How Long Term Outcomes Have Changed Our Approach in the Management of Patients with Spina Bifida

Mark P. Cain, MD, FAAP
Robert Garrett Professor
Chief Of Pediatric Urology
Riley Hospital for Children at IU Health

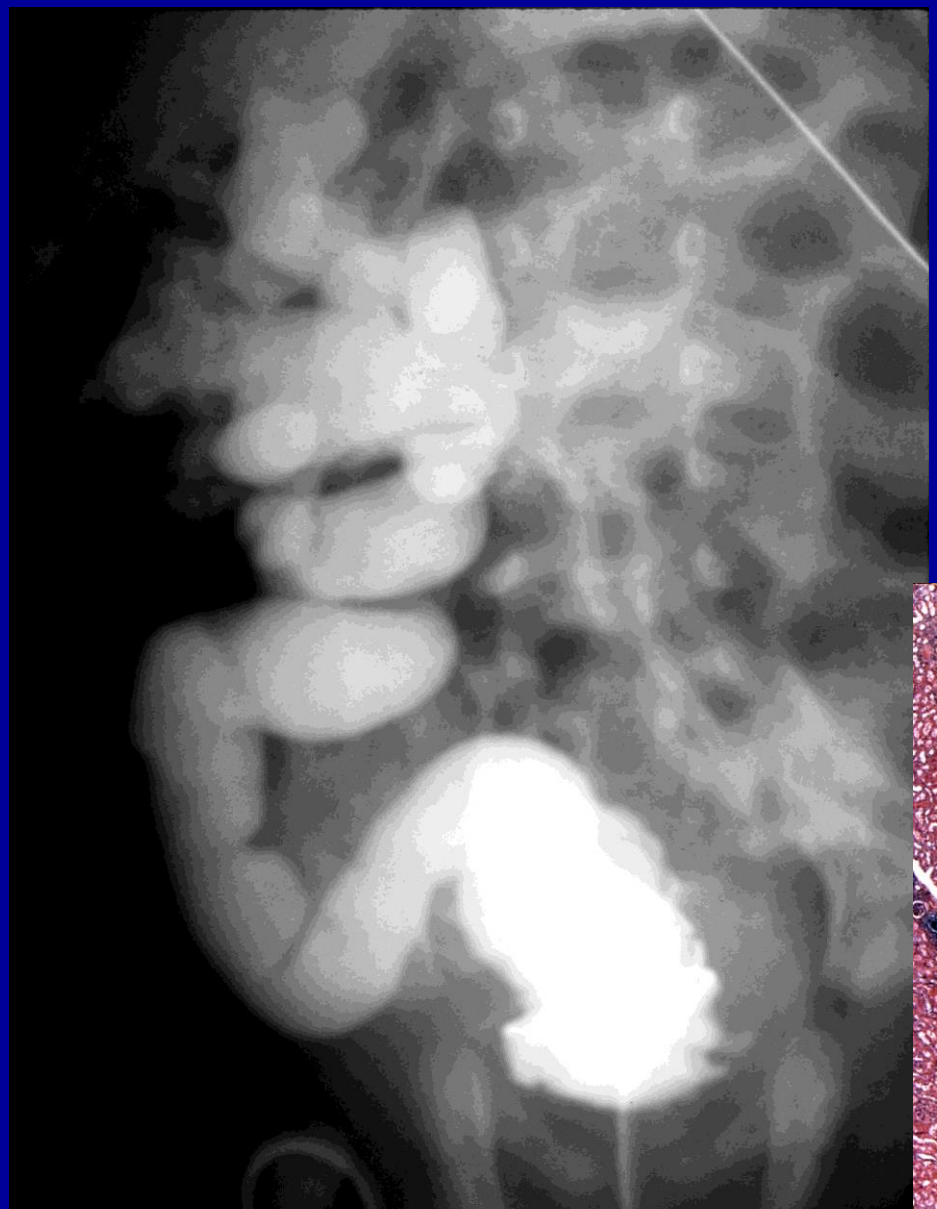
Riley Hospital for Children
Indiana University Health

I have no financial disclosures.

I

Problem: Med/Surgical and Social

VUR and Bladder Pressure = Renal Disaster



Spina Bifida - Urologic Management

Options for Intervention

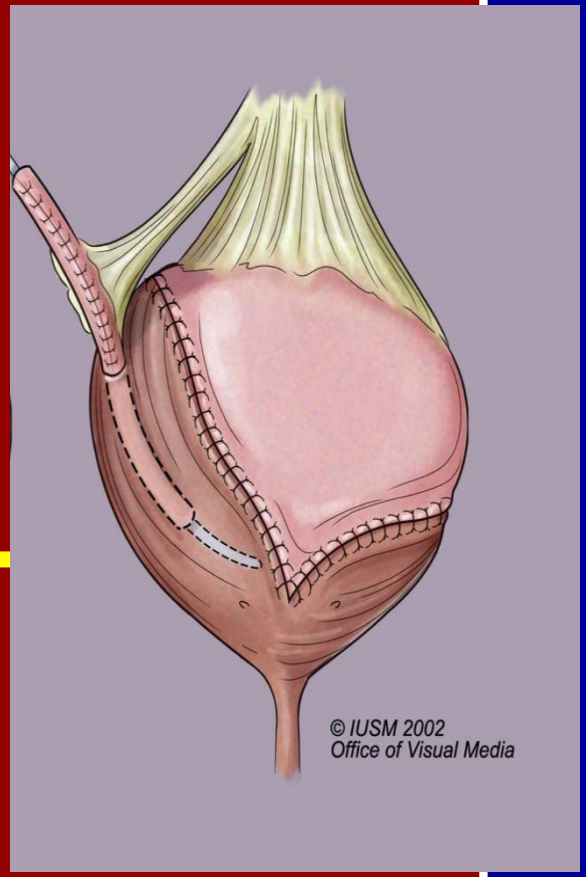
- **Bladder Relaxants**
- **Clean Intermittent Catheterization**
- Urethral Overdilation
- Botox Injection
- Nocturnal Bladder Drainage
- Bladder Electrical Stimulation
- Sacral Spinal Cord Stimulation
- Vesicostomy
- Bladder Reconstructive Surgery

Management of Neuropathic Bladder and Bowel: Historical Surgical Pathway to Continence

Evolution of continence:

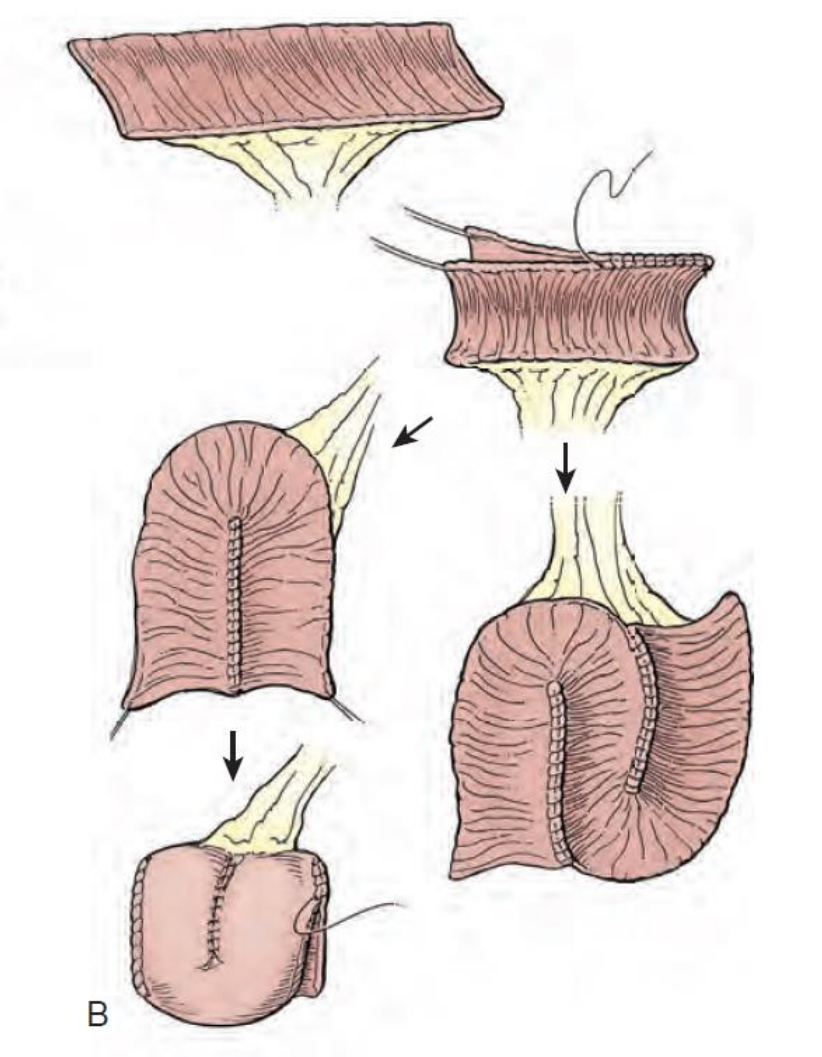
- Intermittent catheterization
- Bladder outlet resistance
- Bladder augmentation

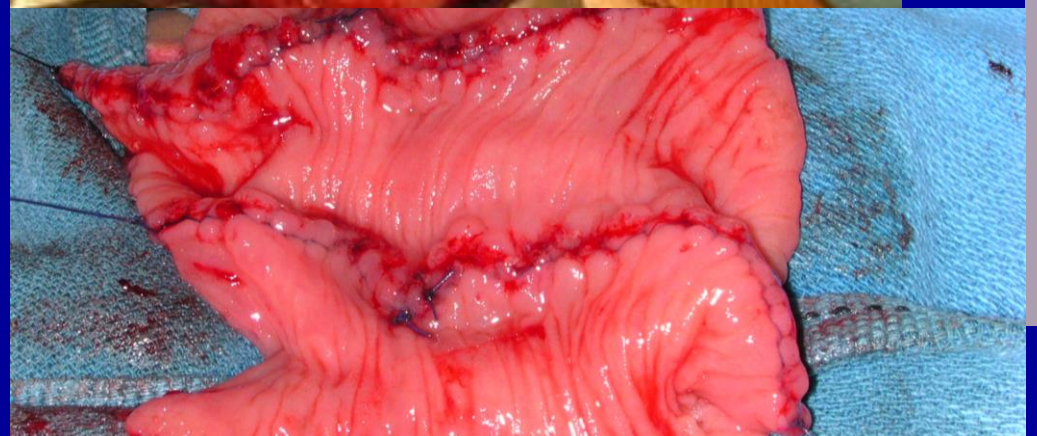
- Mitrofanoff procedure
- MACE procedure

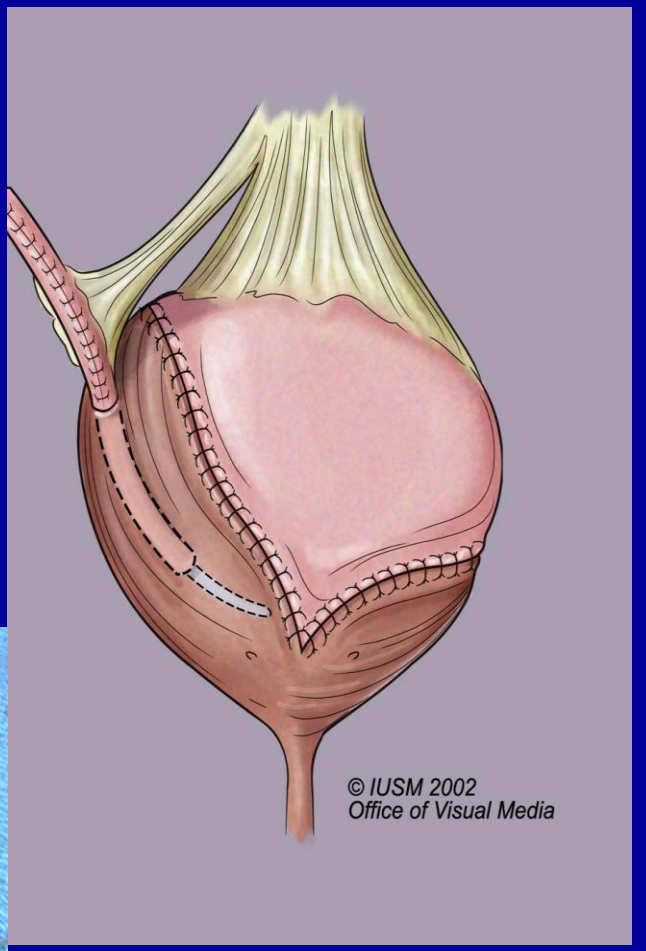


BLADDER AUGMENTATION

ERA OF RECONSTRUCTION







Historical Evaluation of Incontinence and Surgical Results

- Surgeon driven
- Surgeon evaluated
- Parents as proxy for children's outcome
- No validated, disease specific surveys
- No long term follow up into teen/adult life
- Outcomes based on "Pad or diaper use", dry interval (3-4 hours), didn't separate "moving parts" of complex operations

Pediatric Urology at Indiana University

Bowel In LUR : Indiana

1978 – 2018

40 Years

12 Surgeons

>75 Journal articles

>20 Chapters

~ 800 Augments

> 900 Cath Chan.

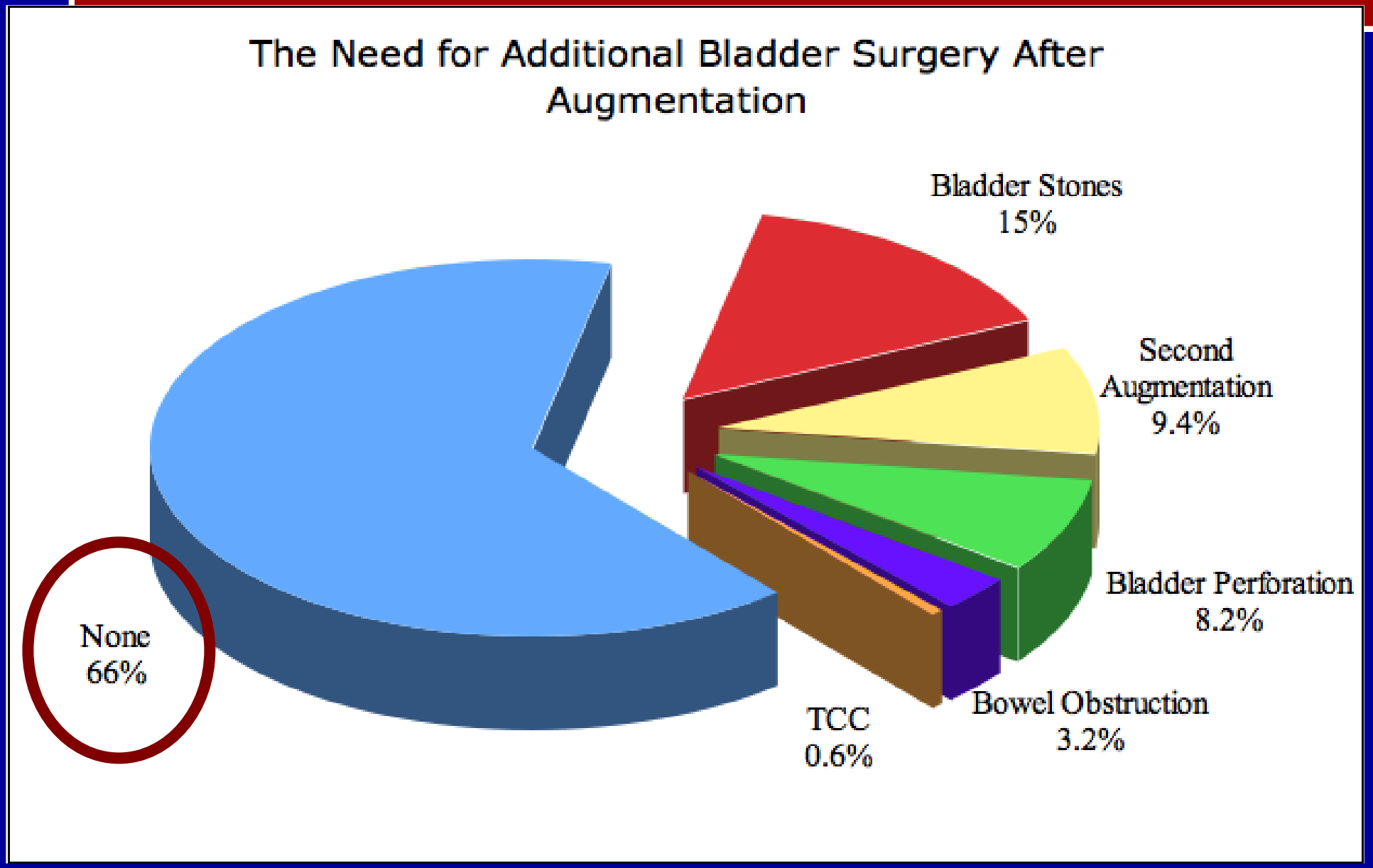
Rick Rink, MD, FAAP, FACS
Mark Cain, MD, FAAP
Martin Kaefer, MD, FAAP
Rosalia Misseri, MD, FAAP
Ben Whittam, MD, MS
Kate Hubert, MD, MPH
Konrad Szymanski, MD, MPH
Shelly King, CNP
Melissa Young, CNP
Hillary Risk, CNP
Taylor Wang, CNP

Bob Garrett, MD, Mike Mitchell, MD, Mike Keating, MD,
Mark Adams, MD, Tony Casale, MD, Kirstan Meldrum, MD

One Surgeon's View On:

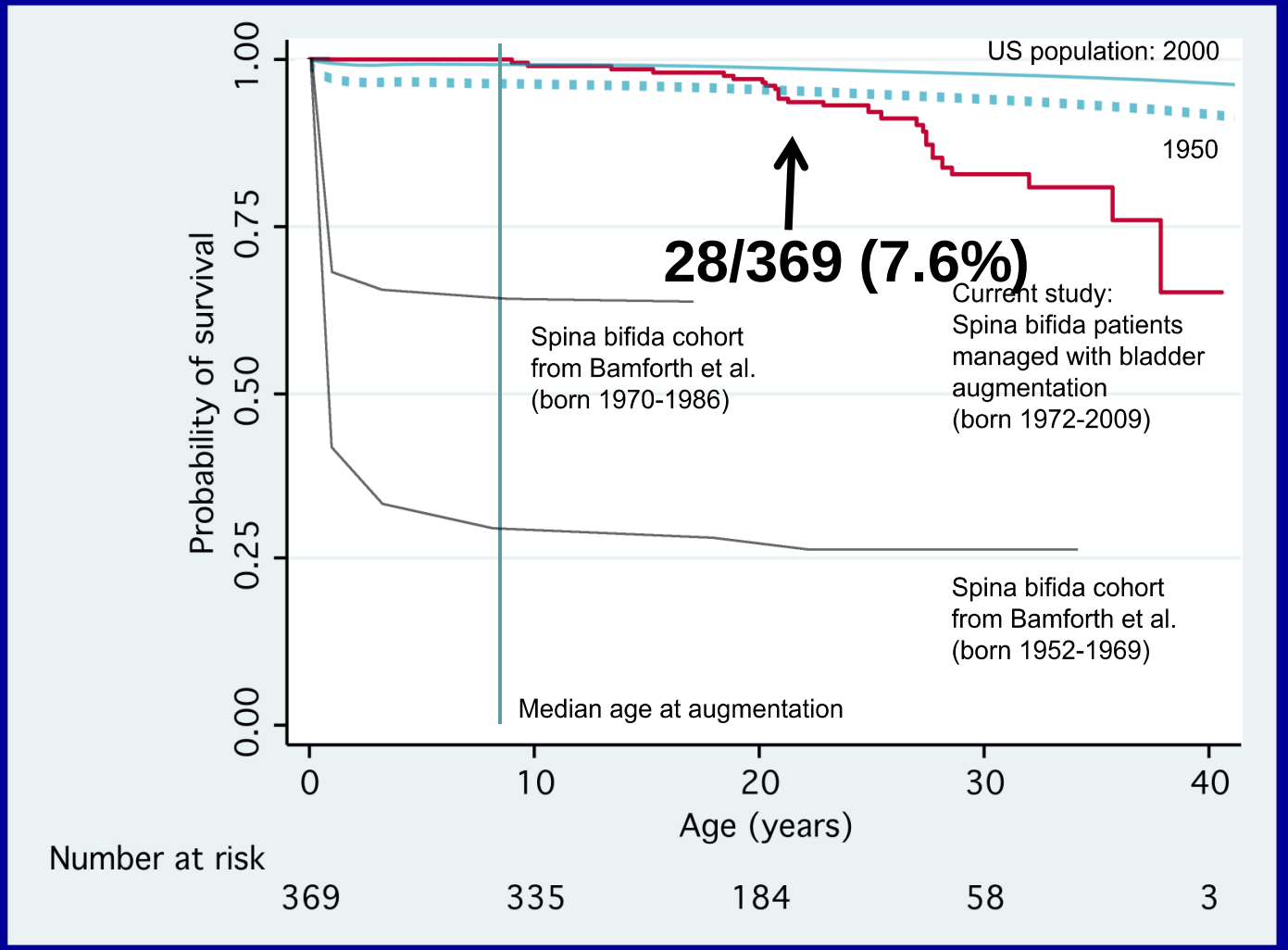
- Why large databases are important…..
- Why long term follow up is important in decision making for childhood diseases….
- Why we need to ask our patients what they want at sequential times over their young lives…..
- Why we need to think differently about health care delivery for chronic and congenital diseases….

Bladder Complications: Indy 500



Metcalfe et al JUrol, 2006

Survival in the Era of Aggressive Bladder Management



Mortality after Bladder Augmentation in Children with Spina Bifida

Konrad M. Szymanski,* Rosalia Misseri, Benjamin Whittam, Cyrus M. Adams, Jordan Kirkegaard, Shelly King,† Martin Kaefer, Richard C. Rink and Mark P. Cain

From the Division of Pediatric Urology, Riley Hospital for Children at IU Health, Indianapolis, Indiana

Causes of death

Cause of death	Total (n=28)
Infection	16
Ventriculoperitoneal/atrial shunt-related infection	7
Decubitus ulcer fasciitis	2
Peritonitis	2
Urosepsis	2
Pneumonia	1
Sepsis (unknown source)	2
Non-infectious pulmonary	3
Congenital cardiac disease	2
Renal failure	2
Ventriculopleural shunt obstruction	1
Iatrogenic cardiac tamponade	1
Motor vehicle collision	1
Suicide	1
Unknown	1

- Non-adherence with CIC
- Pre-existing renal insufficiency

We Are All Victims of Our Survival

- 14th Century – 30% of population died of plague
- 1900s - Pre ABX, Vaccines, Sanitation
  - Infection (pneumonia), TB, GI infections
- 1990 – Modern medicine era
  - MI, CA, Stroke (60%), COPD, Dementia*
- 2018 - prevention + gene therapy
- Spina Bifida patients (and us) now living longer
- Quality of Life matters more, especially later…..

Does Surgery Matter?

Indications Different

Does Continence Matter?

Urologic Management
GOALS OF INTERVENTION:

Renal

Bladder

Prevent Infection

Urinary/Fecal Continence

Transition to Independent Care

QUALITY OF LIFE…

Don’t turn a social problem into a medical problem….

Health Related Benefit after Reconstruction for Urinary/Fecal Incontinence in Children: Parent Perspective

- 300 families evaluated, 120 responses
- Glasgow Inventory (validated) + 6 specific questions
- Statistically ~~significant improved~~ HRB
- What changed life most:
  - 31% MACE, 44% aug/channel, 25% both
- What did parents find most important:
  - 48% stool continence
- 68% needed less/no assistance with care

Strine, Misseri et al, J Urol, 2015

A Piece of My Mind

April 17, 2018

Mentoring Millennials

Jennifer F. Waljee, MD, MPH, MS[1]; Vineet Chopra, MD, MSc[2]; Sanjay Saint, MD, MPH[3]

≫ Author Affiliations | Article Information

JAMA. 2018;319(15):1547-1548. doi:10.1001/jama.2018.3804

Millennials have been shaped by a profound expansion of information technology, enhanced social networking, and a connected global culture. Although sometimes labeled as impatient, distracted, overly socialized, and entitled, millennials could also be characterized as deeply empowered, collaborative, and innovative. These generalizations, however, can lead to conflict and misunderstanding, particularly in environments such as hospitals where apprenticeship and hierarchy are the norm.

Mentorship is the cornerstone of academic medicine. A mentor is defined as an advisor characterized by altruism, expertise, patience, and experience. In many ways, graduate medical education has adapted to millennials through the expansion of online and video-based learning resources, disease-based educational curricula, abbreviated work hours, and team-based care models.[2] However, mentorship strategies for millennial faculty members, residents, and medical students are not well understood. Indeed, we have personally witnessed generational differences leading to frustration, miscommunication, and attrition in these mentor-mentee dyads. Consider 3 common scenarios.

Riley Pediatric Urology Team 2018

Does urinary incontinence affect HRQOL in adults with SB?

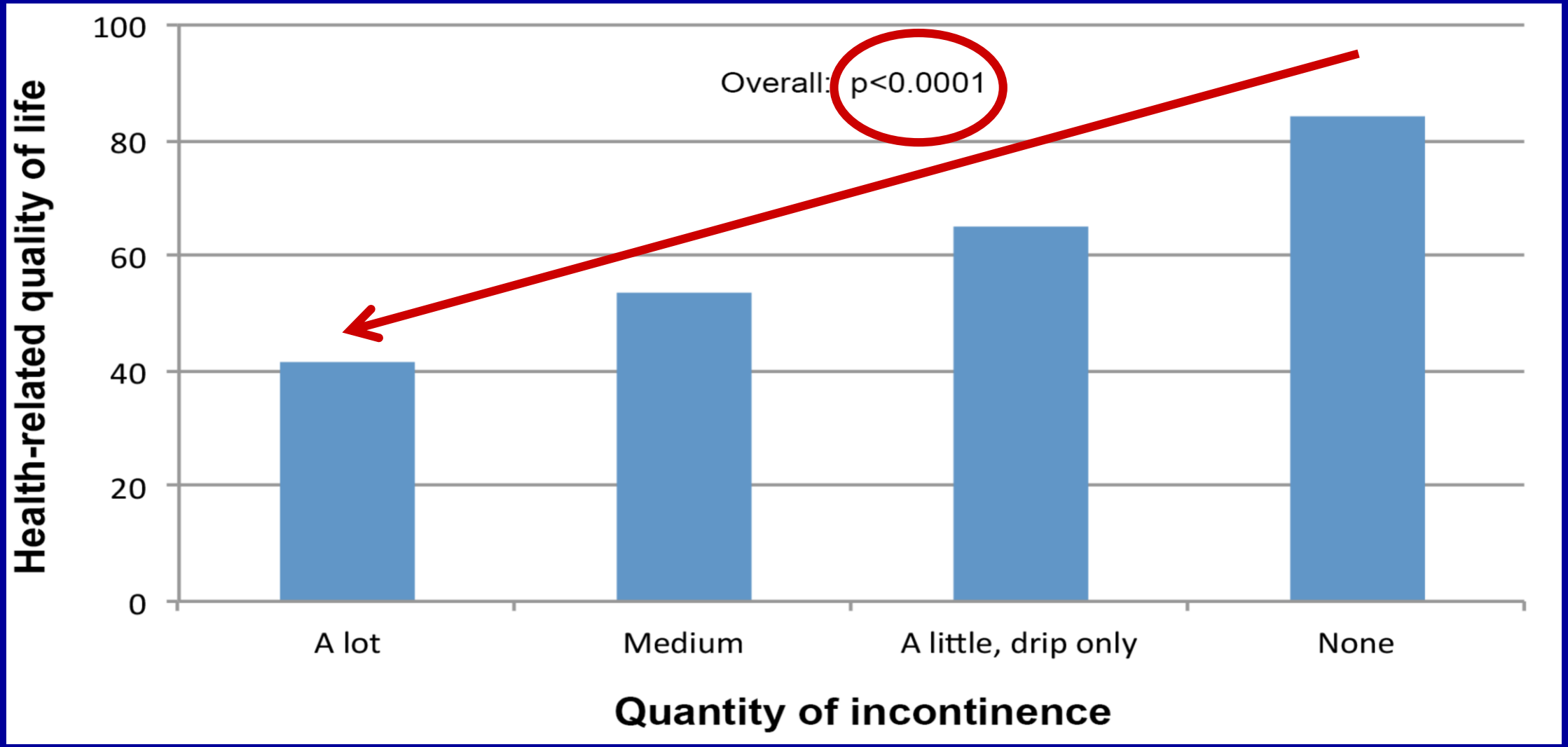
- Two studies report no difference[1,2]
  - Only measured dry interval
  - Did not use SB-specific questionnaire
- Adults with SB recruited online and clinic
  - Daytime UI (dry interval, quantity, undergarments)
  - SB-specific HRQOL (QUALAS-A[3])

1. Lemelle, J. L., Guillemin, F., Aubert, D. et al.: Quality of life and continence in patients with spina bifida. Qual Life Res, 15: 1481, 2006
2. Vu Minh Arnell, M., Seljee Svedberg, K., Lindehall, B. et al.: Health-related quality of life compared to life situation and incontinence in adults with myelomeningocele: is SF-36 a reliable tool? J Pediatr Urol, 9: 559, 2013
3. Szymanski, K. M., Misseri, R., Whittam, B. et al.: QUAlity of Life Assessment in Spina bifida for Adults (QUALAS-A): development and international validation of a novel health-related quality of life instrument. Qual Life Res, 2015

Population characteristics (n=518 Adults; 150 Adolescents; 250 Kids) Validated for each age group

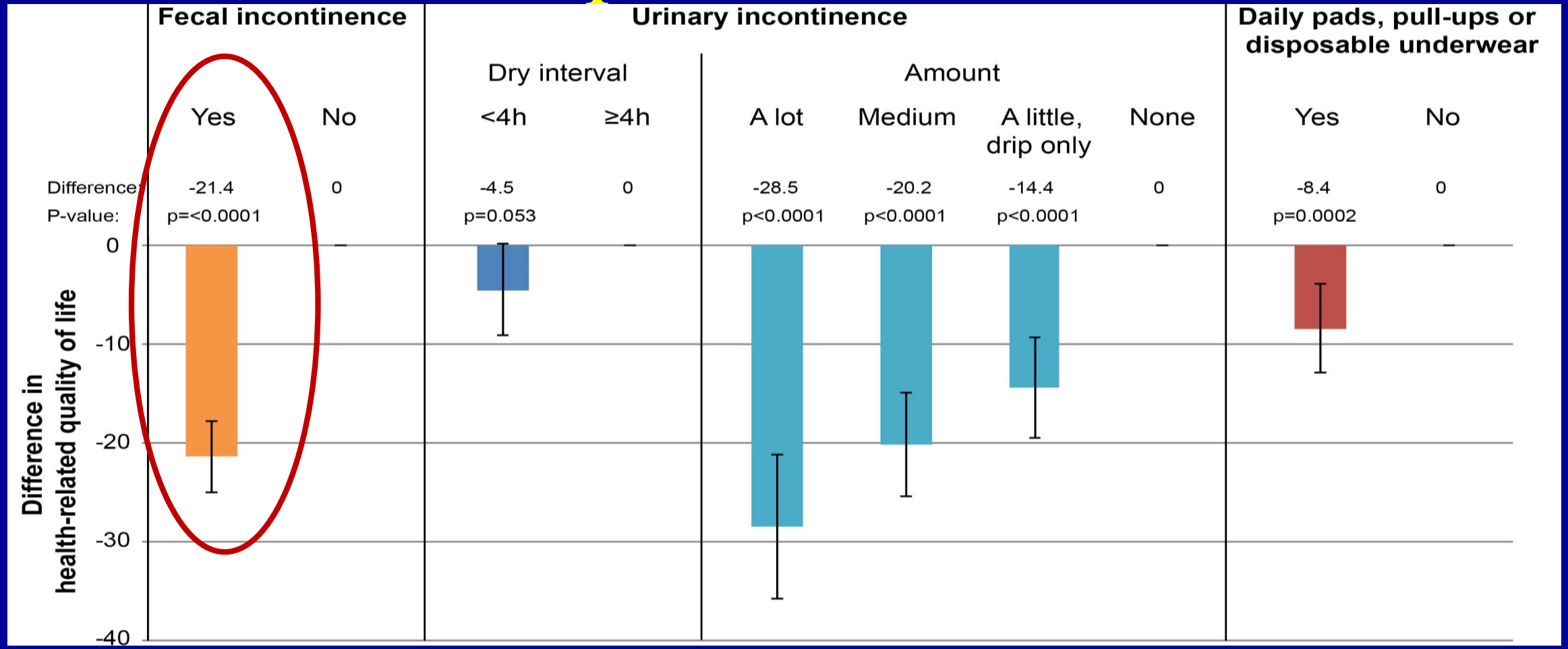
Median age: 31.9 years (78.7% live in US)

HRQOL and urinary incontinence: Does It Matter to Adult Spina Bifida Patients?



Szymanski et al, AUA/JUrol, 2016

Does Urinary/Fecal Incontinence Matter to Adult Spina Bifida Patients?



Szymanski, 2016

HAVE WE HELPED CHILDREN?

Quality of life

QUAlity of Life Assessment in Spina bifida for Adults (QUALAS-A): development and international validation of a novel health-related quality of life instrument

Konrad M. Szymanski, Rosalia Misseri, Benjamin Whittam, Sonia-Maria Raposo, Shelly J. King, Martin Kaefer, Richard C. Rink, et al.

Quality of Life Research
An International Journal of Quality of Life Aspects of Treatment, Care and Rehabilitation - Official Journal of the International Society of Quality of Life Research

ISSN 0962-9343
Volume 24
Number 10

Qual Life Res (2015) 24:2355-2364
DOI 10.1007/s11136-015-0988-5

Springer

Pediatric Urology

Quality of Life Assessment in Spina Bifida for Children (QUALAS-C): Development and Validation of a Novel Health-related Quality of Life Instrument

Konrad M. Szymanski, Rosalia Misseri, Benjamin Whittam, David Y. Yang, Sonia-Maria Raposo, Shelly J. King, Martin Kaefer, Richard C. Rink, and Mark P. Cain

OBJECTIVE To develop and validate a self-reported health-related QUAlity of Life Assessment in Spina bifida for Children (QUALAS-C).

METHODS We drafted a 27-question pilot instrument using a patient-centered comprehensive item generation and refinement process. It was administered to a sample of children 8-12 years old with spina bifida (SB) recruited online via social media and in person at an outpatient SB clinic (January 2013-September 2014). Healthy controls were recruited at routine pediatrician visits. Validation and final questions were determined based on clinical relevance, high loadings on factor analysis, and domain psychometrics. Children with SB also completed the validated generic Kidscreen-27 instrument.

RESULTS Median age of 150 participants was 9.6 years (60.7% male, 72.7% Caucasian), similar to 46 controls ($P \geq .10$). There were 97 online and 53 clinic participants (89.0% and 84.2% of eligible, respectively). Face and content validities of the 2-domain, 10-question QUALAS-C were established by patients, parents, and experts. Internal consistency and test-retest reliability was high for the Esteem & Independence and Bladder & Bowel domains (Cronbach's alpha: 0.72-0.76, ICC: 0.74-0.77). Correlations between QUALAS-C domains were low ($r = 0.51$), indicating that QUALAS-C can differentiate between two distinct health-related quality of life components. Correlations between QUALAS-C and Kidscreen-27 were also low ($r \leq 0.44$). QUALAS-C scores were significantly lower in children with SB than without ($P < .0001$).

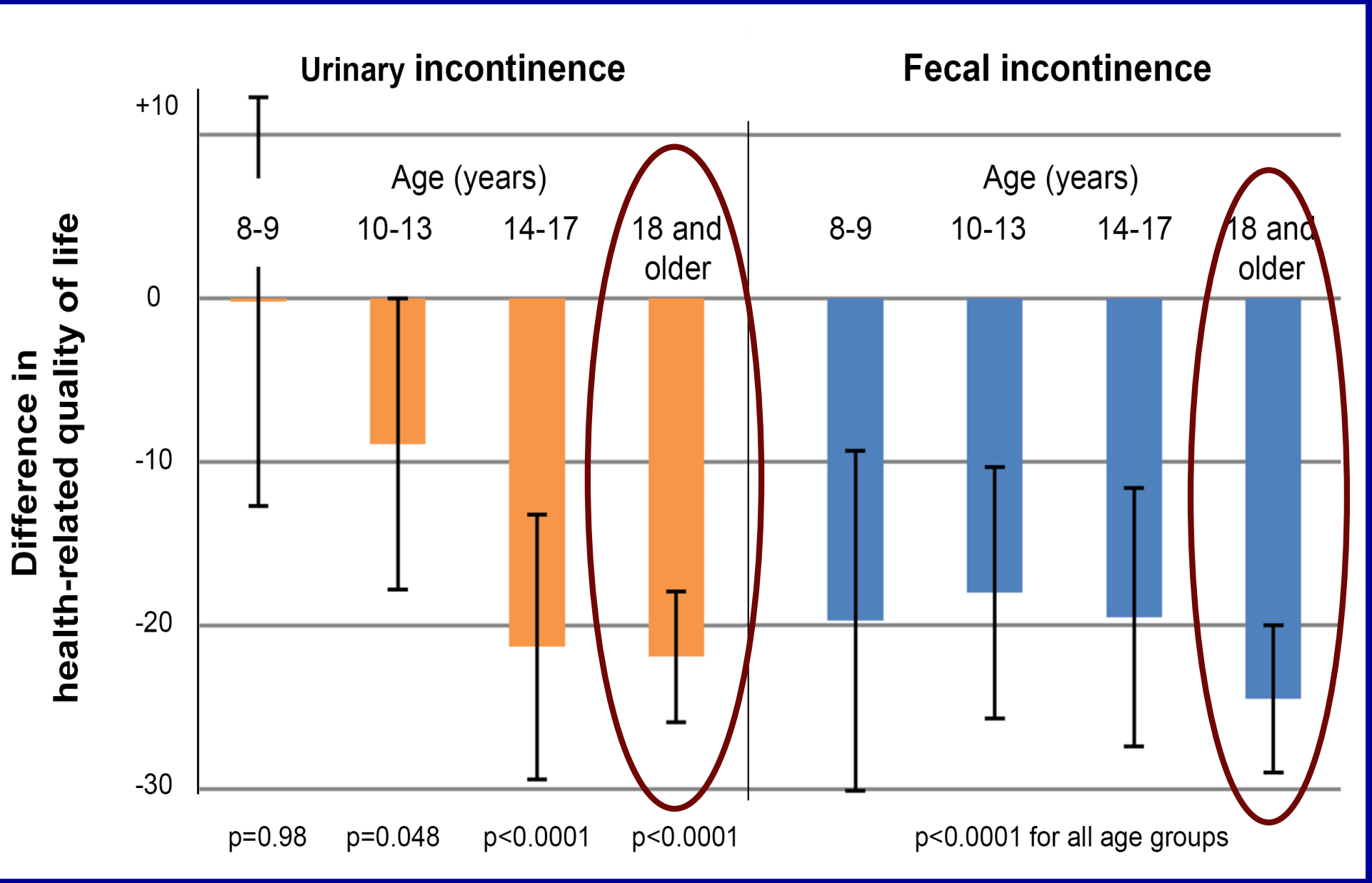
CONCLUSION QUALAS-C is a short, valid health-related quality of life tool for children with SB. It will be useful in clinical and research settings. UROLOGY 87: 178–184, 2016. © 2015 Elsevier Inc.

Spina bifida (SB) is the most common congenital anomaly of the central nervous system, affecting 3.4 per 10,000 live births in the United States.[1] Children surviving infancy face neurological, neurosurgical, orthopedic, and urological challenges. Unfortunately, studies on health-related quality of life (HRQOL) in children with SB tend to be small, single-institutional studies using limited, poorly-validated, or non-validated, instruments.[2-6] Others report patient satisfaction, rather than HRQOL.[7,8] Importantly, generic HRQOL instruments developed for healthy children[9,10] may be unable to capture small, but clinically important, differences because they were not designed to measure the impact of SB on HRQOL.[11] Moreover, no validated and comprehensive SB-specific HRQOL instrument exists which incorporates bladder and bowel domains.

SB-specific HRQOL is a component of quality of life that focuses on an individual's perception of the impact of SB on his/her physical and psychosocial functioning.[11,12] Assessing self-reported HRQOL is particularly important, as those with SB often report better HRQOL than perceived by their parents and caregivers.[13,14] For this reason, instrument development requires input from individuals with SB, their parents, and caregivers.[2,11] An ideal, clinically relevant HRQOL instrument focuses on HRQOL rather than physical function. It should also possess excellent psychometric properties and be condition specific, yet remain short

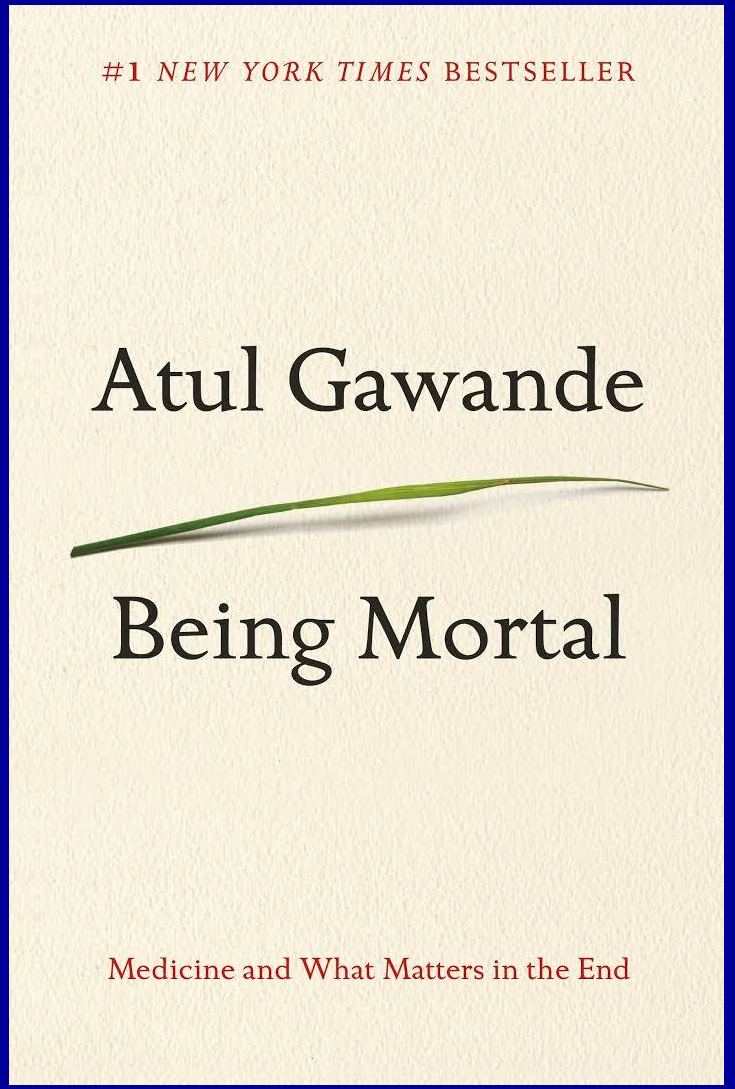
Financial Disclosure: *The authors declare that they have no relevant financial interests.*
Funding Support: *Funding was provided by the Indiana University School of Medicine's Department of Urology.*
From the Division of Pediatric Urology, Riley Hospital for Children at IU Health, Indiana University School of Medicine, Indianapolis, IN
Address correspondence to: Konrad M. Szymanski, M.D., M.P.H., Division of Pediatric Urology, Riley Hospital for Children at IU Health, Indiana University School of Medicine, 705 Riley Hospital Dr. #4230, Indianapolis, IN 46205. E-mail: szymanko@iupui.edu
Submitted: August 6, 2015, accepted (with revisions): September 21, 2015

Does Incontinence Matter at Different Ag



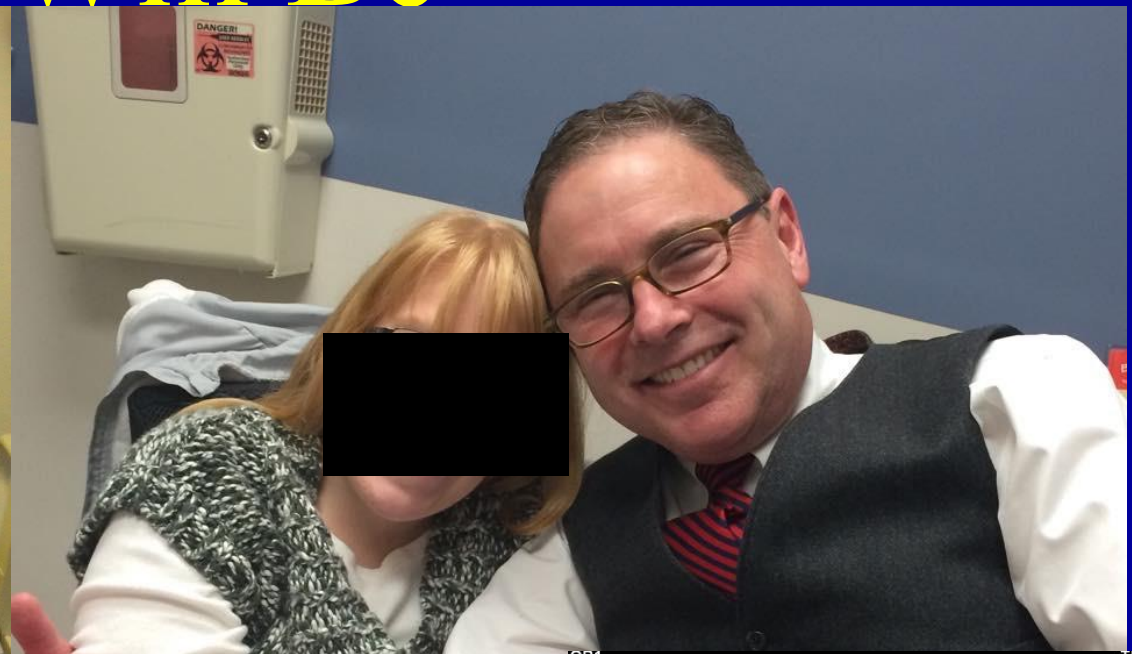
Szymansk
et al, 2017

Quality of Life is Just as Important for Kids as End of Life for Adults



I Didn't Always Think About My Patients As Eventual (Sexual) Adults…..

But They Will Be

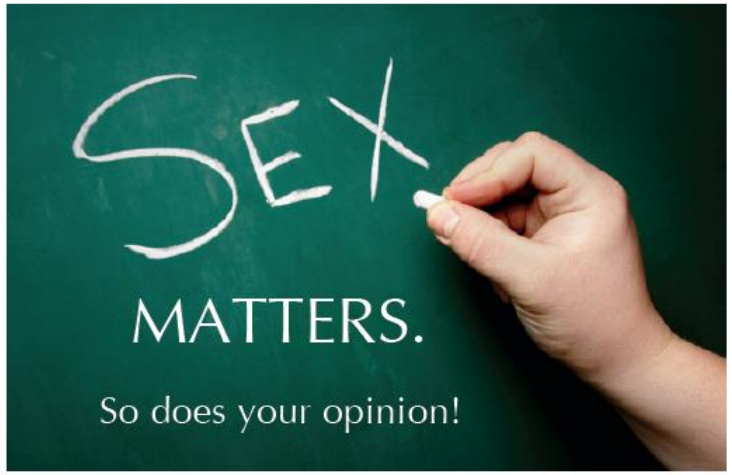


IU Research Regarding Sexuality

International anonymous online survey
Men and women 18+ years old with SB who spoke English
(2016-2017)

Recruitment:

- **SB clinics**
- **Online via social media**



If you are an adult with **spina bifida**, we want to hear from you!

Researchers at Indiana University School of Medicine are interested in learning more about the sexual health of adults with spina bifida.

If you are 18 or older and have spina bifida, you can participate, whether or not you ever had sex. We will ask questions about your background, your experiences with spina bifida, and your sexuality (feelings about sex, sexual behaviors, and desires for a family). It takes only 15-20 minutes.

All your answers are confidential and completely anonymous. No one you know will see your answers. Even the researchers will not be able to identify who you are or your answers. Thank you!

To go to the survey, go to this link: https://goo.gl/NocVRR **or scan this code:**

For IRB Office Use ONLY

Relationships

Single	**50.7%**	
In relationship		**47.8%**
Dating, hanging out		**4.4%**
Dating, in a relationship		**14.5%**
Living together, not married		**10.1%**
Married		**18.8%**
Divorced, separated	**1.5%**	

Lifetime sexual activities (ever events)

Masturbation alone	91.3%
Partnered non-genital contact (Cuddled, kissed, held hands)	85.5%
Partnered genital contact:	
Mutual masturbation	78.3%
Partnered intercourse	75.4%
Vaginal / anal	62.3% / 34.8%

Conclusions

- Partnered sexual activity common
  - May not involve penetrative intercourse
- Better ambulatory status associated with better sexual outcomes and erectile function
- ED is common in men with SB
  - PDE5Is are used frequently
  - May benefit this population

Urinary and Fecal Incontinence During Sexual Activity Is Common and Bothersome Among Adults with Spina Bifida

Joshua D. Roth, Devon J. Hensel, John S. Weiner, Rosalia Misseri, Konrad M. Szymanski

SCHOOL OF MEDICINE
INDIANA UNIVERSITY

Riley Hospital for Children
Indiana University Health

Duke Medicine

Baseline Data

Baseline Incontinence in last 4 Weeks

UI	66.7%	FI	50.7%

Bladder Management

CIC	44.6%
Urostomy	6.8%

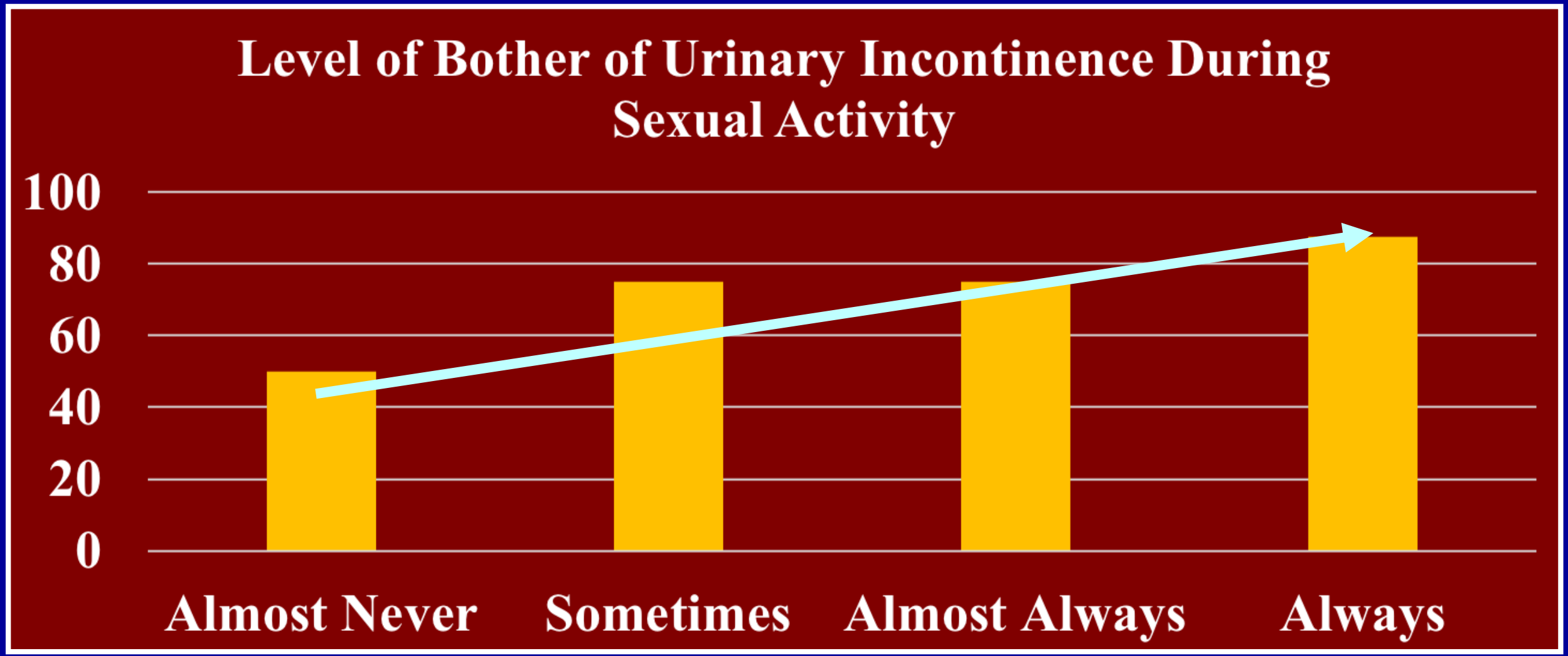
Stool Management

MACE	17.1%
Colostomy	1.1%

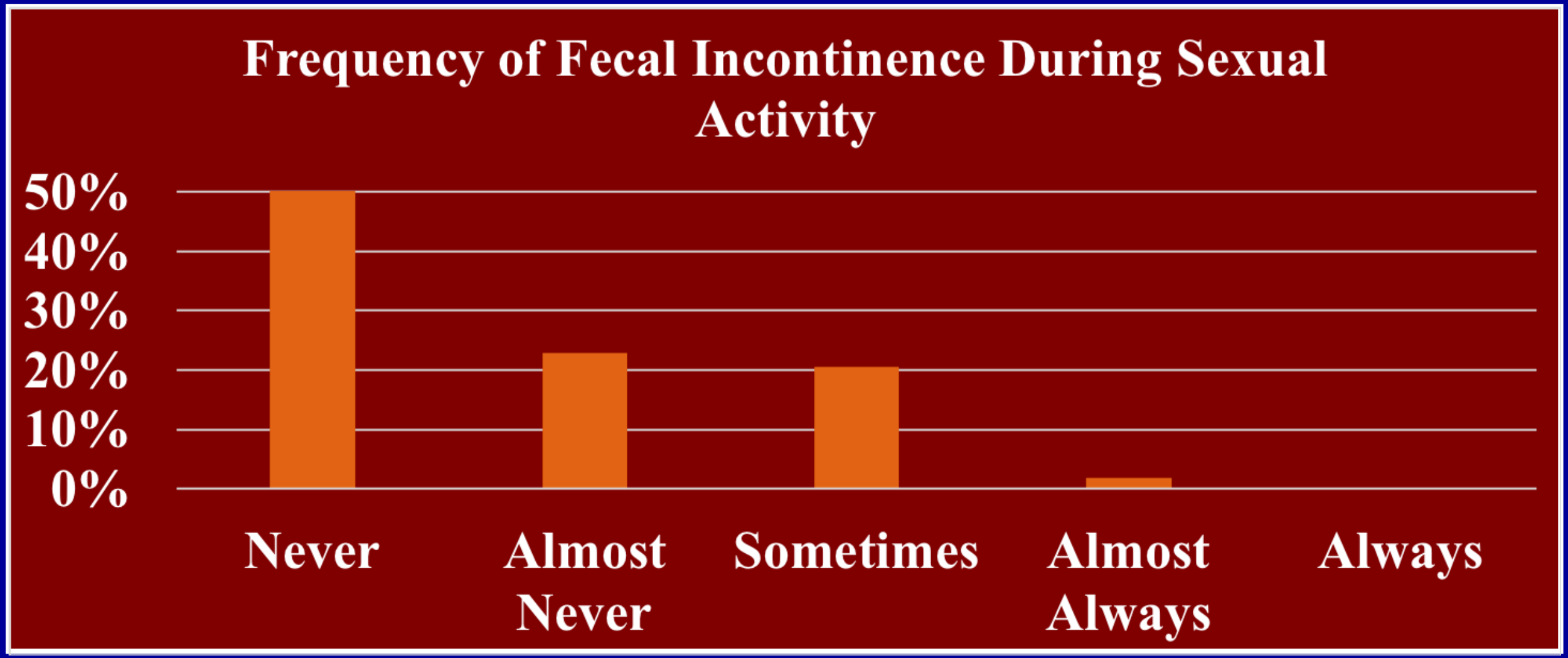
Adults Without Diversion

UI during Sex (64.6%) > FI/S (45.2%)	p<0.001
UI during Sex	
Women > men (76.0% vs. 52.2%)	p=0.002
Greater with underlying UI (79.0% v. 48.7%)	p=0.02
Not associated with age, shunt, ambulation, CIC	p≥0.32

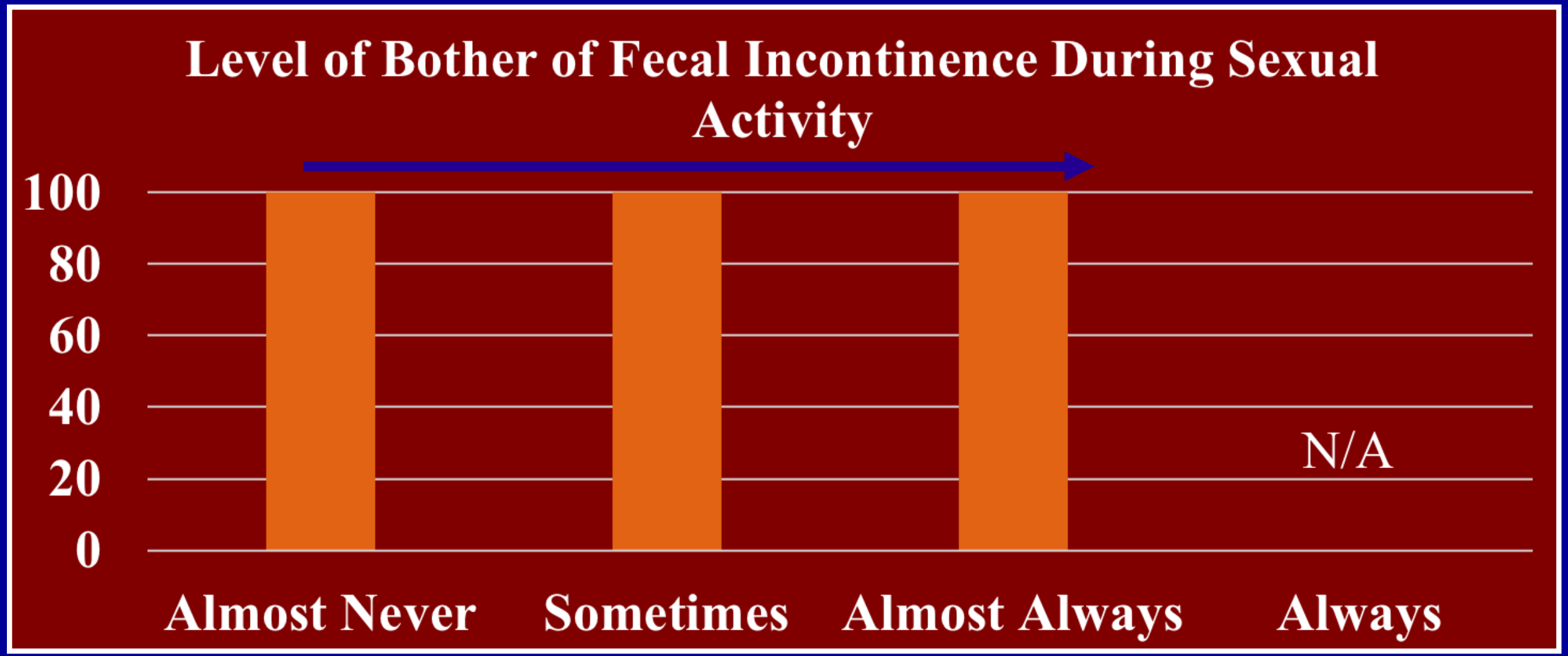
Adults Without Diversion



Adults Without Colostomy



Adults Without Colostomy



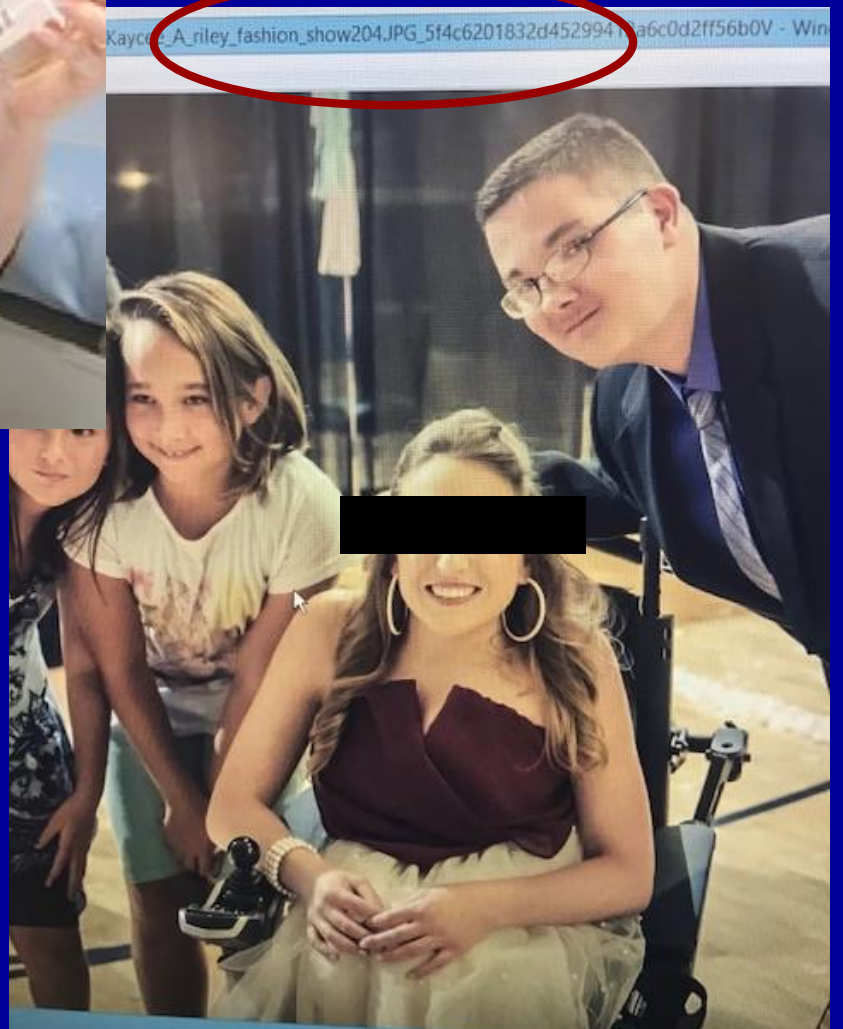
Conclusions

Incontinence during sexual activity is a common problem for adults with SB

Baseline incontinence is an independent, but not absolute predictor of UIS and FIS

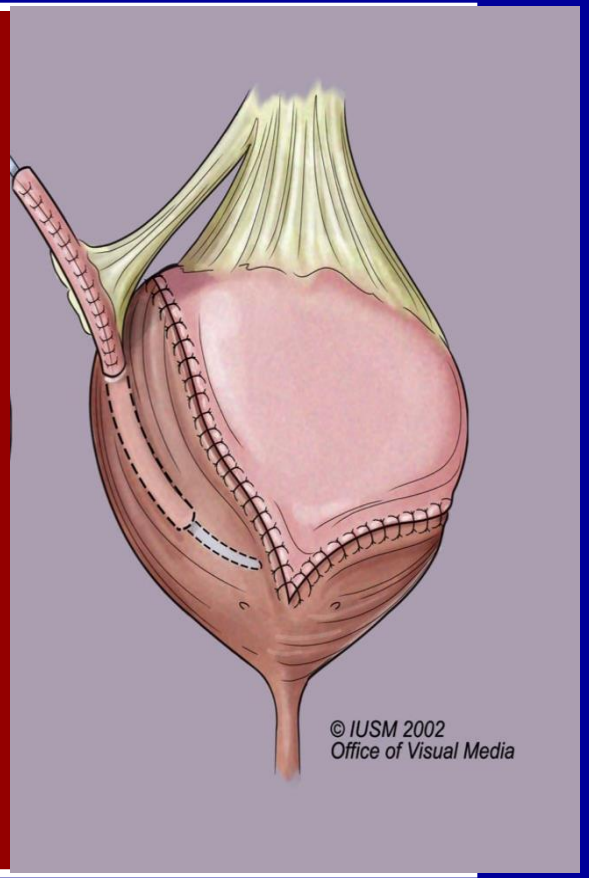
Women are more likely than men to experience UIS, regardless of baseline incontinence

Continence: For QOL, It Matters…

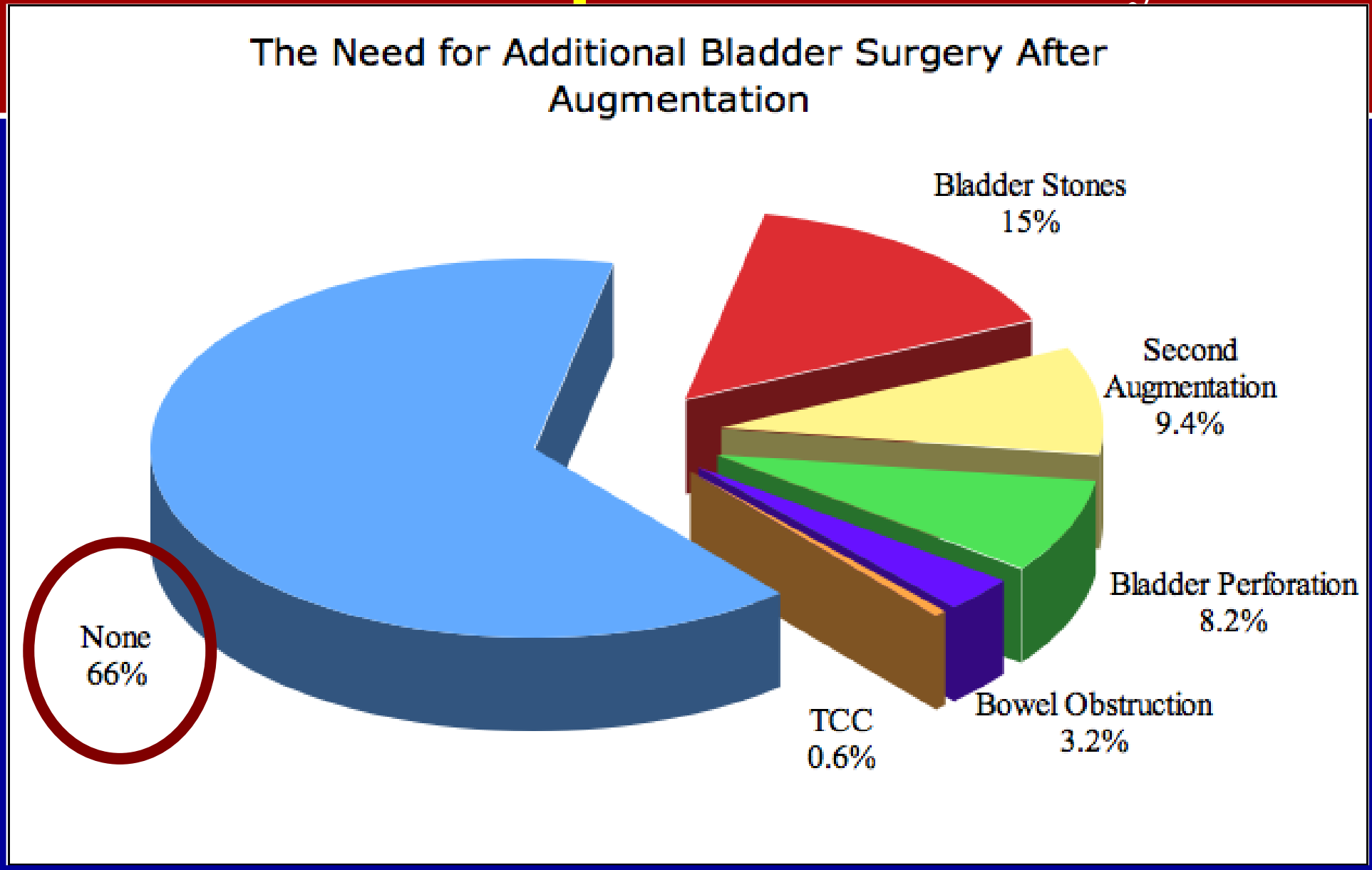


Quality of Life Conclusions: Reconstruction for Continence Matters (Still need Pre/Post Op Longitudinal Study)

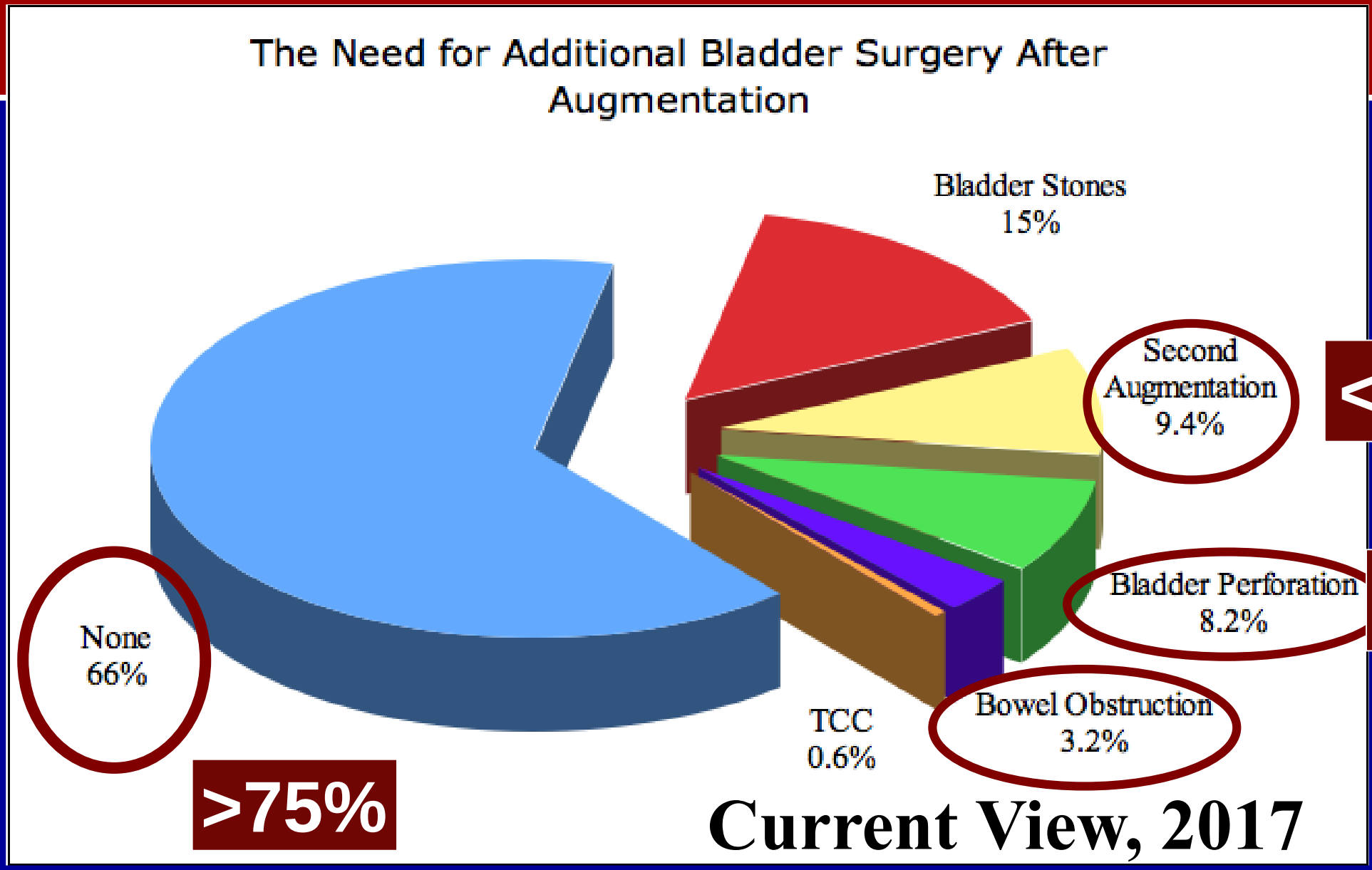
- Intermittent catheterization
- Bladder outlet resistance
- Bladder augmentation
- Mitrofanoff procedure
- MACE procedure



Bladder Complications: Indy 500



Update on Bladder Complications: Indy 500



Additional surgeries after bladder augmentation in patients with spina bifida in the 21st century

Konrad Szymanski, Rosalia Misseri, Benjamin Whittam, Nathan Hollowell, Rachel Hardacker, Carly Swenson, Martin Kaefer, Richard Rink, Mark Cain

SCHOOL OF MEDICINE
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Riley Hospital for Children
Indiana University Health

Methods

Retrospective cohort study of consecutive SB patients after bladder augmentation: 1978-2018

***born after Jan. 1, 1972**

Two cohorts: 1. Entire group
2. Modern (ileocystoplasty since 2000)

Risk factor: Detubularization & reconfiguration (DR)

Methods

Outcomes: Incontinent urinary diversion
Bladder stones
Bladder perforation
Reaugmentation
Laparotomy for bowel obstruction
Bladder tumors (benign and malignant)

Statistics: Survival analysis, Cox regression

Patient Selection

Augmentation database: 784

Born before 1972: - 116

Not SB: - 255

Included in the study: 413

Population characteristics (n=413)

Median age:		**8.5 years**
MMC:		**95.4%**
Segment:	**Ileum:**	**80.9%**
	Sigmoid:	**11.1%**
Urinary channel:		**74.1%**
MACE:		**69.1%**

Results

At median follow-up of 11.2 years
44.1% had 370 additional surgeries

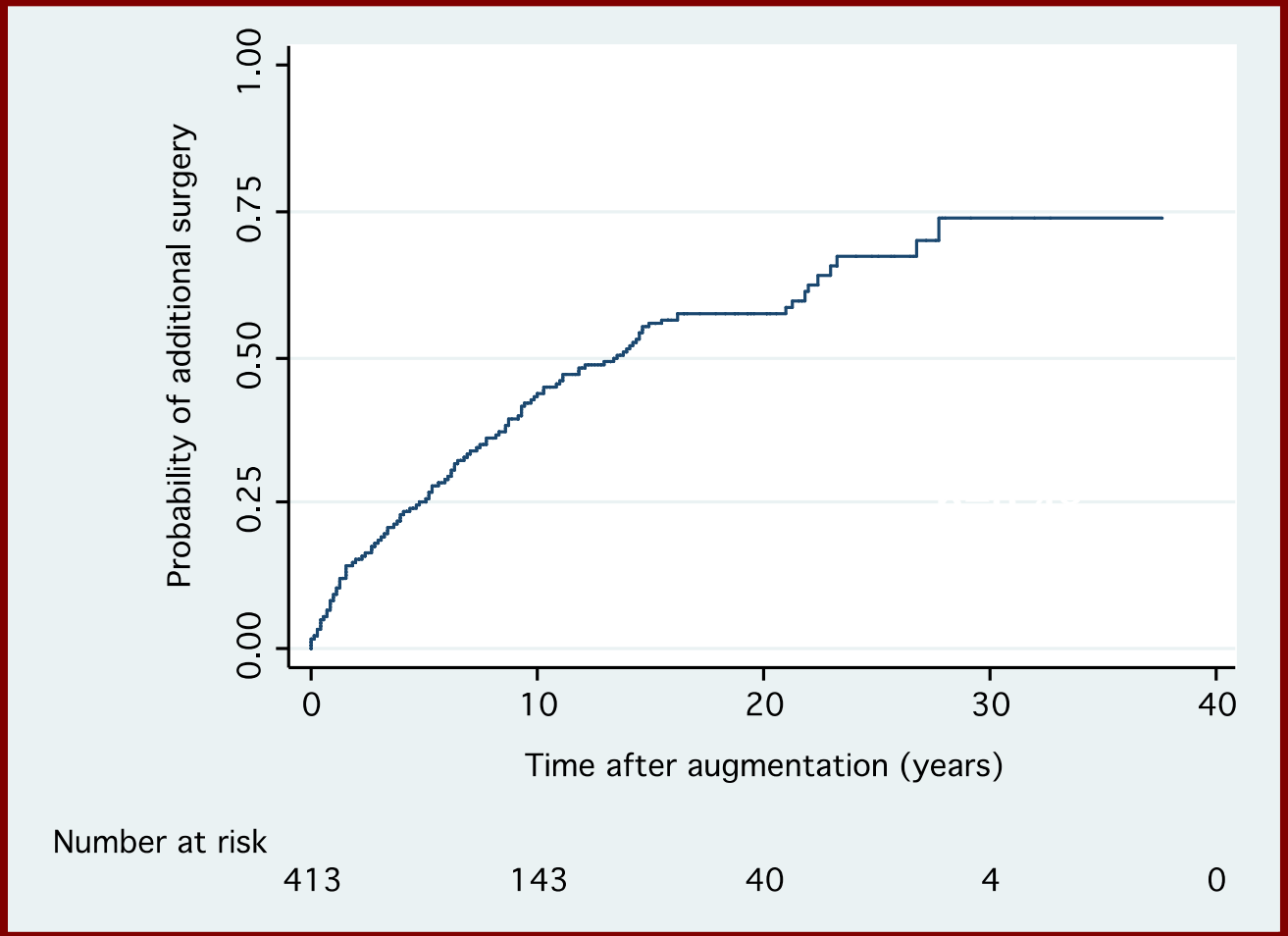
***34.6% were recurrences of the same secondary surgery**

#1. Bladder stones (57.6% of all surgeries)
#2. Bladder perforation

Risk of any subsequent surgery

5 years	25.5%
10 years	43.9%
20 years	57.4%

≥ 1 surgery at 10yr	
2 or more	17.4%
3 or more	9.9%
4 or more	3.8%



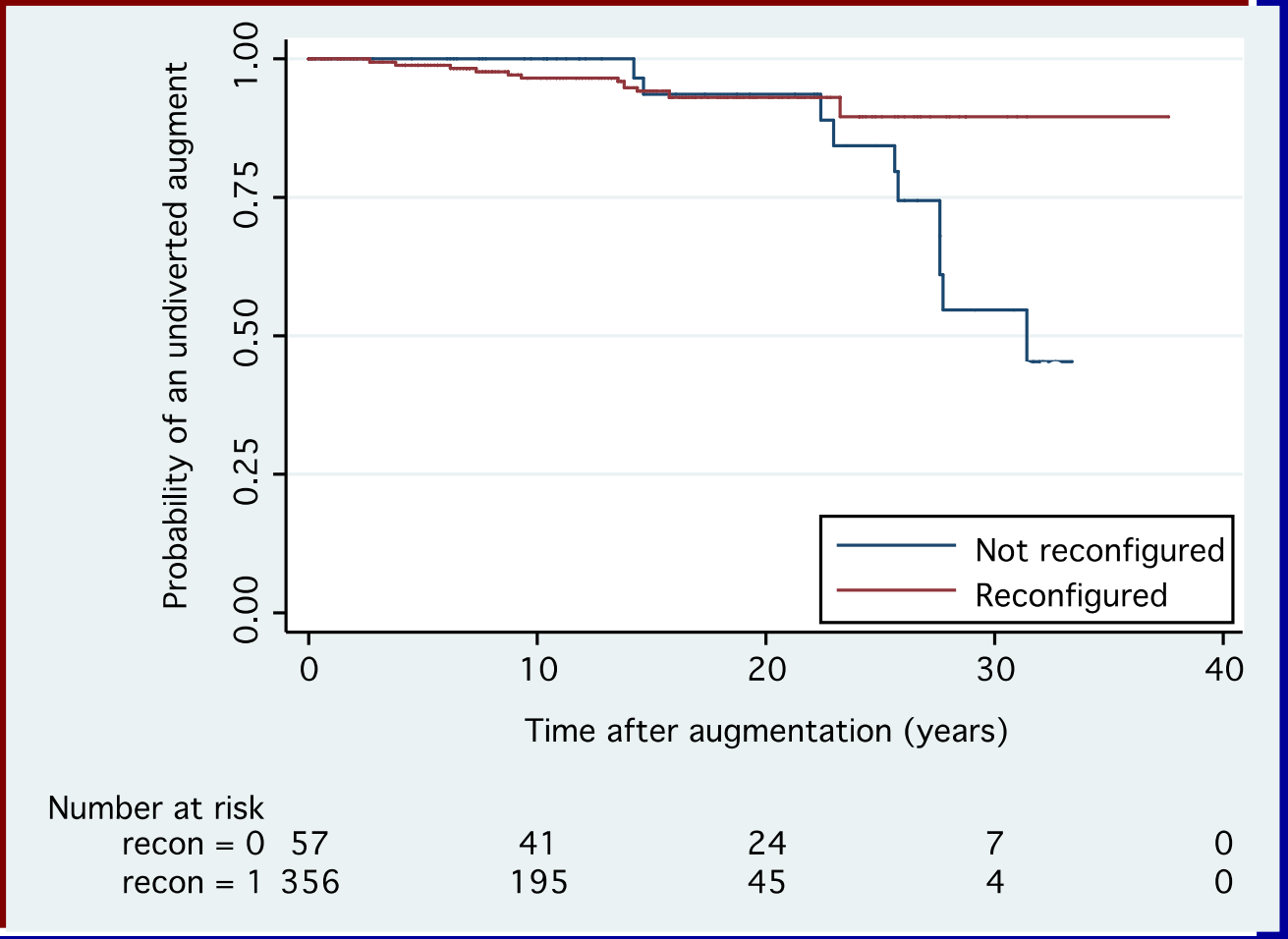
Urinary diversion

10 year risk 2.7%

65.2% ileal conduit

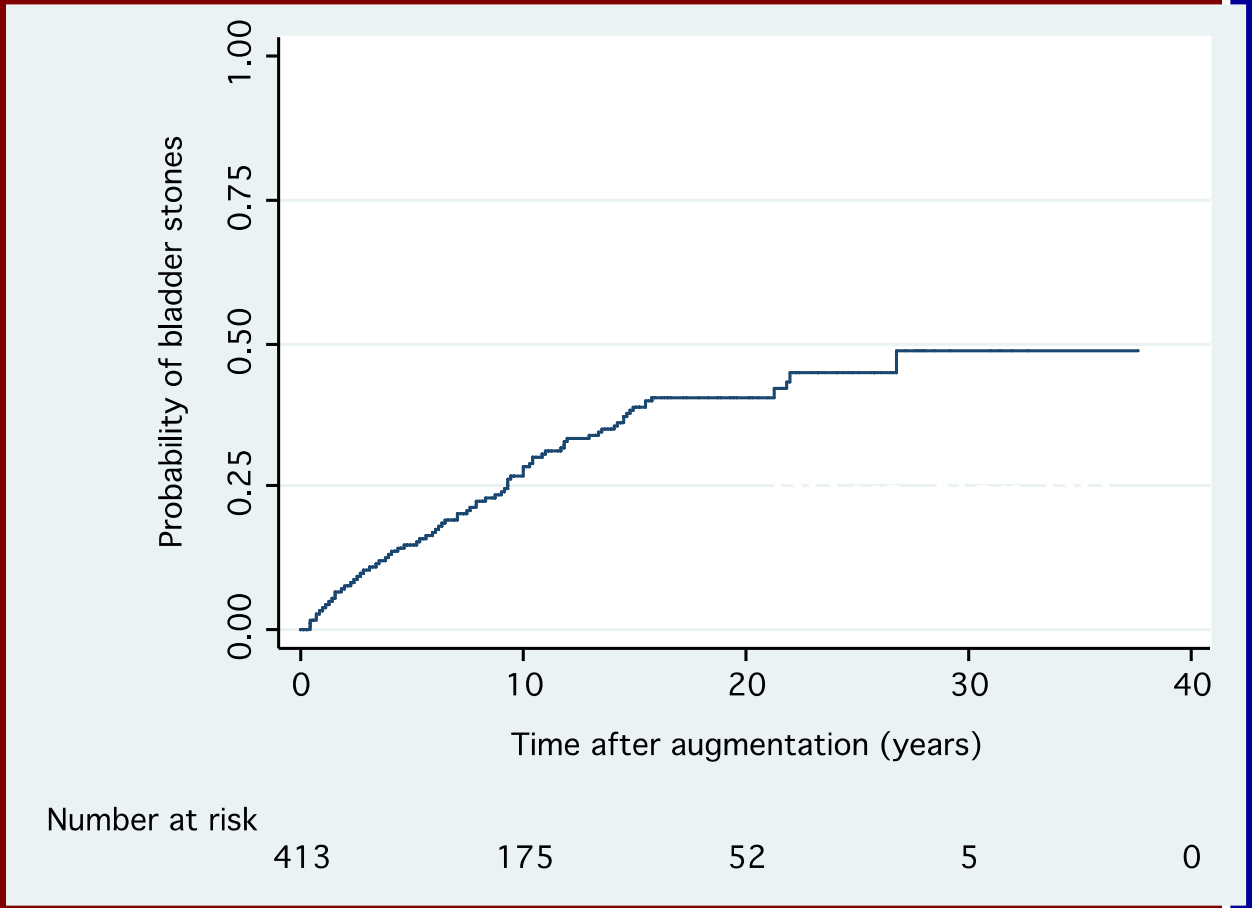
Indications

Perforations	34.8%
Incontinence	34.8%
UT changes	30.4%
Difficult cath.	8.7%



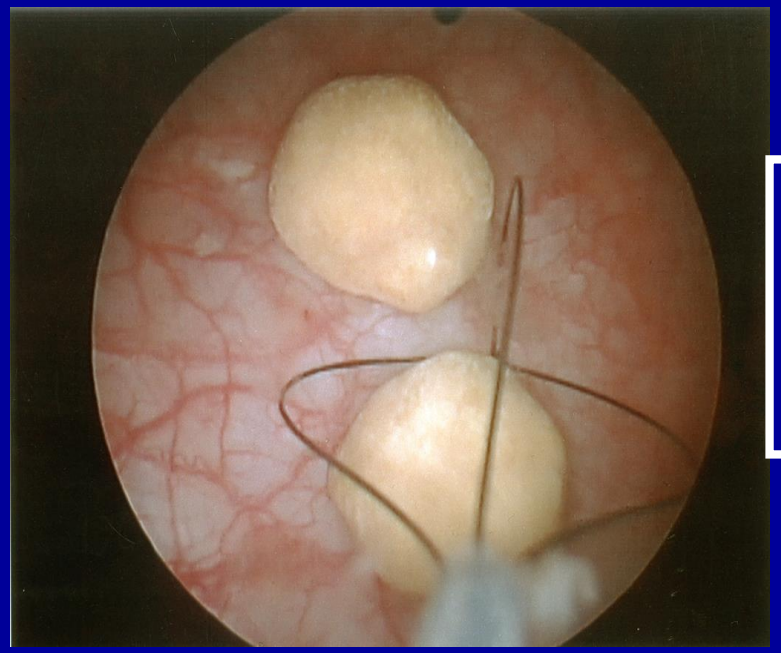
Bladder stones

10 year risk	28.2%
Endoscopic	32.5%
Percutaneous	29.9%
Open	37.6%
10 year recurrence	52.4%

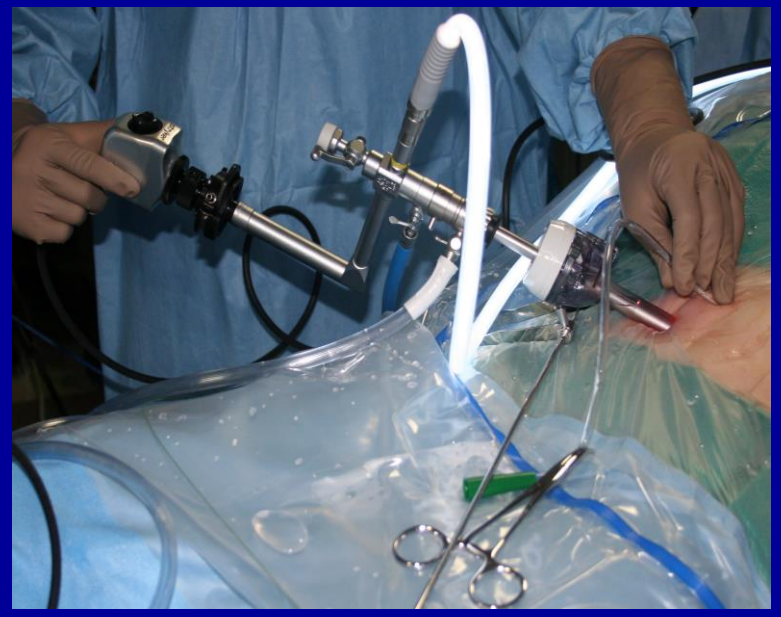


BLADDER STONES

IU Experience: Removal Technique



OPEN cytolithotomy 33.6%

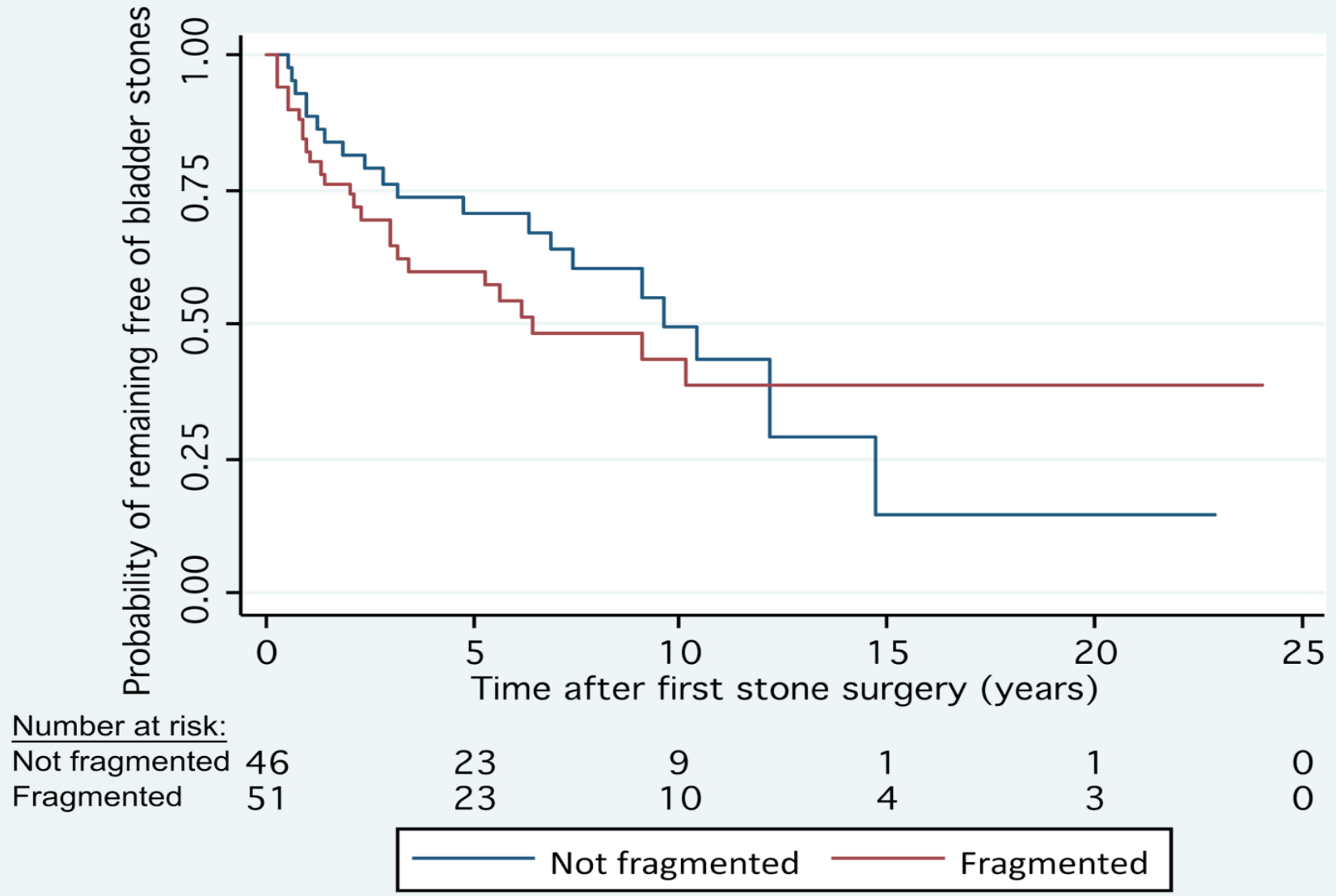


Endoscopic 32.7%

Percutaneous 33.6%

47.7 % had stone fragmented

Stone fragmentation



Bladder perforation

10 year risk

Not DR 23.7%

DR 9.6%

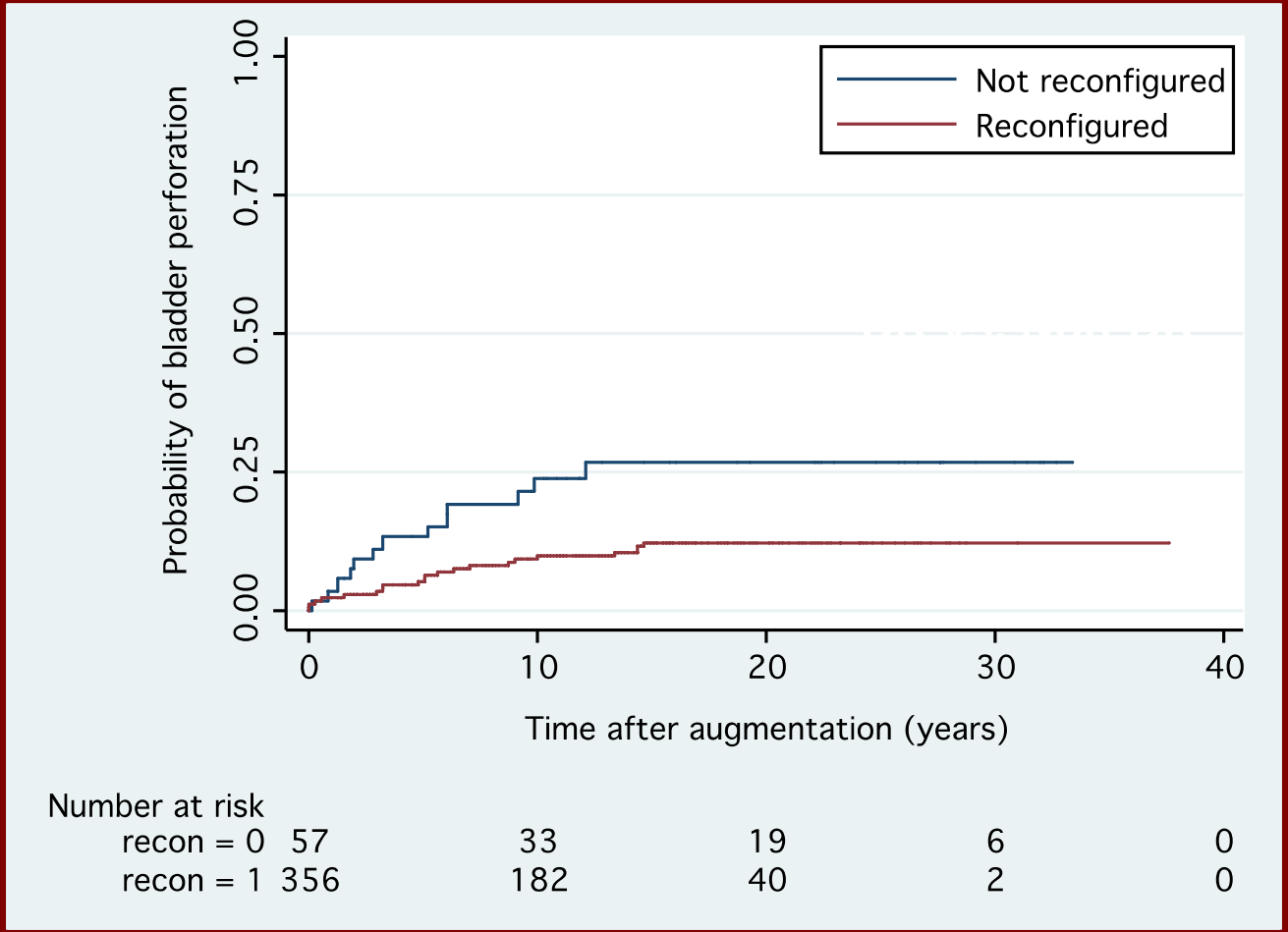
p=0.01

10 year recurrence

Not DR 73.8%

DR 32.1%

p=0.05



Reaugmentation

10 year risk

Not DR 15.2%

DR 5.3%

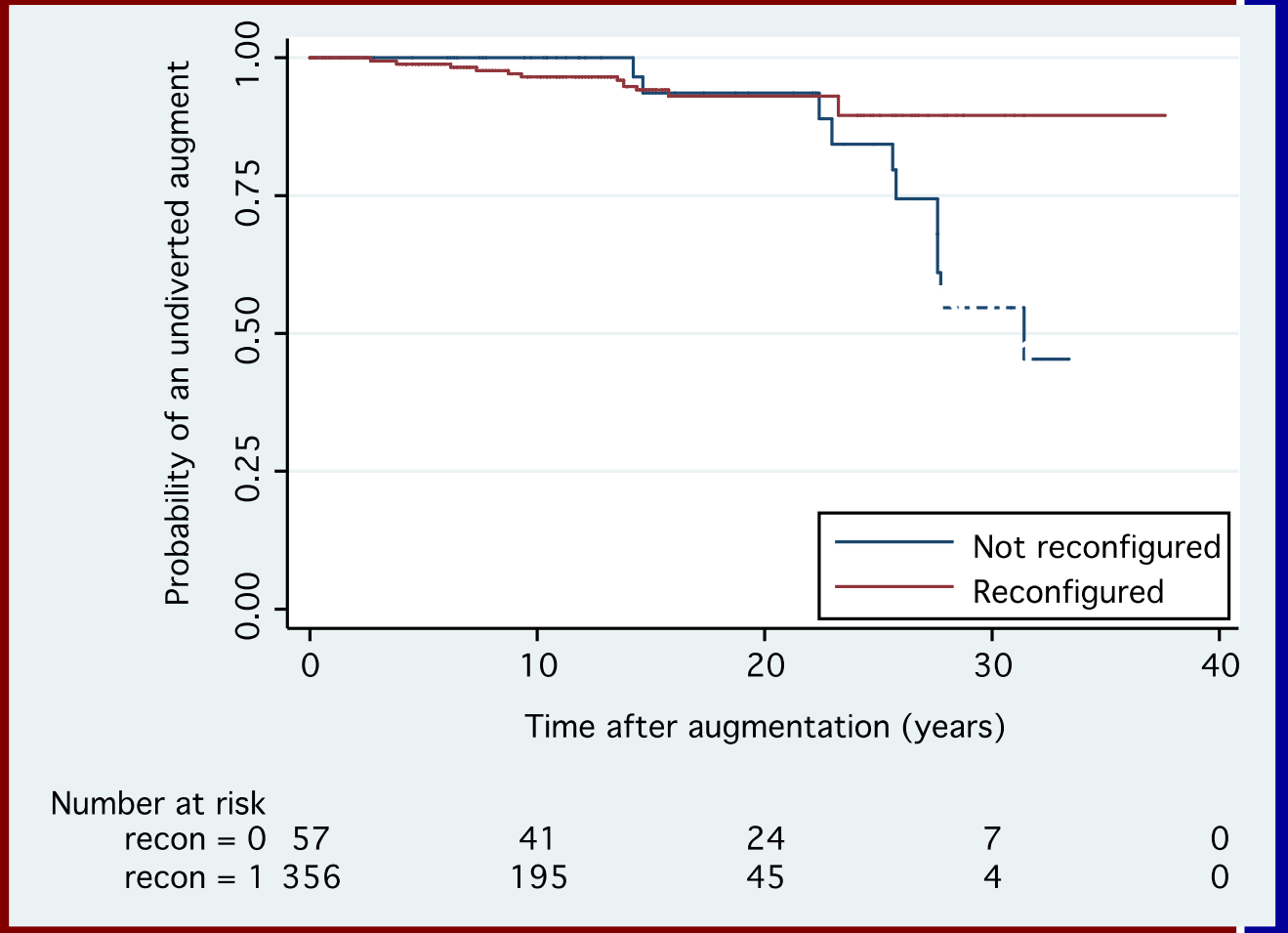
p=0.001

Indications

Contractions 61.5%

Small capacity 53.8%

Incontinence 50.0%

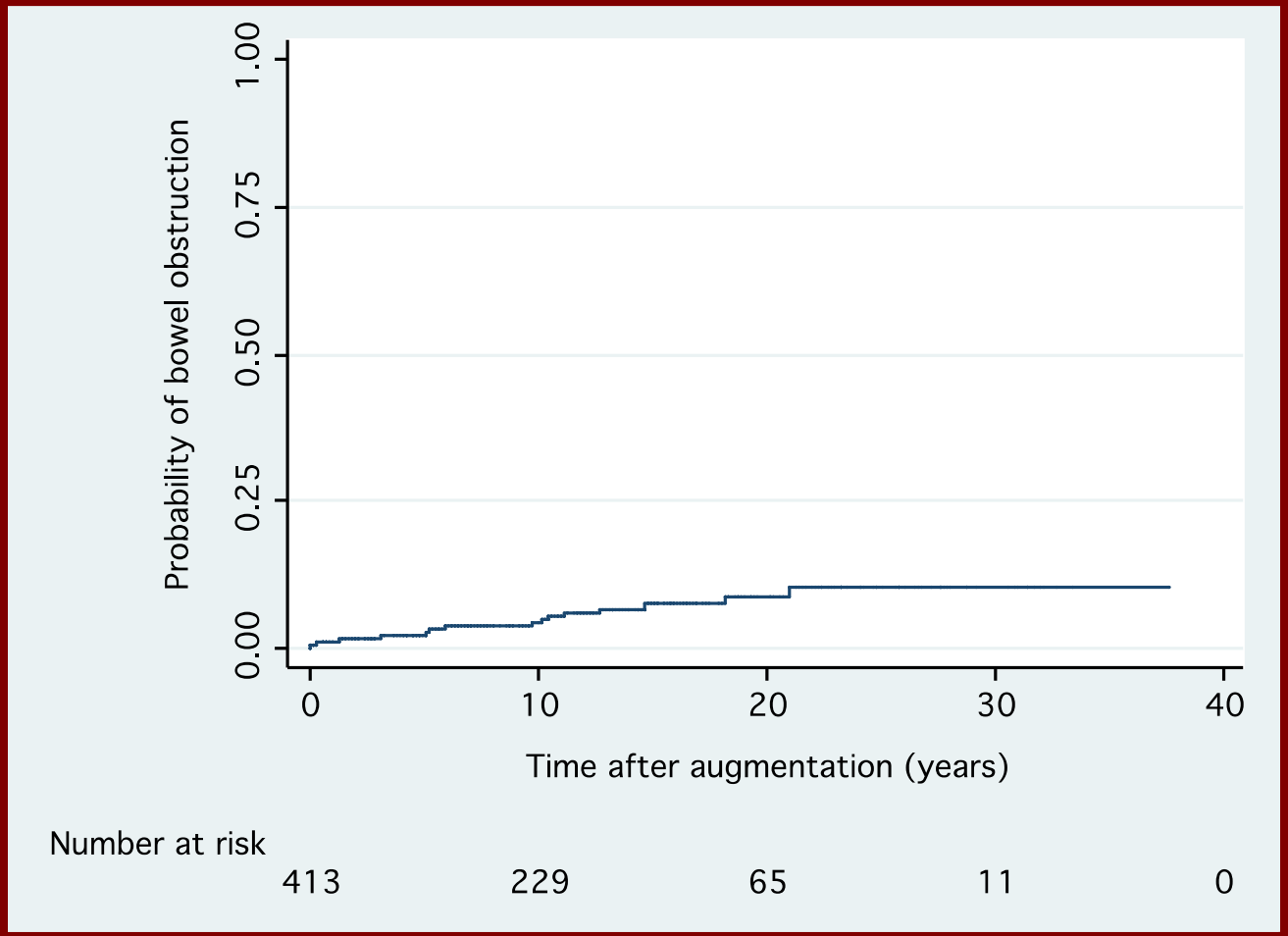


Laparotomy for bowel obstruction

10 year risk 4.5%

91.3% small bowel

10 year recurrence 10.0%

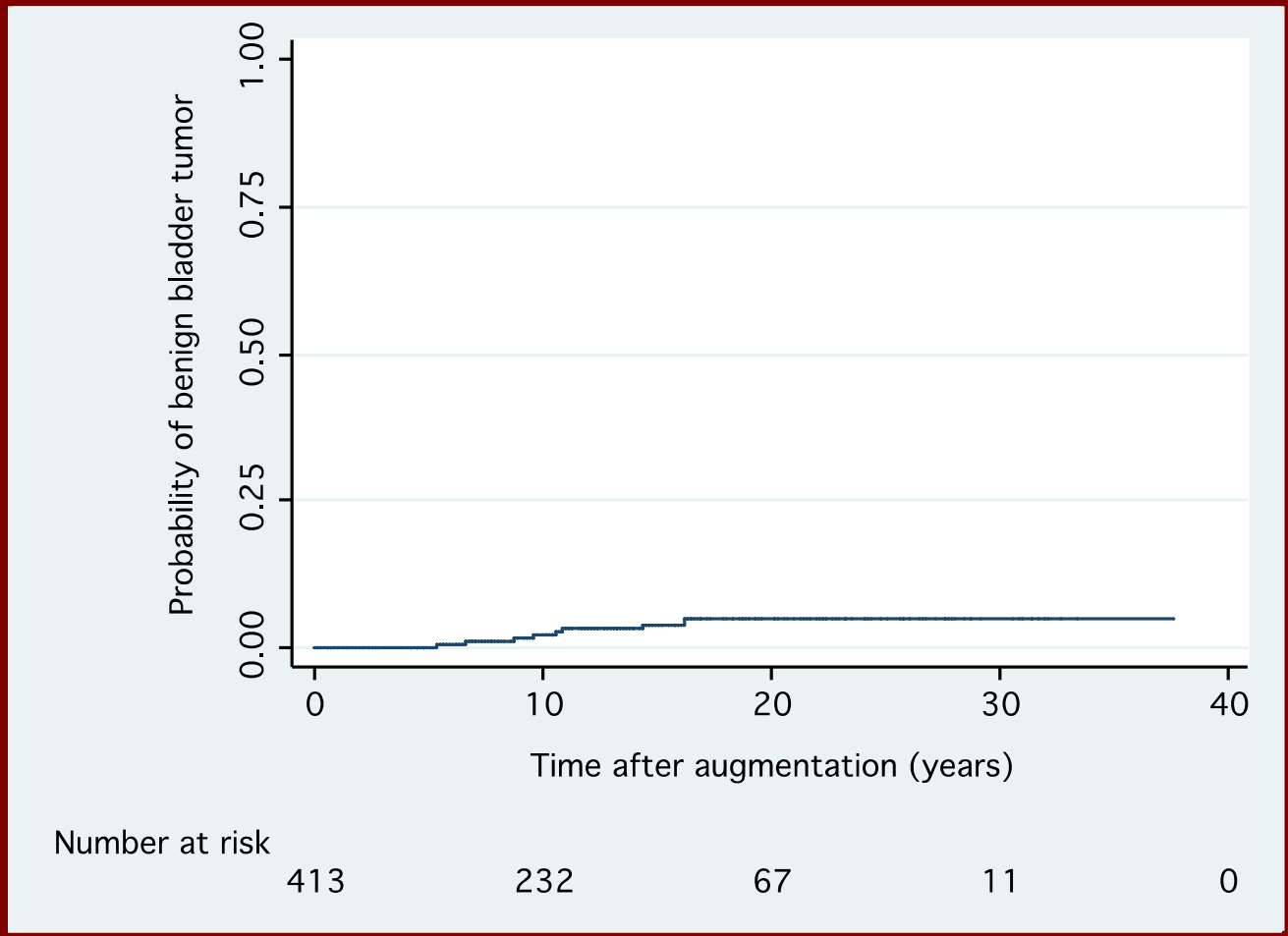


Excision of benign bladder tumor

10 year risk 2.2%

100.0% nephrogenic adenoma

10 year regrowth 48.2%



Bladder cancer

20 year risk 0.0%

No patient was diagnosed with bladder malignancy

****we have 4 CA in database**

Modern cohort (n=222)
Ileocystoplasty since 2000

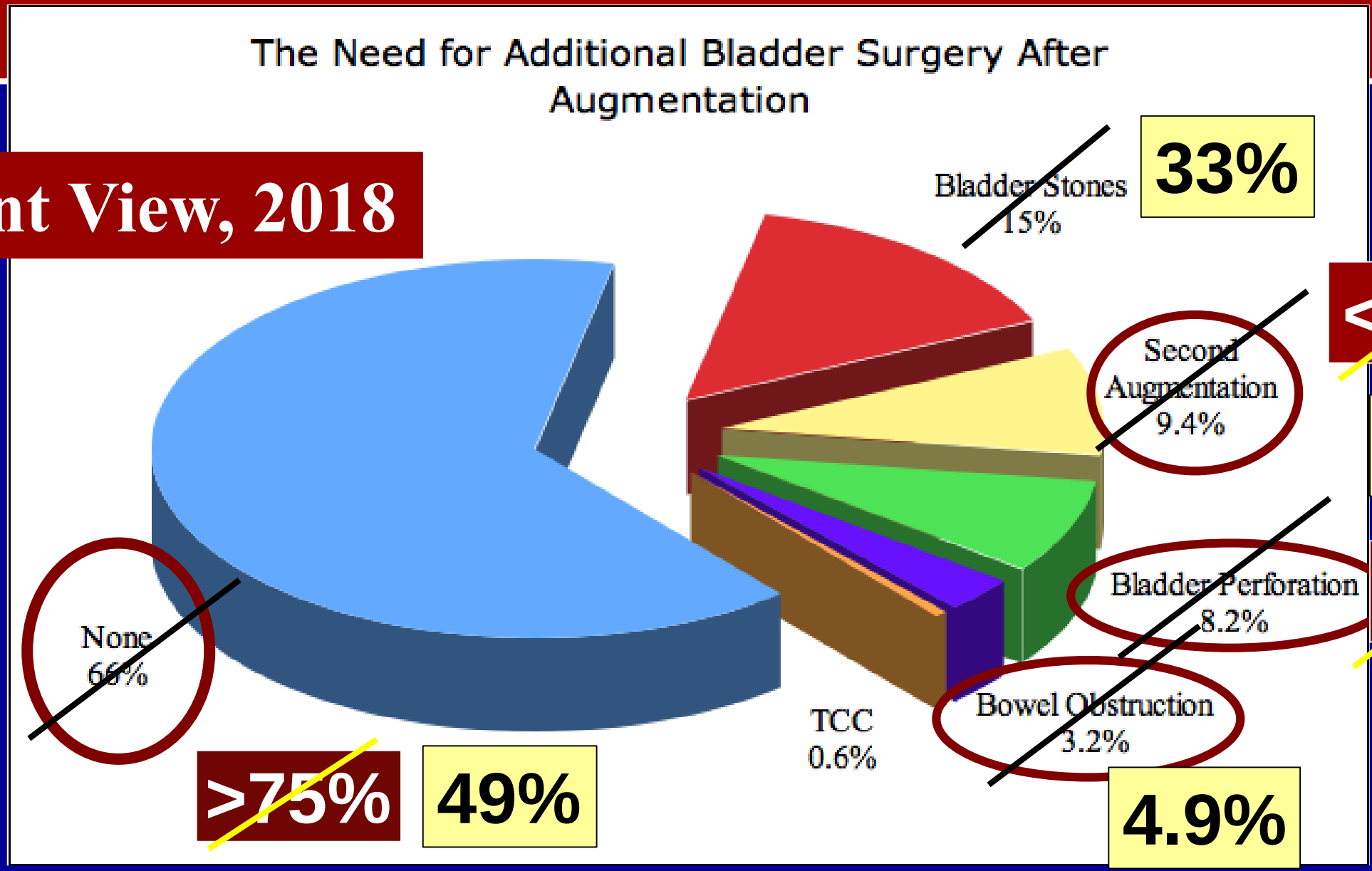
Surgery	10 year risk	10 year recurrence
Urinary diversion	4.0%	
Bladder stone	32.9%	44.5%
Bladder perforation	8.8%	42.2%
Reaugmentation	4.3%	
Bowel obstruction	4.9%	10.0%
Benign tumor excision	4.7%	40.0%
Bladder cancer	0.0%	

Modern cohort (n=222)
Ileocystoplasty since 2000

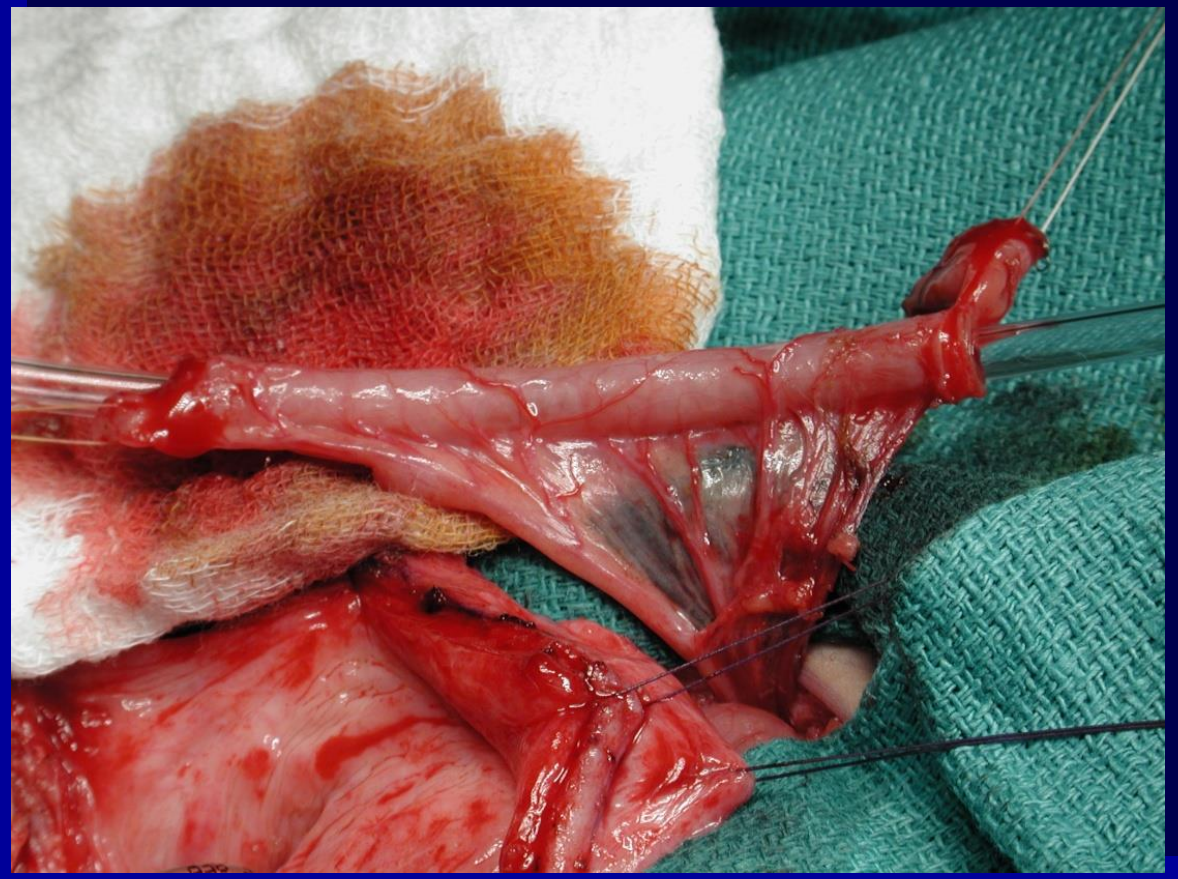
Surgery	10 year risk	10 year recurrence
Urinary diversion	4.0%	
Bladder stone	32.9%	44.5%
Bladder perforation	8.8%	42.2%
Reaugmentation	4.3%	
Bowel obstruction	4.9%	10.0%
Benign tumor excision	4.7%	40.0%
Bladder cancer	0.0%	

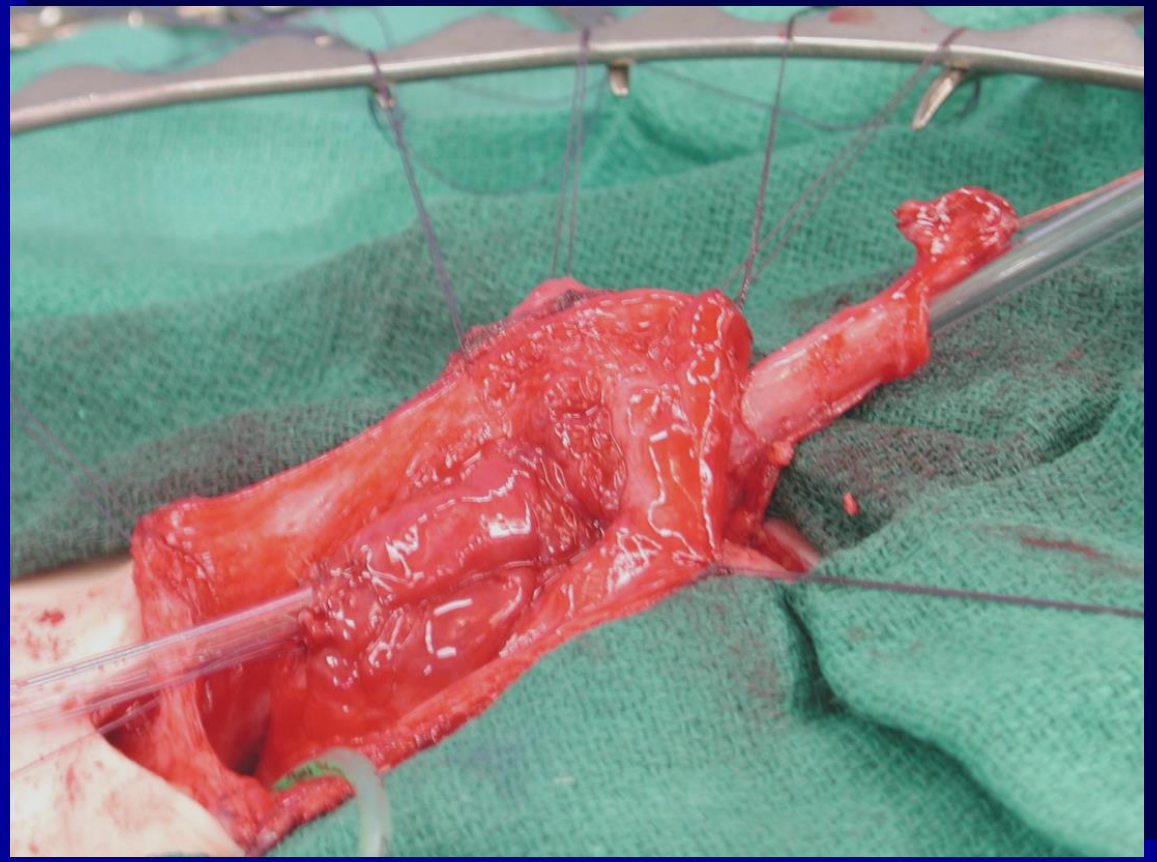
Update on Bladder Complications:

Current View, 2018



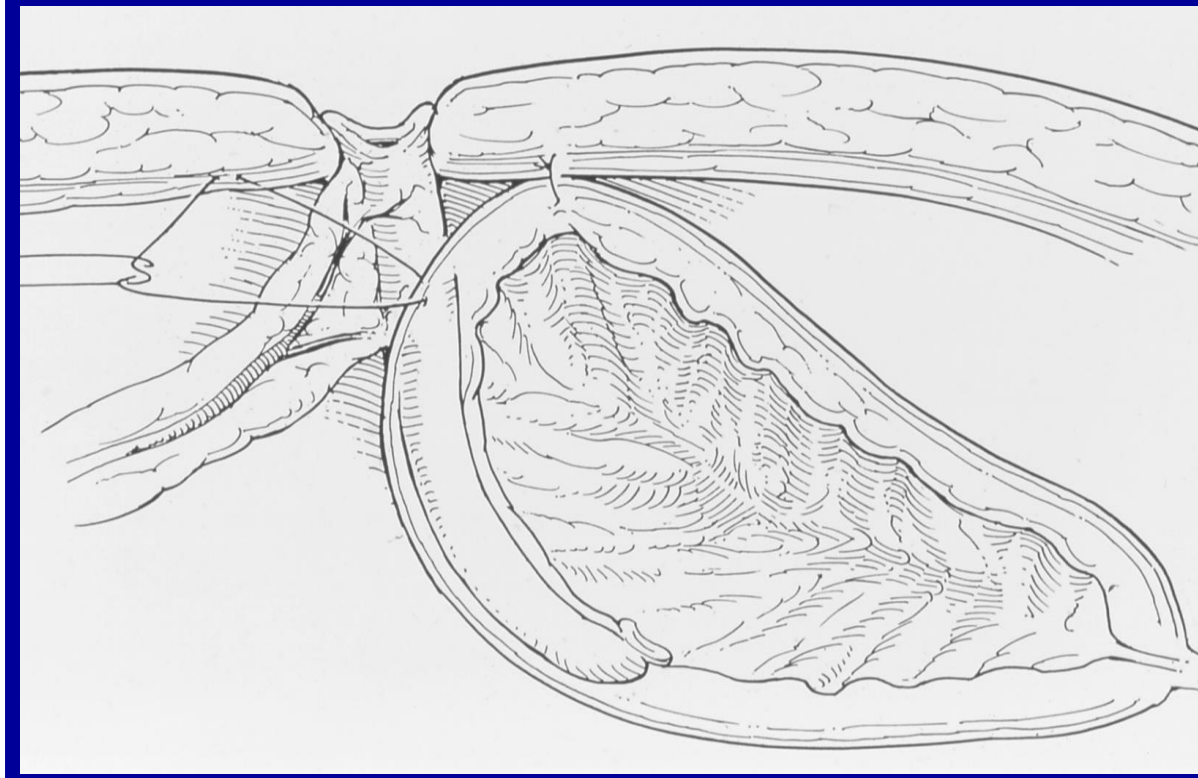
APPENDICOVESICOSTOMY





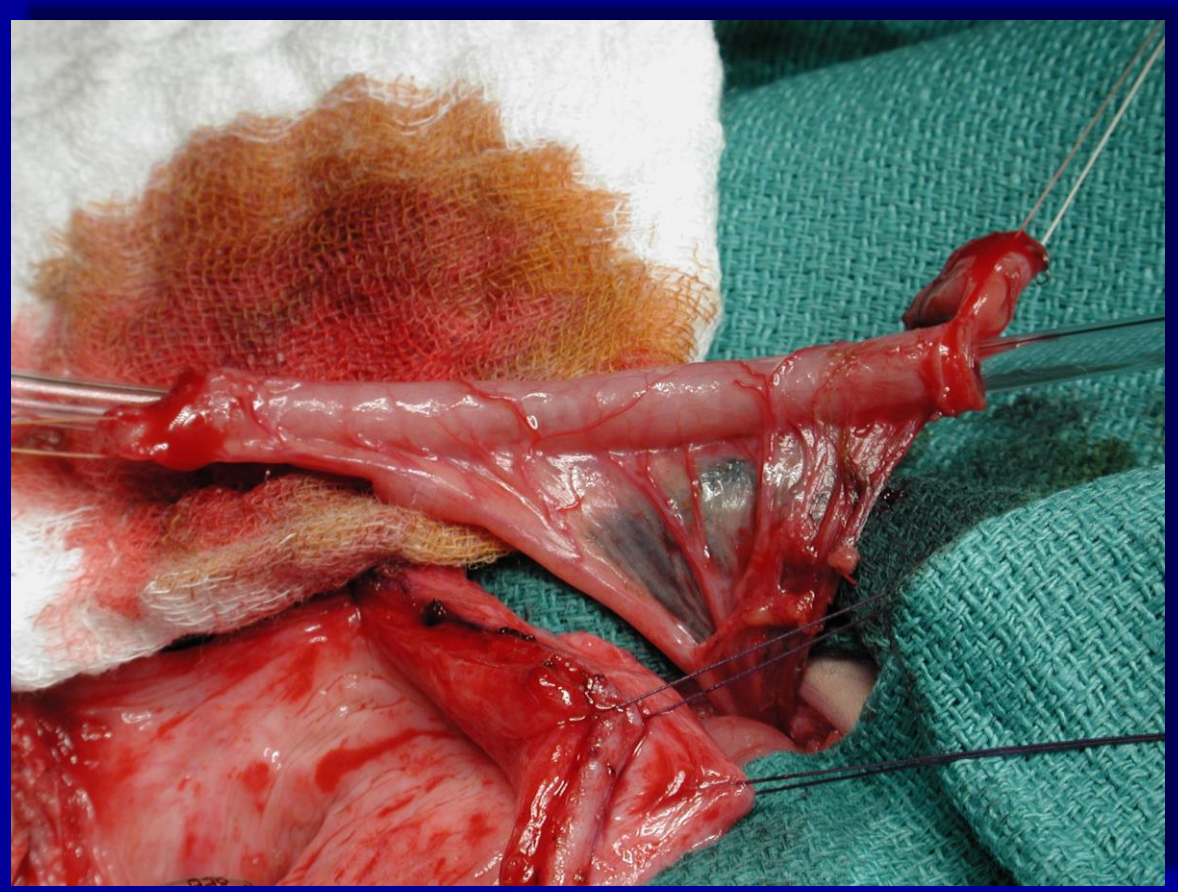
Catheterizable Channels:
Complications

- Stomal Stenosis
- Angulation of Channel
- Leakage
- Trauma
- Obliteration
- Abscess
- Polyp



Short, Straight
Supple, Secure

APPENDICOVESICOSTOMY
VS.
MONTI-YANG



Appendicovesicostomy vs. Monti

0022-5347/99/1625-1749/0
THE JOURNAL OF UROLOGY
Vol. 162, 1749–1752, November 1999
Printed in U.S.A.

APPENDICOVESICOSTOMY AND NEWER ALTERNATIVES FOR THE MITROFANOFF PROCEDURE: RESULTS IN THE LAST 100 PATIENTS AT RILEY CHILDREN'S HOSPITAL

MARK P. CAIN, ANTHONY J. CASALE, SHELLY J. KING AND RICHARD C. RINK

From the Department of Urology, James Whitcomb Riley Hospital for Children, Indiana University Medical Center, Indianapolis, Indiana

100 pts. : 57 Apv, 21 Monti, 21 CV

- 98% continence, 12% stomal stenosis

- 20 secondary procedures

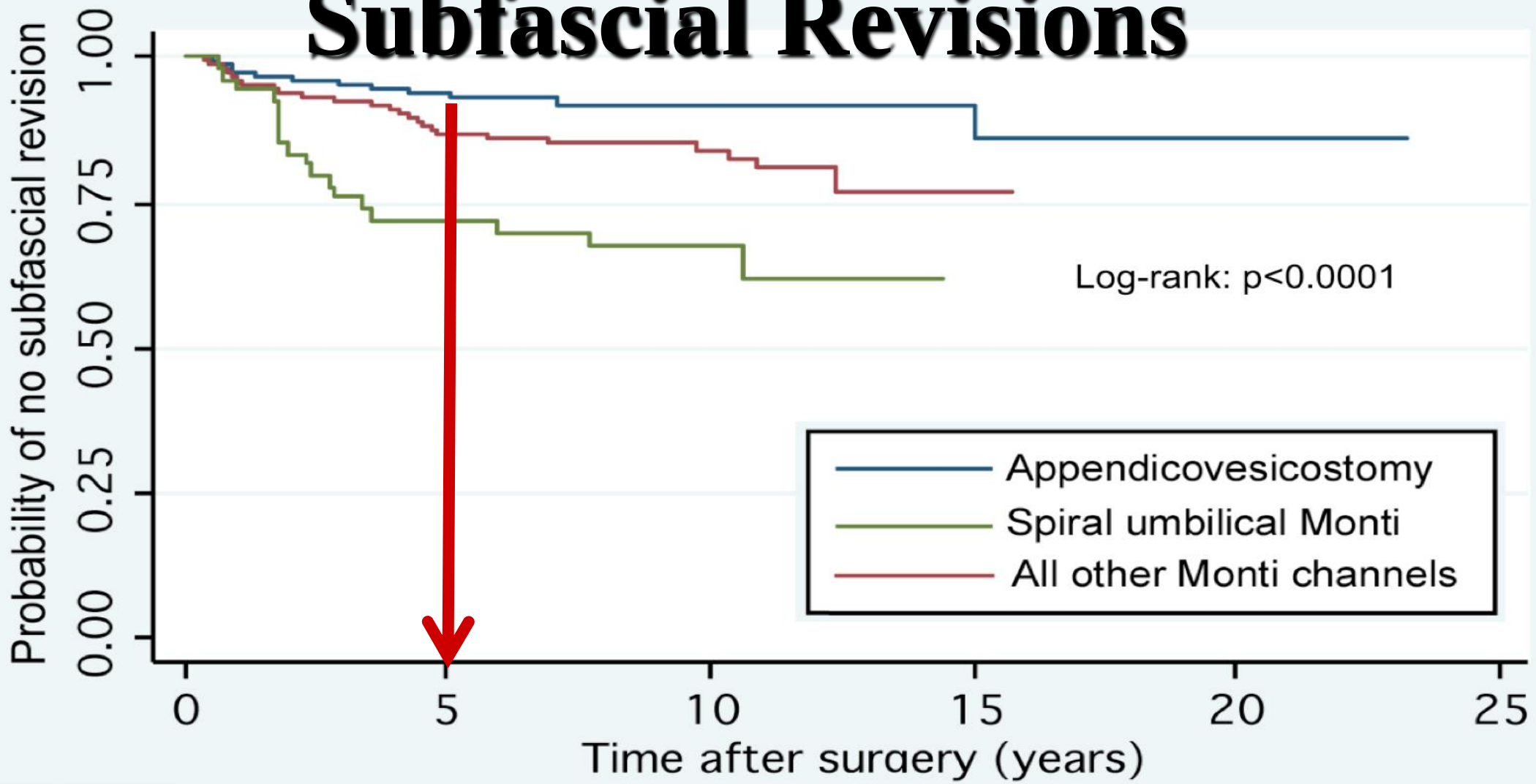
12/57 Apv (21%) F/U 31 mos

2/21 Monti (10%) F/U 9 mos

Subfascial Revisions: Szymanski, J Ped Urol, 2015

Channel type and stomal location	Number	Number of first subfascial revisions	P-value	Median follow-up (years)
APV	215			
Non-umbilical	118	6 (5.1%)	reference	5.4
Umbilical	97	8 (8.3%)	0.41	6.2
Monti				
Traditional	146			
Non-umbilical	96	14 (14.6%)	0.03	8.4
Umbilical	50	6 (12.0%)	0.19	9.9
Spiral	150			
Non-umbilical	94	11 (11.7%)	0.13	4.8
Umbilical	56	18 (32.1%)	<0.001	9.0

Subfascial Revisions



Number at risk:						
Appendicovesicotomy	215	116	41	17	7	0
Spiral umbilical Monti	56	36	16	0	0	0
All other Monti channels	240	134	63	5	0	0

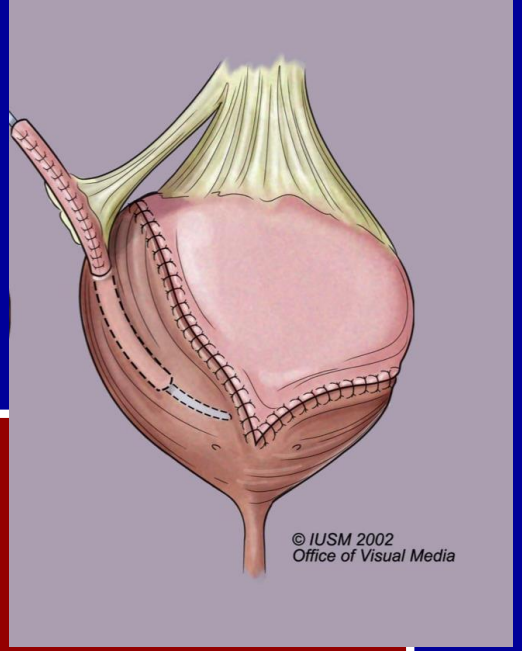
What do we tell the families about risk?

Subfascial Revision = Major Surgery

	5years	10years
Appendicovesicostomy	6.4%	8.6%
Monti Channels	12.9%	15.5%
Spiral to Umbilicus	27.9%	32.3%

Majority of revisions occur in first 5 years

BLADDER AUGMENTATION
Common Sense Follow Up Evaluation



Stones:
- Yearly RBUS and KUB

Recurring UTI:
- RBUS/KUB, review CIC technique, ?cystoscopy

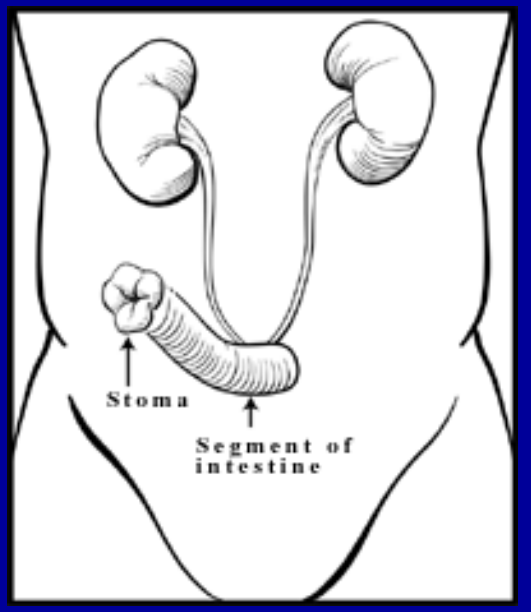
Metabolic:
- Yearly CBC, BMP, Cystatin C, B12 yearly after 5 years
- ? DEXA scan

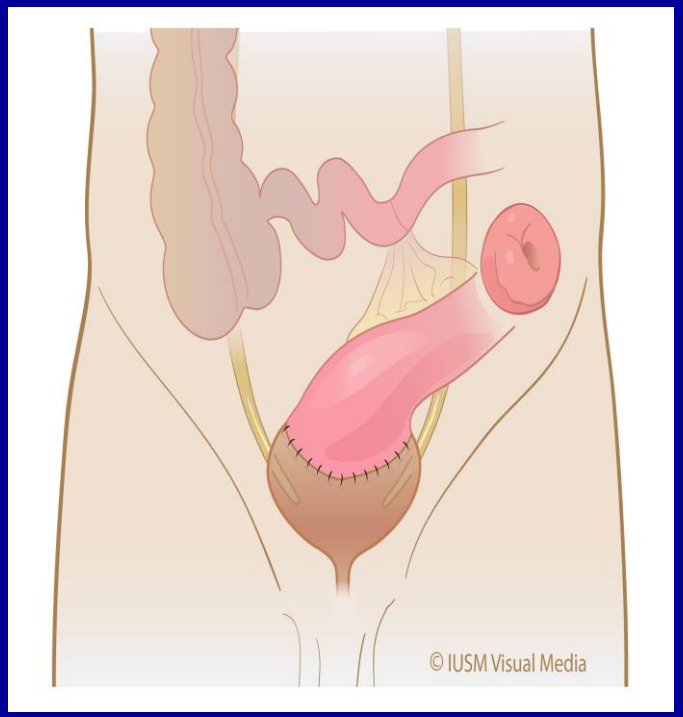
Cancer Surveillance:
- High Risk: Yearly cystoscopy, cytology starting 5 yrs. (?)
- Low Risk: Evaluate aggressively for hematuria, recurrent UTI, pain, abnormal X-ray

Ileal Chimney

- Drains continuously
- Protects kidneys
- Appliance works well

For patients that loose Bladder priveleges:



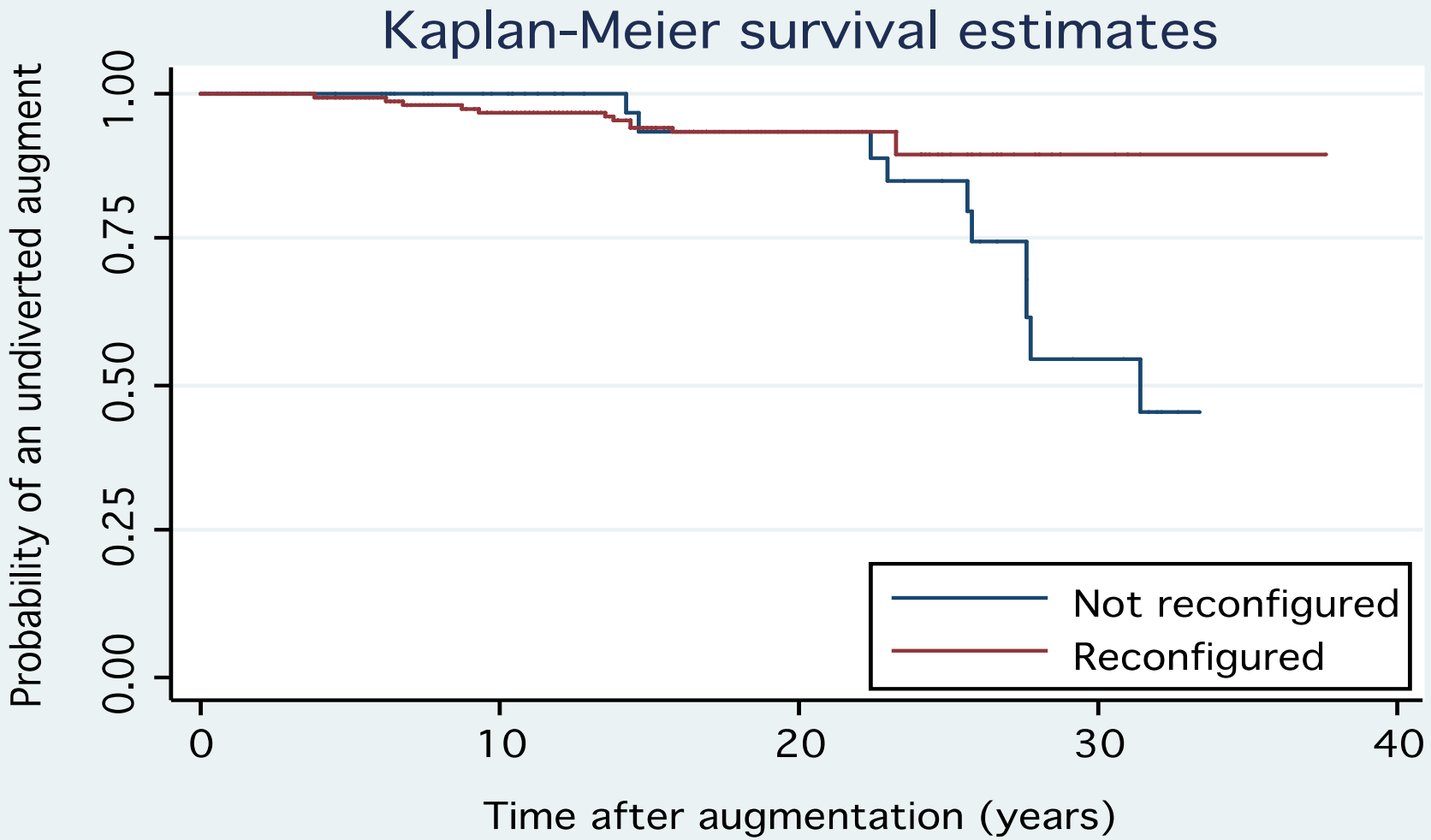


Spina Bifida Urologic Care
What Have We Learned?

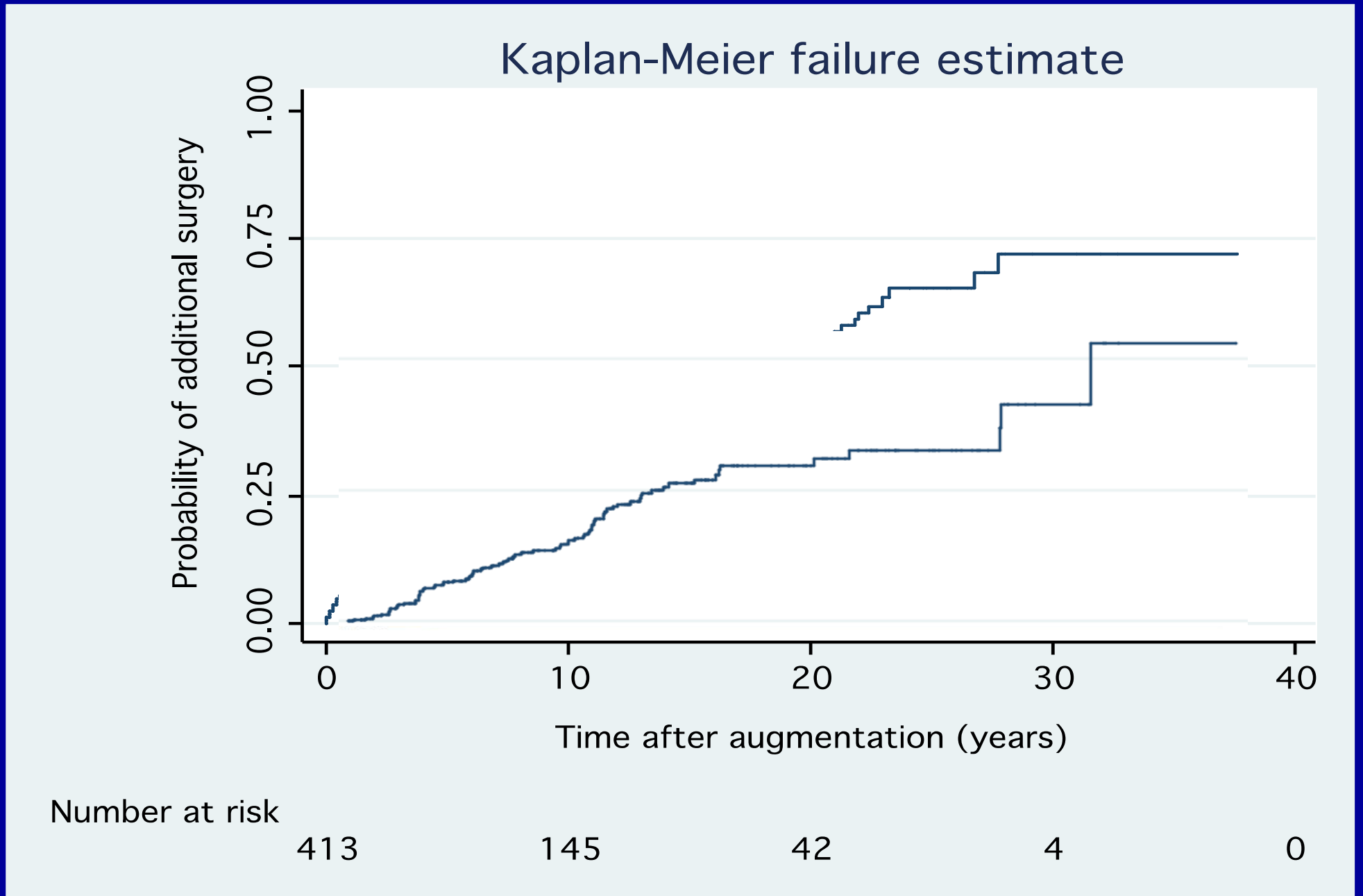
- Our patients want to be continent, but not until later
- Our patients want to be continent of stool, earlier
- Our patients survive into adulthood, and need care
- Our patients are sexual adults, want help early
- Our patients are fertile, and need our help then too
- Our patients are still teaching us medical lessons…..


patagonia

Probablity of Reaugmentation –



Need for Any Surgery…?



Francis Collins, MD, PhD
Director, Human Genome Project

- Prepare for dramatic change
- Your path will not be smooth.
  - Build a strong foundation up front (train for opportunity)
  - Be a responsible skeptic
- Clarify your definition of success.
  - Allow it to change over time
- Define **resume** virtues vs. ***eulogy*** virtues

If We Are Going To Augment the Bladder What Is the Cost To the Patient?

- Metabolic
  - Acidosis; B12 deficiency; Bone Density
- Mucus
- UTI
- Bladder Stones
- Bladder Perforation
- Malignancy risk

BLADDER AUGMENTATION

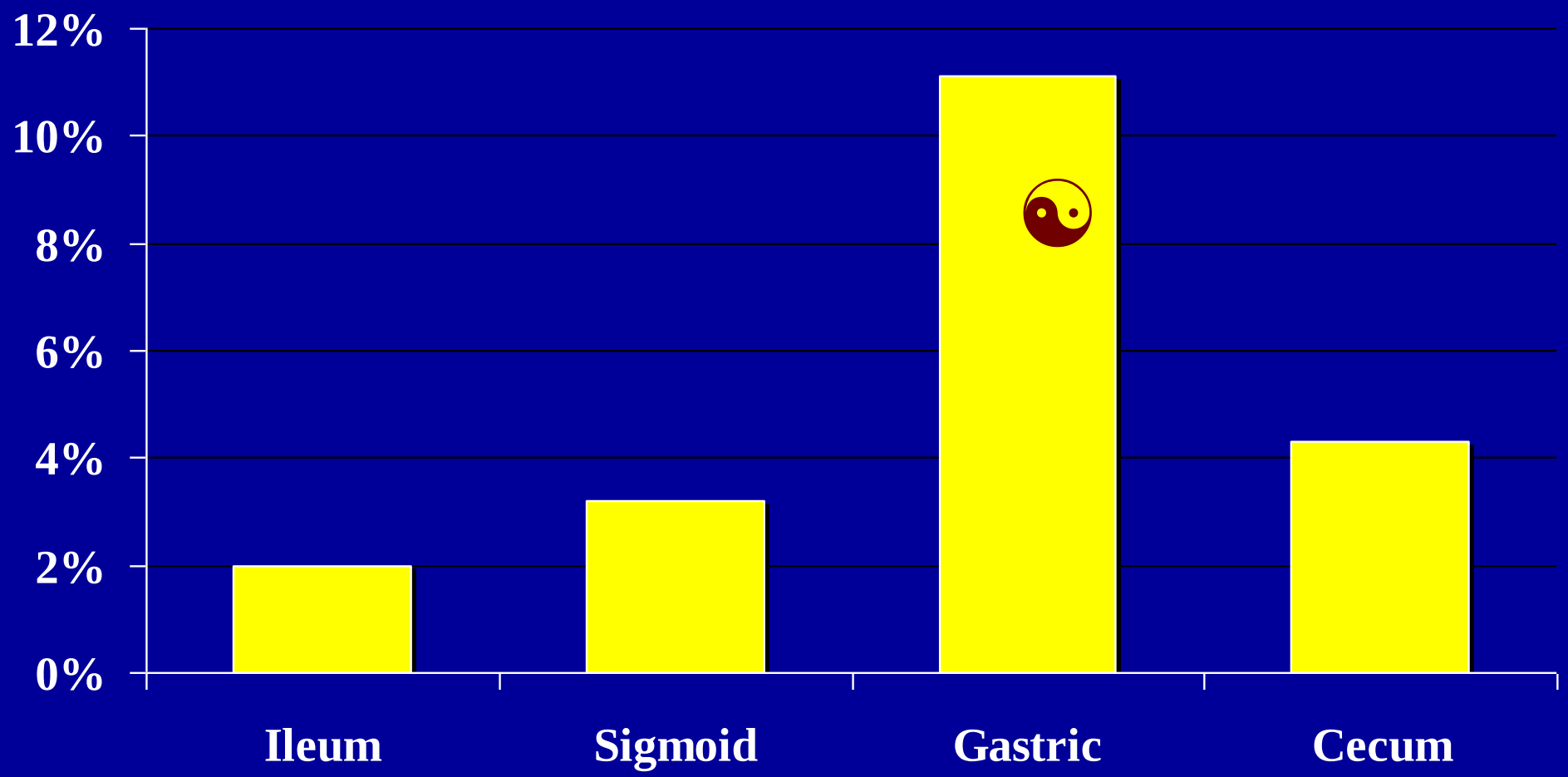
Indy 500 (patients)

Segment	# pts	%
Ileum	297	60
Sigmoid	85	17
Stomach	38	8
Cecal	46	9
Ileum + sig.	8	1.6
Ureter	8	1.6
Ureter +ileum	3	.6
Stomach + bowel	7	1.4

BLADDER AUGMENTATION
Indy 500

- **Bowel Obstruction**
- **3.2% (16 patients)**
- **Mean time from surgery = 51 months**
  - **7 < 2 months**
  - **9 > 12 months**

Risk per Segment
Bowel Obstruction



BLADDER AUGMENTATION

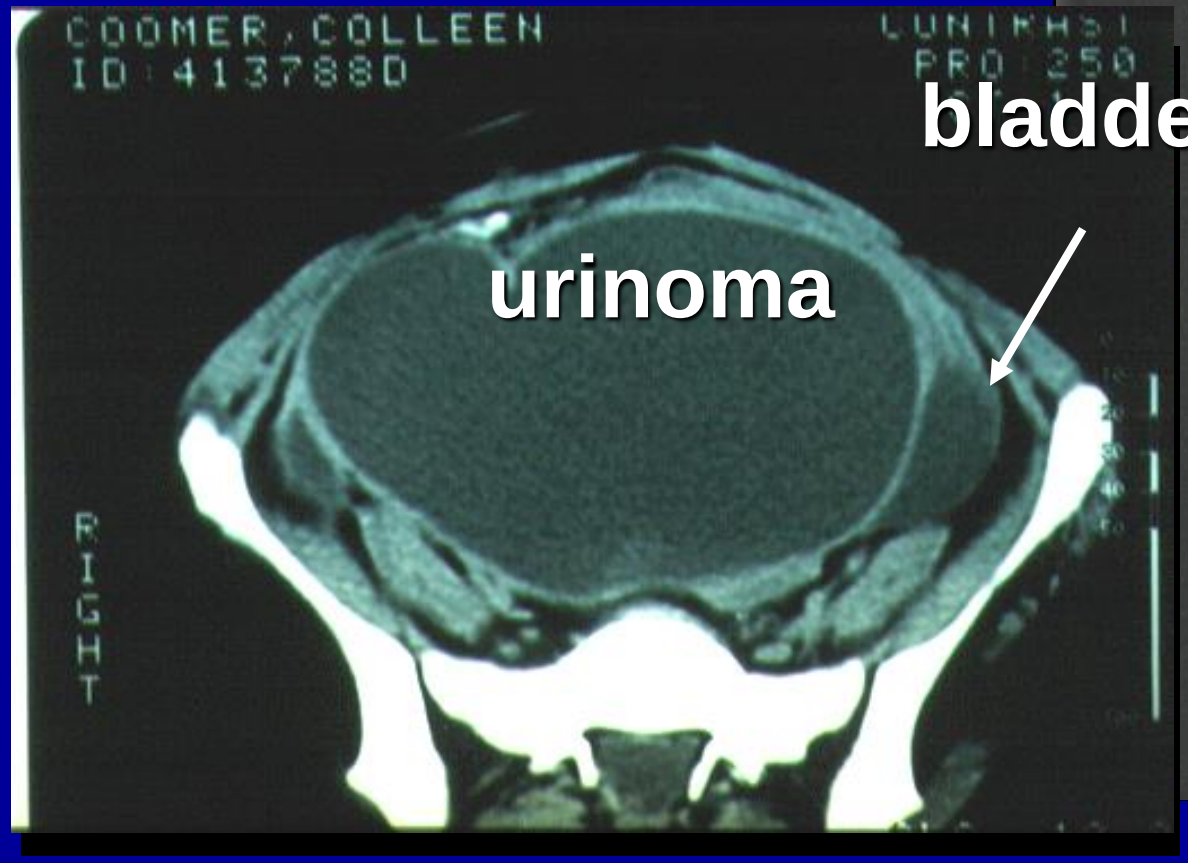
Major Complications

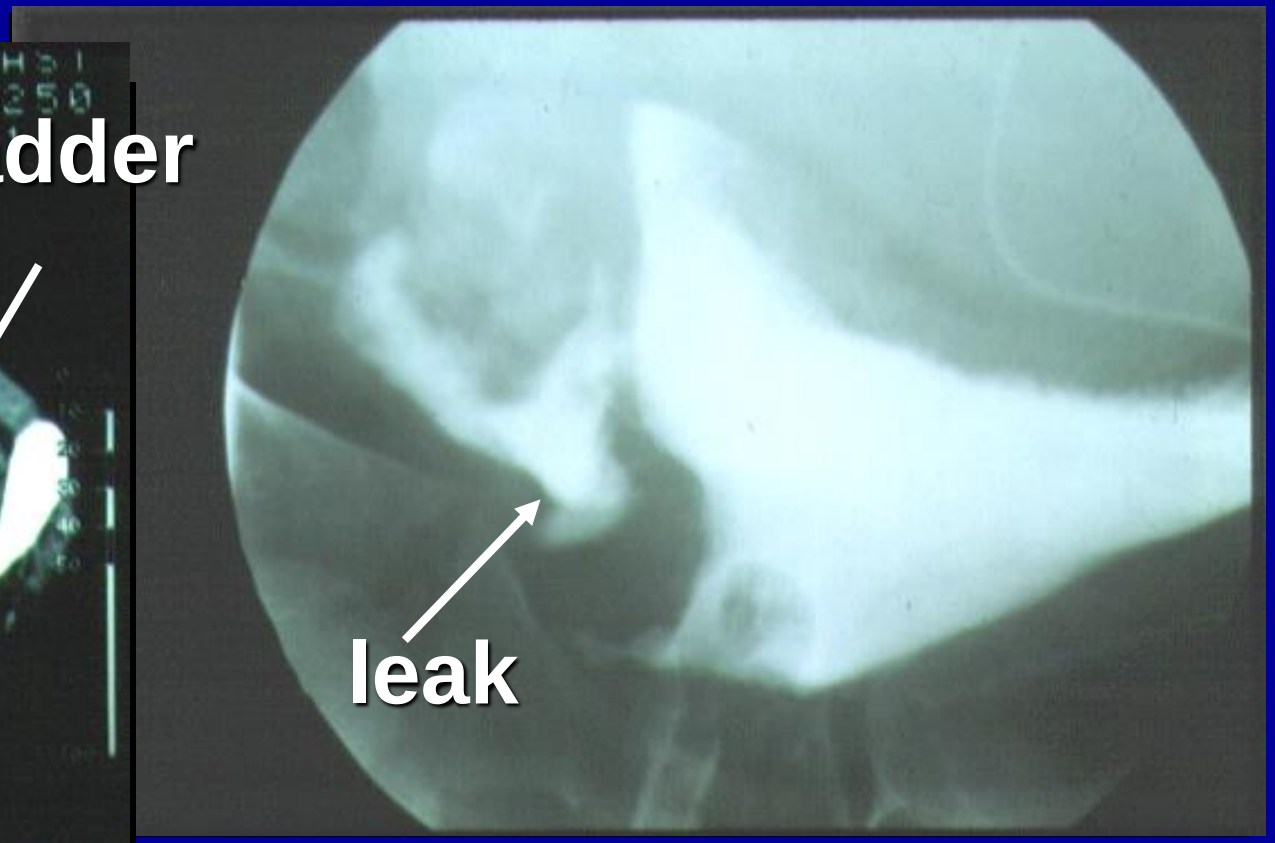
- **Two potentially lethal complications:**

  - **Perforation**
  - **Malignancy**

BLADDER AUGMENTATION

PERFORATION





Indy 500 41 patients - 53 ruptures

SPONTANEOUS BLADDER PERFORATION

500 Patients

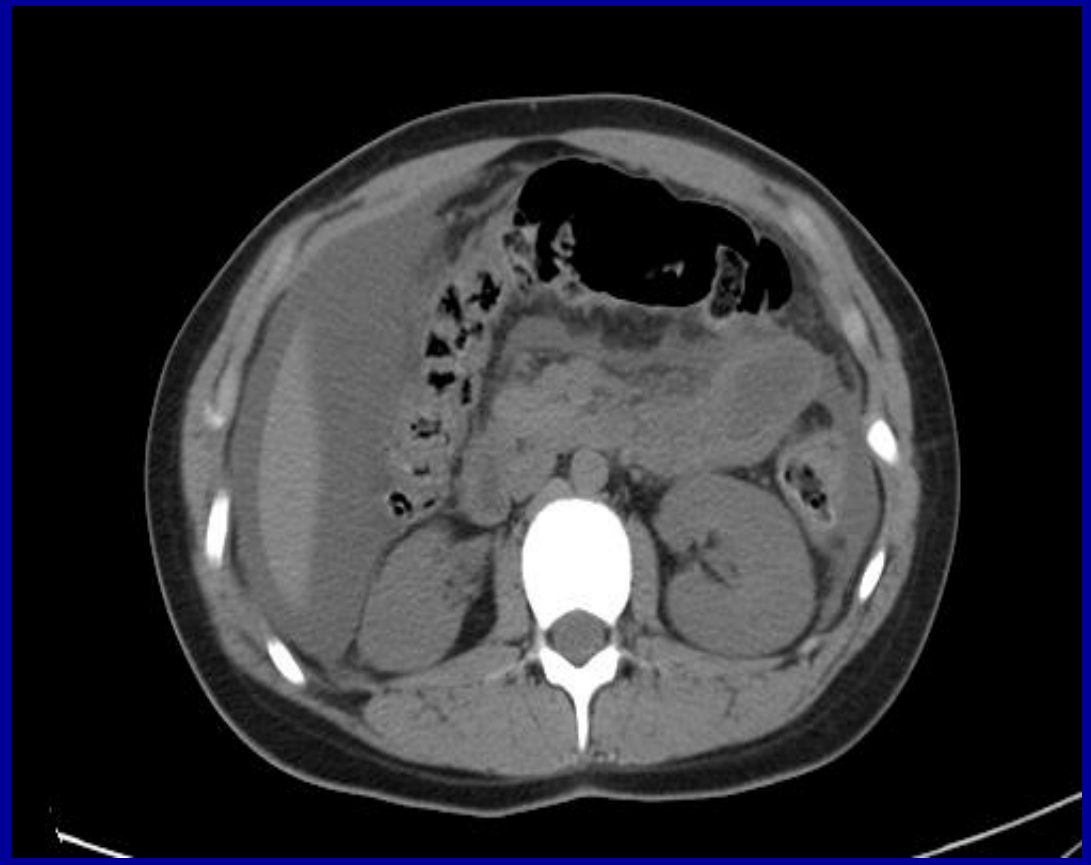
Perforation Risk:

Sigmoid	**16/84**	**(19%)**
Ileum	**23/297**	**(7%)**
Gastric	**2/44**	**(5%)**
Cecal	**2/38**	**(4%)**

Mean time aug–to–perf: 46 months

Overall Risk 43/500 (8.2%); 9 had > 1 perf.

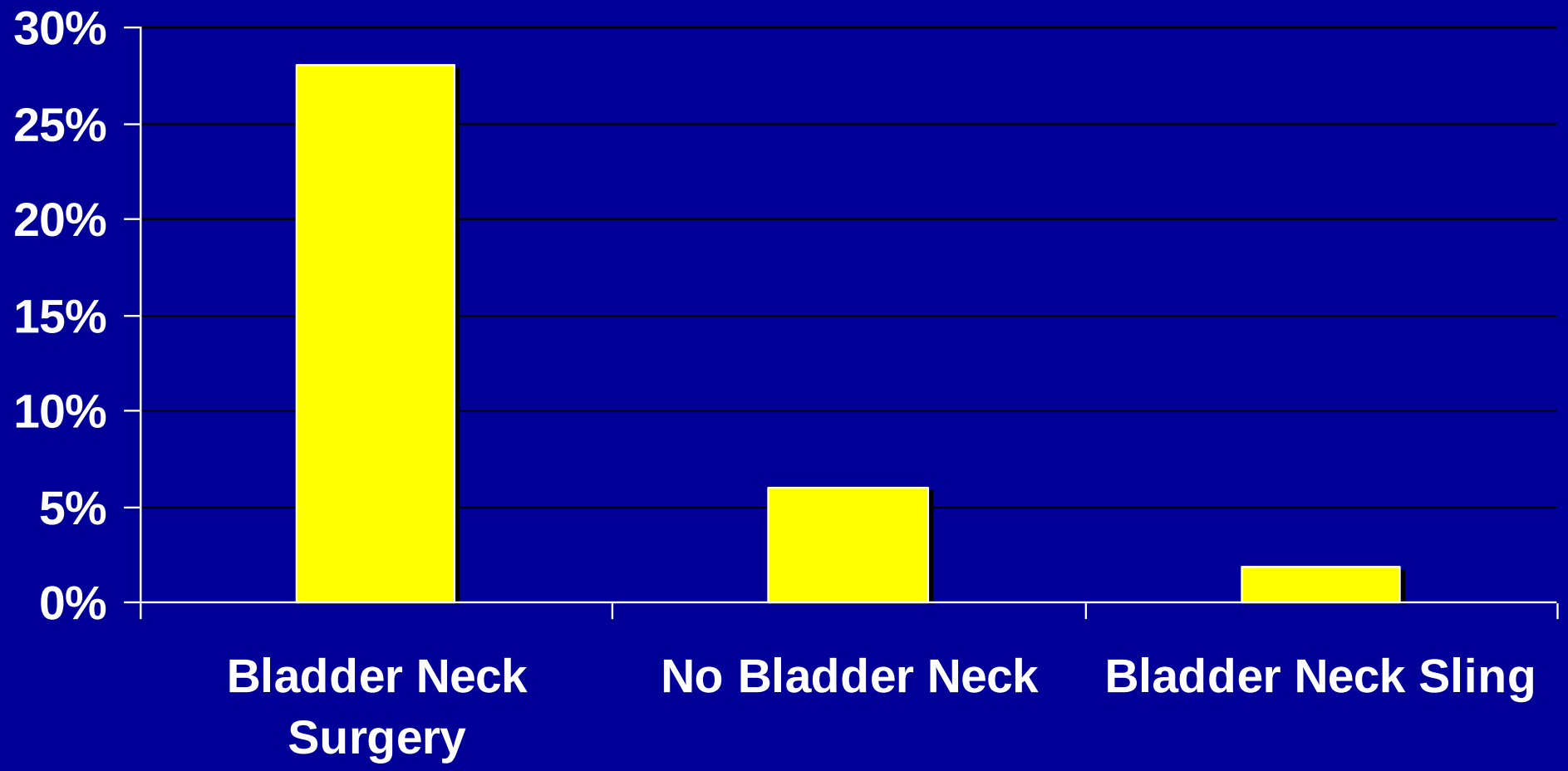
CT Cystogram for Pediatric Rupture



45 ruptured bladder augmentations in >900 pts. (5%)
2 most critical findings: symptoms, increase fluid
Karmazyn et al J Ped Urol, 2015
Poor pt. compliance, abuse increases risk (Husmann, 2016)

Risk with Bladder Neck Surgery

Perforation

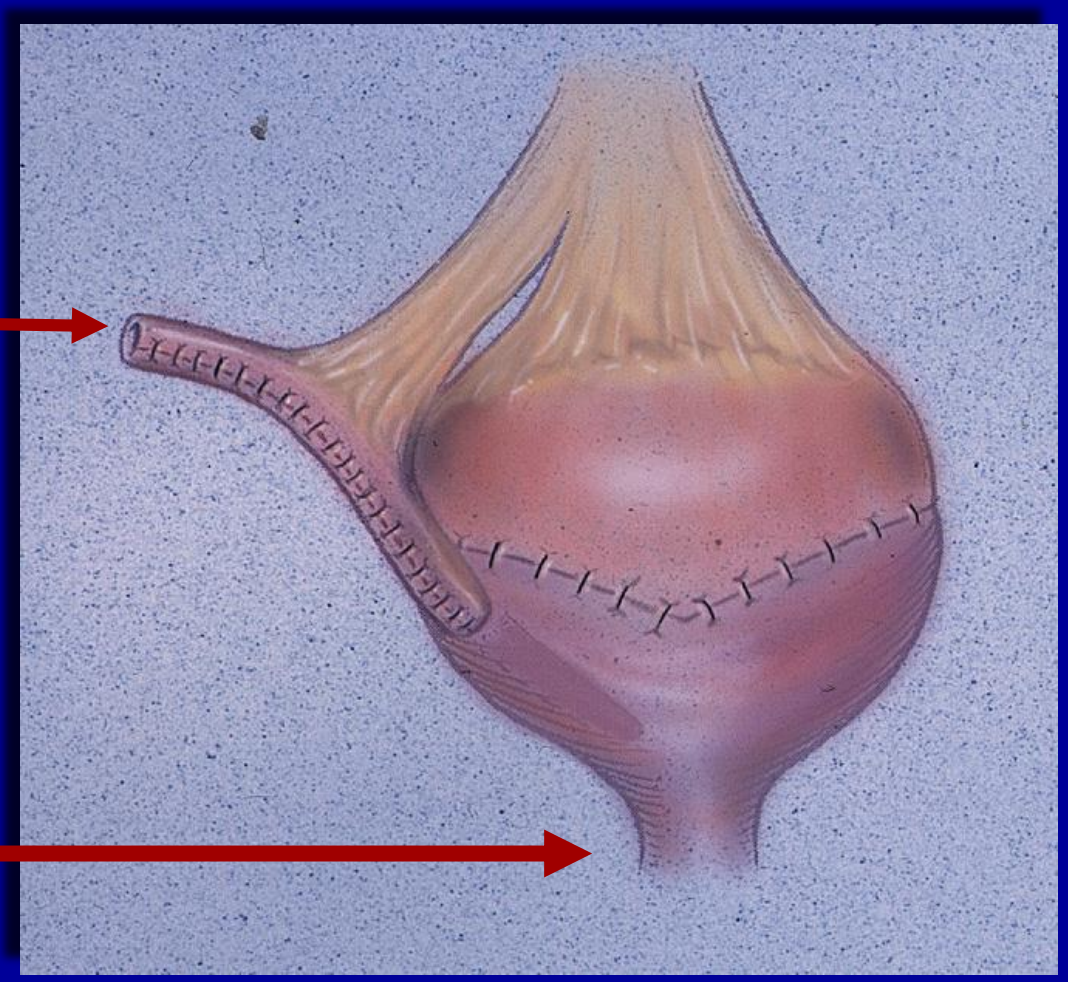


BLADDER AUGMENTATION

Perforation

CIC Catheterizable channel - 4.4%

CIC Native urethra - 12.2%



Management of Bladder Perforation

- Laparotomy, bladder closure, drainage
- Rarely percutaneous drainage and bladder drainage
  - Still need externalization of VP shunt

INDIANA EXPERIENCE: CA

483 PTS. with AUGMENTATIONS

260 > 10 year F/U

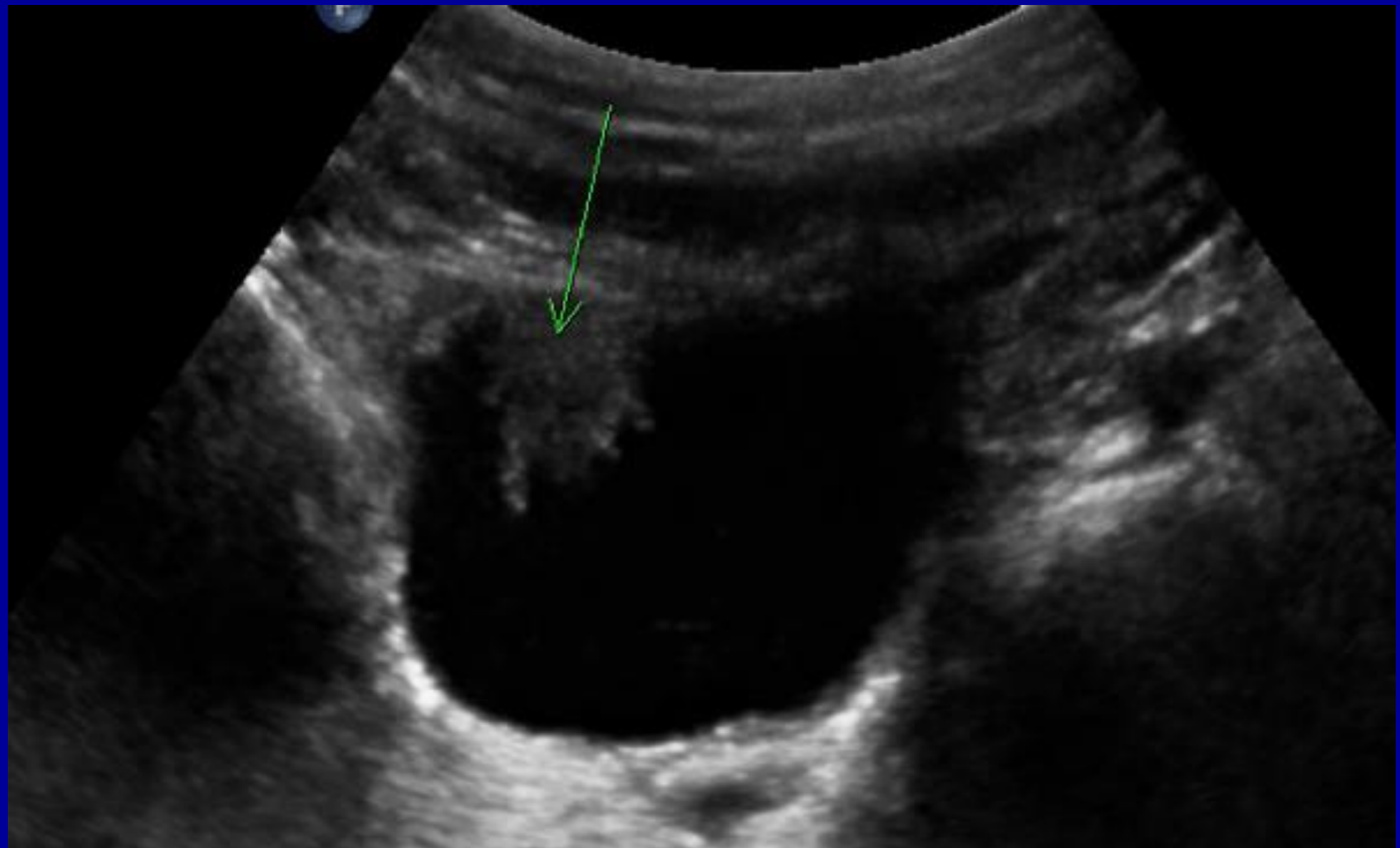
3 patients presented with metastatic TCC

- **Age at Augmentation**
  - 8, 20, and 24 yrs
- **Age at Diagnosis**
  - 29, 37, and 44 y/o respectively
  - mean time from augmentation to TCC = 19 years
- **Type of Augmentation**
  - ileocecal (2) cecal (1)

BLADDER AUGMENTATION

- 4.6% malignancy with augment; 2.6% risk CIC no augment
- 153 patients with augment; 1:1 Control group on CIC only.
- NGB - 97 pts
  - 2% (2 / 97) TCC s/p augment; 3% (3/97) on CIC no aug
  - > 2PPD tobacco use > 25 yrs.
- Exstrophy – 39 pts
  - 8% (3 / 39) adenocarcinoma s/p augment
  - 3% (1/38) adenocarcinoma on CIC without augmentation
- PUV – 18 pts
  - 12% (2 / 18) CA s/p augment; 0/18 on CIC
  - both had renal transplant
  - immunosuppressed - viral cystitis increase risk

Nephrogenic Adenoma – Benign Tumor



BLADDER AUGMENTATION

Recommendations – IU Current

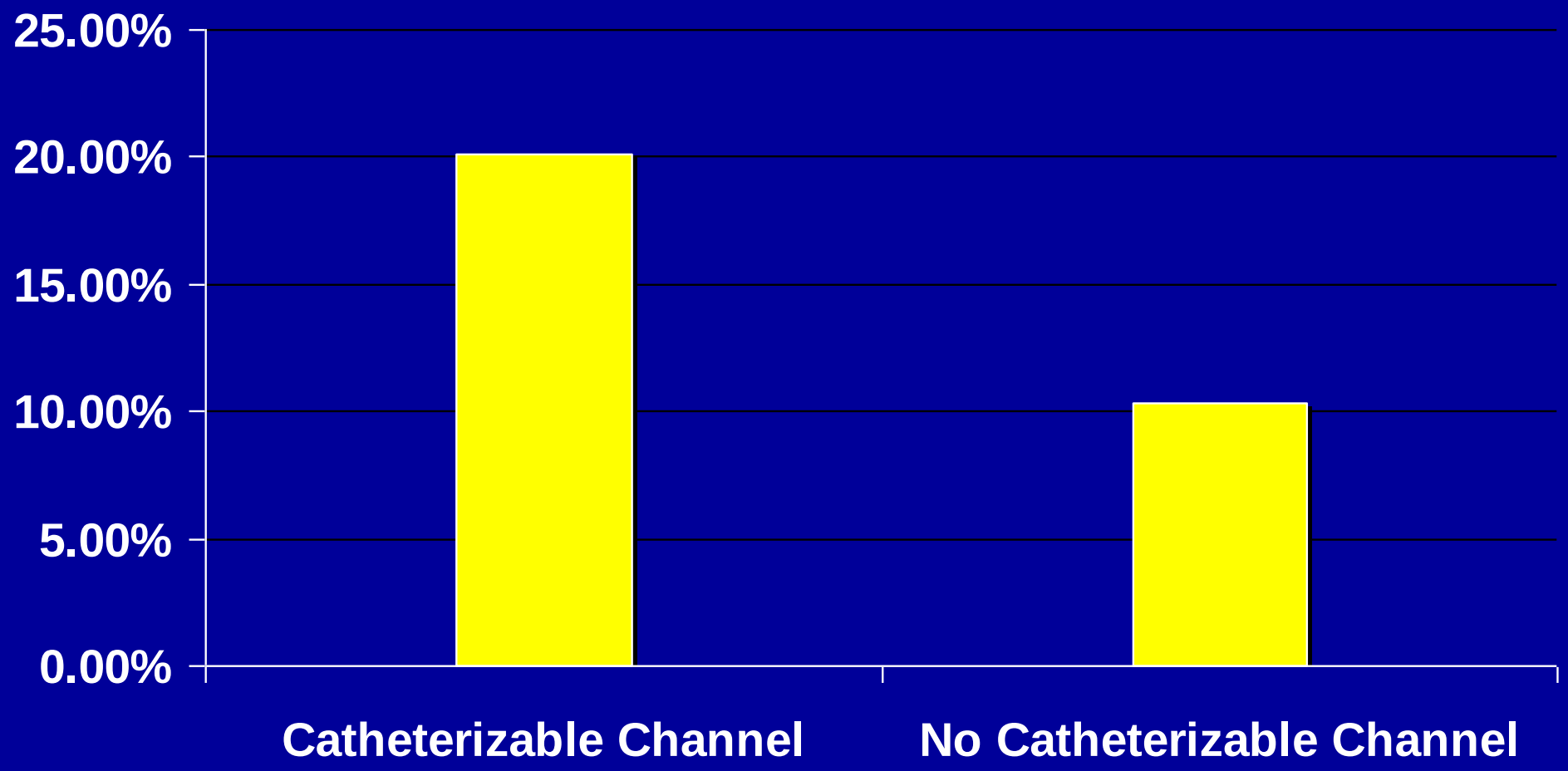
Routine Follow-Up For Cancer Screening

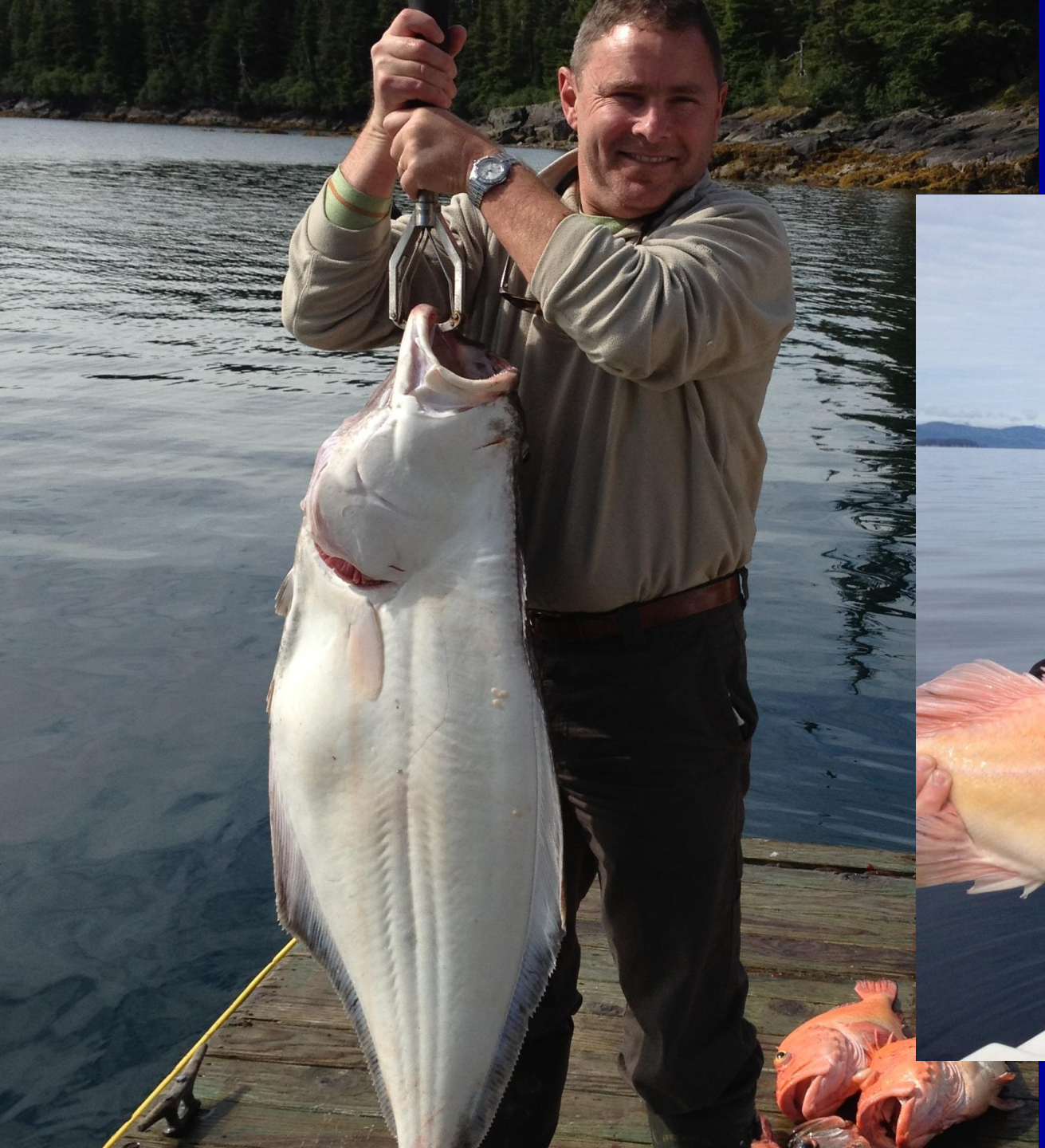
- Renal bladder ultrasound yearly
- KUB yearly
- Cytology:
  - yearly beginning at 5 years **(unhelpful)**
- Cystoscopy:
  - yearly beginning at 7-10 years in high risk **(?)**
  - **hematuria, recurrent UTI, pain, US mass**

BLADDER STONES
#1 Problem

- Not mentioned in early series
- Hendren - 1990 18%
- Hirst - 1991 18%
- Blyth - 1992 30%
- Palmer - 1993 52%
- Boston - 1998 13%
- Indiana - 2006 15%

Stone Risk with Catheterizable Channel



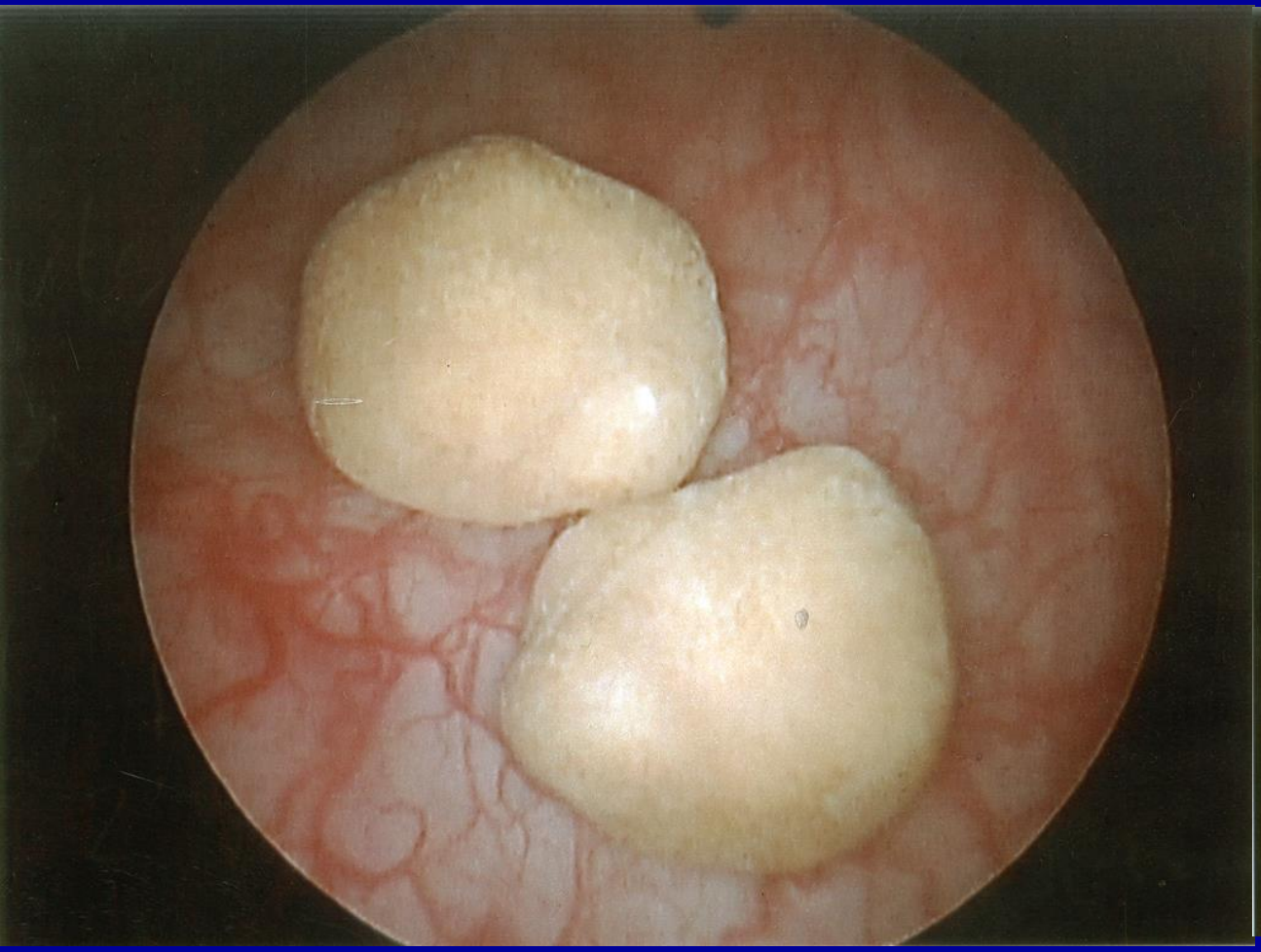


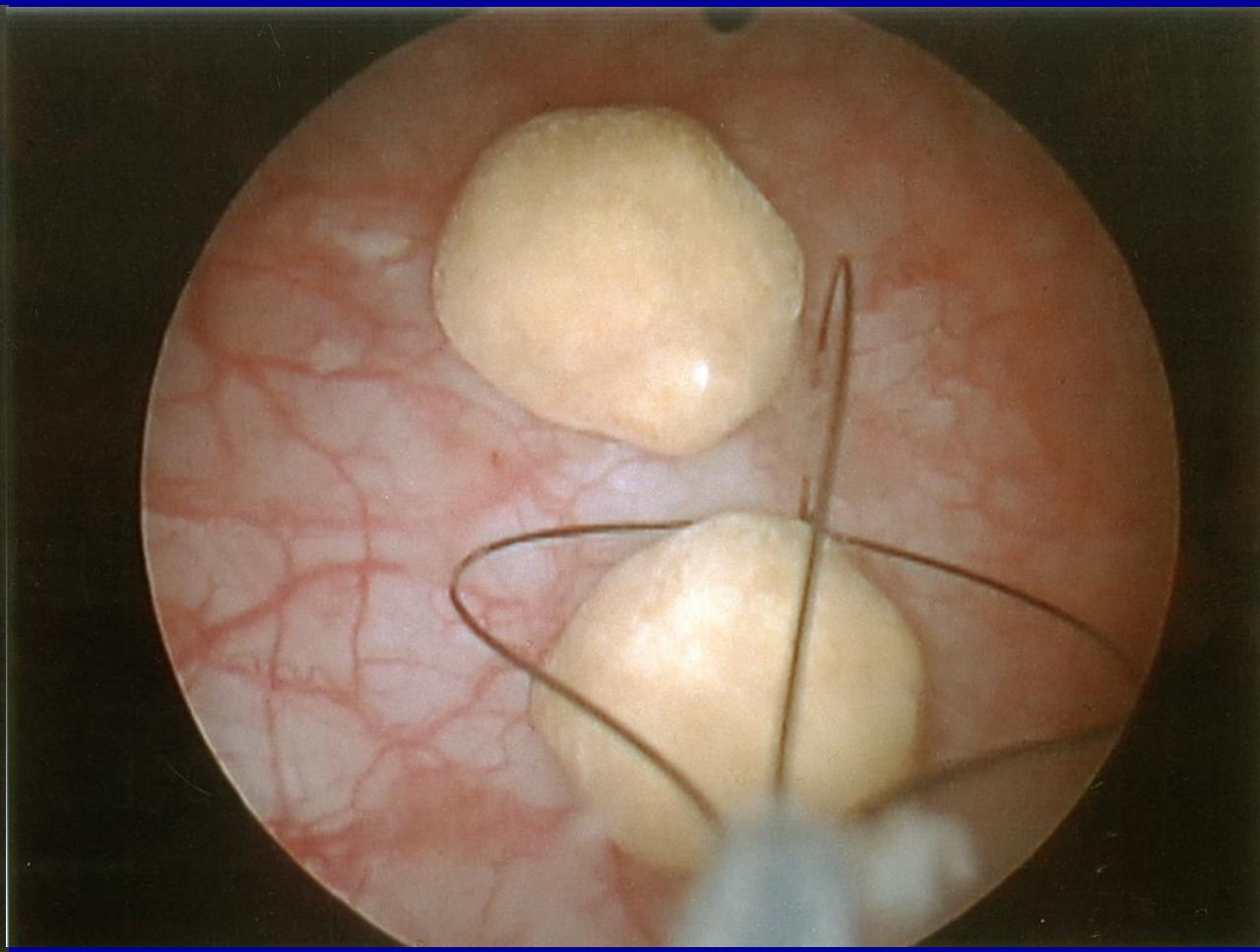

PRO
OUTFITTERS
MONTANA

10-50% Risk

Not all stones Created equal…

Bladder Stones – Percutaneous Removal






INCHES
INOX
Vmade

Stone Recurrence in Augmented Bladder

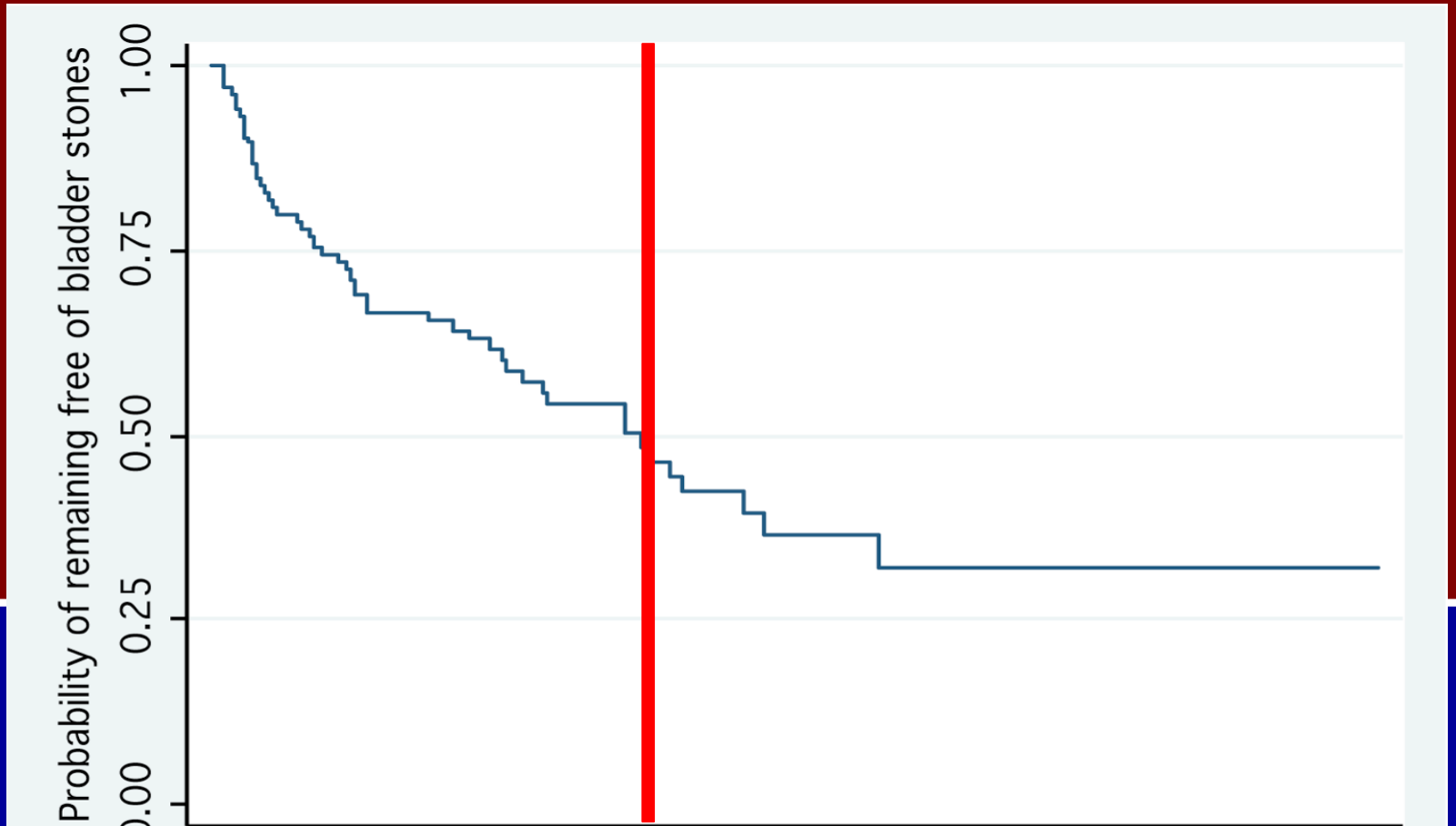
To determine if recurrence correlated with treatment modality:

1.Open vs. endoscopic vs. percutaneous
2.Stone fragmentation

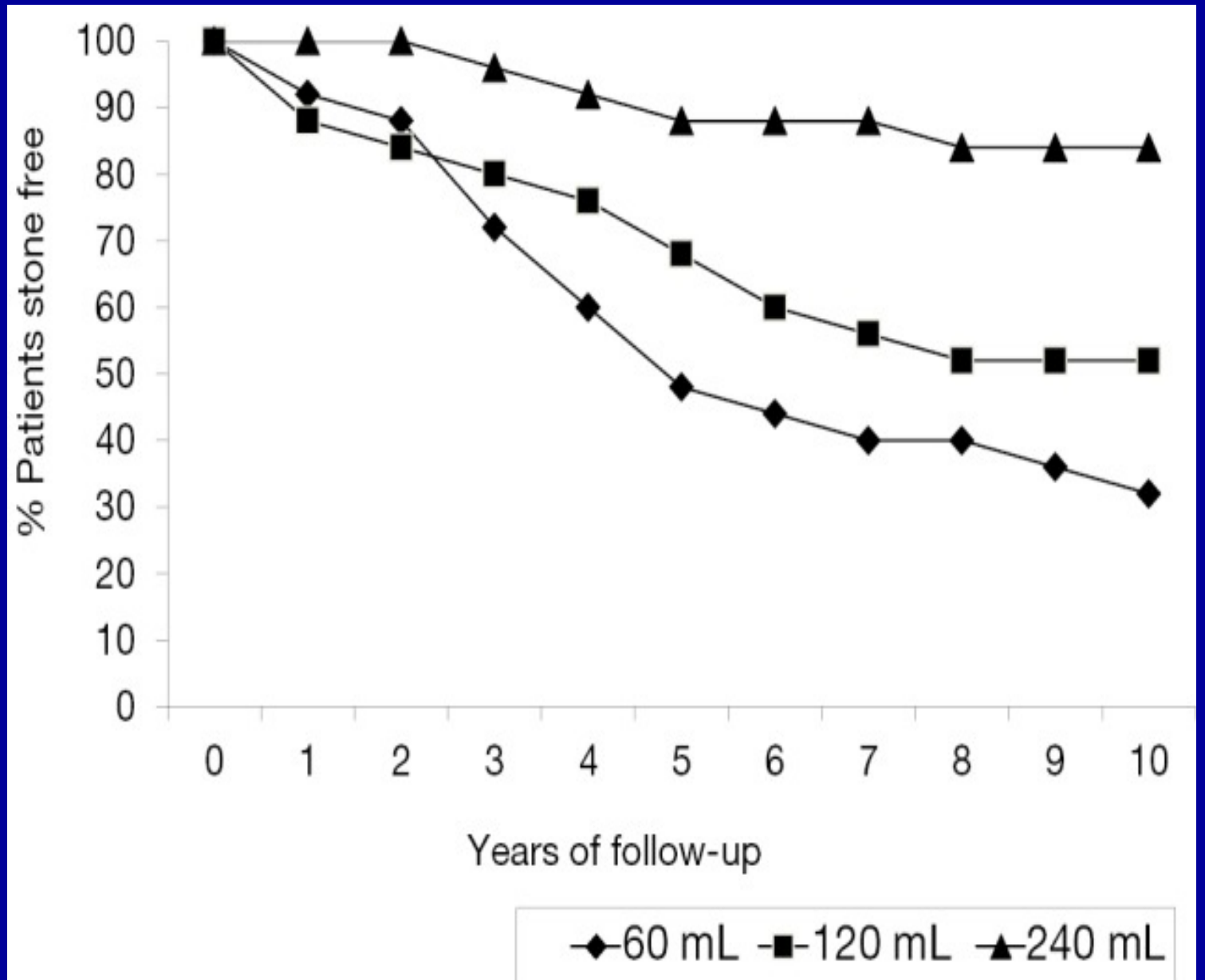
Retrospective review of 107 patients treated for bladder stones at our institution (1981-2013)

Results

Stones recurred in 51 (47.7%) patients at median 9.5 years (range 3mo.-14.7 years)



High Volume Lavage



AUGMENTING THE AUGMENTED BLADDER

Significant Contractions

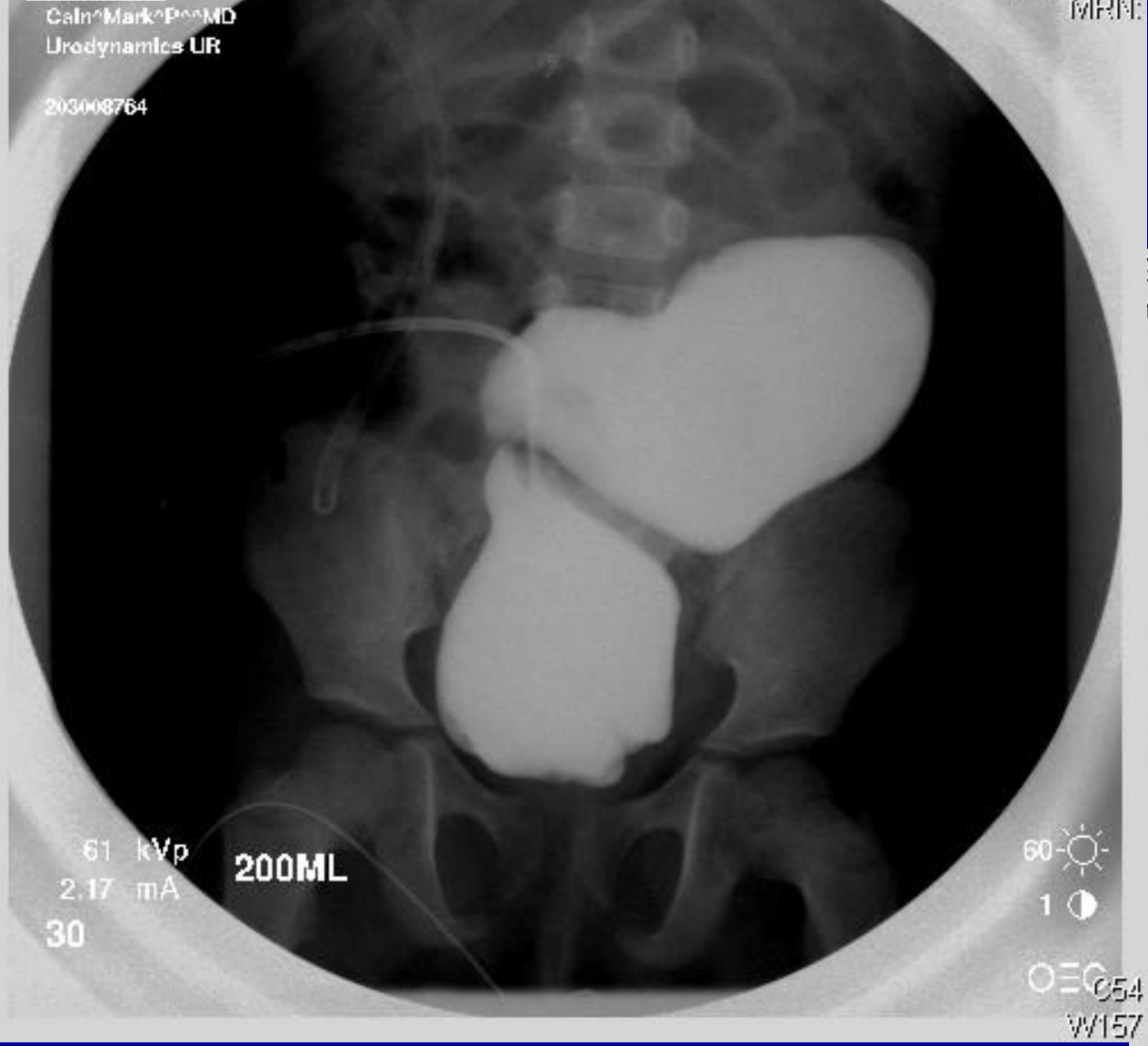
19/323 (5.9%)

Initial Bowel Segment	No. Total	(%)
Sigmoid	12/87	(13.8%)
Gastric	4/39	(10.3%)
Ileocecal	1/48	(2.1%)
Ileal	2/145	(1.4%)

Indy 500 Update:

- 9.4% (47pts) of 500 pts.

Pope, et al
J. Urol, 1998


Caln^Mark^P^^MD
Urodynamics UR
203008764
MRN:
61 kVp
2.17 mA
200ML
30
60
1
C54
W157


Study Date:11/3/2005
Study Time:12:58:33 PM
MRN:
Caln^Mark^P^^MD
Urodynamics UR
203008764
60 kVp
2.12 mA
63
7

Serum B12 Following Augmentation Metabollic Consequences

Ileal Enterocystoplasty and B12 Deficiency in Pediatric Patients

David H. Rosenbaum,* Mark P. Cain, Martin Kaefer, Kirstan K. Meldrum, Shelly J. King, Rosalia Misseri and Richard C. Rink

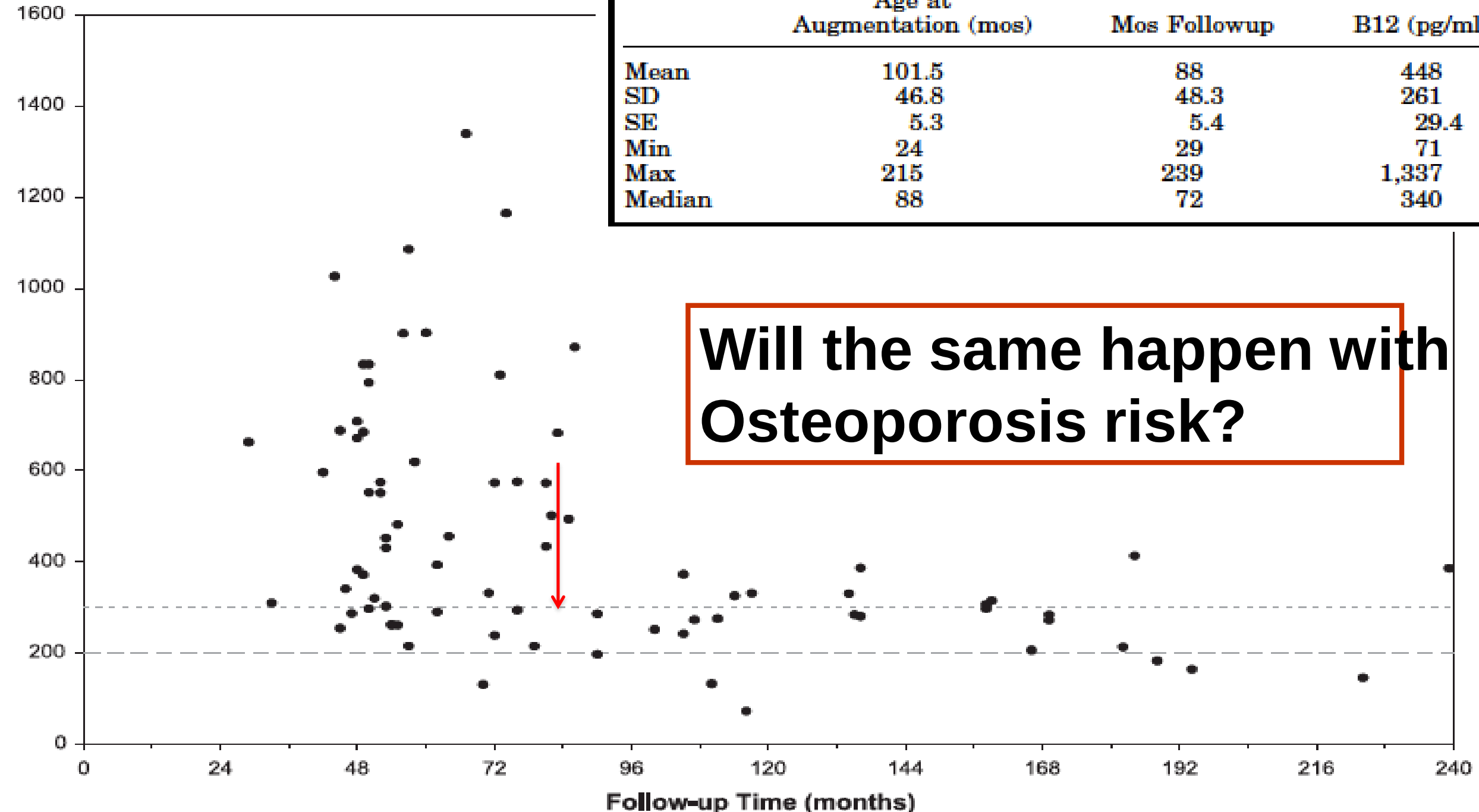
From the Division of Pediatric Urology, Riley Hospital for Children, Indianapolis, Indiana

Purpose: Vitamin B12 deficiency is a feared complication of enterocystoplasty but it has never been demonstrated in pediatric patients who have undergone ileal enterocystoplasty. We reviewed our series of more than 500 bladder augmentations in an attempt to define the timing and risk of vitamin B12 deficiency in pediatric patients after bladder augmentation.
Materials and Methods: From October 2004 to present we obtained serum B12 values in patients who had undergone bladder augmentation at our institution. We looked at patients who had undergone ileal enterocystoplasty and who were 18 years or younger at the time of augmentation. Any B12 value that was obtained while on any form of B12 supplementation was excluded. These criteria resulted in 79 patients with 105 B12 values. B12 values of 200 pg/ml or less were considered "low," and values between 201 and 300 pg/ml were considered "low-normal."
Results: There was a statistically significant correlation between followup time and serum B12 ($p = 0.0001$). The probability of low B12 increased as followup time increased ($p = 0.007$), as did the probability of low-normal B12 ($p = 0.005$). Starting at 7 years postoperatively 6 of 29 patients (21%) had low B12 values, while 12 of 29 (41%) had low-normal values.
Conclusions: Pediatric patients who have undergone ileal enterocystoplasty are at risk for development of vitamin B12 deficiency. These patients are at the highest risk beginning at 7 years postoperatively, and the risk increases with time. We recommend an annual serum B12 value in children beginning at 5 years following bladder augmentation.

Key Words: vitamin B 12 deficiency, urinary bladder, pediatrics

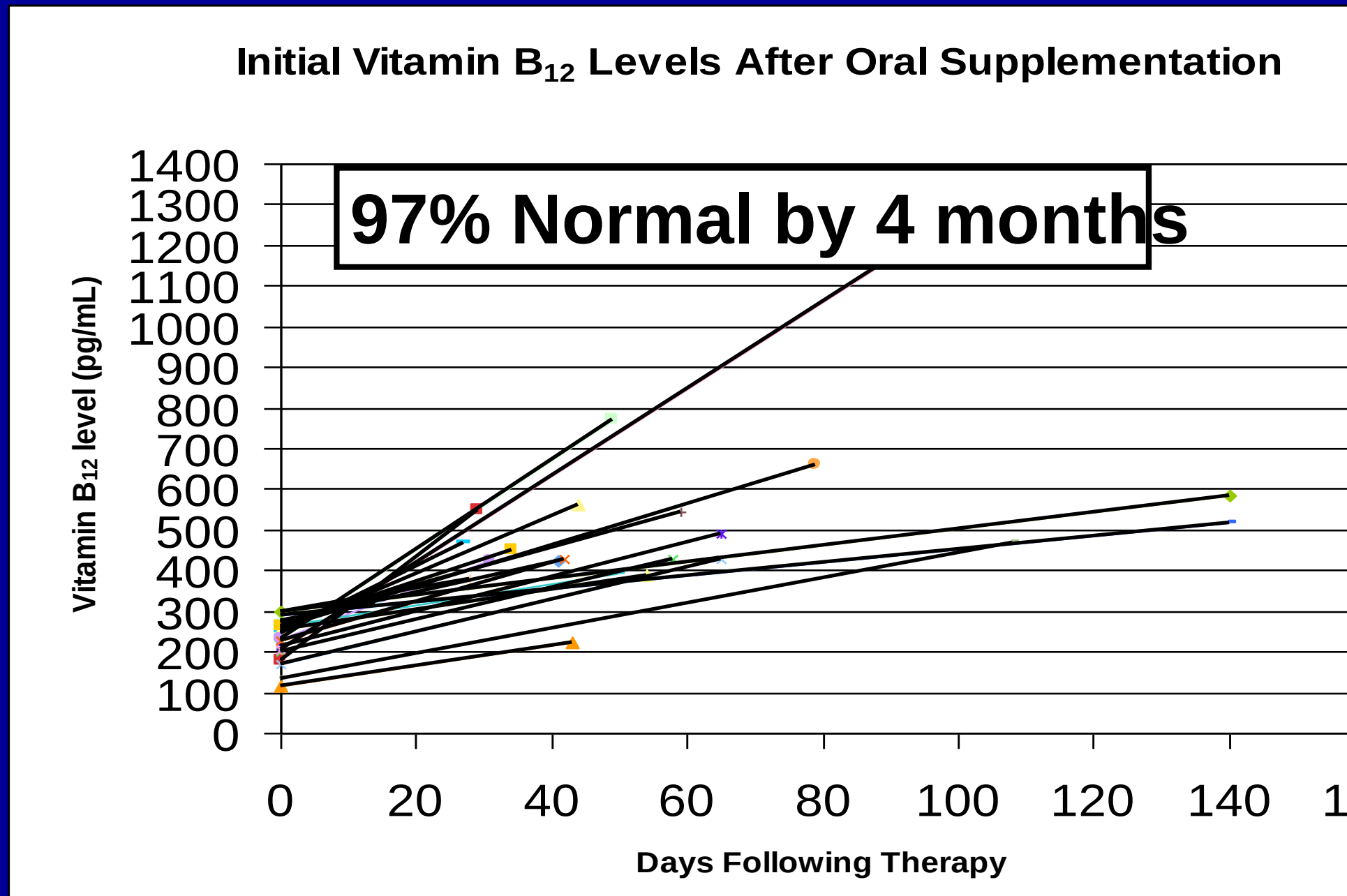
TABLE 1. Statistical data on total B12 values

	Age at Augmentation (mos)	Mos Followup	B12 (pg/ml)
Mean	101.5	88	448
SD	46.8	48.3	261
SE	5.3	5.4	29.4
Min	24	29	71
Max	215	239	1,337
Median	88	72	340



Will the same happen with Osteoporosis risk?

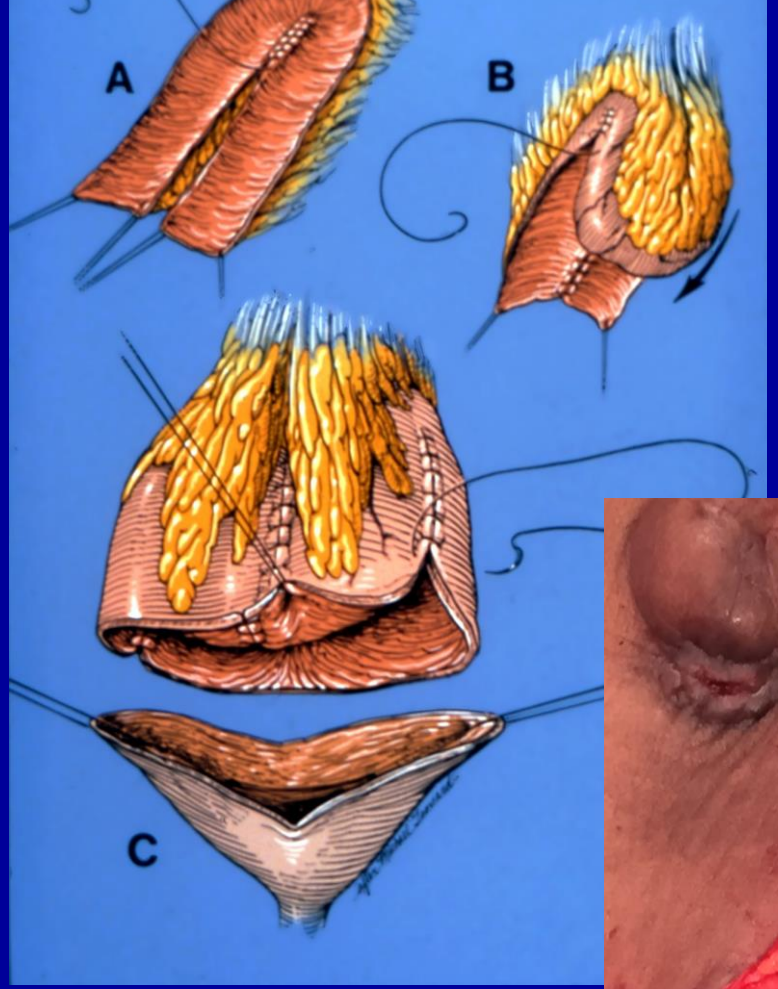
RESULTS

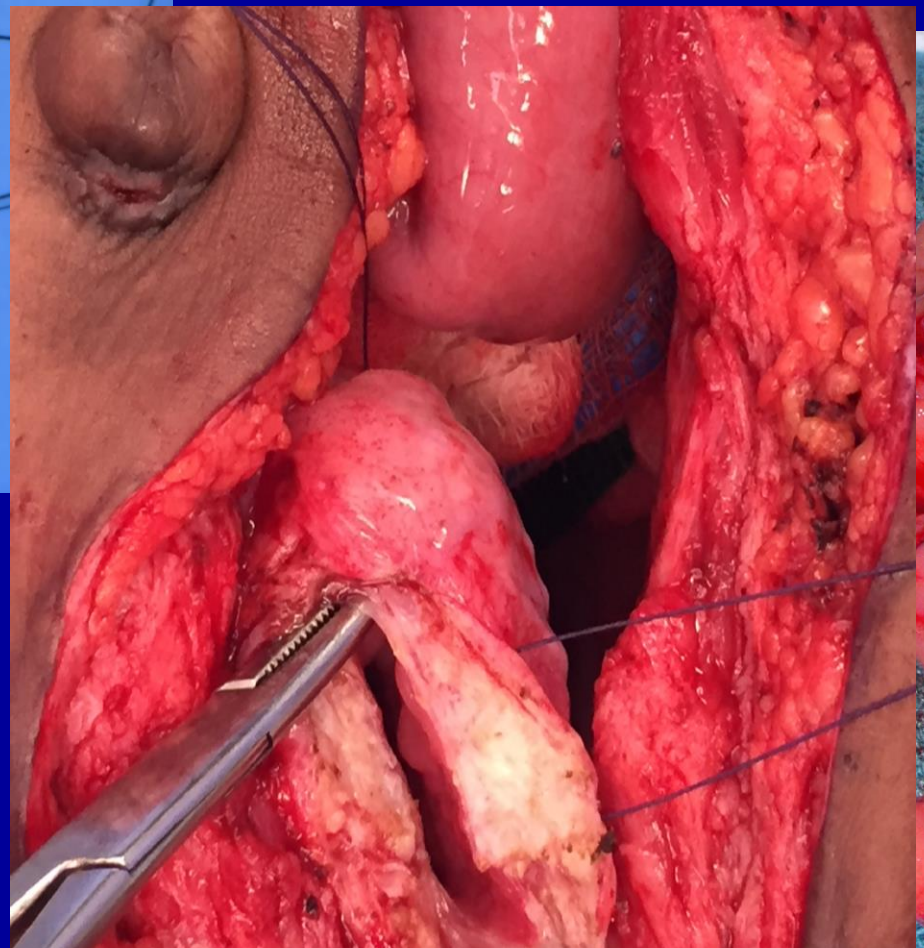

Initial Vitamin B_{12} Levels After Oral Supplementation
97% Normal by 4 months
Vitamin B_{12} level (pg/mL)
1400
1300
1200
1100
1000
900
800
700
600
500
400
300
200
100
0
0
20
40
60
80
100
120
140
160
Days Following Therapy

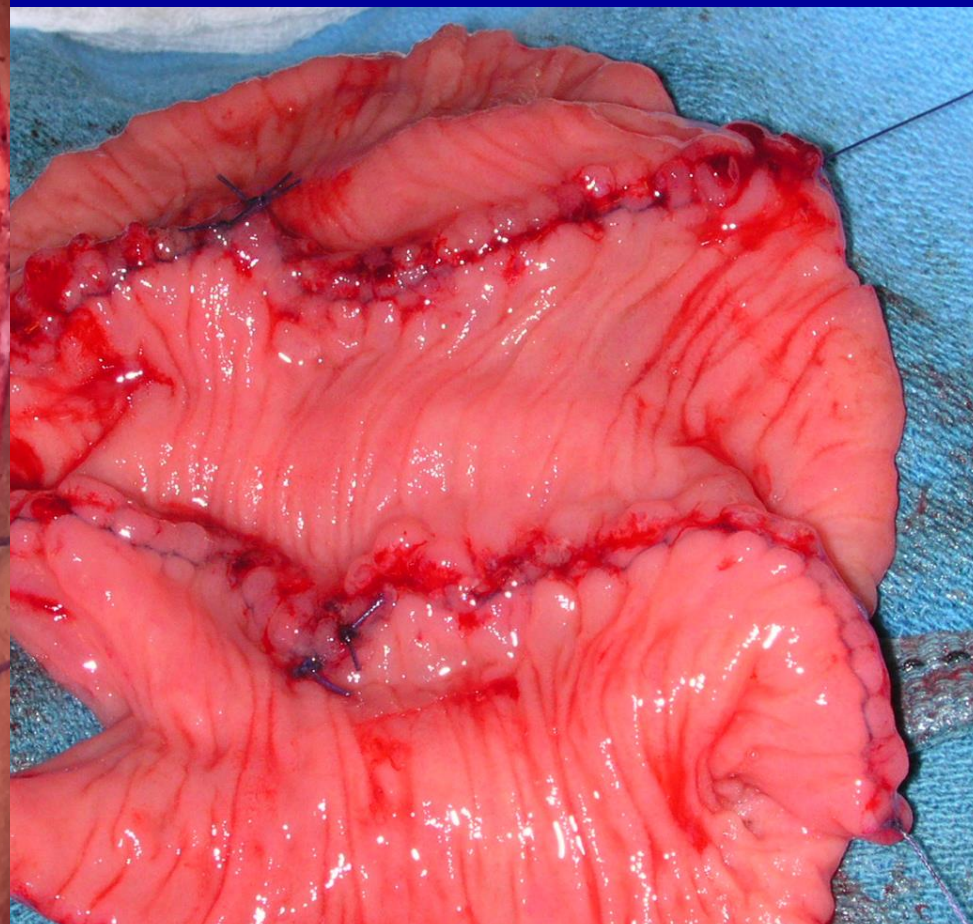
Long term outcomes of Oral B12 = POOR COMPLIANCE…….

- 25/36 patients had documented follow-up with multiple serum B12 levels after initiation of oral therapy (mean F/U 49 months)
- Only 9/25 patients (36%) had normal serum B12 levels on most recent follow-up
- Need to consider injection therapy for low B12?

Still The Gold Standard (For Some Patients)





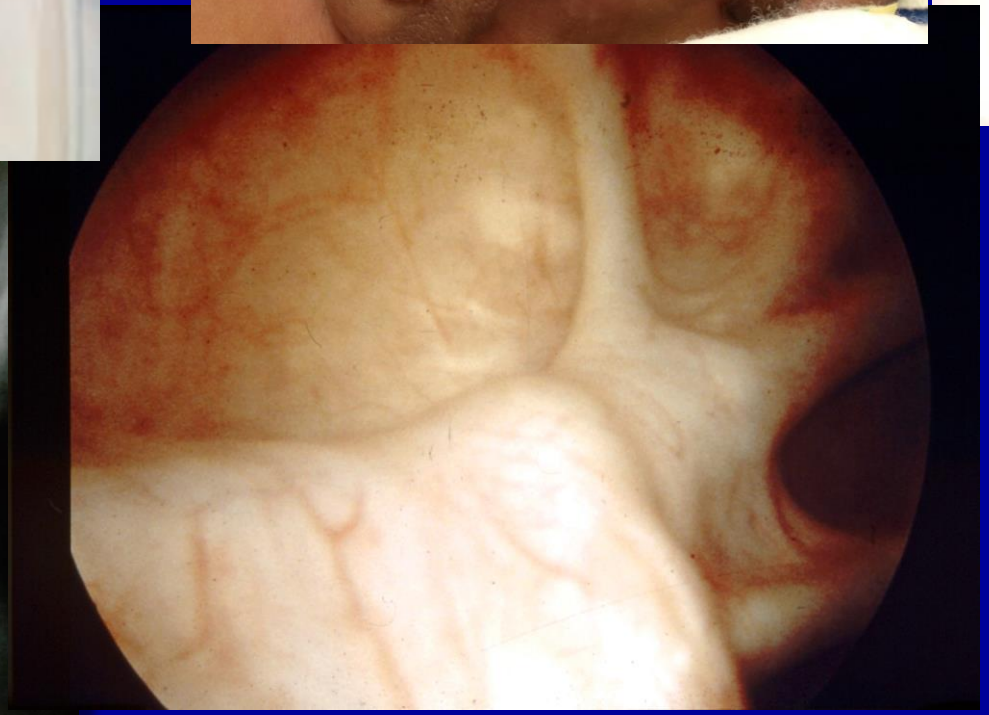


What you need to remember:

- Yearly follow up
  - BMP, CBC, B12, KUB, RBUS
- Prompt evaluation for gross hematuria, abdominal pain
  - Cystoscopy, CT cystogram
- Catheterization problems are an emergency
- Pregnancy and delivery will require your presence
- Involve your pediatric/transitional urologist

Gracias!

Problem: Medical, Surgical and Social



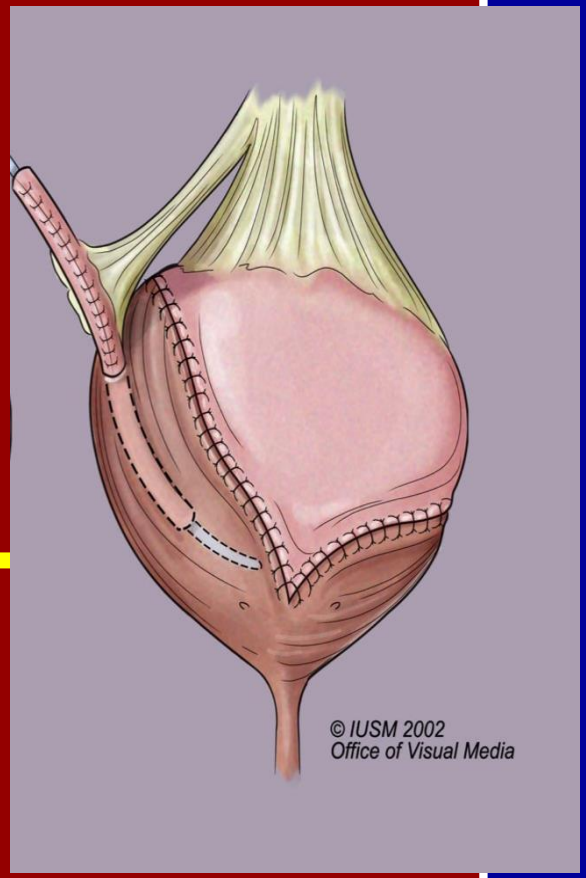
Riley Pediatric Urology Team 2018

Management of Neuropathic Bladder and Bowel: Historical Surgical Pathway to Continence

Evolution of continence:

- Intermittent catheterization
- Bladder outlet resistance
- Bladder augmentation

- Mitrofanoff procedure
- MACE procedure



Ideal BN Procedure: Does Not Exist

- Dry at 3-4 hours +/- CIC
- No change in bladder capacity/compliance
  - ie avoids need for augmentation
- Preserve kidneys, ie allows leakage at high bladder P
- Allow easy CIC per urethra
- Long term durability
- Technically easy to perform

Long-Term Fate of the Bladder after Bladder Neck Procedure

Retrospective review: Bladder neck surgery without augmentation with greater than 4 years follow-up (1997 – 2008)

Primary outcome: incidence of augmentation
Secondary outcome: predictors of augmentation

Urodynamics reviewed and analyzed

- **Bladder capacity, DLPP, and compliance/detrusor pressure**

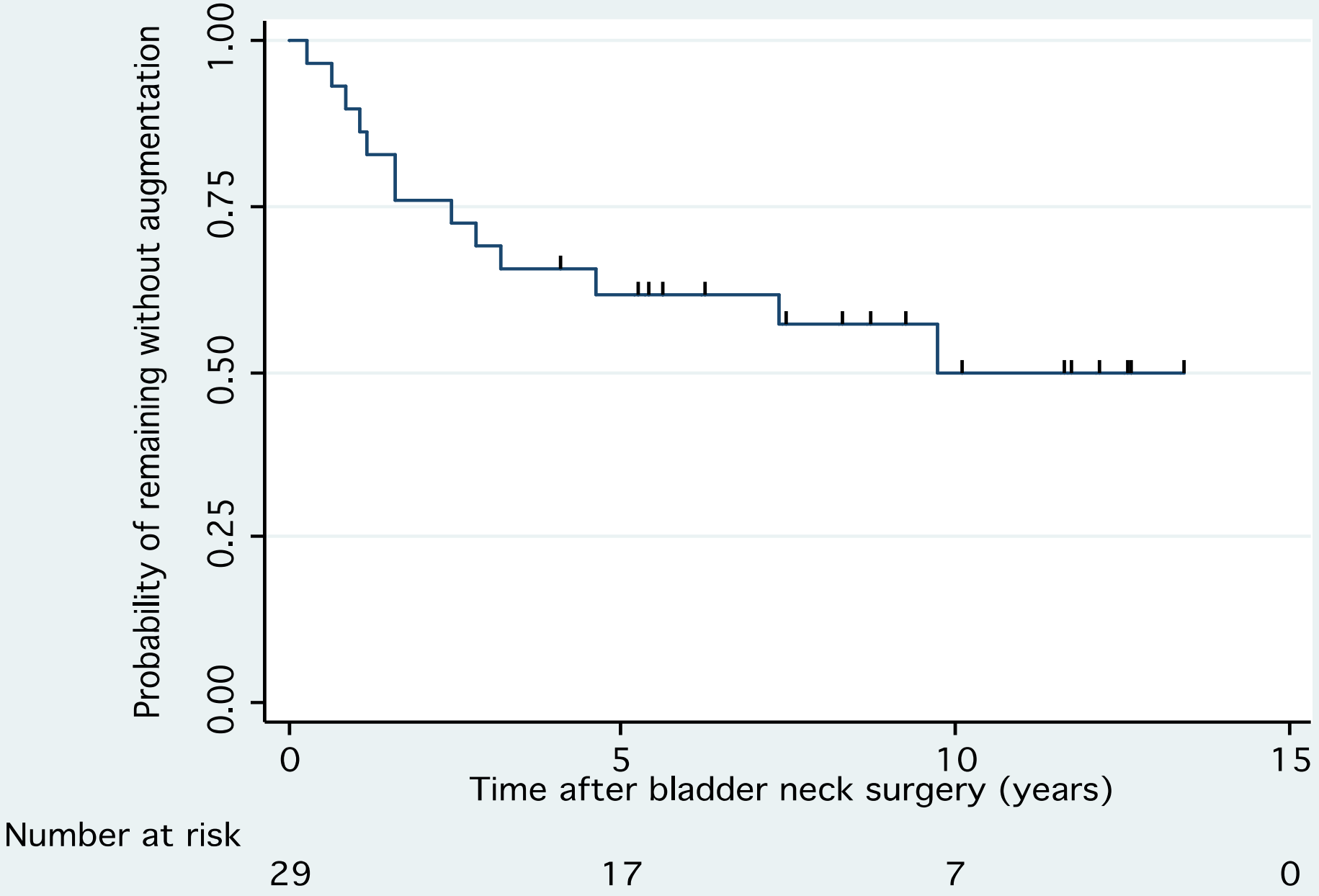
29 patients met inclusion criteria, average follow-up 8.9 years (4.0 – 13.3)

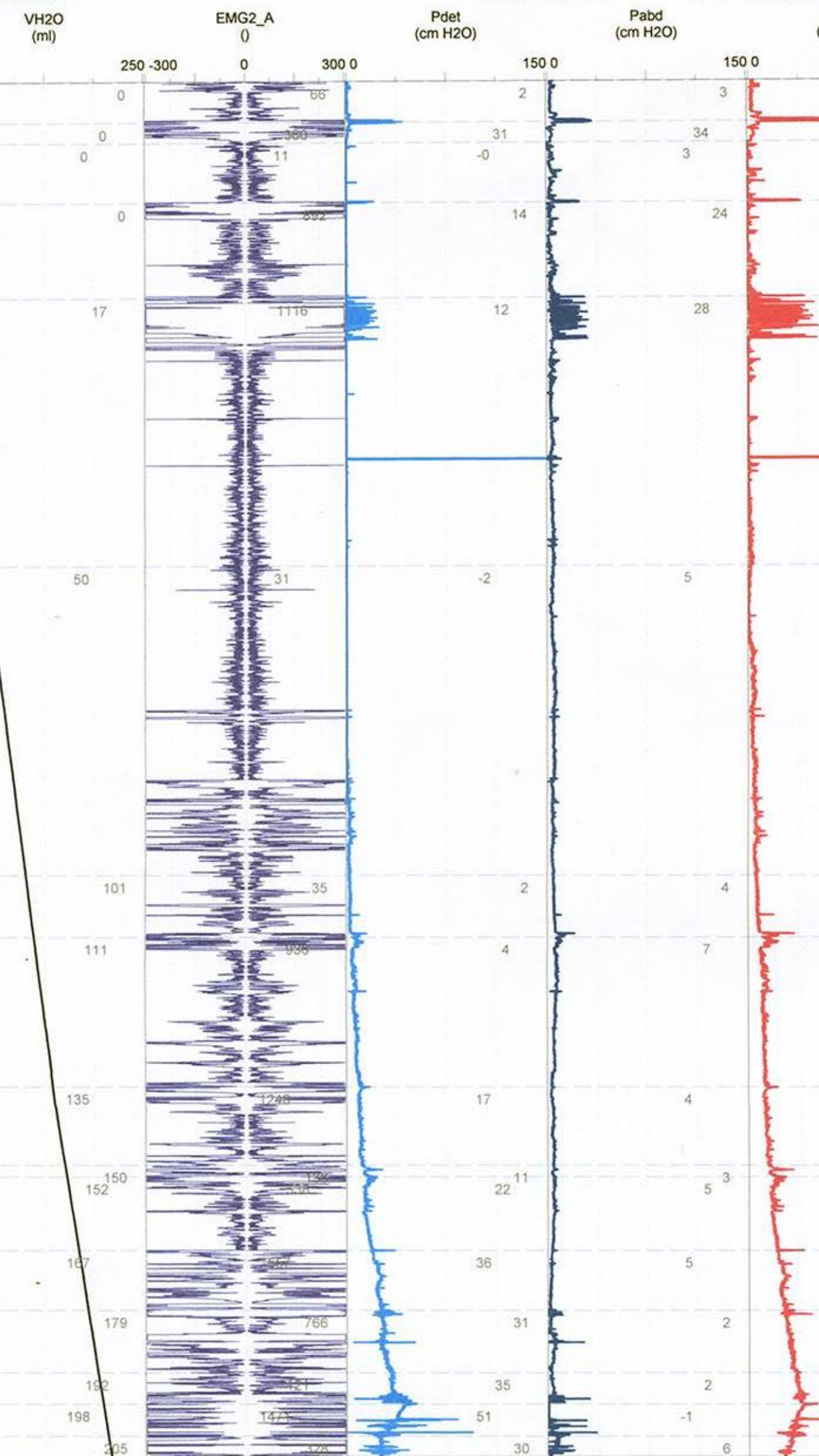
- **16 (55%) avoided augment at last follow-up**
- **13 (45%) underwent delayed augmentation**

Results: Time to Augmentation

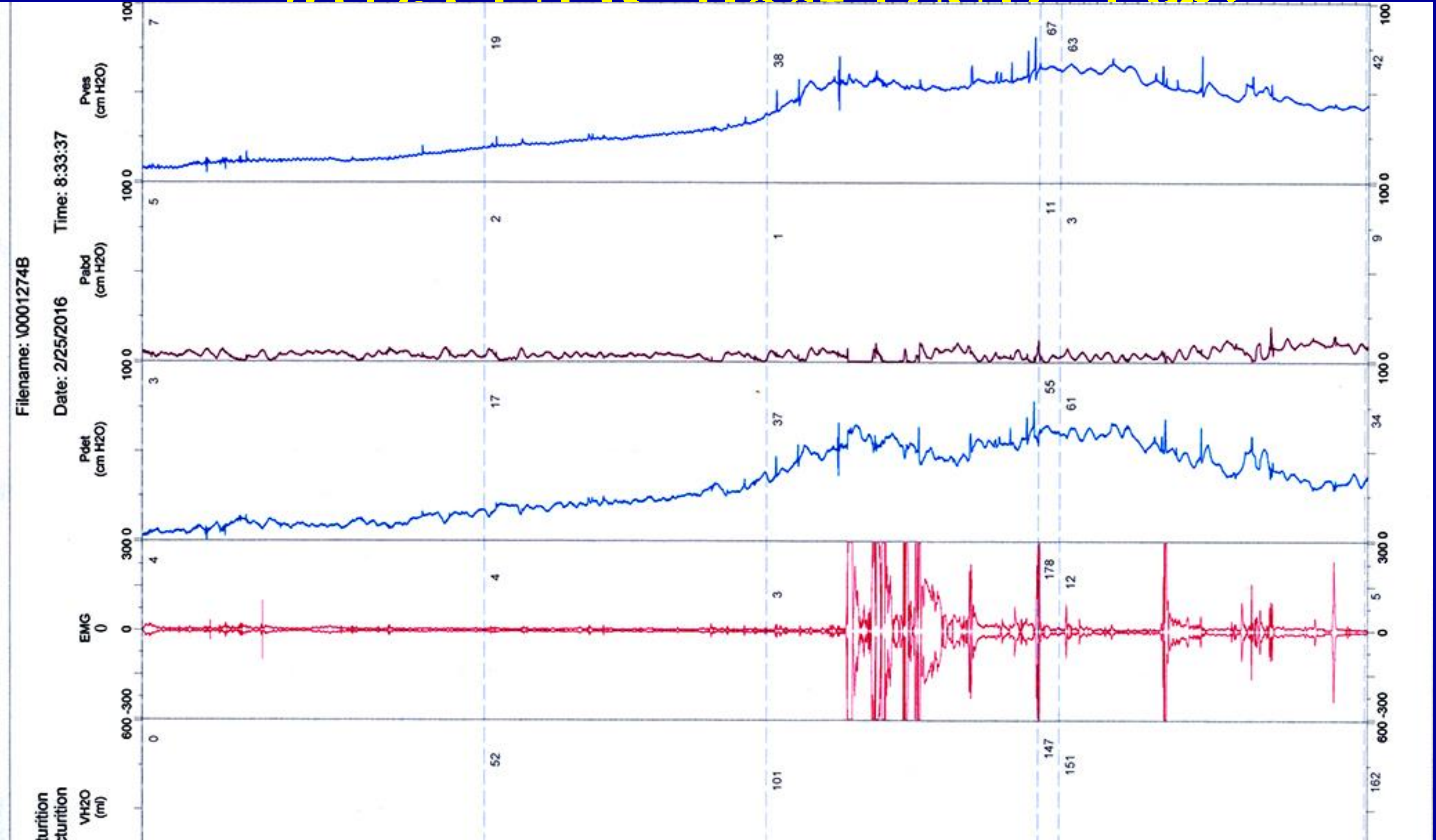
- Average Time to Augment: 3 years
- Indications for Augment
  - New/Worsening Incontinence: 5
  - New VUR/Hydronephrosis: 5
  - New Renal Scarring: 2
  - Small bladder capacity: 6
  - Poor compliance: 7
- All patients with 2+ indications for augmentation
- No significant difference between groups regarding: age, sex, ambulatory status or VP shunt.

Probability of Remaining Augment Free

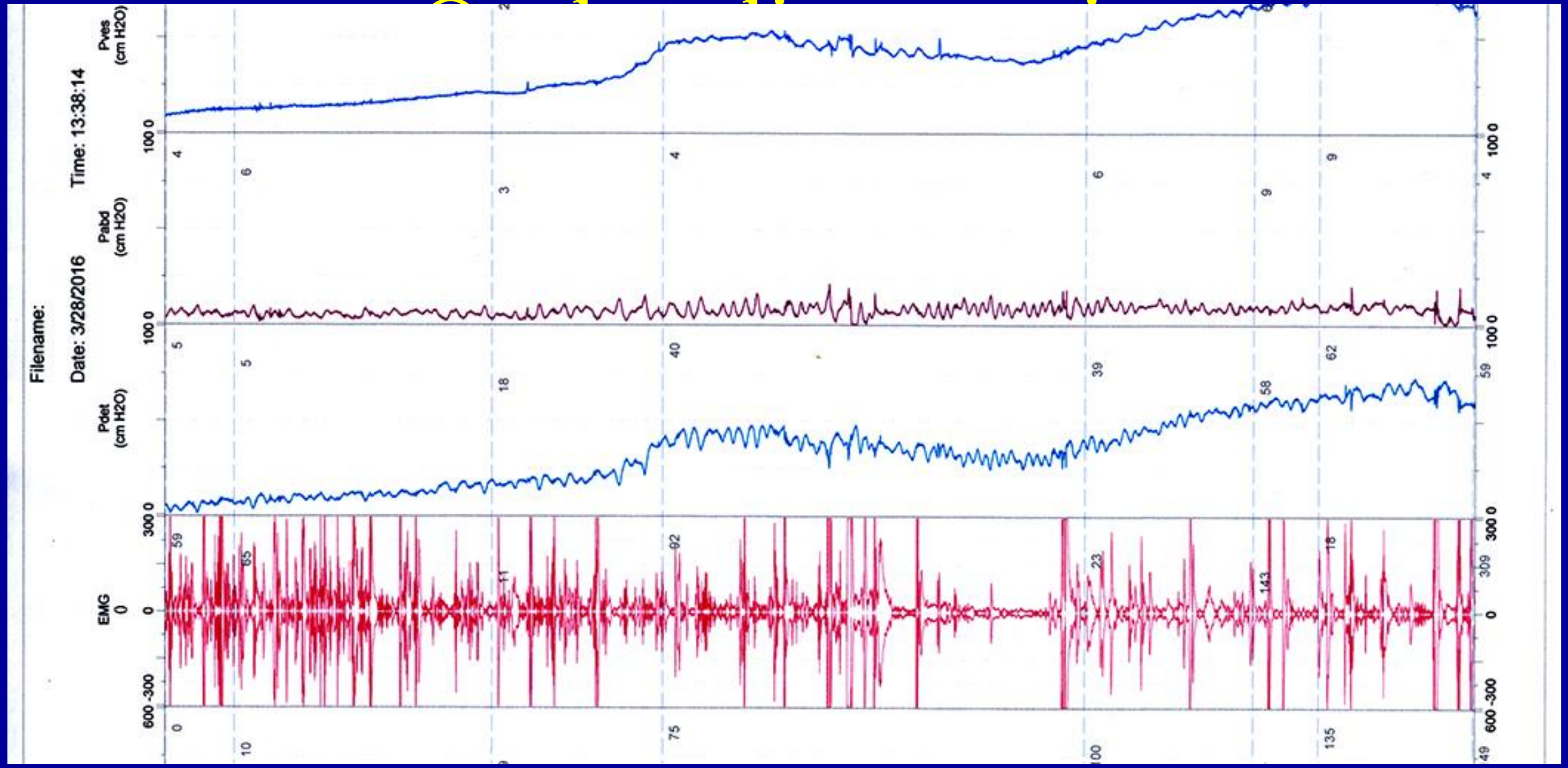



VH2O
(ml)
EMG2_A
()
Pdet
(cm H2O)
Pabd
(cm H2O)
250 -300
0
300 0
150 0
150 0

2016: UDS Post RNP, Dry



2016: UDS Post BNR +



Surgery, and Life in General……

We shall not cease from exploration SEP
And the end of all our exploring SEP
Will be to arrive where we started SEP
And know the place for the first time.

TS Eliot

The Four Quartets

Catheterizable Channels

Indiana Experience 1990-2013

Appendicovesicostomy	215
Monti ileovesicostomy	296
Continent Vesicostomy	33
Total Urinary Channels:	**544**
MACE	415
Total Cath Channels:	**959**

Mitrofanoff Principle

- Mobilization of bladder
- Mobilization of channel pedicle
- Minimize extravesical channel length
- Wide spatulation of channel on both ends
- Wide skin stomal flap
- Secure bladder to posterior abdominal wall
- Cath channel multiple times during procedure

Short, Straight, Supple

(any supple tube)

- Appendix / Ureter 1980
- Fallopian tube
- Vas deferens
- Stomach 1991
- Bladder 1992
- Yang 1993
- Tapered ileum 1994
- Prepuce 1995
- Monti – Yang 1993, 1997

Appendicovesicostomy vs. Monti

0022-5347/99/1625-1749/0
THE JOURNAL OF UROLOGY
Vol. 162, 1749–1752, November 1999
Printed in U.S.A.

APPENDICOVESICOSTOMY AND NEWER ALTERNATIVES FOR THE MITROFANOFF PROCEDURE: RESULTS IN THE LAST 100 PATIENTS AT RILEY CHILDREN'S HOSPITAL

MARK P. CAIN, ANTHONY J. CASALE, SHELLY J. KING AND RICHARD C. RINK

From the Department of Urology, James Whitcomb Riley Hospital for Children, Indiana University Medical Center, Indianapolis, Indiana

100 pts. : 57 Apv, 21 Monti, 21 CV

- 98% continence, 12% stomal stenosis

- 20 secondary procedures

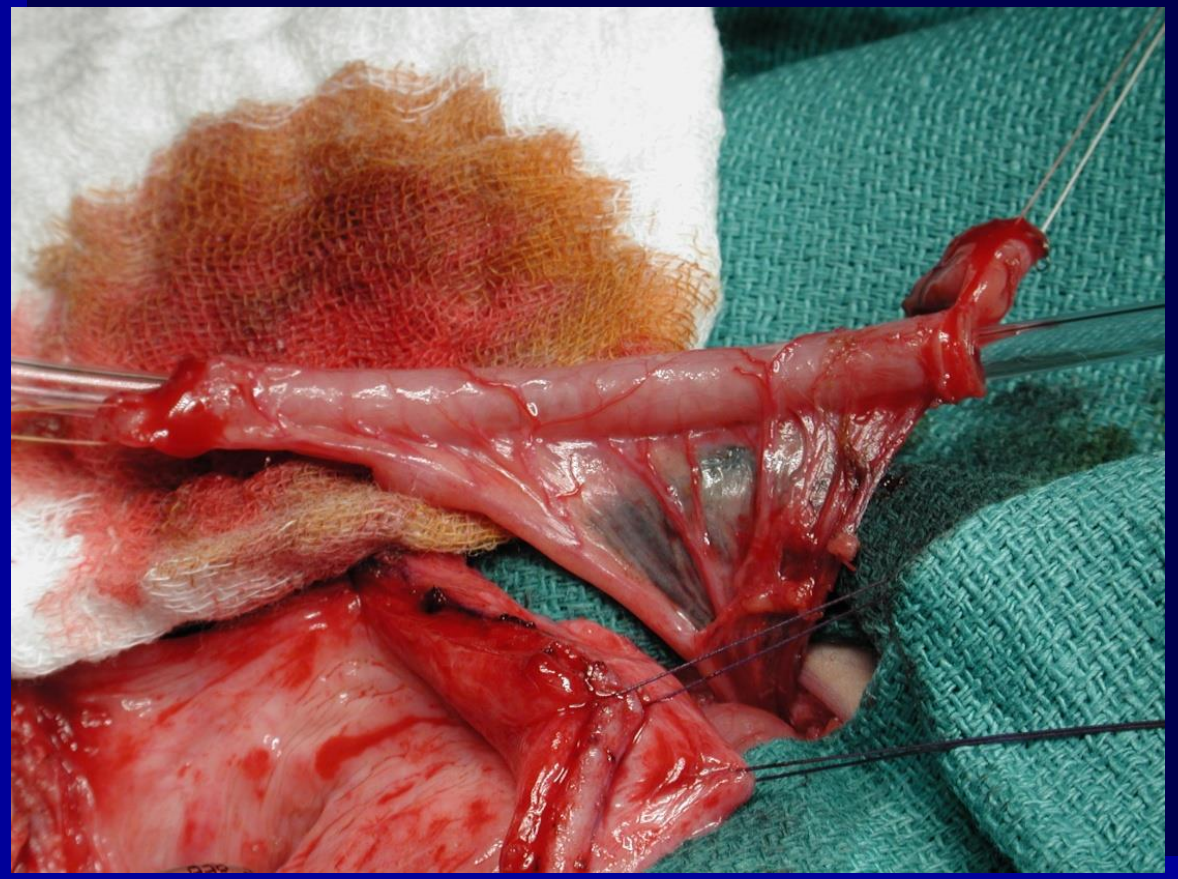
12/57 Apv (21%) F/U 31 mos

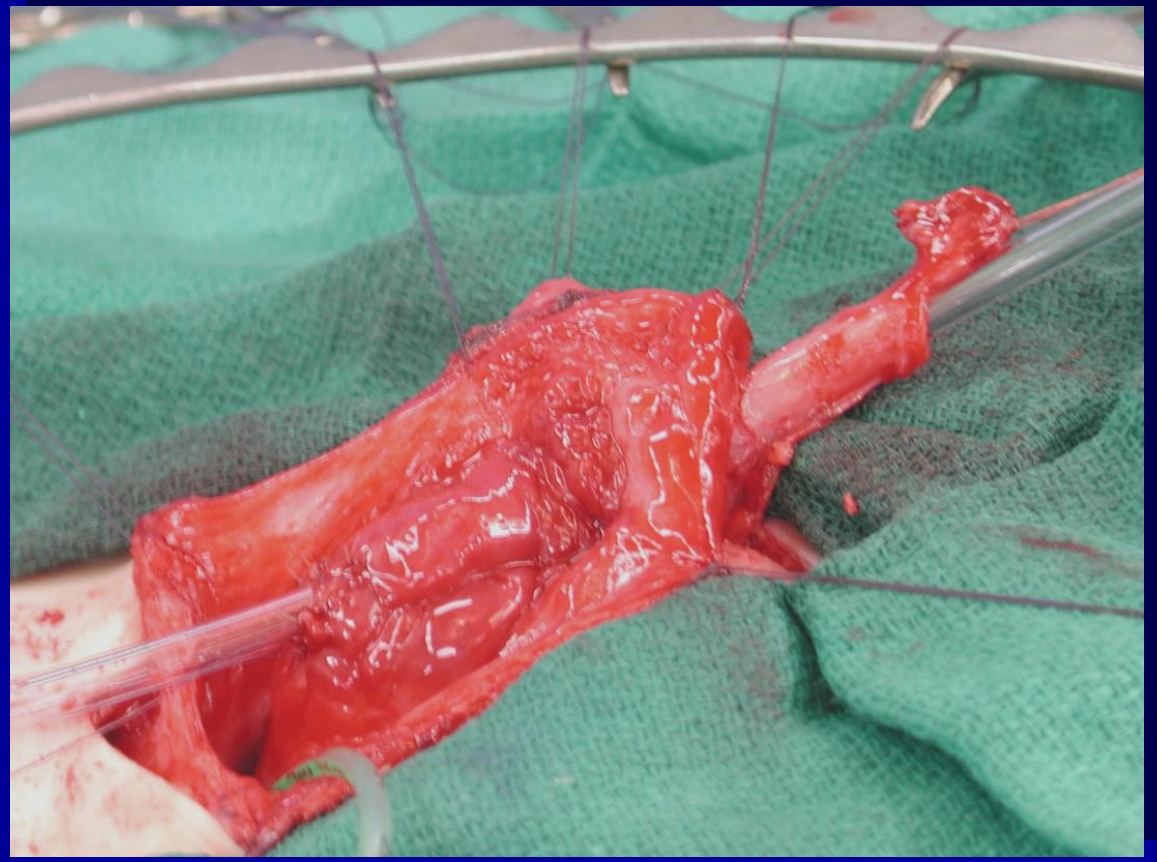
2/21 Monti (10%) F/U 9 mos

The David Bloom Principle:

"Mark, that is all very good, but show me a study with at least 100 patients and at least 10 years of continuous follow up, and then you will have made a real statement (ie become an expert)…."

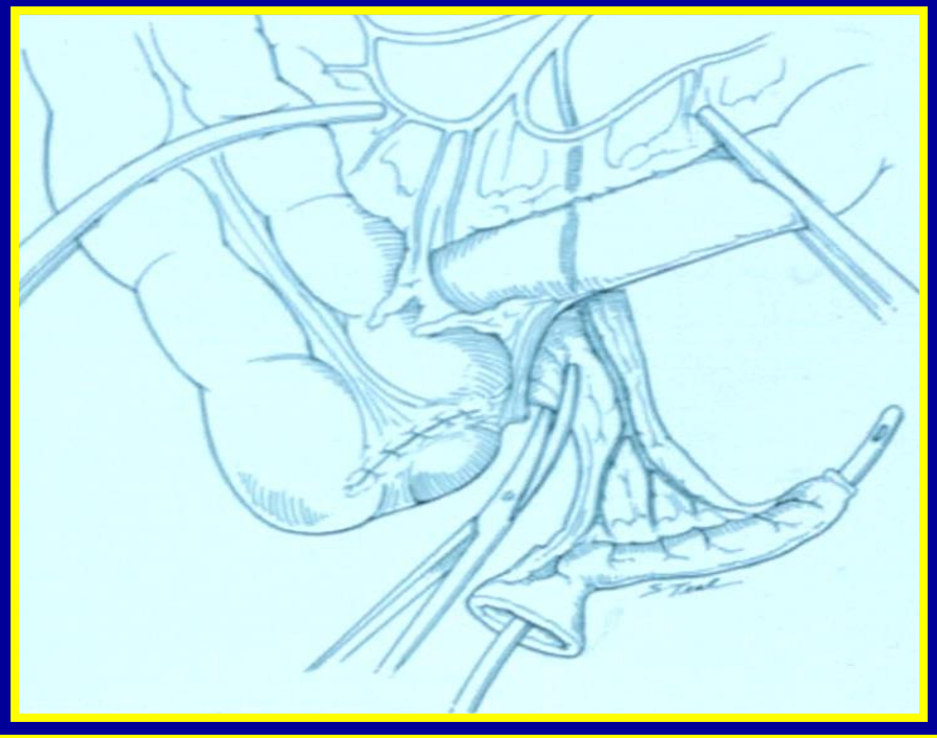
APPENDICOVESICOSTOMY





Mitrofanoff Procedure

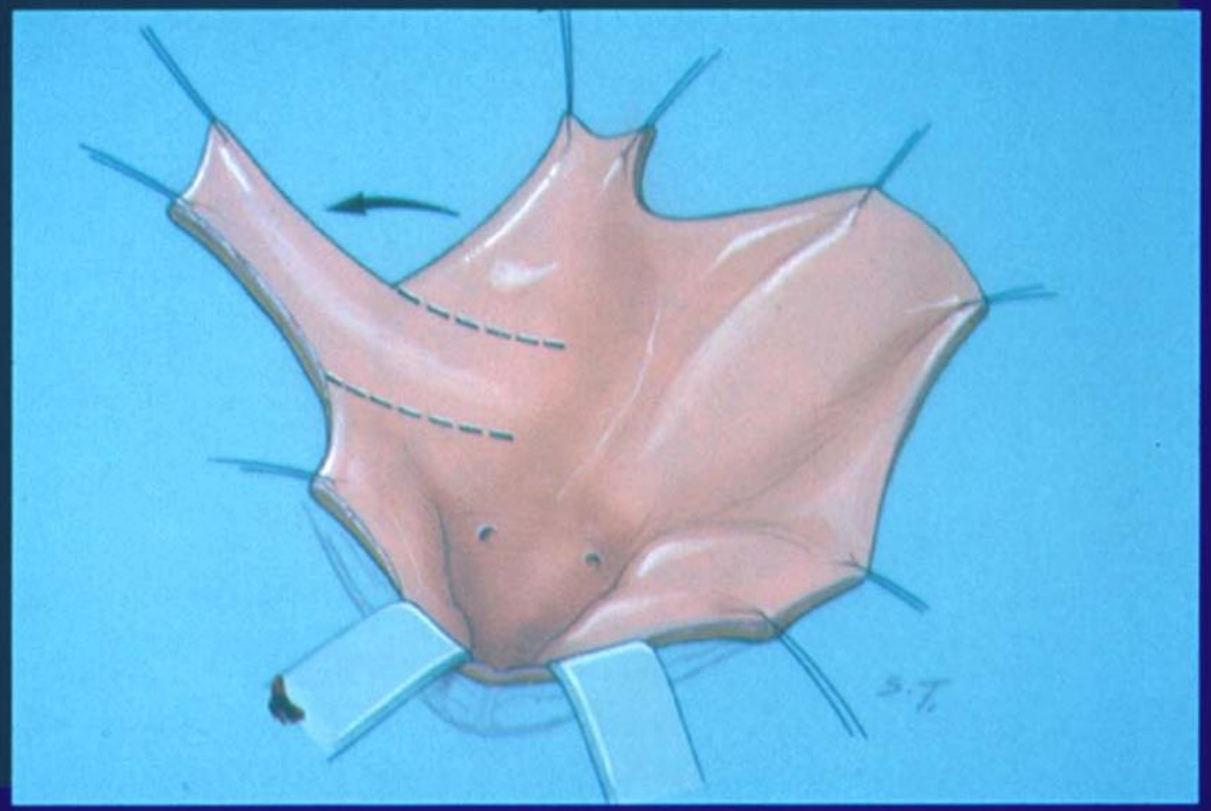
APPENDICOVESICOSTOMY

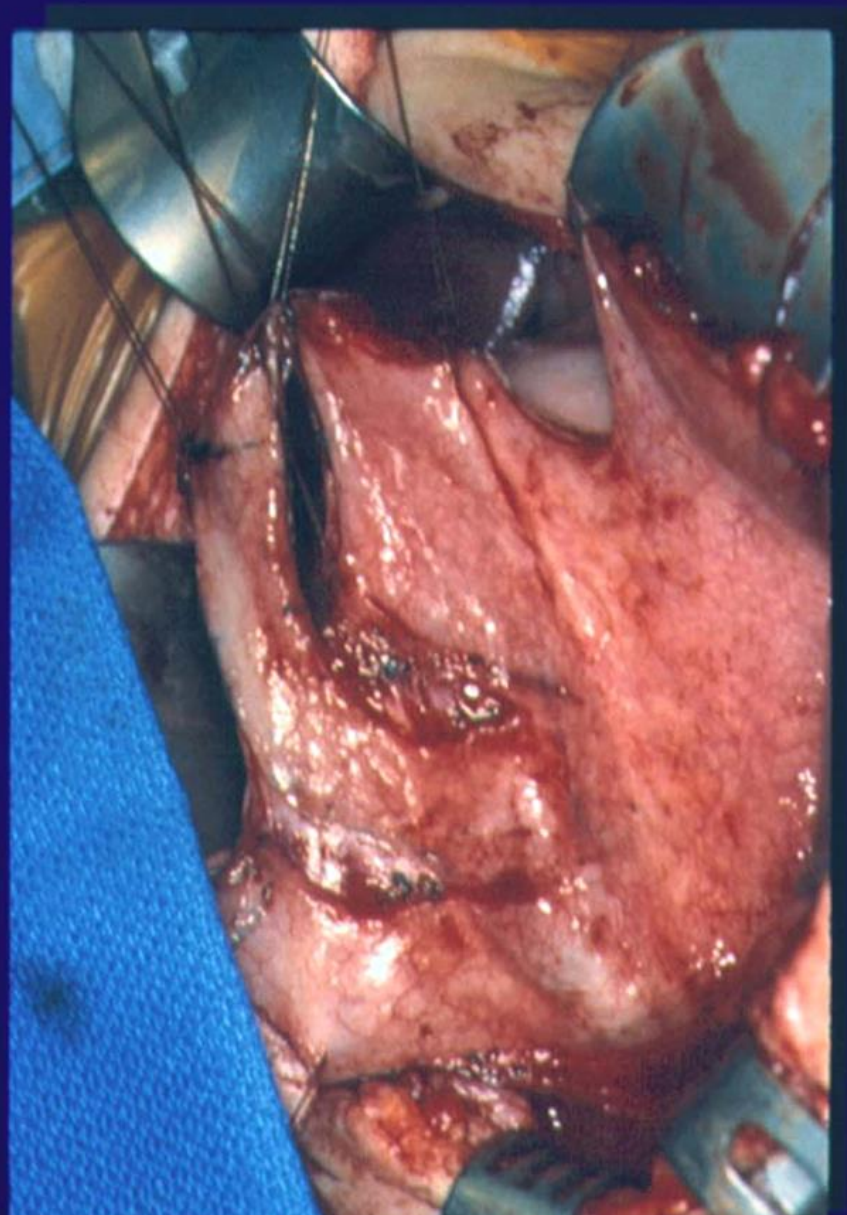


Disadvantages

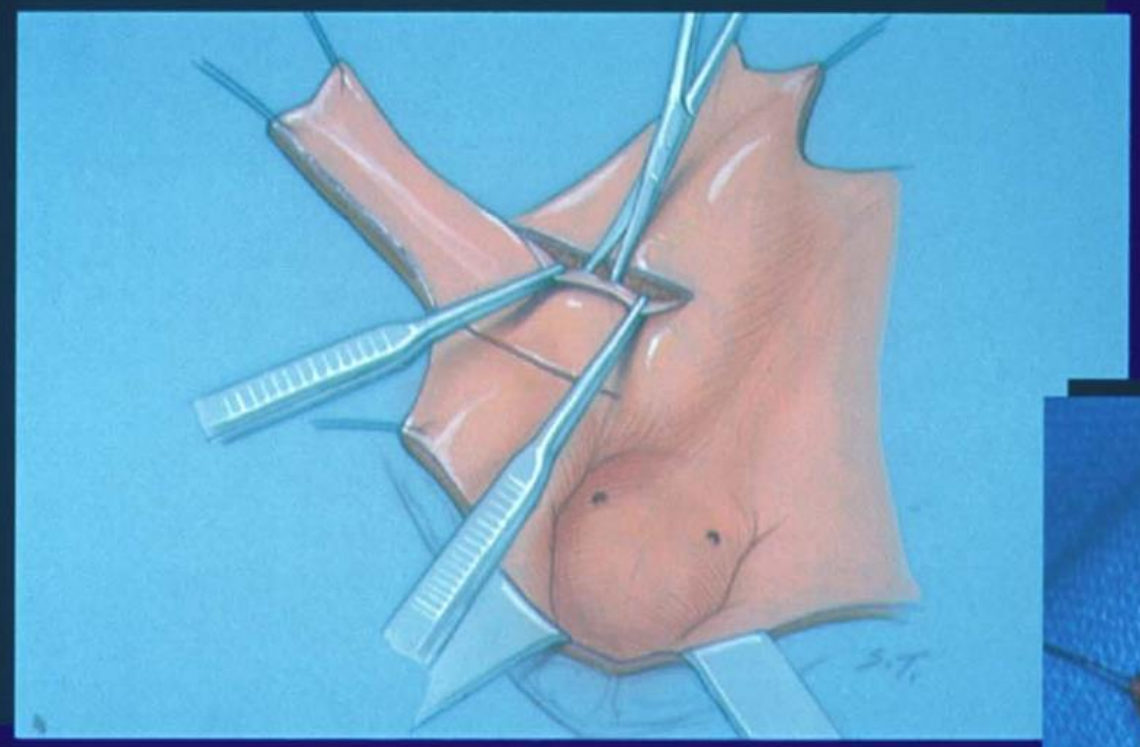
- Fatty mesentery
- Short mesentery
- Used for MACE
- Prior appendectomy
- Not useable

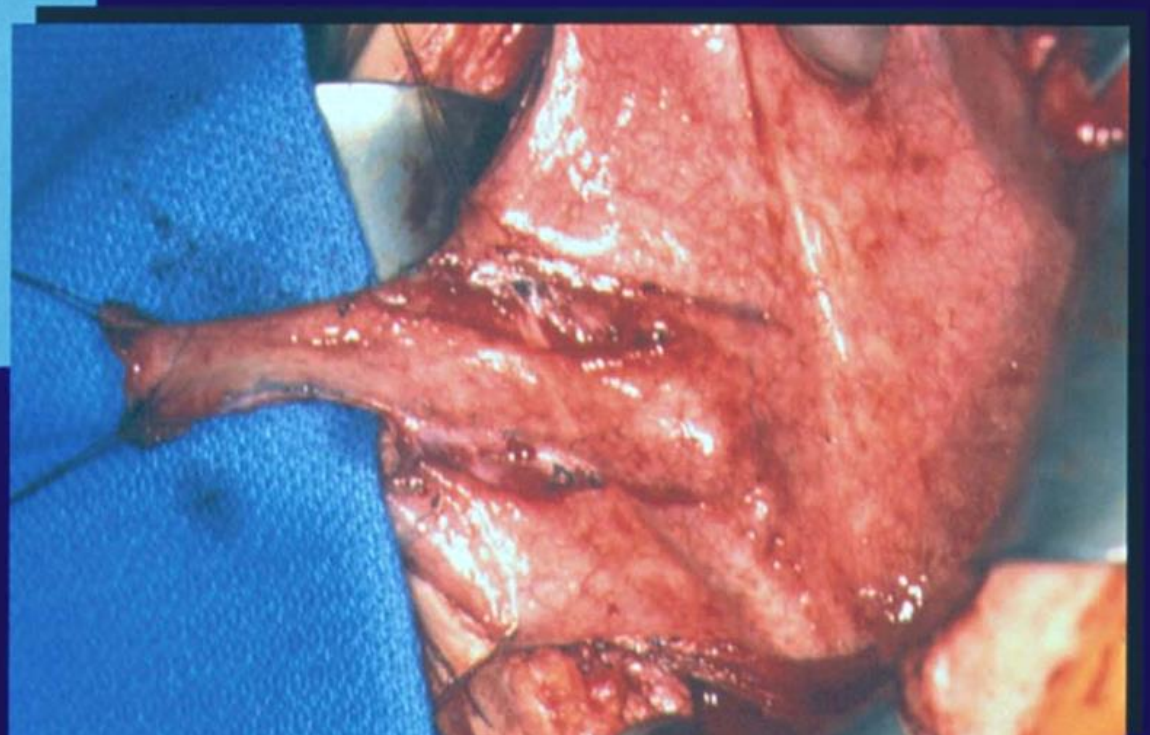
CONTINENT VESICOSTOMY



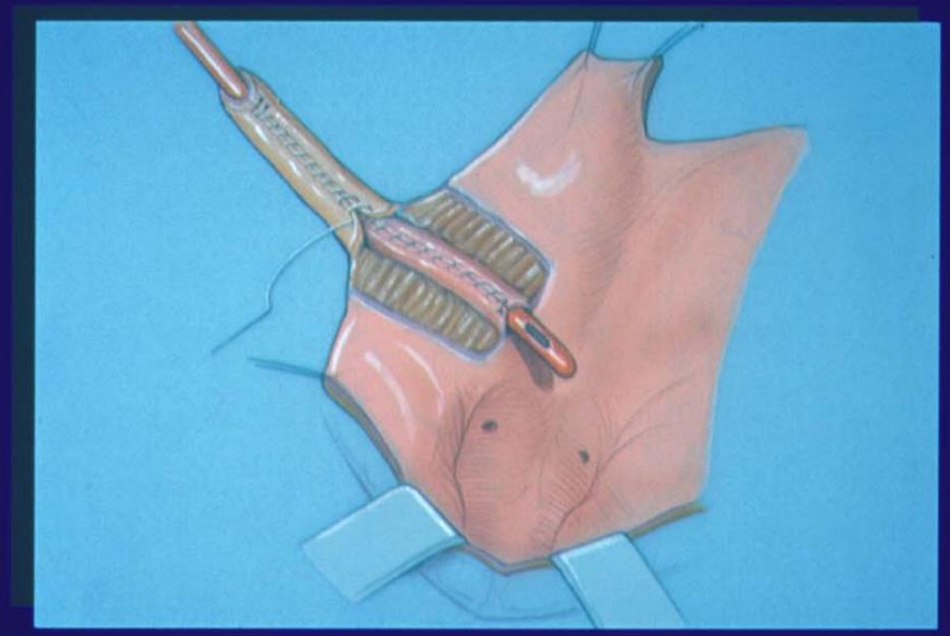


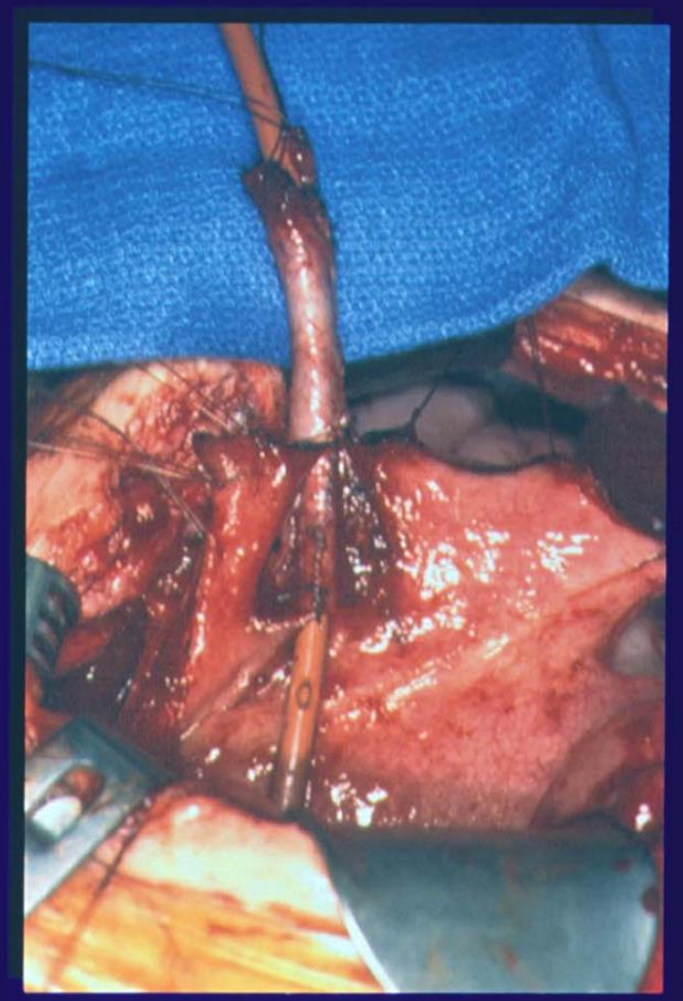
CONTINENT VESICOSTOMY





CONTINENT VESICOSTOMY





continence 100%

Stomal stenosis 45%

CONTINENT VESICOSTOMY

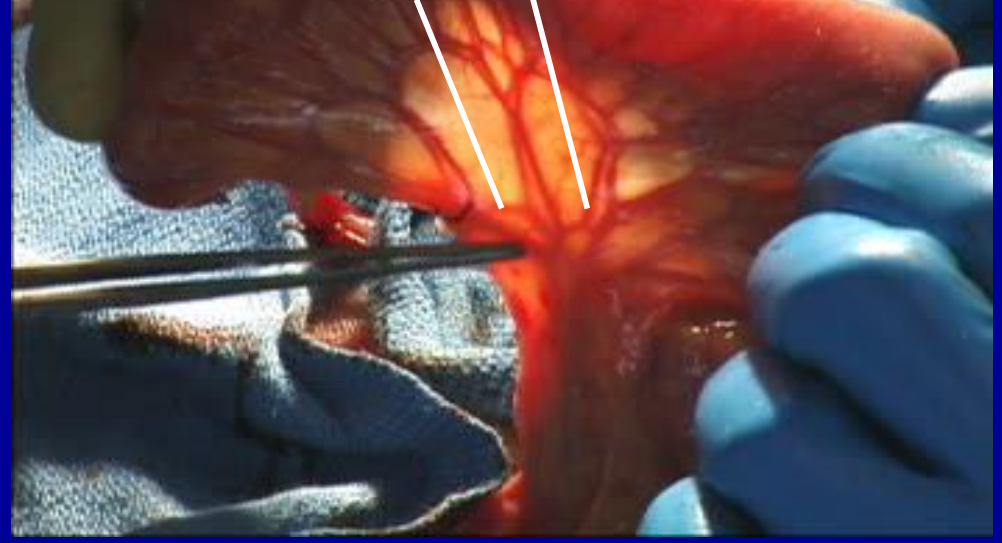
31 patients

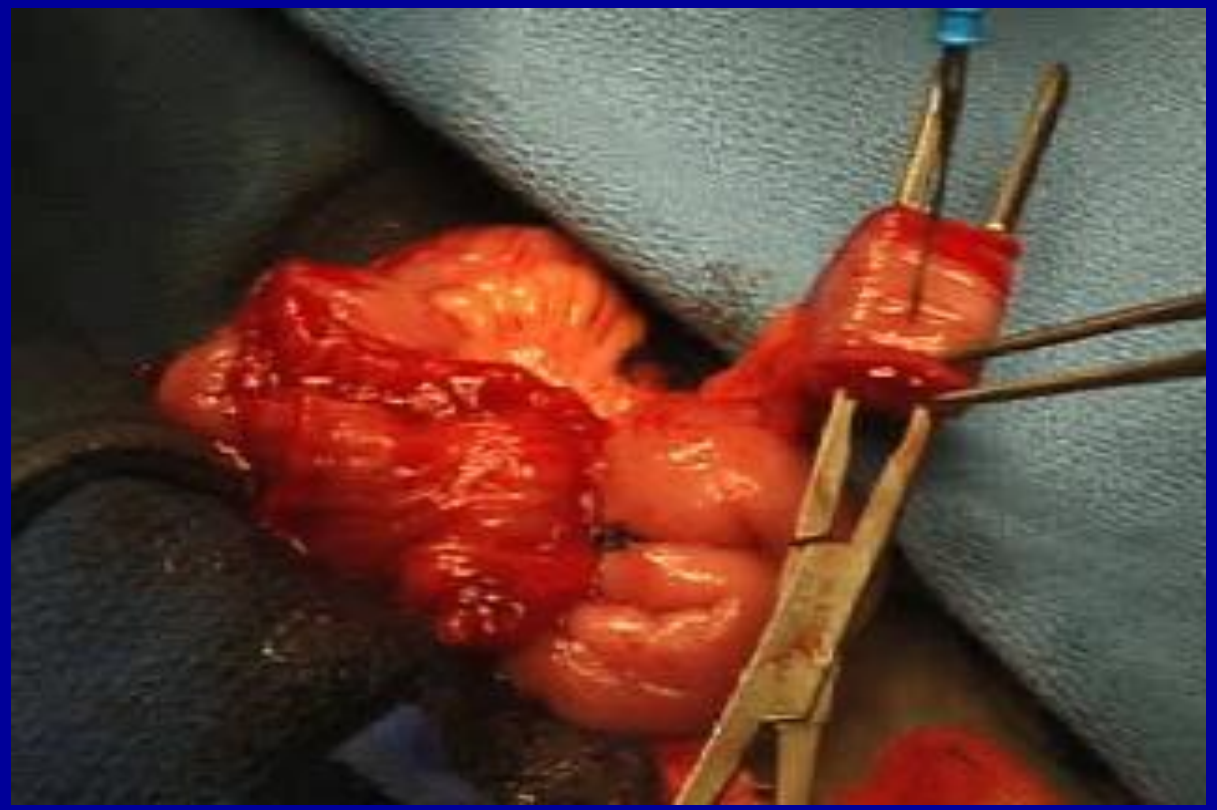
- 17 RLQ, 7 umbo., 7 neoumbo
- 31/31 (100%) continent
- 14/31 (45%) stomal stenosis
- 20/31 (65%) still using CV

Cain, Rink et al

Monti – Yang Tube

1997 -1993



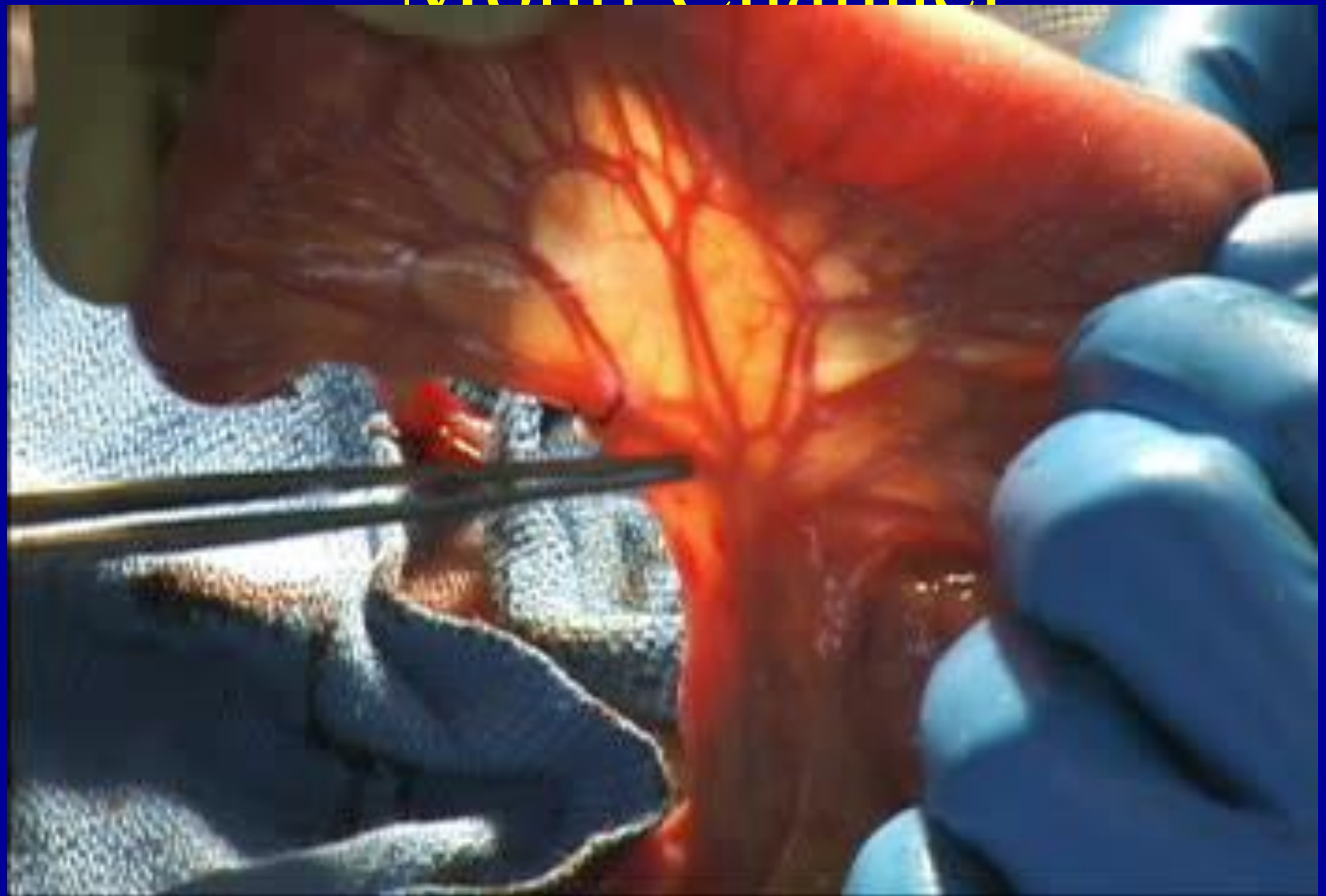


Early Data – Monti Procedure

Study	n	F/U	Continent	Stenosis
Gerharz	16	4.1	81%	n/a
Cain	21	8.5	95%	4 %
Castellan	25	13	93%	0 %
Mcandrew	21	34	n/a	24%
Narayanaswamy	25	25	88%	16%

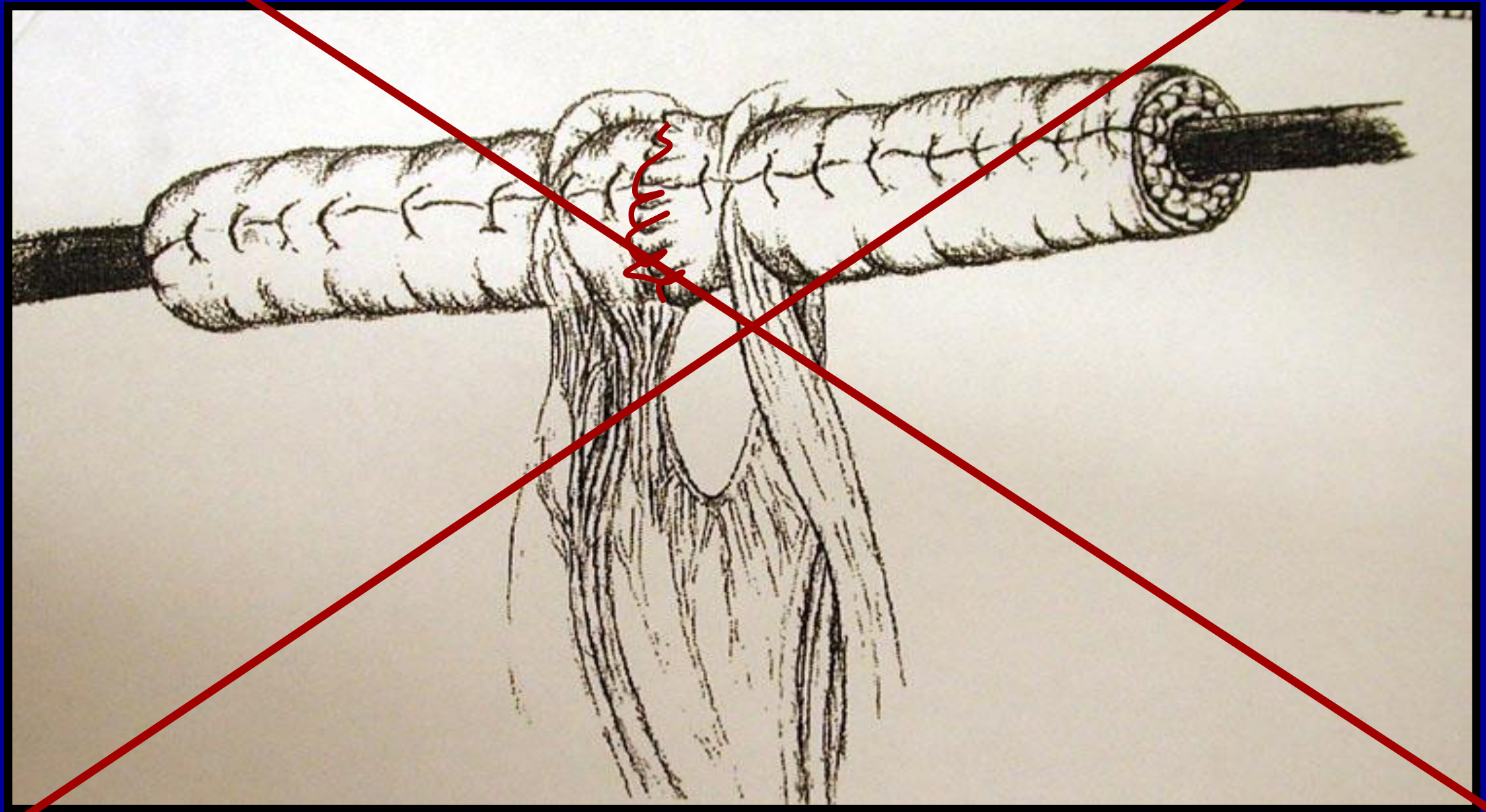
BJU J Urol Urol 1998;2001

Mitrofanoff Procedure
Monti Channel

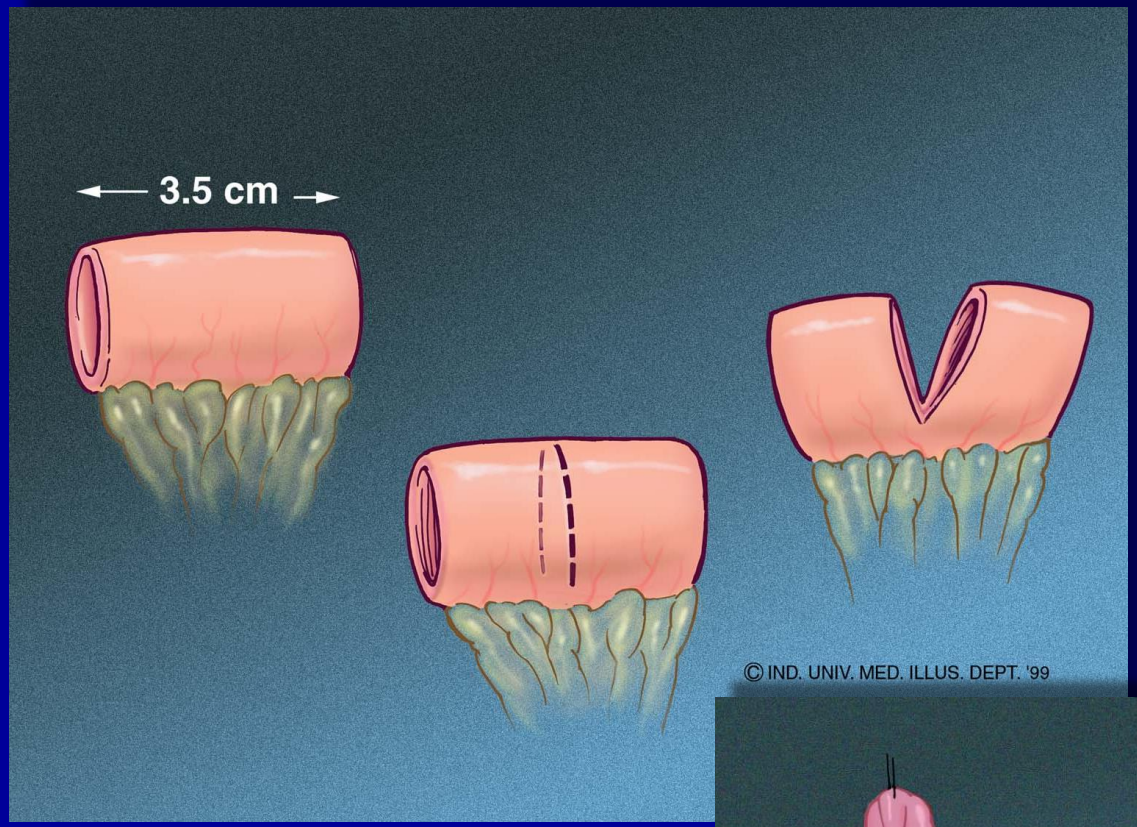


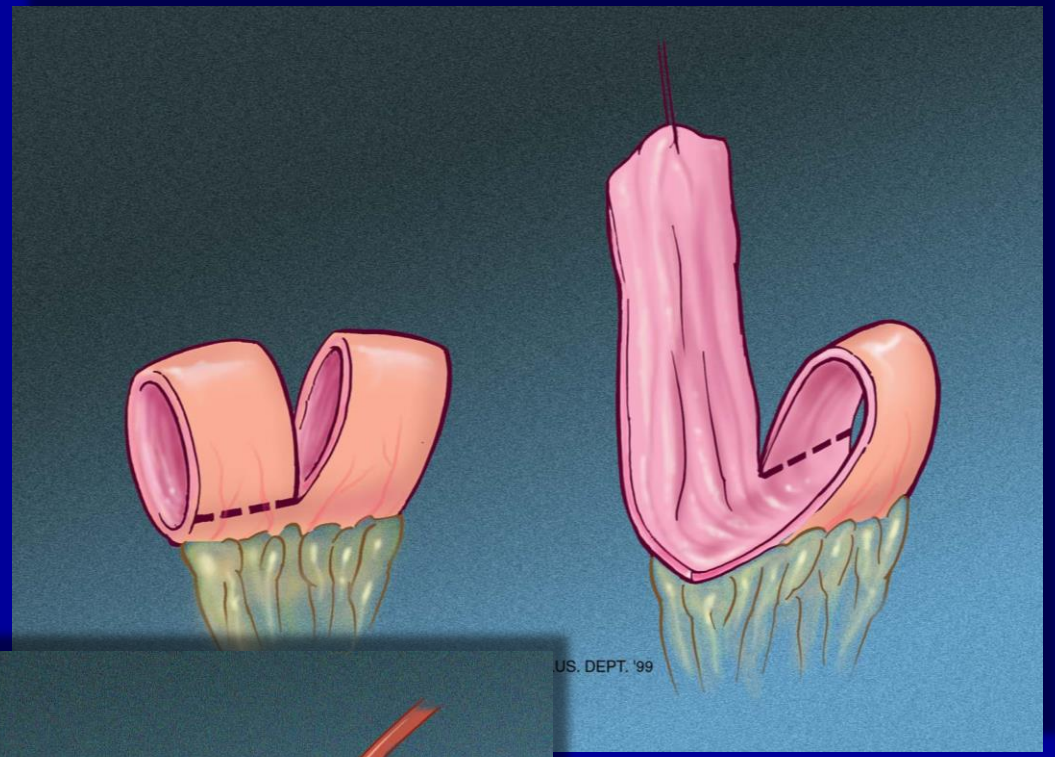
What if you need a longer channel?

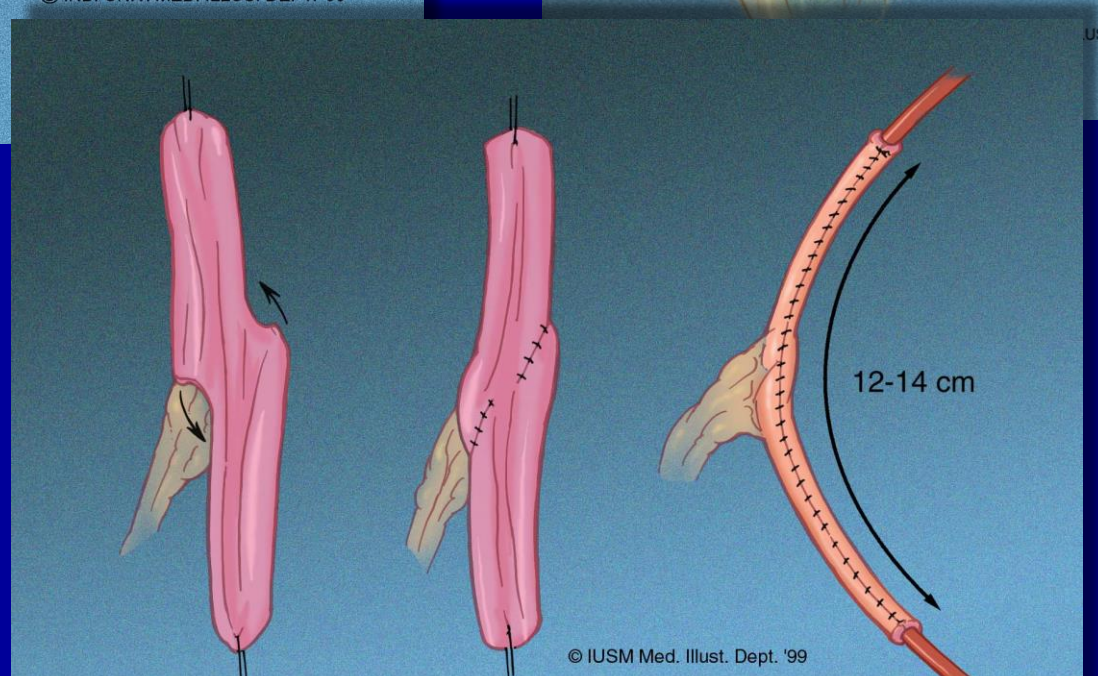
Double Monti



SPIRAL MONTY







Casale

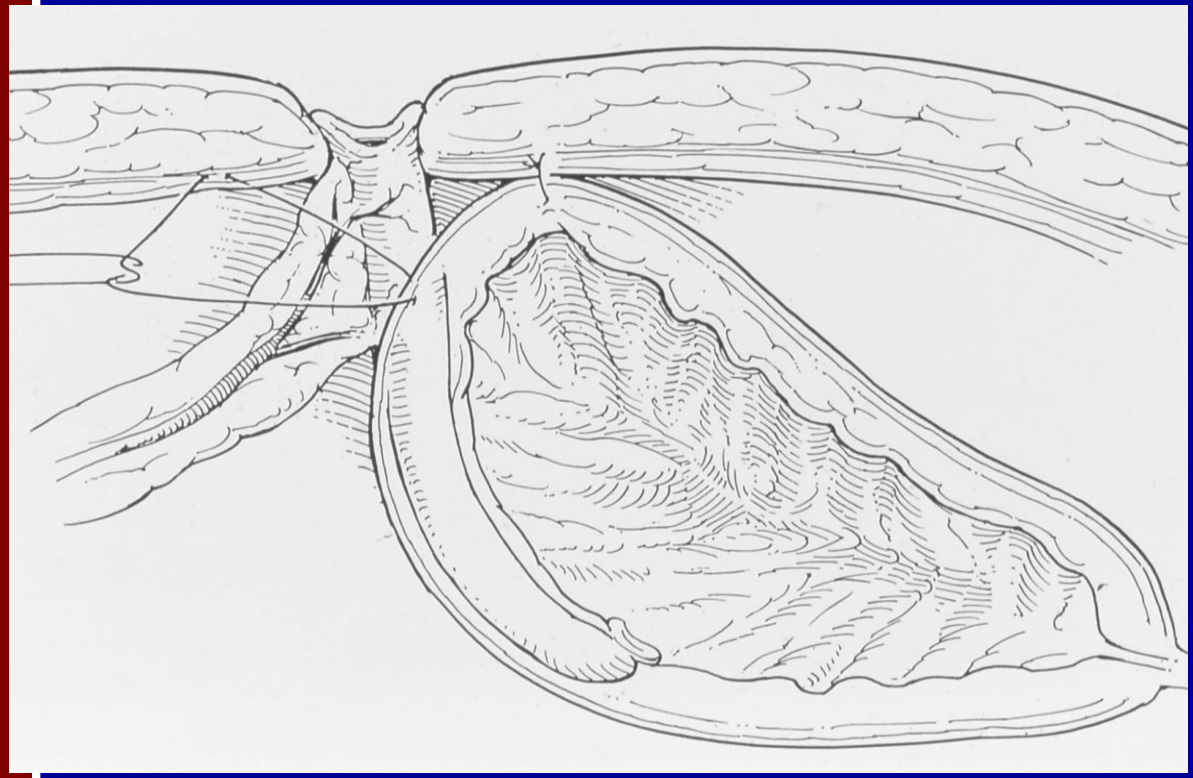
J Ural 1999

Mitrofanoff Procedure

Catheterizable Channels:

Complications

- Stomal Stenosis
- Angulation of Channel
- Leakage
- Trauma
- Obliteration
- Abscess
- Polyp



Short, Straight
Supple, Secure

CONTINENCE:

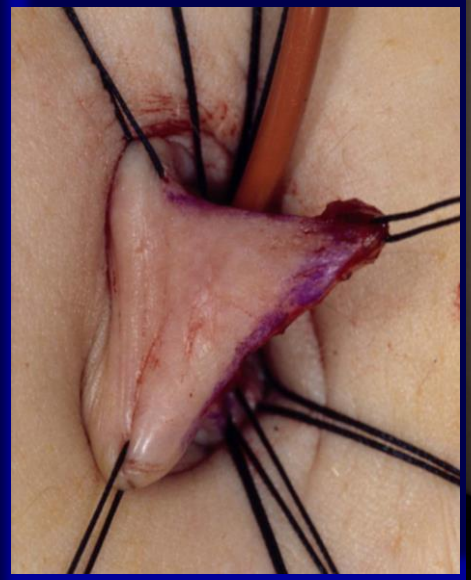
Appendicovesicostomy vs. Monti

Study	n	F/U	Continent	Stenosis
Multiple Authors (Appendix)	408	33mo	94%	14.7%
Indiana Monti	***198***	***39 mo***	***96%***	***11.6%***
Indiana Appendix	***111***	***62 mo***	***96 %***	***12%***

APV and Monti: Channel Complications
Riley Series >500 Procedures

Stomal stenosis	34 (60.7%)
Channel angulation/elongation	9 (16.1%)
Tissue overgrowth/granulation	6 (10.7%)
Prolapse	3 (5.4%)
Polyp	3 (5.4%)
Peristomal abscess	1 (1.8%)
TOTAL	56

Mitrofanoff Stoma



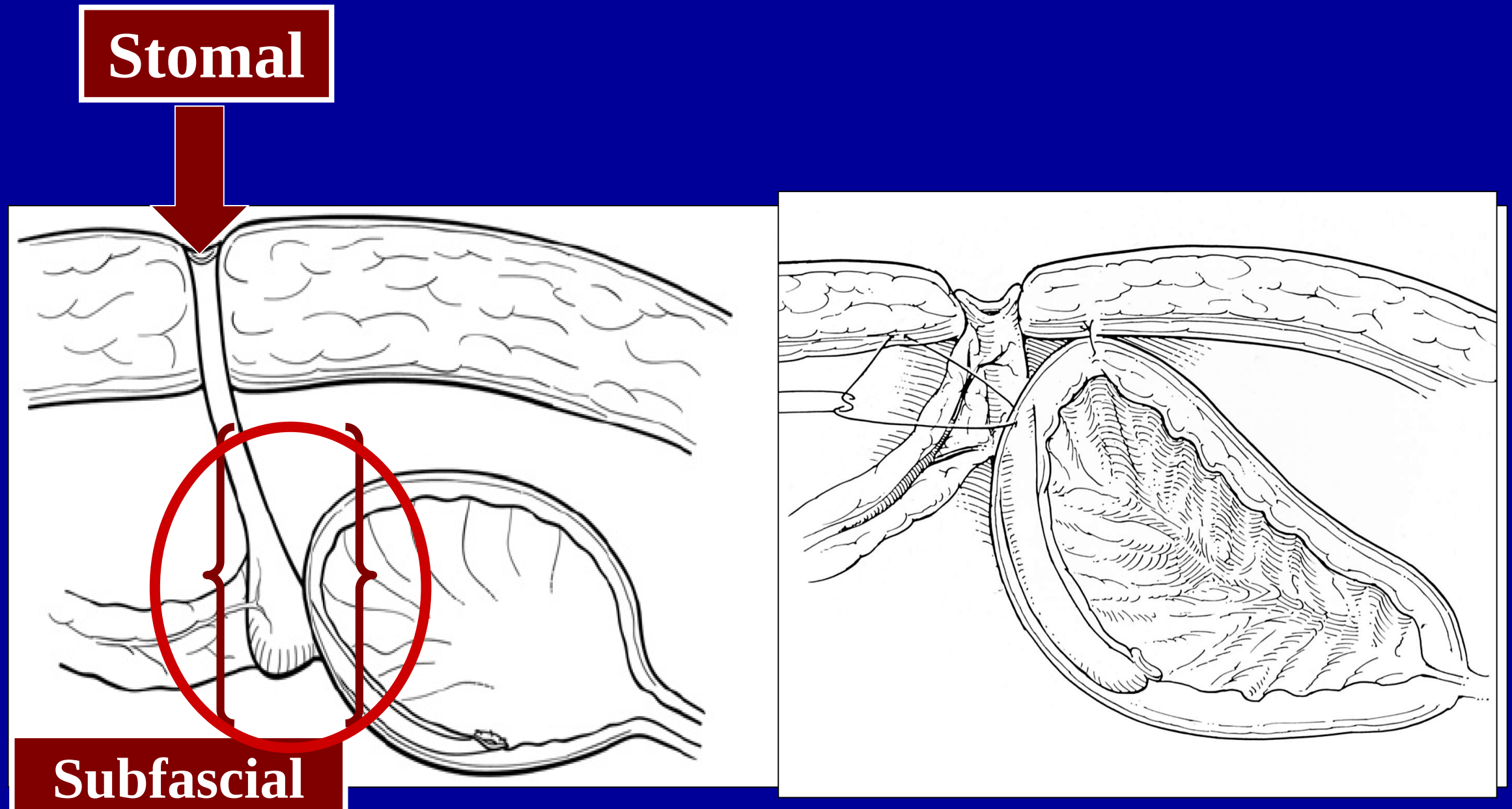
Secondary Procedures

Appendicovesicostomy vs. Monti

	Appendix	Monti
• Follow up	62 months	39 mos
• Early complic.	4%	3.5%
• Skin revision	12%	11.6%
• Bladder revision	5%	8.5%
• Tube replacement	4%	1%
• Cath prob/endoscopy	5%	5.5%
• Leakage	4%	4%

(25%) (25%)

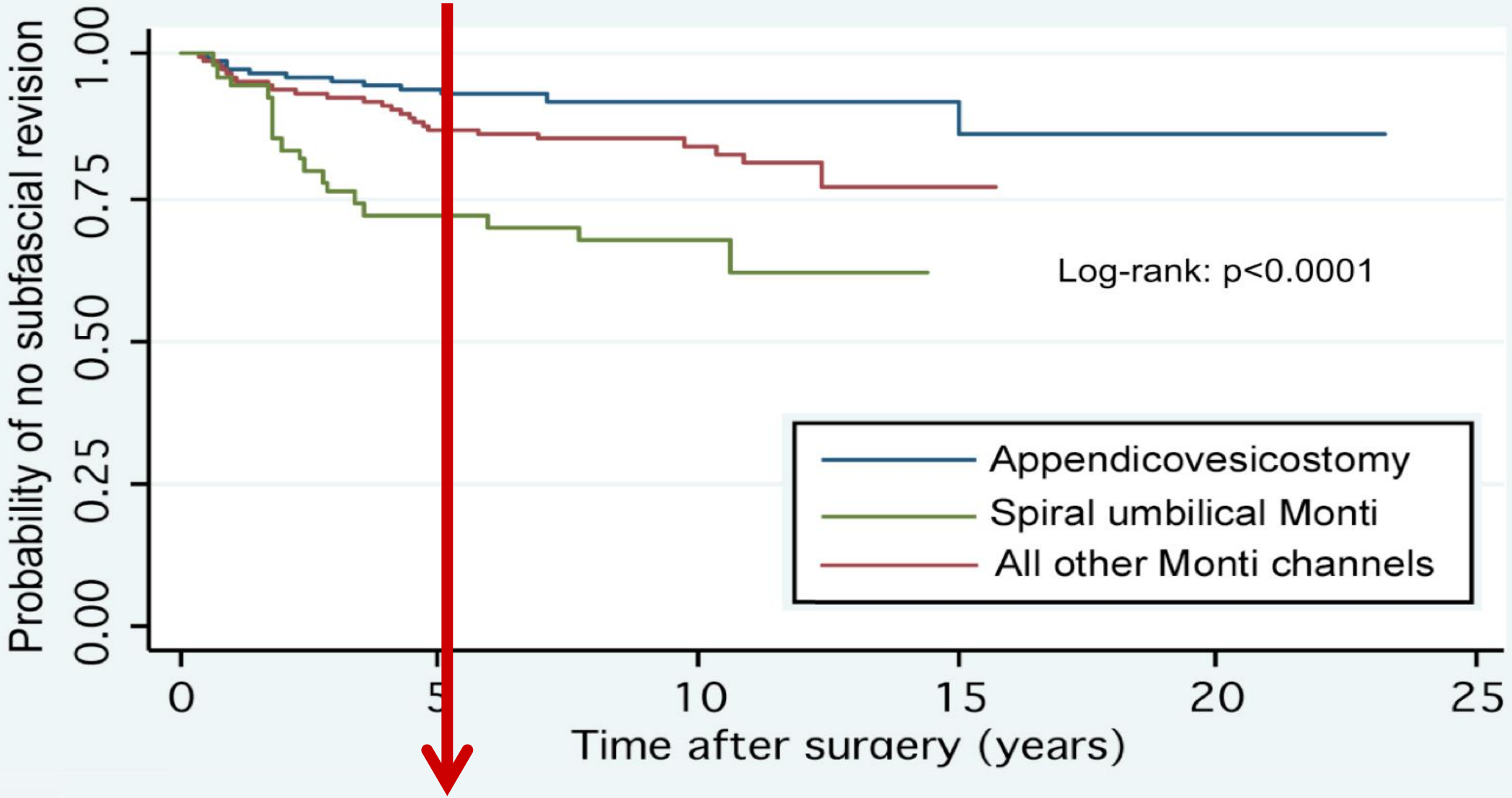
Mitrofanoff Channels



Szymanski, J Ped Urol, 2015

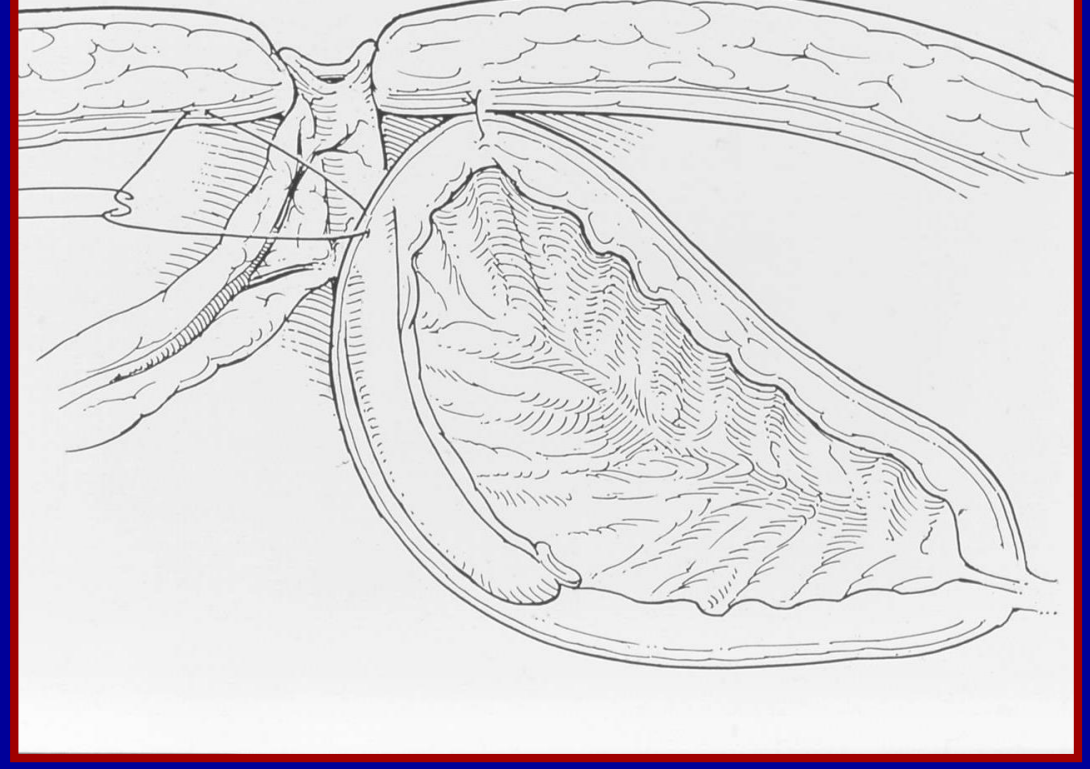
Channel type and stomal location	Number	Number of first subfascial revisions	P-value	Median follow-up (years)
APV	215			
Non-umbilical	118	6 (5.1%)	reference	5.4
Umbilical	97	8 (8.3%)	0.41	6.2
Monti				
Traditional	146			
Non-umbilical	96	14 (14.6%)	0.03	8.4
Umbilical	50	6 (12.0%)	0.19	9.9
Spiral	150			
Non-umbilical	94	11 (11.7%)	0.13	4.8

Subfascial Revisions

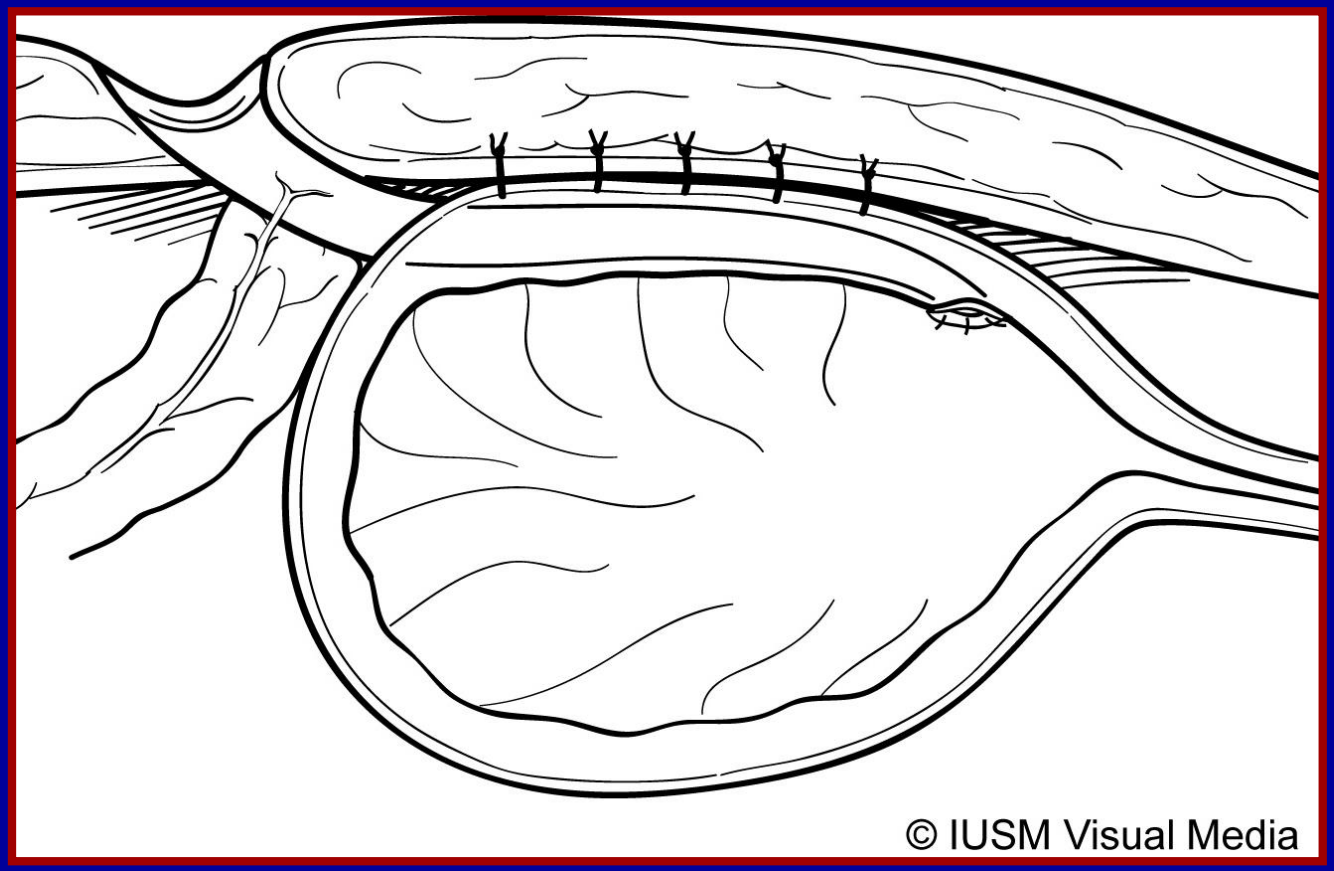


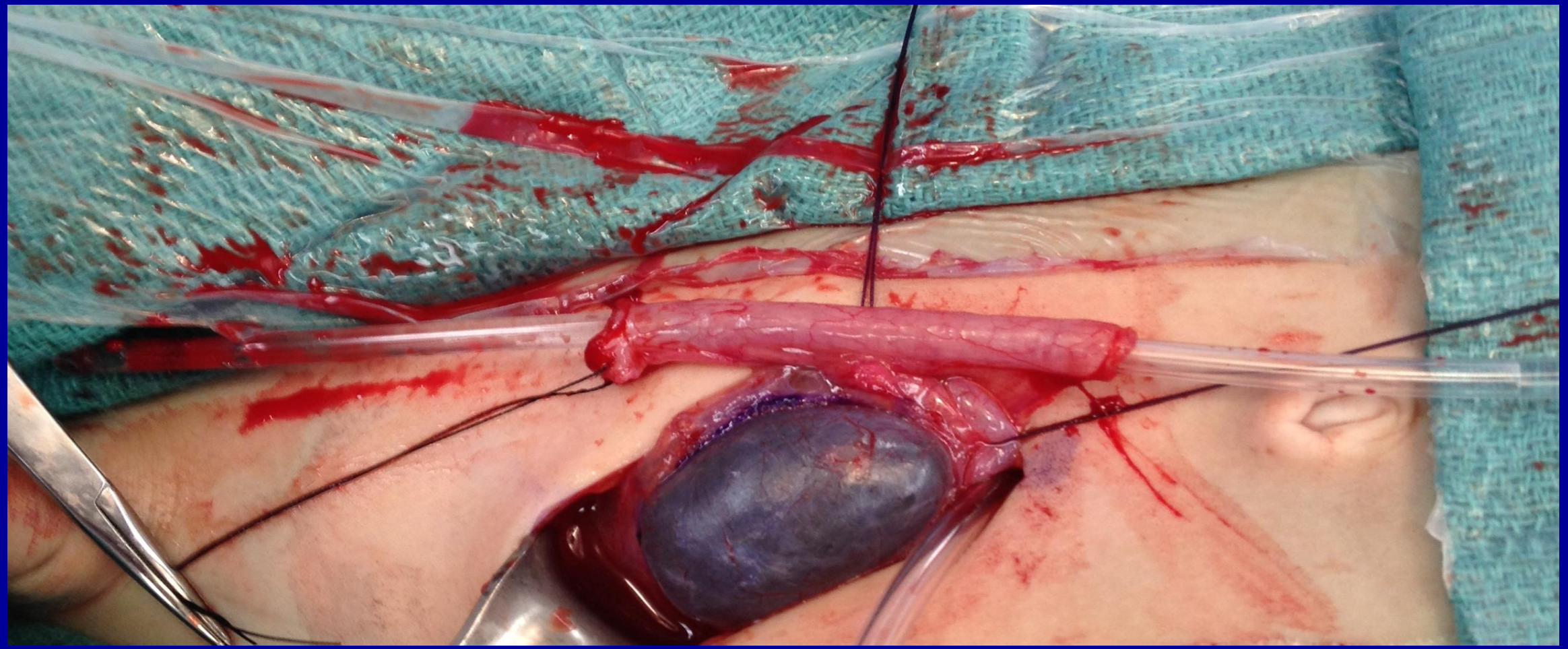
Number at risk:

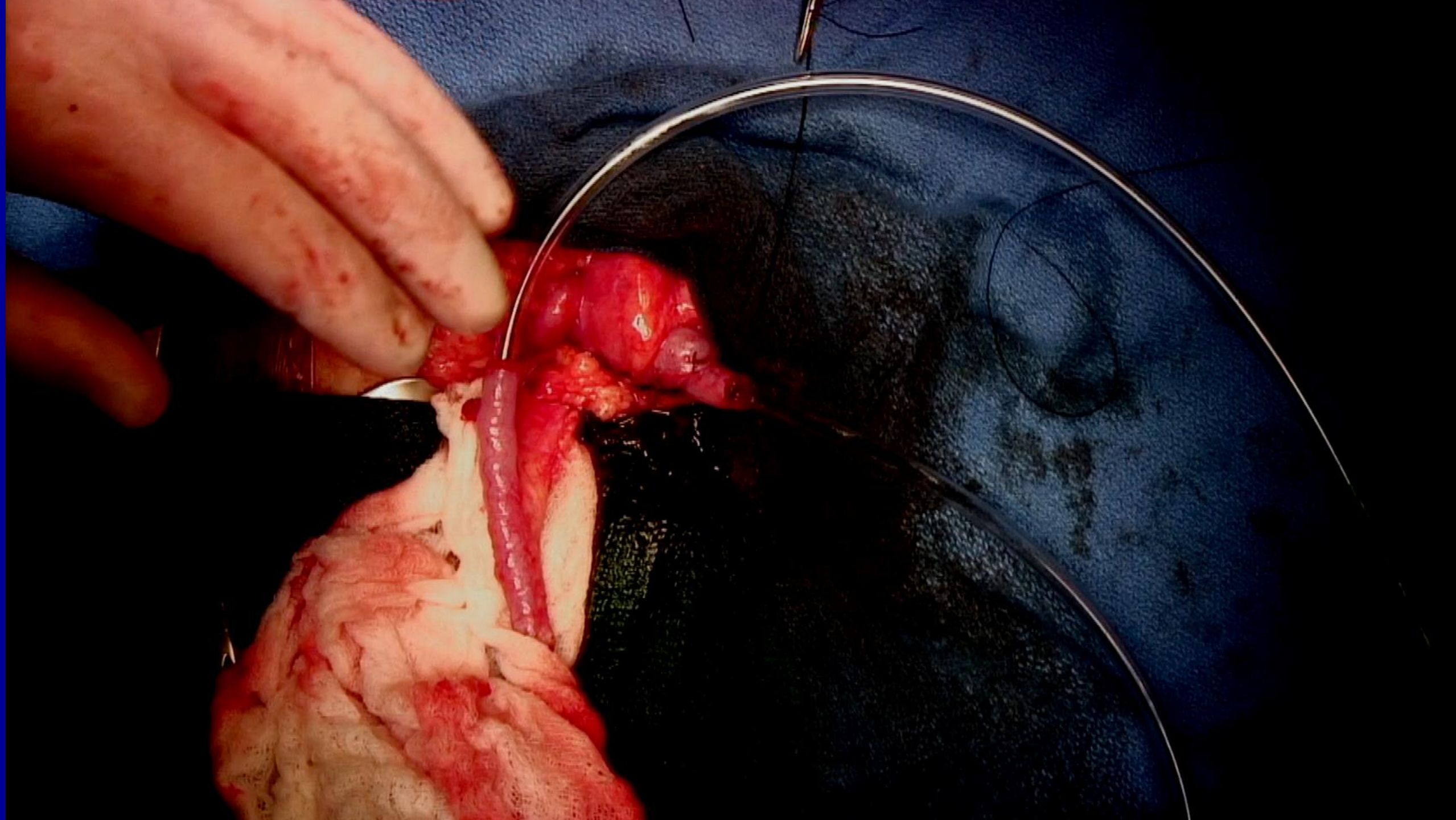
Appendicovesicotomy	215	116	41	17	7	0
Spiral umbilical Monti	56	36	16	0	0	0



Current technique







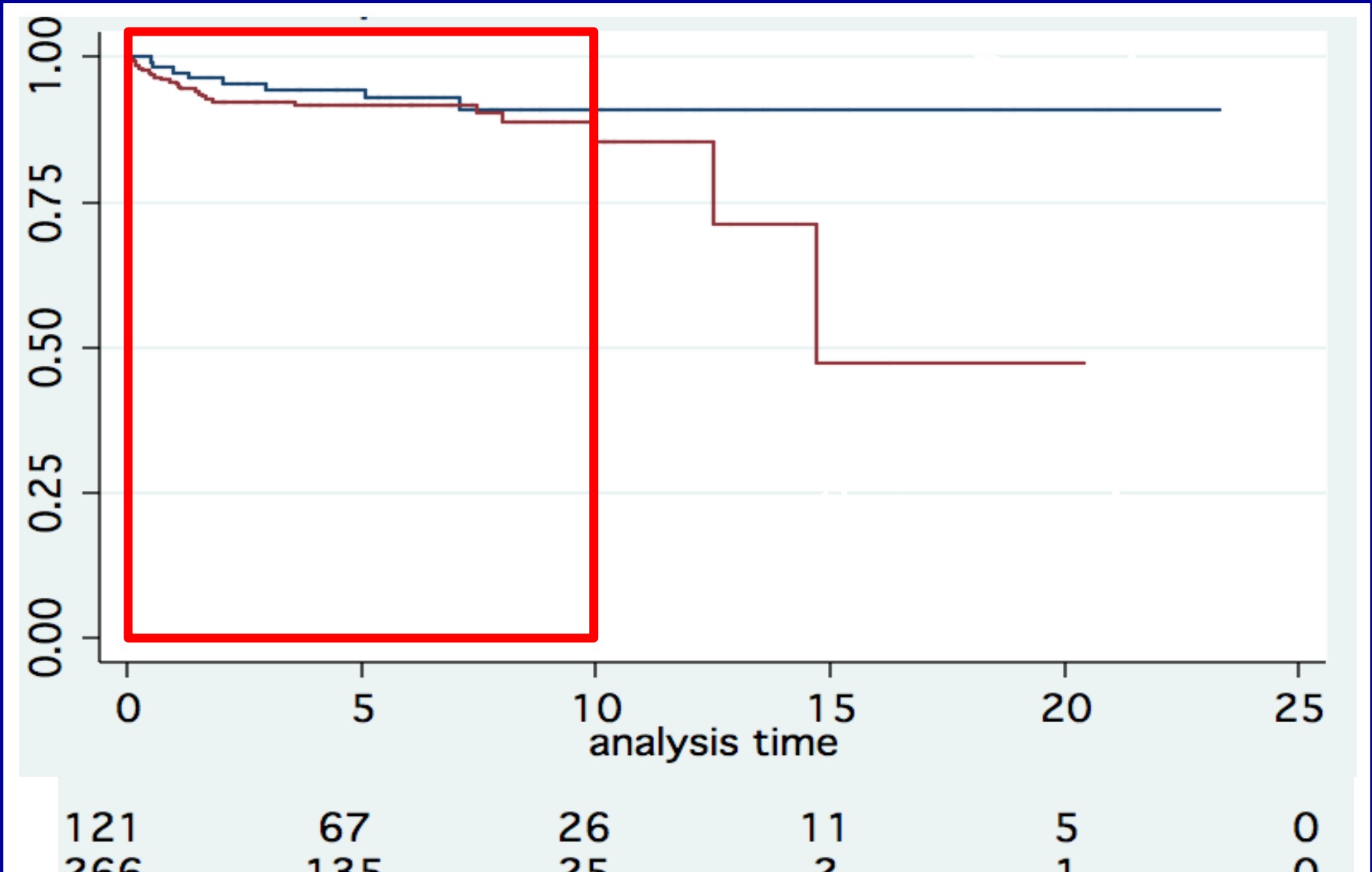
3 Institutions:
675 urinary channels
Does Anterior Position Matter?

Riley (458), Argentina (178), Chile (39)

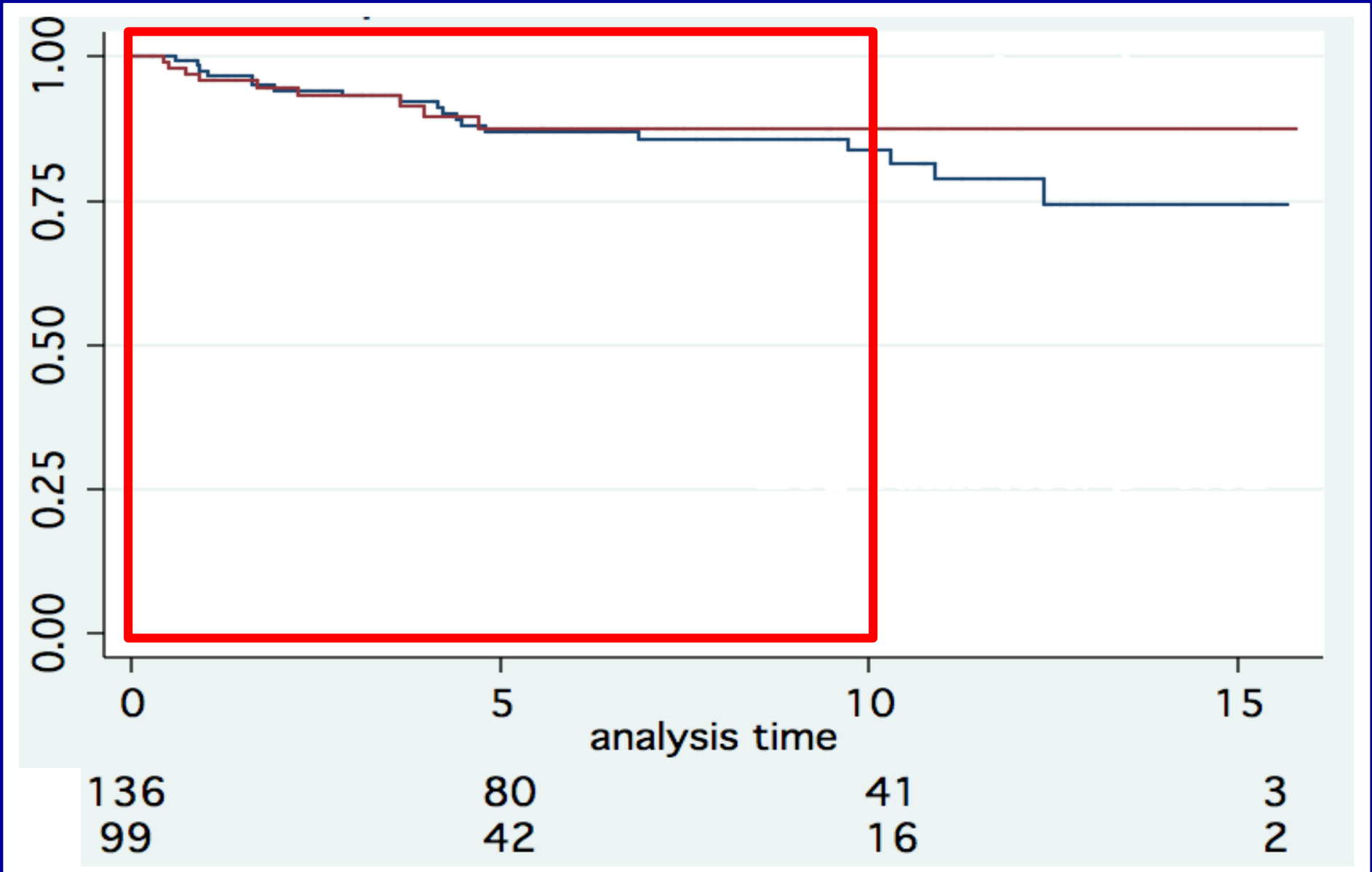
Male 53.8%
VPS 49.7%

Median age at surgery: 8.8 years old
Median follow-up: 6.0 years
***results reliable up to 10 years**

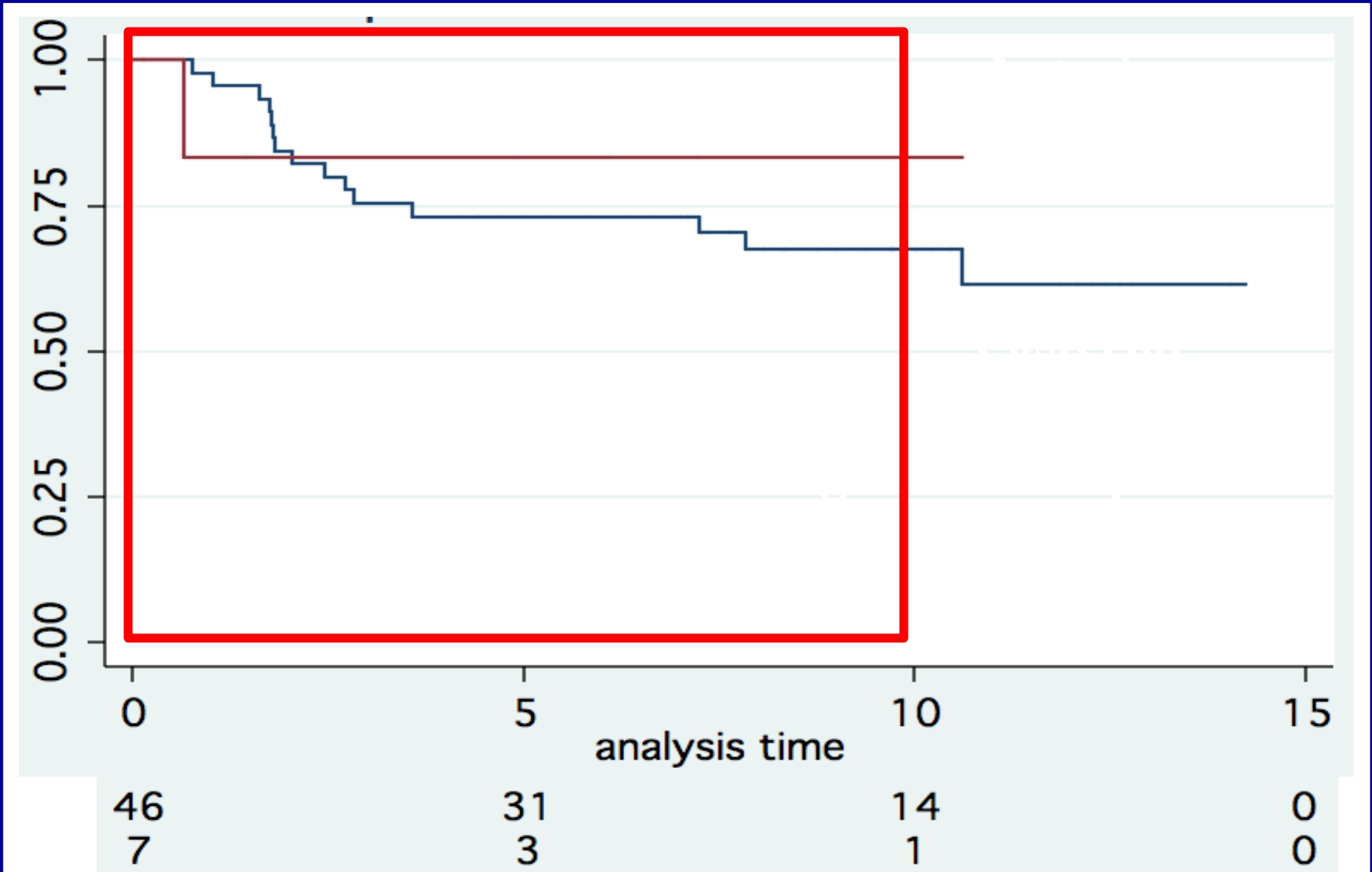
Appendicovesicostomy (n=387)



Traditional Monti (n=235)



Spiral umbilical Monti (n=53)



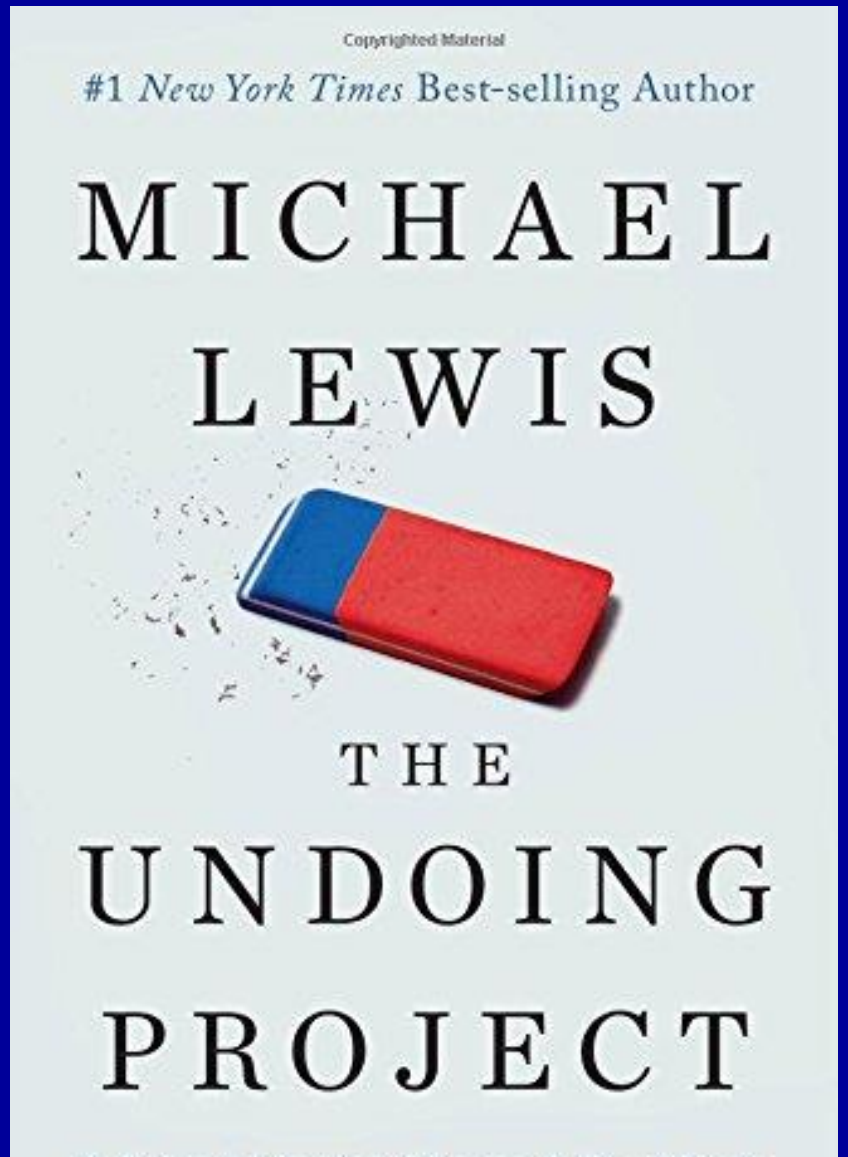
Take Home Message:
Open Mitrofanoof

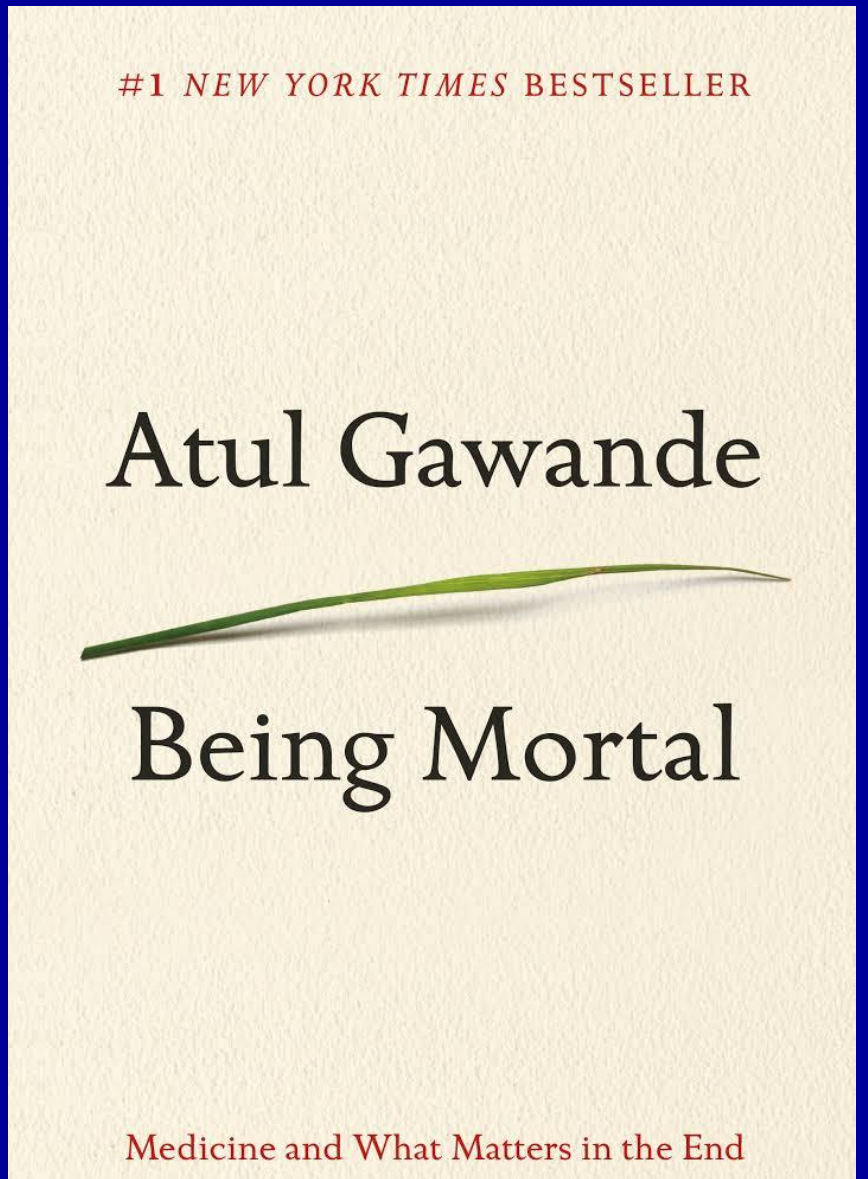
- No difference for anterior vs posterior Mitrofanoff channel with open technique
- Continence rate 95-96%, with ”good” bladder
- Revision rate approximately 25%
- Anterior placed channel using minimally invasive technique should duplicate open experience
  - IN EXPERIENCED HANDS

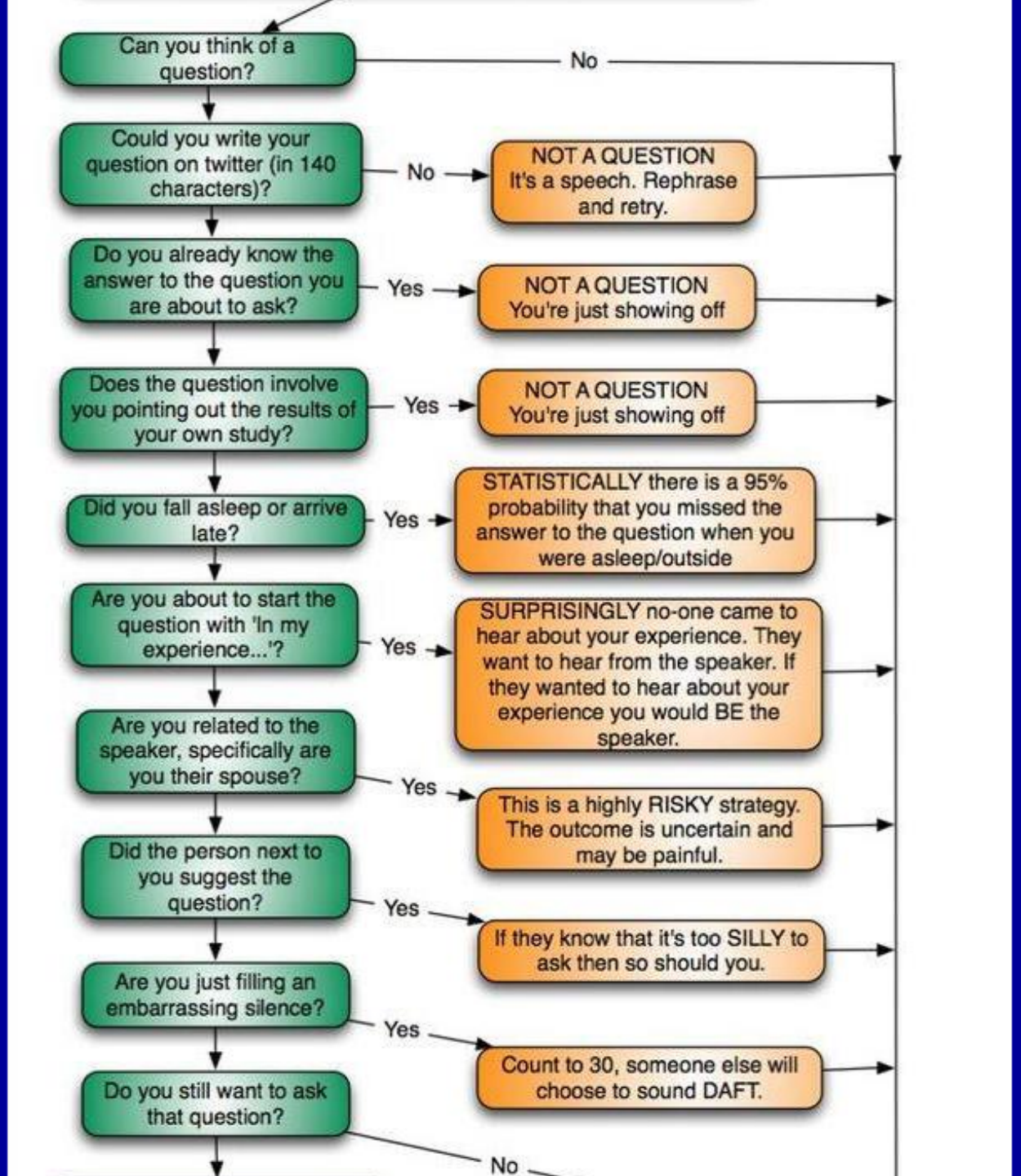
What you need to remember:

- Yearly follow up
  - BMP, CBC, B12, KUB, RBUS
- Prompt evaluation for gross hematuria, acute abdominal pain
  - Cystoscopy, CT cystogram
- Catheterization problems are an emergency
- Pregnancy may require your presence
- Involve your pediatric urologist

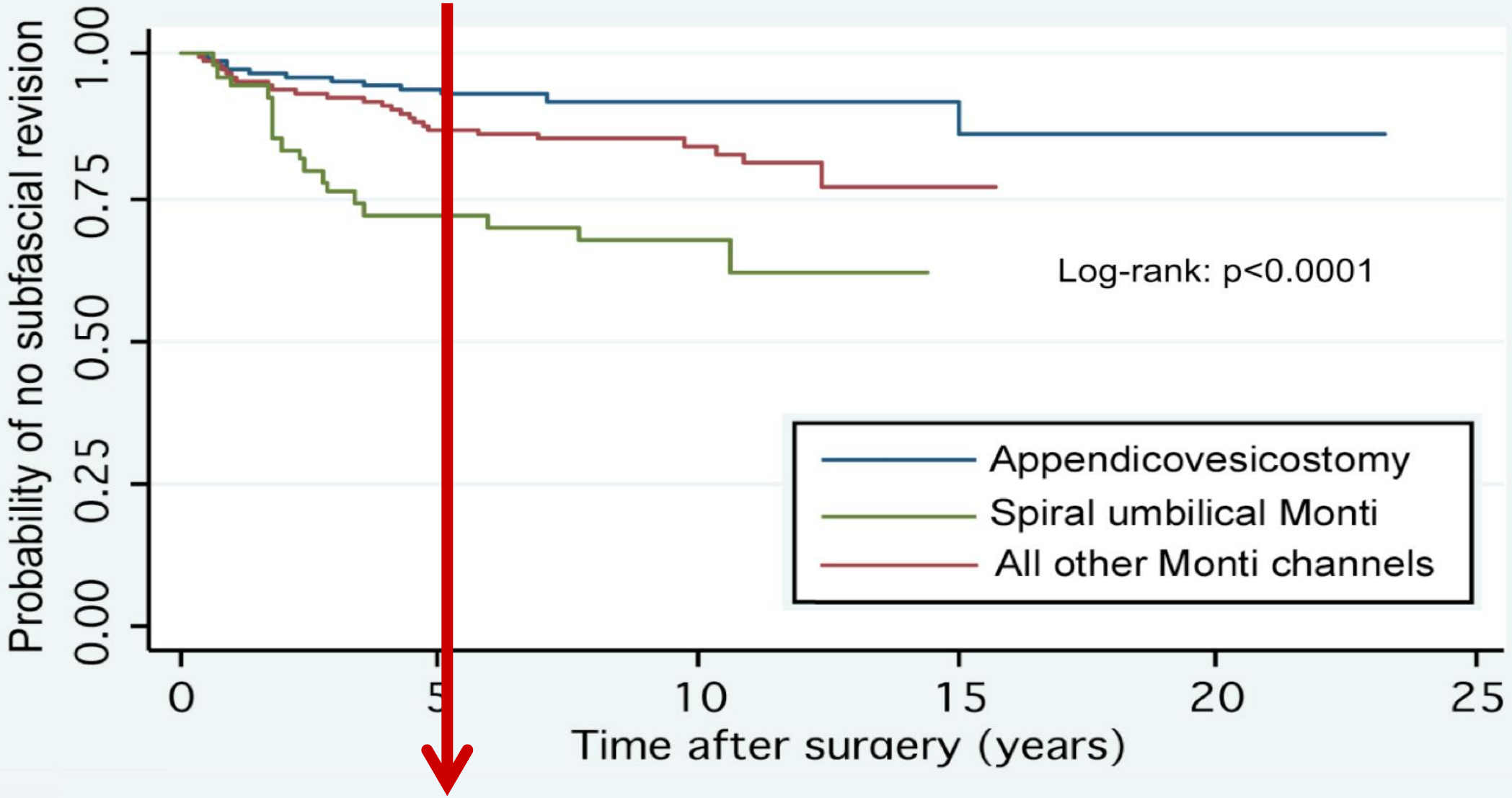
If you learn nothing else from this lecture, read these books..






Can you think of a question?
No
Could you write your question on twitter (in 140 characters)?
No
NOT A QUESTION
It's a speech. Rephrase and retry.
Do you already know the answer to the question you are about to ask?
Yes
NOT A QUESTION
You're just showing off
Does the question involve you pointing out the results of your own study?
Yes
NOT A QUESTION
You're just showing off
Did you fall asleep or arrive late?
Yes
STATISTICALLY there is a 95% probability that you missed the answer to the question when you were asleep/outside
Are you about to start the question with 'In my experience...'?
Yes
SURPRISINGLY no-one came to hear about your experience. They want to hear from the speaker. If they wanted to hear about your experience you would BE the speaker.
Are you related to the speaker, specifically are you their spouse?
Yes
This is a highly RISKY strategy. The outcome is uncertain and may be painful.
Did the person next to you suggest the question?
Yes
If they know that it's too SILLY to ask then so should you.
Are you just filling an embarrassing silence?
Yes
Count to 30, someone else will choose to sound DAFT.
Do you still want to ask that question?
No

Subfascial Revisions



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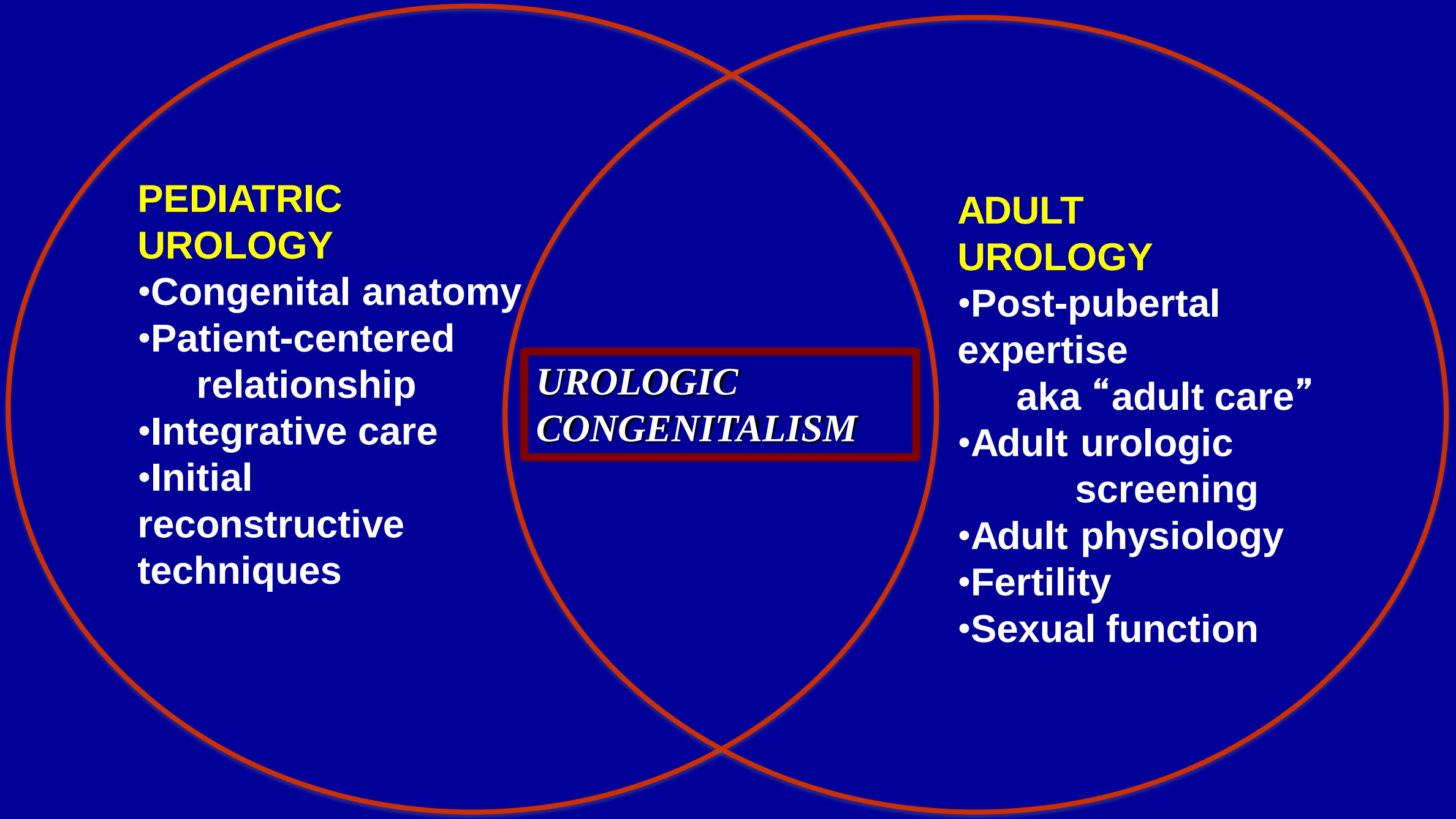
PEDIATRIC UROLOGY
•Congenital anatomy
•Patient-centered relationship
•Integrative care
•Initial reconstructive techniques
UROLOGIC CONGENITALISM
ADULT UROLOGY
•Post-pubertal expertise
aka “adult care”
•Adult urologic screening
•Adult physiology
•Fertility
•Sexual function