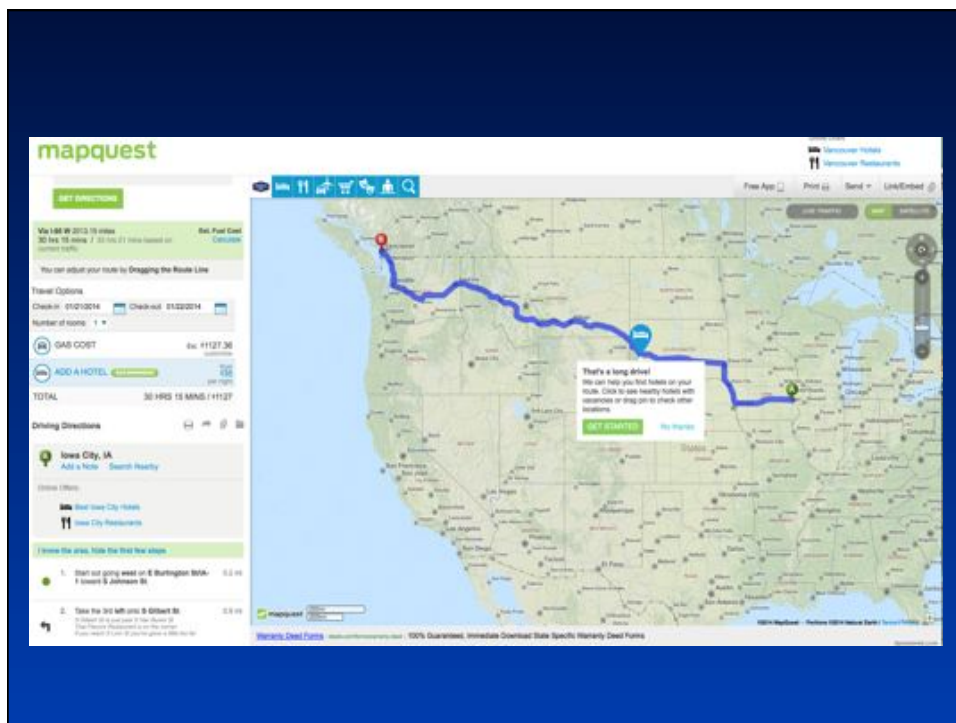
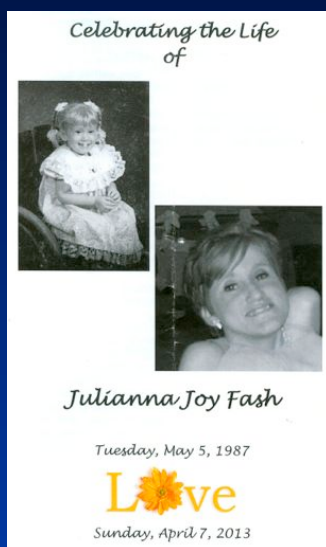




Pediatric Neurogenic Bladder

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Objectives

- Review what's known about:
Diagnosis, Evaluation, Urologic Management, and Outcomes for patients w/ NGB d/t Myelodysplasia
- Discuss what's not known...

NGB in Children

- Spina Bifida
 - * Myelomeningocele (>90%)
 - * Lipomyelomeningocele
 - * Meningocele
- Tethered Spinal Cord
 - * Dermoid cyst
 - * Dermal sinus tract
 - * Intradural Lipoma
 - * Thickend Filum



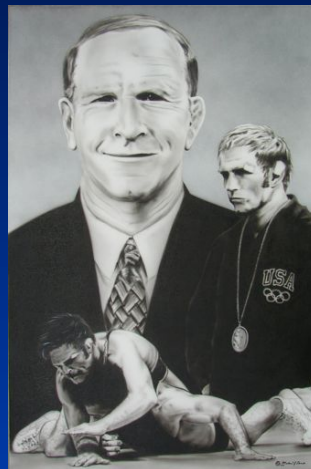
Myelodysplasia Success Story

- Increasing rates of infant survival
 - * VP Shunt 1950s
 - 1956-62 <20% survived past 24 months
 - 1957-75- 54%
 - 1975-2000- 83%
 - 2001-92%
 - * Most now live into adulthood
- Decreasing rates of renal failure and augmentation cystoplasty 27-41% to 11-17% related to:
 - * CIC by Lapedes 1972
 - * Recognition by McGuire of intravesical leak P > 40 cm H2O
 - * Proactive management of 'hostile' UDS parameters
- Prevention / declining incidence of myelodysplasia
 - Folate and/or termination

Who is the legendary U of Iowa
Wrestling Coach?

Answer

- Dan Gable



In 1972, Dan Gable won the Olympic Gold Medal.
How many cumulative points did he lose throughout
his Olympic matches?

- A. 1
- B. 2
- C. 4
- D. 8
- E. None of the above

GABLE

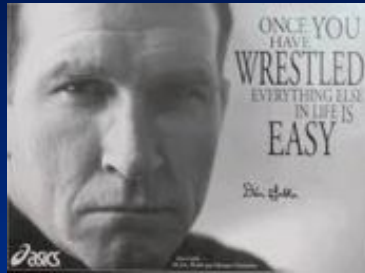
- Did not surrender a single point in the Munich Games
- ‘Top 100 Olympians’ of all times
- 182-1 Combined prep and college record
- Coaching record:
 - * 15 NCAA Titles
 - * 21 yr record of 355-21-5 (94.4%)
 - * 45 National Champions

MMC Success Story ... ?

Question for Andrew MacNeily -

Would you want to tell Dan Gable how you
'successfully' manage patients with MMC?

MMC Success Story ... ?



“Well Sir, I have them stick this tube in their urethra about 5X a day and take this medicine which keeps their pants pretty dry...

most of the time...

and then I get an ultrasound and a UDS once a year to see if there's been any damage...”

MacNeily – Gable Conversation Continued

“Well yes Sir, Mr. Gable Sir, you could consider damage ‘A Loss’...”

“Yes sir, I’ll say it with you sir:

‘Winners try to Win, they don’t try NOT TO LOSE!’,
yes sir”

GABLE

“I’m a big believer in starting with high standards and raising them. We make progress only when we push ourselves to the highest level. If we don’t progress, we backslide into bad habits, laziness and poor attitude.”

Are We Progressing w/MMC?

We've done a lot right, and a lot of good, but we have a long way to go before we get a 'Gold Medal' and we can do better...

Questions

- Is asymptomatic bacteruria really harmless?
- Should we wrap muscle around BNR?
- Which service should be care giver/coordinator for MMC patient at any age? (answer- not pediatric surgeons, orthopedists, neurosurgeons)
- How aggressive should I treat asymptomatic OAB on UDS?
- Would vesicostomy as "short term" solution be better than CIC?
- Does nocturnal catheter increases bacteruria or cancer risk?
- Should botox be more of a first line agent than anticholinergics? How can we make it a clinic procedure?
- What can we inject beside/beyond botox?
- What's our HgA1C for the bladder?
- How does tx. of constipation affect the NGB function?
- Is obesity pre-destined in this population?
- How many get PT and how often and for how many years?
- How could we use peer mentors?
- What do we do for parents / families? What do they need? (they are primary care givers)

Back to a few things we know...

(or at least ask about on examinations)

Sacral Abnormalities

⊕ Dimple, Mass, Skin tag (phallic), Hemangioma, or Hair Tuft

⊕ Location

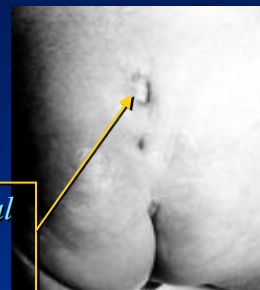
⊕ High- more likely spinal abnormality

⊕ Low- more likely Pilonidal sinus

⊕ Appearance of Gluteal Cleft

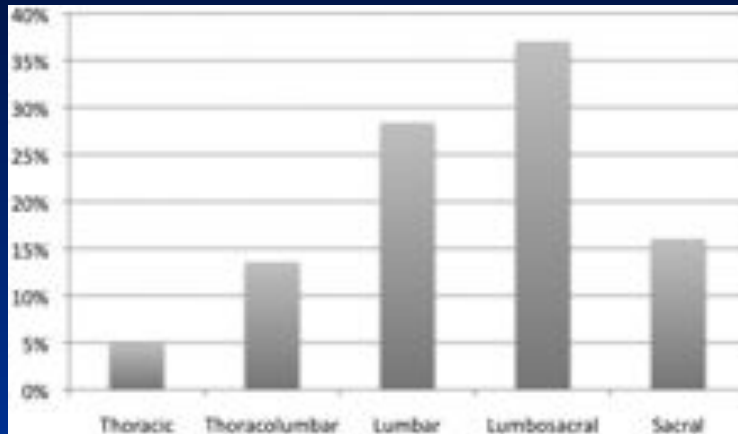
⊕ Symmetric and Midline

⊕ Eval with MRI best



*Dermal
sinus
tract*





*Myelomeningocele levels-
lumbosacral lesions account for most cases*

Metcalf P et al. J Urol 2011;185(suppl6):2547-51

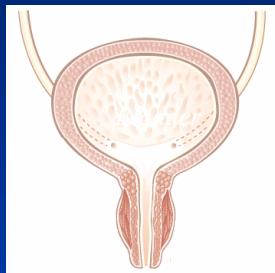
MMC- Vertebral and Spinal Level

	Vertebral Anomaly	Neurologic Level
Thoracic	20%	16%
Lumbar	28%	20%
Lumbosacral	52%	44%
Sacral		20%

N.B.- Bony level doesn't correlate well with neurologic level or ultimate bladder function

Functions of the Lower Urinary Tract

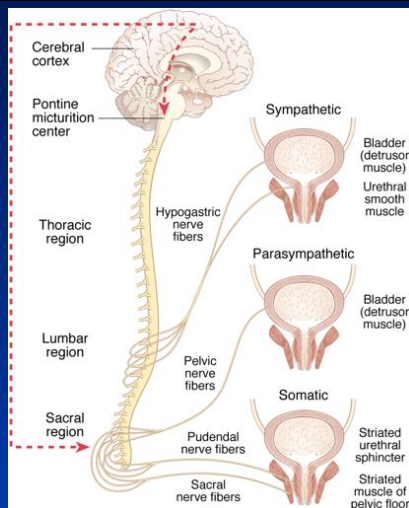
1. Urine storage
- Reservoir: Bladder
2. Urine release
- Outlet: Urethra



Slide from Firouz Daneshgari, M.D.

AUA MOC Review Course 2013

This involves reflexes organized at the spinal and brainstem level
Lower Urinary Tract Innervation



Upper vs. Lower Motor Neuron

- UMN- overactive bladder, no control of sphincter, DSD
 - * Leads to thick walled bladder
- LMN- acontractile detrusor and some denervation of external sphincter
 - * Bladder usually small and smooth walled

Arnold-Chiari Malformation

- 85% of MMC have A-C malformation
 - * Cerebellar tonsils herniate through foramen magnum
 - * Obstructs 4th ventricle and prevents CSF from entering subarachnoid space around brain/cord-hydrocephalus

Fetal MMC Repair

- ⊕ Performed at 3 centers
 - ⊕ CHOP, Vanderbilt, and UCSF
 - ⊕ All in Management of MMC Study
 - ⊕ 'MOMS' Trial (Randomized)

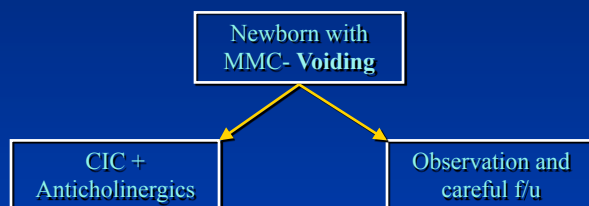


- ⊕ Decreased Need for VP Shunt by 50% (40% vs.82%)
- ⊕ **Bladder and bowel function doesn't appear to be significantly improved**

⊕ Clayton DB et al. J Urol 2011; 1581-85

Newborn Management

- C-section if Dx Known
- Spinal Closure within 24 hrs.
- Renal U/S +/- VCUG or VUDS
 - * Establish a baseline
 - * Up to 20% of Pts will have abnormal imaging as newborn
- Latex precautions



Early Catheterization (CIC)

- ⊕ 80-90% will have NGB dysfunction
- ⊕ No problem with acceptance by child
- ⊕ Parents incorporate CIC into care routine
- ⊕ Potential to prevent need for bladder augmentation
- ⊕ Potential to prevent renal injury

Observation

- Upper tract surveillance with U/S
- UDS
- CIC Indications
 - * UTIs
 - * VU Reflux
 - * Detrusor Sphincter Dyssynergia
 - * Hydronephrosis
- Advantages
 - * Less burden for caregivers
 - * Less bacteria in urinary tract

Follow-up

- Upper Tract Surveillance
 - * U/S- yearly (more often in newborn- q 3mo.s)
- Bladder
 - * VUDS- Newborn
 - Frequency depending on severity
 - Infections or upper tract changes
 - **CONTINENCE**

Components of VUDS

- **Fluoroscopy**
 - * Trabeculations / Morphology
 - * VUR
- **CMG**
 - * Detrusor Activity
 - Overactive
 - Underactive
 - * Compliance
 - * Capacity
 - * Cystometric Leak Point Pressure (> 40 cm H₂O predicts deterioration)
- **EMG**
 - * Needle- most accurate
 - * Patch

Initial UDS Finding

⊕ Detrusor	⊕ Sphincter
⊕ Overactive	⊕ Dyssynergic- 55%
⊕ Stable	⊕ Synergic- 18%
⊕ Areflexic- 37%	⊕ Denervated (absent activity)- 27%

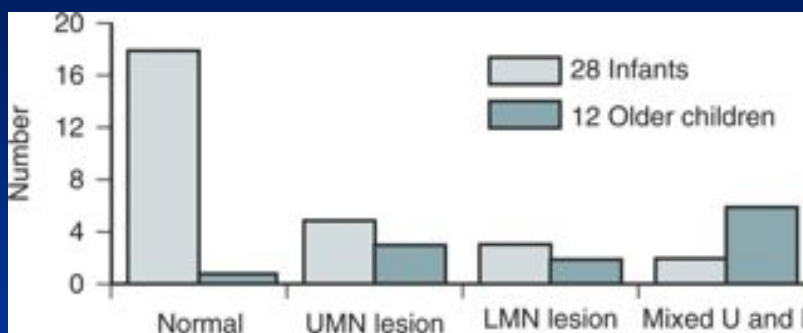
Bauer et al. JAMA 1984; 252:650.

Risk for Urinary Tract Deterioration

- ⊕ Dyssynergic- 71% deteriorated w/in 3 years
- ⊕ Synergic- 17%
- ⊕ Denervated- 23%
- ⊕ Conclusion- DSD patients need intervention

Bauer et al. JAMA 1984; 252:650.

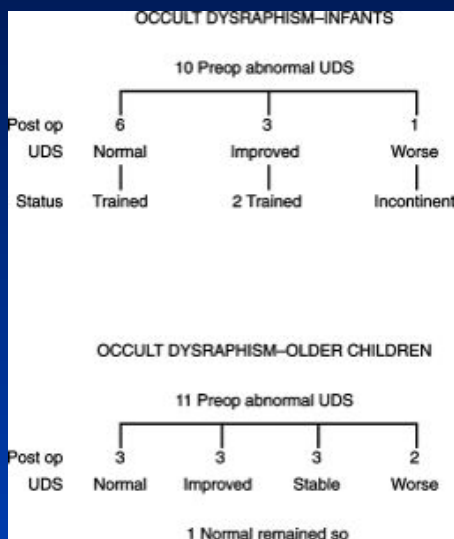
UDS in Tethered Spinal Cord



- Normal UDS in 60% infants
- Drops to 20% at 3 yrs. Suggesting progressive lesion

Keating et al. J Urol 1988; 140:1299.

Urodynamics after SC Untethering



Treatment in infancy improves outcome

Much better prognosis than MMC

Deterioration is uncommon after surgery-

~25% will re-tether (often in age 2-8)

Keating et al. J Urol 1988; 140:1299.

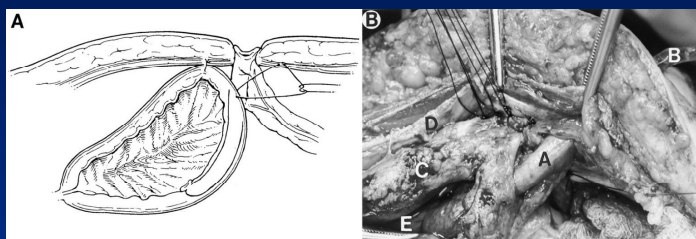
Reflux and NGB

- 5-20% have VUR as infants
 - * Poor bladder compliance, overactivity, or DSD
 - * Rate increases if observed.
- Treatment- Prophylaxis
 - * **Bladder Management- Resolves up to 50%**
 - CIC + Anticholinergics
 - * **Surgical Treatment- Must adequately manage bladder first!**
 - Ureteral reimplantation- 95% success
 - Endoscopic -60-70% success
 - Vesicostomy/Augmentation- If bladder can't be Rx'ed with CIC

Clean Intermittent Catheterization

- Adjust frequency to keep bladder volumes below 40 cm H₂O pressure
- Increasing frequency of CIC associated w/ decreased UTIs (tough concept for parents)
 - * Asymptomatic bacteruria in 70%, but symptomatic UTI <30%
- Adjust size of catheter so adolescent boys using ≥ 12 F

Mitrofanoff- Continent Catherizable Channel



- Facilitates catheterization
- Increases independence
- Durable - ~10% stenosis & 98% continent -mean f/up 52 mo
 - CHOP series: Cooper et al. J Urol, 163, 1922-26, 2000
 - Other series ~15-25% stomal revision rate- may occur years later
 - Welk BK, Afshar K, Rapoport D, MacNeily AE J Urol, 180, 1856-1860, 2008

'PRN' Antibiotics

- Parents become skilled at suspecting when child on CIC has UTI
- Consider Atbx. Rx to be used by parents prn:
 - Decreased resistance likely compared to daily
 - More likely to improve rapidity of treatment
 - Saves parental and physician time and healthcare costs
 - Not evidence based- may be good project...

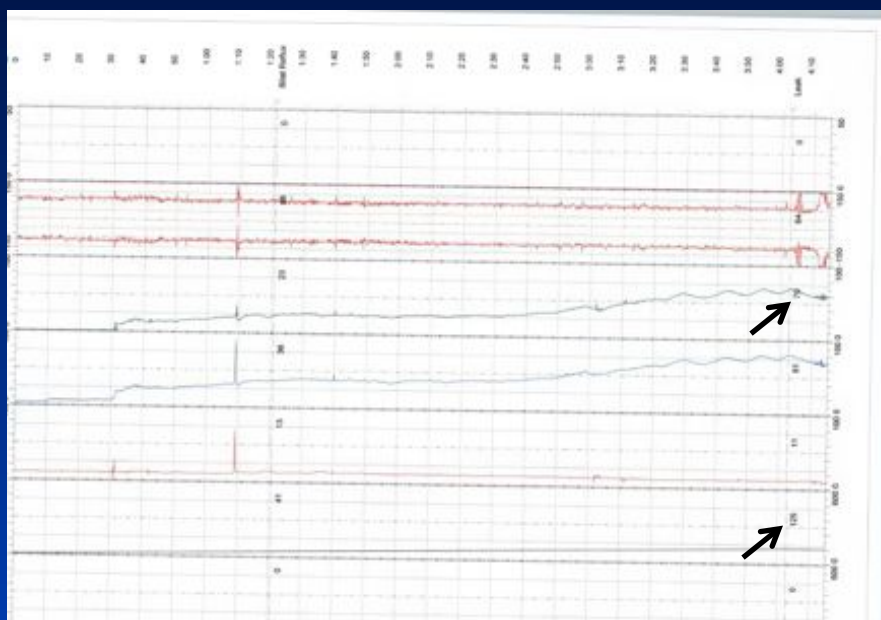
Overnight Catheter Drainage

- Night time is longest interval between catheterization.
 - * Chronic high pressures storage
 - * Catheter overnight alleviates obstruction
 - Tape cath or use Foley- don't use hydrophilic!
 - Double diaper catheter or attach to bag
- Unknowns:
 - Utility as prophylactic measure
 - Replace high pressure and stasis with chronic foreign body/irritation...? Increase risk of Cancer?

Nocturnal Emptying/ Overnight Catheter Drainage

- Koff et al. J Urol 174, 1629-32, 2005
- 19 children
- F/up 23 months
- 79% improved
 - * Hydronephrosis
 - * UTI
 - * Incontinence
 - * Bladder capacity

KM UDS Before Nocturnal Cath

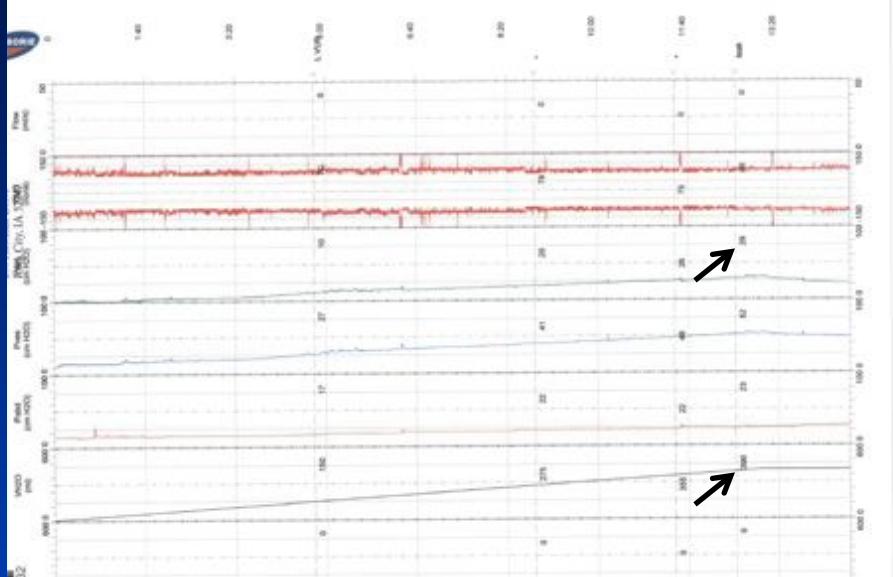


KM Before Nocturnal Cath



2 Months Later...

KM 2 Mo.s After Nocturnal Cath



KM 2 Mo.s After Nocturnal Cath



Take Home Point:
The bladder has an amazing
ability for rehabilitation and
remodeling.

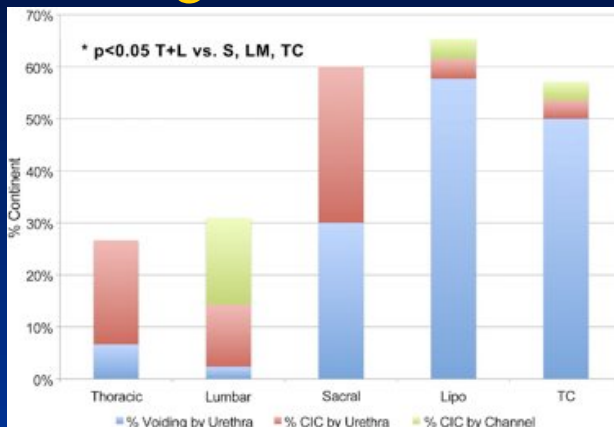
Further Challenge:
Developing environment to
permit this to happen.

Urethral Dilation

- Not widely used
- Elevated DLPP, worsening compliance, hydronephrosis
- Failed CIC/Anticholinergics
- 19 Girls (mean age 19 months)
 - Dilated 18-36 fr
 - DLPP decreased in 75%
 - VUR resolved in 25%
 - Only 10% required Vesicostomy
- Guarding Reflex- Stretch of Urethra inhibits detrusor

Kiddo et al. J Urol 2006; 176:1831.

Voiding and Continence



Better continence with sacral (S) myelomeningocele, lipomeningocele (LM, Lipo) and tethered cord (TC) than in those with lumbar (L) and thoracic (T) myelomeningocele.

Metcalf P et al. J Urol 2011;185(suppl6):2547-51

Non-Surgical Options for Continence

- CIC
- Anticholinergics
 - * Oxybutynin (Ditropan XL) - titrate to effect/ Side effects
 - Up to 30 mg/ day for Adolescents/ Older children
 - * Oxytrol Patch- Fewer side effects (Off label)
 - * Others (Tolteratidine, Trospium, Darifenicen, Solifenicen- Off label)
 - * Combinations of the above
 - * Intravesical oxybutynin
- α -Sympathomemetics
 - Psuedoephedrine
- Botox if fails anticholinergics (or sooner??)

botulinum-A Toxin

- Neurotoxin produced by *Clostridium botulinum*
 - * Inhibits Ach release, and other neuropeptides
 - * Common dose- 200 IU or 10 IU/kg up to max of 300
- 65-87% become dry despite failing CIC/Ach
 - * Duration ~6 months
- Repeat injections efficacious and no increased fibrosis
 - Pascali MP et al. J Urol 2011; 2552-57

Surgical Options for Continence

Pre-operative Evaluation

- Pattern of Incontinence / Dry Interval
- Ability/ Motivation to perform CIC
 - * If unable or unwilling don't operate!
- Kidneys
- Bladder - VUDS
 - * Bladder as a reservoir – assess need for augment
 - Capacity with bladder outlet **occluded**
 - Objective assessment of compliance
 - Detrusor Hyperreflexia
 - * Bladder outlet competency
 - Detrusor LPP
 - Abdominal LPP
 - **BN appearance on cystogram- in one study closed bladder neck was only UDS factor predictive of continence after augmentation cystoplasty**
 - Ghanem MA et al., J Ped Urol 2013; 293

Types of Procedures

- **Lengthen** Urethra
 - Kropp
 - Pipi-Salle
 - YDL
- **Narrow** Urethra / BN
 - Mitchell
 - Keeling
- **Compress** Urethra / BN
 - AUS
 - Fascia Sling/ Wrap
- **Close** Urethra / BN

Surgical Results: Rx of Neurogenic Sphincteric Incontinence

PROCEDURE	% CONTINENCE	% CIC	% AUGMENT	% REVISION
AUS	76-93	63	33-58	19-28
Kropp	78-81	100	88-100	20
Pipi-Salle	50-69	100	75-83	12-17
YDL	64	91	91	45
Sling	40-100	78-100	55-93	15
Injection	5-63	68-100	0-31	31-61

Kryger et al. J Urol. 163:256, 2000

What is Continence?

- Loyd JC et al. J Ped Urol 2013; 567
 - * Literature review
 - * Only 57% clearly defined 'continent'
 - Always Dry (used in 24% of reports)
 - Dry w/ CIC/Void
 - q2 hr; q3 hr; q4 hr
 - No pad use
 - Security pads only
 - No leak on UDS
 - * 63% didn't say how continence status determined
 - * Some authors on repeat studies changed their own definition of continence!

'Second Line' Surgical Options

- Injection of BNR/Sling w/Deflux
 - * 25% dry w/1 or 2 injections
 - Dajusta et al. J Ped Urol 2013; 278
- Bladder neck closure
 - * 96% success rate in 28 patients, f/up 69 mo.s
 - * 3/28 had rupture, 1/28 stone, 82% compliance w/fup
 - Kavanagh A, Afshar K, Scott H, and MacNeily AE, J Urol 2012
- Continent catheterizable resevoir

What to do for a failing bladder on CIC and anticholinergics?

- ~15% have progressive decline in bladder functions
- **RULE OUT: tethering (3-32% of MMC develop), syringomyelia, VP shunt malfunction, partial herniation of brainstem/cerebellum**

What to do for a failing bladder on CIC and anticholinergics?

- Addition of 2nd anticholinergics (po or intravesical)
 - * 85% continence in adults (Amend, 2008)
- Bo-Tox
- Auto-augmentation
- Augmentation cystoplasty
- Vesicostomy
- Diversion

Table 128-1.
Drugs That Affect Lower Urinary Tract Function

TYPE	Dosage ¹	
	Minimum	Maximum
Cholinergic		
Bethanechol (Urecholine)	0.7 mg/kg tid	0.8 mg/kg qid
Anticholinergic		
Propantheline (Pro-Banthine)	0.5 mg/kg bid	0.5 mg/kg qid
Oxybutynin (Ditropan)	0.2 mg/kg bid	0.2 mg/kg qid
Glycopyrrolate (Robinul)	0.01 mg/kg bid	0.03 mg/kg tid
Hyoscyamine (Levsin)	0.03 mg/kg bid	0.1 mg/kg tid
Tolterodine (Detrol)	0.01 mg/kg bid	0.04 mg/kg bid
Trospium (Sanctura)*	10 mg/day	20 mg/day
Solifenacin (Vesicare)*	5 mg/day	10 mg/day
Darifenacin (Enables)*	7.5 mg day	15 mg day
Propiverine (Detrunorm)*	0.1 mg/kg bid	0.4 mg/kg bid
α-Sympathomimetic		
Phenylpropanolamine	2.5 mg/kg tid	2.5 mg/kg qid
Ephedrine	0.5 mg/kg tid	1.0 mg/kg tid
Pseudoephedrine	0.4 mg/kg bid	0.9 mg/kg tid
α-Sympatholytic		
Frazosin (Minipress)	0.05 mg/kg bid	0.1 mg/kg tid
Phenoxybenzamine	0.3 mg/kg bid	0.5 mg/kg tid
β-Sympatholytic		
Propranolol	0.25 mg/kg bid	0.5 mg/kg bid
Smooth Muscle Relaxant		
Flavosate (Urispas)	3.0 mg/kg bid	3.0 mg/kg tid
Dicyclomine (Bentyl)	0.1 mg/kg tid	0.3 mg/kg tid
Other		
Imipramine (Tofranil)	0.7 mg/kg bid	1.2 mg/kg tid

*Studies evaluating safe dosing in children have not been completed.
¹Dosages based on recommendations for a 70-kg person.

Campbell's

Detrusorectomy/ Auto Augmentation

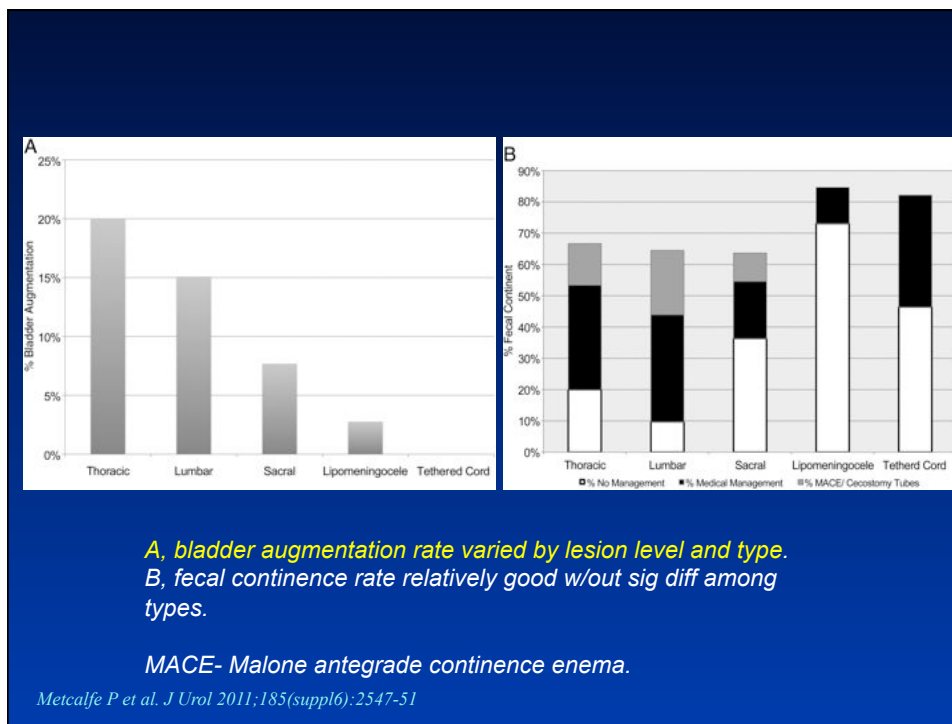
- Snow and Cartwright- 1989, 1992
- Dik et al- 35 pt.s: improved compliance (16) and capacity (13)
- MacNeily, Afshar et al- 17 patients w/ 71% failure rate for upper tract protection and continence
- Veenboer PW et al. J Urol 2012- greater increase in capacity w/ enterocystoplasty



Bladder- Bowel Augmentation

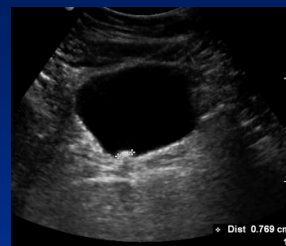
- **Last option** for low capacity, poorly compliant, refractory bladder
- Small or Large Bowel
(rarely gastric augment unless renal failure)
- Excellent increase in Capacity and Compliance
- No need for preop bowel prep
 - Victor D et al. J Ped Urol 2012, 201-204





Complications of Augmentation

- Acid-base
- Vitamin B12 deficiency (begins by 7 yr.s)
- Bone metabolism
- Recurrent UTIs
- Bladder stone formation (~15%)
- Perforation (8%)
 - * May present with vague abdominal pain
 - * CT cystogram more sensitive than cystogram
- Bowel obstruction (3%)
- Cancer Risk (1-2% at 19-32 years)
- Metcalf et al 2006 reviewed 500 augmentations and 1/3 required additional operation at mean of 13 years



Clayton DB, Brock III, JW, Joseph DB Dev Disabilities Research Reviews 2010
Blackburn SC et al J Ped Urol 2012;47-50

Bowel Segment: Advantages and Disadvantages

APPENDIX 1: BOWEL SEGMENT, ADVANTAGES AND DISADVANTAGES		
	Advantages	Disadvantages
Stomach	Decreased infection risk, decreased absorption of urea	Hematuria-dysuria syndrome, hypokalemic hypochloremic metabolic alkalosis, osteomalacia and vitamin D deficiency, B12 deficiency (due to lack of intrinsic factor)
Jejunum	None	Severe hyponatremic, hyperkalemia and hypochloremic metabolic acidosis; profound dehydration
Ileum	Abundant and redundant mesentery which allows for easy manipulation and positioning, allows for low pressure urine storage, lower incidence of ureteral anastomotic stricture	Hyperchloremic hypokalemic metabolic acidosis, B12 deficiency (if terminal ileum is used), relative contraindication if patient has history of pelvic radiation, resection of >100 cm lipid malabsorption, vitamin (A, D, E, K) malabsorption
Ileocecal	Antireflux mechanism	Reduction in stool transit time, increased risk of steatorrhea, increased risk of stone formation secondary to bile acid loss
Colon	Transverse colon can be used if patient underwent prior pelvic radiation	Hyperchloremic hypokalemic metabolic acidosis (chloride absorption and bicarbonate excretion more pronounced than ileum); higher incidence of ureteral anastomotic stricture; sigmoid neobladder, poor urodynamic parameters, high rate of incontinence

Poch MA, Guru KA, and Peabody JO. AUA Update Series. 30(39):374, 2011

Regenerative Medicine

- ⊕ Atala 2006
 - ⊕ 7 Patients underwent augmentation w/ cell-seeded matrix and covered w/omentum
 - ⊕ Results- Modest increase in capacity
- ⊕ Joseph 2009
 - ⊕ Multi-institutional phase II study
 - ⊕ 10 children, subjective improvement in some, but not correlated w/objective UDS improvement- Questionable efficacy

Xiao Procedure

- Xiao et al, J Urol 173; 2111-2116, 2005
 - * Creation of a somatic-autonomic reflex pathway
 - * Lumbar VR to S3 VR microanastomosis- L5 dorsal root left intact as afferent branch
 - * By 1 year, 17/20 improved voiding (scratching thigh) and continence
- Peters et al, 2010
 - * Only 30% w/improved continence and spontaneous voiding- none w/ complete continence
 - * Temporary LE weakness common (89%)
 - * 1 of 9 w/permanent foot drop

Quality of Life

- Strong evidence is lacking regarding improved QOL with continence
 - * 31 pt.s w/ spina bifida
 - * No improvement in QOL scores despite improvement in continence
 - MacNeily, Jafari, Scott, Dalgetty, Afshar, J Urol. 2009
- Need for better instruments
- Need for personal interviews
 - Arnell MVM et al. J Ped Urol 2013; 559

**And now, Fuzzy Memories
by Jack Handey...**

Maybe it's my imagination, but food seemed to taste better when I was a kid. Also, food would sing and dance and play musical instruments, but that could also have been my imagination.

**And now, Fuzzy Memories
by Jack Handey...**

*When I looked up at the scoreboard, there were fifteen seconds left. It seemed like plenty of time, but it wasn't.
Before I could get to the rest room, I had wet my pants.*

Puberty and Beyond

- ⊕ Increased concerns over body image
- ⊕ **Decreased compliance** w/CIC, meds, f/up
 - ⊕ ~2/3 of adults do not seek regular urologic f/up
 - ⊕ >50% stopped CIC and >50% use too small a catheter
 - ⊕ Increased risk of injury w/decreased catheter size
 - ⊕ Adolescent boys should use \geq 12F
- ⊕ Recommendation- yearly u/s and Cr (not evidence based!)
 - ⊕ UDS for new symptoms
- ⊕ **60% of adults incontinent!**
- ⊕ Multiple barriers to transitioning to adult clinics
- ⊕ If born between 1963-1971:
 - ⊕ 33% dead before age 5
 - ⊕ 26% more died over next 35 years
 - ⊕ Epilepsy, PE, CSF shunt failure, urinary sepsis

Sexuality with MMC

- 28-40% of MMC teenagers had sexual encounters
- Women 2.3X more likely than men
- Only 52% satisfied w/sexual lives
 - * Incontinence, low self-confidence, and hydrocephalus risk factors
- Almost all desired to marry and have children
- 80% of men had improved erectile function w/ Sildenafil (Palmer et al, 2000)
- Men w/neurologic lesions S1 or lower more likely to have nl. fet.

MMC and CKD

Age	N	I	II	III	IV	V
31-40	1	0	9	5	0	1
21-30	22	4	20	3	0	0
11-20	14	2	11	1	1	0
0-10	21	1	3	1	0	0

26% have CKD 1-4; 1% have CKD 5

Renal Failure increases with age

Risk factors: VUR, Upper Tract Dilation, High Bladder P, Poor Patient Compliance

Malakounides G et al J Ped Urol 2013;585

Wide P et al J Ped Urol 2012;187

Torre M et al J Ped Urol 2011; 39

Veenboer PW PLOS 2012; 1

Cancer and MMC

- Iowa study
 - * 8 pt.s w/mean age 41
 - * 88% presented w/T3 stage
 - * Median survival – 6 months
 - * **Only 1 of 8 had previous augment!**
 - * All had chronic UTIs
- Unclear if screening cysto and cytologies useful
 - Husman DA et al. J Urol 2009;403

Questions?

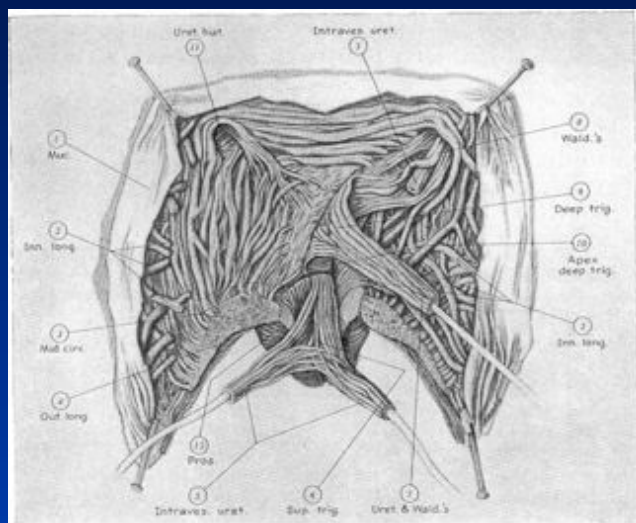


Thank You!

Bowel Function

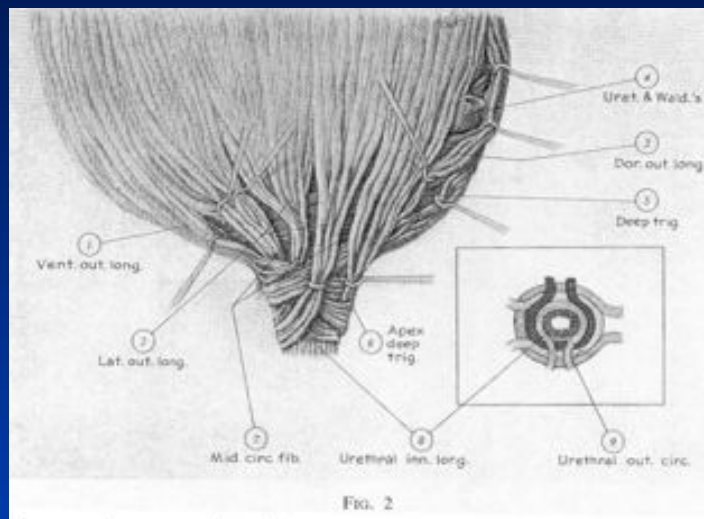
- Bowel incontinence not associated with attainment of urinary continence
- Treatment Options:
 - * Diet / Fiber-Bulking
 - * Miralax
 - * Suppositories
 - * Enemas
 - * MACE / Cecostomy tube
 - 60-90% continence rate w/no difference b/t techniques
 - 300-1500 ml of H₂O, Glycerol, Miralax, Mineral Oil
 - Leakage ~15%

BN: Anatomy and Physiology



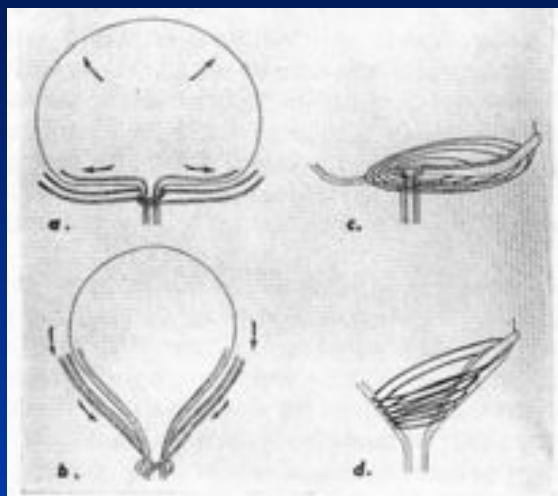
Tongue and

BN: Anatomy and Physiology



Tanaka and

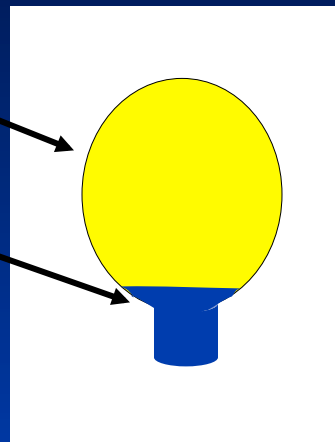
BN: Anatomy and Physiology



Tanaka and

BN: Receptor Pharmacology

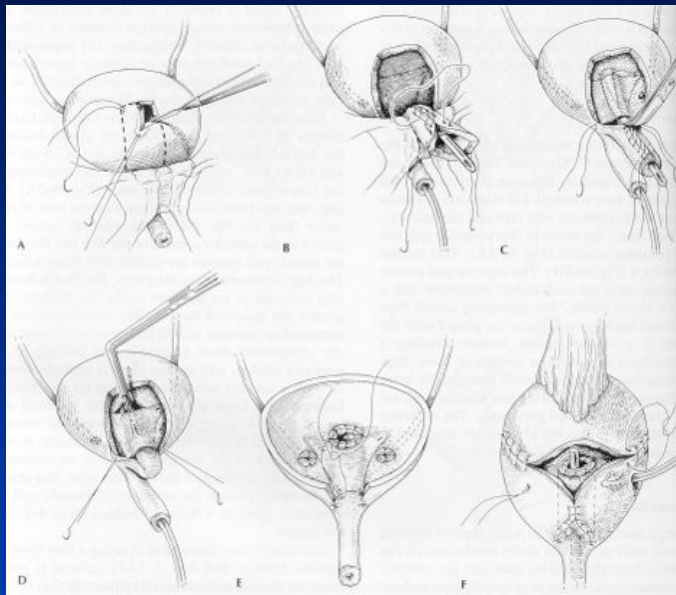
- Bladder Body
 - β2-Adrenergic
 - m-ACh
- Bladder Base
 - α-Adrenergic
 - Nitric Oxide



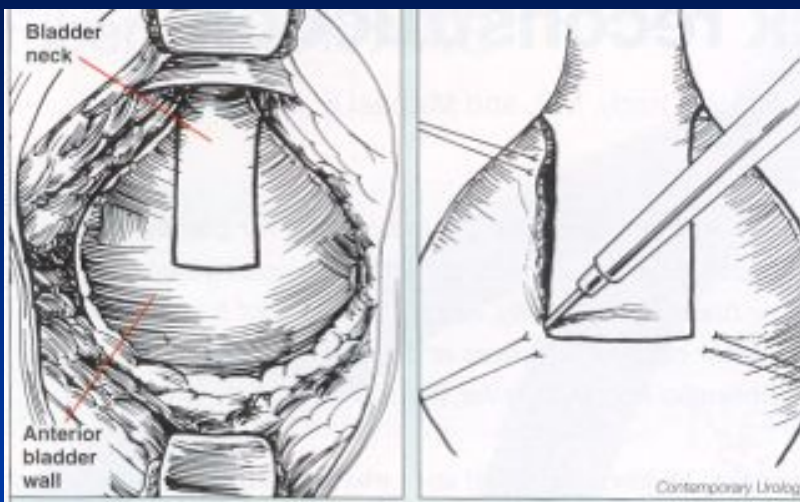
Pre-operative Evaluation

- Ability/ Motivation to perform CIC
- Dry interval/ pattern of incontinence
- Upper tract Imaging
- VUDS
 - * Bladder as a reservoir
 - Capacity with bladder outlet **occluded**
 - Objective assessment of compliance
 - Detrusor Hyperreflexia
 - * Bladder outlet competency
 - DLPP
 - ALPP
 - BN appearance on cystogram

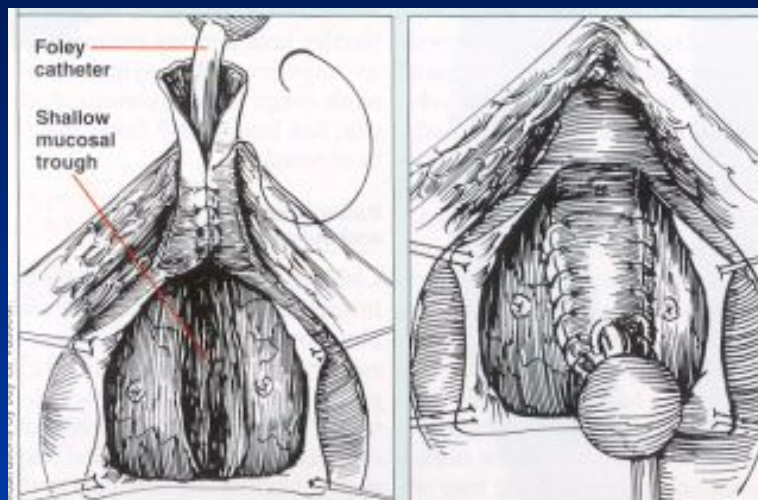
Kropp Procedure



Modified Kropp



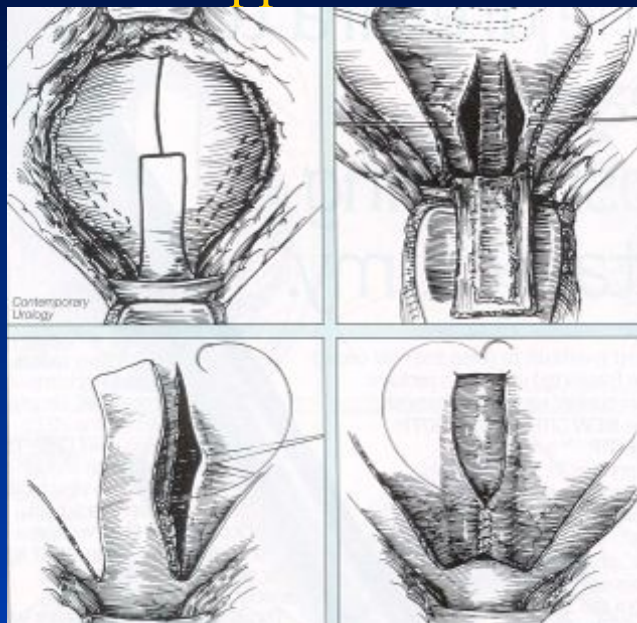
Modified Kropp



The Kropp Procedure

- 24 pts (25 procedures)
 - 14 female/ 10 male
 - 22 neurogenic/ 1UG sinus/ 1 trauma
- Continence 80%
- Complications
 - * Diff Cath -11/24, Perf (9/24), Stone (8/24), re-op tube(4/24), re-op augment (11/24)
- Recommended that all have augmentation
 - » Nill et al. J. Urol. 1990
- Perhaps too effective

Pippi-Salle



Pippi-Salle

- 17 pts (10 female:7 male)
 - * 13 neurogenic
 - * 4 exstrophy
- Augment 13/17
- Continence 12/17 (70%)
- Complications
 - * Fisutla 2/17, 2/17 New reflux, 2/17 unable to cath
- Augmentation should be performed when there is diminished bladder compliance
 - » Pippi Salle et al. J Urol. 1997

Young-Dees-Leadbetter

- 38 pts (25 female:13 male)
- Neurogenic Incontinence
- 35/38 Augmentation
- Continence
 - 23/38 (60%) -initial
 - 30/38 (79%)- after 2^o procedures
- Complications
 - 4-difficult cath, 5-perf, 11-revision
- 2 pts. void spontaneously
- Rec- AUS over YDL

» Donnhoo et al. J Urol. 1999

Fascial Slings

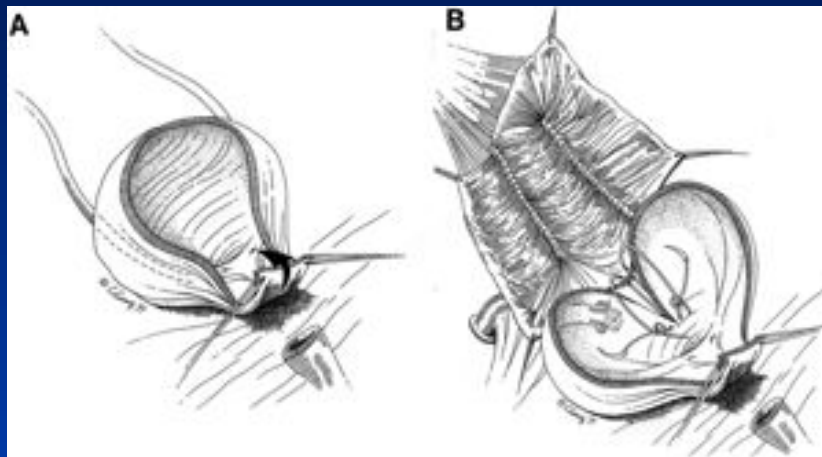
- 18 pts- 16 MMC/ 2 Spinal cord injury
 - 10-F/ 8-M
 - Mean age 14.4 (8-18)
 - 2 previous Augment/ 4 concomitant Augment
- Sling position:
 - F-BN / Urethra
 - M- Distal prostatic urethra
- 14/18 dry (1 pad or less)
- No change in mean DLPP (23.2 vs 23.2)
- Increase in mean ALPP (41 vs 66)

» Austin et al. J. Urol 165:2369, 2001

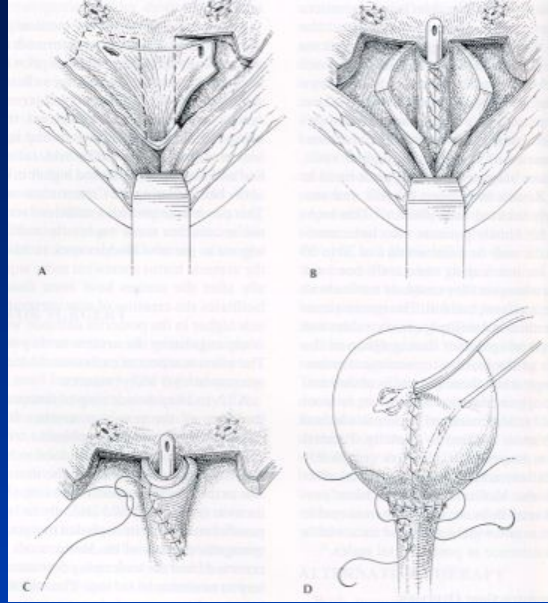
AUS

- Poor results if prior BN surgery
- Infection or Erosion-10-20%
- High revision rate due to mechanical problems- 25-85% (earlier series worse)
- Good continence rate- 85-97%
- Compatible with voiding or CIC (1:4)
- 40-60% will ultimately require augmentation
 - May be performed simultaneously
 - DH will lead to poor compliance

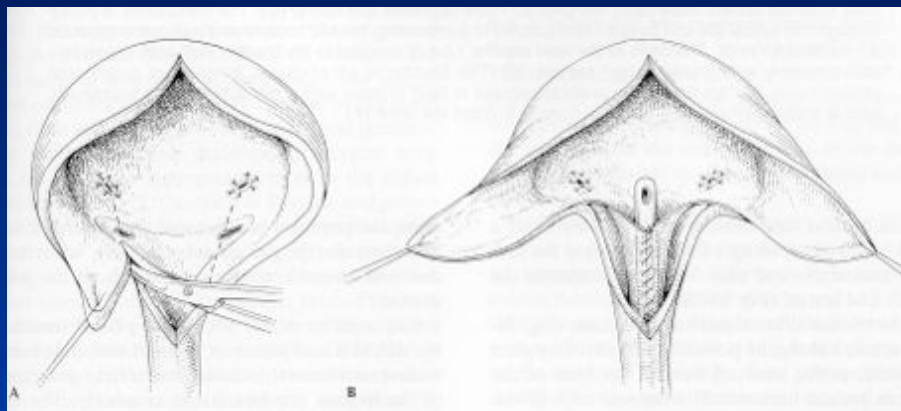
Bladder Neck Closure



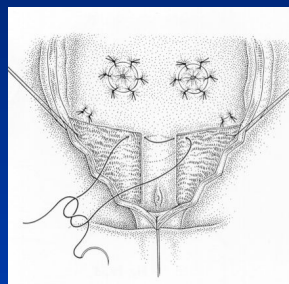
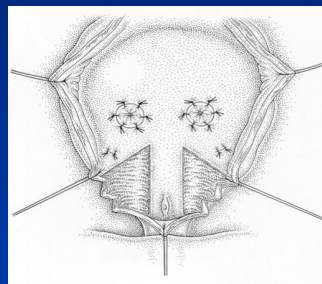
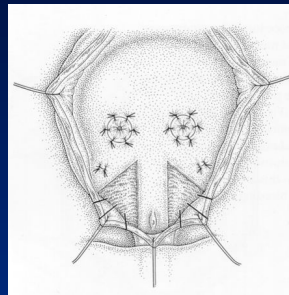
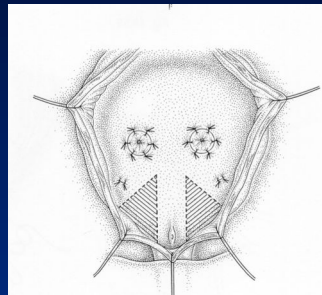
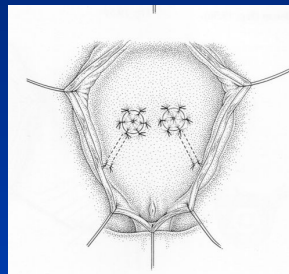
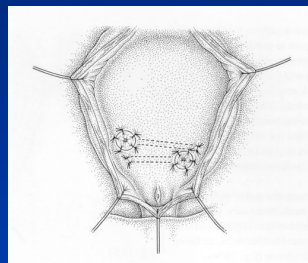
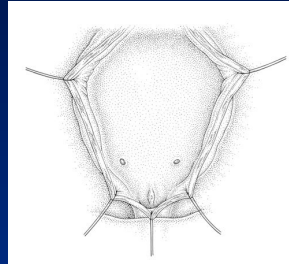
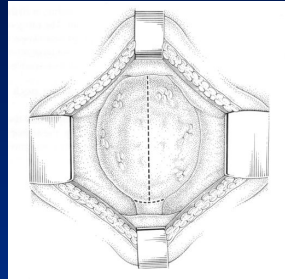
Young-Dees BNplasty

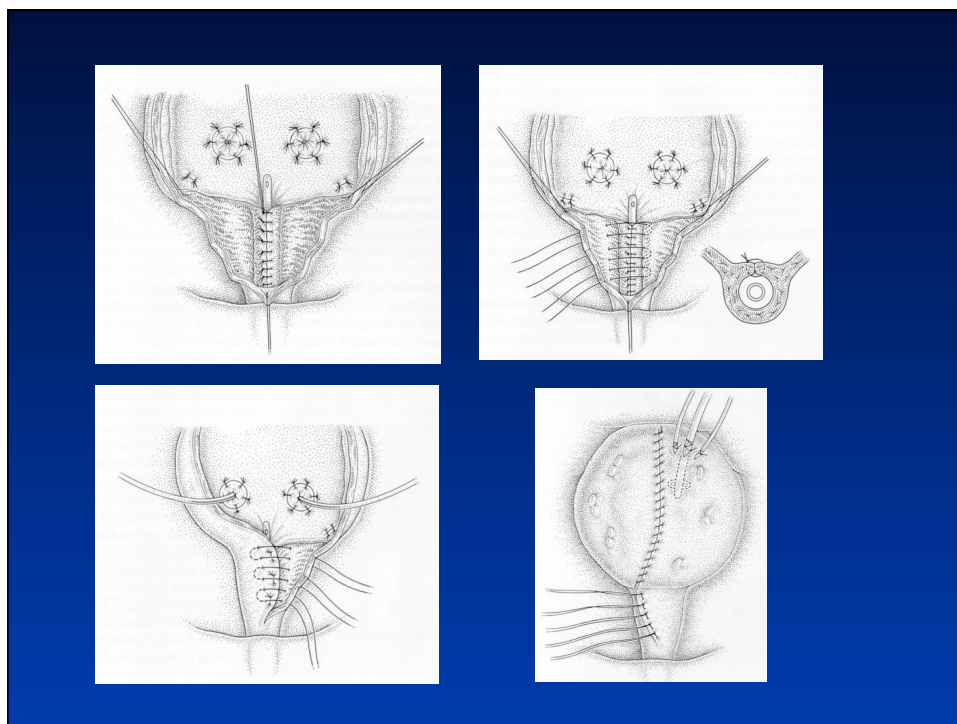


Leadbetter Procedure



Modified YDL





MMC/ Exstrophy/ Bladder Ca

- Recent publications have suggested that bladder augmentation increases risk of bladder cancer
 - Sorgel et al. J Urol. 172:1649, 2004
 - Castellan et al. J Urol. 178:1771, 2007
 - Recommended yearly cysto and screening for bladder Ca.
- Newborns with bladder exstrophy have been found to have polyps with cystitis glandularis
 - Novak et al. J Urol. 174:1522, 2005
- Known increase in risk for bladder cancer in SCI patients (indwelling cath)

MMC and Bladder Ca: U of Iowa Series

- 1995-2006
 - * 10 patients
 - 8 Female
 - 2 Male
- Median Age 37 (20-60)
- Presentation
 - * Gross hematuria
 - * Difficulty cathing/ changing cath
 - * UTIs/ Urosepsis
 - * Sterile Pyuria

Austin, Elliott, Cooper. J Urol.178:798, 2007.

MMC and Bladder Ca: Univ. of Iowa Series- II

- Path
 - * TCCa – 60%
 - * AdenoCa – 20%
 - * Squamous Cell Ca – 20%
- Stage
 - * Ta/T1- 0%
 - * T2 - 10%
 - * T3/4 - 90%
 - * Nodes (+) - 40%

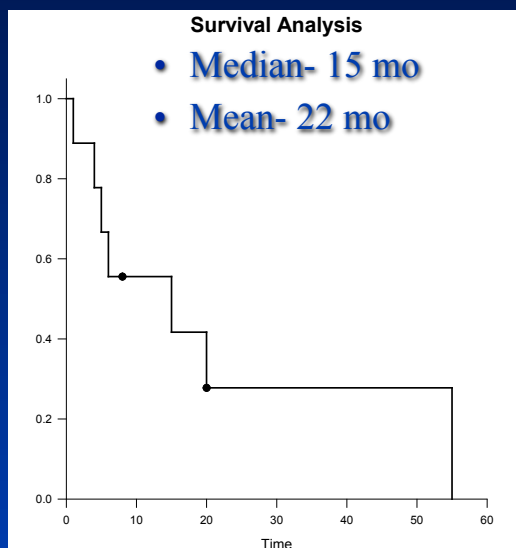
MMC and Bladder Ca: Univ. of Iowa Series- III

- Treatment
 - * Radical Cystectomy -80%
 - * Radiotherapy- 20%
 - * Died at presentation (age 20-F)
 - * Sigmoidectomy- 10%
 - * Chemotherapy 30%

MMC and Bladder Ca: Univ. of Iowa Series- IV

- Median Survival – 15 months
- Only 1 patient had T2N0 disease and is alive and disease free at 20 months
- 6/10 patients had regular, yearly visits with a urologist
 - * UA, Labs, Ultrasound
- **Only 2/10 (20%) had bladder augmentation (8,14 yrs)**

MMC and Bladder Ca: Univ. of Iowa Series- V



Combined Published Experience: MMC and Bladder Cancer

- Review of all published cases (additional 13 published)
- 23 patients
 - * 8 Male
 - * 14 Female
- Age
 - * Median 35 years (20-73)
 - * Mean 38 years
- Augmentation – **10 pts (43%)**
 - * Colon-3
 - * Ileum-3
 - * Stomach-4

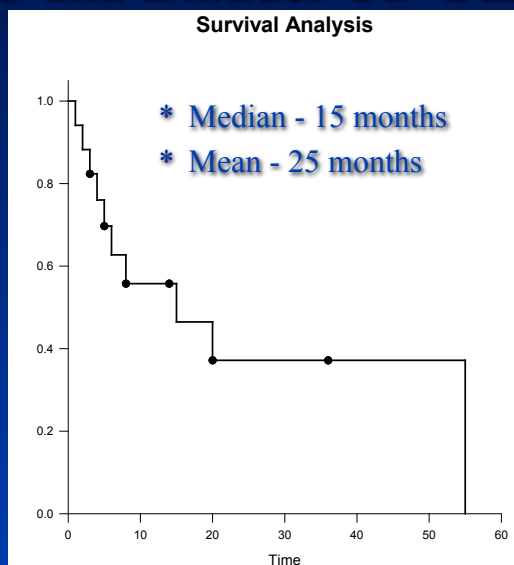
Combined Published Experience: MMC and Bladder Cancer-II

- Histology
 - * TCCa – 57%
 - * Squamous CCa – 17%
 - * AdenoCa – 13%
 - * **Gastric Ca – 13% (3/4 Gastrocystoplasty)**
- Tumor Stage (15pts)
 - * T1 – 6% (Gastric Ca)
 - * T2 – 14%
 - * **T3 – 40%**
 - * **T4 – 40%**
 - * **(+) Nodes – 33%**

Combined Published Experience: MMC and Bladder Cancer-III

- Years From Augmentation to Diagnosis
 - * Median 13.5 years (8-21)
- Survival
 - * Median - 15 months
 - * Mean - 25 months

MMC and Bladder Ca- Survival



MMC- Iowa's Screening Program

- 20 years old NGB on CIC
- > 10 years out from bladder augmentation
 - * Yearly visits
 - * Cystoscopy
 - * Renal/Bladder ultrasound
 - * Urine cytology.
 - * PRN visits for gross hematuria, UTIs, difficulty cathing, or new bladder Sxs.

Is Screening Going to Help?

- Low incidence
 - * Bowel Augmentation 1.2- 3.8 %
 - * Gastrocystoplasty 1-10%
- Gastrocystoplasty- 2/4 patients had cysto within 1 year of diagnosis
- MMC patients- 6/10 followed yearly by urologist
- Is more than annual visits going to be necessary?
Will it lead to a stage migration?

Summary

- Atypical symptoms/ Presentation
 - * **Need to suspect the diagnosis in young adults**
- Aggressive tumor/ advanced stage
- Poor Survival
- Bladder augmentation may be a risk factor
- Need lifelong follow-up