized in Table 1.

Fascial sling for neurogenic incontinence was first reported by McGuire and colleagues, and is currently the most commonly used procedure in the United States for this condition in children. Technical aspects in published series vary; for example, McGuire and colleagues described a pubovaginal fascial sling elevated until the bladder neck was observed cystoscopically to close. Others have placed the sling around the bladder neck/proximal urethra in a U or X configuration or 360° wrap, adjusting tension “loosely,” “snug,” or “tight” without cystoscopy. Increase in DLPP averaged approximately 10 to 15 cm H2O, regardless of technique.

Outcomes are listed in Table 2.

The artificial sphincter compresses the bladder outlet circumferentially. Continence rates greater than 80% are reported for functioning devices, but concerns for mechanical failure and/or erosion have limited the use of the artificial urinary sphincter (AUS) in children. Long-term follow-up indicates most failures occur within the initial 3 to 5 years of placement, with a mean device survival time of 12 years.

BN closure is not generally considered first-line therapy for neurogenic incontinence in the United States, although historically Paul Mitrofanoff developed appendicovesicostomy in the mid-1970s to facilitate CIC after closing the outlet. He reviewed his outcomes in 22 patients with a mean follow-up of 20 years. Of these, 5 (23%) had persistent incontinence requiring reclosure, 4 of whom were said to have developed high detrusor pressures, but no mention was made of urodynamic findings or anticholinergic use in this series. Others reported BN closure, with simultaneous augmentation in most, achieving continence in more than 80% of patients at a mean of 20 months follow-up. Some children had urinary leakage from the stoma, whereas others experienced difficulty with catheterization because of stomal stenosis, a potentially serious complication after closing the bladder outlet.

### Changes in Bladder Pressure After Outlet Procedures

These bladder outlet procedures have been associated with increased detrusor pressures, resulting in trabeculation, vesicoureteral reflux, and/or hydronephrosis. However, risk for adverse changes is unknown and factors, including preoperative urodynamic parameters, that might predict likelihood for their development have not been well defined. Further complicating evaluation is lack of universal criteria to diagnose outlet insufficiency in neurogenic bladders, for pre- and postoperative anticholinergic use and CIC intervals, to define postoperative continence, and to determine need for simultaneous or subsequent augmentation.

#### Table 1

<table>
<thead>
<tr>
<th>Authors</th>
<th>Number of Patient (Male/Female)</th>
<th>Mean Follow-up</th>
<th>Number of “Dry” (%)</th>
<th>New Vesicoureteral Reflux (%)</th>
<th>Augmentation</th>
<th>Total (%)</th>
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<tbody>
<tr>
<td>Nill et al</td>
<td>24 (10/14)</td>
<td>1.5–7 y</td>
<td>20 (83)</td>
<td>10 (42)</td>
<td>“all”</td>
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<td>Belman, Kaplan</td>
<td>18 (10/8)</td>
<td>ns</td>
<td>14 (78)</td>
<td>4 (22)</td>
<td>16</td>
<td>7 (94)</td>
</tr>
<tr>
<td>Mollard et al</td>
<td>16 (0/16)</td>
<td>12–36 mo</td>
<td>13 (81)</td>
<td>ns^</td>
<td>7</td>
<td>7 (44)</td>
</tr>
<tr>
<td>Snodgrass</td>
<td>22 (13/9)</td>
<td>ns</td>
<td>20 (91)</td>
<td>9 (50)^</td>
<td>19</td>
<td>19 (86)</td>
</tr>
<tr>
<td>Salle et al</td>
<td>17 (7/10)</td>
<td>26 mo</td>
<td>12 (70)</td>
<td>2 (12)^</td>
<td>12</td>
<td>13 (76)</td>
</tr>
<tr>
<td>Hayes et al</td>
<td>28 (12/16)</td>
<td>28 mo</td>
<td>18 (64)</td>
<td>ns^</td>
<td>23</td>
<td>23 (82)</td>
</tr>
</tbody>
</table>

Abbreviation: ns, not stated.

^ Reimplant for refluxing or insufficient intraureteric distance for tube in 12 patients.

^ Six underwent simultaneous reimplant.

^ One of ten despite reimplant; one of seven without reimplant.
Accordingly, variation most likely exists in not only surgical procedures but also patient selection for outlet enhancement as well as their postoperative management.

**Lessons from Artificial Urinary Sphincter**

The AUS was introduced in 1972 and used to treat incontinence from various causes. Although continence rates exceeded those achieved by other available means, mechanical failures, erosion, and upper tract changes were noted complications. A report by Roth and colleagues in 1986 highlighted the development of hydronephrosis in a series of children implanted with AUS for neurogenic incontinence, occurring in 11 (25%) of 44 with a functioning device (excluding 1 with female epispadias). In 4 patients, 3 who had prior augmentation, dilation was attributed to urinary retention from noncompliance with CIC, and resolved with resumed catheterization and “neuropharmacology,” although 1 patient was recognized later to have decreased bladder compliance. Another patient with a known “hypertonic bladder” preoperatively awaited augmentation. The remaining 6 were believed to have new onset of decreased bladder compliance based on either cystometry or voiding cystourethrogram showing trabeculation. Of these, 3 had no preoperative urodynamic evaluation and 3 relied on combined valsava voiding and CIC. No mention was made of anticholinergic use or changes in medical regimen in response to hydronephrosis, but augmentation was recommended for each. This report also failed to disclose medical therapy and prior or simultaneous augmentation in the other 33 children without hydronephrosis, and did not report on overall follow-up for their patients.

That same year, another report found new loss of bladder compliance or UBC developed in 6 of 35 children treated with AUS for neurogenic incontinence. However, 3 of these were also noted to have spinal cord tethering with new leg spasticity, 2 of whom had detethering with resolution of bladder changes, whereas the other 3 were treated with a trial of anticholinergics before augmentation. Similar to the report by Roth and colleagues, medical management for the entire group, prior or simultaneous augmentation, and postoperative follow-up were not described.

Light and Prieto evaluated 15 patients with myelodysplastic syndromes who had detrusor hyperreflexia or areflexia with decreased compliance after AUS, selected from a nonspecified population of patients at Baylor, where the device was developed. Anticholinergics improved findings in 11 patients, indicating functional rather than structural bladder changes. Only 2 of these patients with adverse

<table>
<thead>
<tr>
<th>Authors</th>
<th>Number of Patients (Male/Female)</th>
<th>Mean Follow-up (mo)</th>
<th>Number of “Dry” (%)</th>
<th>Augmentation Prior/ Simultaneous</th>
<th>Subsequent</th>
<th>Total (%)</th>
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<tr>
<td>Gormley et al</td>
<td>10 (0/10)</td>
<td>49</td>
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<td>Bauer et al</td>
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<td>12</td>
<td>8 (73)</td>
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<td>4 (36)</td>
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<tr>
<td>Elder</td>
<td>14 (4/10)</td>
<td>12</td>
<td>13 (93)</td>
<td>13</td>
<td>13 (93)</td>
<td></td>
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<tr>
<td>Decter</td>
<td>10 (4/6)</td>
<td>26</td>
<td>5 (50)</td>
<td>6</td>
<td>3 (90)</td>
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<tr>
<td>Walker et al</td>
<td>17 (8/9)</td>
<td>16</td>
<td>16 (94)</td>
<td>11</td>
<td>11 (65)</td>
<td></td>
</tr>
<tr>
<td>Perez et al</td>
<td>36 (13/23)</td>
<td>17</td>
<td>22 (61)</td>
<td>35</td>
<td>35 (90)a</td>
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<tr>
<td>Kurzrock et al</td>
<td>24 (15/9)</td>
<td>9–14</td>
<td>19 (79)</td>
<td>24</td>
<td>24 (100)</td>
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<td>Barthold et al</td>
<td>27 (7/20)</td>
<td>≥12</td>
<td>10 (37)</td>
<td>20</td>
<td>22 (81)</td>
<td></td>
</tr>
<tr>
<td>Austin et al</td>
<td>18 (8/10)</td>
<td>21</td>
<td>14 (78)</td>
<td>6</td>
<td>6 (33)</td>
<td></td>
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<tr>
<td>Bugg, Joseph</td>
<td>15 (1/14)</td>
<td>10–36</td>
<td>9 (60)</td>
<td>15</td>
<td>15 (100)</td>
<td></td>
</tr>
<tr>
<td>Castellan et al</td>
<td>58 (15/43)</td>
<td>50</td>
<td>51 (88)</td>
<td>58</td>
<td>58 (100)</td>
<td></td>
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<tr>
<td>Snodgrass et al</td>
<td>30 (18/12)</td>
<td>22</td>
<td>17 (57)</td>
<td>0</td>
<td>1 (3)</td>
<td></td>
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<tr>
<td>Chrzan et al</td>
<td>89 (46/43)</td>
<td>72</td>
<td>42 (47)</td>
<td>11</td>
<td>20 (22)b</td>
<td></td>
</tr>
</tbody>
</table>

a Series comprised of 39 patients; 36 with neurogenic bladder. Diagnosis unclear in 4 patients without augmentation.
b Simultaneous detrusorrhaphy was present in 59 patients; only 19 patients had sling alone.
pressures that did not respond to anticholinergics underwent augmentation.

These articles raised concern that urinary continence achieved through outlet procedures may be associated with decreased bladder compliance. However, the risk for change cannot be determined from this earlier era in which the AUS was implanted without preoperative urodynamic evaluation, sometimes in already trabeculated bladders, in patients who often had no systematic follow-up while asymptomatic and were often not being treated with anticholinergics or CIC. Furthermore, the question arose as to whether diminished bladder capacity was masked by the incompetent outlet or was caused by a change in bladder dynamics after outlet surgery. For example, McGuire and colleagues noted that urodynamic evaluation with BN occlusion in five patients leaking at 50 mL showed decreased compliance in three and a flat filling curve in the other two. However, despite subsequent statements that preoperative urodynamic evaluation with the BN occluded by a Foley balloon is therefore “mandatory,” no publication seems to report urodynamics with versus without BN occlusion in children experiencing sphincteric insufficiency.

Instead, two centers used preoperative urodynamics with BN occlusion in their decision making before AUS placement and evaluated postoperative results. de Badiola and colleagues found preoperative compliance of less than 2 mL/cm H2O predicted need for postoperative augmentation, performed a mean of 14 months after AUS implantation. From a population of 23 children with neurogenic outlet incompetency, augmentation occurred in 7 (30%), whereas another 5 (22%) had increased postoperative capacity and the other 11 were only described as continent without BN occlusion in children experiencing sphincteric insufficiency.

In contrast, in a report focused on AUS device survival, Levesque and colleagues also performed preoperative urodynamics with BN occlusion. Criteria for augmentation included pressure greater than 20 cm H2O at 50% or less predicted or functional capacity for age, and were met by 15 (37%) of 41 patients, 13 of whom did not have UBC or decreased compliance detected with preoperative urodynamics.

Preoperative UD findings did not reliably correlate with postoperative need for augmentation, which is not surprising given the variations in performing cystometry, which included not only whether the outlet was occluded but also use of slow vs rapid “provocative” filling rates, and even gas-versus-fluid infusions. Additionally, variations in postoperative bladder-emptying routines (valsalva vs CIC), anticholinergics, and UD and radiologic testing further influenced impressions of bladder changes. Another confounding factor was the lack of definition for what constituted need for augmentation.

After reviewing experience reported by de Badiola and colleagues and Levesque and colleagues, Kronner and colleagues declared that determining which patients need augmentation during or after AUS is “challenging,” admitting that decision making in their center was based on surgeon judgment without relying on formal urodynamic criteria. This approach resulted in 35 (44%) of 80 children with neurogenic incontinence undergoing simultaneous augmentation, and subsequent augmentation in another 15 because of decreased compliance and/or upper tract changes, for a total rate of 63%. As in other studies from this era, technical aspects of urodynamics varied among patients, some having gas-versus-liquid cystometry performed with or without BN occlusion. Postoperative intervals for radiologic imaging, and CIC and anticholinergic regimens were not specified. Nevertheless, a trend toward increasing augmentation was seen compared with prior reports.

**SLINGS AND AUGMENTS**

Kryger and colleagues reviewed publications regarding AUS, finding reported overall augmentation rates between 33% and 58%. Slings gained popularity as an alternative to the AUS and bladder neck reconstructions and, although they do not compress the outlet to the same extent as AUS, simultaneous augmentation rates increased.

In a review of published series, Perez and colleagues stated that 70% of patients undergoing slings for neurogenic incontinence were augmented. Their own enterocystoplasty rate was 90%, partly because continence seemed improved, and also because of concern that preoperative urodynamics may not detect adverse bladder characteristics in the presence of outlet incompetency.

This article’s authors attempted urodynamics using a Foley balloon adjacent to a urodynamic catheter and found that the outlet was often poorly occluded. After the death of two patients from bladder rupture after enterocystoplasty performed for continence, and realizing that indications for cystoplasty were not clearly defined, the authors decided to perform slings without augmentation in all patients diagnosed with neurogenic outlet incompetency. Sphincteric insufficiency was defined as DLPP less than 50 cm H2O in patients with areflexia (with and/or without anticholinergics)
and a nontrabeculated bladder, and/or stress incontinence. Postoperative continence was categorized as “dry” when there was no, or rare, pad use; “improved” when 1 to 2 pads per 24 hours were used; or “wet” when more than 2 pads per 24 hours were used. In an initial series of 30 consecutive children, with data prospectively maintained in a database, 83% were dry (57%) or improved (27%), which reflect similar results to those in the literature.

One boy was augmented 15 months later. He originally was dry with CIC and only underwent appendicovesicostomy. Immediately postoperatively he became incontinent, with repeat urodynamics showing decreased capacity and sphincter insufficiency. A sling was performed, but he remained incontinent and capacity continued to decrease despite anticholinergics. MRI showed no apparent cord tethering, and upper tracts remained normal. BN closure with augmentation was performed. Another boy with a failed sling had BN closure and augmentation by another surgeon, whereas a girl complaining of pain in the pubic region 4 years after sling placement underwent augmentation by another surgeon despite being continent with stable findings of a smooth bladder with end filling pressures of 15 cm H2O.

Subsequently, the authors used standardized questionnaires to assess patient-reported continence, anticholinergic use, CIC interval, and health-related quality of life (HRQOL) in two cohorts: one after sling placement plus augment and the other after sling alone. Medication requirements were significantly greater in those without augmentation, and mean CIC interval was 3 hours without augment versus 3.5 hours after augment. However, continence and HRQOL were the same in both groups.

The authors recently reviewed urodynamic results after sling without augmentation, comparing preoperative, initial postoperative (within 12 months after surgery), and most recent studies (at least 18 months postoperatively), with mean follow-up 39 months. Of 26 patients, 73% had DLPP less than 25 cm H2O before sling. Postoperative urodynamics within 12 months showed increased pressures and/or UBC in 8 patients (31%). After adjustments in anticholinergics, 6 of these had decreased end-filling pressures on their latest urodynamic evaluation, whereas the other 2 remained stable. All other patients similarly remained stable or had decreased pressures on their latest urodynamic evaluation after sling placement. No trend toward loss of bladder compliance was seen, and no patient developed trabeculation, hydronephrosis, or reflux. The authors concluded that early adverse changes after surgery should prompt adjustments in medical therapy rather than augmentation.

The authors’ experience agrees with the impression of Light and Prieto that adverse bladder changes occur in a minority of patients early after surgery, and are most often responsive to anticholinergics and CIC, sometimes with overnight catheter drainage. The anticholinergic regimen they use consists of oral oxybutynin, 0.2 mg/kg, three to four times daily (or extended-release oxybutynin twice daily). When used, intravesical oxybutynin, 5 to 10 mg, is instilled twice daily in preteens and teens, respectively. Children with pressures greater than 40 cm H2O may require augmentation, a finding at last urodynamic evaluation in less than 10% of our patients.

**SUMMARY**

Initial care of newborns with spina bifida centers on preventing bladder and upper tract damage from DLPP of greater than 40 cm H2O. The authors recommend using urodynamic-based management to select patients with elevated pressures for anticholinergic therapy and CIC, using diapers and observation with biannual renal sonography for the remainder.

At the age of toilet training, children who have urodynamic evidence of UBC and/or rising pressure during filling are started on anticholinergics and CIC, or have their dosage increased until pressures less than 40 cm H2O and areflexia are achieved. Sphincter incompetency is diagnosed in incontinent children with pressures less than 40 cm H2O.
and areflexia and/or stress incontinence. The authors currently use Leadbetter/Mitchell bladder neck revision and a 360° tight rectus fascial sling with appendicovesicostomy to provide continence and facilitate CIC.

Augmentation is indicated in patients with hydronephrosis and/or reflux and end-filling pressures or DLPP greater than 40 cm H2O despite medical management to the point of patient tolerance. A minority of patients, not yet well-defined, will also need augmentation after bladder outlet surgery for similar postoperative indications.

REFERENCES