



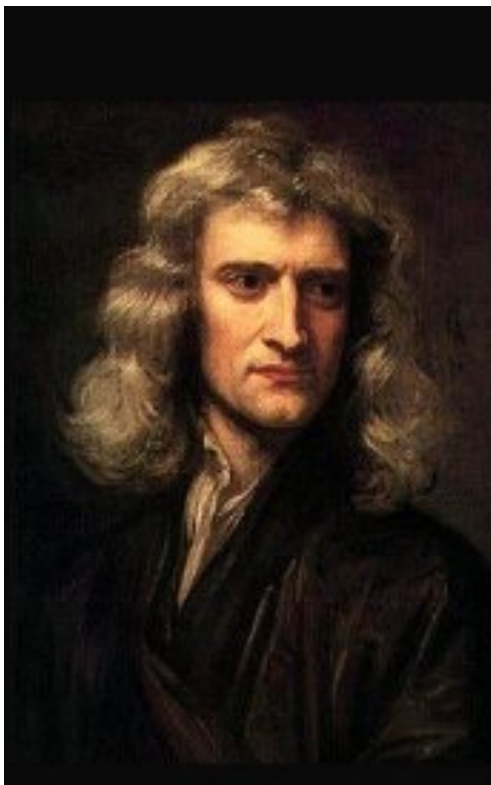
Bowel & Bladder Management in Spina Bifida

Brian VanderBrink, M.D.

Division of Urology

November 11, 2023





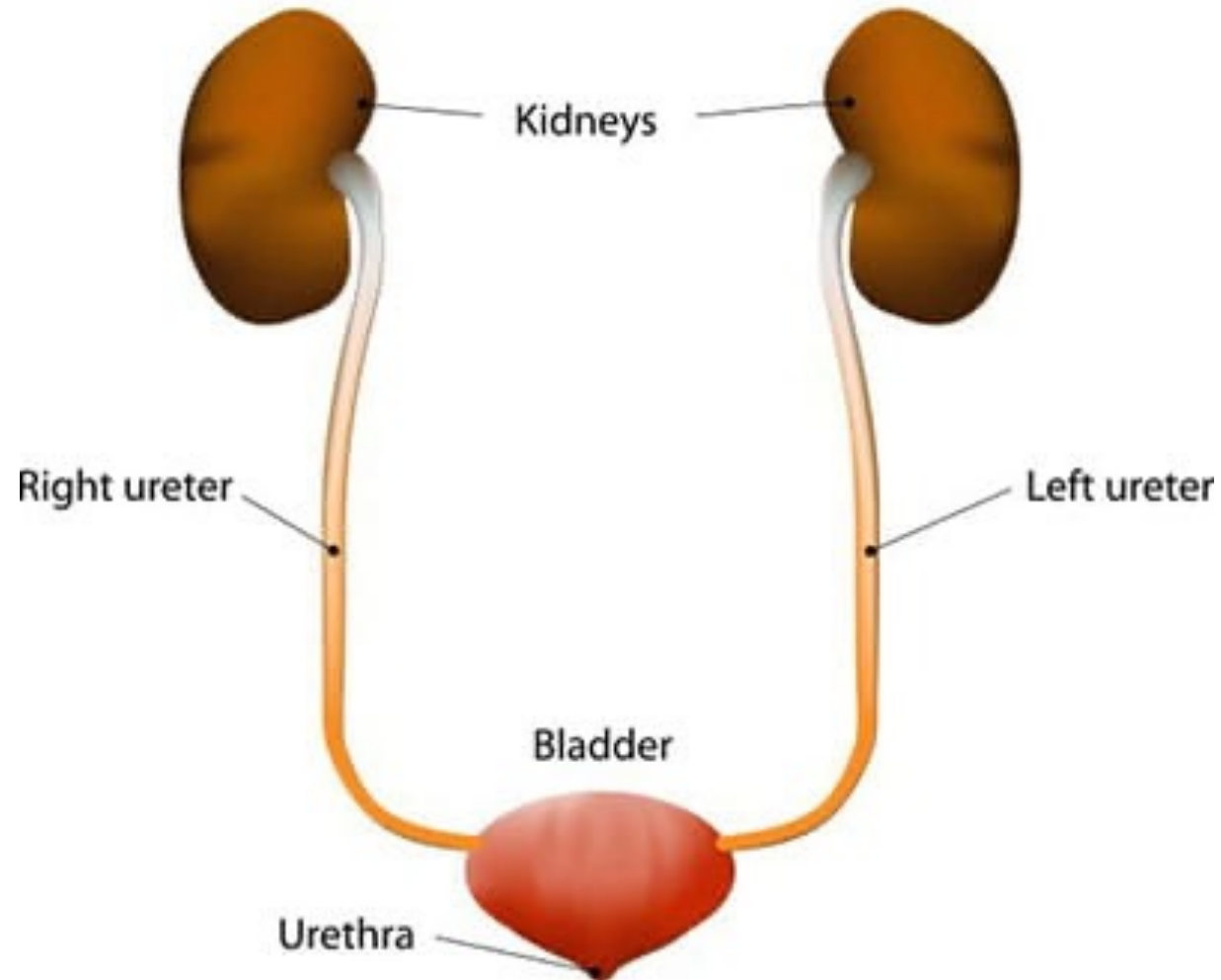
If I have seen further than others, it is by standing
upon the shoulders of giants.

(Isaac Newton)

Urinary Tract Anatomy

**The kidneys
are
essential
for life**

**The kidneys
are
connected
to the
bladder**

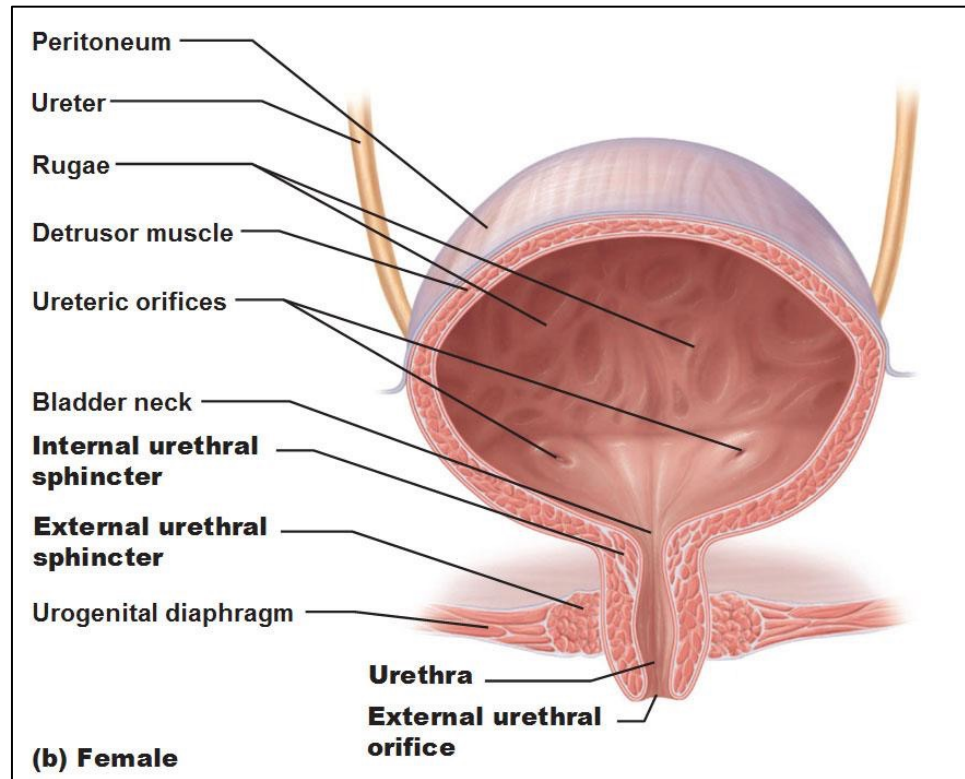


Bladder Function/ Urinary Continence

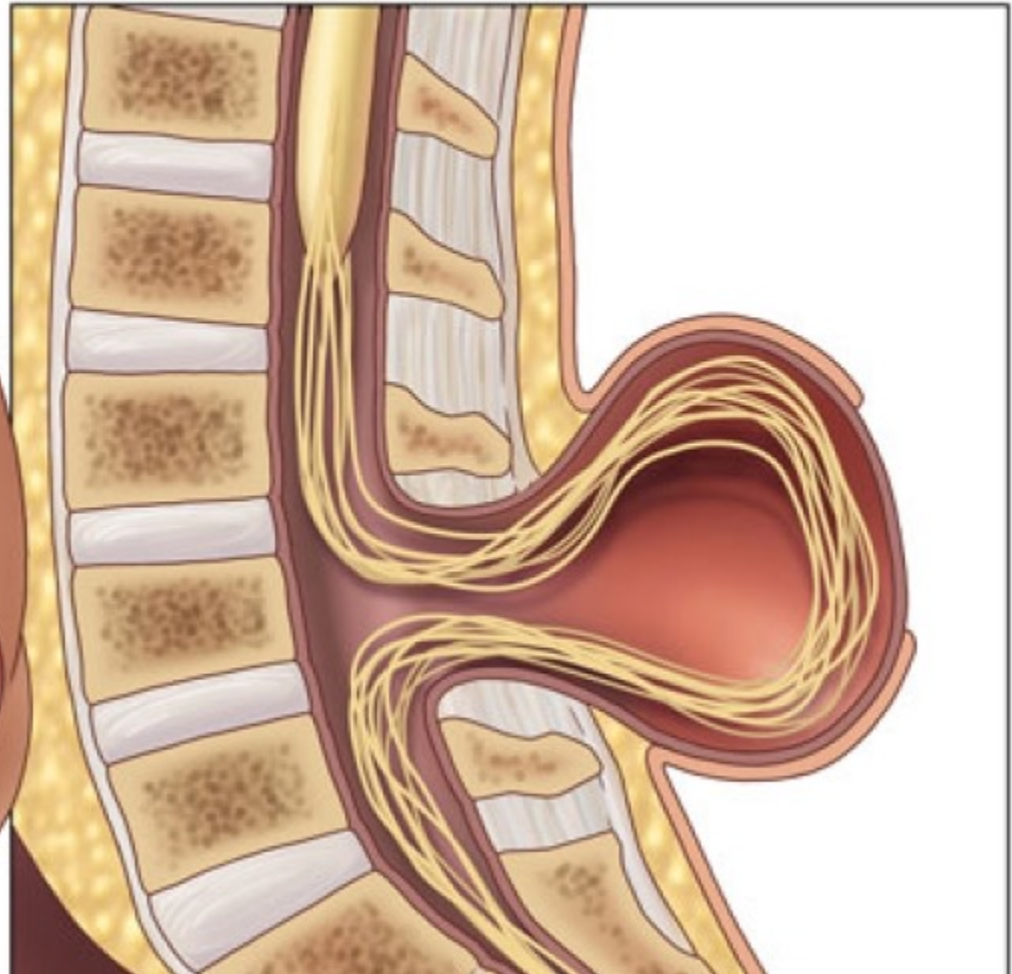
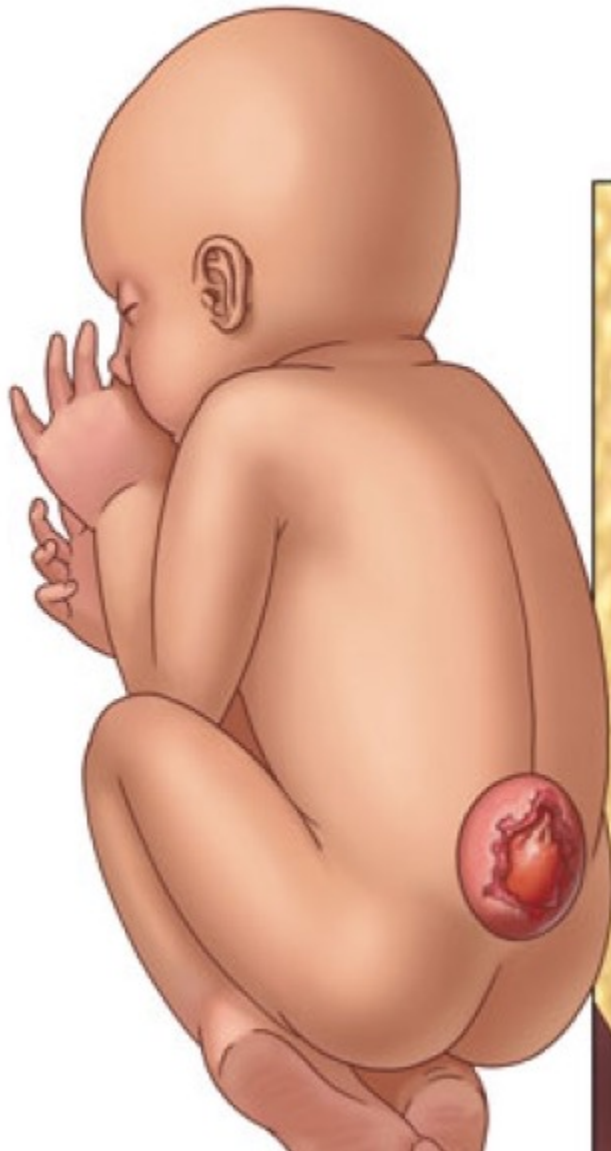
We need a sphincter that provides resistance to leakage of urine

We need a bladder big enough to store urine at low pressure to prevent back pressure to kidneys

We need coordination between the bladder muscle and sphincter muscle by nervous system



Bladder Innervation



Neurogenic Bladder

- What is it?
- The CDC Spina Bifida website defines it:
“Having a neurogenic bladder means an individual lacks bladder control due to a brain or nerve condition.”

Neurogenic Bladder

- Is there a cure?
- Because there are no current ways to fix nerve function that is damaged there is no cure
- Goals of any treatment should be to minimize its impact to health and lifestyle

Goals of Management of The Urological Tract

Appropriate Diagnosis of
Urological Conditions



Birth

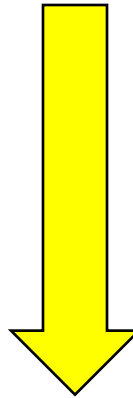
Education of Family

Initiate therapy **as indicated** to Protect
the Kidneys



Achieve Social Continence

Surgery **ONLY IF** needed



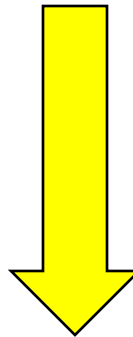
School Age



Enable self care



Maximize socialization, entry into
workforce & body image



Young Adult



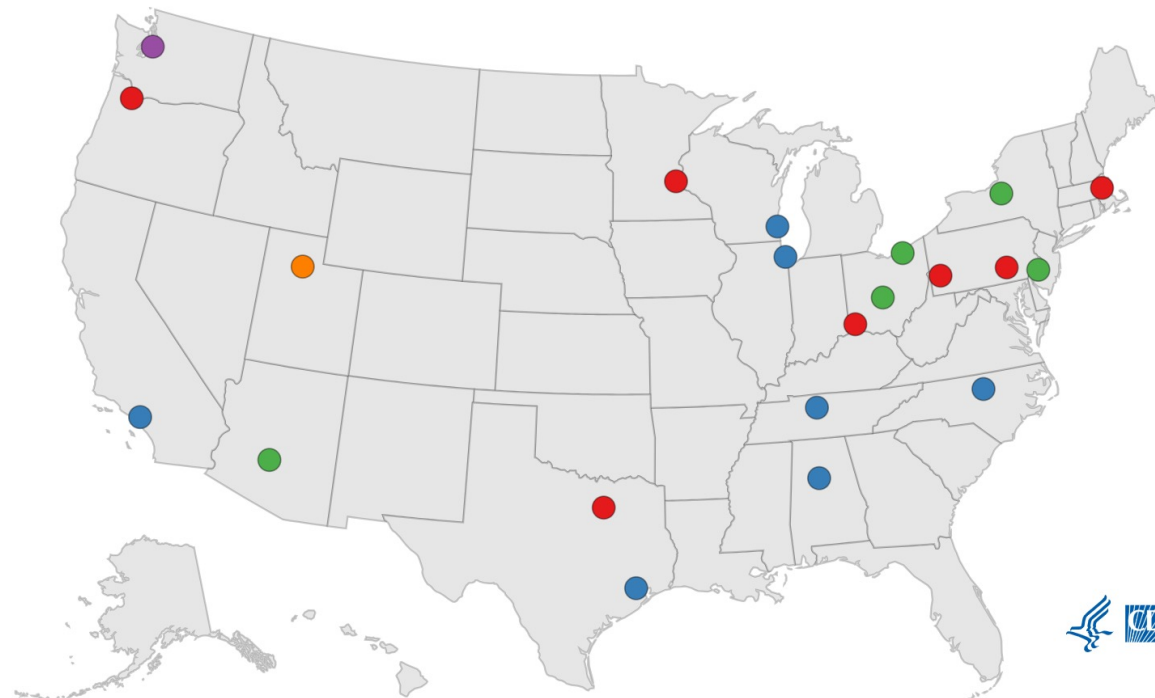
Individualized Care

- No two children are alike
- No two families are alike
- Treating all children the same is inappropriate



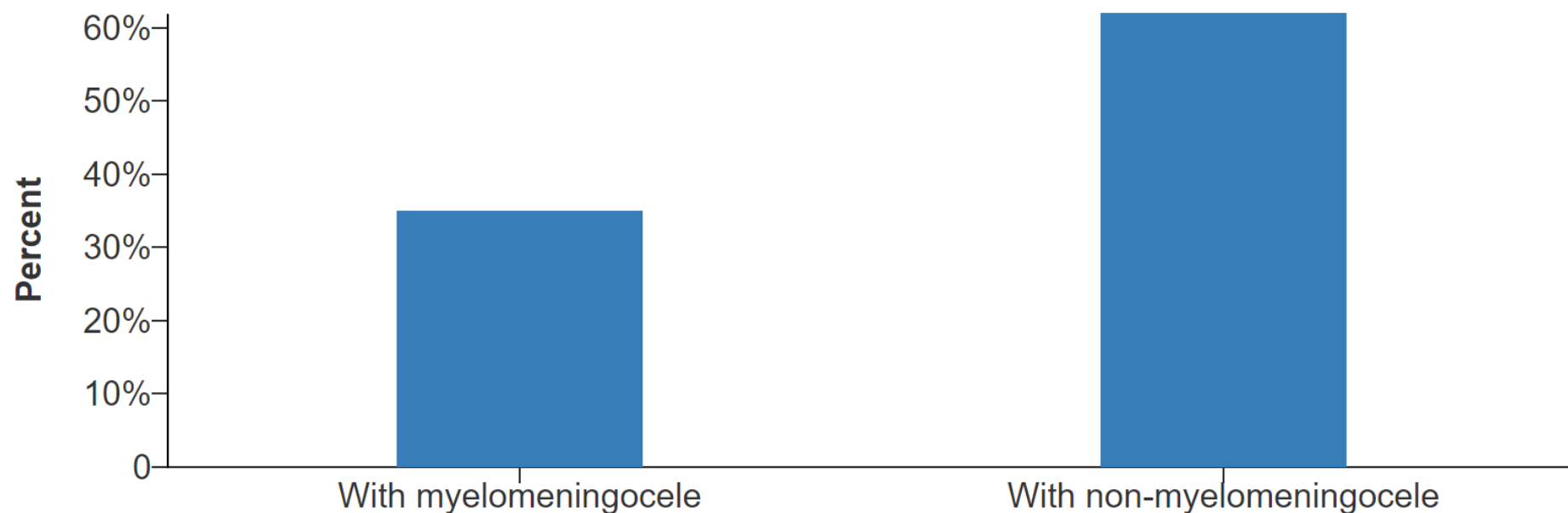
Spina Bifida Registry

- SBA established the National Spina Bifida Patient Registry (NSBPR) in concert with CDC
 - Survey of spina bifida clinics across the United States
- Information gathered:
 - Past surgeries
 - Continence
 - Mobility
 - Employment status
 - Education attained



Bladder Continence by Type of Spina Bifida (regardless of impairment)*

- Among Registry participants ages 5 years and older with myelomeningocele, 34% are continent of bladder.
- Among Registry participants ages 5 years and older with non-myelomeningocele, 62% are continent of bladder.



Source: <http://www.cdc.gov/ncbddd/spinabifida/nsbprregistryfindings.html>

Urology Management Options

- Surveillance/Observations Who decides which One?
Doctor?
Patient?
Parents?
- Medications
- Catheterizations
- Surgery



Beginning a Cath program – Who Decides?

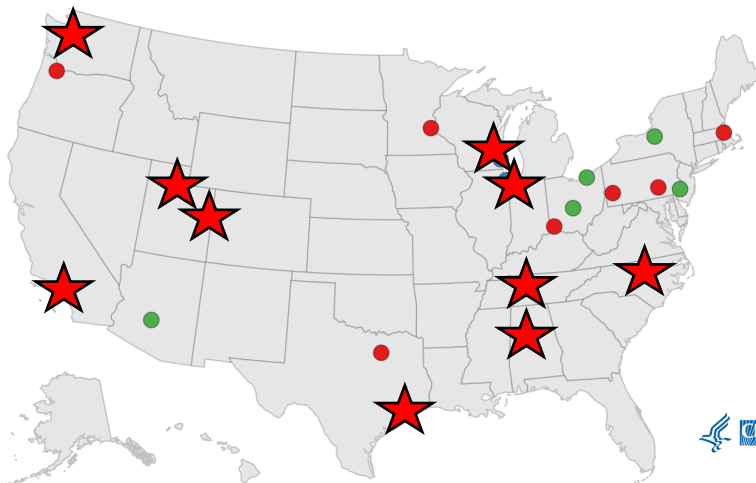
- Little controversy and resistance to understanding why to start
 - When kidneys are in peril
 - When UTIs happen
 - When urinary continence is not achieved and continence is desired
- Controversial; Physician and Institutional Bias; Parent Preference
 - Abnormal urodynamics that may threaten the kidneys
 - “Proactive” approach
 - The young child with no UTI, No problems on imaging, Too young to know if continence is achievable

Beginning a Cath program – Who Decides?

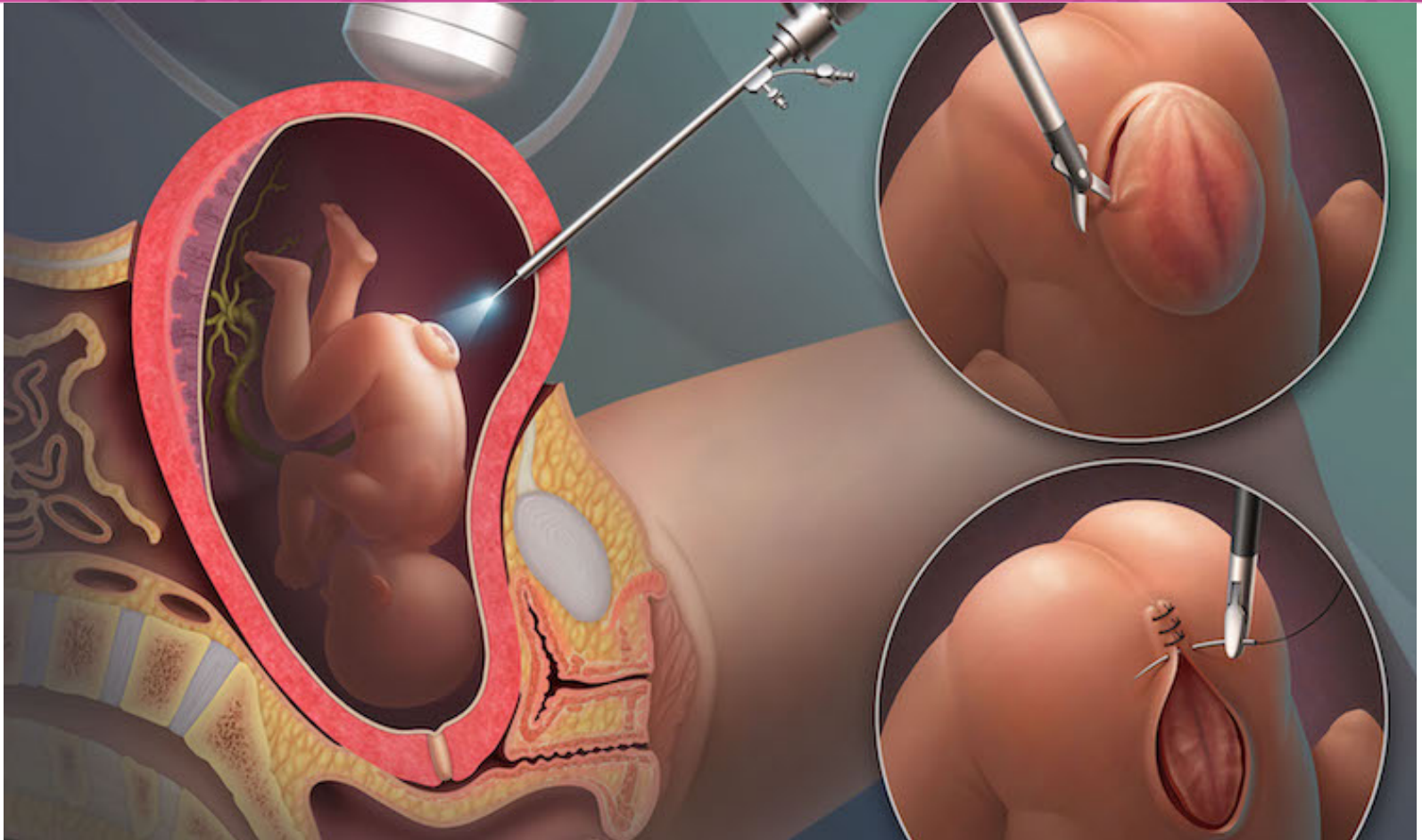
- Cincinnati Children's Approach – 2023
 - Shared decision making process but generally as Urologist in our Spina Bifida clinic I favor a proactive approach
- Personal observations
 - Easier for families to assimilate
 - Bladders requiring less surgical interventions/bladder augmentations compared to delayed interventions

UMPIRE Protocol

- Urologic Management to Preserve Initial Renal Function Protocol for Young Children with Spina Bifida (UMPIRE)
- UMPIRE program began in 2014 – 10 centers
 - Following the first group of infants until they are 10 years of age
 - Also adding more newborns with myelomeningocele as they are born



Fetal Surgery/Repair for Spina Bifida



How does bladder work with fetal repair?

Is Bladder better with Prenatal MM Repair?

Bladder Function After Fetal Surgery for Myelomeningocele

PEDIATRICS Volume 136, number 4, October 2015

- MOMS trial-Rates of Catheterization
 - 38% in Prenatal surgery group
 - 51% in Postnatal surgery group



Is Bladder better with Prenatal MM Repair?

Effect of Prenatal Repair of Myelomeningocele on Urological Outcomes at School Age



John W. Brock III, John C. Thomas,* Laurence S. Baskin, Stephen A. Zderic, Elizabeth A. Thom, Pamela K. Burrows, Hanmin Lee, Amy J. Houtrow, Cora MacPherson and N. Scott Adzick for the Eunice Kennedy Shriver NICHD MOMS Trial Group

Rates of Intermittent Catheterization

No./Total No. Prenatal Surgery (%)	No./Total No. Postnatal Surgery (%)
48/78 (61.5)	68/78 (87.2)

Bladder Management and Continence Outcomes in Adults with Spina Bifida: Results from the National Spina Bifida Patient Registry, 2009 to 2015

John S. Wiener,* Kristina D. Suson, Jonathan Castillo, Jonathan C. Routh, Stacy T. Tanaka, Tiebin Liu, Elisabeth A. Ward, Judy K. Thibadeau, David B. Joseph and the National Spina Bifida Patient Registry

	5 – 11 yr	12 – 19 yr	20+ yr	P-value
BLADDER MANAGEMENT				
NO MANAGEMENT (INCONTINENT)	12.6%	6.1%	5.0%	<0.0001
SPONTANEOUS VOID	19.8%	15.4%	10.4%	<0.0001
CLEAN INTERMITTENT CATH	64.6%	76.2%	76.2%	<0.0001
INDWELLING CATHETER	1.9%	1.8%	3.6%	0.0007
VESICOSTOMY	3.1%	1.8%	1.6%	0.0033
UROSTOMY BAG	0.1%	0.7%	4.2%	<0.0001
CREDE	0%	0.2%	0.5%	0.0239
CONDOM CATHETER	0%	0%	0.3%	0.0037

What to expect if Cath Program is started?

- Those who urinate normally don't do it 1-2 times/day
 - So don't expect typical recommendation to be 1-2 times a day cathing the bladder
- More regular emptying = Better bladder health
 - Less urine in bladder can lead to less UTI
 - Less urine in bladder means less leakage
 - Less urine can also mean less pressure on the kidneys

What to expect if Cath Program is started?

- Hopefully improvement in condition leading to starting it!
 - Decreased UTIs
 - Resolution of dilated kidneys and/or reflux
 - Less leakage
 - Improved renal function
- You will see urine and examine it....sometimes to a fault!
 - Challenging to have urine be crystal clear and “pure” when a plastic tube is placed several times a day into urinary tract

BACTERIA IN BLADDER FROM CATHING

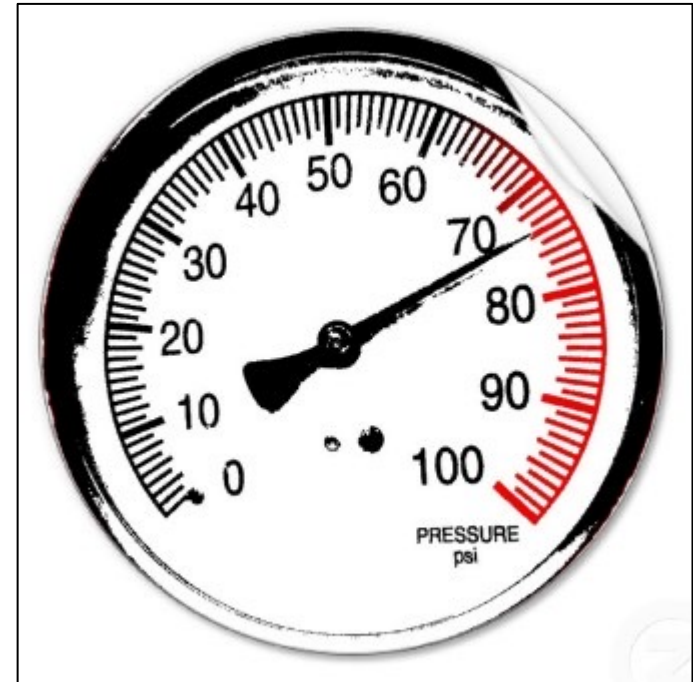
Bacteriuria in children with neurogenic bladder treated with intermittent catheterization: Natural history

- 14 spina bifida pts on CIC followed weekly with Urine cultures for 6 months
- **70% of cultures positive**
- Symptomatic UTI developed in 5/323 patient weeks during surveillance

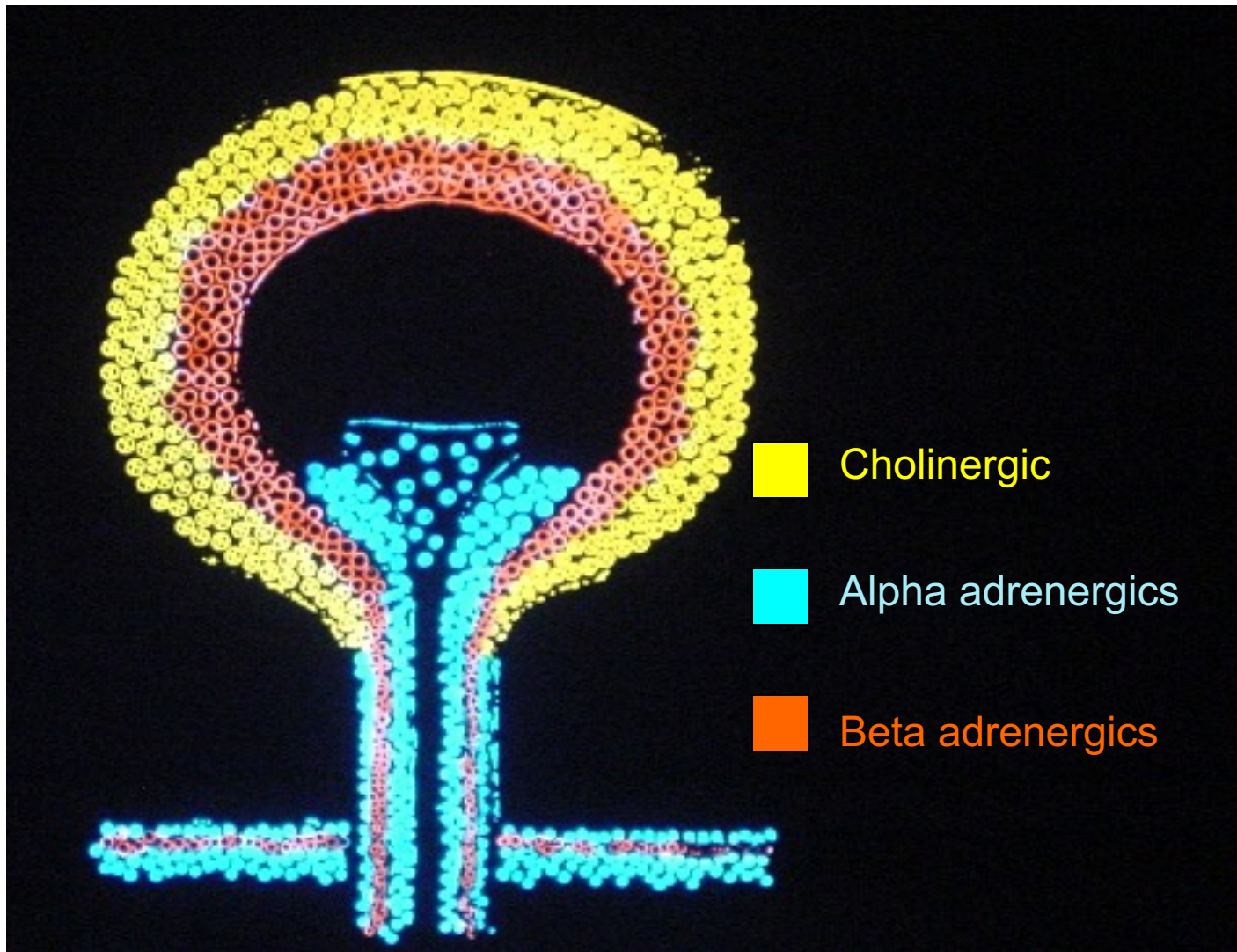
What if Cath Program alone doesn't fix “it”?

- Medications

- Bladder Pressures can hurt kidneys and bladder leading to change in function
- More urine in Bladder increases Bladder Pressure
- Frequent emptying can lower pressure.....
- But medication to lower pressure is needed at times because cathing cannot solve everything in every situation



Bladder Receptors



How do you lower pressures with oral meds?

- Anticholinergics

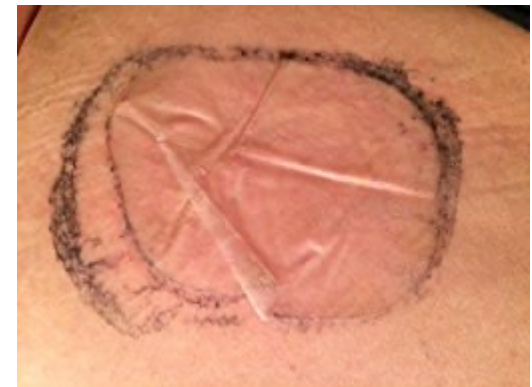
- Ditropan[®], Oxytrol[®],
Gelnique[®], Detrol[®],
Enablex[®], Vesicare[®],
Sanctura[®]



- Beta 3 Receptors

- Mybetriq[®], Gemtesa[®]

- These can be given by mouth, across skin or directly into the bladder



CHOLINERGIC BLADDER RECEPTORS

ANTICHOLINERGIC MEDICATIONS

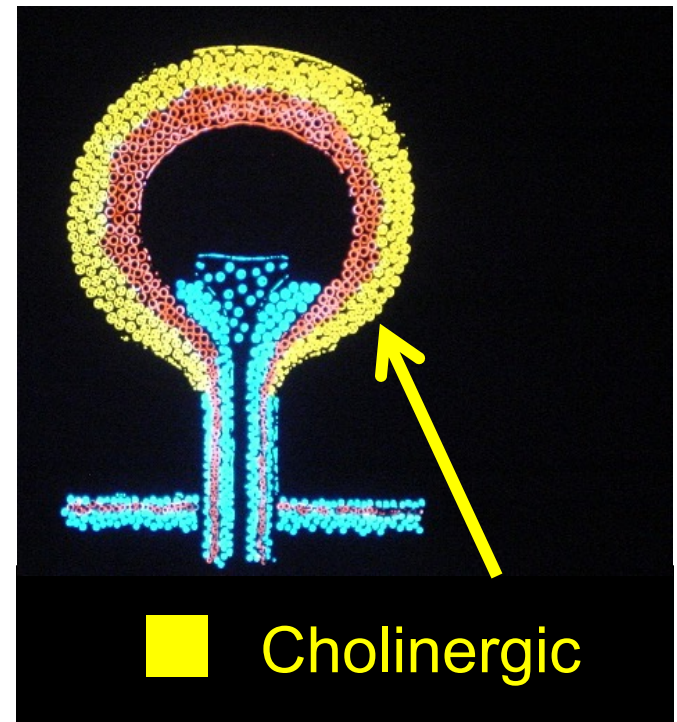
- **Stop contraction of the bladder muscle**

Examples:

Oxybutinin

Solifenacin

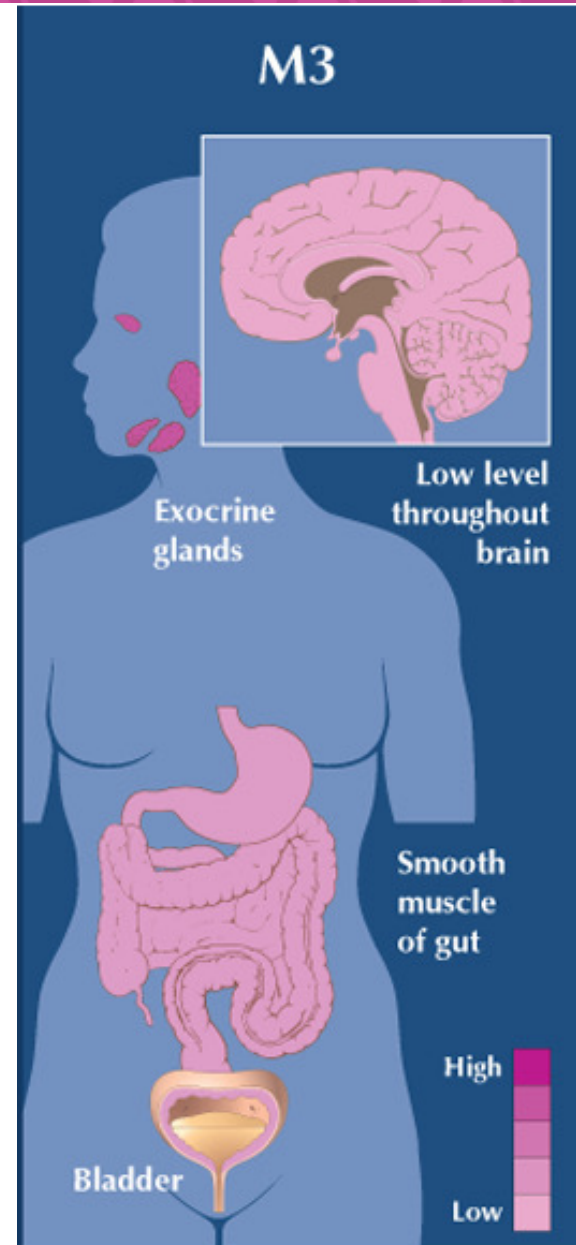
Tolterodine



Which medication is best?

- Need to try to find out which works best....remember no cookie cutters!
- One may work better and another may not
- One may give side effects and another may not
- That said Ditropan has longest track record and most commonly prescribed

Cholinergic Receptors – M type



I've heard that Ditropan can cause damage to my child?

- There is concern that the anticholinergic class of medications can have effects on the brain
 - Hard to ascertain in spina bifida where cognitive abilities may be altered from disease/VP shunt

Behavioral Effects of Long-Term Antimuscarinic Use in Patients with Spinal Dysraphism: A Case Control Study

THE JOURNAL OF UROLOGY®

Vol. 190, 2228-2232, December 2013

- 32 children from Holland divided between on medication and those without
 - No significant differences in behavior were found between children with and without long-term use of antimuscarinics



BETA-3 ADRENERGIC AGONISTS

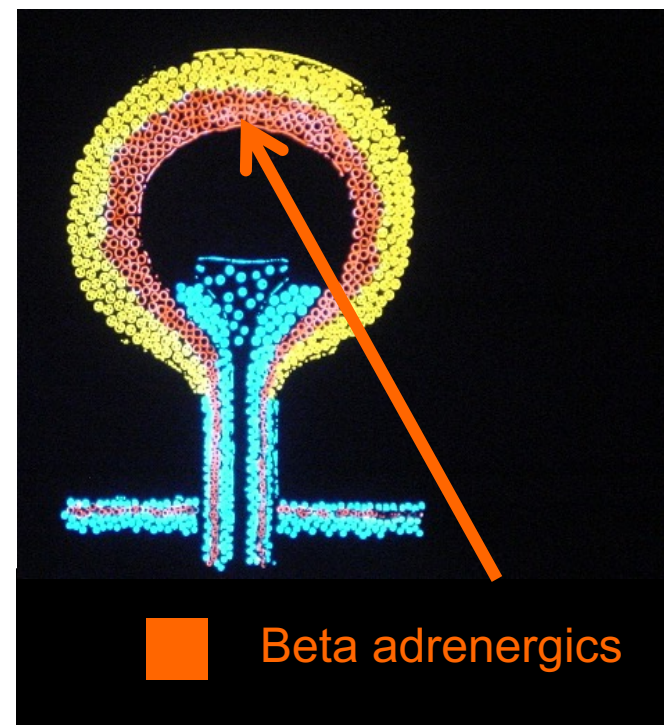
SYMPATHOMIMETIC MEDICATIONS

- **Increases relaxation of the bladder muscle**

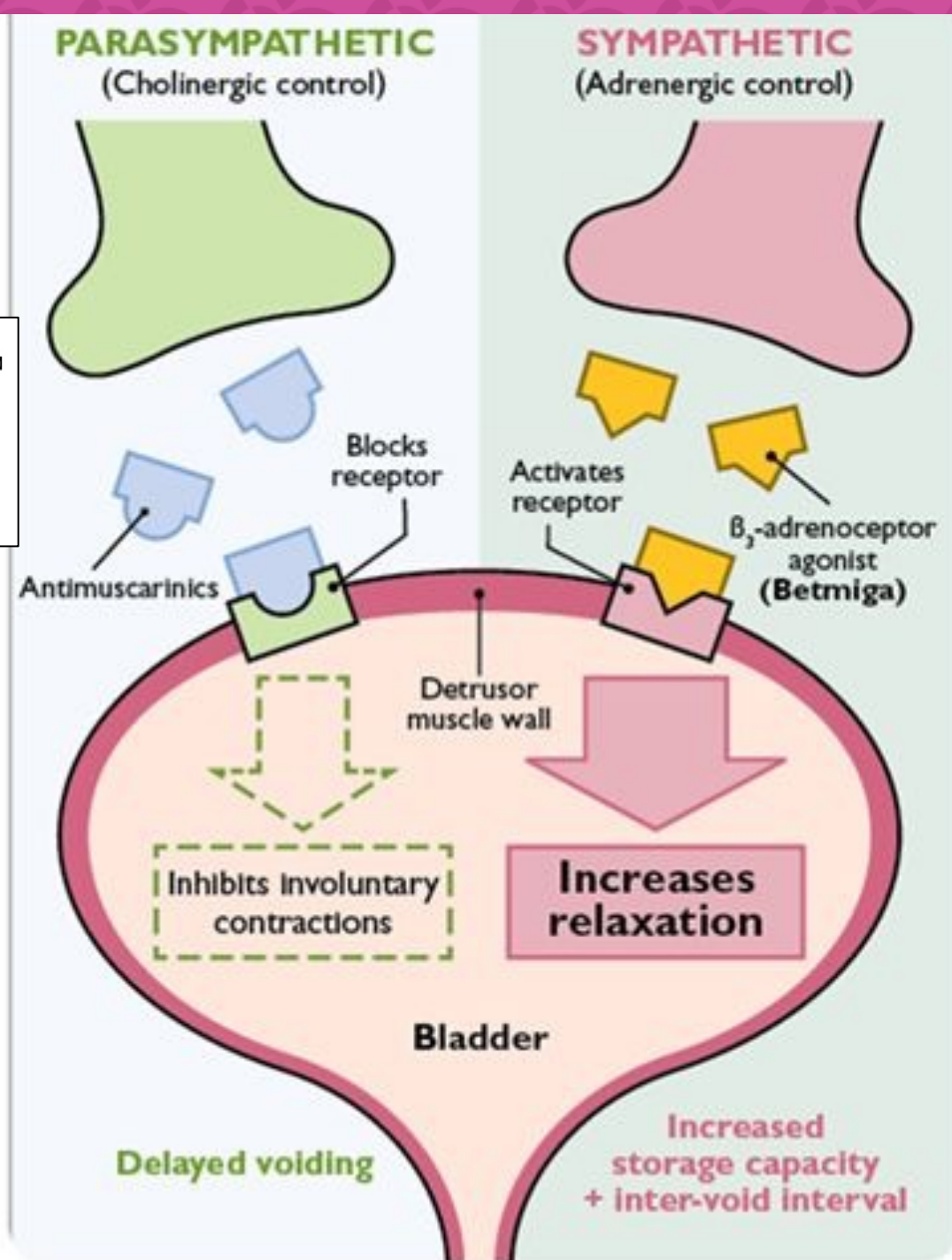
Examples:

Mirabegon

Virabegron



 MyrbetriqTM
mirabegron



Can we try more than one medication?

- 2023 study between Cincinnati Children's and CHOP (Philadelphia)



+



- Improvement was seen in many parameters:
 - Increased bladder capacity (30% increase from baseline)
 - Decreased Bladder Pressures (60% response)
 - Improved Urinary continence between caths (70% response)

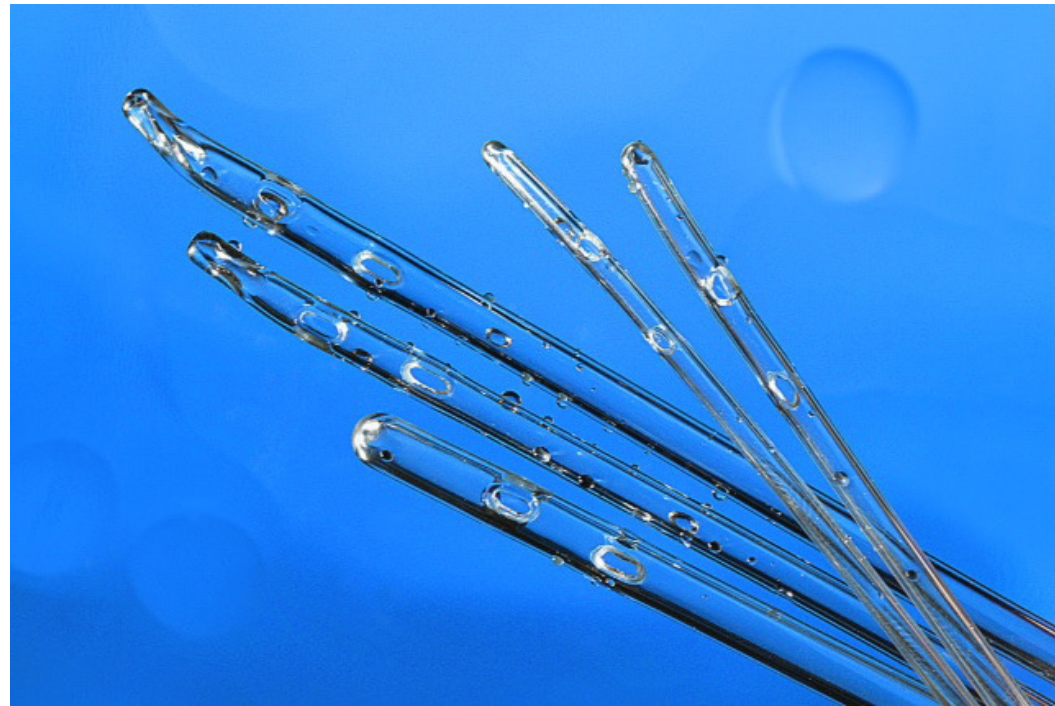
SYNERGY
 $1+1=3$

What if we aren't dry with cathing and medicines?

- Wait
 - Some kids may get drier as they get older
 - No way to reliably predict
- Make sure medication is optimized
 - Right dosage for weight, tolerability
- Make sure we are catheterizing regularly with correct size catheter

The right catheter size

- Appropriate size catheter will work best
 - We still don't use the same size toothbrush we learned to brush our teeth with
 - So why still use the same size catheter you learned to cath with as a baby!



The Colon and Bladder are Neighbors...

Sometimes the Colon is not a friendly Neighbor



Success!!!!

- If catheterization is needed or pursued and the goals are met with catheterizing through the urethra

- **NO UROLOGY SURGERY MAY BE NEEDED**

- When the child is older and mature enough they can be taught to catheterize themselves
 - If body habitus, developmental readiness and dexterity permit



How will we teach them to do it?

- Start Early

- Assess developmental appropriateness
- This is started too late too often



- Begin with have them gather supplies

- Allows them to recognize the multiple items needed to cath
- “What size catheter do we use?”
- “What kind of prep/wipe do we use?”

Fundamentals



SOAP AND WATER

The most underestimated way to kill germs!



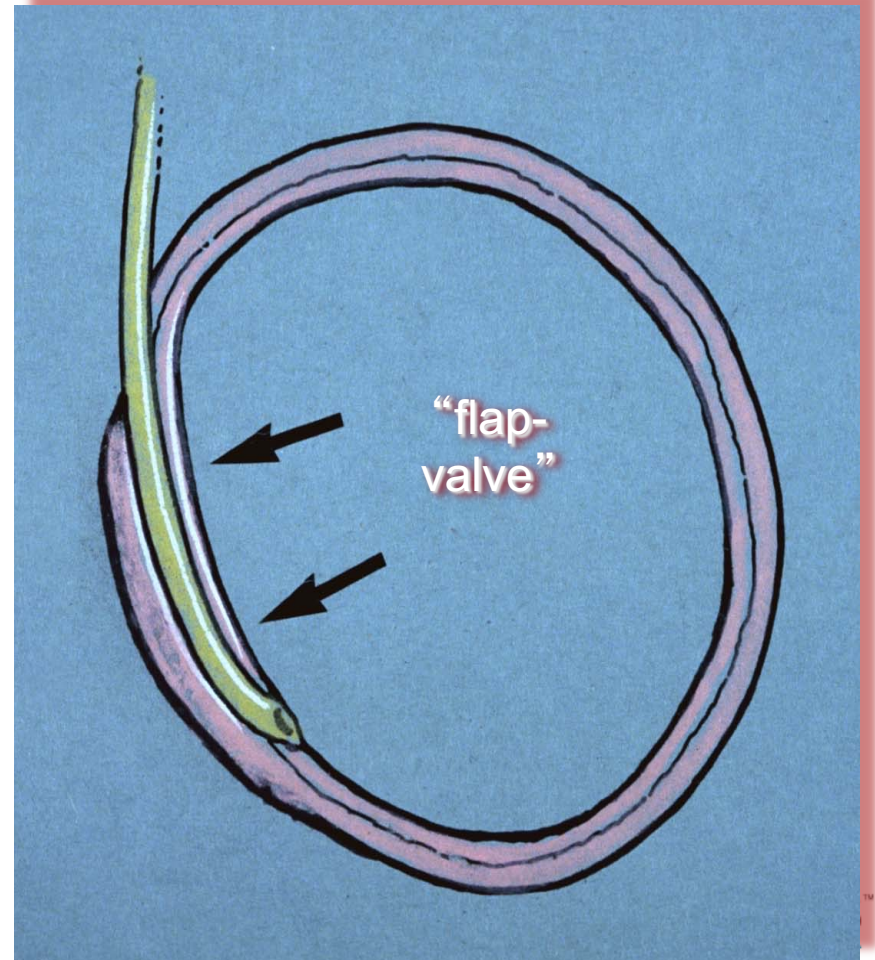
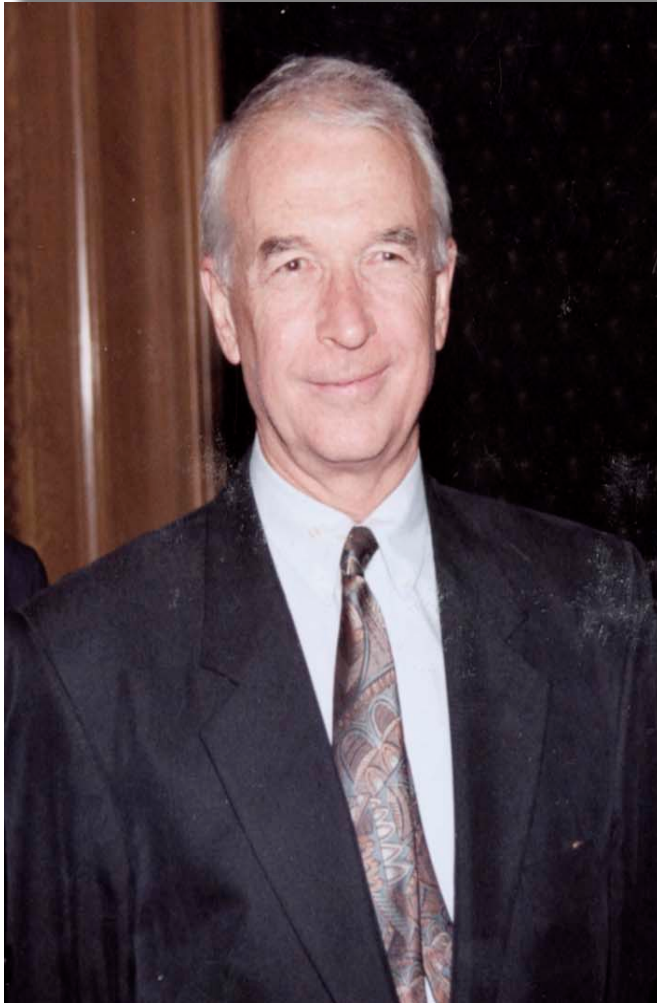
Fundamentals



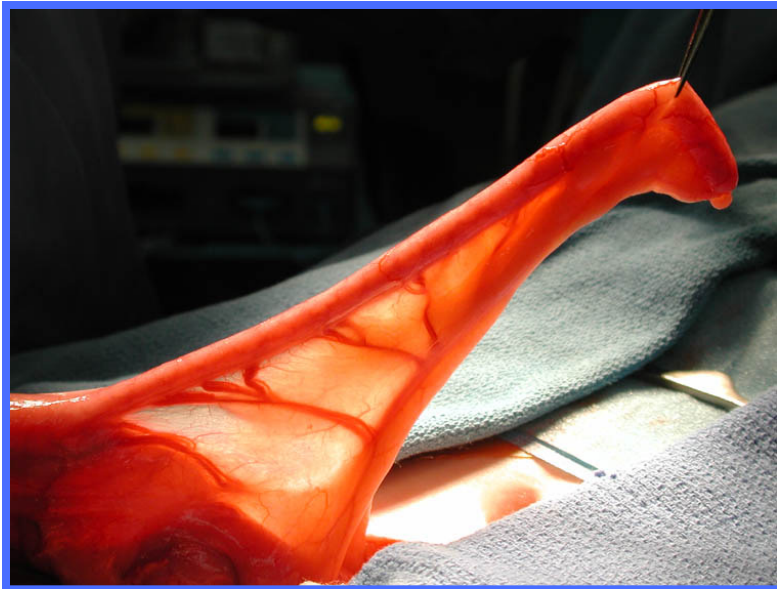
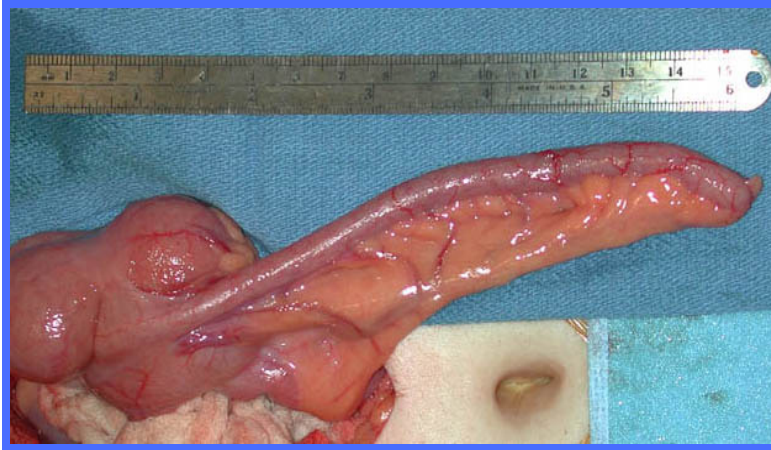
What if we can't teach them to cath per urethra?



MITROFANOFF 1980



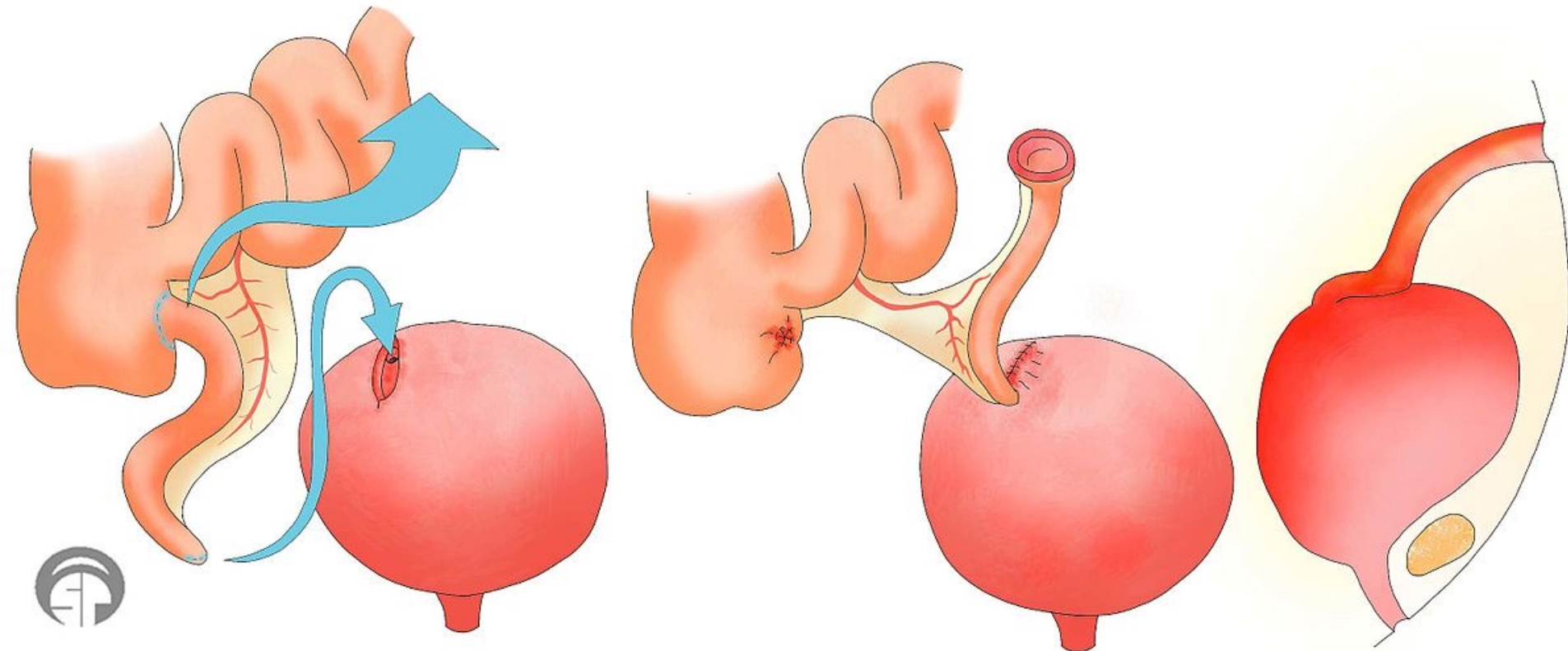
Appendix as the catheterizable channel “Gold Standard”



Advantages

- readily available
- already tubular
- good blood supply
- removal has no effect on bowel

APPENDICOVESICOSTOMY / MITROFANOFF PROCEDURE

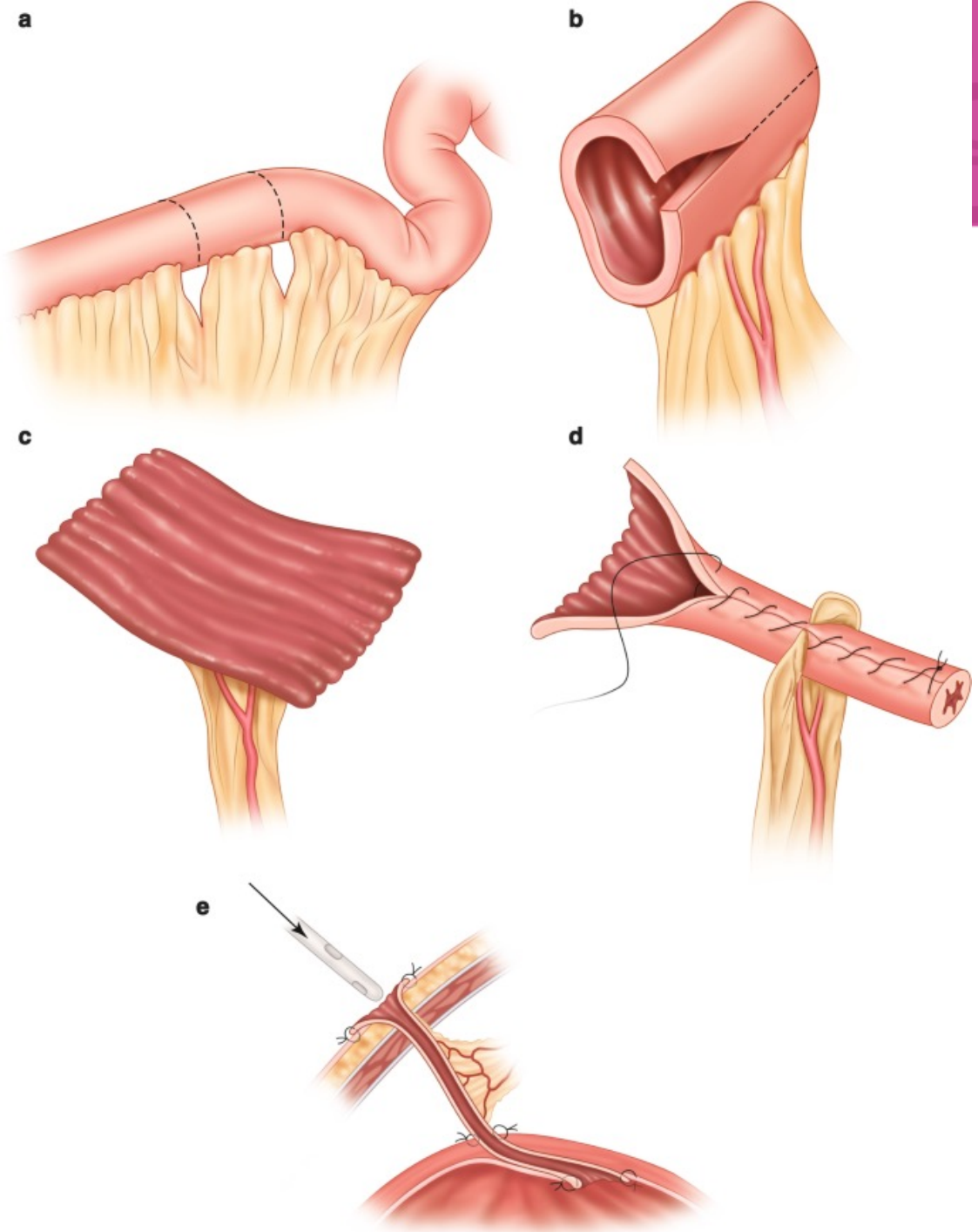
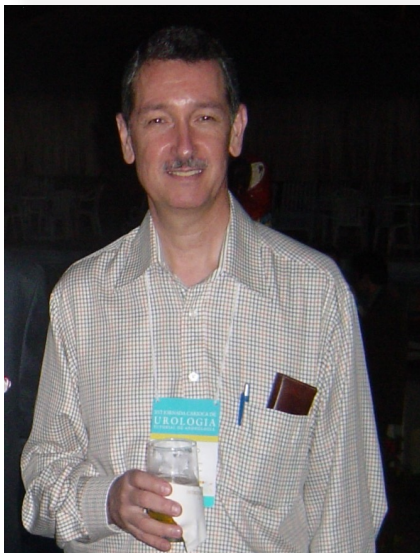


APPENDICOVESICOSTOMY

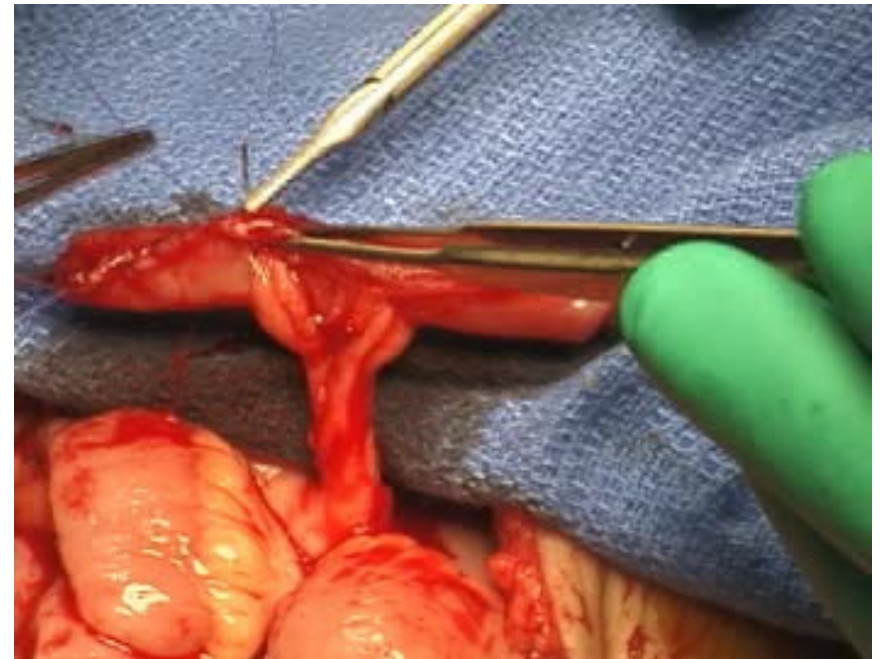
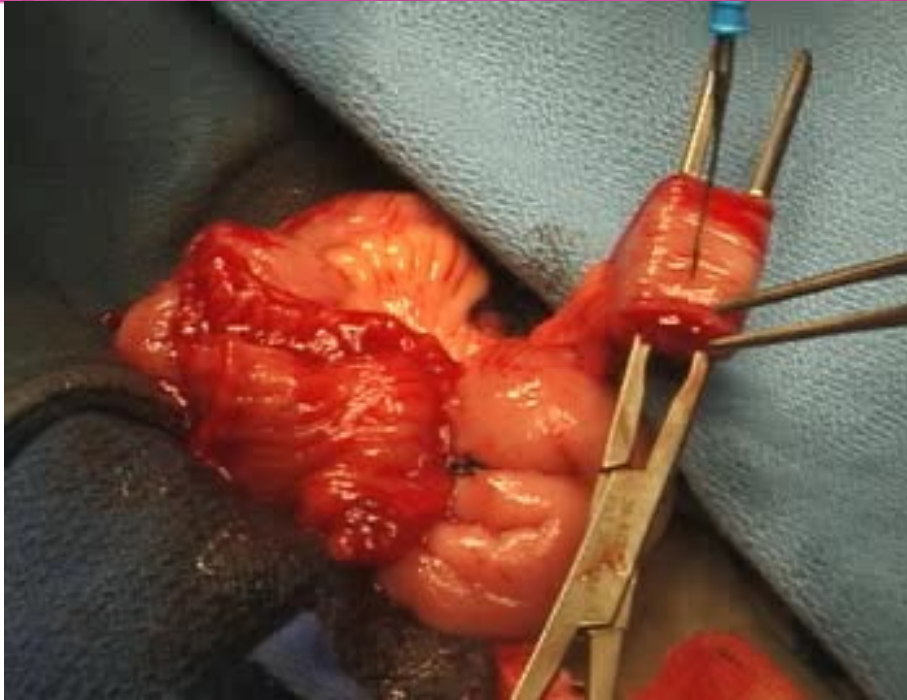


What do you do
if there is
No or insufficient length
appendix available?

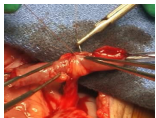
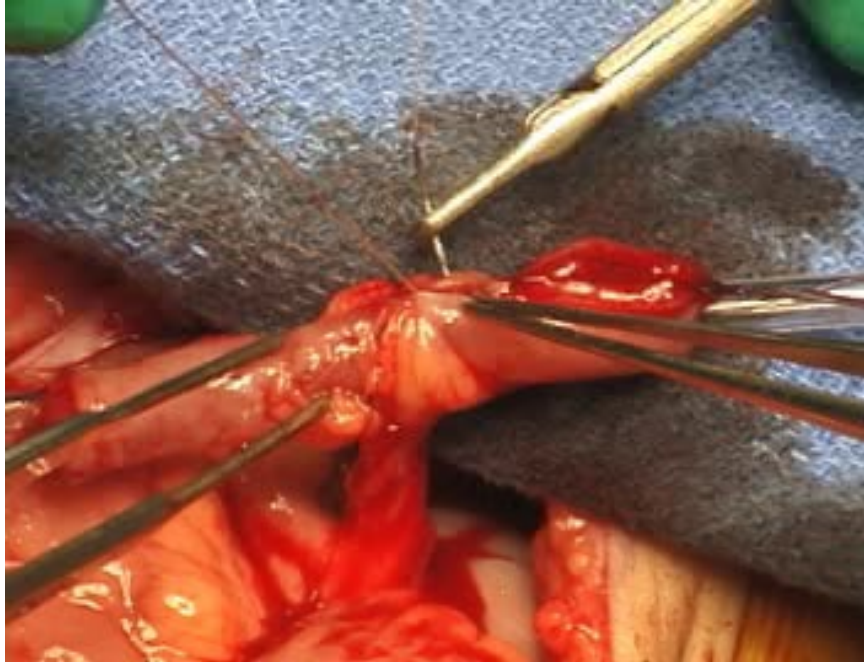
Monti Procedure



MONTI - YANG

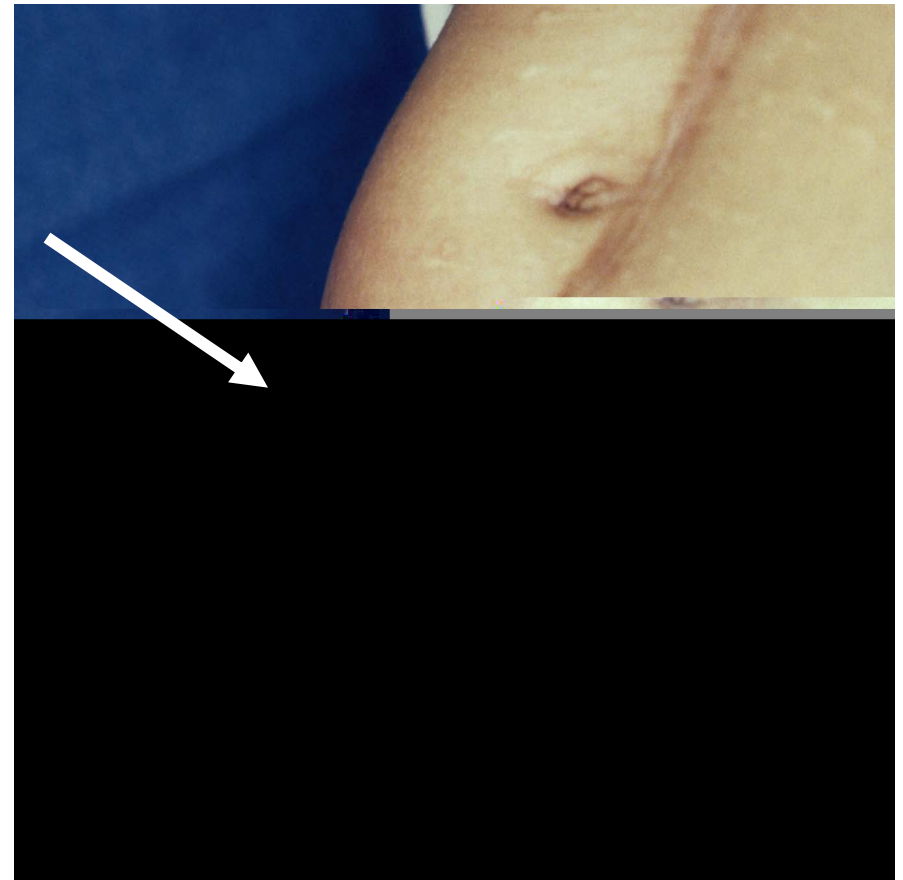


MONTI - YANG



What problems happen with a Mitrofanoff?

Complications



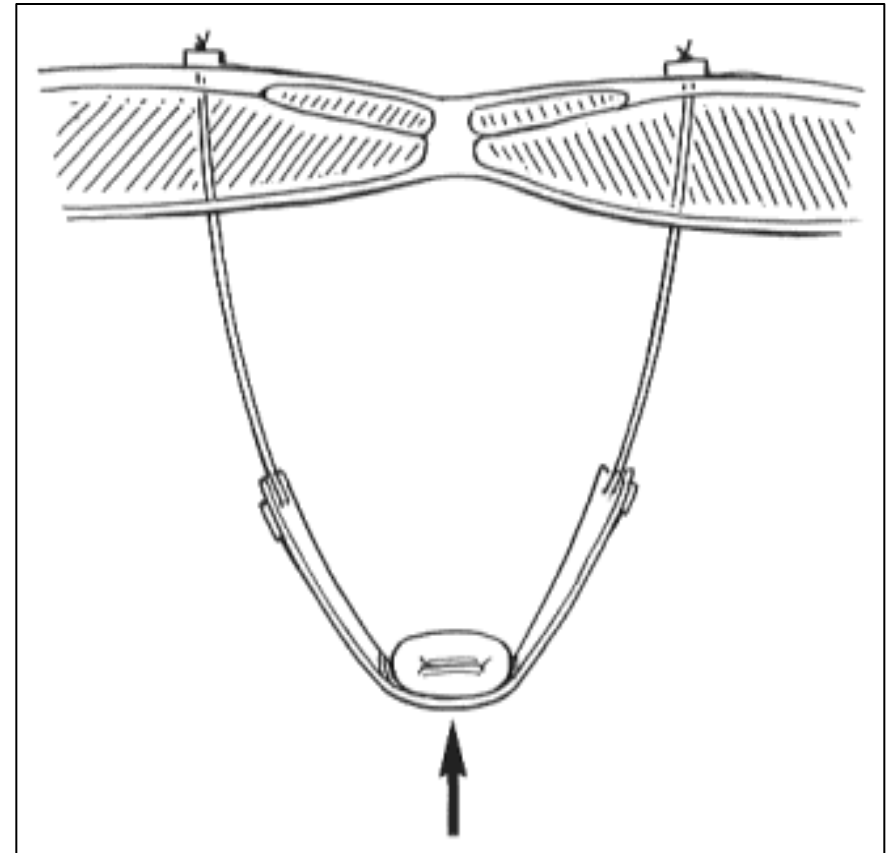
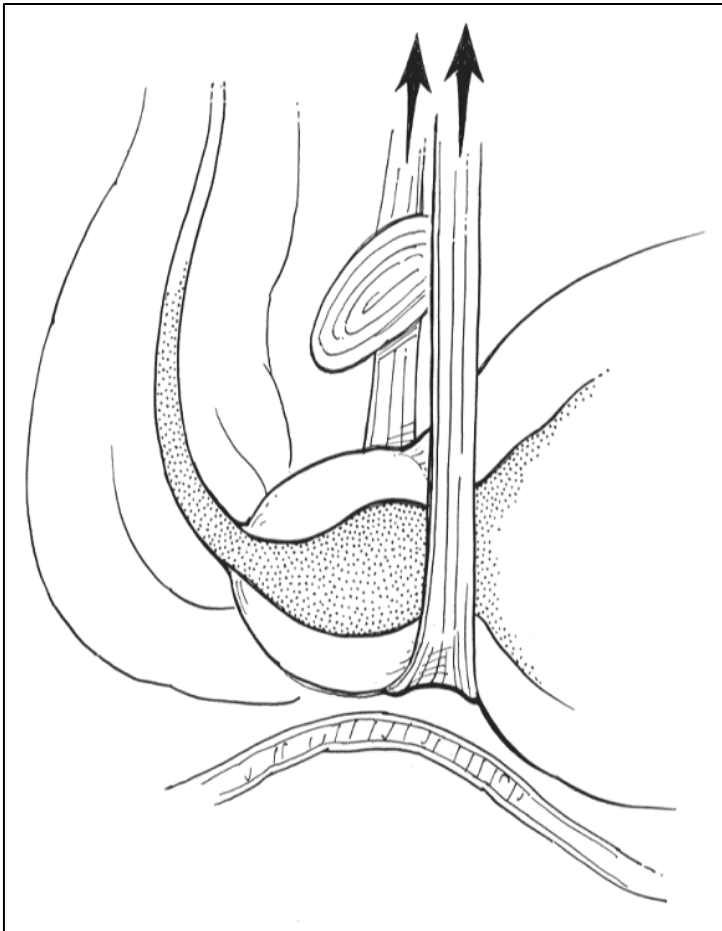
What if we still aren't dry and we want it NOW?

- Surgery
 - Bladder outlet/sphincter may not have enough resistance to keep urine in between caths
 - Bladder may not be large enough or low pressure enough to store urine to stay dry between caths

Bladder Outlet Procedures

- **Bladder Neck Sling**
- **Bladder Neck Reconstruction**
- **Artificial Urinary Sphincter**
- **Bladder Neck Closure**

Sling procedures

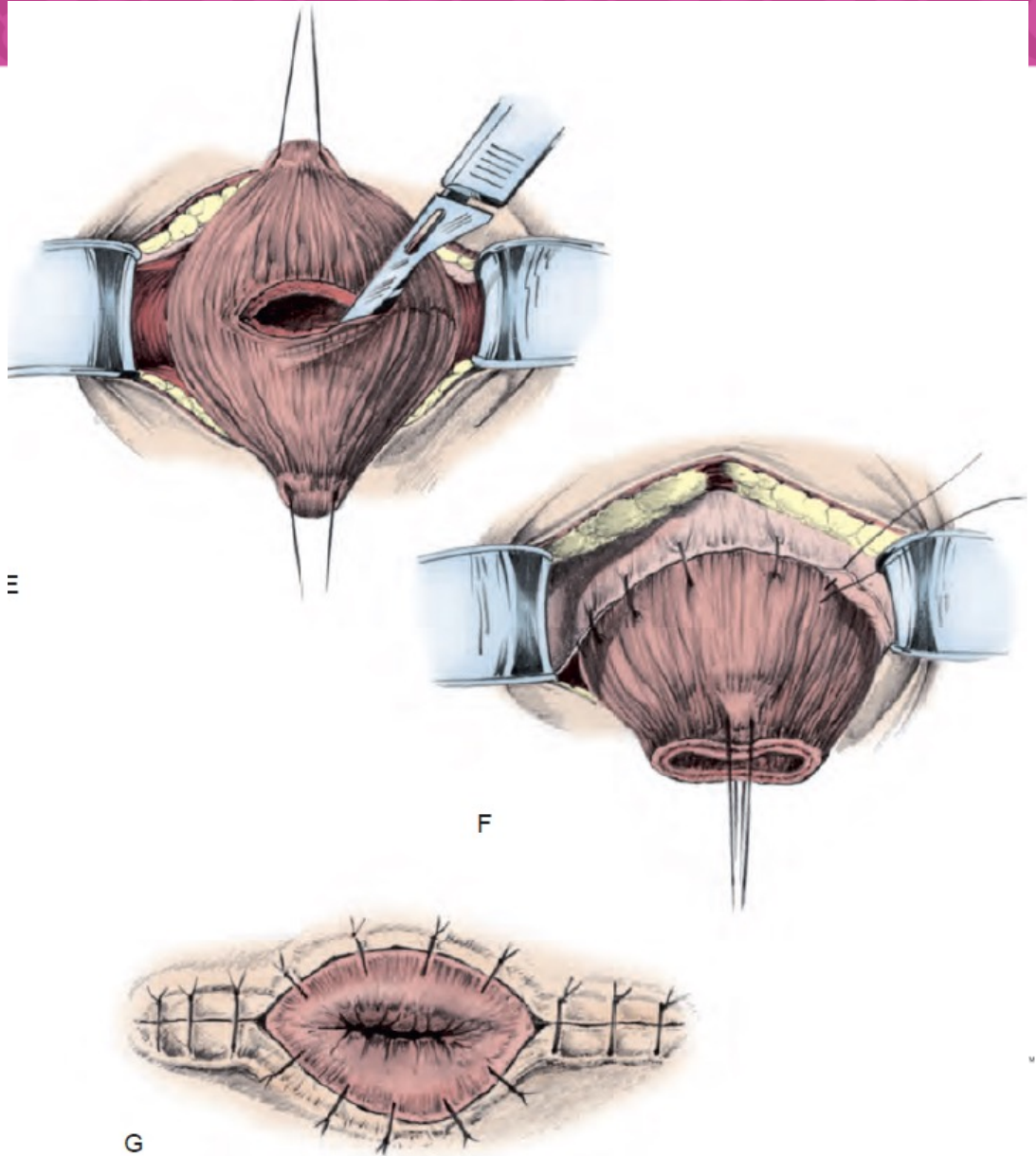


Surgeries Aimed at Lowering Bladder Pressures

- Vesicostomy
- Bladder BoTox
- Bladder Augmentation

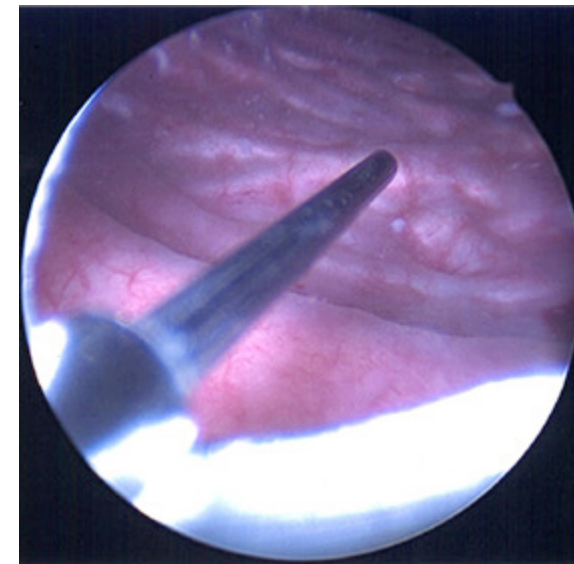
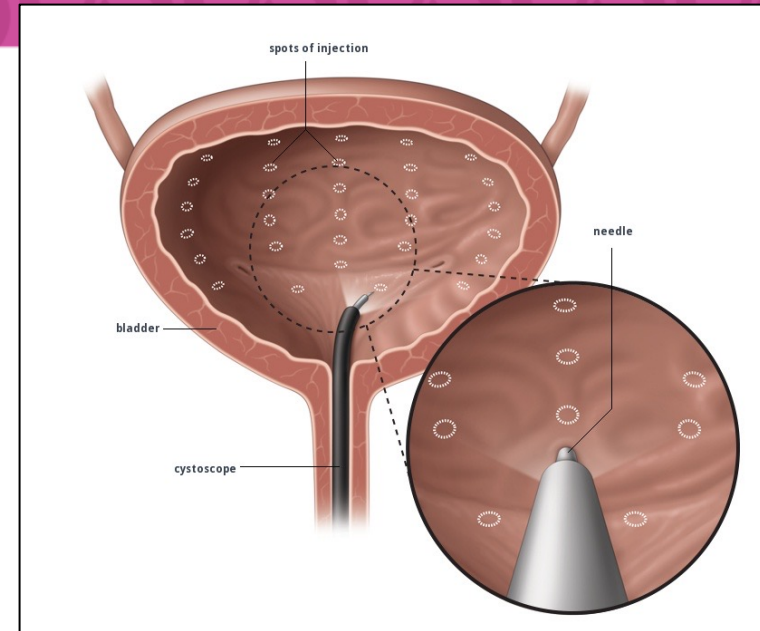
VESICOSTOMY

- Younger patients
- Diaper dependent
- Reversible



BOTOX

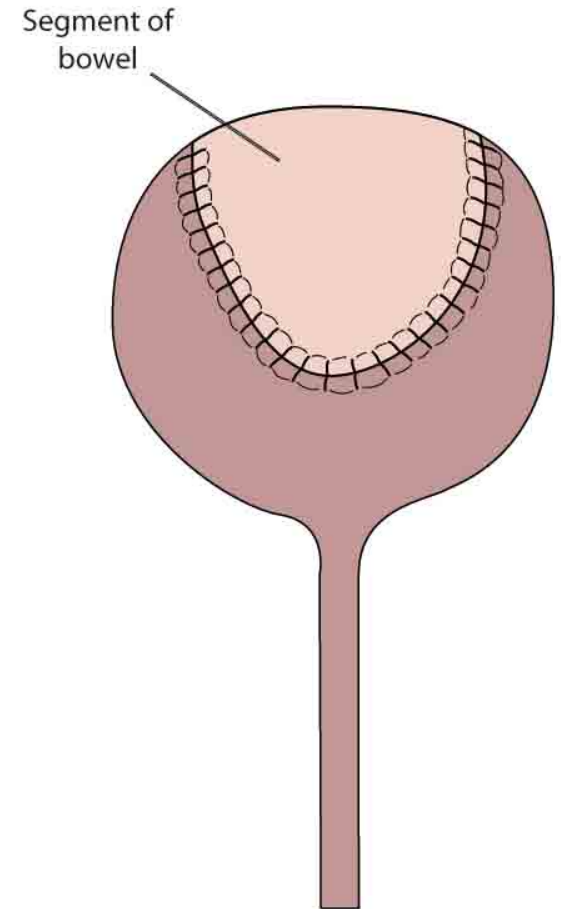
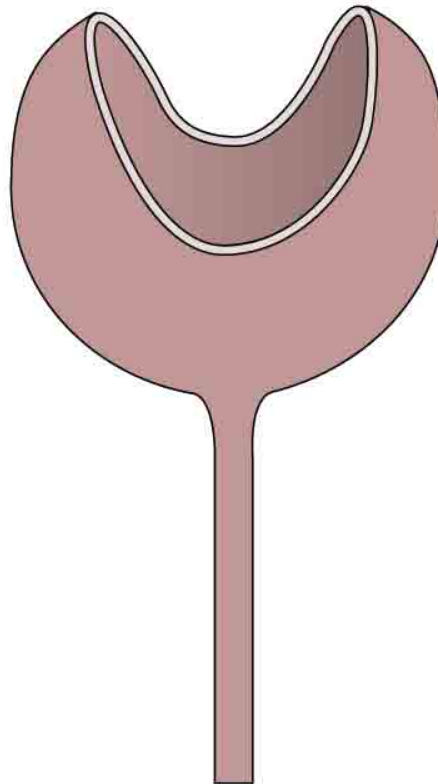
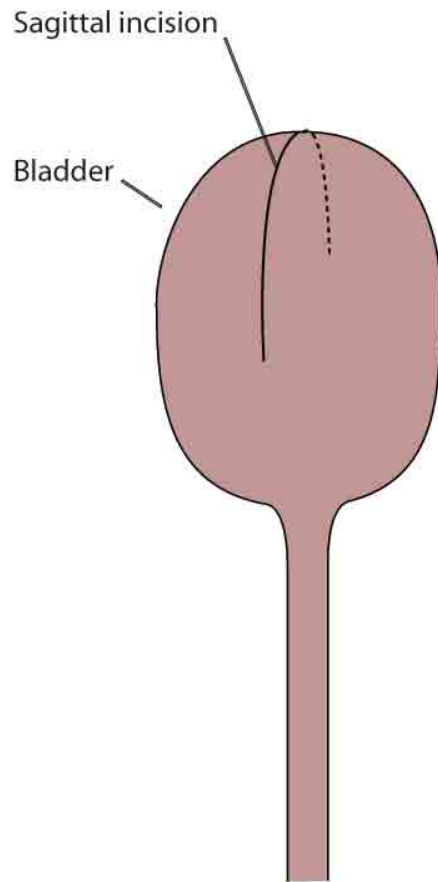
- Medicine injected directly into the bladder with a telescope into bladder (cystoscope)
 - Interferes with bladder muscle ability to contract
 - Wears off after 3-6 months (variable)
 - Need for catheterization in nearly all cases due to “paralysis” of bladder
- Can be repeated but requires cystoscope and this usually needs anesthesia in pediatric age group



BLADDER AUGMENTATION

- Used when medications unsuccessful
- Used when BoTox fails
- Very reliable method to increase capacity and lower pressure in bladder when maximal medication fails.....
 - ...But requires lifelong follow-up and need for additional surgery in many

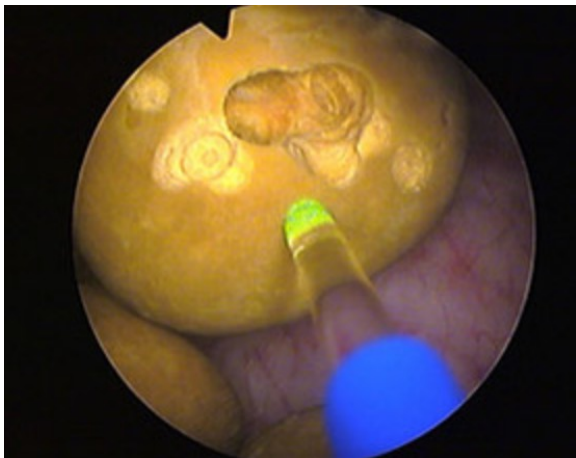
BLADDER AUGMENTATION



Bladder Augmentation

What problems happen with a Bladder Augment?

Complications

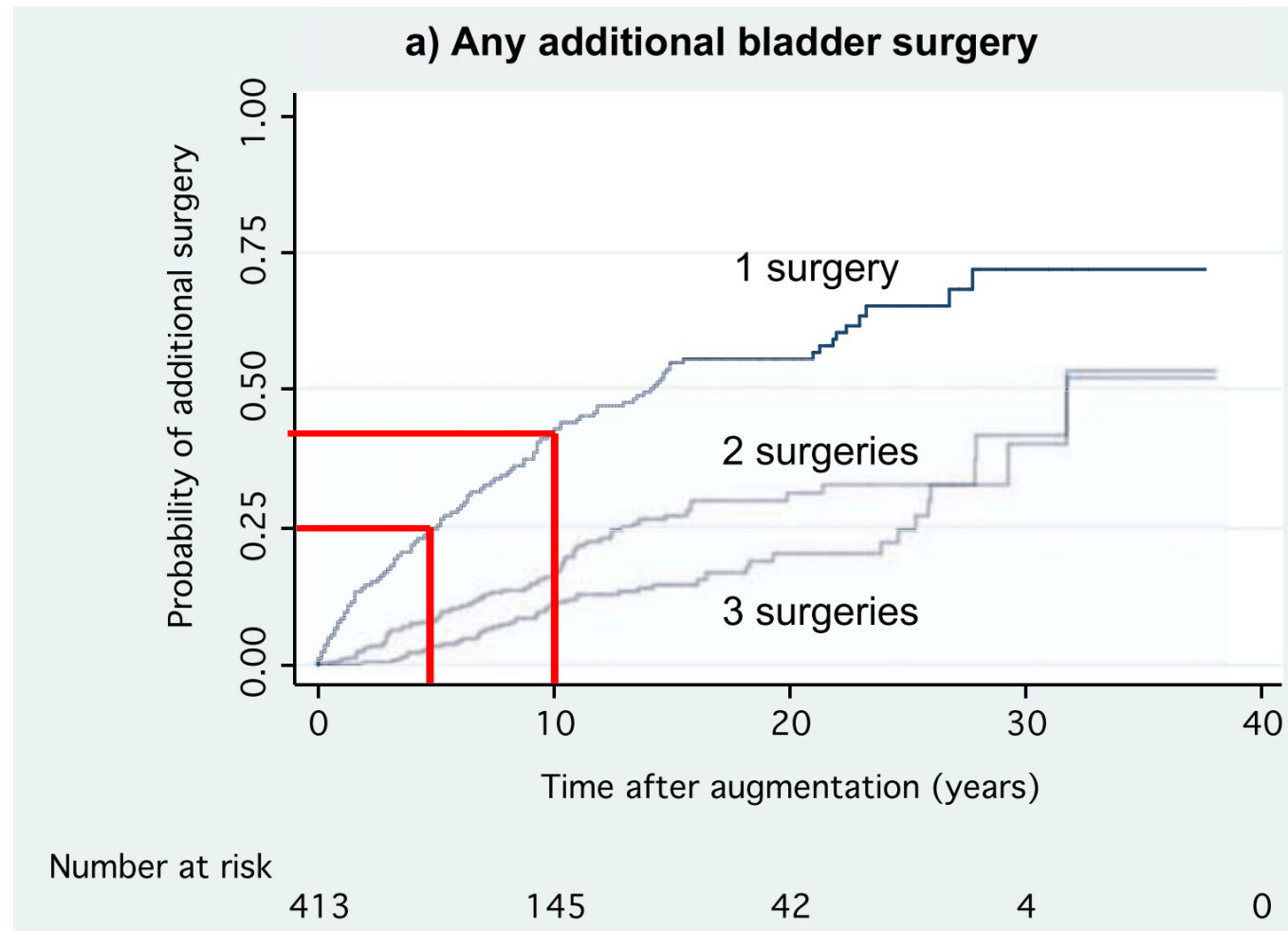


Additional Surgeries after Bladder Augmentation in Patients with Spina Bifida in the 21st Century

Konrad M. Szymanski,* Rosalia Misseri, Benjamin Whittam, Nathan Hollowell, Rachel E. Hardacker, Carly R. Swenson, Martin Kaefer, Richard C. Rink and Mark P. Cain

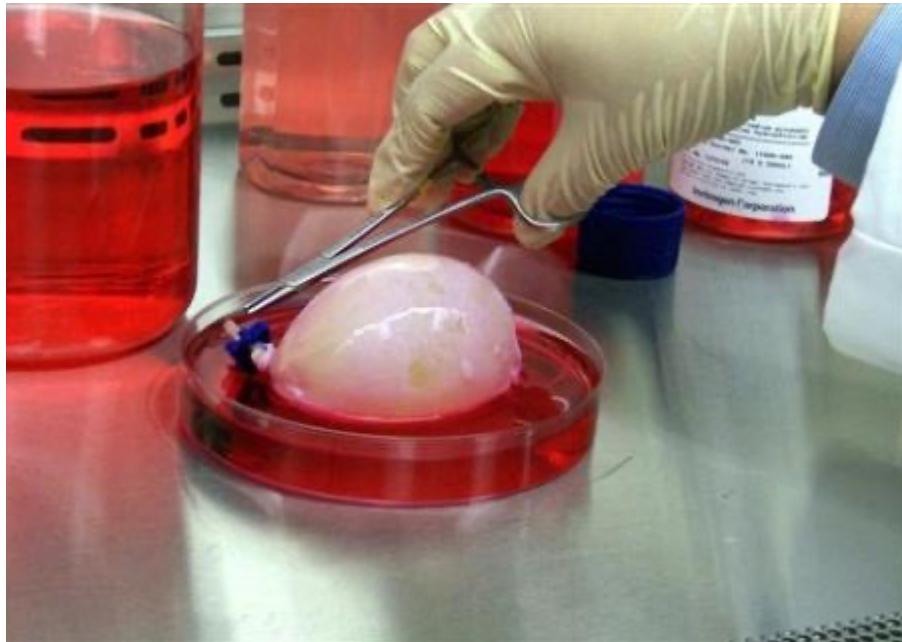
Division of Pediatric Urology, Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, Indiana

- 400+ children with augment
- Half followed for 11+ years
- 25% and 43% risk of additional surgery at 5 and 10 years, respectively.



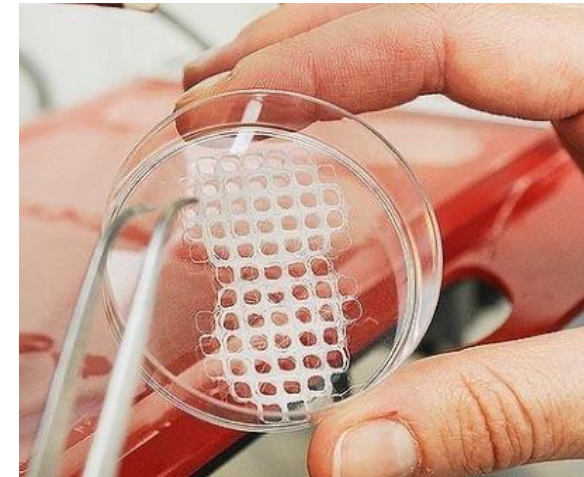
BLADDER AUGMENTATION

What is the future?



Autologous Cell Seeded Biodegradable Scaffold for Augmentation Cystoplasty: Phase II Study in Children and Adolescents with Spina Bifida

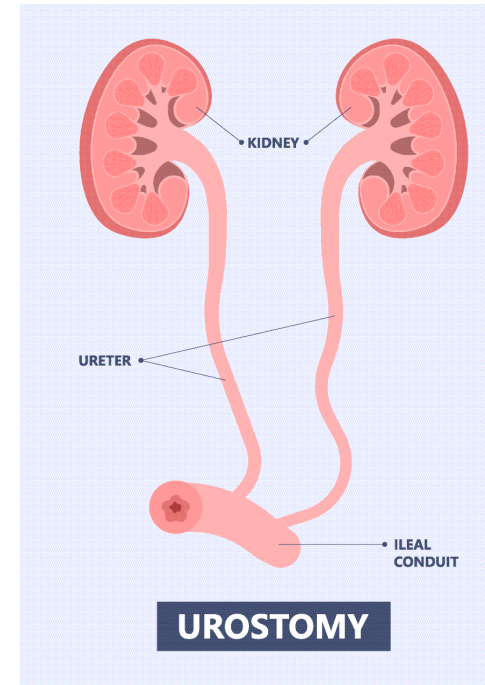
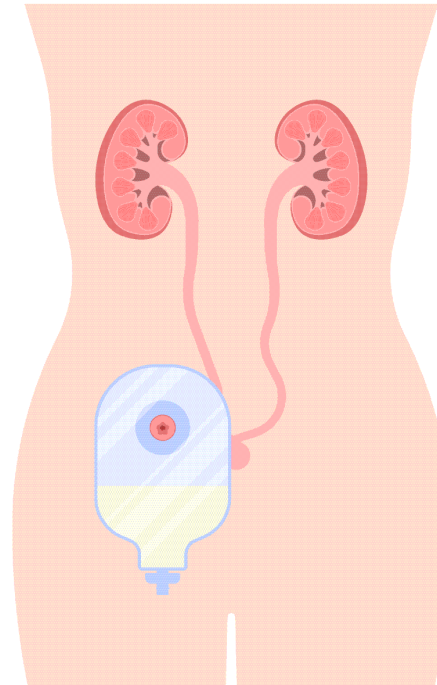
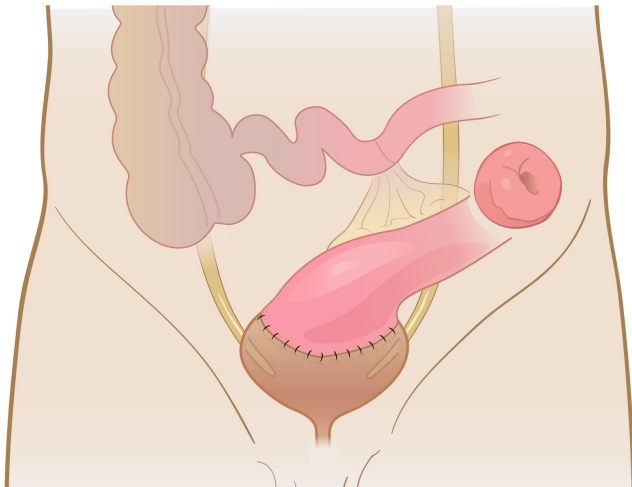
- Research study was performed in 10 children with neurogenic bladder due to spina bifida requiring enterocystoplasty despite maximum medical therapy
- Following open bladder biopsy, urothelial and smooth muscle cells were grown ex vivo and seeded onto a biodegradable scaffold to form a regenerative augment as the foundation for bladder tissue regeneration.
- **Results:**
 - Bladder pressures improved at 1 and 3 years, although the difference was not clinically different.
 - There was no clinical or statistical improvement in bladder capacity in any patient.
 - Serious adverse events of bowel obstruction and/or bladder rupture occurred in 4 patients.
 - **5 patients eventually underwent traditional bladder augmentation**



Urinary Diversion

ILEAL CHIMNEY

ILEAL CONDUIT



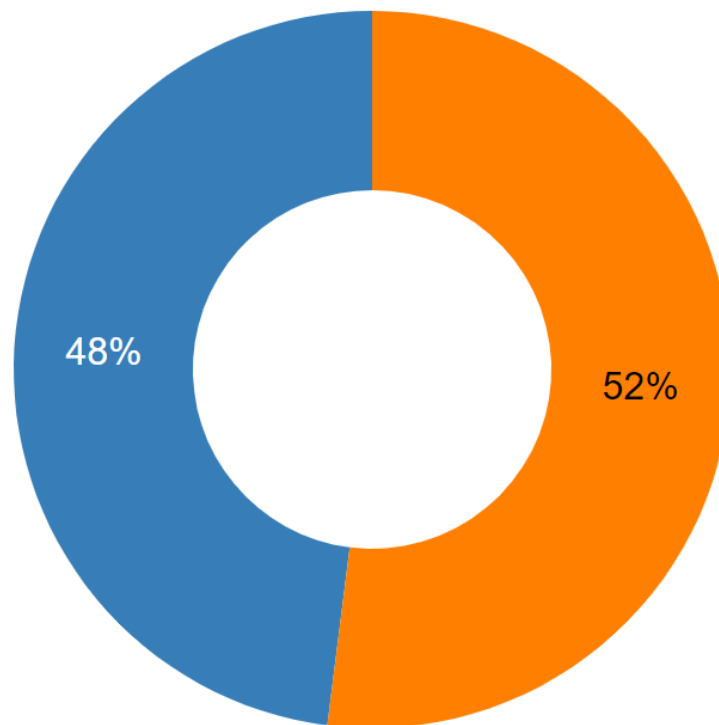
Neurogenic Bowel Management



Fecal Continence in Spina Bifida

Based on 7,589 Registry participants, ages 5 years and older with impaired bowel function.
Continence is defined as not having any daytime leakage of stool in the last month.

- 47% continent
- 53% incontinent



**Bowel
Continence
among
Impaired
Individuals**

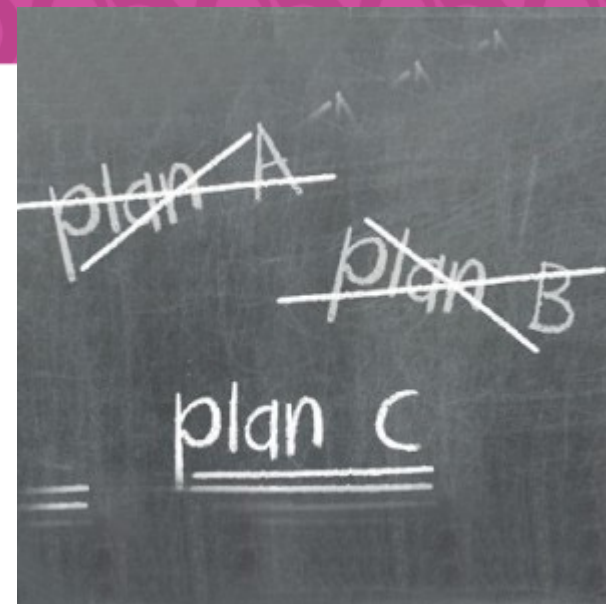
- Continent
- Incontinent

Source: Spina Bifida Registry survey

<http://www.cdc.gov/ncbddd/spinabifida/nsbprregistryfindings.html>

Bowel Management

- Multiple Therapies available
- Trial and Error Approach
- Start Simple and work up to more complex
- Goal is to have the child be independent with the program



Bowel Management Options

- Behavioral/Dietary
- Medications
- Enemas
- Surgery



Bowel Management

- Dietary Impact
 - Fiber and fluids are key to success
- Toddlers: Encourage raw fruits and vegetables
- Mix prune juice with apple juice



Bowel management

- Toilet sitting
 - Takes advantage of reflex that when food enters stomach it induces a colon contraction
- 15-20 minutes after eating
 - Sit with well supported feet
 - Grunt or bear down
- This approach is called habit training
 - Pick a time that works for you



Bowel management

- Consider stimulants either orally or rectally
 - Dulcolax
 - Laxative - Senna
- Miralax
 - Takes days to work
 - Not predictable for maintenance in everyone
 - Softens stool and may make it hard to push o
- Cone Enema
 - High volume irrigations



High volume Enema Program



PERISTEEN



NAVINA





What if enemas work but we can't teach them to do enema alone?



CHAIT TUBE



Mini-ACE

MiniACE®

Body Exterior

Soft External Bolster

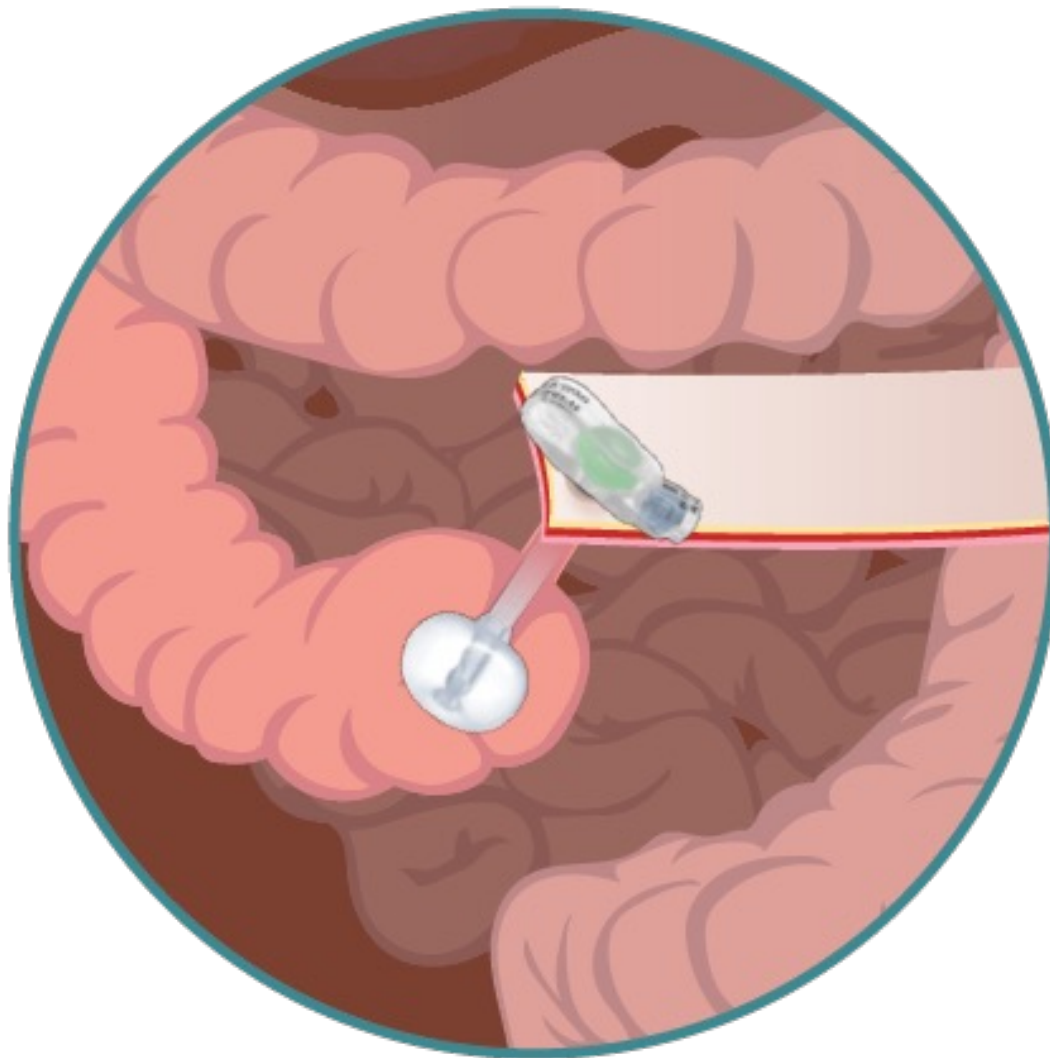
Anti-Leakage Valve

Secure Stoma Seal

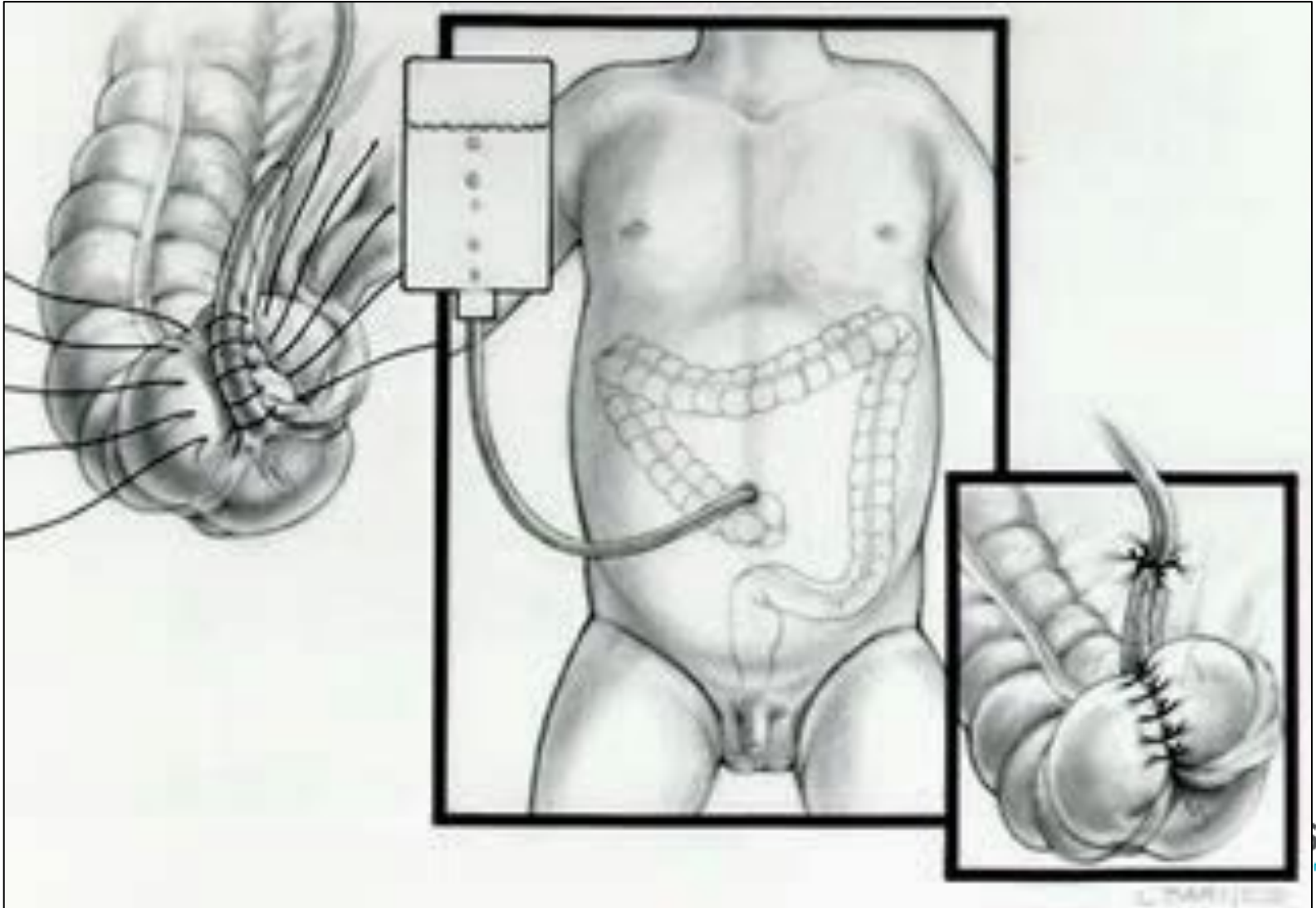
Exclusive AMT Balloon
Expands Horizontally

Protected Tip

Body Interior

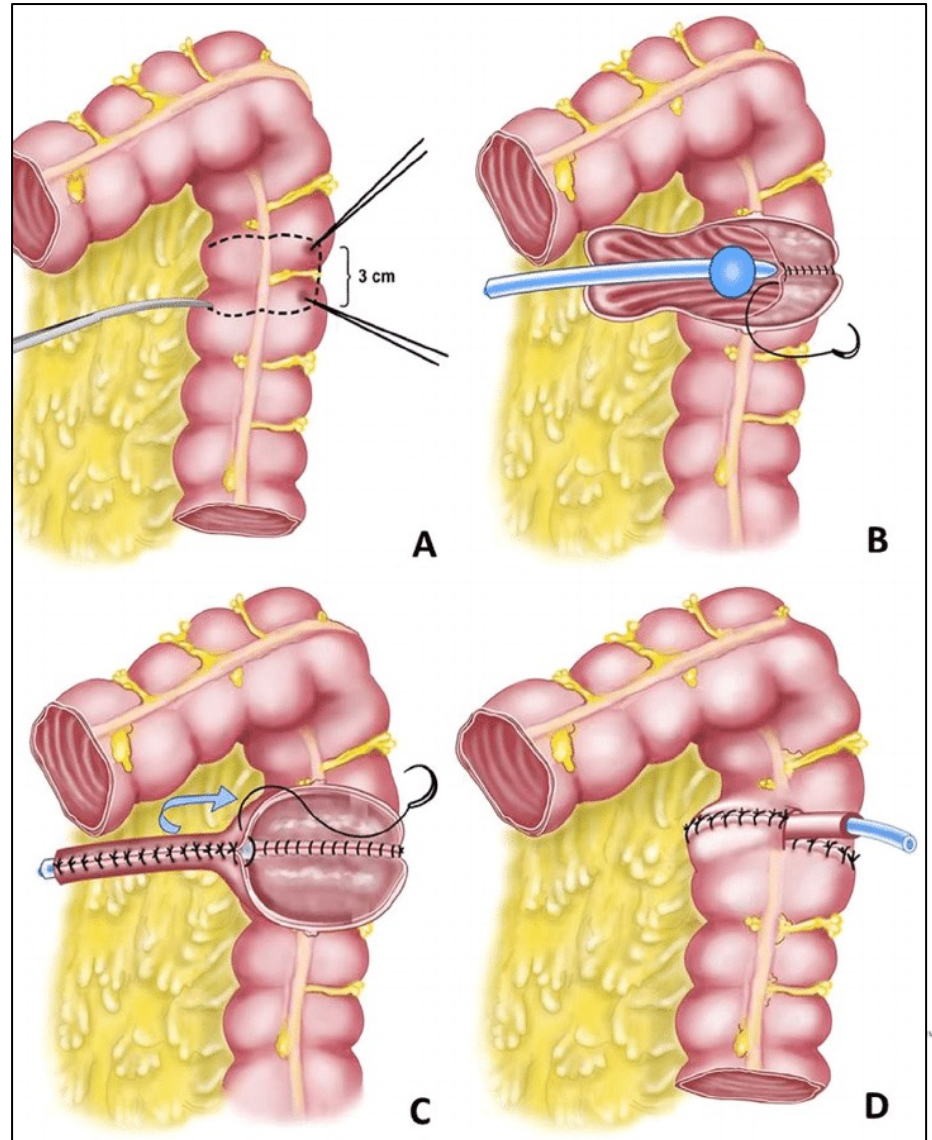


Malone Antegrade Continence Enema (MACE)



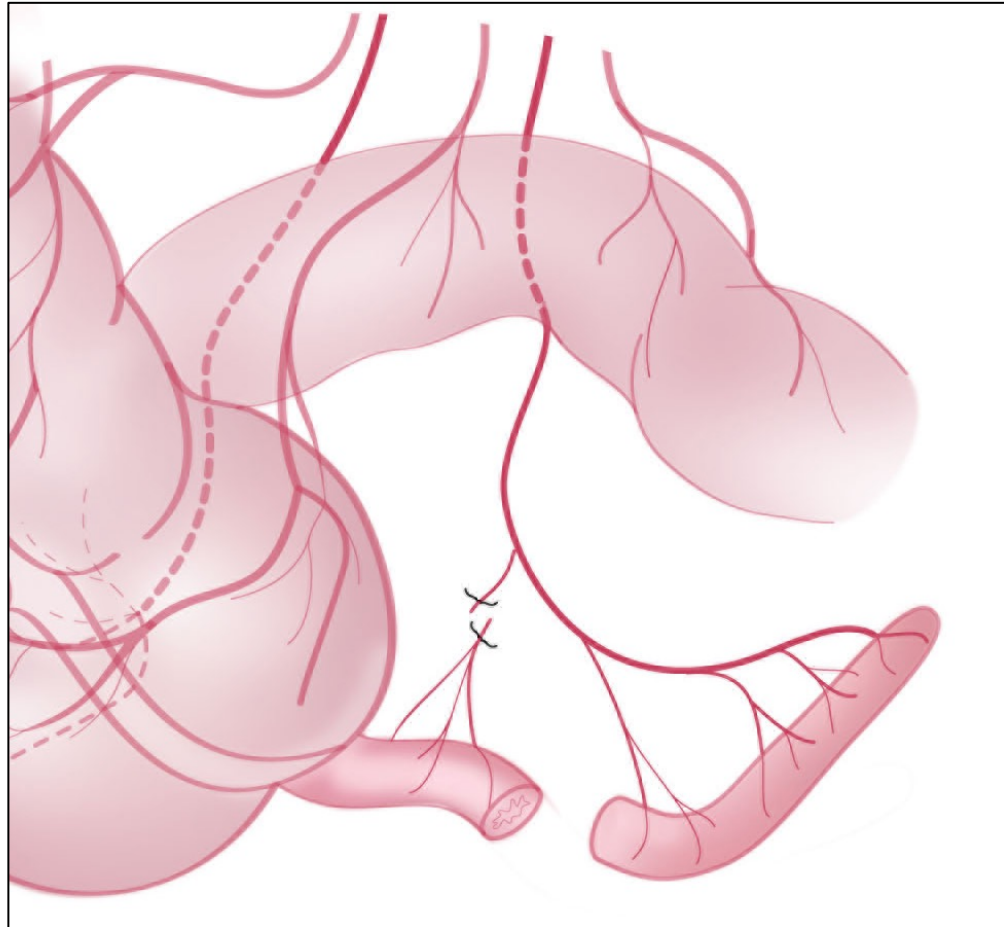
When Appendix is Absent/Unusable

- NeoMalone

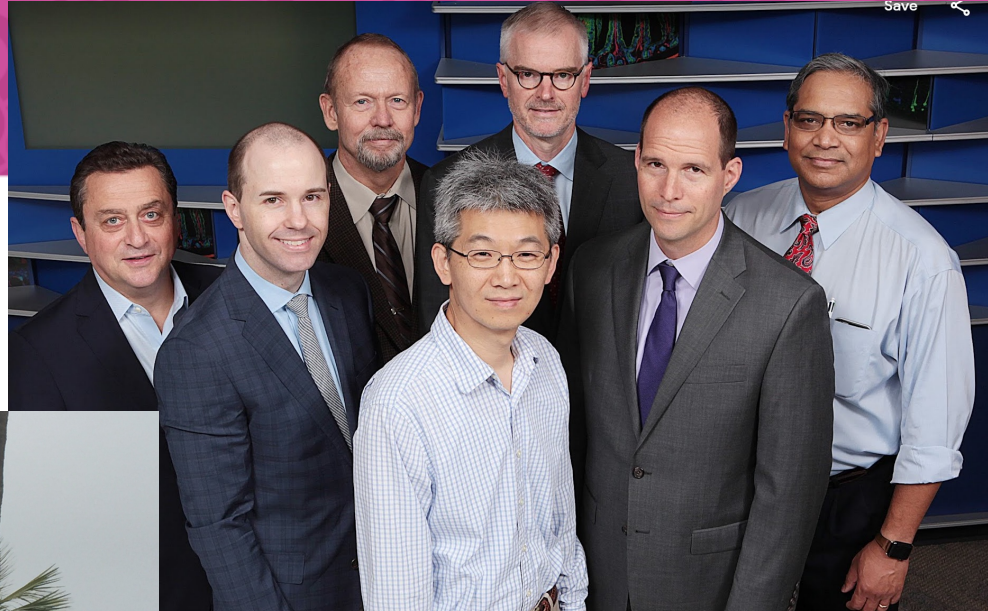


Reconstructive surgeries

- Malone should not be performed until assessment of bladder complete and possible need for Mitrofanoff excluded
- At times the appendix can be divided to make both channels and avoid need for more involved surgery



Our Teams



Questions ?

