

**Abstracts of the  
2023 World Congress on Spina Bifida Research & Care  
Tucson, Arizona | March 22–25, 2023**

**Urology**

## Prevalence of detrusor overactivity in myelomeningocele patients with sleep disordered breathing

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**Background:** The correlation between sleep apnea syndrome, irritative bladder symptoms and detrusor overactivity (DO) have been well known in adults. Recent literature demonstrated high rates of sleep disordered breathing in myelomeningocele children. We performed an exploratory analysis to examine prevalence of DO and sleep apnea in this population.

**Methods:** A retrospective chart review was performed on children with myelomeningocele with a screening polysomnography from 2005 to 2020. Clinical variables such as demographics, Apnea-Hypopnea Index (AHI), urodynamic findings and urologic use of anticholinergics, were recorded.

**Results:** We identified 26 patients; 15 were male. The median age at time of polysomnography was 1.3 years old. Most were white and non-Hispanic. Average AHI was 5.34. 6 had normal polysomnographies. Of these, 2 (33%) had a urodynamic diagnosis of DO. None were placed on anticholinergics. 20 patients with abnormal polysomnographics had average AHI of 6.76. 11 (55%) had DO on urodynamics, and 2 were given anticholinergics. There was no association between diagnoses of sleep disordered breathing and DO (Fishers's exact test,  $p=0.64$ ). 12 patients were diagnosed with obstructive sleep apnea or combined obstructive and central sleep apnea; 8 (67%) had a diagnosis of DO. There were four patients who were treated for their sleep apnea and had corresponding pre-treatment and post-treatment polysomnographies with urodynamic studies. 3 were treated with supplemental oxygen with average AHI improvement of 3.3. These 3 all had pre-treatment detrusor overactivity; 2 had resolved DO on follow-up studies while 1 required bilateral ureterostomies for poorly draining upper urinary tracts. One patient had worsening AHI after treatment with tonsillectomy, but their DO resolved post-surgery.

**Conclusions:** While our initial analysis did not show association between treatment of sleep disordered breathing and DO in this population, we believe there may still be a potential relationship between these variables. A prospective study protocol including post-treatment polysomnography with urodynamics can help further define this relationship.

## Use of prophylactic intravesical gentamicin instillations in adults and children with neurogenic lower urinary tract dysfunction

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**Background:** Myelomeningocele patients with neurogenic lower urinary tract dysfunction (NLUTD) may experience recurrent urinary tract infections (UTIs) refractory to traditional preventative measures given their complex anatomy, bladder management needs, and comorbidities. Intravesical gentamicin instillation has been described as a prophylactic measure to prevent UTIs in patients with NLUTD who self-catheterize or use indwelling bladder catheterization in small samples. We describe a large cohort of patients utilizing intravesical gentamicin with the hypothesis that it reduces UTI burden among both children and adults with NLUTD.

**Methods:** A retrospective cohort study of all patients using prophylactic gentamicin instillations at an adult and an academically affiliated pediatric hospital was performed from years 2007 to 2022. Participants were evaluated 5 years before gentamicin initiation to present day. UTIs were defined as presence of urologic symptoms and positive urine culture or UTI treatment. The primary outcome was change in UTI frequency after initiating gentamicin.

Kaplan Meier curves were generated to compare time to first UTI since initiating gentamicin stratified by adult and pediatric cohorts.

**Results:** Sixty-five patients with NLUTD receiving care at a pediatric hospital (n=22) and adult hospital (n=43) were reviewed. Average length of time using gentamicin instillations was 36.2 months (38.5), with 41 patients continuing to use the instillations at the time of analysis. Overall, there was a significant reduction in the average number of UTIs per year after initiation of gentamicin (1.24 [SD=1.2] vs. 1.01 [SD=1.8]; p=0.01) The number of UTI-related hospitalizations was unchanged. On survival analysis, there was no significant difference (p=0.64) in time to first UTI after starting gentamicin between pediatric and adult cohorts (3.3 months, 2.6 months respectively).

**Conclusions:** Gentamicin instillations significantly reduced the number of UTIs experienced by patients with NLUTD. Prospective studies in patients with myelomeningocele are needed to further define the ideal administrative protocol and patient characteristics for optimal therapeutic success.

## Time to define constipation for our spina bifida patients

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**Background:** Individuals with spina bifida often have neurogenic bowel, characterized by constipation and/or fecal incontinence. Neurogenic bowel is managed primarily by urology providers in multidisciplinary spina bifida clinics in the United States. Accepted definitions of constipation in the literature are not clinically representative of individuals with neurogenic bowel. The lack of a disease-specific consensus on constipation in this population has inhibited progress towards improved bowel management research. We aimed to generate a consensus

definition of constipation in individuals with neurogenic bowel secondary to spina bifida.

**Methods:** A multi-disciplinary group, comprised of clinicians, researchers, caregivers of individuals with spina bifida, and adults with spina bifida, were tasked with creating a consensus definition for constipation. Published literature and experience from experts and patients within the group were used to create the definition. A consensus was reached.

**Results:** The group determined that constipation was best defined by the presence of positive signs and the lack of negative signs of bowel function. Signs that the bowel is working well (positive signs): no appetite changes, proper weight gain, mushy/creamy stool consistency, no acute changes to stooling pattern. Signs that the bowel may not be working well (negative signs): abdominal distension, irritability, poor appetite/change in appetite, change in stooling pattern, change in stool consistency (pellet looking, or very loose), straining to pass stool, bubbly liquid stool, vomiting, blood in the stool, change in urinary continence (leakage of urine or urinary retention), increased urinary tract infections.

**Conclusions:** The proposed definition, which is a constellation of signs for constipation unique to individuals with neurogenic bowel secondary to spina bifida, should be considered for use in clinical and academic environments.

## Prospective evaluation of a pediatric urodynamics protocol before and after limiting urine cultures

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**Background:** No consensus exists on the utility of urine culture (UCx) during urodynamics (UDS) in children. We aimed to determine the rate of symptomatic UTI after UDS with (1) routine UCx for all patients and (2) after limiting UCx to only a subset of patients who may be at higher risk.

**Methods:** A 2-part prospective study was undertaken at one pediatric hospital, Phase 1 (7/2016-6/2017) with routine UCx during UDS and Phase 2 (7/2019-6/2020) limiting UCx to those (1) <2 years old (2) with immunosuppression (3) with symptomatic UTI

in the previous 30 days or (4) with history of resistant organism in the urine. The primary outcome was symptomatic UTI after UDS, defined as UCx  $\geq 10^4$  CFU/mL and fever  $\geq 38.5^\circ\text{C}$  or *new* urinary symptoms within seven days. Per protocol, positive cultures in asymptomatic non-CIC patients received short empiric treatment.

**Results:** 1,154 UDS were included: 553 in 483 unique patients during Phase 1 and 601 in 533 patients during Phase 2. Age, sex, race, ethnicity, bladder management, and indication for UDS did not differ significantly between Phases. All 553 UDS in Phase 1 had concomitant UCx, compared to 34% (204/601) in Phase 2. The proportion of positive UCx was 39% (218/553) in Phase 1 and 35% (72/204) in Phase 2. The proportion of asymptomatic patients treated empirically was effectively stable at 7% (41/550) and 6% (33/598), reflecting consistency of asymptomatic bacteriuria in this population. Three patients developed symptomatic UTI in each period, resulting in a stable post-UDS UTI rate of 0.5% in both Phase 1 (3/553) and Phase 2 (3/601).

**Conclusions:** As the rate of symptomatic UTI after UDS was  $<1\%$  before and after limiting UCx, we conclude that routine UCx during pediatric UDS can be safely limited and possibly eliminated. We will continuously study our limited-UCx UDS protocol and make iterative changes as indicated.

## Preliminary results of UMPIRE protocol deviations among infants with spina bifida

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**Background:** The UMPIRE Protocol was designed to represent an expert-informed protocol hypothesized to lead to improved urologic outcomes. The quality improvement design allows for protocol deviations based on the assumption that deviation trends will reflect clinical realities not adequately captured by the protocol. Documenting and analyzing

these deviations can inform which aspects of the protocol are working well and which merit revision. We explored the deviations to describe protocol adherence and patterns of deviations in anticipation that this information will lead to future protocol improvements.

**Methods:** We analyzed deviation data accrued between 2/2015 and 11/2020, by clinical domain (e.g., creatinine measurement, urodynamics, ultrasound completion). A committee of UMPIRE clinicians reviewed deviations to assess the clinics' categorization of "clinician-", "patient-", or "system-related" and, by group majority, to either concur or re-classify the deviation. The committee also evaluated the clinical narrative and further categorized deviations as justified, partially justified, or not justified. Descriptive statistics were used to quantify and describe trends.

**Results:** A total of 1,203 deviations were reviewed for 426 patients. The domains with the most common deviations were blood pressure (29%), creatinine (27%), urodynamics (20%), and renal ultrasound (8%). Additional clinically-significant deviations were noted to occur in patients with hostile bladder (1%) or vesicoureteral reflux (3%). The majority of deviations were not clinically justified (50-99% by domain), and were attributable to clinician decision (46-100% by domain).

**Conclusions:** The frequency and categorization of deviations (clinicians, systems, and patients) vary by clinical domain, and the majority of deviations were categorized as not justified. Further examination of these data is ongoing and will inform protocol revisions and improved adherence.

## Early administration of botulinum toxin in children with neurogenic detrusor overactivity: A single center experience in children 5 and under

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**Background:** Botulinum toxin A is a second line therapy for children with neurogenic detrusor overactivity (NDO) as young as age 5. Less is known about the effects of botulinum toxin on NDO and

bladder compliance (BC) in younger children. We hypothesized that children ages 5 and under would have a greater improvement in BC and resolution of NDO compared with older children.

**Methods:** We performed an IRB approved retrospective review of all patients who underwent intravesical botulinum toxin injection at our institution from 2013 until 2020. Data collected included demographics, urodynamics parameters, and clinical evaluations before and after the first injection and after the most recent injection. Primary outcomes were resolution of NDO and improvement in BC based on Detrusor Leak Point Pressure or End Fill Pressure if no leak was recorded. Patients with insufficient data were excluded. A McNamar's test was used to examine BC changes over time. Regression tree analysis was used to examine predictive factors of improvement in compliance.

**Results:** Sixty-one patients were identified (35 males (57%); median age at first injection 12.5 years). Both age groups (5 years and under and greater than 5 years) demonstrated significant improvement in BC after first and most recent injection. Multivariable modeling demonstrated age 5 or less as the strongest predictor for improved BC after most recent botulinum toxin A injection.

**Conclusions:** Early utilization of botulinum toxin A in patients with neurogenic bladder is associated with greater improvement in long term BC compared to older patients. Further studies are needed to verify this finding in a larger cohort. Future studies may identify if there is a difference in progression to bladder augmentation in this cohort compared to older children.

### Validity of repeated urodynamics in infants with spina bifida within 6 months without initiation of anticholinergic therapy

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**Background:** The bladder management of spina bifida patient requires diagnostic testing for risk assessment. Urodynamics are used to assess the bladder characteristics and dynamics to ensure a safe reservoir for storage of urine. Findings on cys-

tometry may lead to changes in therapy when abnormalities are identified, but at times testing is repeated prior to intervention in order to confirm results. We sought to understand the reproducibility of urodynamics in an infant spina bifida population that had repeat testing within 6 months of their initial testing under the same clinical management. We hypothesized that there would be little differences in results despite young age.

**Methods:** A retrospective review was conducted for patients with spina bifida that underwent repeat urodynamic testing in 6 month or less without any change in bladder management from March 2010 to March 2020. Urodynamic variables including baseline pressure, EFP, LPP, compliance were compared between the tests. Uninhibited detrusor contractions (UDC) were quantified and compared between studies using both historical and current International Children's Continence Society definitions. The urodynamic variables were normalized to the percent expected bladder capacity when appropriate.

**Results:** Eighteen patients were identified that had repeated urodynamic testing within 6 months of their initial testing without starting any medications for review. There were no differences in any of the variables between the two urodynamic studies when adjusting for the change in bladder capacity over time. There were no differences in cystometric volumes for 20, 30, or 40 cm H<sub>2</sub>O between the two studies. There were less UDCs present when using the historical definition compared to the current definition within a study, but between studies there was no difference in UDC frequency, amplitude, or volume when they first occurred.

**Conclusions:** The presence of UDCs and the leak point pressures are consistent between repeated urodynamic studies in infants with spina bifida. As a result, there may be a limited role or benefit of repeating urodynamics in these patients when anticholinergics have not been started.

### Use of clean intermittent catheterization in children aged two years and under with myelomeningocele: Findings from the UMPIRE protocol

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**Background:** Clean intermittent catheterization (CIC) is commonly used in bladder management for individuals with myelomeningocele (MMC). Optimal use of CIC in infants and young children has not been established. We examined CIC use in children aged 2 years and under enrolled in the Urologic Management to Preserve Initial Renal Function Protocol for Young Children with Spina Bifida (UMPIRE) protocol.

**Methods:** The UMPIRE protocol requires initiation of CIC among all newborns with MMC soon after birth and weaning if volumes remain low. Clinical indicators for resuming CIC in the protocol are: hydronephrosis, high-risk bladder, grade 5 vesicoureteral reflux, or clinical discretion. We evaluated hydronephrosis, urodynamics, and vesicoureteral reflux, and anticholinergic use at routine time points until 2 years of age (0, 6, 12, 18 and 24 months). We compared clinical and sociodemographic characteristics between the baseline and 2-year visit using Chi-square tests of independence.

**Results:** 232 participants were included and 17% required CIC at 3 months of age. The proportion requiring CIC increased to 26% at 12-15 months and 35% at 2 years of age. CIC use at 2 years was not associated with sex, race/ethnicity, insurance, prenatal closure, level of lesion, shunt, or hydronephrosis, but was associated with presence of vesicoureteral reflux ( $p=0.02$ ) and anticholinergic use ( $p<0.001$ ).

**Conclusions:** Following our strict protocol, less than one-fifth of newborns with MMC required CIC, but that proportion doubled within the first two years of life. Because bladder characteristics can change over time, findings indicate that routine surveillance to monitor clinical changes may help inform re-initiation of CIC.

## Bladder neck reconstruction in neurogenic bladder: With increased continence comes increased responsibility

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**Background:** Neurogenic incontinence with incompetent outlet remains a surgical dilemma—how to achieve success without compromising renal and overall health. We aim to characterize continence and secondary effects after modified Mitchell urethral lengthening/bladder neck reconstruction (MMBNR) with sling in pediatric neurogenic bladder.

**Methods:** A single-institution, retrospective cohort study of patients after primary MMBNR between 2011-2019 was performed. Demographics, urodynamics, operative details, complications, continence, and additional procedures were collected. Descriptive statistics were utilized. Modifications that differ from previous descriptions include: oblique low anterolateral incision on the bladder to identify the trigone and incompetent bladder neck prior to opening the posterior urethra; focal endopelvic fascia incision *after* full thickness posterior plate creation to minimize blunt dissection for sling placement; voiding is not permitted—a continent catheterizable channel is required. Ureteral reimplantation is not routinely performed.

**Results:** Twenty-five patients (13 females) had MMBNR with sling at a median age of 10 years (interquartile range [IQR] 8, 11). Bladder augmentation was performed concurrently in 14/25 (56%) patients. At a median of 5.0 (IQR 3.9-7.5) years follow-up after MMBNR, 9/11 (82%) without bladder augmentation and 13/14 (93%) with bladder augmentation had no leakage per urethra during the day without further continence procedures. Of the 3 patients with persistent incontinence, two achieved continence with bladder Botox injection (overall continence 24/25, 96%). New and recurrent vesicoureteral reflux was noted in 5 and 1 patients, respectively. Two patients, both continent after MMBNR and with lapses of medical compliance, required subsequent bladder augmentation for pressures and 1 other will likely require it. None have required bladder neck closure or revision.

**Conclusions:** MMBNR with sling provides promising continence outcomes per urethra. The promise of being dry however must be balanced with a heightened awareness to medical compliance and potential secondary effects.

### **Feasibility of enhanced recovery after surgery (ERAS) implementation: Pilot phase outcomes of a multicenter, prospective study**

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**Background:** Pediatric Urology Recovery After Surgery Endeavor (PURSUE) is a multicenter prospective study of Enhanced Recovery After Surgery (ERAS<sup>®</sup>) in patients undergoing lower urinary tract reconstruction, common in patients with spina bifida. The study design includes a pilot phase to verify protocol implementation. We hypothesized that ERAS could be effectively implemented at participating centers with  $\geq 70\%$  protocol adherence at  $\geq 75\%$  of centers.

**Methods:** Local context at each center determined ERAS implementation through structured multidisciplinary meetings with key stakeholders, iteration, and overcoming barriers. After enrollment of the first 5 patients, an audit committee reviewed protocol adherence. Pilot outcomes included documenting involvement of core disciplines during startup, enrolling a minimum of 2 patients in first 6 months, completing 90-day follow up for first 5 patients, defining barriers to implementation, and assessing protocol adherence.

**Results:** 30 pediatric urologists (median 4 per center) participated in ERAS implementation across 8 centers, each enrolling 5 patients (total 40 patients).

Implementation meetings included 10 disciplines and 5 specialties. Sites held a median of 7 meetings (range 2-17) with median 5 participants (range 2-35). Median implementation duration (first meeting to first patient enrolled) was 64 days (range 15-306) and pilot duration (first to fifth patient) was 150 days (range 22-952). All 8 sites reviewed compliance and discussed challenges with the audit committee. Median protocol adherence across all sites was 16/20 (80%) process measures (range 15-18), with 7/8 (88%) centers achieving  $\geq 70\%$  protocol adherence.

**Conclusions:** Successful implementation of ERAS across 8 pediatric urology centers was possible through multidisciplinary collaboration, structured approach, adaptation to local context, and strong auditing process.

### **Comparison of surgical complications between incontinent ileovesicostomy and bladder augmentation in spina bifida patients at a single children's hospital**

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**Background:** An ileovesicostomy (IV) is an incontinent urinary diversion that is offered as an alternative to a bladder augmentation (BA) in a subset of neurogenic bladder patients at-risk for renal deterioration, and who are unable to catheterize due to behavioral, mental, physical, or social barriers. Complications following an IV and BA have never been directly compared. In this study, we compared the incidence and rate of surgical complications between IV and BA in our spina bifida (SB) patients. We hypothesize that IV results in fewer surgical complications due to its incontinent mechanism that does not rely on catheterization.

**Methods:** We conducted a retrospective study of our pediatric patients undergoing IV or BA from 2002 to 2021. Inclusion criteria included having a primary diagnosis of SB. Charts were reviewed for complications requiring subsequent surgery related to the original reconstruction. We compared the overall in-

idence and rate of surgical complications using Fisher's exact test and Wilcoxon-test. Statistical significance was set a priori at  $p < 0.05$ .

**Results:** 11 patients with IV and 62 patients with BA met study criteria. There was a difference in the overall incidence (36% vs. 76%,  $p=0.014$ ) and rate of surgical complications per patient year (0.14 vs. 0.40,  $p=0.04$ ), with IV patients requiring less surgical interventions. However, there was no difference in the incidence of small bowel obstruction, stone-related surgery (which includes both bladder and kidney stones), stomal revision, or bladder perforation.

**Conclusions:** IV and BA are two distinct surgical options for managing SB patients with refractory neurogenic bladders. Our study demonstrates that IV patients were less likely to experience surgical complications.

### Continent catheterizable channels: Does “hitching” the bladder reduce complications?

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**Background:** Continent catheterizable channels (CCC) are a mainstay for reconstruction in patients with neurogenic bladders. It has been suggested that stabilization of the bladder to the anterior abdominal wall can reduce complications. At our institution, hitching is only performed if intraoperative anatomy suggests the need for channel stabilization. We reviewed outcomes of CCCs to determine if “hitching” the bladder reduced complications.

**Methods:** A retrospective cohort of patients with CCC created between 2/2005-6/2019 was performed. Patients were excluded if the channel was implanted into augmented bowel or they had < 6 months of follow-up. Patients were divided into those performed with “hitching” and those without. Complications including subfascial revision for difficulty catheterizing, channel incontinence and stomal stenosis were compared.

**Results:** A total of 105 patients had CCC created during the study period with a median follow up of 5.9 (range 0.6 – 14) years. A total of 21/105 (20%) channels were hitched to the abdominal wall. There

were no significant differences in baseline characteristics. The rate of subfascial revision or need for channel replacement due to difficulty with catheterization was 9/105 (8.6%). The rate of revision or replacement was 1/21 (4.8%) in the hitched group versus 8/84 (9.5%) in the non-hitched group ( $p=0.684$ ). The channel incontinence rate was 3/105 (2.9%). The rate of channel incontinence was 0/21 (0%) in the hitched group versus 3/84 (3.6%) in the non-hitched group ( $p=1.0$ ). The stomal stenosis rate was 23/105 (21.9%) with 5/105 (4.8%) going on to a stomal-level surgical revision. The rate of stomal revision for stenosis was 1/21 (4.8%) in the hitched group versus 4/84 (4.8%) in the non-hitched group ( $p=1.0$ ).

**Conclusions:** Based on retrospective review of patient outcomes and surgical technique, routine hitching of the bladder to the abdominal wall with CCC was not associated with different complications or the need for future revision.

### Single-port laparoscopic-assisted mace creation: Technique video

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**Background:** Patients with spina bifida frequently have neurogenic bowel requiring management with a bowel regimen, of which the Malone antegrade continence enema (MACE) is an effective technique that affords patient independence for a thorough regular colon cleanout. Creation of a MACE catheterizable channel has been done with open and laparoscopic techniques, but never before described as a single port technique when including an imbricated tunnel to limit the likelihood of stool incontinence from the channel.

**Methods:** We describe a technique for creating a MACE channel via a single umbilical incision. A 12mm umbilical laparoscopic port is placed, allowing introduction of the 5mm laparoscope. Once the appendix is identified, a 5mm grasper is passed alongside the camera to grasp the tip of the appendix. The appendix is pulled through the port and then the port is removed. The fascial incision can be widened slightly at this point to permit delivering the cecum into the surgical field. The cecum is imbricated over the base of the appendix, creating a tunneled continence mechanism. The cecum is dropped



back into the abdomen, the fascial widening is reapproximated, and the spatulated appendix tip is anastomosed to the umbilicus.

**Results:** This technique is a simple and quick means that permits creation of a MACE channel, including a cecal imbrication for continence, via a single umbilical incision. The single laparoscopic incision in the umbilicus and placement of the channel in the umbilicus allow for a cosmetic result in that the incision and channel are both effectively hidden in the umbilicus.

**Conclusions:** An imbricated MACE channel can be created via a single-port laparoscopic-assisted technique through the umbilicus, providing a functional, continent, and cosmetic catheterizable channel for antegrade enemas in neurogenic bowel patients.

### Does the indication for bladder augmentation in spina bifida patients effect the long-term risk of adverse events?

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**Background:** Bladder augmentation (BA) is a standard in management for children with spina bifida (SB) who develop neurogenic bladder dysfunction refractory to medical management. Two indications for BA include to preserve renal function in children with hostile bladders (high intravesical pressures with poor bladder compliance), and to achieve social continence. No prior study directly compares the long-term risks of adverse events and subsequent surgeries based on the initial indication for BA in patients with SB. We hypothesized that SB patients requiring BA for a hostile bladder have higher risk for adverse events and subsequent surgery.

**Methods:** Following IRB approval, we retrospectively reviewed all BA under 20 years of age at our institution between July 2002, and April 2021. Only patients with a primary diagnosis of SB were included. Primary outcome was rate of adverse events after 30 days postop (using Clavien-Dindo grade) and subsequent surgery per patient year based on the in-

dication for surgery. A patient was assigned to the hostile bladder group if the preoperative urodynamic study demonstrated an elevated detrusor leak point pressure (DLPP) or end filling pressure (EFP) at or greater than 40cm H<sub>2</sub>O.

**Results:** A total of 60 patients met study criteria; 32 patients in Group 1 had a hostile bladder, while 28 patients in Group 2 had a BA solely to improve urinary continence. Median age of surgery was 8.6 years and median follow-up time from surgery was 8.4 years. We found no significant difference in the adverse events per year (0.4 vs. 0.68, p=0.25) or subsequent surgeries per year (0.62 vs. 0.54, p=0.86) between Group 1 and 2.

**Conclusions:** Our experience with BA in SB patients demonstrates that the initial indication for surgery did not impact the rate of long-term adverse events or subsequent surgery in the pediatric population.

### Differences in self-perceived health and quality of life between patients with acquired and congenital neurogenic bladder

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**Background:** We aim to compare decisional regret and quality of life (QoL) in patients with spina bifida (SB) and spinal cord injury (SCI) following urinary diversion

**Methods:** Patients with neurogenic bladder secondary to SB and SCI were identified within a neurogenic bladder cohort at a single institution. Demographic and clinical data were recorded and analyzed. Subjects completed PROMIS-10 Global Health<sup>®</sup> before and after diversion, and following enrollment also filled the Decisional Regret Scale (DRS) and SF-Qualiveen<sup>®</sup> forms. Patients' ambulatory status, continence, bladder management, and original urostomy were recorded. The groups were compared using two-sample t-tests or Wilcoxon's Rank Sum tests for continuous variables and Fisher's exact test for categorical variables. Statistical analysis consisted of linear regression models with an analysis of covariance.

**Results:** Twelve patients in each cohort completed all survey data. Groups were similar with respect to demographic and preoperative functional characteristics. In this small cohort, we did not observe differences between groups regarding DRS scores nor in potential impacts of diversion in PROMIS® sub-components. However, we demonstrate higher baseline pain levels, lower baseline physical and mental health scores, and a lower level of QoL in the acquired group.

**Conclusions:** Patients with acquired spinal injury demonstrate baseline differences in reported pain, physical and mental health, and overall QoL, when compared with those with congenital malformation, which may impact perioperative expectations. In this small cohort, we failed to demonstrate a difference between groups in decisional regret following urinary diversion.

### **Experiences and barriers of home bladder manometry in the pediatric neurogenic bladder population: Findings from stakeholder perspectives**

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**Background:** Patients with neurogenic bladder are at risk of developing renal deterioration secondary to increased intravesical pressures. To date, invasive urodynamics is the gold standard test to properly assess bladder dynamics. Home bladder manometry is a low-cost and simple method to evaluate bladder pressures and volumes during clean intermittent catheterization. Previous literature has shown that home manometry measurements correlate with urodynamic pressures and hydronephrosis on ultrasound; however, no studies have evaluated the challenges and barriers faced by caregivers with the process.

**Methods:** We performed a qualitative study through interviews and focus group discussions with twenty-three stakeholders. Stakeholders included parents/caregivers of pediatric spina bifida neurogenic bladder patients, providers, and nursing staff. Grounded Theory Methods were used to analyze transcripts and identify preliminary concepts that described at-

titudes towards the current home bladder manometry process.

**Results:** 10 (43%) interview participants were parents, 9 (40%) providers, and 4 (17%) nursing staff. Median age was 39 years (range 26-66). The six themes identified during discussions were an evaluative measure, patient-specific characteristics, sources of error, materials/technology, home environment/context, and education/learning experience. All parents expressed understanding and agreement with the purpose and importance of home manometry. Emergent concepts identified as targets for improvement were the need for standardization of teaching processes with focus on hands-on practice and visual demonstration, standardized expectations for frequency of performing home manometry keeping in mind that it takes an extra set of hands to perform, and the need for appropriate materials including extension tubing with compatibility to catheters in current use.

**Conclusions:** Home bladder manometry is a feasible and beneficial way for neurogenic bladder patients to monitor their bladder pressures and volumes at home. To improve the process, there is a need for standardization of instructions and modification of materials to improve patient/family experiences.

### **Characterization of intradetrusor botulinum toxin use in spina bifida patients over time**

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**Background:** Intradetrusor onabotulinum toxin A (Botox) injection has become increasingly common in bladder management of patients with neurogenic bladder. As practitioner's comfort with Botox has increased over time, it is suspected that this treatment maybe offered earlier in the disease course. We aimed to characterize changes in Botox use over time in spina bifida patients at our institution. We hypothesized that the age of patients receiving Botox has decreased in recent years.

**Methods:** We identified all patients with spina bifida who underwent intradetrusor Botox injections between 2013 and 2019. To evaluate a change in

practice over time, clinical characteristics were retrospectively obtained for patients who had first injection from 2013-2015 and from 2018-2019. Descriptive statistics are reported using parametric methods.

**Results:** 23 patients underwent first Botox injection in 2013-2015, and 20 patients underwent first injection in 2018-2019, for 7.6 patients per year in the early cohort and 10 patients per year in the more recent group. Mean age at first Botox was 10 years (y) (range 3.2-17.6) in the early group and 8.8y (range 2.7-17.6y) ( $p=0.403$ ) in the recent group. Mean follow-up from first Botox was 7.2y (range 1.7-8.9y) and 3.3y (range 1.2-4.4y) in the early and recent groups, respectively. In the early group, 9 (39%) patients proceeded to augmentation within a mean time of 1.5y (range 0.2-4.9y). In the recent group, 2 (10%) patients proceeded with augmentation within a mean time of 1.7y (range 1.1-2.2y). 8 (34.7%) patients continued regular Botox in the early group compared to 6 (28.6%) patients in the recent group.

**Conclusions:** In this limited sample, age of Botox patients did not decrease over time as hypothesized. However, in the recent cohort, more patients per year underwent Botox per year and fewer patients proceeded to augment.

### Long-term follow up of myelomeningocele patients with history of intradetrusor botulinum toxin: Where are they now?

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**Background:** Intradetrusor onabotulinum toxin A (Botox) injection has become a common tool in bladder management of children with neurogenic bladder. However, it remains unclear whether repeat Botox injections are a viable option for long-term management or part of a stepwise approach aiming to delay bladder augmentation. We sought to characterize the current bladder management in patients with long-term follow up after Botox injection. We hypothesized that a significant portion of patients will have discontinued Botox and progressed to reconstructive surgery.

**Methods:** Patients with myelomeningocele who underwent Botox between 2013-2015 were retrospectively identified. Demographic information and clinical characteristics were recorded. Date of last follow-up and current bladder management were recorded.

**Results:** 23 patients were identified. Median age at first Botox was 9.6 years (y) (IQR 5.8-14.8). Median follow-up from time of first Botox was 7.4y (IQR 6.7-8.5). 8/23 patients remain on Botox with CIC. 2 of these patients continue Botox under anesthesia, but 5 have transitioned to office Botox in the adult clinic. 15/23 patients were no longer undergoing Botox treatments. 9/15 patients underwent subsequent augment or vesicostomy, and 6/15 patients have been deemed to have "safe" bladders that have not required further treatment. For patients who progressed to reconstructive surgery, 7/9 had elevated bladder pressures on urodynamics. Median time to surgery after first Botox was 0.9y (IQR 0.2-2.7). 5/8 patients still getting Botox also remained on scheduled anticholinergic medications. No patients had evidence of worsening upper tract function on last follow-up.

**Conclusions:** Most patients who have had intradetrusor Botox injections for management of neurogenic bladder associated with myelomeningocele are no longer undergoing Botox at long-term follow up. However, only a portion of these patients required anatomy-altering reconstructive operations. Additionally, in the proportion that have continued Botox injections, transition to injection without anesthesia appears feasible.

### Patient perspectives on clinician communication about sex and sexual function in young men with spina bifida

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**Background:** Young men with spina bifida (SB) express interest in sexual activity, but their experiences discussing sexual health with their physicians is unknown. Disease specific sexual health questionnaires

are also lacking. This study aimed to assess the perspectives of young men with SB on: (1) Physician counseling on sexual health; and (2) Utility of the International Index of Erectile Function (IIEF) questionnaire in young men with SB.

**Methods:** Semi-structured qualitative interviews were conducted with men  $\geq 18$  years old with SB and focused on: 1) Physician-patient interactions about sex; 2) Suggestions for improving these interactions; 3) Attitudes toward the IIEF; and 4) Suggestions for improving the IIEF. Iterative analysis of transcripts was performed by three coders followed by thematic analysis of codes.

**Results:** Twenty patients participated with a median age of 22.5 years (range 18-29). 80% had myelomeningocele. Most reported romantic interest in women (85%), and were not sexually active (65%). Barriers and facilitators of successful physician-patient interactions were identified. Patient barriers included general discomfort with talking about sex. Patient facilitators of effective interactions included patient comfort with their urologist and discussing sex in relation to disability. Suggestions for improving discussions with physicians and the IIEF were provided (Table). Most felt that the IIEF was applicable, yet limited for those who did not define themselves as sexually active.

**Conclusions:** Young men with SB are interested in discussing sexual health with their physicians. Physician level opportunities for improving discussions as well as the IIEF were suggested.

### **National survey among catheter users: Examining catheter usage and coverage by health plan**

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**Background:** Approximately 70% of individuals with a neurogenic bladder utilize urinary catheters on a regular basis. Inconsistencies exist between healthcare insurers and plans regarding coverage of

urinary catheters and their supplies (i.e. gloves, lubricant). The Spina Bifida Association, Christopher & Dana Reeve Foundation, United Spina Association, Paralyzed Veterans of America, and our university partnered to survey a large group of individuals who use urinary catheters regularly to better understand to what extent catheters are being covered by health insurers and to ascertain what users' out-of-pocket costs are for catheters and their supplies. Also, inquiry into the success of using an insurance appeal for coverage was done.

**Methods:** Survey content was generated by members of the above organizations and was programmed into Qualtrics. Branching logic was utilized with the maximum number of questions an individual would be asked at 29 and an expected completion time of 10 minutes. The survey was translated and administered in Spanish and English. The survey was distributed via each non-profit's registered email list and social media. The survey was open from 1/19/21-2/15/21 and only included individuals who used catheters themselves or were the primary caretaker of an adult or child who uses catheters regularly. To assess, the association between insurance groups and out-of-pocket costs we used two-sample t-tests were used to compare means, and Mann-Whitney U tests were used to test the equality of between-group medians. Kruskal Wallance test was used to assess differences between groups of catheter users in data that did not meet normality assumptions. Results were deemed statistically significant when the p-value was  $<0.005$ .

**Results:** 2366 individuals who responded met inclusion criteria. Straight catheters were the most used catheter representing 63% of respondents as users. The average number of catheters used per month for all types of catheters besides Foley and Condom Catheters was 136/month. 36% reported only having a public/government health plan and 25% reported only having an employer-provided commercial plan, the remainder had both. Of those with public/government insurance, 8% reported that their health plan did not cover any costs for their catheters. For those with commercial coverage, 17% reported their plan did not cover any costs. 42% of those with United Health Care (13% of all who were commercially covered) reported their plan did not cover any costs. Out-of-pocket costs varied significantly amongst insurance plans. Mean annual out-of-pocket costs for catheters and supplies were \$1,464 for those who pay anything after their deductible is met. Appeals to

insurance companies for coverage were denied 30% of the time with 61% of these denials stemming from one insurance company.

**Conclusions:** Catheter users are covered by a variety of health plans with the greatest proportion covered by public/government plans which also cover the largest percentage of catheter and supply costs. Large variations in coverage and out-of-pocket expenses exist for users dependent on the health plan.

### Comparison of quality of life and bladder symptoms in adolescents and young adults with spina bifida who catheterize via urethra versus catheterizable channel

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**Background:** Clean intermittent catheterization (CIC) via catheterizable channel is thought to improve quality of life (QOL), compared to CIC via urethra, but data are conflicting. We assessed the association between QOL and CIC route among adolescents and young adults (AYA) with spina bifida (SB), hypothesizing QOL would be better for those who catheterize via channel than urethra.

**Methods:** We performed a retrospective cross-sectional analysis of clinical questionnaires administered to patients  $\geq 12$ -years-old on CIC between June 2019 and March 2020 in a SB clinic. Questionnaires were completed in English or Spanish independently or with caregiver assistance. Medical records were reviewed for demographic and clinical characteristics. The primary exposure was CIC route (urethra or channel). The primary outcome was general QOL, measured by the Patient Reported Outcome Measure Information System Pediatric Global Health 7 (PGH-7). Secondary outcomes were bladder-specific QOL and bladder symptoms, both measured by the Neurogenic Bladder Symptom Score (NBSS), with lower symptom score signifying fewer bladder symptoms. Multivariable linear regression models

assessed associations between CIC route and questionnaire scores.

**Results:** Of 162 patients on CIC, 146 fully completed PGH-7 and NBSS and were included. 73% catheterized via urethra and 37% via channel. Median age was 17.5 years, 58% of patients were female, 80% had myelomeningocele. Urinary incontinence was more common in patients who catheterized via urethra (60%) compared to channel (33%). CIC route was not significantly associated with PGH-7 scores in bivariable and multivariable analyses. Compared to CIC via urethra, CIC via channel was significantly associated with higher bladder-specific QOL and lower bladder symptom scores on adjusted analyses.

**Conclusions:** AYA with SB who catheterized via channel had better bladder-specific QOL and less bladder symptoms than those who catheterized via urethra. There was no association between CIC route and general QOL.

### Who is managing the bowels? A survey of clinical practice patterns in spina bifida clinics

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**Background:** Neurogenic bowel dysfunction (NBD) affects 80% of individuals with spina bifida. Performing and disseminating research on NBD to reach the appropriate audience is difficult given the variability among medical specialties managing NBD. This study aimed to identify which medical specialties and types of providers are currently managing NBD in the United States.

**Methods:** A survey was developed and sent to 75 spina bifida clinics. Surveys queried which specialty was primarily responsible for medical and surgical management of NBD and any others that assist in NBD care. The license and certification level of the providers were collected. Descriptive statistics were performed to describe the results.

**Results:** The response rate was 68%. Urology was the leading specialty primarily responsible for NBD management (39%) followed by rehabilitation medicine and developmental pediatrics (22% and 20%, respectively). Physicians were the primary providers

of care followed by nurse practitioners (54% vs 31%). Urology performs 65% of NBD surgeries.

**Conclusions:** Multiple specialties and providers are involved in NBD management with variation among clinics. The development of improved NBD care should include a spectrum of specialties and providers. Dissemination of research should be aimed at multiple specialty groups.

### Utilization of a patient portal questionnaire in the pediatric spina bifida population to capture NSBPR data points

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**Background:** The National Spina Bifida Patient Registry (NSBPR) collects data annually for all patients included within the registry. There are many of these data points that can be considered patient reported outcomes. However, often most of the information reported within the registry is through the provider's documentation and does not necessarily include the patient's or family's perspective. The adoption of EHR patient portals allows new modalities to communicate with patients and their families. Understanding acceptance by parents to use these modalities and their willingness to respond to surveys can help providers choose optimal ways in which to deliver a questionnaire.

**Methods:** A patient-entered questionnaire was built within our EHR to capture data points that are part of the annual NSBPR survey. All patients seen within our institution's multidisciplinary spina bifida clinic since 6/1/2020 were reviewed. A patient portal message with the spina bifida questionnaire attached was sent to all patients that had an activated patient portal account and spoke English as a primary language in the week preceding the clinic visit. The questionnaire was built with nested logic to limit unnecessary questions for families. Information was collected regarding whether the message was read and if the questionnaire was completed.

**Results:** There was a total of 353 patients that were seen in the spina bifida clinic during the study period. 304 (85.4%) had an activated patient portal account. 26 of these patients did not receive a survey due to English not being the primary language. A

total of 601 questionnaires were sent prior to the clinic visit. Of these surveys, 455 (75.7%) were read and 288 (47.9%) were completed. 63.2% of families that read the message went on to complete the survey. For the messages that were not read initially, at the follow-up visit 17.4% filled out the questionnaire. For the families that read the initial message only, 39.7% filled out the questionnaire at their next visit. Of those that filled out the questionnaire initially, 69.4% filled out the questionnaire at the next visit.

**Conclusions:** Questionnaires sent through the electronic health record have good utilization in the spina bifida population. There is a proportion of families that did not read the message or fill out the survey initially that subsequently filled out the questionnaire at follow-up visit. This strategy offers the opportunity to gain more information from the patients and families themselves. This may potentially lead to more reliable and accurate data collection for this national registry.

### Comparing binary & ordinal definitions of urinary and stool continence outcomes: Data from the National Spina Bifida Patient Registry

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**Background:** The National Spina Bifida Patient Registry (NSBPR) is a multi-institutional patient registry of individuals living with spina bifida (SB). The annual NSBPR questionnaire utilizes ordinal categories for bladder and bowel continence. Previous studies have used binary classifications to define continence as having an incontinent event “never” or “less than monthly.” Binary continence status may not fully reveal the range of patient experience or capture changes to and from continence after interventions. Our aims were to **1)** compare the use of ordinal continence categories with the binary continence categories; **2)** explore the correlation of continence with undergarment usage; and **3)** assess continence status following continence surgeries.

**Methods:** Univariate logistic regression was used to analyze data from the most recent clinic visit for NSBPR participants using both ordinal categories of incontinence and binary definitions. Continence surgical outcomes were analyzed for those with data at least three months post-operatively. Chi-square tests evaluated associations among categorical variables. Univariate and ordinal logistic regression models were used to test associations of ordinal continence status with patient and condition factors. Statistical tests were 2-sided; p values <0.05 were considered significant.

**Results:** Analysis of 7217 individuals using ordinal outcomes showed little difference from binary definition. Among those reporting never being incontinent of urine or stool, 14% reported the use of protective undergarments. Of 495 individuals who had bladder outlet surgery, 38% reported never being incontinent. Of 1396 individuals who had appendicostomy surgery, 48% reported bowel never being incontinent.

**Conclusions:** Analysis of ordinal categories did not markedly change the proportion of patients reporting bladder and bowel continence, indicating the binary definition of continence can capture changes in continence in the SB population. Undergarment choice is a poor surrogate for reported continence. After bladder and bowel continence surgery, nearly half reported never having an incontinent episode.

## Improvement in urodynamic parameters after secondary spinal cord detethering

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**Background:** Secondary tethering of the spinal cord (sTSC) can occur following primary repair of a spinal dysraphism. One of the earliest harbingers of sTSC is change in bladder dynamics seen on videourodynamic studies (VUDS). We sought to compare differences in VUDS pre and post complex tethered cord release surgery.

**Methods:** An IRB approved registry of urology patients was queried to identify all children with a spinal dysraphism who had undergone complex tethered cord release from 6/2015 through 6/2022 with both pre and post-operative VUDS within 12 months of surgery. Data was gathered via prospectively maintained flowsheets. Fischer’s exact test and Welch’s t-test were used to determine pre and postoperative differences in VUDS.

**Results:** A total of 60 patients were included. Mean age was 3.1 years (SD=2.9) and 35 were male (58.3%). VUDS were performed 3.6 months prior to surgery (SD=2.6) and 6.0 months post-operatively (SD=2.7). There was a statistically significant increase in the number of patients who reached 75% of expected bladder capacity (EBC) (p=0.03). There was a statistically significant decrease in storage pressure at 25% EBC (p=0.05), 50% EBC (p=0.05) and at actual capacity (p=0.04) (Figure 1). Detrusor sphincter dyssnergia was noted in 27 patients preoperatively (45%), and 19 patients postoperatively (31.7%) (p=0.159). Of the 42 patients who had detrusor overactivity (DO) preoperatively, 27 continued to experience it postoperatively; meanwhile, 7 new patients developed it postoperatively (p=0.091). Of the 8 patients who had vesicoureteral reflux preoperatively, 5 continued to exhibit it postoperatively, and 2 new patients developed reflux postoperatively (p<0.001). There was a statistically significant decrease in the number of patients with upper urinary tract dilation pre and postoperatively (preoperative n=14 (23.3%), postoperative n=8 (13.3%), p=0.007).

**Conclusions:** Changes in bladder function can indicate the presence of sTSC. Spinal cord detethering improves some VUDS parameters within 12 months of surgery. Further studies are warranted to better understand the pre-operative characteristics and post-operative expectations for complex tethered cord release following sTSC.

### Variability in cystatin C and creatinine based equations for chronic kidney disease staging in adult spina bifida patients

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**Background:** Estimated Glomerular Filtration Rate (eGFR) is the standard method for determining kidney function and chronic kidney disease (CKD) stage classification. In the Spina Bifida (SB) population, eGFR estimates from a creatinine-based equation may not accurately represent kidney function due to the lower muscle mass in this population. We hypothesized that estimating equations utilizing Cystatin-C (CysC) would yield different CKD classifications than Creatinine (Cr) – based equations for the adult SB population.

**Methods:** We conducted a retrospective chart review of all patients seen in an adult urology clinic with a diagnosis of spina bifida from 2004-2021 that were managed in our urology clinic with creatinine and cystatin C values available. All patients were > 18 years old. A cross-sectional study of patients with a CysC and concurrent Cr measurement was performed; the most recent lab values were utilized if multiple existed. Three adult eGFR equations were utilized. Patients were staged based on CKD classification (GFR > 90, GFR 60-90, GFR <60). A Chi-square test was conducted to determine the difference in CKD classification based on each equation.

**Results:** A total of 105 SB patients (63% female) were included in cross-sectional analysis. The mean age was 33 ± 6.5 years. Medical comorbidities including diabetes (12.4%), hypertension (28.6%), obesity (34.2%) were noted. Table 1 shows the difference in eGFR and CKD classifications between equations. On Chi-square analysis, CKD classifica-

tions were significantly different by the type of equation used ( $p = 0.007$ ). Pair-wise comparison showed that significance was driven by the difference between cystatin C and creatinine equations ( $p = 0.002$ ) and the cystatin C and combined equations ( $p = 0.05$ ).

**Conclusions:** In an adult SB population, CysC-based equations result in lower eGFRs and different CKD classifications. Determining an appropriate eGFR measurement is essential to assessing and managing renal function in these medically complex patients.

### Comparing cystatin C-estimated and creatinine-estimated glomerular filtration rates in patients with thoracic versus sacral motor levels of spina bifida

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**Background:** Myelomeningocele (MM) type spina bifida is associated with a high risk of development of kidney disease secondary to a neurogenic bladder. Creatinine is routinely used as a marker of kidney function in the spina bifida population, while to our knowledge fewer clinics monitor kidney function using Cystatin C. We aim to examine differences between creatinine-estimated glomerular filtration rate (Cr-eGFR) and cystatin C-estimated GFR (CysC-eGFR) in patients with thoracic versus sacral level MM given expected differences in muscle mass based on spinal cord motor level within the same population.

**Methods:** Concurrently obtained creatinine and cystatin C levels for 57 adults with MM (thoracic n=44 [77%]; sacral n=13 [23%]) were evaluated using a retrospective chart review (2005-2018). Calculated Cr-eGFR and CysC-eGFR were compared.

**Results:** Mean Cr-eGFR was significantly higher for thoracic (140.8 mL/min [SD=23.9]) versus sacral MM (112.0 mL/min [SD=22.6]) ( $p=0.0003$ ). There was no significant difference in CysC-eGFR between thoracic MM (124.8 mL/min [SD=17.9]) and sacral (116.6 mL/min (SD=23.7) motor levels. The mean difference between Cr-eGFR and CysC-eGFR in thoracic MM (24.2 mL/min [SD=16.3]) was sig-



nificantly greater than in sacral MM (-12.8 [SD=15.7]) ( $p<0.0001$ ).

**Conclusions:** These data suggest that Cr-eGFR may overestimate kidney function in patients with thoracic MM, as there was a significantly higher discrepancy between Cr-eGFR and CysC-eGFR in thoracic motor levels when compared to sacral motor levels of MM. This suggests there is little difference in screening patients with sacral MM with creatinine versus cystatin C to evaluate for underlying renal disease, however, routine monitoring of patients with thoracic level MM with cystatin C should be considered.

### Using machine learning of urodynamics to predict clinical outcomes in patients with spina bifida

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**Background:** Urodynamic (UDS) testing is fundamental for guiding neurogenic bladder management in patients with Spina Bifida. UDS findings correlate with the development of adverse urinary outcomes. However, associations have been based on single UDS variables without encompassing the multitude of parameters provided by UDS testing. Furthermore, despite the relevance of accurate UDS readings, heterogeneity in UDS interpretation may hinder optimal management. Machine learning offers the potential to address these limitations by standardizing UDS interpretation and accounting for the complexity of UDS data. Here, we aim to use machine learning to predict clinical outcomes based on urodynamic data in patients with Spina Bifida.

**Methods:** Clinical and UDS data were retrospectively extracted from patients at a single pediatric urology clinic who underwent UDS testing between May 2012 and September 2020. Logistic regression models were generated to predict getting a symptomatic UTI within 1 year of UDS testing and/or hydronephrosis within 2 years of testing. Models were

trained on 75% of the data and 25% of the data was used for performance evaluation. Performance was compared between models with both UDS and clinical features and clinical features only.

**Results:** This study included 305 patients with total of 509 UDS tests. There were 191 instances of patients developing a UTI within 1 year of UDS testing and 185 instances of patients developing hydronephrosis within 2 years of testing[RF1]. Machine learning models that incorporated UDS data predicted the development of UTIs (AUC=0.65, [0.53-0.74] 95%CI) and hydronephrosis (AUC=.063m [0.53-0.73] 95%CI). Models that did not include UDS data failed to predict either UTIs or hydronephrosis.

**Conclusions:** Machine learning models based on urodynamic data can predict the development of future UTIs and hydronephrosis. Models are improved by inclusion of urodynamic data. Machine learning models may help guide clinical decision making to prevent adverse urinary outcomes in individuals with spina bifida.

### Radiation safety in pediatric videourodynamics: Introducing a standardized imaging protocol

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**Background:** A standardized protocol for imaging in children with spina bifida (SB) during VUDS has not been established. We previously demonstrated our utilization of fluoroscopy during VUDS was consistent with the ALARA principle but was not standardized. The aim of this study is to implement a standardized imaging protocol for children with SB during VUDS and compare to our prior findings.

**Methods:** This is an IRB approved retrospective study from 2013 through 2022 of consecutive SB patients undergoing VUDS by a single provider. Patients were categorized into three age groups; group 1 (0-2YR), group 2 (2-10YR), group 3 (>10YR). Radiation data was reported as mean air kerma (AK), dose area product (DAP), exposure time (seconds) and effective dose (ED). Risk was reported as lifetime attributable risk (LAR). All patients undergoing VUDS after May of 2020 underwent the following imaging capture protocol: scout film, bladder capacity at 25%, 50%, 75% and 100%, +/- image with evidence of detrusor overactivity, +/- image

with plateau in pressure to assess for reflux, image with voiding to assess bladder neck/spincter. Comparison was made to our prior findings.

**Results:** 67 patients were assessed and compared to our original cohort. Pre and post protocol data points are shown in table 2. ED increased with age in both cohorts. Groups 1 and 2 in the protocol cohort were found to have statistically significant higher ED ( $p < 0.0001$ ,  $p = 0.0020$ ). There was no statistically significant difference of the ED of group 3. The LAR for the protocol VUDS groups 1-3 was 0.003, 0.002, and 0.006, respectively; corresponding to a 0.3%, 0.2%, and 0.6% chance, respectively, of developing cancer as a result of the radiation exposure from a VUDS.

**Conclusions:** Our findings support that our proposed imaging protocol for SB VUDS remains consistent with the ALARA principle. While greater patient numbers are needed, this preliminary data shows no statistically significant difference in risk from radiation exposure compared to our initial findings.

### **Role of renal ultrasonography during the first year of life in myelomeningocele patients: Data from the Urologic Management to Preserve Initial Renal Function (UMPIRE) protocol**

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**Background:** Various schedules for renal/bladder ultrasound (RBUS) in patients with myelomeningocele (MMC) have been arbitrarily defined and utilized. Newborns enrolled in the CDC-funded

Urologic Management to Preserve Initial Renal Function (UMPIRE) Protocol study undergo RBUS every 3 months during the first year of life, then annually thereafter. Our objective was to define the probability of a positive RBUS in the first year of life given a positive or negative initial RBUS at birth. We hypothesize the incidence of abnormalities found on RBUS is low enough to warrant fewer screening ultrasounds during this critical growth period.

**Methods:** Serial RBUS performed over the first year of life for newborns enrolled in UMPIRE were analyzed. Dilating hydronephrosis (SFU grades 3-4) or an increase of  $\geq 2$  SFU grades were deemed positive. Hierarchical Bayesian logistic models were used to estimate the probability of a positive RBUS.

**Results:** In total, 421 infants with 1595 RBUS at birth, 3, 6, 9, and 12 months were analyzed. Thirty-one infants collectively had 62 positive RBUS results. Those with a negative RBUS at birth had a low probability of significant findings at months 3 (0.01%, 95% CI 0-22.5%), 6 (0.02%, 95% CI 0-32.66%), 9 (0.02%, 95% CI 0-35.6%), and 12 (0.02%, 0-33.10%). Infants with a positive RBUS at birth had a high probability of being positive: at months 3 (55.2%, 95% CI 0.5-99.3%), 6 (67.5%, 95% CI 0.8-99.6%), 9 (70.2%, 95% CI 0.9-99.6%), and 12 (67.9%, 95% CI 0.8-99.6%). Using the 3-month RBUS as the screening ultrasound rendered similar probability trends.

**Conclusions:** In infants with MMC it is safe to perform less frequent surveillance imaging during their first year of life. If an infant's RBUS is negative for hydronephrosis at birth and/or 3 months of age, there is a low probability of significant hydronephrosis developing later during the first year of life.

### **Can contrast enhanced urodynamics sonography replace fluoroscopy during video urodynamics? A structured comparison pilot**

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**Background:** Video urodynamics (VUDS) synthesizes anatomical and functional details as one component of medical and surgical decision-making in

patients with neurogenic bladder (NGB). The resultant radiation exposure limits availability and use as surveillance. We aim to explore contrast-enhanced urodynamics sonography (CEUDS) using intravesical microbubble contrast during urodynamics (UDS) as a radiation-free replacement for VUDS.

**Methods:** A pilot study to compare VUDS to CEUDS was developed. Children with NGB secondary to spina bifida or caudal regression were prospectively enrolled to undergo CEUDS immediately following standard of care VUDS. Three UDS runs were performed, of which the last two were used for analysis. Images were obtained at set percent estimated bladder capacity (0, 25, 50, 75, 100, etc.) and with observed urodynamics “events” to assess for open bladder neck or reflux. Each tracing was classified as safe, intermediate, or hostile using the National Spina Bifida Patient Registry (NSBPR) definitions by a blinded urologist. Concordance between VUDS and CEUDS tracings was recorded.

**Results:** In total, 8 patients 3-to-10-years-old were enrolled and completed testing. The UDS tracings were comparable between VUDS and CEUDS, with the classification matched in 100% of VUDS-CEUDS pairs [Safe (5), Intermediate (2) and Hostile (1)]. Layering of microbubbles over residual contrast and rupture of bubbles after gentle warming were encountered on one occasion each and rectified. Based upon matched UDS tracings and quality of imaging, the UDS infusion pump did not interfere with microbubble quality and there was no suggestion of transducer interference by microbubbles.

**Conclusions:** Substitution of intravesical microbubble contrast for standard fluoroscopy during UDS appears to provide comparable urodynamic information based upon these pilot data. Further studies will validate these observations and their radiologic correlates for confident decision-making with various diagnoses, ages, and body types.

## Diagnostic test characteristics of ultrasound-based hydronephrosis for chronic kidney disease in children and adolescents with spina bifida: Results from the UMPIRE and NSBPR cohorts

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**Background:** Renal ultrasounds (RUS) are performed in patients with spina bifida (SB) to screen for hydronephrosis as a marker of kidney health. We determined the diagnostic test characteristics of ultrasound-based hydronephrosis for low kidney function defined by estimated glomerular filtration rate (eGFR).

**Methods:** We performed a retrospective cross-sectional study using two cohorts of children and youth with SB. One cohort is from the Urological Management to Preserve Initial Renal Function Protocol for Young Children with Spina Bifida (UMPIRE; 2016-2022), and the second is from the National Spina Bifida Patient Registry (NSBPR; 2009-2021). Eligibility criteria included patients 1-to-18-years-old with available eGFR data (ie, serum creatinine and height/arm-span measurement) within 6 months of a RUS. We excluded NSBPR patients <6-years-old to address potential duplication across cohorts (as some participants are in both NSBPR and UMPIRE). The primary outcome of interest was low eGFR<90mL/min/1.73m<sup>2</sup> using the bedside

Schwartz formula. Hydronephrosis was dichotomized into any/none. We calculated sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) with 95% confidence intervals.

**Results:** In UMPIRE, 276 patients were included with median age 2.4 years, 52% male sex, 100% myelomeningocele diagnosis, and 23% having eGFR<90. Any hydronephrosis versus none conferred a sensitivity/specificity/PPV/NPV of 24/75/22/77%, respectively. In NSBPR, 2000 patients were included with median age 11.9 years, 49% male sex, 79% myelomeningocele diagnosis, and 14% having eGFR<90. Any hydronephrosis versus none conferred a sensitivity/specificity/PPV/NPV of 41/82/27/89%, respectively.

**Conclusions:** In two cohorts of children and youth with SB, hydronephrosis conferred a sensitivity of 24-41% for a creatinine-based eGFR<90mL/min/1.73m<sup>2</sup>. This low sensitivity suggests that RUS alone is a poor screening marker of kidney health and it may help to combine with routine serum testing.

## Urological outcomes of 5-year-old patients after open prenatal spina bifida aperta repair

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**Background:** To investigate urological outcomes after open prenatal spina bifida aperta repair (OPSBAR) at the age of five years.

**Methods:** Patients with OPSBAR at the Zurich Center for Spina Bifida were followed in a standardized program. Data obtained at follow-up visit was collected in a RedCap database. Patients five years or older were considered for this study.

**Results:** 54 patients with OPSBAR were at least 5 years old at the time of analysis. Twelve were excluded due incomplete data (11) and denied research

consent (1). The 5-years follow-up of the 42 included patients took place at a mean age of 5.0 years. 16% (7/42) showed normal cystometry and were able to void volitionally, six being fully continent and one diaper dependent. 81% (34/42) were on clean intermittent catheterization (CIC). 24 of these 34 (71%) were on anticholinergics. CIC was started in the neonatal period in 18 patients. Ten were placed on CIC in the following eleven months (mean 6.8 months) and eight after the age of one year (mean 30.9 months). A hyperactive/high pressure bladder was the reason to start CIC in 28 patients (usually combined with instillation of intravesical anticholinergics). In eight patients, the reason was an acontractile bladder. In one patient, CIC could be stopped at the 2 year follow-up visit. In another, CIC was stopped after vesicostomy placement.

In six patients (14%), CIC became necessary only after neurosurgical resection of inclusion cysts (mean 28.3 months). Five showed a persistent acontractile bladder and one a hyperactive bladder. Performed surgeries were Botulinum toxin injection in the detrusor (7), bladder sling with appendicovesicostomy (1) and vesicostomy (1).

**Conclusions:** Only a minority of patients with OPSBAR shows a normal bladder function at 5 years. However, the percentage is higher than reported percentages after postnatal repair. A non-negligible number developed neuropathic bladder dysfunction after inclusion cyst resection.

## Cystatin C-based estimated glomerular filtration rate equation has higher sensitivity in detection of kidney dysfunction than equations containing creatinine in spina bifida and other non-weight-bearing conditions

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**Background:** Individuals with spina bifida are commonly non-weight-bearing (NWB), and muscle mass differs from the general population. Muscle mass is a main determinant of serum creatinine (sCr), but not cystatin C. We sought to compare eGFR equations that include serum creatinine, cystatin C, or both, in NWB individuals and matched ambulatory controls.

**Methods:** EMR was reviewed for individuals with available sCr and cystatin C and a NWB diagnosis. A matched control group was identified without NWB diagnosis. eGFR was calculated using the 2021 CKD-EPI equations. Demographics and renal function were compared using descriptive statistical analyses. Renal dysfunction was defined by renal ultrasound abnormalities (length more than two standard deviations below the population mean, abnormal echogenicity or thickness, evidence of scarring) or proteinuria.

**Results:** The NWB (n=102) and control populations (n=204) had similar demographics. In the NWB population, 60% of whom had spina bifida, eGFR differed when calculated using CKD-EPI<sub>Cr</sub>, CKD-EPI<sub>Cr+Cys</sub> and CKD-EPI<sub>Cys</sub> (107, 93, 80 mL/min/1.73 m<sup>2</sup>, respectively, p<0.001). eGFR did not differ by equation in the control group. In the patient subset with evidence of renal dysfunction, the NWB population again had different eGFR when using CKD-EPI<sub>Cr</sub>, CKD-EPI<sub>Cr+Cys</sub> and CKD-EPI<sub>Cys</sub> (p<0.001), but the control population did not. 58% of NWB individuals with imaging or urinalysis evidence of kidney dysfunction were reclassified into a lower eGFR category when using CKD-EPI<sub>Cys</sub> relative to CKD-EPI<sub>Cr</sub>.

**Conclusions:** Calculation of eGFR using CKD-EPI<sub>Cys</sub> was more sensitive in identification of kidney dysfunction relative to the CKD-EPI<sub>Cr</sub> equations. Comparison of these equations against gold standard GFR measurement is warranted to confirm which most closely approximates true GFR.

## Anorectal manometry characteristics of pediatric spina bifida

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**Background:** Myelomeningocele, commonly known as spina bifida, occurs when the caudal neural tube fails to fuse during fetal development. It is one of the most common fetal malformations occurring in approximately 1:3000 live births worldwide. Symptoms that result from spina bifida include bowel and bladder dysfunction, paraplegia, hydrocephalus, and orthopedic anomalies. Fecal incontinence

associated with spina bifida has been shown to negatively impact quality of life for these patients.

Comprehensive bowel management in children with spina bifida allows for predictable, independent bowel movements, and social continence. Development of a bowel management treatment plan in children with spina bifida often includes anorectal manometry testing.

There is limited pediatric data on the anorectal manometry characteristics of patients with spina bifida. The aim of our study was to assess the anorectal manometry characteristics of patients with spina bifida compared to pediatric controls with functional constipation.

**Methods:** This retrospective study compared 38 children with spina bifida to 243 controls with functional constipation followed by Children's Hospital Los Angeles between April 2018 and April 2021. Anorectal manometry was performed using a Medtronic ManoScan™ High Resolution AR Manometry catheter and analyzed with ManoView™ AR Analysis Software.

**Results:** Patients with spina bifida were found to have lower baseline sphincter pressures, lower maximum sphincter squeeze pressures, and prolonged rectoanal inhibitory reflex (RAIR) recovery compared to controls (p value <0.05). Spina bifida patients were also more likely to have unsuccessful balloon expulsion during simulated defecation. There were no differences in sensation (first sensation, urge, and discomfort) in the two groups.

**Conclusions:** Obtaining acceptable ranges for anorectal manometric data specific to spina bifida patients may allow for improved bowel management program development, as well as improve detection of spinal cord anomalies in patients with intractable constipation. Our study found that patients with spina bifida were more likely to have lower baseline sphincter pressures, lower maximum sphincter squeeze pressures, and prolonged RAIR recovery. There were no significant differences in sensation between groups suggesting that fecal incontinence is likely related to low sphincter tone and control of the sphincter in spina bifida patients as opposed absence of sensation.

## Gynecological care needs for women with spina bifida establishing with adult gynecology

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**Background:** This study aims to 1) describe the demographic and clinical characteristics of women with spina bifida (SB) establishing adult gynecological care and 2) assess the frequency at which age-based USPSTF preventative health guidelines for gynecological care are met within this population and 3) to identify factors associated with being up-to-date with recommended age-based guidelines.

**Methods:** Demographic and clinical data of patients with SB who established care with an adult gynecologist affiliated with a medical home clinic for adults with intellectual/developmental disabilities were collected via chart review of the electronic

health record (EHR). Data were collected from EHR visit notes and the problem list at the time of the initial gynecology visit and reported in medians and frequencies. Clinical data included SB condition specific characteristics, gynecological history and exam findings, and completion of recommended USPSTF preventative health guidelines for gynecological care. Logistic regression was used to determine associations between characteristics and preventative health outcomes.

**Results:** Sixty women with SB established adult gynecological care during the study period, which represents 68% of all eligible patients at the medical home. The most common reasons for establishing care were for well-woman care (n=31, 51.7%) and menses concerns (n=21, 35%). Of the 44 who underwent a pelvic exam at this first visit, 43% had normal findings.

Only 23.3% (n=14) had a previous HIV screen, 43.3% (n=26) had begun the HPV vaccine series, and 26.7% (n=16) had cervical cancer screening. Age at which care was established was directly associated with HIV screening (p=0.021) and inversely associated with starting the HPV series (p=0.005). Sexual activity history was associated with higher rates of cervical cancer screening (p=0.014)

**Conclusions:** This study highlights the need for more comprehensive and accessible gynecological care for patients with SB in order to improve preventative health outcomes in this population.