

Adult Spina Bifida Transition Clinic: Bridging the Gap of Care



Objectives for Presentation

- Understand definition of spina bifida (SB)
- Understand basic urologic surgeries common in SB
- Understand the importance of transitional care from pediatric to adult care
- Become an advocate for transitional care



2

Spina Bifida (SB)

- Spina bifida is a neural tube defect that affects the spine and is usually obvious at birth.
- Three Types of Spina Bifida
 - Myelomeningocele
 - Meningocele
 - Spina Bifida Occulta




3

Initial Neurogenic Bladder & Bowel Management

- Neurogenic Bladder
 - Clean Intermittent Catheterization (CIC) per urethra
 - Indwelling urethral catheter (Foley)
 - Suprapubic tube (SPT)
 - Risks
 - Other options
- Neurogenic Bowel
 - Medications
 - Cone Enema
 - Other options



4

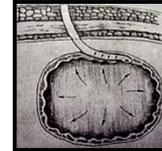
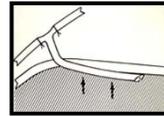
Common Surgical Procedures/Anatomy of SB Patients

- Urinary Tract
 - Continent Catheterizable Channel
 - Bladder Augmentation
 - Urinary Diversion +/- Cystectomy
 - Artificial Urinary Sphincter (AUS)
 - Bladder Neck Procedures
- Gastrointestinal
 - Malone Antegrade Contience Enema (MACE)



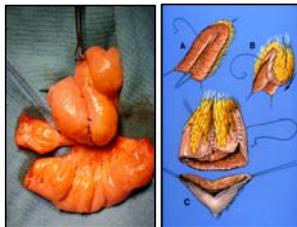
Continent Catheterizable Channel

- Mitrofanoff principle (MONTI)
- Appendicovesicostomy (APV)



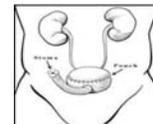
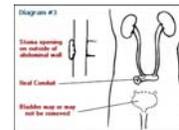
Bladder Augmentation

- Why?
 - Small Bladder Capacity
 - Non-compliant bladder
- How?
 - Bowel/Stomach (rare) piece of tissue is sown to native bladder



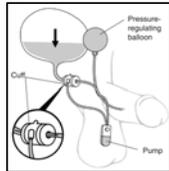
Urinary Diversions and Cystectomy

- Ileal & Colon Conduits
- Indiana pouch
- Removal of bladder



Artificial Urinary Sphincter & Bladder Neck Procedures

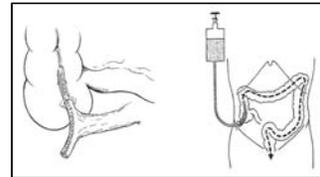
- First used in 1973
- AMS 800 prosthesis (American Medical Systems, Minnetonka, MRN)
- Bladder sling
- Bladder neck closure



9

MACE

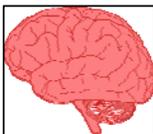
- When other conservative management has failed
- Help restore normal bowel function
- Manage constipation
- Gain fecal continence



10

Transition Readiness Assessment Questionnaire

- Validated Tool
- Cognitive Development



(Roth, 2019)



11

Audience Poll

- Responses are:
 - 1. No, I do not know how
 - 2. No, but I want to learn
 - 3. No, but I am learning to do this
 - 4. Yes, I have started doing this
 - 5. Yes, I always do this when I need to

(Roth, 2019)



12

Transition Readiness Assessment Questionnaire (TRAQ)

- Do you know what to do if you are having a bad reaction to your medications?
- Do you follow up on any referral for tests, checks-ups or labs?
- Do you call the doctor about unusual changes in your health? (For example: Allergic reactions?)
- Do you apply for health insurance if you lose your current coverage?
- Do you know what your health insurance covers?
- Do you manage your money & budget household expenses? (For example: use checking/debit card)
- Do you keep a calendar or list of medical and other appointments?
- Do you make a list of questions before the doctor's visit?
- Do you tell the doctor or nurse what you are feeling?
- Do you answer questions that are asked by the doctor, nurse, or clinic staff?

(Roth, 2019)



13

TRAQ SB Results

- Cross-Sectional Study from August 2017-April 2018
- 43 adult patients 18-25 years old participated over 9 months
- Average age 21
- 46.5% were Female
- 3.0 average daily medications taken
- 72.1% shunted hydrocephalus
- 90.7% myelomeningocele
- 72.1% had catheterizable channels or MACE
- 48.8% had bladder augmentation
- 25.6% no genitourinary reconstructive surgery

(Roth, 2019)



14

TRAQ Control/College Students Results

- 100 total participants
- Average age 20
- 53% were female
- 0 daily medications



(Roth, 2019)



15

Expecting too much?

- No significant difference between TRAQ scores
- "Appointment Keeping"
- "Readiness to transition"
- SB patients more likely to answer "No, I don't know how" r/t transitional tasks
- SB patients and college students not fully prepared for transition
- Compared to other adults with chronic health diseases, SB adults had lower TRAQ scores → less ready to transition

(Roth, 2019)



16

The Importance of Adult Transition Clinic



17

Importance of Clinic

- 7 in every 10,000 births
- > 166,000 people living currently with MMC
- 85% survival rate into adulthood
- Largest group of adults with congenital urologic problems in the United States
- Renal function of aging patient
- Only pediatric urology group in the state of Indiana
- 40 percent of our own pediatric patients failed to transition

(Roth, 2019)(Szymanski, 2017)(Misseri, 2015)



18

Risk Factors For Transition

- Minority Race (African American, Hispanic)
- Low Income
- English as second language

(Wood, 2016)



19

Setting Appropriate Transitional Expectations

- Current urologic status
- Explore the patient's goals, options, limitations
- New or persistent concern
- Testing
- Treatment options

(Wood, 2016)



20

Life Long Follow Up

- Yearly urological visit
- Renal bladder ultrasound
- Kidneys, ureters, bladder (KUB) plain xray
- Serum lab work panel
 - Complete metabolic panel
 - Complete blood count
 - Cystatin C
 - Vitamin B 12
- If eGFR is:
 - **60-90**
 - voiding/cath diary check compliance
 - Ultrasound to check for renal changes (hydronephrosis)
 - Urodynamics
 - See every 6 months
 - **<60 referral to Nephrology**
- Urinary tract changes



21

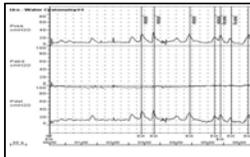
Cystoscopy Recommendations

- Surveillance of Bladder Augmentations
 - Gastric Augmentation: Yearly
 - Colon Augmentation: Age 50 then every 10 years
- Other Reasons
 - Recurrent Hematuria
 - Recurrent UTI
 - Issues with accessing channel / difficulty cathing the channel



22

Urodynamics



- Recommended if:
 - 3 or more UTI's per year
 - Change in continence/incontinence
 - Desire for more or difference continence
 - Change in ultrasound i.e. new or worsening hydronephrosis
 - Status post any urologic procedures
 - Botox
 - Bladder neck



23

Quality of Life

- Poor urinary and stool continence
- Skin breakdown
- Urinary Tract Infections
- Mental Health



(Wood, 2016)



24

High Risk Population

- Chronic Comorbidities
 - Diabetes
 - Hypertension
 - Hypercholesterinemia
 - Hypothyroidism
 - Obesity
- Peripheral vascular disease
- Pregnancies
- Mental health
- Sexual



25

Barriers to providing and receiving healthcare



- Transportation
- Insurance
- Communication of surgeries/anatomy
- Provider Comfort
- Aging Caregivers

(Wood, 2016)



26

What if no transition?

- Preventable complications
- 34% of hospital admissions were avoidable
- Cost of the preventable complications has been estimated around \$364 million dollars over a two year period

(Roth, 2019) (Wood, 2016)(Szymanski, 2018)



27

History of the Riley Adult Transitional Clinic

- 2007 – Founded by Dr. Rosalia Misseri, Urologist
- 2017 – Multidisciplinary format Urology, Physical Medicine and Rehab, Neurosurgery
- Currently serving around >250 unique adult patients including diagnosis of:
 - Spina Bifida
 - Cerebral Palsy
 - Bladder Extrophy
 - Cloaca Anomalies
 - Imperforate Anus
 - Posterior Urethral Valves
 - Prune Belly Syndrome



28

Current Adult Clinic Model

- Neurosurgery
- Physical Medicine and Rehab
- Social Work
- Urology



A Day In Clinic

- 3 Urological providers
 - 3 Urology RN's in clinic and to perform urodynamics
 - 2 Medical Assistants to room patients
- 1-2 Neurosurgical providers
- 1-2 Physical Medicine Physicians
- On call social worker (call when needed)
- Other Additions
 - Tables that lower to the ground to allow for easy transfer
 - Radiology located on same floor



Variety of Clinic Patients

- Spectrum of patients
- Pandora's Box
- Walkie-talkie versus inability to communicate
- Caregiver, extended care facility, living situations all different
- Multiple hats to wear during appointment



What was good then does not mean it works now



- Parents/Children
- Aging child, changing body habitus, worsening chronic health issues, dexterity issues
- Aging parent/Death of care giver
- Bladder Augmentation maintenance/cathing care
- Ileal conduit/empty bag



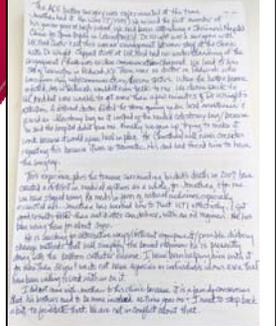
Goals for Clinic



- Increase and promote independence
- Maintain healthy kidneys and bladder
- Early detection of complications of prior interventions
- Improve quality of life
- Provide patient centered care



Patient Stories



- Patient being transported to clinic in family vehicle; stop suddenly; fell forward in wheel chair; broke both legs
- Patient with wound on foot for >2 years
- Aging 40's yo male patient; parents are in 70-80's, declining health for both patient and parents; emotional; difficult discussions
- Mother concerned for independence; didn't come to appointment but sent a handwritten letter
- Patient with recent bladder augmentation; parent moved patient group home; patient died



Riley Patient Testimonies

- **Has the Adult Transition Clinic (ATC) Improved your life? If yes, how/why?**
 - "It has saved my life since I was born"
 - "Yes, I get the specialty care I need."
 - "Yes"
 - "Yes helps to see different doctors at once"
 - "Yes, I get help when I need it by phone or clinic. Excellent health care"
 - "Yes! Much easier than multiple appointments multiple places"
- **What does the ATC do well?**
 - "They helped me by putting a vesicostomy in me."
 - "The care is excellent. The Dr's are very helpful."
 - "Everything"
 - "Doctor takes the time to listen and develop solutions."
- **What can the ATC do to improve?**



Riley Parent Testimonies

- **Has the Adult Transition Clinic (ATC) Improved your life? If you, how/why?**
 - "Helps my child by keeping on top of all kidney conditions"
 - "Yes"
 - "Yes more doctors in one place"
- **What does the ATC do well?**
 - "Caring staff"
 - "Makes it easy to see everyone at one visit"
 - "Thorough and take time with patients"
- **What can the ATC do to improve?**
 - "Nothing"
 - "None I can think of"
 - "Maybe more interaction with other patients, resources they need as adults"



References

- Centers for Disease and Control. Spina Bifida.
<https://www.cdc.gov/ncbddd/spinabifida/facts.html>
- Gearhart, Rink, RC, Mouriquand. Pediatric Urology 2014 p.740-750
- Koyle MA, Golda N, Hillis C, Oldershaw B et al. A proposed solution to a urological tightrope walk: The challenge of transition of spina bifida patients from pediatric to adult care in Ontario. Can Urol Assoc J 2016;10(9-10):306-10
- Misseri R. Approach to the myelodysplasia patient. Transition and lifelong care in congenital urology. 2015 p.11-20.
- Roth JD, Szymanski KM, Ferguson EJ, Cain MP, Misseri R. Transition young adults with neurogenic bladder—Are providers asking too much? Journal of Pediatric Urology 2019; 04.013
- Szymanski KM, Cain MP, Hardacker TJ, Misseri R. How successful is the transition to adult urology care in spina bifida? A single center 7-year experience. Journal of Pediatric Urology 2017;13:40.e1-40.e6.
- Urology Care Foundation
<https://www.urologyhealth.org/search?keywords=bladder+neck+closure&RowsPerPage=10&x=0&y=0>
- Wood HM, Loftus CJ. Congenital causes of neurogenic bladder and the transition to adult care



37

Questions & Answers



38