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Poster Presentations

All Areas of Basic Science Research

Early Postnatal Ablation of Inhibitory Neurons in the Spinal Cord Confers Spina Bifida-like Motor Dysfunction in a Chick Model

Khan, Md. Sakirul I., Nabeka, Hiroaki, Islam, Farzana, Shimokawa, Tetsuya, Xuan, Li, Yamamiya, Kimiko, Doihara, Takuya, Matsuda, Seiji
Ehime University Graduate School of Medicine

Background Spina bifida aperta (SBA) causes lifelong neurological complications, especially affecting motor function. It has been difficult to gain insight into the mechanisms underlying the pathogenesis of this intractable disease due to the lack of appropriate animal models. We aimed to clarify the underlying mechanisms of SBA-related motor dysfunction in chick model of SBA.
Methods The roof plate of the neural tube in chick embryos was incised longitudinally using a microknife to produce SBA chicks. The procedure was performed when embryos were at developmental stages 17–21, on day 3 of incubation. After characterizing the chicks with SBA, the interplay between parameters of inhibitory and excitatory signals in the spinal cord were assessed at different points from embryonic to post-hatching periods.
Results Chicks with surgically-induced SBA exhibited various neurological disorders consistent with SBA in human neonates: (i) disappearance of voluntary leg movements within a few weeks of hatching; (ii) complete loss of motor coordination (sit to walk) by 4 days post-hatching; and (iii) paralysis of the lower limbs by 10 days post-hatching. These neurological disorders were concomitant with the loss of GABAergic neurons, important inhibitory interneurons, in the lumbar cord. In fact, the reduction in inhibitory synaptic boutons on motor neurons resulted in a lower ratio of inhibitory-to-excitatory inputs in SBA chicks. We also found that the GABAergic interneurons degenerated via apoptosis. This study revealed that disrupted lumbar cord motor networks influenced the onset of loss of motor coordination and voluntary leg movements in chicks with SBA.
Conclusions This study illuminates the involvement of GABAergic neurons in the pathogenesis of SBA. Our results suggest that novel therapeutic approaches for the management of SBA may be found by focusing on GABAergic system regulation.

Towards understanding the Molecular Basis of Folate Non-responsive NTD in Human Population

Mayanil, Chandras[1,2], Tsurubuchi, Takao[4,1], Ichi, Shunsuke[3,1], Mohanty, Vineet[1,2], Tomita, Tadanori[1,2], McLone, David[1,2]
[1]*Ann and Robert H Lurie Children's Hostpital of Chicago*
[2]*Northwestern University Feinberg School for Medicine*
[3]*Japanese Red Cross Medical Center*
[4]*Department of Neurosurgery, Graduate School of Comprehensive Human Sciences, University of Tsukuba*

Background Maternal folate intake has reduced the incidence of human neural tube defects by 60–70%. However, 30–40% of cases remain non responsive to folate intake. The main purpose of this study was to understand the molecular mechanism of folate non-responsiveness in a mouse model of neural tube defect and test the hypothesis in NTD affected human pregnancies.
Methods Neurosphere cultures, immunostaining, quantitative RT-PCR, and enzyme linked immunosorbent assay (ELISA) methods were used in this study.
Results *Fkbp8*$^{-/-}$ derived neural stem cells showed (i) increased noggin expression; (ii) decreased Msx2 expression; (ii) premature differentiation-neurogenesis, oligodendrogenesis (Olig2 expression) and gliogenesis (GFAP expression), increased UTX expression and decreased H3K27me3 polycomb modification. Exogenous folic acid did not reverse these markers. MM-AFSCs showed increased methylation and decreased acetylation marks on histone H3 levels with a concomitant decrease in KDM6B, compared to normal controls. Anencephaly-AFSCs, showed increased methy-

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lation as well as acetylation marks. MM-amniotic fluid and serum showed high levels of BMP4, whereas Shh levels were high with anencephaly associated pregnancy.
Conclusions Folate non-responsiveness could be attributed in part to increased noggin expression in *Fkbp8-/-* embryos, resulting in decreased Msx2 expression. Folate treatment further increases Olig2 and noggin expression, thereby exacerbating ventralization. Levels of epigenetic marks, such as H3K4me, H3K27me3, H3K9Ac and H3K18A, in cultured AFSCs in combination with levels of key developmental molecules, such as BMP4 and Shh, in amniotic fluid and maternal serum, are potential biomarkers for early detection and identification of folate responsive and non-responsive NTDs in human population.

Family/Caregiver Issues

Bowel Management for Spina Bifida: An Unofficial Guide by and for Parents and Adults with Spina Bifida spinabifidabowelmanagement.com

Payne, Colleen[1,2,3,4]
[1]*Bowel Management for Spina Bifida web site*
[2]*Spina Bifida Association of Kentucky*
[3]*Spina Bifida Association of America*
[4]*Commonwealth Council on Developmental Disabilities*

Background Bowel management is one of the most important but least talked about topics in the care of SB children and adults. Often, the people who live this every day are the experts. We are not medical professionals; we are parents and adults with SB who have gone through the school of personal experience and are happy to share what we learned.
Methods More than 2,000 parents and adults with SB share experiences and knowledge on the Bowel Management for Spina Bifida Facebook page. To create the accompanying web site, I surveyed group members and compiled the best tips, products, and collective wisdom.
Results We established best practices for each age range. The group recommends a cone enema program starting at age 3, and the web site includes a thorough parent's guide to starting one. An overview of all bowel management options helps parents and adults with SB make informed decisions. Practical sections of the web site include Signs of Constipation, Clean Outs, and Bowel Bootcamps.
Conclusions Through the Bowel Management for SB Facebook Group, we learned this is an area where parents and adults with SB feel lost. As the healthcare field works toward better quality of life for people with SB, the community begs for help with bowels. This web site, based on the experiences and wisdom of people who live it every day, provides a base of knowledge as well as practical tips that empower our community to improve their lives through bowel management. Note: The web site is scheduled to be completed in the next month and has not yet been made public. Although this is not a typical research project, Lisa Raman and Pat Beierwaltes suggested healthcare professionals would be interested in the community-generated content.

General Care

Spina Bifida Health Management Plan: Parent Wallet Card

Baillie, Sharon
NSW health

Background The Spina Bifida Service (SBS) has clinics monthly at the John Hunter Children's' Hospital in the Hunter New England Local Health District.
Families with children attending the Spina Bifida clinic raised concerns in a service review about their experiences when they presented at Emergency Department (ED). Standard procedures currently in place for clients such as a clinical alert and the daily fax report of ED presentations was not successful in preventing adverse outcomes for children with Spina Bifida. SBS became aware of two serious incidences following ED presentations and including one that would require a "London protocol".
Methods SBS identified gaps in the delivery of service and recognised the risk to children and their families attending ED. The SBS identified the need for Medical staff to have Specific information and contacts at hand to consult with and Clients to be supported in providing the necessary medical information.
A wallet sized card containing specific SB information with SBS contact details was developed and trialled with families.
Results Principles of co-design were implemented in consultations with key stakeholders throughout the project.
SBS engaged in quality improvement processes and standard service agreements and the SBS card was approved for distribution. Trial consultations of the card are favourable at this time.

Conclusions The process undertaken to develop a SB card was developed as a direct response to the potential risk of adverse outcomes for SB clients when they attended ED.
Client engagement in the process of developing the SB card suggests clients will feel empowered in situations that have usually made them feel most vulnerable.
The clear reference guide, process and contacts for assistance are all likely factors to assist ED staff in fast paced ED environments.

Transition to an Advance Practice Registered Nurse Managed Pediatric Multidisciplinary Spina Bifida Program

Doton, Katherine A.
Dartmouth Hitchcock Medical Center

Background Multidisciplinary Spina Bifida Programs are at risk for extinction across the United States-victims of budget cuts, low reimbursement rates, and staffing costs, despite longstanding evidence to support this model. In 2012, Dartmouth Hitchcock Medical Center made a decision to transition from a surgeon led to an APRN managed model of care.
Methods Researchers utilized a mixed methods design-systematic review, validated Likert scale survey tool, Brio data retrieval, and salary cost comparison to assess effectiveness, patient satisfaction, and cost savings.
Results Although literature supports the unique role of the APRN in multidisciplinary complex, chronic care, data less than five years old that is specific to pediatric Spina Bifida is limited. Patient satisfaction response was positive, but was limited by low response rate. Effectiveness, defined by monthly average of unplanned readmissions, assessed utilizing a post-intervention Brio search of Spina Bifida related ICD-9 code diagnoses, was encouraging, but was limited by short duration of surveillance, and absence of pre-intervention data. A comparison of national average physician and APRN salaries indicates potential for significant cost savings.
Conclusions Due to the many variables associated with analysis of total cost, additional research is indicated. As healthcare evolves away from a traditional fee for service model, toward value-based patient and family centered care, population health, and cost efficiency, implications for transition to APRN managed complex chronic care are promising, however researchers recommend a large scale randomized controlled study to evaluate outcomes and costs of the intervention compared to a control group of pediatric patients with complex, chronic illness receiving traditional, physician led, departmentalized care.

Improving Safe Discharges in an Outpatient Multi-Disciplinary Clinic

Forte, Amy, Neises, Lori
Cincinnati Children's Hospital

Background Written discharge instructions to patients can prevent medication errors and reduce hospital readmissions. Families attending a multi-disciplinary clinic at the Center for Spina Bifida at Cincinnati Children's Hospital Medical Center expressed confusion regarding follow up plan upon receiving multiple after visit summaries (AVS) at the conclusion of their visit. The clinic's nurse coordinators identified a safety concern and proposed developing a single AVS that includes discharge instructions from each specialty provider.
Methods Upon meeting with all members of the multi-disciplinary clinic, the nurse coordinators presented the concern and proposed a method for delivering a multi-disciplinary AVS to patients upon completion of their visits. The team agreed upon a process for the nurse coordinator to obtain discharge instructions from each provider and document in a single AVS. Information provided to nurse coordinator from provider includes laboratory and imaging orders, medication changes, referrals, and return plan. The discharge plan is categorized by provider specialty on the AVS as well as contains a summary statement of the return plan.
Results All 330 patients seen since the process was implemented in January 2015 have received the AVS. Rapid cycle feedback obtained from patients and providers showed no significant barriers. The medical team reports satisfaction with implementation of the AVS and has noted improvement in adherence to treatment recommendations. Qualitatively, the AVS has improved communication of the follow up plan to families and scheduling agents. Initial feedback from families suggests increased understanding of the medical plan.
Conclusions Providing patients with a multi-disciplinary AVS is feasible and easily implemented in a busy, multi-disciplinary clinic. It also has the potential to improve communication between providers and patients and increase adherence to the follow up plan.

Using a Pediatric Passport to Effectively Collaborate Care in the Multidisciplinary Spina Bifida Clinic

Greene, Roseann[1], Wolf, Jennifer[2], Meyer, Theresa A.[2], Prodoehl, Deborah[1], Rosen, Colleen[1], Moylan, Susan[2]
[1]*Pediatric Neurosurgery, Ann & Robert H. Lurie Children's Hospital of Chicago*
[2]*Pediatric Urology, Ann & Robert H.Lurie Children's Hospital of Chicago*

Background The coordination of care for patients with Spina Bifida can be very challenging. At our institution, a multidisciplinary specialty outpatient clinic includes all the services for the Spina Bifida population, including Neurosurgery, Orthopedics, and Urology. Patient/Family surveys demonstrated opportunities for improved communication among providers and families regarding follow up appointments and scheduled testing. Previously, parents were informed by the providers when to schedule follow up. By the time parents approached the scheduler, they could not accurately recall this information. A Patient Passport tool was created to increase communication between providers, families and the scheduler.
Methods We instituted a pre-clinic chart review and created the Patient Passport Tool in January 2015. A folder including The Passport was provided to every patient at registration for their Spina Bifida clinic visit. This folder accompanied the family through clinic visits with each specialty. Each provider completed the passport for their discipline, noting the next visit and any testing that needed to be completed before the next visit. At the end of the clinic visit, the passport was given to the scheduler, who was then able to schedule follow up, medical imaging and testing before the family left the clinic. After trialing this Passport, a paper survey was given in clinic to parents to assess their response to this new tool.
Results The survey was completed by 247 families. The results of this parental survey revealed that 98% (242/247) of patients knew when next to return to clinic, with 94.6% (226/239) of those responding approving this new tool.
Conclusions In conclusion, the Patient Passport allows for enhanced communication between the providers, patient families and scheduling. This promotes timely scheduling of return visits and testing on the date of service.

Prevalence of Chronic Constipation in Infants with Spina Bifida

Huang, Gene O., Elizondo Sáenz, Rodolfo A., White, Jeffrey T., Castillo, Jonathan, Castillo, Heidi, Tu, Duong
Texas Children's Hospital/Baylor College of Medicine

Background It has been well-established in the spina bifida literature that bowel dysfunction can adversely affect quality of life. Although fecal incontinence and constipation in children and adults with spina bifida have been extensively studied, little to no data exists as to the constipation that is present during infancy. The purpose of our study was to define the prevalence of chronic constipation at 12 months of life in the spina bifida population.
Methods In this single-institution, retrospective review, we studied infants seen in our multidisciplinary spina bifida clinic between the years of 2014 and 2016. The 12 month visit was reviewed to determine if a bowel regimen was in use. Infants requiring scheduled regimens were considered to have chronic constipation. Demographic data, clinical characteristics, and gross motor quotients (GMQ), a measure of gross motor milestones for age, were reviewed.
Results Forty-two patients were identified, of which 14 (33.3%) were chronically constipated and required scheduled bowel regimens. Of the 28 without chronic constipation, 17 (61.7%) reported having a prescription for constipation, as needed. Males were more likely than females to be chronically constipated (50.0% vs 20.8%, $p = 0.047$). Infants with thoracic or lumbar level functional deficits were more likely to be chronically constipated than those with sacral deficits (47.8% vs 15.8%, $p = 0.028$). Patients with chronic constipation were noted to have lower GMQs (54.7 ± 15.8 vs 69.69 ± 24.8, $p = 0.030$).
Conclusions Our data show that chronic constipation affects a third of infants with spina bifida at 12 months of age. The prevalence of constipation in the first year of life for the general pediatric population has been reported to be 2.9%. Further study and follow-up will add to this potentially helpful information for counseling parents of children with spina bifida.

Hot Topics: A Case Presentation of a Deep Partial-thickness Burn from a Laptop Computer

Lyons, Julia
Holland Bloorview Kids Rehabilitation Hospital

Background Holland Bloorview Kids Rehabilitation Hospital provides care to nearly 400 children with spina bifida in a multidisciplinary clinic. Team members regularly monitor and provide education regarding the risks associated with insensate skin; however, much of the focus has been related to the risk of decubitus ulcers from pressure, friction and shearing. Little education had previously been delivered relating to the risk of tissue trauma from thermal sources.
Methods Two teens with thoracic myelomeningocele presented to the spina bifida clinic within three months with burns from laptop computer use. This is a case presentation on the second injury, which resulted in a deep partial-thickness burn. In this case, a teen had been unwell and spent several hours in bed on his laptop. He reports he shifted position regularly, but is unsure how long he had the laptop on his thighs before noticing the burn. Wound healing is chronicled with an overview of nursing interventions.
Results This teen sustained a deep partial-thickness burn to the thigh from a laptop computer achieved adequate burn healing after 48 days without infection. Due to insensate skin, he fortunately had no reports of pain. He was left with a flat, discoloured scar once healed. The offending laptop was returned to the manufacturer for evaluation, but was found that the heat emitted was within industry standards and was considered safe.
Conclusions As a result of these two burn injuries, clinical care has been altered to include education on tissue trauma from unsuspecting causes, particularly portable electronic devices. Children are discouraged from using laptops on their laps. Members of the multidisciplinary team ask children and parents *how* electronic devices are being used and encourage them to protect insensate skin through positioning and the use of insulating material. No further burns have been reported to clinic staff since these initial two cases.

A Review of the Multidisciplinary Care Model for Patients with Spina Bifida (SB) or Cystic Fibrosis (CF)

Ouyang, Lijing[1], Valdez, Rodolfo[1], Thibadeau, Judy[1], Ashby, Jasmine[2]
[1]*CDC*
[2]*Ohio State University*

Background The coordinated care provided by practitioners of various health disciplines to a patient is known as the multidisciplinary care model (MDCM). It has been recommended for the care of patients with SB or CF. We reviewed literature on MDCM and other care delivery models for patients with SB and CF and the associations of these models with health outcomes, care cost, and care organization.
Methods The search terms in this review included Spina Bifida, Cystic Fibrosis, multidisciplinary care model and pediatric care. We searched for articles published after 2006 and key sentinel articles published before that year. All literature was retrieved from Google Scholar and PubMed.
Results A total of 24 articles were synthesized in this review (13 articles on CF and 11 articles on SB). The most common form of care for patients with CF or SB was the multidisciplinary clinic. SB studies mostly focused on single specialty clinics while CF studies included comparisons among clinics in the care network. Studies on SB clinics show that MDCM was associated with better health outcomes and higher patient satisfaction. Studies on CF clinics suggest that quality of patient care and better outcomes were positively associated with adherence to CF care guidelines regardless the care model used. Various care models such as the hybrid model can have comparable or better health outcomes than MDCM. We found no information on the cost of MDCM relative to other health care models.
Conclusions A majority of the literature retrieved indicates that MDCM brings health benefits to patients with SB or CF. More studies are needed on the cost and effectiveness of this model when addressing the needs and constraints faced by both the patients and the health care systems.

Arm Span vs Segmental Height to Assess BMI in Latinos with Spina Bifida

Smith, Kathryn[1,2], Van Speybroeck, Alexander[1,2], Macias, Kristy[1], To, Hao Quoc[1], Pubols Neighbors, Cindy[3]
[1]*Children's Hospital Los Angeles*
[2]*Department of Pediatrics, Keck School of Medicine, University of Southern California*
[3]*Uniform Services University*

Background Obesity rates for individuals with spina bifida have been reported ranging from 28–50% among children and 34–64% among adolescents and adults. The purpose of this study was to prospectively evalu-

ate differences in calculated BMI using arm span vs. height/linear length in a cohort of patients from a primarily Hispanic cohort ages 2–18.
Methods Weight was measured using a standing, wheelchair or infant scale. Height/length was measured using a stadiometer or a tape measure. Arm span was measured using a tape measure affixed to a length of PCV pipe, with a level attached to assure that it was maintained in the horizontal position. Neural tube defects were categorized as Lesion Level 1- LL1 (high lumbar motor level 1–3 and thoracic), Lesion Level 2- LL2 (low lumbar motor level 4–5) or Lesion Level 3- LL3 (sacral level). Within and between group differences were compared for BMI, level of defect, race, age and method of height/length and arm span measurement.
Results A total of 206 patients participated, 52% male; 82% were Latino. There were 28% with LL1, 38% with LL2 and 34% with LL3. In general, the most significant differences tended to occur in the older age groups 10–17 and the higher lesions levels. LL1 males and females ages 10–17 showed significant variance that persisted when stratifying by Hispanic ethnicity. For the LL3 significant differences were found in males in all age groups and females ages 1–9; these differences persisted when stratifying by Hispanic ethnicity as well.
Conclusions This study provides important evidence of the challenges associated with ascertaining BMI given the variability in relationships between the use of arm span and measured height/length in calculating BMI.

Design and Methodology for Updating the Spina Bifida Association (SBA) Guidelines for Spina Bifida Health Care Services Throughout the Lifespan

Herron, Adrienne R.[1,2], Brei, Timothy[4,5], Beierwaltes, Patricia[3], Chelliah, Jerome B.[5], Dosa, Nienke[6], Ramen, Lisa[5], Struwe, Sara[5], Thibadeau, Judy[2], Dicianno, Brad E.[7]
[1]*Oak Ridge Institute for Science and Education*
[2]*National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention*
[3]*Minnesota State University-Mankato*
[4]*Seattle Children's Hospital*
[5]*Spina Bifida Association*
[6]*SUNY Upstate Medical University*
[7]*University of Pittsburgh Medical Center*

Background To describe the process underway for updating and expanding the *Guidelines for Spina Bifida Health Care Services Throughout the Lifespan* 3rd ed. published by SBA in 2006.
Methods In 1990, the SBA via its Professional Advisory Council published the original *Guidelines for Spina Bifida Health Care Services throughout the Lifespan*; these guidelines have be updated twice (1995 and 2006). In 2014, SBA and CDC convened a steering committee to assess the need for updating the guidelines. The 2006 guidelines were reviewed, and the committee concluded that an update and expansion was necessary. The 4th edition of the guidelines will address 26 topic areas, some of which were not covered in the 2006 guidelines: Transition; Self-Management; Mental Health; Quality of Life; Men's Health; Respiratory & Sleep; Nutrition; Physical Activity; Preventative Health Care; Secondary Conditions; Endocrine; and Epidemiology.
The proposed guidelines will be developed based on review of current evidence and clinical consensus in topic specific workgroups. The workgroups will use Single Text Procedure and Nominal Group Technique, review the current guidelines, develop clinical questions, identify gaps in research, and generate suggested updates. Vetting of the suggested guidelines by the spina bifida community will occur at the 2017 Spina Bifida World Congress.
Results The 4th edition of the guidelines will be the next step in implementing evidence based practices that contribute to improved care for people living with spina bifida.
Conclusions The use of multi-disciplinary groups along with the methodology by which the guidelines will be established will provide the spina bifida community with reliable and accessible guidelines that offer the most current and relevant evidence and consensus based approaches to optimize outcomes and improve care for all people living with spina bifida.

Genetics/Epidemiology

Increase in Hospitalizations of Children with Spina Bifida in U.S. Hospitals

Briggs, Virginia
MCPHS University

Background Spina Bifida (SB) is a common birth defect that involves incomplete closure of the spinal canal before birth, resulting in permanent physical ability of-

ten accompanied by developmental delays. Health care needs are concentrated in the areas of urology, orthopedics and neurosurgery, and usually last a lifetime. Previous research has shown an increase in hospitalizations of medically complex children, despite decreases in incidence rates. The goal of this study was to examine the hospitalization rates of children with spina bifida, specifically.

Methods Rates were calculated using the Healthcare and Utilization Project's Nationwide Inpatient Sample (NIS), a national discharge database. From 2000 to 2008, patients with SB were identified using ICD-9 diagnosis codes. National hospitalization rates for SB patients were calculated per every 100,000 using weighted estimates from the NIS and U.S. Census estimates for each year. Trends in rates were compared between four age groups; $<$ 5 years, 5–9 years. 10–14 years and 15–19 years.

Results A significant increase in hospitalizations in children 0 to 5 years was found (B $= 0.28$; $p < 0.05$) between 2000 and 2008. Rates ranged from 63.5 per 100,000 in 2000 to 89.1 per 100,000 in 2008. While overall hospitalization rates were significantly lower in the older groups of children (5–9, 10–14 and 15–19 years) compared to the younger group (0–5 years), there was no significant change over time in any of the older groups.

Conclusions Advances in medicine have resulted in longer life-expectancy and thus increased medical management of individuals with spina bifida, particularly in the early childhood years. As a result, the prevalence of children with spina bifida may be increasing in U.S. hospitals. Additional research is needed to determine what types of health care services are being received and if the services are best serving this unique population.

Current Estimates of the U.S. Spina Bifida Population in the United States

Briggs, Virginia
MCPHS University

Background Spina bifida is a birth defect resulting in deficits of neurological functions. Individuals diagnosed with spina bifida often require a lifetime of medical care to manage this condition. Currently, the number of people living with spina bifida in the United States is unknown. The purpose of this study is to provide estimates of the population size, and the distribution by gender and U.S. region.

Methods Population estimates from the National Center of Health Statistics, United States Division of Vital Statistics from 1960 to 2015 and spina bifida birth rates reported by the Centers of Disease Control were used to calculate the number of spina bifida births over a 55 year period. Mortality rates for those affected by spina bifida were used to estimate the number of deaths, and thus the number of spina bifida cases $\leqslant$ 55 years old, estimated to be alive in 2015.

Results In 2015, the estimated number of individuals living with spina bifida $\leqslant$ 55 years of age in the U.S. was 48,372 (18,467 male; 20,005 female). Of the nine regions of the United States analyzed, the South Atlantic and Pacific regions had the highest proportions of spina bifida cases (19.7% and 16.3%, respectively).

Conclusions Understanding the approximate size and distribution by age, gender and U.S region of residence may assist health care providers in planning services for this changing population.

Trends in Multivitamin Intake Among Women of Reproductive Age – United States, 2006–2015

Wong, Eugene, Rose, Charles E., Flores, Alina L., Yeung, Lorraine F.
US Centers for Disease Control and Prevention

Background The United States (U.S.) Public Health Service recommended that all women of childbearing age consume 400 μg of folic acid daily in addition to a varied diet to reduce the risk of having a pregnancy affected by a neural tube defect (NTD). Women can ensure they are consuming the recommended amount by taking a multivitamin (MV), containing 400 μg of folic acid, daily.

Methods To analyze trends in daily MV intake among women aged 18–44 years, we used data from the Porter Novelli Health Styles surveys of adults in the U.S. (n $=$ 8547). We assessed the change in prevalence of MV use in 2006–2015 by age (18–24, 25–34, 35–44 years), race/ethnicity (White, non-Hispanic (NH); Black, NH; Hispanic; other), and in 2006–2012 by pregnancy status (pregnant, trying to get pregnant, non-pregnant) using log-binomial regression to generate prevalence ratios (PR).

Results Daily MV consumption decreased from 33.5% to 26.3% over the years 2006–2015 for women aged 18–44 years (PR $= 0.72$, $p < 0.001$). This decrease was similar ($\chi^2 = 2.95$, $p = 0.23$) across all age groups. Decreases in daily MV intake were found among White-NHs (n $= 5509$, PR $= 0.75$, $p < 0.001$),

Hispanics (n = 973, PR = 0.39, $p < 0.001$), and other (n = 915, PR = 0.58, $p < 0.001$). Black-NHs (n = 1150) had a stable daily MV intake (25.4% vs. 27.4% 2015, $p = 0.28$). Prevalence of daily MV intake varied for pregnant women, 52.8% in 2006 and 71.0% in 2012 (n = 185, $p = 0.21$), and remained fairly stable for women trying to get pregnant, 40.2% in 2006 and 38.3% in 2012 (n = 477, $p = 0.19$). For non-pregnant women, the prevalence of daily MV intake decreased over the same period, 31.3% in 2006 and 21.3% in 2012 (n = 5594, PR = 0.59, $p < 0.001$).

Conclusions Generally, daily MV intake in women decreased over the past decade. Development of innovative, engaging messaging and targeted public health interventions for increasing folic acid intake is needed to reduce NTD risk.

Neurosurgery/Neuroscience

Neural Tube Defects in Costa Rica and Folate Fortification: Impact of 20 Years of a Public Health Strategy on Neurosurgical Outcomes

Caceres, Adrian[1,2], Segura, Juan Luis[1,2], Zamora, Justiniano[1,2], Jimenez, Edgar[1,2], Trejos, Humberto[1,2]
[1]*National Childrens Hospital of Costa Rica*
[2]*University of Costa Rica*

Background Folate supplementation in the prevention of Neural Tube Defects (NTD) has been widely recognized as an effective preventive strategy in reducing the rate of spina bifida however there has been little description of the impact in the severity of the malformations in those born after this policy has been established.

Methods A retrospective review of patient records (1991–2016) recollected the pre and post fortification anatomical, functional level, defect size and the presence of associated kyphotic deformity were recorded as well as the presence of symptomatic Chiari malformation as manifested by respiratory stridor, swallowing anomalies and neurovegetative disfunction as a manifestation of tonsillary herniation and other craniovertebral junction anomalies.

Results A decrease in the incidence of NTD from 0.8 to 0.4 per 1000 live births has been stable for the past 20 years, however those patients still born and affected with NTD had evidence of a decrease in the severity of the primary neurulation morphogenetic anomalies as evidenced in a three dimentional axis by a caudalization of the defects, a reduction of their diameter and an almost total disappearance of kyphotic deformities. Concurrent with these lesser impact in the lumbosacral spine region, we also noted a decrease on the number of patients who presented with a symptomatic Chiari malformation at birth as well as the number of patients who eventually required a VP shunt following closure of their NTD.

Conclusions The impact of folate supplementation through a government mandatory policy not only decreases the number of NTD incidence but also has an favorable impact in the morphological deformities both at the primary site of the NTD and as well on the associated anomalies in the hindbrain with a lesser degree of symptomatic Chiari malformations and reduced need to treat hydrocephalus by VP shunting.

Prenatal Repair of Myelomeningocele: A Model for Neurodevelopmental Follow-up

Castillo, Jonathan[1], Castillo, Heidi[1], Ostermaier, Kathryn[1], Whitehead, William[2], Olutoye, Oluyinka[3], Cass, Darrell[3], Tu, Duong[4], Huang, Gene O.[4], Fremion, Ellen[1], Sanz Cortes, Magdalena[5], Belfort, Michael[5], Voigt, Robert[1]
[1]*Pediatrics-Texas Children's Hospital*
[2]*Neurosurgery-Texas Children's Hospital*
[3]*Fetal Surgery-Texas Children's Hospital*
[4]*Urology-Texas Children's Hosptial*
[5]*Obstetrics and Gynecology-Texas Children's Hospital*

Background Following the Management of Myelomeningocele Study (MOMS) a number of centers are able to offer in-utero closure; concurrently, a call has gone out for greater standardization of services. The purpose of our study was to explore the nature of multidisciplinary myelomeningocele (MMC) care after prenatal repair and describe a comprehensive approach to medical and neurodevelopmental follow-up.

Methods We report our experience with in-utero repair. The Capute Scales, consisting of the Cognitive Adaptive Test (CAT) and Clinical Linguistic & Auditory Milestone Scale (CLAMS) were performed as a neurodevelopmental outcome measure at 12, 24, and 36 months of age.

Results Developmental pediatrics provides prenatal counseling, as well as NICU and outpatient care coordination. Of 444 patients with spina bifida followed in our multidisciplinary clinic, 21 (mean age = 21 months; range 4–46) had undergone in-utero repair at our center. Of these, 5 (23.8%) were repaired fetoscopically. Eleven (52.4%) were male, 90.5% were

White, and 9.5% were Black; of these patients four (19%) were also Hispanic. The mean age at delivery was 35.25 wks (SD 2.8). Seven (33.3%) had undergone endoscopic third ventriculostomy (ETV), 2 (9.5%) had a shunt, and none have undergone an untethering procedure. Ten patients have had their 12 month visit; mean CAT/CLAMS scores were 93.8 (SD 12.0) and 91.1 (SD 19.6) respectively. Eight patients have had their 24 month visit with mean CAT/CLAMS scores of 83.5 (SD 11.1) and 90.6 (SD 13.4) respectively. Only 2 patients have had their 36 month visit with mean CAT/CLAMS scores of 70.5 (SD 27.6) and 88 (SD 14.1) respectively.

Conclusions Along with low shunt rates, our data indicate that a cohesive MMC care team of specialists provides a structure to support needed neurodevelopmental monitoring. We offer our experience and early outcomes in the hope of contributing to the advancement of optimal practice criteria.

Endoscopic Third Ventriculostomy Training to Treat Spina Bifida Related Hydrocephalus in Africa: A Literature Review

Burckart, Caryssa[2], Castillo, Heidi[1], Castillo, Jonathan[1]
[1]*Texas Children's Hospital*
[2]*Liberty University*

Background In Africa, endoscopic third ventriculostomy (ETV) has been identified as a promising alternative to shunt surgery, requiring minimal follow-up and a low rate of complications. However, there is a lack of professionals trained in neuroendoscopic management of hydrocephalus. The purpose of this study was to explore educational efforts to upscale the use of ETV in Africa.

Methods A PubMed ®search was performed for articles on neurosurgical management in patients with spina bifida. Articles were identified by searching the terms "ETV", "Africa", and "hydrocephalus." Articles were excluded if they did not refer to hydrocephalus among individuals living with spina bifida. Articles meeting inclusion criteria were classified according to management method.

Results A total of 43 articles were identified and 27 met inclusion criteria. Of these, only three (11.1%) described efforts to train local medical professionals, including surgeons and nurses on performing ETVs. Two educational programs were conducted across nine African nations (i.e. Ethiopia, Kenya, Nigeria, Rwanda, Sudan, Tanzania, Uganda, Zambia, and Zimbabwe). The programs encompassed training on techniques utilizing ETV with and without Choroid Plexus Cauterization for hydrocephalus associated conditions including myelomeningocele (14%), post-infectious hydrocephalus (60%), and other (26%). Training regarding the need for long term follow-up programs were also addressed.

Conclusions It has been shown that ETV has advantages over shunt placement for the management of hydrocephalus in low income settings and that efforts to train local surgeons in its use have been successful. However, few academic centers have developed educational programs along with their outreach efforts in the region. Thus, capacity building efforts must be enlarged in order to meet the growing needs of individuals with hydrocephalus in Africa.

Long-term Follow Up of 136 Patients with Myelomeningocele, Predictors of Good Prognosis

Collange, Nelci Z.[1,2], Coelho, Giselle[1,4], Grecco, Luanda A.[1,3]
[1]*CENEPE, Pediatric Neurosurgical Center*
[2]*UNIFESP-Universidade federal de São Paulo*
[3]*Cenepe Reabilitação*
[4]*Hospital Santa Marcelina-Itaquera*

Background In the world, the neural tube defect. (NTD) are detected in 300.000 new cases a year. After 2004, Brazilian law became mandatory fortification of corn and wheat flour with folic acid (0,15 mg folic acid/100 g) and iron. The lack of antenatal diagnosis of the myelomeningocele (MMC) drives the patients be delivered in a non-specialized centers. The aim is to analyse retrospectively the results of 136 MMC patients and predictors of good prognosis.

Methods Retrospective study of a convenience sample of cases followed in a general hospital, pediatric neurosurgery out patients clinic. Complications, associated malformations, other surgeries and gait were analyzed. Pearson's Chi Square test was used to analyze the association between categorical variables.

Results The level of MMC was significantly associated with ambulation, (X2 = 21.9, V = 0:40, p = 0.001) and scoliosis (X2 = 17.2, V = 0:36 p = 0.001), club foot deformity (X2 = 10.2, V = 0 27, p = 0.0016). Although not associated incidence of hydrocephalus and Chiari malformation, the level of the lesion showed a significant association with symptomatic Chiari malformation (X2 = 9.3, V = 0:26, p =

0.02) and the VP shunt revision (X2 = 9.4, V = 0:26, $p = 0.002$).The incidence of infectious in the neonatal period was associated with the transfer between hospitals before surgery (X2 = 7.1, V0.23, $p = 0.029$) and death (X2 = 8.7, V = 0:25 $p = 0.013$).

Conclusions The thoracic level was associates with more shunt revision and symptomatic Chiari malformation. The predictive factor associated to infection was transfer between hospitals after birth and before surgery and this is the only preventable cause of infection detected by this retrospective analysis.

Cryopreserved Human Umbilical Cord (HUC) vs. Acellular Dermal Matrix (ADM) for In-Utero Spina Bifida Repair

Mann, Lovepreet K.[1], Won, Jong H.[1], Snowise, Saul[1], Onanian, Marisa T.[2], Tseng, Scheffer C.[3], Grill, Raymond[4], Papanna, Ramesha[1]
[1]*University of Texas McGovern Medical School*
[2]*Columbia University*
[3]*Ocular Surface Center*
[4]*University of Mississippi*

Background To investigate the acute inflammatory response and regenerative ingrowth of native cells at repair site after HUC vs. ADM sutured over spina bifida lesions (SB) in a retinoic acid (RA) rat model.

Methods Pregnant time-dated Sprague-Dawley rats were gavaged with RA (40 mg/kg) on gestational day 10 (GD10) to induce SB in fetuses. Laparotomy and hysterotomy were performed on GD20 and HUC or ADM were sutured over the SB. After 48–52 hours, fetuses were harvested and fixed in 4% paraformaldehyde. Trichrome staining was used to assess cellular migration within the patches. Immunofluorescence was then performed to assess inflammatory response using myeloperoxidase (MPO:neutrophils). To evaluate regenerative ingrowth of cells, native cell markers including cytokeratin (Cyt 5/6: epidermal), glial fibrillary acidic protein (GFAP: astrocytes) and chondroitin sulfate-56 (CS-56: meningeal layer) were used. TUNEL (Apoptosis) staining were also performed.

Results Fetal survival with intact patches was equal: HUC 8/12 vs ADM 7/12, ($p = 0.5$). Neutrophils were lower in the HUC compared to ADM. Epidermal cells were present on the outer surface in both groups, however, the cells were positive on the inner surface of the patch in ADM (5/7) compared to none in HUC ($p = 0.03$). Meningeal cells were positive in the inner surface of both patches, whereas, these cells were present on the outer surfaces of 5/7 ADM compared to none in HUC ($p = 0.03$). Astrocyte cells were attached to the inner surface of 6/7 ADM compared to only 2/8 HUC; p =0.01. These findings suggest disorganized cell growth in ADM. TUNEL positive cells were lower in the HUC compared to ADM ($p = 0.03$).

Conclusions Cryopreserved human umbilical cord promotes lower acute inflammatory response, better organized cellular migration and lower cell death as compared to acellular dermal matrix at the in-utero spina bifida repair site.

Conventional vs Cryopreserved Human Umbilical Cord Patch Based Repair for In-utero Spina Bifida In A Sheep Model

Papanna, Ramesha[1], Mann, Lovepreet K.[1], Won, Jong H.[1], Morales, Yisel[1], Fletcher, Stephen[1], Jain, Ranu[1], Lopez, Suzanne[1], Moise Jr., Kenneth J.[1], Grill, Raymond[2]
[1]*University of Texas McGovern Medical School*
[2]*University of Mississippi*

Background The objective of this study was to evaluate the efficacy of in-utero repair of spina bifida using single layer HUC patch compared to conventional repair (CR) in order to decrease scar formation and improve offspring spinal cord function in a sheep model.

Methods On gestational day (GD) 75, spina bifida without myelotomy (remove dura matter only) was created in the seven timed pregnant ewe (11 fetuses). Repeat hysterotomy was performed on GD 95 and the surviving fetuses were randomly assigned to CR or HUC-suture repair group. The ewes were delivered at GD 140 by C-section and neurological assessments of the newborns were performed on day 2 of life using Texas Spinal Cord Injury Scale (TSCIS) to assess the gait, proprioception and nociception (Range: 0–20). MRI with digital tensor imaging (DTI) of the spinal cord was performed to quantify the disruption of spinal tracts.

Results There were 7/11 (64%) fetuses with spina bifida that survived to the repair stage; of which 3 underwent CR and 4 were repaired with HUC; all seven survived to delivery. The combined hind limbs TSCIS scores for the control lambs were 20 ± 0, HUC-repair were 16 ± 7 and CR were 12.7 ± 4 (Control vs. CR $p = 0.03$; CR vs. HUC $p = 0.5$ and Control vs. HUC $p = 0.4$). CR repair had predominant loss of proprioception and mild to moderate ataxia. HUC repaired lambs had normal proprioception and mild ataxia in the

hind limbs. DTI showed disruption of dorsal column in the spinal cord in 3/3 CR (3/3) compared to 1/4 HUC repair ($p = 0.1$). Histology showed thickened arachnoid layer with activation of myofibroblast in CR 3/3 compared to 0/4 in HUC ($p = 0.03$) suggesting scar formation in meningeal layers.
Conclusions In our spina bifida sheep model, HUC preserved the integrity of meningeal layers after in-utero repair as compared to CR repair, with improvements in the spinal cord function.

Analysis of Footpath Temporal Variability During Gait of a Low-lumbar Level Myelomeningocele Patient

Rodrigo, Silvia E.[2], Lescano, Claudia N.[1,2]
[1]*National Council of Scientific and Technical Research Council (CONICET)*
[2]*National University of San Juan*

Background Considering the variability of gait patterns seen in Myelomeningocele (MMC) patients and based on the term motor synergy – an organization of variables from the central nervous system for stabilizing a task-specific parameter, such as footpath during gait – the behavior of this parameter for a low-lumbar level MMC patient is analyzed. The aim is to explore the applicability of the footpath as control variable of a robotic orthosis for gait rehabilitation of these patients.
Methods An adult female MMC patient participated in our research, previous signed consent approved by Ethics Committee of FLENI (Argentina). The patient is within Group 2 set up by Gutierrez (2003) from the MMT (0–5 scale), showing the ankle plantar-flexor muscles 0 and 3–4 MMT values for right and left legs, respectively. During gait data registering she walked at a self-selected speed without aids. From independent gait trials the anteroposterior (AP) and vertical (V) components of the right malleolus (footpath) trajectory at sagittal plane were obtained and then analyzed their intra-variabilities through a linear rescaling of the footpath time axis.
Results The footpath (average $\pm$ SD) of MMC patient showed an increased variability for the swing phase of the gait. Greater AP component variability as regards the V one was also seen at late-stance phase.
Conclusions The greater footpath AP variability seen during MMC patient's gait could be linked to a limitation of ankle plantar-flexor muscles to provide body COM forward propulsion at late-stance phase (Winter, 2009). As well, the increased variability at swing phase could relate to a distinct combination of muscle activation patterns, as is seen in spinal injury patients (Ivanenko et al., 2003). In future works we propose to deeper explore these issues, in the search for clues to design a suitable control scheme of a robotic orthosis for gait rehabilitation of MMC patients.

Improvement of Gait and Bowel/Bladder Function after Minimally Invasive Lateral Spinal Shortening for Recurrent Tethered Cord Syndrome Secondary to Spina Bifida

Wali, Arvin R., Steinberg, Jeffery, Martin, Joel, Gonda, David, Taylor, William
University of California, San Diego

Background Recurrent tethered cord syndrome (TCS) is a complication associated with pediatric spina bifida. Spine shortening via vertebral osteotomy (SSVO) is a novel approach for treatment of recurrent TCS. Current reports of this technique describe a posterior spinal shortening approach, performing a corpectomy, laminectomy, facetectomy, and pedicle resection. We report the first patient treated via a minimally invasive lateral approach for SSVO in recurrent TCS, with improved symptomatology at 3 and 6 month follow up.
Methods A 22 year-old female presented with progressive lower extremity weakness, bowel and bladder incontinence, and back pain secondary to recurrent TCS. Her history included spina bifida, two prior childhood repairs of myelomeningocele and two prior detethering procedures. Minimally invasive lateral SSVO was offered to the patient.
Results Using a minimally invasive, lateral retropleural approach with partial corpectomy of the T12 vertebral body, 18 mm of spinal shortening was achieved. Post-operatively; the patient remained at her neurologic baseline. At 3 and 6 month follow up, the patient demonstrated reduction in lower extremity radicular pain and improved control in bowel and bladder function. At 6 month follow up, posterior-anterior and lateral thoracic-lumbar xrays demonstrated intact instrumentation without complication and partial fusion formation.
Conclusions This case report represents the first minimally invasive lateral approach for SSVO for recurrent TCS. Improved neurologic outcome at 3 and 6 month is reported, attributed to spinal cord tension reduction. Additional studies are warranted to further assess this approach for recurrent TCS.

Nutrition/Bone Health

BMI Evaluation in Spina Bifida During Development

Clayton, Gerald H.[1,2], Blanchard, Alesia[1,2], Watne, Laura[1], Campbell, Kristen[2], Pyle, Laura[2], O'Keeffe, Shannon[1], Wilson, Pamela[1,2]
[1]*Children's Hospital Colorado*
[2]*University of Colorado, School of Medicine*

Background Pediatric obesity is very prevalent and can have important functional and health consequences especially in disabled youth. However, little accurate data is available to understand the patterns of obesity for disabled children or to make comparisons with their neurologically intact peers. Understanding these patterns would set the precedent for establishing anticipatory guidance standards for clinics working with the pediatric Spina Bifida (SB) population.
Methods We retrospectively analyzed patients diagnosed with Spina Bifida (N = 272; 455 visits) enrolled in the CDC National Spina Bifida Registry ages 3 months to 33 years. BMI Z-scores were calculated and used to create growth curves for this population using the Cole LMS method. The impact of spinal lesion level, ambulation status and gender on the BMI/age relationship were evaluated using a multivariable random intercept and slope model and spaghetti plots. Initial explorations of these relationships in a large multi-site dataset are explored using similar methods.
Results Prior to age 12 there is little association between BMI and age (R2 = 0.0016) however this changes 24-fold in later years with increased mean BMI (R2 = 0.0381; $p = 0.0001$) and variability (SD = 3.827/6.044). There is a significant association overall between age and BMI z-score ($p = 0.0496$) but no association between z-score and lesion level ($p = 0.2504$), gender ($p = 0.0812$) nor ambulation status ($p = 0.1320$) and BMI z-score.
Conclusions This SB population is significantly overweight as compared to a cohort of normally developing peers. No developmental difference in gender is noted in the mean BMI's, until after puberty. Surprisingly, lesion level and ambulation status have little impact on BMI/age relationships. There may however be more variability at higher lesion levels. This work provides preliminary information for development of SB anticipatory growth guidelines. Limitations to these results include limited sample size.

A Community Based Accessible Gardening Campaign to Promote Folic Acid Awareness

Davis, Kristen, Boyer, Jill, Lindenmayer, Barbara, Whitney, Christine, Dosa, Nienke
SUNY Upstate Medical University

Background A folate rich diet helps children with spina bifida and their families to manage chronic constipation, reduce recurrence risk in pregnancies, and prevent obesity. Gardening is a lifetime fitness activity that has been shown to increase children's knowledge about food and willingness to eat fruit and vegetables.
Methods Patients/families at the Spina Bifida Center of Central New York were invited to participate during clinic encounters and via our quarterly newsletter and Facebook page. A picnic was scheduled in late Spring. This get together included brief presentations by our clinic team. A second "rain date" picnic allowed for planting of a late season crop of folate rich foods. Each family received a raised gardening table; adaptive gardening tools, information about the Spina Bifida Association of America's Folic Awareness Campaign, seed packets for folate-rich vegetables, and recipes for high fiber and folate-rich meals. Families identified spaces in their community for placement of the raised gardening beds. Families shared photos of their gardens via Facebook and our clinic newsletter. These photos and information about our folic acid awareness campaign were also shared with Community Supported Agriculture coalitions and at our local Farmer's Market.
Results Participants were young families recruited via the clinic newsletter (7), clinic visits (2) and our Facebook page (1). Families willingly shared photos of the gardens on social media. As an outgrowth of this campaign we discovered a community garden coalition that is interested in making neighborhood gardens accessible to people with disabilities.
Conclusions Collaboration between spina bifida centers and community garden coalitions is a strategy for sustaining the folate awareness program in an inclusive setting, thereby promoting social participation and benefitting diverse populations.

Orthopedics

Functional Status of Lower Limbs in Patients with Orthopedic Manifestations of Myelomeningocele

Cheverda, Andriy[1], Guk, Yuriy[1], Zyma, Andrii[1], Martsyniak, Stepan[1], Kinchaya, Tamara[1], Stewart, David[2], Demyan, Yuriy[1], Oliinuk, Yurii[1]
[1]*State institution "Institute of traumatology and orthopaedy of National academy of medical sciences of Ukraine"*
[2]*Children's Bone and Spine Surgery*

Background Myelomeningocele is an important cause of static-locomotor disorders of children. Injuries of varying degrees of paralysis or paresis of the lower limbs, dysfunction of the pelvic organs, limbs and spine deformation, which significantly affects the quality of life and ability to movement and maintenance depends on the level of spinal cord lesion.
Methods Material of the study were 81 patients in aged from 5 to 21 years (average age was 10 years and 8 months). To determine the level of spinal cord lesion we used Sharrard's (1964) as modified Bartonek et al. (1999) classification. According to the Hoffer et al. (1973) classification of ability to movement was evaluated.
Results All patients included in the study are distributed according to the level of functional muscular strength by five clinical groups with a certain of level of functional muscular strength by Sharrard's(1964) classification with Bartonek modification (1999) and estimated the actual degree of movement of patients by Hoffer classification.
Comparing the actual degree of movement in groups of patients to the theoretically expected degree of movement of patients was conducted. It is shown that children with the same level of functional insufficiency of the lower limbs do not always achieve the expected degree of movement, and depending on the impact of confounding factors such as intellectual (mental) level of the child, the progression of hydrocephalus, obesity, severe orthopedic pathology, presence of spasticity and others can reach a higher or worse degree of movement.
Conclusions The level of the spinal cord lesion that determines the level of muscle function of the lower limbs is an important factor in the ability of patients to movement with the effects of myelomeningocele. Patients with the same level of loss of the muscle function does not always achieve the expected of the level of movement.

Other

The BCH-SBPR: A Multi-Disciplinary Registry Collecting Longitudinal Data on Patients with Spina Bifida

Alkhawaldeh, Mohammad Y.[1,2]
[1]*Boston Children's Hospital*
[2]*University of Massachusettst*

Background Spina Bifida (SB), which means "cleft spine," refers to the incomplete development of the brain, spinal cord, and/or meninges. In the U.S. alone, approximately 1500 infants are born with SB each year. An estimated 166,000 individuals with SB live in the United States. The BCH-SBPR was established in August 2015 to help increase knowledge about new procedures, surgeries and treatment options, growing up with Spina Bifida, and to guide healthcare practices by prospectively studying a cohort of children born with this condition.
Methods Study subjects include children whose ages range from birth to 35 years with one of the following six SB diagnoses: Myelomeningocele, Meningocele, Lipoma of Spinal Cord, Fatty Thickened Filum, Split Cord Malformation, and Terminal Myelocystocele will be eligible to participate.
Results Six hundred and thirty-three patients were enrolled in this registry. Six hundred and thirty-three patients were enrolled in this registry. Sixty-six patients were randomly selected to conduct a descriptive analysis. The mean age was 9.7 (7.63 SD). The majority of the patients' age were between 5 and 13 years old (40 %). The female participants (58.3 %) were slightly more than male participants (41.7%). The majority was white (56.7%) and None-Hispanic or Latino. 40% of the patient relaid on public insurance. The majority had lumbar lesions (61.90%), 26.9% had tethered cord, and 7.93% had sacral lesions, and 3.17 had thoracic lesions. This is an ongoing research project therefore, further analysis of descriptive and inferential statistics will be conducted early December 2016.
Conclusions This Registry will provide valuable longitudinal clinical data from approximately 700 patients with SB conditions. It will include subjective and objective data about innovative procedures, surgeries and treatments that are only performed in few clinics across the United States. The collected data will be used for quality improvement and research projects.

Testing VO_{2peak} in Wheelchair-using Youth with Spina Bifida: Arm Cranking or Wheelchair Propulsion?

Bloemen, Manon[1,2], de Groot, Janke[1,2], Backx, Frank[2], Benner, Joyce[1], Westerveld, Rosalyn[1], Takken, Tim[2]
[1]*HU University of Applied Sciences Utrecht*
[2]*University Medical Center Utrecht*

Background Arm cranking is mostly used as the gold standard to measure VO_{2peak} in wheelchair-using people, however specificity is lacking. A lab test using wheelchair propulsion, for example the graded wheelchair propulsion test (GWPT), or a field test like the Shuttle Ride Test (SRiT) seems more adequate. There are no clinimetric properties available for wheelchair-using youth with SB, so the aim of this study is to analyze the validity and reliability of the GWPT and SRiT in wheelchair-using youth with SB.
Methods Fifty-three wheelchair-using children with SB (13.6 y, $\pm$ 3.9) participated. An incremental arm cranking protocol, the GWPT and the SRiT were assessed for validity. For reliability, the GWPT and SRiT were assessed twice. A mobile gas analyses system was used for cardiorespiratory responses. The clinical outcome measure of the SRiT is the number of shuttles. Validity was analyzed by paired t-tests. We also tried to predict VO_{2peak} by the number of shuttles. Reliability was analyzed by the $ICC_{agreement}$ (ICC 2.1.A), the Smallest Detectable Change (SDC) and the coefficients of variations (CVs).
Results VO_{2peak} and HR_{peak} were higher during the GWPT compared to the arm cranking protocol. We found no significant differences in VO_{2peak} and HR_{peak} between the GWPT and SRiT. VO_{2peak} could not be predicted adequately using the number of shuttles, as the individual prediction intervals varied widely. The ICCs were excellent for the GWPT for both the VO_{2peak} and HR_{peak} (ICC $>$ 0.90) with a questionable SDC (VO2piek 5.18 ml/kg/min). The ICCs were good to excellent for the SRiT for the number of achieved shuttles, VO_{2peak} and HR_{peak} (ICC 0.77–0.98). The SDC was small for the shuttles (1.5) and CVs varied from 6.2% and 6.4% for absolute and relative VO_{2peak} respectively.
Conclusions Wheelchair propulsion, with the use of a mobile gas analyses system, has the preference to measure VO_{2peak} in wheelchair-using youth with SB.

A Quality Improvement Nursing Intervention for Neurogenic Bowel Management in the Texas Children's Spina Bifida Clinic

Bolin, Rhonda G., Morrison Jacobus, Melissa, Perez, Ana
Texas Children's Hospital Clinical Care Center

Background The Texas Children's Hospital Spina Bifida Clinic sees over 400 patients for routine visits. Starting with prenatal visits, parents are informed of issues regarding incontinence, constipation and the eventual need for a bowel program. Physicians query families over time regarding readiness for bowel control, and then recommend an individualized bowel plan with the family. The spina bifida nurses intervene and provide individualized bowel management training and education. Despite attempts at education in the clinic setting, many families struggle to institute a bowel program in their homes. We propose a comprehensive quality improvement nursing approach to patient education with the hope of improving outcomes.
Methods A cross sectional cohort of children ages 3–19 years will be included. A bowel continence readiness nursing assessment tool will be used for patients instructed to begin a daily bowel management program. Nurses will provide one on one patient/family teaching in the clinical setting. Families will view a video discussing bowel function as a component of this teaching. Telephone follow-up will be provided to assess barriers. After a couple of months, families will be instructed to again complete the readiness assessment tool. Pre and post readiness scores will be compared with clinical bowel continence outcomes. Successful bowel regimens are measured by one or less accidents per month, decreased constipation and no visits to the ER due to bowel issues.
Results Assessments and evaluations are ongoing and results will be available on final poster.
Conclusions Through nursing assessment, planning, intervention and evaluation, we are able to measure patient's readiness for and success with bowel continence. Using novel approaches to nursing intervention, we have successfully used quality improvement methods to help children achieve the important goal of bowel continence.

Latin American-Descent Families' Educational Needs: Opportunity for International Advocacy and Collaboration to Promote Skin Health

Castillo, Jonathan, Perez, Ana, Morrison Jacobus, Melissa, Bolin, Rhonda G., Fremion, Ellen, Ostermaier, Kathryn, Castillo, Heidi
Texas Children's Hospital

Background In the US nearly 16% of children are children with special health care needs (CSHCN) and 13% of these live in Limited English Proficiency (LEP) homes. Many studies document that language barriers affect access to quality health care, patient safety, and patient-physician communication. Concurrently, research shows that culturally and linguistically appropriate, interventions for CSHCN with LEP families is effective in improving outcomes (Mosquera, et al. 2016). In Houston, where at least 145 languages are spoken at home, many families affected by spina bifida hail from and/or partly reside in Latin America. The purpose of our study was to describe and evaluate a locally-adapted, culturally sensitive, application of a Spina Bifida Association (SBA) sponsored skin care bundle among a primarily Spanish-speaking population.
Methods Within a large urban medical center, a patient education bundle made available by the SBA in collaboration with the Centers for Disease Control was rolled out as part of a quality improvement initiative to improve skin health.
Results Among our clinic (N = 313 pts), with a baseline skin injury rate of 13.1% (n = 41), risk factors associated with injury were identified. After staff training on the use of the educational materials made available through the National Spina Bifida Patient Registry (NSBPR), quality improvement methodology was effective in the outreach to Hispanic families which constitute 56.5% of patients.
Conclusions Employing quality design, the implementation of a culturally sensitive tool to assess risk of skin injury and provide education in Spanish is feasible. Lessons learn are shared in the hopes of aiding the up-scaling of this linguistically appropriate intervention across other NSBPR sites, as well as, at institutions caring for individuals living with spina bifida throughout Latin America.

Factors Associated with Bowel Continence in a Pediatric Spina Bifida Clinic

Davis, Richard Drew[1,2], Rocque, Brandon G.[1,2], Hopson, Betsy D.[2,1], Graham, Anna[2], Carroll, Rachel[1,2], Law, Charles[3,2,1]
[1]*University of Alabama At Birmingham*
[2]*Children's of Alabama*
[3]*United Cerebral Palsy of Greater Birmingham*

Background Neurogenic bowel is reported to occur in nearly 80% of individuals with spina bifida, and medical management represents a substantial portion of clinic visits. The purpose of this study is to evaluate the status and success of bowel management in our spina bifida clinic population.
Methods We collected data on stool continence and bowel management for subjects age 5 and older in our institutional pediatric spina bifida clinic using the National Spina Bifida Patient Registry and retrospective medical record review. We then performed univariate (Chi square, t-test) and multivariate (logistic regression) analyses to determine association between clinical and demographic variables and stool continence. We report interventions for subjects reporting a bowel management regimen.
Results A total of 459 individuals were included (52% female). 78% have a diagnosis of myelomeningocele, and 69% are white, non-Hispanic. 141 (48%) of individuals have stool incontinence at least monthly. Comparing subjects who have stool incontinence at least monthly to those with less frequent incontinence, increasing age (OR 1.098, $p = 0.001$), diagnosis of myelomeningocele (OR 3.3, $p = 0.003$), and thoracic lesion level (OR 6.5, $p = 0.001$ vs sacral) were associated with more frequent incontinence. An active bowel program was reported by 210 patients (46%), though there was no association between having a bowel management program and improved bowel continence. The most common components of a bowel management program were antegrade enema (31%), oral stimulant (24%), cone saline enema (16%), and oral stool softener (12%).
Conclusions We have identified factors associated with bowel incontinence in individuals with spina bifida, and characterized bowel management techniques. Given the lack of bowel program reported in 54% of subjects in spite of significant clinical efforts, this data will form the basis for a focus on improved bowel management in our clinics.

Healthcare Expenditures for the First Year of Life for United States Infants Born with Spina Bifida

Do, ThuyQuynh, Thibadeau, Judy, Ouyang, Lijing, Grosse, Scott
Centers for Disease Control and Prevention

Background Individuals with spina bifida (SB) face a variety of medical, physical, and neurocognitive challenges. Medical and surgical advances have significantly improved life expectancy ($\sim$ 90% reach adulthood). Several studies have explored the healthcare cost burden of SB during childhood, with the highest expenditures occurring during the first year of life (infancy). The mean direct lifetime cost per infant with SB is estimated to be $791,900 or $577,000 excluding caregiving costs (Grosse et al., 2016). Although most surgeries occur during infancy, little information exists on the type of procedures performed. The objective was to compare healthcare expenditures for infants with SB by infancy stage, procedure type, and patient setting.
Methods Data from the 2012–2014 MarketScan Commercial Claims and Medicaid databases were analyzed for infants born between January 1, 2012 and December 31, 2013 diagnosed with SB having 12 months of continuous insurance coverage. Separate analyses were performed to calculate mean and median net payment reimbursement over the first year of life and by each infancy stage.
Results A total of 252 infants with SB were included (privately insured = 95; Medicaid = 157). Over the course of the first year of life, the mean and median net payments for a privately insured infant were $84,347 and $54,042, respectively. Mean and median Medicaid net payments were $30,933 and $9,083. Top procedural categories differed by insurance. Both privately insured and Medicaid insured babies had surgical procedures involving urinary and nervous systems. More than 50% of surgical claims occurred between newborn and 1–3 months stages. Total net payment was highest during infancy stages, 4–6 months and 10+ months.
Conclusions Expenditure comparisons of insurance reimbursement are important in planning for resources and understanding healthcare utilization for an infant with SB.

The Prevalence of Skin Ulcers in Pediatric and Adult Spina Bifida Clinics

Fremion, Ellen[1,2], Dalton, Evan[1], Saha, Sharmistha[1,2], Castillo, Jonathan[2,1], Castillo, Heidi[2,1], Ostermaier, Kathryn[2,1]
[1]*Baylor College of Medicine*
[2]*Texas Children's Hospital*

Background Skin ulcers have gained recognition as a common morbidity and cause for hospitalization in the spina bifida (SB) population. Factors associated with skin ulcers have been described primarily in pediatric hospitals and clinics, but the differences in skin ulcer prevalence and associated risks between pediatric and adult clinic settings is less defined.
Methods In a one-year cross-sectional study, we examine skin ulcer prevalence and associated factors in patients presenting for their annual visit in a SB pediatric multi-disciplinary clinic (ages birth to 18) and the adult medical home clinic that receives the majority of the pediatric clinic's transition referrals (ages 18 and above).
Results Three hundred and fifty-six pediatric and 96 adult patients were studied (452 total). Sixty-five patients presented with a skin ulcer (14%) and 12 presented with multiple ulcers (3%). The prevalence of skin ulcers was increased in the adult clinic compared to the pediatric clinic (28% vs. 11%), and adults with skin ulcers were more likely to have multiple ulcers (26% vs. 13%). Factors associated with the presence of a skin ulcer included: higher SB lesion levels, decreased ambulation, African-American race, older age, and the presence of a shunt.
Conclusions The increased prevalence of skin ulcers in adult versus pediatric SB patients, and specific factors associated with skin ulcers, highlight the need for clinical guidelines, patient education, and self-management intervention for skin ulcer treatment and prevention. These interventions should address the specific care needs and associated factors for individuals with SB across the lifespan.

Case Studies: Nutrition for Youth with Spina Bifida Undergoing Surgical Interventions for Scoliosis

Meneses, Veronica M., Wittenbrook, Wendy, Torres, Angelica, Edwards, Latoya
TSRH

Background Youth with spina bifida are at increased risk for neuromuscular scoliosis. Progression impacts

multiple organ systems, including respiratory, feeding, and cardiac, with a significant role for nutrition across systems. Definitive treatment consists of spinal fusion by an orthopedist, with initial placement of halo for preliminary treatment with traction in some cases. Youth undergoing these procedures have distinct needs related to nutritional status, given risks of infections, complications and poor wound healing. Moreover, these youth often have pre-existing nutritional problems such as dysphagia, poor weight gain or obesity, chronic wounds, and psychosocial stressors.

Methods A biopsychosocial approach identifies the overall health and well-being of youth prior to surgery along with barriers to adherence to nutritional treatment. An interdisciplinary team including the Dietitian, Nurse, Developmental Pediatrician and Orthopedist evaluates across systems and formulates a comprehensive care plan. Youth receive integrated evaluation and intervention pre-, intra-, and post-operatively. Youth/family receive education with respect to how nutrition influences risks, recovery, and post-operatve functioning. Additionally, shared decision-making promotes adherence to the plan of care. Case Studies will present different scenarios for youth with spina bifida undergoing surgical interventions for scoliosis, exemplifying a model of care. Cases will include an underweight, early school-age youth with severe dysphagia as well as a youth with morbid obesity, Type II Diabetes Mellitus, and a chronic sacral wound.

Results A biospsychosocial interdisciplinary model prioritizing education and shared decision-making can optimize outcomes.

Conclusions Key approaches:

1. Biopsychosocial model.
2. Interdisciplinary co-management.
3. Education and shared decision-making.

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Clinician Self Knowledge: Aging with Spina Bifida

Still, Monica A.
SBAA

Background The population of people born with Spina Bifida is aging yet it appears that medical knowledge of the aging process of Adults with SB has not increased as the population has. Many adults with SB see providers who have little to no experience with this population. Do the clinicians feel that they lack the knowledge to provide their best practice to this population? Do they have resources?

Methods This researcher set up a Spina Bifida Awareness vendor at a UPMC Skin Care Conference. Skin care was chosen because of the impact wounds have on the SB adult community. UPMC was chosen because it is a teaching hospital, services a broad area of western PA, northern WV, and eastern Ohio. It has facilities that provide services along the entire continuum of medical care. It also has an active SB clinic for adults. A random sampling of 50 out of 80 attendees was chosen based on who approached the vendor table first. Each were given a questionnaire to complete. There was a 100% rate of return for the questionnaire and 98% of the attendees approached the vendor table.

Results 62% had taken care of an adult with SB. 96% were nurses. Of those who had cared for adults with SB 87% felt that they needed more knowledge and 93.5% did not know of resources. Of those who had no experience with adults with SB 63% felt they needed more knowledge and 74% did not know of resources.Life expectancy was placed at a higher age by those who had exposure to adults with SB.

Conclusions This is a very small sampling of the medical community. Clinicians were unaware of resources despite the presence of an active adult clinic. Clinicians who had provided care to adults with SB understood that the life expectancy normal. So, why does the myth of a shortened life expectancy persist? More research has to be done on how aging and spina bifida effect each other. Those results need to be distributed beyond the medical community that is traditionally seen as providers for those with SB.

A Review of Developmental Assessments in Spina Bifida: What This Tells Us About Service Delivery in the Australian Context

Williams, Tracey, Elliott, Rebecca, Botman, Antoinette
The Children's Hospital at Westmead

Background Historically, the Spina Bifida (SB) service at the Children's Hospital at Westmead has undertaken developmental assessments with children with SB and hydrocephalus at regular time-points (at 18, 30, and 42 months).

The first objective of this study was to determine the proportion of children with SB and hydrocephalus who scored below average in their cognitive skills on these developmental assessments. The second objective was to consider the clinical utility and implications for service delivery of conducting such assessments.

Methods All developmental assessments conducted by our SB service (from 2005 to present) were anal-

ysed. Data pertaining to the number and type of assessments, age at assessments, and assessment results were recorded. Results were categorised to indicate whether a child was considered to be 'competent', 'at risk' or 'delayed' in their cognitive skills at each time-point.
Results The data revealed that at any time-point at least 43% of children with SB were deemed to be 'at risk' or 'delayed' in their cognitive skills. There was inconsistency in the administration of assessments in terms of the frequency; the children's age; and in the assessment tools used. Conclusions could not be drawn about the most appropriate ages to conduct developmental assessments.
Conclusions The large portion of children who scored 'at risk' or 'delayed' with respect to their cognition demonstrates the need for regular developmental assessments to be conducted in SB services. These data are important to ensure referral to early intervention and appropriate planning prior to school commencement. Our research has also highlighted the need for consistency of the developmental assessments tools used over time, and the need for stricter adherence to assessment time-points in order to be able to investigate the developmental trajectory and needs of children with SB further.

Physical Activity and Social Participation

Physical Behavior in Wheelchair-using Youth with Spina Bifida

Bloemen, Manon[1,2], van den Berg, Rita[3], Tuijt, Matthijs[1], Nooijen, Carla[3], Takken, Tim[2], Backx, Frank[2], Vos, Marleen[1], de Groot, Janke[1,2]
[1]*HU University of Applied Sciences Utrecht*
[2]*University Medical Center Utrecht*
[3]*Erasmus University Medical Center*

Background Extensive literature shows associations between physical behavior (expressed in terms of activities and intensity) and health-related outcomes. Wheelchair-using youth are of high risk for developing unfavorable physical behavior so the goal of this study is to assess the physical behavior of wheelchair-using youth with SB.
Methods Accelerometry-based VitaMove data of 34 (13.7 ± 3.2 years) and Actiheart data of 36 (13.5 ± 3.6 years) wheelchair-using children with SB were used to assess physical behavior. The children used a wheelchair for daily life, long distances or sports. The VitaMove measures the type of activities. The VitaMove-data were compared to available reference data from typically developing peers. Sedentary activities were defined as sitting and lying. Dynamic activities were walking, running, wheeling, (hand)biking and non-cyclic moving. The Actiheart measures the intensity based on the heart rate. The % of heart rate reserve was used to classify the intensity into categories ranging from very light intensity to (near to) maximal intensity, based on the American College of Sports Medicine.
Results Wheelchair-using children with SB showed significantly more sedentary activities (94.3% versus 78.0%, $p < 0.05$) and significantly less dynamic activities (5.0% versus 12.2%, $p < 0.05$) compared to typically developing peers.
Wheelchair-using children with SB spent 90% of the wear time (IQR 8%) sitting or lying during a school day compared to 96% (IQR 10%) during a weekend day ($p < 0.01$). Moreover, the intensity of the activities was significantly higher during a school day compared to a weekend day.
Conclusions Wheelchair-using children with SB are significantly more sedentary and less dynamic-active compared to typically developing peers. Physical behavior during a school day is significantly more favorable compared to a weekend day. Our results show the necessity to improve physical behavior in wheelchair-using children with SB.

Predictors of Permanent Disability Among Adults with Spinal Dysraphism

Davis, Matthew C., Hopson, Betsy D., Blount, Jeffrey, Carroll, Rachel, Wilson, Tracey S., Powell, Danielle, McLain, Amie B., Rocque, Brandon G.
University of Alabama at Birmingham

Background Predictors of permanent disability among individuals with spinal dysraphism are not well established. We examined potential risk factors for self-reported permanent disability among adults with spinal dysraphism.
Methods One hundred eighty-eight consecutive individuals followed in an adult spina bifida clinic completed a standardized National Spina Bifida Patient Registry survey. Chi-square tests and logistic regression were used to identify factors independently associated with self-identification as "permanently disabled."
Results A total of 106 (56.4%) adults with spina bifida identified themselves as permanently disabled. On

multivariate analysis, relative to completion of primary and/or secondary school, completion of technical school (OR 0.01 95% CI 0–0.40 $p = 0.021$), some college (OR 0.22 95% CI 0.08–0.53 $p < 0.001$), college degree (OR 0.06 95% CI 0.003–0.66 $p = 0.019$), and holding an advanced degree (OR 0.12 95% CI 0.03–0.45 $p = 0.019$) were independently associated with disability. Relative to myelomeningocele, diagnosis of closed spinal dysraphism was also independently associated with disability (OR 0.20, 95% CI 0.04–0.90, $p = 0.036$). Additionally, relative to no stool incontinence, stool incontinence occurring at least daily (OR 6.41, 95% CI 1.56–32.90, $p = 0009$) or more than weekly (OR 3.43, 95% CI 1.10–11.89, $p = 0.033$) were both independently associated with disability. There was a suggestion of a dose-response relationship with respect to the influence of educational achievement and frequency of stool incontinence on likelihood of permanent disability.

Conclusions Our findings suggest that level of education and degree of stool incontinence are the strongest predictors of "permanent disability" among adults with spinal dysraphism. These findings will be the basis of efforts to improve community engagement and to improve readiness for transition to adult care in a multidisciplinary pediatric spina bifida clinic.

Effectiveness of Wheelchair Skills Training in Children

Sol, Marleen[1,2], de Groot, Janke[1,3], Verschuren, Olaf[2,3], Visser-Meily, J.M.A.[2]
[1]*HU, University of Applied Sciences Utrecht*
[2]*Center of Excellence for Rehabilitation Medicine, University of Medical Center Utrecht and De Hoogstraat Rehabilitation*
[3]*Partner of Shared Utrecht Pediatric Exercise Research (SUPER) Lab, Utrecht, The Netherlands; Child Development and Exercise Center, Wilhelmina Children's Hospital, University Medical Center Utrecht*

Background The majority of children with spina bifida use a wheelchair in daily life, community or sports settings. Recently, several facilitators and barriers towards physical activity were described by children with spina bifida and their parents. One of the facilitators mentioned was wheelchair skills training (WST), where children learn to become independent and confident in using their wheelchair. In adults, WST has proven to be beneficial towards gaining wheelchair skills and confidence. The benefit of WST in children remains unknown. Therefore the purpose of this study was to evaluate the effectiveness of WST in children.

Methods Children who use a manual wheelchair were recruited via a patient organization or special needs school. All children followed four to six WST sessions by an experienced trainer over a six month period. A repeated measures design was used to assess the pre-post training results on the Utrecht Pediatrics Wheelchair Mobility Skills Test (UP-WMST). In addition, patient reported outcomes from children and parents were recorded.

Results Thirty-five children, including 12 children with spina bifida, started the WST program. The mean time to complete an item on the UP-WMST showed an improvement on 13 of the 15 items, with a significant improvement in six of these items. In addition, the total ability to perform an item showed a strong significant improvement. Similar improvements were found in children with spina bifida and children with other diagnoses. All children and parents reported an improvement in wheelchair skills and all but one child noticed an increased confidence in wheelchair use.

Conclusions These preliminary findings show the overall positive results of the WST program on the UP-WMST, where children not only improved their ability to conquer obstacles but also increased their time to complete most items. These results are endorsed by the positive findings of children and parents on the effect of the WST program.

Transition/Adult Care

Adults with Myelomeningocele Living in Southern Sweden-: How Are We Doing?

Alriksson-Schmidt, Ann[1,2], Josenby-Lundkvist, Annika[3], Lindquist, Barbro[5], Westbom, Lena[4]
[1]*Lund University, Dept. of Clinical Sciences, Orthopedics*
[2]*Skåne University Hospital*
[3]*Lund University Department of Health Sciences*
[4]*Lund University, Department of Clinical Sciences Lund, Paediatrics*
[5]*The Habilitation Center, Hospital of Halland*

Background Children with myelomeningocele (MMC) in Sweden are eligible for coordinated multidisciplinary healthcare through the pediatric habilitation services. The same coordinated care is less available in adulthood. The problems related to the transition from pediatric- to adult healthcare are vast, and

include both personal and environmental factors. How these challenges manifest socially and medically in adulthood are unknown. We investigated health, medical status, physical functioning, health-related quality of life (HRQoL), and social outcomes in adults with MMC.

Methods Convenience sample of 51 adults with MMC (males = 53 %; median age = 29 years). Self-reported HRQoL was measured with the EQ5D-5L. Three structured questionnaires (general, physical therapy, and urology) were used to assess social-medical, physical functioning, and bowel and bladder function. In most cases, physical- and urotherapists performed the assessments and the principal investigator (neuropaediatrician) performed medical record reviews.

Results Fourteen were enrolled in school, 16 were working, 7 participated in government sponsored daily activities, and 14 did not participate in either category. Eleven of the participants had a driver's license. Twenty-seven percent reported a medical and/or gross motor decline respectively, and 23 % a decline in uro-bowel function over the past 3 years. Fifty-seven percent had normal renal function (based on relative eGFR), and 2 had undergone kidney transplants. Only 9 participants reported no urine or fecal leakage. Based on the medical record reviews, only 25% were considered to have received satisfactory medical follow-ups and 51% needed referrals or further medical follow-ups.

Conclusions Much work still remains to be done to maximize health and participation in adults with MMC.

Four Cohorts of Adolescents with Myelomeningocele from Western Sweden

Bondjers, Lisa, Abrahamsson, Kate, VuMinh Arnell, Magdalena, Olsson, Ingrid
Inst Clinical Sciences

Background The number of children born with myelomeningocele (MMC) has decreased during the last decades. It is important to assess the outcome as to the different medical aspects, in order to point at the need for an adequate follow-up by a multisdisciplinary team also in adulthood.

Methods In retrospective studies we have identified all adolescents with MMC and lipoMMC born 1986–1989, 1990–1993, 1994–1997 and 1998–2001, living in western Sweden (about 1.6 million inhabitants) on July 1st2004, July 1st2008, July 1st2012, July 1st2016, respectively.

Results There were in all 143 adolescents with MMC or lipoMMC in the four cohorts, with half the number in the last cohort compared to the previous ones. Seventy-five per cent had operated hydrocephalus, one third had untethering operations, and 38% had been treated for scoliosis. The majority had motor impairments – one third could walk independently indoors. One tenth had epilepsy. Clean intermittent catheterisation (CIC) was used by 92%, and in the last cohort the majority had started CIC in the neonatal period. None had kidney failure, and in the last cohort all had normal renal function. More than half used enemas on a regular basis. Comparisons will be presented between the different cohorts.

Conclusions Regular follow-up, including assessment of bladder and kidney function is important during childhood in order to preserve function and increase independence. Dedicated urotherapists play an important role in this. Life-long follow-up by multidisciplinary teams is necessary for adults with MMC. The complex medical situation, often in combination with cognitive difficulties, makes it necessary to coordinate medical services.

Bridging the Gap: The Value of Connecting with Spina Bifida Adults on Their Own Terms

Bruegel, Karlin
Todos Santos Institute

Background The long-term health of people with spina bifida (SB) depends on self-care behaviors that are outside the immediate control of medical practitioners. This paper presents a new framework for achieving improved autonomous self-care and health management in adults with SB, based on data from an ongoing online survey of 500+ respondents with SB. Data from this survey indicate that how people with SB conceptualize their health status may be substantially different from what the acute care model (ACM) assumes. These findings are discussed in terms of practical implications for clinical encounters and communication with adults with SB.

Methods As part of an extended online survey regarding health experiences, behaviors and outcomes of adults with SB, respondents were asked to rate their overall health status, based on 1) their personal evaluation of their overall health status, and 2) their evaluation of their overall health status compared to other adults with SB.

Results Based on preliminary analysis, adults with SB appear to anchor their perception of their health status

differently when comparing themselves to other adults with spina bifida than when the question is framed in terms of the conventional acute care model of health care. Subsequent conversations with adults with SB suggest that they make self-care decisions in terms of this implicit comparison with other adults with SB.
Conclusions Based on preliminary findings from the current survey, adults with SB appear to conceptualize their health status differently from what the acute care model (ACM) assumes. Since health professionals typically operate within the ACM, these preliminary findings have potential practical implications for clinical encounters and communication with adults with SB.

Pregnancies in Adults with Spina Bifida-A Single Institution Overview

Hopson, Betsy D.[1], Dew, Morgan [1], Arynchyna, Anastasia[1], Biggio, Joseph[2], McLain, Amie B.[2], Rocque, Brandon G.[1], Blount, Jeffrey[1]
[1]*UAB-Children's of Alabama*
[2]*UAB*

Background Successful implementation of CIC and effective treatments for hydrocephalus have extended the lifespan for individuals with spinal dysraphism such that decisions regarding elective pregnancies for patients with SB are increasingly important. The purpose of this descriptive study is to characterize the pregnancy history of women followed at an adult multi-disciplinary that carried successful pregnancies to term between 2009–2016.
Methods We have identified a cohort of female patients seen in the adult SB clinic who completed successful pregnancies. Included variables for chart review include the presence of gestational complications, a history of U.T.I.s, maternal surgical history, gravida/para status frequency of visits, reported obstetrical complications, and neurosurgical symptoms. A survey of standardized questions was developed to interview the patients via phone to learn more about their pregnancies and the complications which include dysraphism specific variables: consumption history of folic acid, mobility changes during pregnancy, epidural recommendations, pregnancy complications, and delivery method and complications.
Results A total of 193 individual charts were identified of whom 63% were female. Thirteen patients had pregnancies but 3 were excluded based on a diagnosis of split cord malformation and advanced maternal age. Ten met all inclusion criteria for the study of whom 6 had open MMC and 4 had lipomyelomeningocele. The records of these patients will be reviewed and the survey will be administered.
Conclusions Very limited data on pregnancy in Spina Bifida exists. This cohort will yield data of potentially great value in beginning to approach this very difficult and important problem.

Transition of Adolescents and Young Adults with Spina Bifida into Adult Primary Care and Specialty Health Care Services: A Systematic Review

Fortuna, Suzanne M.
UHCMC/RBC Hospital/Ursuline College

Background Problem Notable increased life expectancy for adolescents living with Spina Bifida (SB) resulting in the need for transition services into adult primary and specialties care. Many professional groups have highlighted the need for adult transition services, but few programs exist.
Purpose The purpose of this evidence-based review was based on the PICOT question: Does participation in an adult Spina Bifida transition program effect quality of life and emergency department visits compared with no participation over a 5 year period?
Methods Six different databases were searched using well defined inclusion and exclusion criteria. Ten key words were used in similar fashion in the different databases. Search resulted in 50 articles and 8 articles kept. The Level of Evidence for the 8 articles were: 5 articles Level IV and 3 articles Level VI.
Results Many factors contribute to transition challenges. Urologic issues contributed to higher ED visits but it was not clear if related to education or convenience. Care coordination was considered necessary, but different themes emerged regarding why assistance was provided or not. Results from QOL questionnaires found differences between self-reported QOL indicators and health related QOL indicators.
Conclusions An advanced-practice nursing role could include navigation coordination to assist transition from pediatric to adult based care. The development of transition programs would allow for improved access to care and health outcomes in the SB population.

A Condition-Specific Transition Clinic Model for Adolescents with Spina Bifida

Fremion, Ellen[1,2], Morrison Jacobus, Melissa[2]
[1]*Baylor College of Medicine*
[2]*Texas Children's Hospital*

Background The condition-specific health needs for children and adolescents with spina bifida (SB) are most commonly addressed in annual visit to an academic multidisciplinary clinic. Preparing for transition to adult care, condition-specific adolescent health needs, and self-management are difficult to address in this clinic model due to time constraints and clinic scope. We present a SB condition-specific transition readiness clinic model that (1) is based on the chronic care model, (2) works in conjunction with a multidisciplinary clinic, and (3) facilitates transition planning.
Methods Adolescents with SB ages 14–18 referred from the SB multidisciplinary clinic are seen every 3 months in the SB transition clinic by an internal medicine-pediatric physician, a nurse, and a social worker to develop transition plans, address chronic condition management, and work-on patient/family-centered self-management interventions.
Results In the first year, 68 of 100 patients referred to the SBTRC from the multidisciplinary clinic have established care SB transition clinic. The triple aim framework will be used to evaluate the effectiveness of this model. Namely, (1) care experience measured by patient satisfaction with transition, (2) population health measured by improvement of self-management skills, and (3) cost/gaps in care measured by time lapse between pediatric and adult care.
Conclusions Transition care plans, self-management support, care coordination, transition clinic visits, patient-centered care, and involvement of adult-centered healthcare providers have been identified as transition facilitators for adolescents with SB. This SB condition-specific transition clinic presents a feasible model for incorporating these essential elements for transition in collaboration with an existing multidisciplinary clinic.

TRAQ Validity in Spina Bifida Population

Getz, Marta
Cincinnati Children's Hospital Medical Center

Background Youth with Spina Bifida (SB) experience gaps in care transitioning from pediatric to adult care. The AAP recommends all youth are assessed for transition readiness. The Transition Readiness Assessment Questionnaire (TRAQ) is a validated tool used to assess typically developing adolescent's readiness to transition. Limited research has been conducted to determine its effectiveness in those with special needs, such as SB.
Methods The social worker (SW) at the Center for Spina Bifida at Cincinnati Children's Hospital Medical Center began administering the TRAQ to all patients age 14 and older at clinic visits.
SW identifies patients during pre-clinic conference. Nurse coordinators designate time for SW to meet with patient during visit. Due to patient's difficulty understanding written questions in pilot testing it was determined TRAQ needed to be administered orally by SW. Upon completion SW collaborates with patient developing a tailored transition goal. SW documents areas of strengths (based on scores 4–5), weaknesses (scores 1–3) and the agreed upon goal is documented in SW note in Epic. SW reviews progress on goal at follow up visit.
Results Forty-seven of the 60 patients approached from Jan 2015-Oct 2016 completed the TRAQ. The mean age was 17. Most patient reported being fully ready to transition on items related to "Talking with Providers" and "Managing Daily Activities." In the 19 patients who have repeated data collection all of them have increased TRAQ scores.
Items frequently scored lower on the TRAQ are related to appointment scheduling and preparing for appointment.
During oral administration patients engage and are able to answer questions mostly independently.
Conclusions The TRAQ was helpful in assessing transition readiness in adolescents with SB, however due to cognitive deficits modification of the TRAQ was necessary to obtain an accurate assessment. Additional evaluation of the validity of the TRAQ is needed in patients with cognitive deficits.

Risk Factors for Metabolic Syndrome in Adult Spina Bifida Patients

Kielb, Stephanie, Liu, Joceline, Dong, Caroline
Northwestern University Feinberg School of Medicine

Background Spina bifida (SB) is one of the most common birth defects and 75–85% of these patients will now reach adulthood. Preliminary studies have shown increased rates of obesity in these patients due to a combination of altered body composition and limited

mobility. Traditional measures of body mass index are typically inaccurate to diagnosis obesity in this population. We seek to investigate the prevalence of risk factors for metabolic syndrome (MetS) in these patients compared to the general population (GP).
Methods We performed a retrospective chart review of 209 adult SB patients seen at an Interdisciplinary SB Clinic between 2007–2014. We analyzed individual rates of MetS risk factors in our study population and compared these to age-matched GP prevalence.
Results Of 210 patients (121 female), mean age was 32.7 (20–80) years with 171 (81.4%) patients falling in the 20–39 range. 168 (80.0%) patients had BMI data, 199 (94.8%) had blood pressure data, 68 (32.4%) had HgA1c data, and 40 (19.0%) had lipid data. Compared with age-equivalent GP, 74 (44.0%) patients versus 30.3% GP met MetS criteria by BMI, 78 (39.2%) versus 11.9% GP were hypertensive, 11 (16.2%) versus 21.1% GP were insulin-resistant, and 28 (70.0%) versus 23.3% GP were dyslipidemic.
Conclusions Excluding insulin resistance, adult SB patients have a higher prevalence of risk factors for MetS relative to the GP. Given previously established inaccuracy of BMI measurement in SB patients and our limited HgA1c and lipid data, disease burden is almost certainly underestimated in this population. We are currently collecting prospective data including waist circumference and other metabolic parameters to further ascertain accurate prevalence rates of MetS in this high-risk cohort. We hope our preliminary data argues the necessity of comprehensive medical follow-up as these patients continue to come of age.

High Rates of Admission Seen with Adult Spina Bifida Patients Presenting to the Emergency Room

Kielb, Stephanie, Matulewicz, Richard S., Nettey, Oluwarotimi S.
Northwestern University Feinberg School of Medicine

Background The majority of spina bifida patients are now living to adulthood, but finding adult providers to care for these patients is a known transitional challenge. We evaluated the use of the emergency room (ER) by these patients, the presenting complaint, rates of admission, and admitting diagnosis.
Methods A retrospective review of all patients with a diagnosis of spina bifida and neurogenic bladder presenting to the Northwestern University Hospital ER from 2008–2015 was conducted. Information on chief complaint, admitting diagnosis, admission disposition, and imaging utilization was collected.
Results Two hundred and thirty-one patients were identified that met study criteria. The chief complaint was abdominal pain/nausea/vomitting/diarrhea in 33, headache/shunt issues in 30, skin issues or pressure sores in 25, chills/fevers of unknown origin in 22, urinary tract infection (upper or lower) in 22, back/flank pain in 16 (with known nephrolithiasis in 5), catheter issues in 13, chest pain/cough in 12, post-operative complications in 8, and dialysis complications in 7, with the remainder comprising a variety of complaints. Of the 231 SB patients, 199 (90%) were kept overnight, 179 (77%) for inpatient hospitalization and 13% for overnight observation. The most common admitting diagnoses were urinary tract infection, abdominal pain, cellulitis, and osteomyelitis. Of these 231 patients, 154 (68%) had a urine culture sent regardless of chief complaint.
Conclusions Adult SB patients present to the emergency with varied chief complaints. The overwhelming majority are admitted for in-patient management. The most common chief complaints were abdominal pain, headache/shunt issues, and skin issues/pressure sores. The majority of patients had a urine culture sent despite often non-urologic chief complaints.

Transition Planning Process in a Comprehensive Spina Bifida Clinic

Sherburne, Eileen, Larson, Heidi
Children's Hospital of Wisconsin

Background The population of children with chronic illnesses and specifically Spina Bifida has grown over the last 20 years resulting in children transitioning to adulthood and requiring adult centric health care. It can be a challenge for pediatric providers to prepare not only themselves but also patients and families for this transition to increased self-management and the adult health care setting. A multidisciplinary work group has developed and implemented a transition planning process promoting self-management for children with chronic illnesses including Spina Bifida. The goal is to determine best practices for a standard transition process.
Methods The IHI Model for Improvement was used for this ongoing project. A package of materials was developed for champions in the pilot group of four specialty clinics including Spina Bifida clinic who provide care to children with chronic illnesses. Education was provided to the champions that included use of the Epic transition checklist flowsheet as well as avail-

able national resources for use to support the evolving transition process. The checklist is also used for ongoing anticipatory guidance for providers and children/families.
Results Evaluation included determining the number of patients who had documentation within the transition checklist flowsheet during a clinic visit. The pilot clinics have demonstrated regular use of the checklist varying from 45% to 90% although data has not stabilized. Champions have initiated follow-up phone calls to examine feedback about the transition process from young adults.
Conclusions A well-designed ongoing process is a necessary program component for building self-management skills and ensuring successful transition of children with chronic health conditions to the adult world.

Urology

Factors Associated with Transition from Caregiver to Self Clean Intermittent Catheterization in Spina Bifida

Dangle, Pankaj P.[2], Atchley, Travis[1], Hopson, Betsy D.[2], Graham, Anna[2], Arynchyna, Anastasia[2], Rocque, Brandon G.[2], Joseph, David B.[2]
[1]*University of Alabama School of Medicine*
[2]*UAB-Children's of Alabama*

Background In patients with spina bifida (SB), clean intermittent catheterization (CIC) is the mainstay of treatment to prevent renal damage. In a separate analysis, 71.9% of our patients have transitioned to self-CIC by age 10. We reviewed patients $\geqslant$ 10 years to determine which factors may contribute to the delayed or lack of transition in some patients given the greater majority have already begun self-CIC.
Methods We retrospectively reviewed all patients in our pediatric and adult University of Alabama at Birmingham and Children's of Alabama SB registry who were $\geqslant$ 10 years at last visit. We evaluated univariate correlations followed by multivariate stepwise regression on variables with a p-value < 0.05. Patients with incomplete data were excluded.
Results We identified 696 total patients. 287/696 (41.2%) patients were $\geqslant$ 10 years and were on CIC. Of 287 patients, 175 (61.0%) self-CIC. Female gender, myelomeningocele (vs. lipoma, meningocele, etc.), Medicaid insurance, thoracic-level spinal lesions, wheelchair requirement, use of anti-muscarinics, and history of ventriculoperitoneal (VP) shunt were all associated with delayed or lack of transition to self-CIC via univariate analysis. In multivariate analysis, thoracic-level spinal lesions ($p = 0.037$, OR 0.435) and Medicaid insurance ($p = 0.001$, OR 0.241) were independently associated with delayed or lack of transition to self-CIC.
Conclusions Thoracic-level spinal lesions and Medicaid insurance contribute to delayed or lack of transition to self-CIC. Additionally, other non-modifiable risk factors (gender, primary diagnosis, wheelchair use, anti-muscarinic use, and VP shunt) may influence this transition and ultimately functional independence in our SB population.

Practice Patterns for Management of Spina Bifida Patients with Urinary Stones: Single Institutional Experience

Chaudhry, Rajeev, Ost, Michael, Schneck, Francis, Stephany, Heidi, Cannon, Glenn
Children's Hospital of Pittsburgh

Background Patients with spina bifida (SB) are at increased risk of stone formation with high recurrence rates. Management of stones should include a complete metabolic evaluation and, if necessary, pharmacotherapy. The objective of our study was to determine the practice patterns of stone management in this complex patient population.
Methods We performed a retrospective review of pediatric patients with SB who have diagnosis of urinary stones and have undergone definitive stone management. We evaluated referral patterns, types of management and compliance to therapy. In our institution, medical management of stones is performed by nephrologists based on our referral.
Results Fourteen patients with SB had kidney and/or bladder stones that underwent definitive stone surgery, for a total of 28 stone procedures. Of these 14 patients, 7 patients were referred to nephrology for stone management; the remainder was managed by urology. Of those referred to nephrology, 57% made the consult visit, and only 1 patient returned for subsequent follow-up visits. Only 21% of the cohort completed 24hr urine studies, and these patients were all seen by nephrology. One patient was placed on medical management by nephrology, none by urology. Sixty-four percent of patients had recurrence of their stones within 1 year of treatment.
Conclusions Medical management of stones in the SB population remains to be a challenge. Lack of referral

to pediatric nephrology, compliance with appointments and incomplete metabolic work-up seem to be issues in this population. Larger scale studies are warranted to determine the better ways to manage these patients.

History of Urologic Management of Spina Bifida Pre and Post Introduction of Clean Intermittent Catheterization

Chaudhry, Rajeev, Ost, Michael, Cannon, Glenn, Schneck, Francis, Stephany, Heidi
Children's Hospital of Pittsburgh

Background The urologic management of spina bifida (SB) has evolved over the last century, with greater emphasis on medical management and intermittent catheterization. We review the history of urologic management of spina bifida pre and post introduction of clean intermittent catheterization (CIC).
Methods A literature search of medical textbooks, journal articles and historical texts was performed pertaining to history of management of SB.
Results Prior to the advent of CIC, upper tract disease occurred in 40-90% of patients by age 10. Intervention consisted of treating the urologic complications and urinary diversion was the primary treatment option as the bladder was considered unsuitable for use. The development of video urodynamics in the 1960s provided better understanding of the bladder and sphincter physiology in diseased states. In 1972, Lapides et al. first described the CIC technique for bladder management. This provided a reduction in upper tract disease, increased utilization of bladder augmentation and overall increased survival. This also led to the development of continent catheterizable channels, as well as improvements in the augmentation techniques and outcomes. In 1981, McGuire et al. linked elevated detrusor leak point pressures $>$ 40 cm H2O with increased risk of upper tract damage. This landmark study still influences contemporary management of SB patients. There is now an increased emphasis on optimizing bladder function via medical management and pharmacotherapy in order to potentially decrease or eliminate the need for reconstructive intervention all together.
Conclusions Surgical and medical management of SB patients has evolved. The introduction of CIC is still one the greatest advancements in urologic management. As we continue to better understand the neurogenic bladder, we hope to make even greater strides in management over the next 100 years.

Bladder and Bowel Continence in the Spina Bifida Population

Clayton, Gerald H.[1,2], Blanchard, Alesia[1,2], Weitzenkamp, David[2], Teravist, Erin[1], Korber, Marguerite[1], O'Keeffe, Shannon[1,2], Steiner, Julie[1,2], Wilson, Pamela[1,2]
[1]*Children's Hospital Colorado*
[2]*University of Colorado, School of Medicine*

Background Development of an independent means of continence as a child matures is critical to achieving functional and social independence as an adult. This is even more relevant in the disabled population where such factors are known to be significant impediments to transition into an independent adult lifestyle. We aim to identify factors of significance that influence the ability of Spina Bifida children to maintain bowel and bladder continence.
Methods Using generalized linear mixed model analyses (N $=$ 273, 463 visits) subjects enrolled in the CDC National Spina Bifida Registry, we evaluated the impact of lesion level, shunt surgery frequency, bladder and bowel management method, gender, race and ethnicity on levels of incontinence in this population. Primary dependent outcome measures were frequency of bowel and bladder incontinence. Concurrent infections and illness were considered in defining incontinence.
Results Bowel and bladder continence is affected by both management method ($p < 0.0001$) and gender ($p = 0.002$) where women likely have lower frequencies of incontinence. Intermittent catheterization and spontaneous voiding associate with lower bladder incontinence frequencies however management method relationships for bowel are unclear. Higher lesion levels are associated with elevated bowel incontinence frequencies ($p = 0.0136$) but not bladder. Ethnicity and shunt surgery frequency did not play a role however race impacted bowel incontinence ($p = 0.0102$).
Conclusions In this population bowel and bladder incontinence frequency is significantly impacted by gender and management method however lesion level influences only bowel incontinence frequency. Further analysis will divulge the impact of other surgeries and allow for the completion of the continence model. Additional enrollment in other populations will be required to develop a more complete and generalizable model.

The Role of Robotic Surgery in Complex Surgical Reconstruction of Neurogenic Bladder in Myelomeningocele Patients: Comparison of Open versus Robotic Approach

Cornwell, Laura[1], Madris, Brandon[2], Riddell, Jonathan[1]
[1]*SUNY Upstate Medical University*
[2]*St George's University*

Background Augmentation cystoplasty is a well-described surgical option for patients with refractory neurogenic bladder. Traditionally the procedure is done in an open fashion and is accompanied by other reconstructive endeavors such as appendicovesicostomy, bladder neck sling, and antegrade continence enema. As with many open procedures, application of minimally invasive technology may reduce post-operative discomfort and other complications. This study assesses the impact of robotic technology on the early post-operative course.
Methods We performed a single institution review of all augmentation cystoplasties performed from 2011 to 2015 and classified cases as either robotic or open. Primary outcomes included procedure length, number of days admitted, time to return of bowel function, and post-operative narcotic requirements with comparison between groups.
Results Thirty-two patients, mean age 9.7 (3–31), underwent augmentation cystoplasty: 22 open and 10 robotic. The groups were comparable in age, gender, neurogenic bladder etiology, and number of concomitant procedures. 16 (50.0%) had history of myelomeningocele (MMC) or segmental agenesis. Open procedures were found to be shorter in length (368.7 vs 565.6 minutes, $p < 0.001$) but required significantly more post-operative narcotics (8.1 vs 4.4 PO morphine equivalents/kg, $p = 0.048$). This narcotic requirement was more pronounced when epidural anesthesia use was excluded (9.0 vs 4.4, $p = 0.031$). Open vs robotic comparison demonstrated no advantage in admission length (10.0 vs 10.3 days, $p = 0.902$) or time to return of bowel function (5.2 vs 7.0 days, $p = 0.062$).
Conclusions Robotic technology significantly decreases post-operative narcotic requirements for patients undergoing complex reconstruction of the urinary tract, especially for those who are not a candidate for epidural anesthesia (such as MMC).

Pediatric IC: Education for Community Based Collaboration

Fortuna, Suzanne M.[1,2]
[1]*UHCMC/RBC Hospital*
[2]*Ursuline College*

Background Children with Special Health Care needs (CSHCN) are increasingly mainstreamed in school and community settings requiring collaborative team approach to handle complex medical care. Children with complex urologic conditions that require intermittent straight catheterization, many whom are born with spina bifida, neurogenic bladder is a common sequelae. The community based providers can collaborate with urologic professionals to optimize medical management in this population.
Methods Education to improve understanding and maintain consistent practices among health care professionals is essential to maintain optimal urologic health in those with complex urologic conditions. The authors provide insightful steps to intermittent straight catheterization methods and guidelines to promote expert urologic management among community caregivers.
Conclusions Collaboration is key among community providers to optimize urologic health for persons with complex urologic conditions requiring intermittent catheterization routinely in community settings. The urologic Nurse can be the liason to ensure adequate education among caregivers to those with complex urologic healthcare needs.

Robotic-Assisted Lower Urinary Tract Reconstruction in Spina Bifida Patients with Previous Abdominal Surgery

Gargollo, Patricio C.[1], Jacobs, Micah[2], Granberg, Candace F.[1]
[1]*The Mayo Clinic*
[2]*University of Texas Southwestern Medical Center*

Background We describe our experience with robotic assisted complex lower urinary tract reconstruction (CUTR) in spina bifida patients with previous open abdominal surgery (PAS).
Methods All patients with a history of spina bifida and neurogenic bladder and any previous open abdominal surgery undergoing robotic assisted CUTR were included in the study. CUTR was defined as bladder neck reconstruction (BNR) or continent catheterizable conduits (CCC) or both or redo surgery at the bladder

neck for persistent incontinence. Patient demographics, surgery performed, operative techniques, operative times and outcomes were assessed.
Results Thirty-two patients met inclusion criteria during the period analyzed. Twenty-two of these patients had multiple laparotomies for ventriculoperitoneal (VP) shunt revisions and 10 had major previous abdominal surgery including laparotomy with other adjunct procedures in 8 and laparotomy with colostomy in 2. There were 3 conversions. Mean operative time was 8.7 hrs (4.5–12.2 hrs). Mean length of stay (LOS) was 52.3 hrs (23–92 hrs). The first 16 cases took significantly longer than the last 16 cases (mean 11.6 hr vs mean 6.9 hr $p = 0.002$). The distribution of types procedures performed was the same in comparing the first 16 and the last 16 cases. Patients with multiple VP shunt revisions had statistically significant higher conversion rates and higher mean operative times when compared to other patients ($p = 0.01$ and $p = 0.002$ respectively). Patients with multiple VP shunt revisions had statistically longer LOS when compared to other patients ($p = 0.02$).
Conclusions Robotic assisted CUTR in neurogenic bladder patients with previous open abdominal surgery is safe and feasible. Longer operative times should be expected early in a surgeon's experience. Multiple VP shunt revisions have higher conversion rates to open and longer operative times when compared to other indications for previous surgery.

Effect of Bladder Reconstruction on VP Shunt Failure Rates in Spina Bifida

Gonzalez, Dani., Cooper, Jennifer, McLeod, Daryl J.
Nationwide Children's Hospital

Background It is well established that children with spina bifida are at an increased risk for renal injury. In some cases, bladder augmentation is necessary. The majority of patients with spina bifida also require ventriculoperitoneal (VP) shunt placement. There are concerns that bladder augmentation surgery may increase the risk of VP shunt malfunction and/or failure. The aim of this study was to assess whether bladder augmentation affects the rate of VP shunt failure in this population.
Methods Using the Pediatric Health Information System, we studied patients born between 1992–2014 with spina bifida who underwent surgical closure within the first 4 days of life and a VP shunt placement procedure. Using conditional logistic regression, we compared age- and hospital-matched patients who did and did not undergo a bladder augmentation to determine the difference in their rates of VP shunt malfunction or replacement/revision.
Results A total of 4192 patients with spina bifida who underwent both surgical closure and VP shunt placement were identified. Of these, 203 patients with bladder augmentation could be age- and hospital-matched to 593 patients without bladder augmentation at a ratio of between 1:1 and 4:1. The median age at VP shunt placement was 7 days and the median age at bladder augmentation was 7 years. VP shunt malfunction, replacement, or revision occurred within 2 years in 49 (6.2%) of patients, the majority of whom were in the group who underwent bladder augmentation (87.8%, $p < 0.001$). After adjusting for confounders, undergoing bladder augmentation was independently associated with VP shunt malfunction, replacement, or revision (HR: 33.5, 95% CI: 13.15–85.44, $p < 0.001$).
Conclusions Bladder augmentation appears to be associated with increased rates of VP shunt malfunction and replacement/revision. Additional studies are necessary to better define this relationship and identify risk-reduction techniques.

Efficacy of Peristeen Transanal Irrigation System for Neurogenic Bowel in the Pediatric Population

Gordon, Tiffany D., Vandersteen, David
Gillette Children's Specialty Healthcare

Background Fecal incontinence and chronic constipation are significant medical and social concerns for patients with neurogenic bladder. Peristeen has been utilized outside the US for the management of these patients since 2006.Peristeen was approved for use in the United States in 2012.The purpose of this study is to determine efficacy of Peristeen.
Methods Patients were referred for Peristeen from our Urology providers as well as from providers outside of our system. All patients referred for Peristeen were included in the study. There were no exclusion criteria. Patients were assessed using the Neurogenic Bowel Dysfunction Score (NBDS) upon entry, at 3 mos, 6 mos, and 1 year Patients were also assessed regarding prior bowel management programs and treatment failures.Patients were assessed for prior bowel management, treatment failures, and complications of treatment. The data were acquired prospectively and reviewed retrospectively.
Results Fifty-three patients were referred for Peristeen. Six patients were denied/awaiting approval for

treatment. Eight patients were approved for Peristeen but have deferred therapy. Thirty-six patients Peristeen teaching; and are the study group. All patients failed one or more bowel management programs (BMP), including 2 patients with prior ACE procedure, 18 patients with prior enema programs and 18 patients on oral agents only. NBDS improved in 80% at 3 m f/u, and in 86% at 6 m f/u. Detailed analysis within the NBDS demonstrated more improvement in number of incontinent bowel episodes and time spent doing BMP.4% of patients failed treatment. Of patients with prior ACE, 100% had success with Peristeen. There were no complications of treatment.

Conclusions Peristeen is an effective and safe means of managing NB in the pediatric population. Results improve between 3 and 6 months of initiation of therapy. Prior failure of ACE irrigation or other enema programs did not predispose to failure of Peristeen.

Surgical Outcomes of Open Ureteral Reimplantation in Patients with Spinal Dysraphism: A Single Institutional Experience

Hugar, Lee A.[1,2], Chaudhry, Rajeev[2], Stephany, Heidi[2], Ost, Michael[2,1], Schneck, Francis[2], Cannon, Glenn[2]
[1]*University of Pittsburgh Medical Center*
[2]*Children's Hospital of Pittsburgh*

Background Open ureteral reimplantation (OUR) is a common and highly successful urologic procedure for vesicoureteral reflux. However, spina bifida (SB) patients present with unique challenges when undergoing OUR compared to the general pediatric population. In this study, we aim to describe outcomes of patients with SB undergoing open ureteral reimplantation.

Methods A retrospective review of all patients undergoing OUR at a single institution between the years 2008–2015 was performed. Collected variables included demographics, neurological lesion level and type, grade and laterality of reflux, and postoperative outcomes.

Results Of 408 patients undergoing ureteral reimplantation at our institution over an eight-year span, 10 had spinal dysraphism (2.45%). The median age at surgery was 148.9 months, or 12.4 years, and the median body mass index was 21 kg/m2. Half of the cohort was female, and 70% of patients had a lumbar lesion. Seventy percent of patients underwent primary OUR; mostly for high grade bilateral vesicoureteral reflux with recurrent pyelonephritis (5 of 7 patients, 71%). Of the primary OURs, four were performed in a Cohen cross-trigonal fashion and three were done using the Politano-Leadbetter technique. Three patients underwent redo-OURs for stricture. Median length of stay for patients was 5 days. Median follow up was 33 months.

Conclusions Open ureteral reimplantation is a challenging procedure in SB, but can be done safely for the appropriate indications with overall acceptable long term outcomes.

Dedicated Neurogenic Bowel Provider within a Multidisciplinary Myelomeningocele Clinic: Is there an Advantage?

Manz, Betty Jo, Lingsch, Tammy, VanderBrink, Brian
Cincinnati Children's

Background Bowel dysfunction is common in patients with myelomeningocele and may negatively impacts the quality of life. Many therapeutic options exist to improve clinical situation and these recommendations are made by a variety of individuals: primary care doctors, gastroeneterologists, urologists and colorectal specialists. Historically in our center the urology team simultaneously monitored and managed the bladder and bowel dysfunction. In 2013 we decided to separate these important clinical care duties within our Division by having separate scheduled appointments with different providers. The impetus behind this process was that there may be better attention to bowel dysfunction rather than secondarily addressing it following urinary tract management. We report our early experience in this care delivery model.

Methods New and returning patients to our myelomeningocele clinic since 2013 were scheduled to have a bowel management appointment by an experienced nurse practitioner in addition to their scheduled urology appointments. Detailed parent and patient reported bowel habit history were taken. Radiographs were conducted during clinic follow-up to mitigate impact of neurogenic bowel.

Results Two hundred twenty-nine patients were evaluated between January 2013-Sept 2016. There were a total of 527 encounters for these patients. Education about bowel dysfunction was delivered in all patients at same time as assessing for its presence. Dietary recommendations and oral medications were first line interventions in symptomatic patients. Enemas were reserved for refractory cases of constipation or fecal incontinence.

Conclusions Bowel dysfunction can be successful addressed within the framework of multidisciplinary myelomeningocele clinic. Patients, families and providers subjectively found advantages to separate bowel mangement provider model compared to prior urology provider addressing bladder and bowel dysfunction.

Considerations of Urinary Bacterial Colonization in Patients with Myelomeningocele at Time of Urologic Testing

Preece, Janae[2], Baxter, Cheryl[1], Riesenberg, Stephanie[1], Haynes, Andria[1], Ching, Christina[1]
[1]*Nationwide Children's Hospital*
[2]*Children's Hospital of Michigan*

Background Patients with myelomeningoceles have multiple risk factors for urinary tract infection (UTI), including regular urologic testing with urinary tract manipulation. We examined the results of routine urinalysis and urine culture performed at the time of urologic testing in our myelomeningocele population and the risk factors for infectious complications after testing.
Methods Urine was collected by void or clean intermittent catheterization (CIC) from patients with myelomeningocele undergoing renal ultrasound or urodynamics (UDS) with IRB approval. Patients with augmentation cystoplasty were excluded. Results were compared between patients on CIC and those voiding.
Results Sixty patients were included: 52 on CIC and 8 freely voiding. Mean age for all patients was 12.6 years (range 0.2 to 39.7 years); 35% were male. Patients on CIC were significantly older than those freely voiding (mean age 13.6 vs. 6.6 years respectively, $p = 0.04$). There was no difference in gender between groups. More patients on CIC were colonized (54% vs. 38%), however this was not statistically significant. 19% of those on CIC had an UTI at presentation while none of the voiders did. Patients on CIC tended to have multiple organisms in their urine if colonized or infected. The only post-instrumentation complication was pyelonephritis in 1 of the 3 colonized freely voiding patients undergoing UDS. None of the colonized patients on CIC undergoing UDS had a complication.
Conclusions Patients with myelomeningocele are more likely to require CIC as they age. Over half of patients on CIC are colonized, though colonization in voiding patients predisposes to post-UDS complications. This information can guide care for patients at time of urologic testing and dictate which studies are cancelled in order to prevent post-instrumentation pyelonephritis.

Healthcare Provider Experiences in Transitioning Spina Bifida Patients from Pediatric to Adult Care

Slocombe, Kimberly[1], Agrawal, Shree[2], Wilson, Tracey S.[3], Kielb, Stephanie[4], Hadley, Wood[1]
[1]*Cleveland Clinic*
[2]*Case Western Reserve university*
[3]*University of Alabama at Birmingham*
[4]*Northwestern University*

Background The lack of precedent in transition from pediatric to adult care has posed a unique challenge to managing patients with spina bifida (SB). The purpose of this study is to generate a current summary of practices in management of adults with SB across urology providers in the US.
Methods For a national survey of practicing urologists treating patients with SB, we identified 174 physicians, who are current members of the AUA mailing list for urologic congenitalism/SBA network. Survey responses were assessed for implementation/barriers to interdisciplinary adult SB clinics, continuity of care, long term follow-up, and transitioning care.
Results The survey response rate was 40%, which included urologists specialized in pediatrics, genitourinary reconstruction, female pelvic medicine, and reconstructive surgery. SB clinics included adult multidisciplinary care (14%), adults seen in regular clinic (34%), combined adult-pediatric multidisciplinary care (20%), and pediatric multidisciplinary care (28%). Ideal transition from pediatric to adult care was considered to be at 18 (24%) or 21 (22%) years old. Adult patients with SB not experiencing an acute issue were surveilled annually via upper tract imaging with renal ultrasound (91%), serum creatinine to monitor renal function, and cystoscopy to monitor bladder function. 80% of practicing urologists perform urodynamic testing only when there is a change in condition. Providers for patients with SB from 2006 (AHRQ survey) and 2016 report in addition to urology, neurosurgery/neurology (59% vs. 87%), social work (68% vs. 84%), and orthopedics (59% vs. 73%) are essential to the care of adults with SB.
Conclusions This national survey identified several areas for improvement in the care of adults with SB including national provider resources/standardized guidelines, increased collaboration, access to care, and a need for an advanced training pathway.

Highly Elevated Cystatin C GFR in Pediatric Myelomeningocele Patients: Does it Indicate Nephropathy?

Liss, Zachary, VanderBrink, Brian, Jackson, Elizabeth, Minevich, Eugene, Reddy, Pramod, Noh, Paul, DeFoor, W. Robert
Cincinnati Children's

Background Serum Cystatin C is becoming more widely used to estimate Glomerular Filtration Rate (GFR) in spina bifida patients with neurogenic bladder. A highly elevated GFR is difficult to interpret and perhaps suggestive of hyperfiltration injury to the kidneys. It has been our practice to assess for proteinuria in this patient population. The purpose of this study is to review the results of this practice to see if nephropathy is a concern with an elevated Cystatin C GFR.

Methods A retrospective cohort study was conducted of all MM patients with an available Cystatin C GFR at a single pediatric institution between the ages of 2 and 12 years. Data abstracted included patient demographics, Cystatin C GFR, urine protein data, and urine specific gravity. High GFR was defined as greater than 150 ml/min. Proteinuria was defined as presence of greater than trace protein on urine dipstick or a quantitative urine protein to creatinine ratio exceeding 0.21. Patients were excluded if the urinalysis had a low specific gravity that could possibly cause a false negative for proteinuria.

Results A total of 261 patients with MM were identified. Median age was 7 years. 54 patients (21%) had a Cystatin C GFR greater than 150 ml/min. Eighteen had a specific gravity less than 1.010. Thus, 36 patients were included in the study group. Of these patients, 30 (83%) had a negative urinalysis for protein and/or a negative quantitative protein to creatinine ratio. Only 6 patients (17%) had evidence of proteinuria.

Conclusions An elevated Cystatin C GFR (CGFR) was discovered in 20% of the myelomeningocele patients in our series. Our preliminary data shows that the vast majority do not show evidence of proteinuria. Further evaluation and longer follow-up is necessary to assess if those patients with proteinuria or a dilute urinalysis are at higher risk for hypertension, upper tract deterioration, and/or worsening bladder dynamics.