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Abstracts

Urology Session 1

Baseline Urinary Tract Imaging Infants with Spina Bifida

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Background Urologic complications cause significant morbidities in Spina Bifida (SB) and predispose to chronic kidney disease (CKD). Optimizing urologic management may improve continence, reduce surgical need, and prevent CKD. To define optimal SB urologic management using a proactive approach, Centers for Disease Control and Prevention (CDC), collaborating with nine SB centers, developed a single consensus-based protocol for the urologic care of children with SB from birth-5 years.
Methods Enrollment in the National SB Patient Registry Urologic and Renal Protocol for the Newborn and Child began in February of 2015. This analysis includes 101 infants with at least one of the following: baseline renal ultrasound (RUS), cystogram, and dimercaptosuccinic acid (DMSA) renal scan. Hydronephrosis (HN) was graded using the Society of Fetal Urology classification. Vesicoureteral reflux (VUR) was graded 1-5. DMSA renal scarring was graded using the Randomized Intervention for Children with VUR (RIVUR) scale.
Results In the 101 infants (54% boys; 53% white, 30% Hispanic, 11% black), RUS demonstrated 62% no left HN, 18% Grade (G) 1 HN, 12% G2, 2% G3, and 1% G4. On the right, 63% had no HN, 18% G1, 11% G2, 2% G3, 0% G4. 23% of infants had bilateral HN G1 or above. DMSA scan (30 infants) showed 97% of left and 93% of right kidneys had no segments affected. No child had >G1 scarring. Cystograms (72 infants) revealed 92% with no left and 89% with no right reflux. Of those with left reflux, 3% had G5 and 1% each had G 1-3. Of those with right reflux, 6% had G1, and 1% each had G2-3 and 5. 6% of infants had bilateral VUR.
Conclusions In SB infants, baseline RUS demonstrated normal renal parenchyma and no or minimal HN. Baseline renal scarring and VUR are uncommon (< 1 0%). These minimal findings on baseline imaging underscore the importance of a proactive protocol to manage urologic care in children and preserve renal parenchyma and reduce progressive CKD.

Examining the Role of Screening Renal Ultrasound in Spina Bifida During the First Year of Life

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Background Infants and children with Spina Bifida are routinely followed with renal ultrasound (RUS) surveillance, in order to identify changes that may suggest deterioration of bladder dynamics. However, the ideal frequency and time interval for screening remains unclear and trends among practices vary widely. We sought to characterize ultrasound findings at our institution and how they affected management.

Methods A retrospective review of infants with Spina Bifida on a standardized clinical management protocol between 2014 and 2016 at our multidisciplinary clinic was performed. Patients were seen in our clinic at 3, 6, 9, and 12 months of life. RUS was performed with each visit. Patient data and clinical course were reviewed.
Results A total of 32 patients were identified, of which 29 (90.6%) had myelomeningocele and 3 (9.4%) had lipomyelomeningocele or lipomyelocele. The finding of unilateral or isolated SFU grade 1 hydronephrosis uniformly resolved within 3 to 6 months. One (3.1%) lipomyelomeningocele patient developed bilateral SFU grade 1 and 2 hydronephrosis at 9 months that worsened and led to the diagnosis of tethered cord syndrome. One (3.1%) myelomeningocele patient was hospitalized for a urinary tract infection at 3 months and found to have new bilateral hydroureteronephrosis and de novo vesicoureteral reflux.
Conclusions In our single -institution cohort of infants with Spina Bifida, patients screened with quarterly RUS in the first year of life rarely had abnormal findings that led to alterations in our urologic management. The isolated finding of SFU grade 1 hydronephrosis appeared to be negligible, similar to what is reported in non-Spina Bifida infants. Because other diagnostic urologic procedures are often performed in the first year of life, close clinical follow-up with less screening RUS may be feasible. Further studies in larger populations may reveal the ideal surveillance schedule for RUS.

Cause-specific Mortality in Patients with Spina Bifida (SB) and End Stage Renal Disease (ESRD)

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Background Patients with SB are prone to progressive renal damage due to neurogenic bladder. The most severe form of renal damage, ESRD, is assocaited with high mortality rates. We examined the actual causes of deaths in ESRD patients with SB in a national data system.
Methods Using hospital inpatient data (1977–2013) from the United States Renal Data System (USRDS), we identified persons with SB using the ICD-9-CM code 741. For each SB case with ESRD we identified 5 cases of ESRD without SB, matched by age at first ESRD service, year at first ESRD service, gender, and race/ethnicity. We then compared the cause-specific mortality for these two groups in five major categories: cardiovascular disease (CVD), infection, malignancy, other known causes (miscellaneous), and unknown/missing. We used Cox regression to analyze cause-specific hazard ratios for death, adjusting for demographic and clinical covariates.
Results In the study period we identified 2368 ESRD patients with SB. On average, they developed ESRD at age 34 years. Compared to ESRD patients without SB, ESRD patients with SB were more likely to have urological causes for their ESRD (28.8% vs. 2.7%); less likely to receive a renal transplant (36.1% vs. 50.9%); equally likely to survive on dialysis (13.4%); more likely to die of infection (10.1% vs. 4.5%), CVD (18.6% vs. 16.7%), miscellaneous causes (10.0% vs. 5.8%), and unknown causes (10.3% vs. 7.6%). About 80% of deaths by infection among ESRD patients with SB were unrelated to dialysis. Hospitalizations related to urinary tract infections (UTIs) were positively associated with the risk of death by infection for ESRD patients with SB.
Conclusions Compared to ESRD patients without SB, those with SB were substantially more likely to die from infections, mostly from causes unrelated to dialysis. UTI hospitalizations were associated with increased probability of death by infection in these patients.

Ultrasound Estimated Bladder Weight Correlates with Videourodynamic Studies in Neurogenic Bladder Dysfunction

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Background This retrospective study was designed to evaluate which lower urinary tract ultrasound parameter(s) could predict the results of invasive urodynamic testing which are the current gold standard in the evaluation of bladder dysfunction in children with Spina Bifida.
Methods 58 children with Spina Bifida undergoing video urodynamic evaluation and a renal bladder ultrasound as their standard of care were evaluated. Quantitative and qualitative ultrasound parameters were then correlated with the videourodynamic study results which served as the gold standard.
Results For those bladders with an ending storage pressure above 15 cm H_2O there were increases in

these ultrasound measured parameters: I) bladder mass ($p = 0.00019$), II) bladder/body mass ratio ($p = 0.0059$), and III) wall thickness ($p = 0.01$). We defined the storage cost as the final storage pressure divided by the percentage of expected bladder capacity attained. Using a cutoff value of 15, the bladder mass differed significantly between these two populations ($p = 0.001$). For an ultrasound determined bladder mass of less than 40 g, the sensitivity for predicting a storage cost < 15 cm H_2O was 66% with a specificity of 78%. The sensitivity for predicting a storage cost $<$ 20 cm H_2O was 78% with a specificity of 70%.

Conclusions Bladder weight is independent of luminal volume, can be normalized to body weight, and may serve as a clinically valuable tool for noninvasive screening to define a subset of patients with neurogenic bladder with a higher likelihood of having abnormal videourodynamic results.

DMSA in Young Children with Myelodysplasia: Should this be Routinely Obtained?

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Background Infants with neurogenic bladder due to myelodysplasia (MD) are often evaluated for renal abnormalities using DMSA scan. DMSA scans are expensive and expose children to radiation. We sought to identify risk factors for development of DMSA abnormalities and hypothesized that these could improve the utility of DMSA imaging.

Methods We reviewed records of MD patients from Jan 2008 to Oct 2016. We included those who underwent initial DMSA as part of a routine evaluation less than 2 years of age. We evaluated for association between hydronephrosis (HN), vesicoureteral reflux (VUR), detrusor end fill pressure (EFP) > 40 cm H2O, history of febrile UTI and subsequent DMSA abnormality.

Results 136 patients identified underwent initial DMSA at median age of 4.7 mo (IQR 3.3–8.9 mo). Median number of DMSA scans was 2 (IQR 1–3). 12 had initial abnormal DMSA. 21 developed abnormal DMSA in follow up for a total of 33/136 (24%) with abnormal DMSA. Compared to the 103 with normal DMSA scans, the 33 with abnormal DMSA scans were more likely to have VUR (76% vs 28%, $p < 0.001$), HN (73% vs 24%, $p < 0.001$), EFP > 40 cm H2O (64% vs 23%, $p < 0.001$), and febrile UTI (85% vs 37%, $p < 0.001$). On multivariable cox proportional hazard analysis, VUR (HR $= 4.7$, $p < 0.001$), febrile UTI (HR $= 2.6$, $p = 0.02$), and EPF > 40 cm H2O (HR $= 2.6$, $p = 0.01$) were independently associated with subsequent abnormal DMSA. Only 1/33 (3%) with abnormal DMSA did not have prior VUR, HN, febrile UTI, or high EFP; this patient had HN within 3 months of the abnormal DMSA.

Conclusions Children with MD who have VUR, EFP > 40 cm H2O, and febrile UTI are at significantly increased risk for DMSA abnormalities and for patients without any of these risk factors DMSA had very limited utility. Given the cost and radiation exposure, DMSA should be used selectively in this patient group and reserved for those with risk factors.

Agreement Between Electronic Medical Records and Self-Reported Urologic Domains in the National Spina Bifida Patient Registry

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Background CDC National Spina Bifida Patient Registry (NSBPR) sites can collect continence and bladder/bowel management data via patient self-report (SR) and/or review of provider notes in the electronic medical record (EMR). Anecdotally, we have noted discrepancies between SR and the EMR for these prospectively collected data. We examined SR/EMR agreement for 10 urologic domains captured for the NSBPR in our MM clinic.

Methods NSBPR participants with an annual visit between 12/1/14–1/31/16 were identified. Data from urology provider notes in our hospital EMR was categorized into domains corresponding to 10 NSBPR urologic data points. Overall agreement (number of agreeing pairs/number of complete pairs) and strength of agreement, indicated by Cohen's Kappa Coefficient, were reported. Analysis of incontinence was limited to toilet trained children $\geqslant$ 5yo. Analyses were further stratified by demographic factors.

Results Our cohort included 176 patients (median age 11.4 years, range 0.4–24.1), 52% female, 80% white and 27% Hispanic. Diagnoses included Myelomeningocele (MM) (41%), Fatty filum (30%) and Lipoma (19%). Bladder management included clean intermit-

tent catheterization (CIC) in 47%, volitional voiding in 38% and no established program in 14%. Overall SR/EMR agreement was $\geqslant$ 90% for 8/10 domains (range 69–99%).
Overall SR/EMR agreement was lowest for daytime urinary and stool incontinence (DUSI). Sub-analyses demonstrated MM's with twice as much SR/EMR disagreement about DUSI as non-MMs; the same was true for children on CIC when compared to volitional voiders.
Conclusions We found strong agreement ($\geqslant$ 90%) between SR NSBPR urologic domains and the hospital EMR in all categories except for DUSI. Disagreement about DUSI was more likely in complex patients: MMs and those on CIC. Future studies are needed to determine what provider or patient factors contribute to discordant reporting.

Lipomyelomeningocele for the Urologist: Should We View it the Same as Myelomeningocele?

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Background Preserving renal integrity and achieving continence are primary urologic objectives for lipomyelomeningocele (LMM) and myelomeningocele (MM). We have viewed LMM and MM the same: at risk for issues in the short and long term. Unlike MM, many LMM present with a cutaneous lesion and no evident neurological, orthopedic or urological dysfunction. We hypothesize that anatomic and functional factors, as well as early and complete tethered cord release (TCR), determine urologic outcomes. We examine predictors for long -term continence/CIC after primary TCR.
Methods We identified 143 patients who had a TCR for LMM between 1995 and 2010. Patients with concomitant anorectal/genitourinary anomalies, filar lipoma, fatty filum, previous TCR, and follow up $<$ 1 year were excluded. Analysis was limited to patients who were either toilet trained or older than 6 years at last follow up. Ability to achieve urinary continence, with or without CIC, was the primary outcome.
Results A total of 56 patients (27 males) with LMM met inclusion criteria. Median age at surgery was 4.4 months (range 1–224 months); average follow-up of 10 years (range 1.3–19.1 years). 68% were asymptomatic at presentation (cutaneous lesion/ incidental finding on screening). Presentation included urologic symptoms in 9% (5/56), with a median age at surgery of 108 months in this subgroup. At last follow-up, 86% of the patients were continent spontaneously (38) or were dry on CIC (10). 8 were incontinent at last follow-up, with 3 on CIC. Of the 5 who presented with urologic symptoms, all are continent but 4/5 (80%) require CIC. Overall 23% of patients require CIC. Continence at last follow-up was not significantly associated with any anatomic, surgical or functional variable.
Conclusions Among patients with primary TCR for LMM, prospects for continence were excellent. Families can anticipate 23% likelihood of CIC over the long term, considerably less than in MM.

Early Postnatal Bladder Function in Fetoscopic Myelomeningocele Repair Patients

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Background In-utero repair of myelomeningocele (MMC) via hysterotomy has demonstrated neurologic and orthopedic benefits, when compared to postnatal repairs. Urologic benefits, however, have yet to be proven. Our institution has pioneered an endoscopic approach for repair using an exteriorized uterus. Our aim was to review early postnatal bladder function in these patients.
Methods A prospective urologic assessment of patients undergoing fetoscopic MMC repair and receiving subsequent care at our facility, was performed. Patients were managed and urodynamic studies risk-stratified according to the Centers for Disease Control and Prevention Urologic and Renal Protocol for the Newborn and Young Child with Spina Bifida. Per protocol, urodynamic studies were performed at three months of age.
Results Fetoscopic MMC repair was performed in seven patients at a mean gestational age of 24.6 $\pm$ 1.2 (SD) weeks. Mean gestational age at birth was 35.8 $\pm$ 3.6 (SD) weeks. One (14.3%) patient required an endoscopic third ventriculostomy. Mean urologic follow-up was 11.3 $\pm$ 7.7 (SD) months. No patients had hydronephrosis or bladder thickening at birth. Detrusor

overactivity was observed in five (71.4%) patients. Impaired compliance was seen in five (71.4%) patients. No patients had a detrusor leak point pressure of $>$ 40 cm H_2O or evidence of detrusor sphincter dyssynergia. Two (28.6%) patients had vesicoureteral reflux, three (42.9%) had an open bladder neck, and none had trabeculated bladders. Two (28.6%) patients had abnormal but safe, and five had intermediate-risk bladders. No patients required catheterization upon discharge from the hospital.
Conclusions In our early experience with fetoscopic MMC repair, postnatal bladder function does not appear to be any worse than that of previously reported in -utero or postnatal closures.

Urology Protocols in Patients That Have Undergone *In Utero* Repair of Myelomeningocele: Room for Improvement

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Background Variability exists on what urologic testing is deemed necessary to provide optimal care for patients with myelomeningocele. *In utero* surgery has resulted in motivated families of affected fetuses travelling to centers that perform such surgeries and are perceived to be more compliant with provider recommendations. We sought to determine how often *in utero* repaired patients will complete physician recommended urology testing in the setting of multidisciplinary myelomeningocele clinic.
Methods We retrospectively reviewed our *in utero* repair of myelomeningocele patient cohort from 2011–16 to determine how often patients underwent urology testing outlined in our institutions urologic protocol. Our protocol involves obtaining renal ultrasound and VCUG at birth; renal ultrasound every 3 month in first year of life with decreasing frequency in subsequent years; annual urodynamics, and annual follow-up VCUG if vesicoureteral reflux detected until resolution.
Results We identified 39 infants underwent *in utero* repair. 100% of patients underwent renal ultraosund prior to discharge. Six patients did not undergo urology follow-up at our hospital. Renal ultrasounds were obtained according to protocol every three months in first year of life in 31 of the 33 followed patients. VCUG and urodynamics were performed in the first year of life in 32 and patients, repsectively. Successful adherence to the urology testing protocol in the second, third, fourth and fifth year in eligible patients occurred in 66%, 66%, 83% and 50%, respectively.
Conclusions Urologic testing was reliably completed according to protocol at birth however with decreasing reliability as patients aged for a variety of reasons. We continue to review our protocol to refine it while minimizing cost and burden of testing.

Deferment in Urinary Self-catheterization Among Individuals Living with Spina Bifida: A Call to Action

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Background A few early studies have indicated that self-catheterization can be achieved by five years of age in patients with myelodysplasia. However, in recent decades there has been a lack of educational initiatives targeting promotion of self-catheterization. The purpose of our study was to explore and describe the current age of self-catheterization among a diverse patient population.
Methods From July 2015 to Sep. 2016, self-catheterization skill, age at initiation, and method were studied among patients $\geqslant$ 2 years of age. Among a subset of patients, fine-motor ability was evaluated using the Beery -Buktenica Developmental Test of Visual-Motor Integration and cognitive abilities were evaluated through neurodevelopmental assessments.
Results 188 patients (92.6% myelomeningocele, 7.4% lipomyelomeningocele) indicated that some form of catheterization was utilized. Mean age of the sample was 10.5 years (SD 4.5); 52.1% were male, and 64.4% were Hispanic. Higher level of lesion was associated with a lower rate of self-catheterization. One hundred and one (53.7%) patients reported to self-catheterize (91.1% myelomeningocele, 57.4% male, and 62.4% Hispanic) with a mean age of self-catheterization of 9.5 years (SD 3.1) and a range of 2 to 17 years of age. A Mitrofanoff channel was used by 25.7% (50% male; 38.5% Hispanic) of patients who self-catheterized, with a mean age of independence of 9.45 years (SD 2.6). The mean age of independence for urethral self-catheterization was 9.7 years (SD 3.4).
Conclusions The data demonstrated that a higher level of lesion and female gender were associated with a

lower rate of self-catheterization. Concurrently, Hispanic ethnicity was associated with a lower incidence of Mitrofanoff use. The average age of self-catheterization emerged as a target for culturally-appropriate educational interventions to foment greater early independence.

Conservative Use of CIC in Newborns with Myelodysplasia: Report of Clinical Outcomes

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Background Some centers start all newborns with myelodysplasia (MD) on CIC from birth while others treat selectively. We report outcomes of a protocol where infants were started on CIC only when indicated and hypothesized that a minority of patients will require CIC before continence is desired.
Methods From January 2009 to October 2016, newborns with MD were started on the protocol. Per protocol, urodynamics (UD) and DMSA were obtained at 3–6 mo, 1 yr, 3 yrs, then as needed. Renal ultrasound was obtained q3 mo until 1, q6 mo until 2, then annually. We included only patients with initial visit before 6 mo of age. Indications to start CIC included recurrent febrile UTI, upper tract changes, detrusor end fill pressure (EFP) $>$ than 40 cm of H2O, or desire to achieve continence.
Results 105 patients were followed for a median of 3.4 yrs (IQR 2.0–5.0 yrs). 2 started CIC as newborns due to findings on imaging or UD.
45/105 (43%) developed febrile UTI for a 5 yr cumulative incidence of 59% (95% CI 45–73%). 36/105 (34%) had VUR diagnosed. 24/105 (23%) had DMSA abnormalities for a 5 yr cumulative incidence of 30% (95% CI 20–44%). 36/105 (34%) developed EFP $>$ than 40 cm H2O. 5/105 (5%) had vesicostomy at median age of 2.4 yrs (IQR 1.1–2.8 yrs). 36/105 (34%) started CIC at median age of 1.3 yrs (IQR 0.6–3.0 yrs) for UTIs, high EFP, or upper tract changes. Median age to start CIC for continence was 4.0 yrs (IQR 3.4–4.5 yrs) in 13. 56 have yet to be started on CIC at median age of 2.6 yrs (IQR 1.2–3.9 yrs). VUR, EFP $>$ 40 cm H2O, and febrile UTI were associated with abnormal DMSA ($p < 0.001$).
Conclusions Around 1/3 of patients on our protocol were started on CIC before continence was desired due to concerning urologic findings. Close clinical follow up is needed for patients on a protocol such as ours.