Anal canal and sphincter function in children with Hirschsprung disease after definitive surgery

- Dr Tan Wei Sheng

- 4 June 2022

- MAPS 2022 Meeting

Introduction

Hirschsprung disease (HD)

- Most common bowel dysfunction following surgery for HD are **faecal incontinence** and **constipation**. [1-4]
- Reported rate of
  - Incontinence : **27 - 48%**
  - Constipation : **5 - 33%**
  - Soiling : **35 -48%**

1. Hadzic N, Quaglia A, Portmann B, et al. Hepatocellular carcinoma in biliary atresia: King's College Hospital experience. J Pediatr 2011;159:617-22.e1.
2. Marty TL, Seo T, Matlak ME, et al. Gastrointestinal function after surgical correction of Hirschsprung's disease: long-term follow-up in 135 patients. J Pediatr Surg 1995;30:655-8.
3. Rescorla FJ, Morrison AM, Engles D, et al. Hirschsprung's disease. Evaluation of mortality and long-term function in 260 cases. Arch Surg 1992;127:934-41; discussion 41-2.
4. Rintala RJ, Pakarinen MP. Long-term outcomes of Hirschsprung's disease. Semin Pediatr Surg 2012;21:336-43.

Introduction

Anorectal/bowel function

- Clinical assessment alone is highly subjective.
- Anorectal manometry provides an **objective method to assess anorectal functions**.

Introduction

Aim:

- To investigate **anorectal manometry findings** and **bowel function** outcomes of patients with HD after surgery.

Introduction

Hypothesis

- **Anal resting pressure** and **recto-anal inhibitory reflex** in patients with Hirschsprung disease post definitive surgery are associated with **bowel dysfunction**.

Methods

- Prospective, observational study
- Single-centre study
- Convenience sampling method

Methods

- Inclusion criteria
  - All histologically confirmed HD patients who had definitive surgery from January 2019 to December 2020 in Hospital Tunku Azizah (HTA) were recruited.

- Exclusion criteria
  - Patients who were still on a stoma
  - Patients with ongoing perineal complications

Methods

Definitions

- Length of HD[1]
  - **Short segment HD** : Aganglionosis till descending-sigmoid junction
  - **Long segment HD** : Aganglionosis from des-sigmoid to caecum
  - **Total colonic HD** : Aganglionosis of entire colon with small bowel extension.

1. Kawaguchi, A. L., Y. S. Guner, S. Sømme, A. C. Quesenberry, L. G. Arthur, J. E. Sola, C. D. Downard, R. M. Rentea, P. A. Valusek, C. A. Smith, M. B. Slidell, R. L. Ricca, R. Dasgupta, E. Renaud, D. Miniati, J. McAteer, A. L. Beres, J. Grabowski, S. D. S. Peter and A. Gosain (2021). "Management and outcomes for long-segment Hirschsprung disease: A systematic review from the APSA Outcomes and Evidence Based Practice Committee." J Pediatr Surg **56**(9): 1513-1523.

Methods

- **Definitive surgery**
  1. Mini laparotomy assisted Transanal pullthrough
  2. Duhamel procedure

- **Transition zone pullthrough**
  - Presence of both hypertrophic nerve bundles and ganglion cells within proximal margin of resected bowel

Methods

- Bowel function assessment:
  - **Paediatric Incontinence/Constipation Scoring System (PICSS) questionnaire**
  - **Anorectal manometry assessment**

At least 6 months after definitive surgery or stoma closure.

Methods

- PICSS questionnaire

  - Validated questionnaire developed by Fichtner-Feigl et al. to simultaneously evaluate **constipation, incontinence, and their combinations** in paediatric patients (Age 1-12 years old) [1].

  - Consist of 13 questions with corresponding scores.

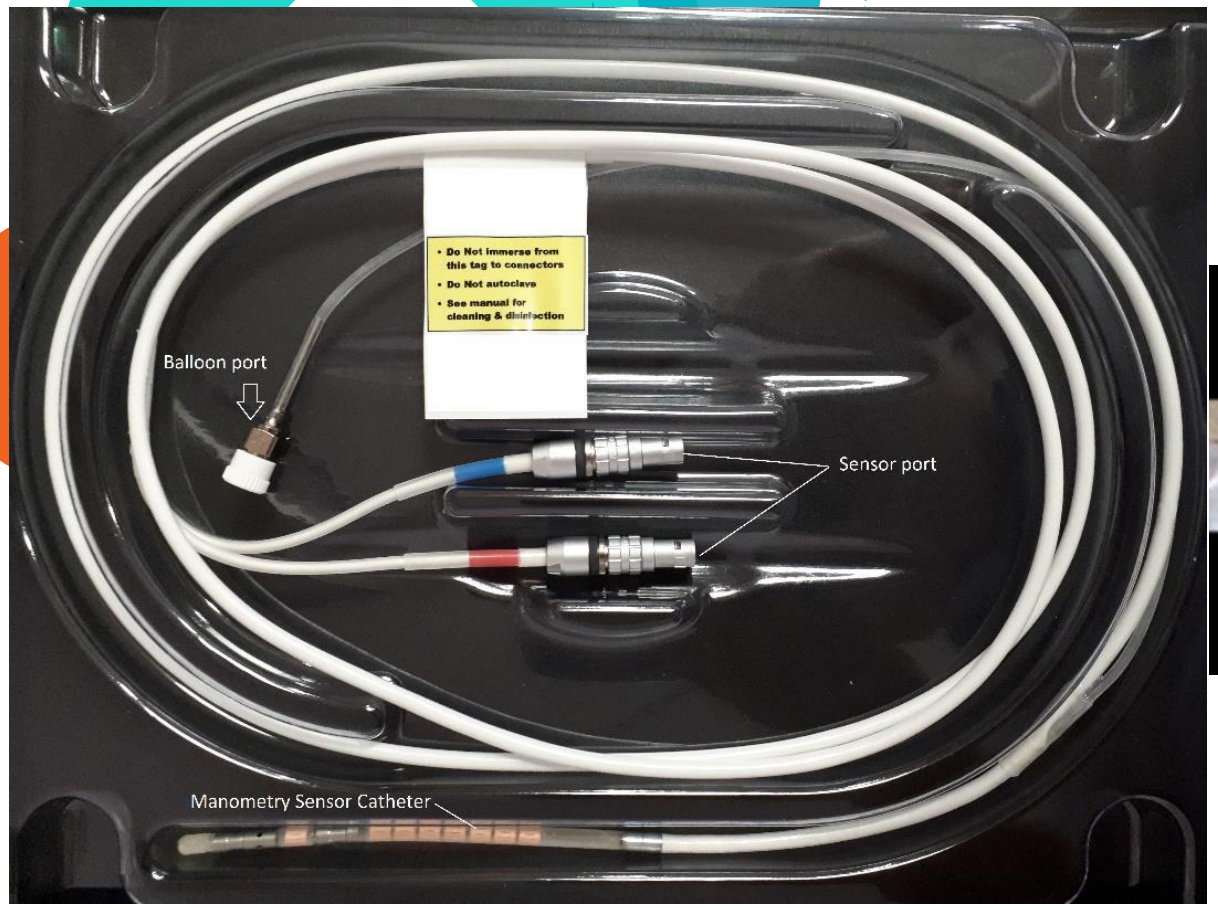
  - Final added score interpreted along corresponding age-specific graphs to group patients into **Normal, Constipation, Incontinence, or Mixed.**

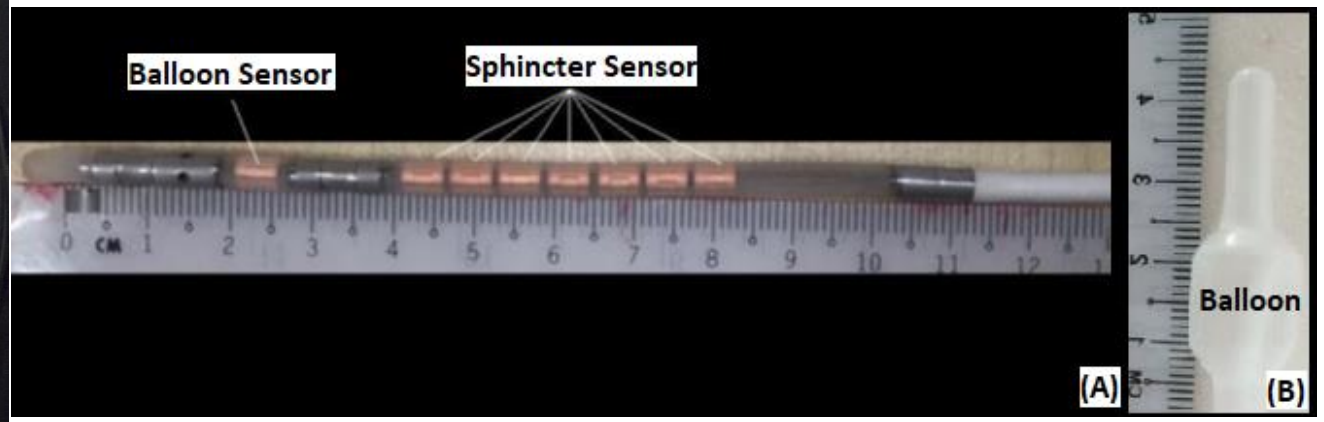
1. Fichtner-Feigl S, Sailer M, Höcht B, et al. Development of a New Scoring System for the Evaluation of Incontinence and Constipation in Children. coloproctology 2003;25:10-5.

Methods

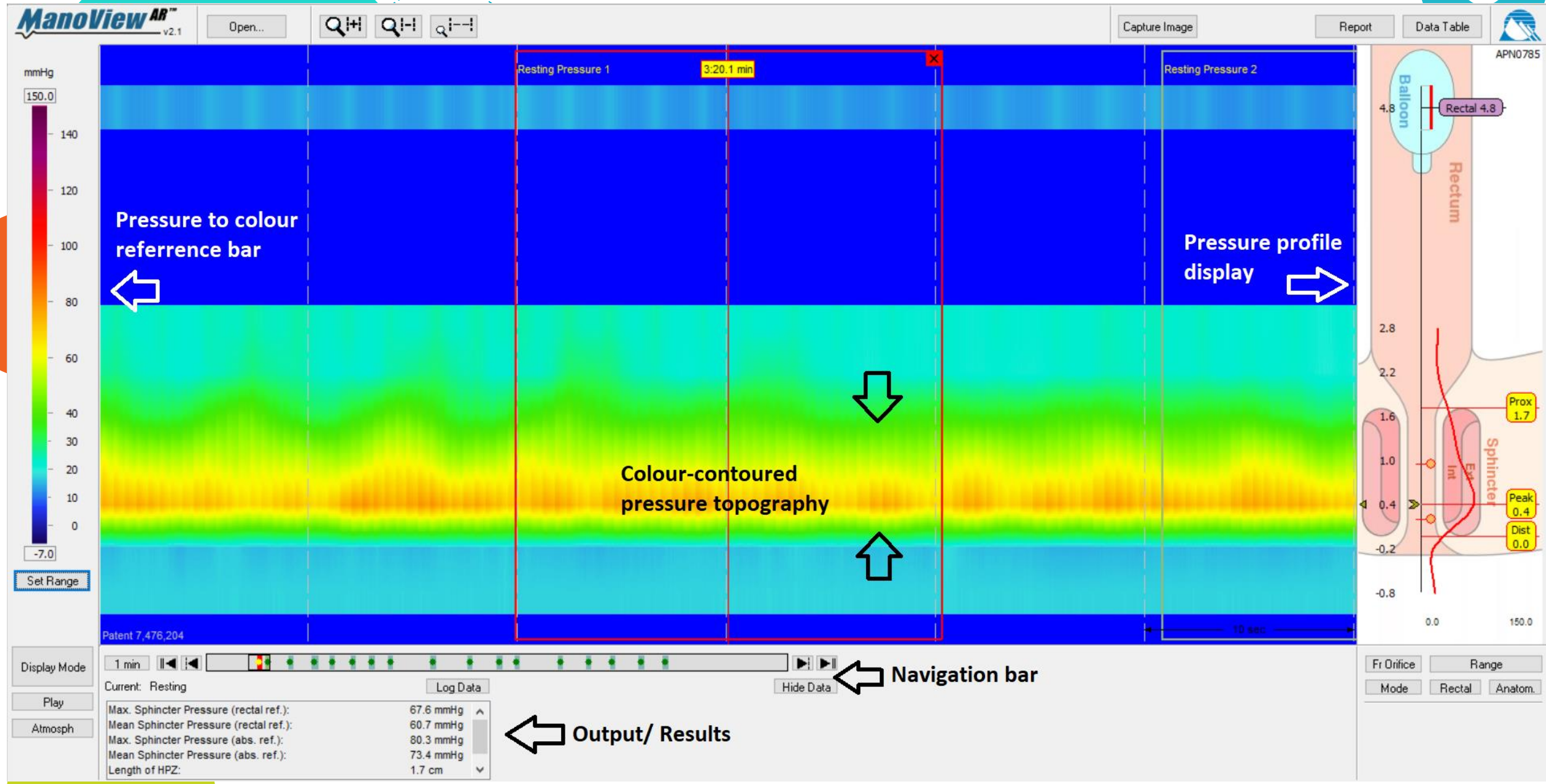
- Anorectal manometry
  - High resolution anorectal manometry (HRAM)
    - 8-channel solid state anorectal catheter
    - Adapted and modified protocol recommended by British Society of Paediatric Gastroenterology, Hepatology and Nutrition – Motility working group (BSPGHAN- MWG) [1].

1. Athanasakos E, Cleeve S, Thapar N, et al. Anorectal manometry in children with defecation disorders BSPGHAN Motility Working Group consensus statement. 2020;32:e13797.





Anorectal Manometry Catheter and Balloon.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3:20.1 min
Resting Pressure 2
Pressure to colour referrence bar
Pressure profile display
Colour-contoured pressure topography
Balloon
Rectal 4.8
Rectum
4.8
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-0.2
-0.8
Prox 1.7
Peak 0.4
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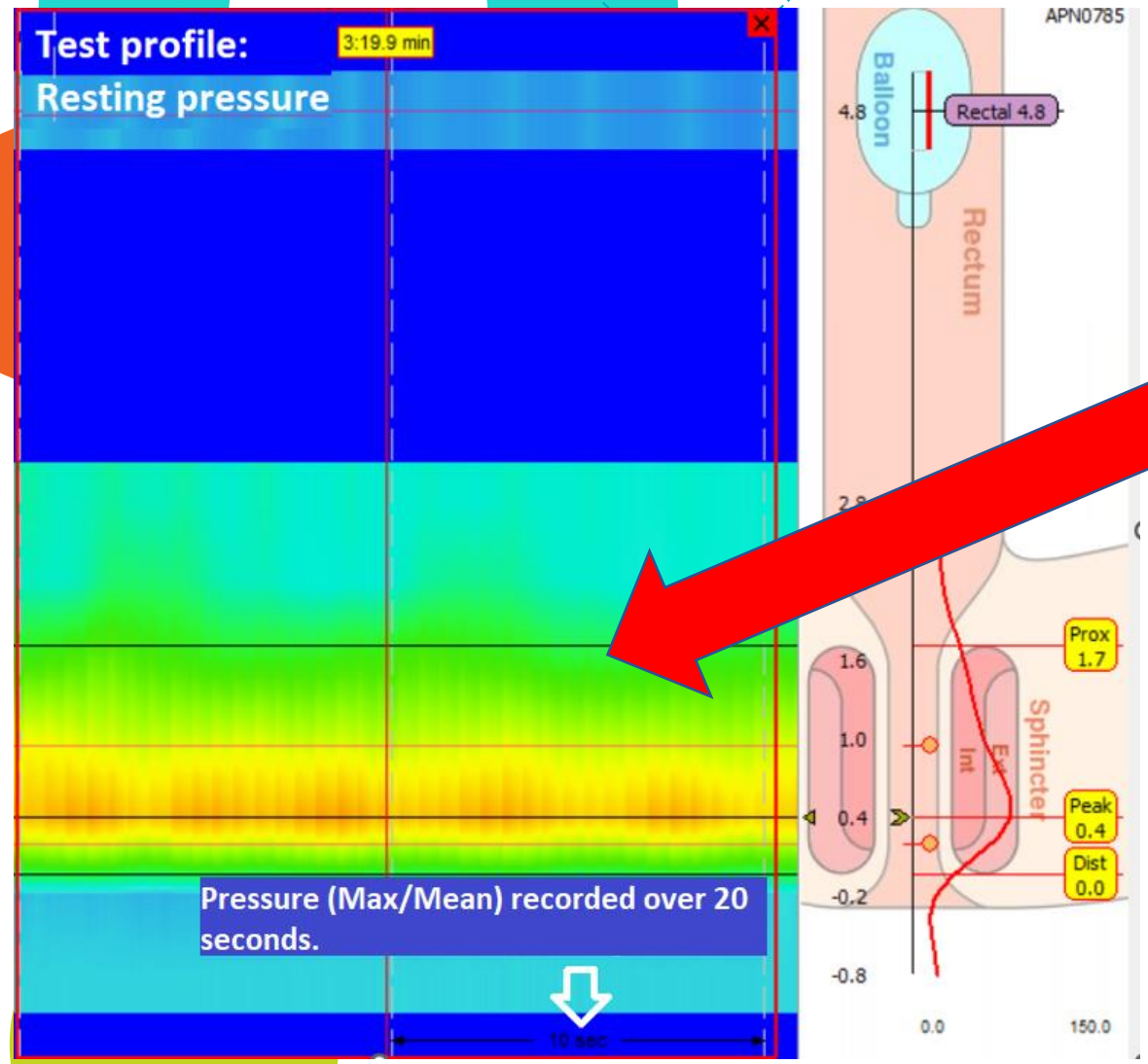
Hide Data
Navigation bar
Max. Sphincter Pressure (rectal ref.): 67.6 mmHg
Mean Sphincter Pressure (rectal ref.): 60.7 mmHg
Max. Sphincter Pressure (abs. ref.): 80.3 mmHg
Mean Sphincter Pressure (abs. ref.): 73.4 mmHg
Length of HPZ: 1.7 cm
Output/ Results
Fr Orifice
Range
Mode
Rectal
Anatom.

Methods



- Anorectal manometry

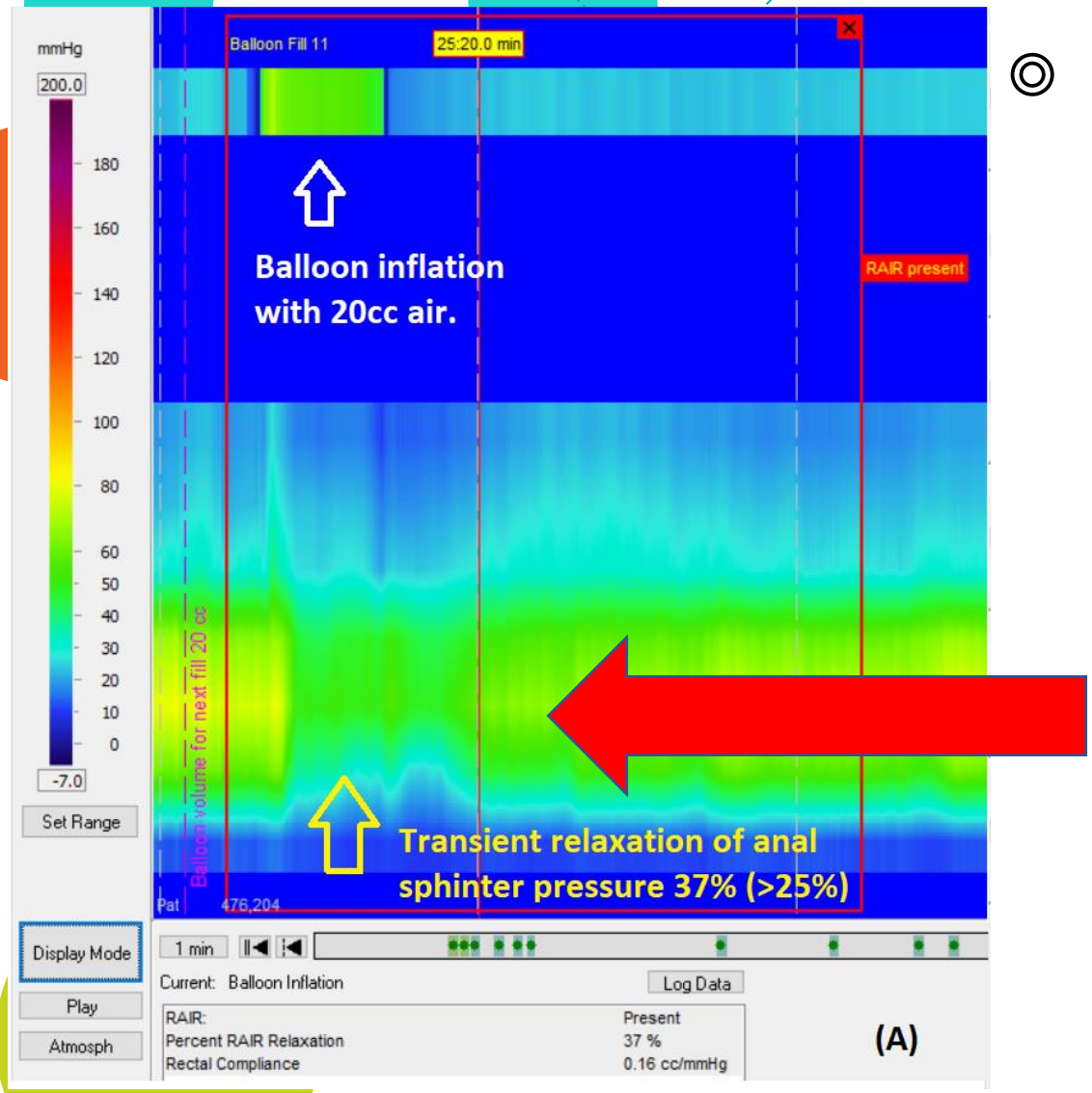
I. Anal resting pressure (ARP)

- Maximum anal resting pressure
- Mean anal resting pressure
- High-pressure zone (HPZ; Functional anal canal length)

II. Recto-anal inhibitory reflex (RAIR)

- Transient drop of anal sphincter pressure of at least 25% from baseline
- Balloon volume required documented

Methods



- Anorectal manometry

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   - Maximum anal resting pressure
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Methods

- Data analysis
  - All data recorded and analysed with IBM SPSS 23
  - Continuous data presented as median with range.
  - Categorical data as numbers and frequencies.

  - **Pearson chi-square test** was used for categorical variables
  - **Kruskal-Wallis test** was used for continuous non-parametric data

  - p value < 0.05 considered significant.

Methods

- Ethics approval
  - Presented and approved:

  - NMRR
    - ID: NMRR-19-3157-51556 S2 R3

  - UM MREC
    - MREC ID: 20191029-7949

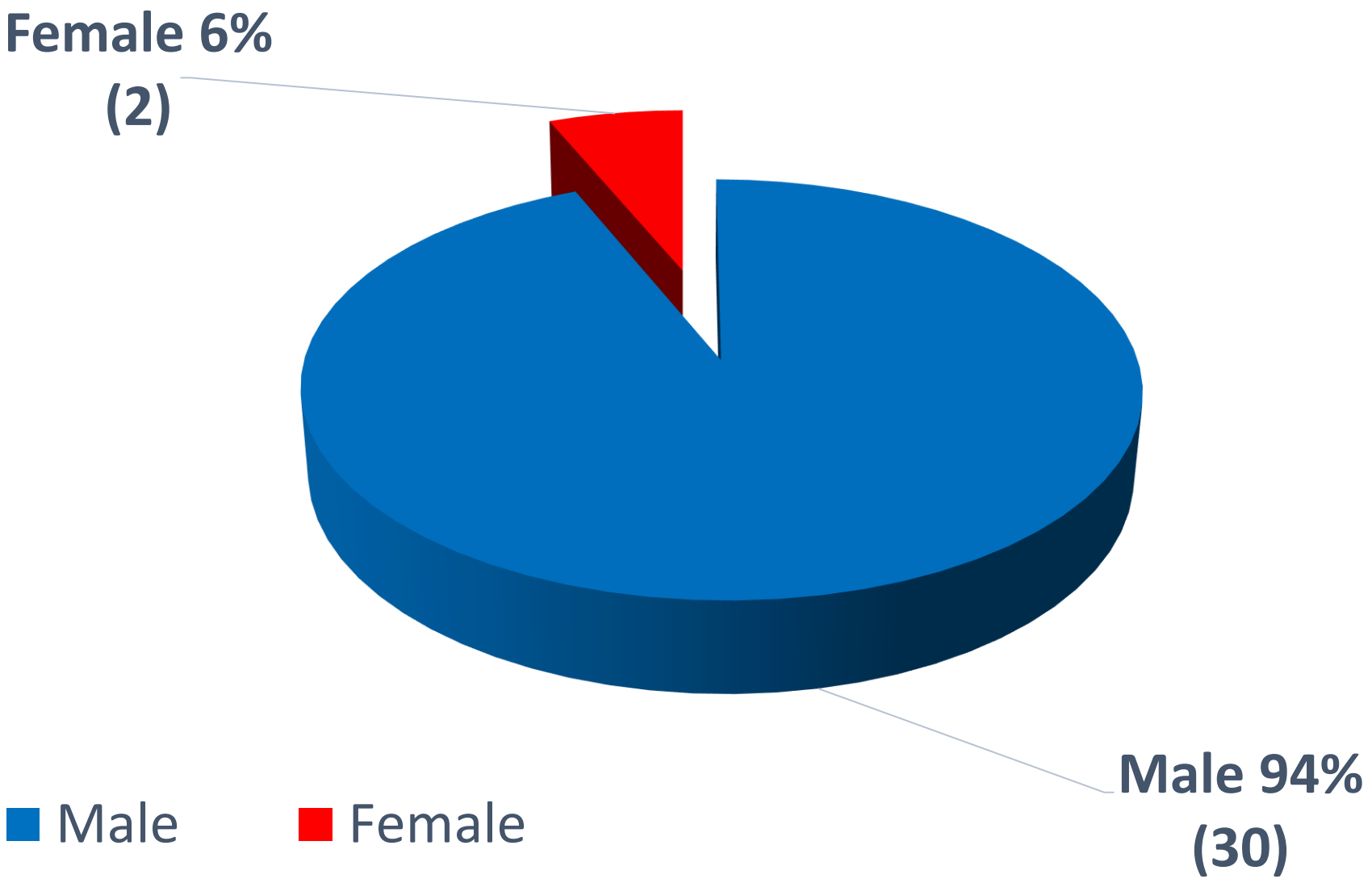
Results

- 54 eligible patients identified
- Only **32 patients** completed study

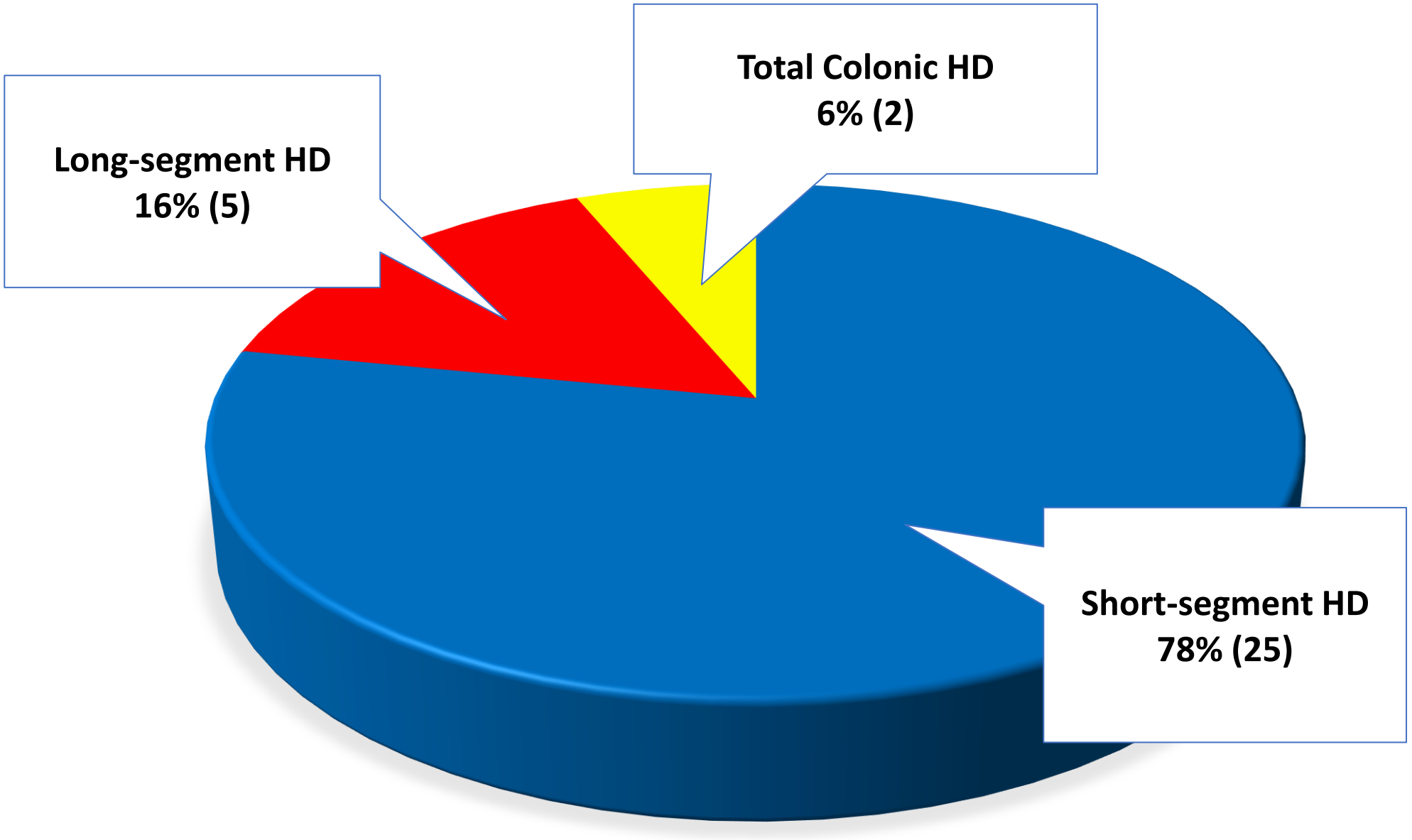
- Median age at time of study participation
  - **26.5 months** (13.8 months to 13 years)

- Median age at definitive surgery
  - **7.3 months** (1.5 months to 12 years)

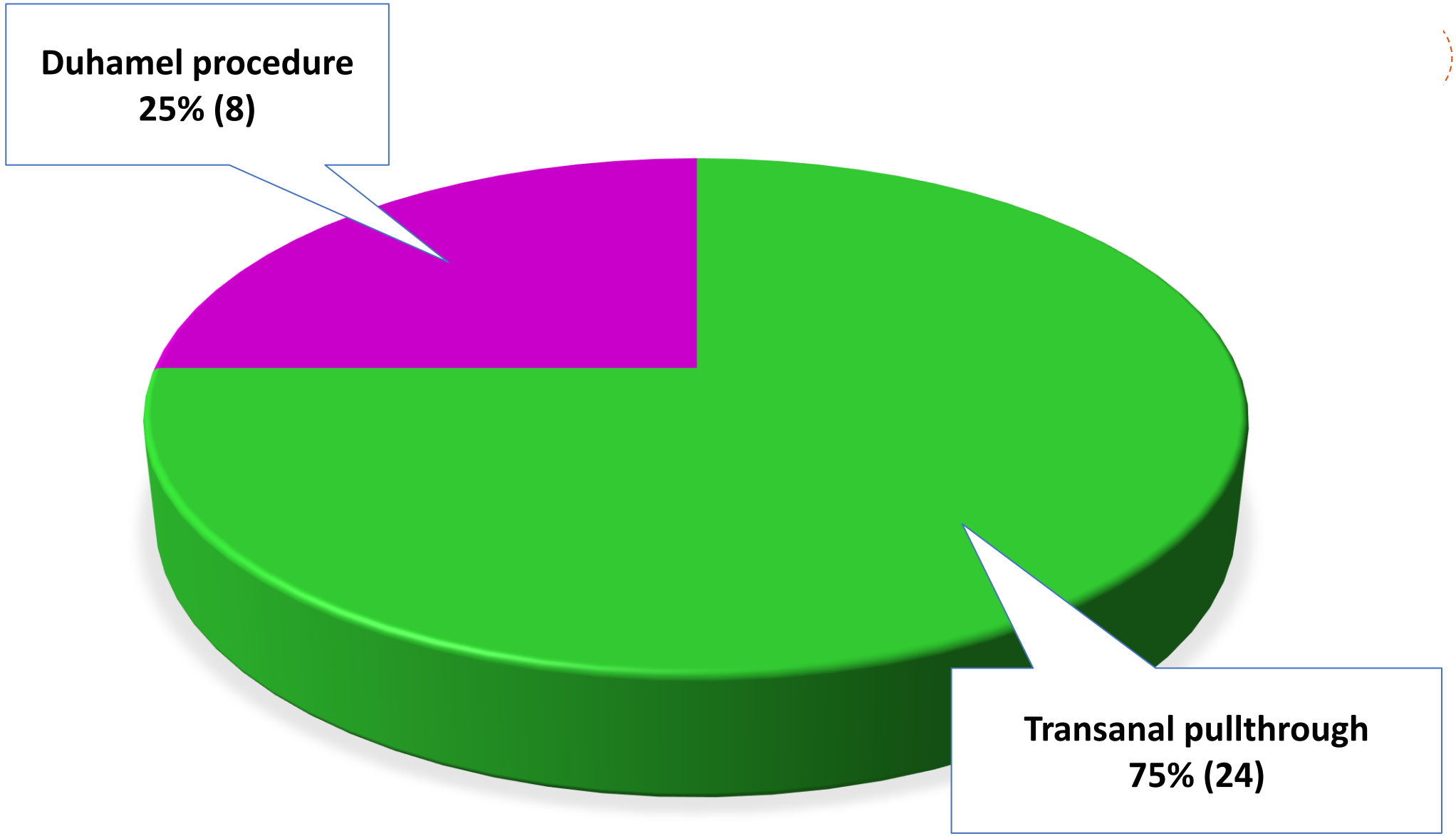
Gender distribution



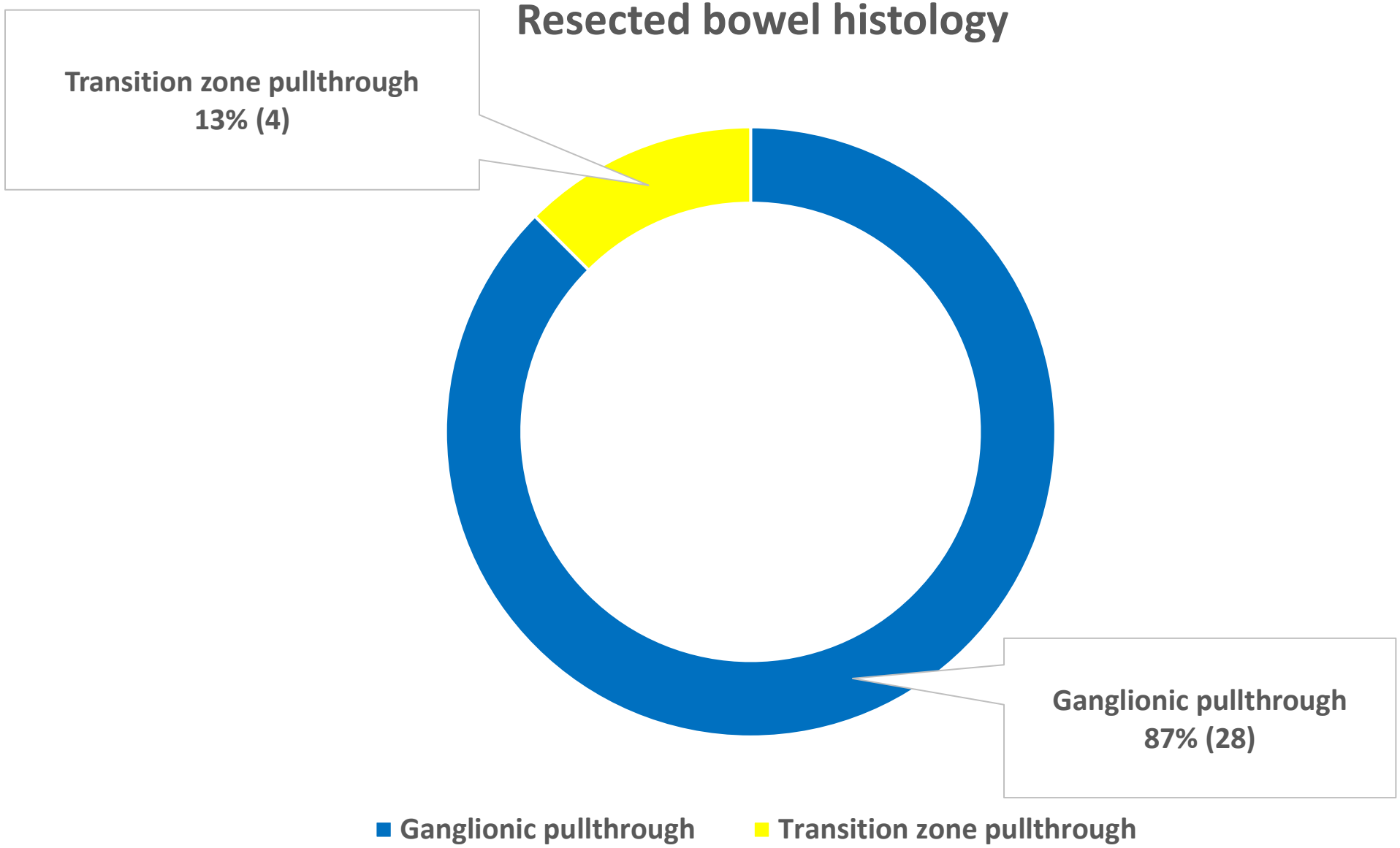
TYPE OF HIRSCHSPRUNG DISEASE

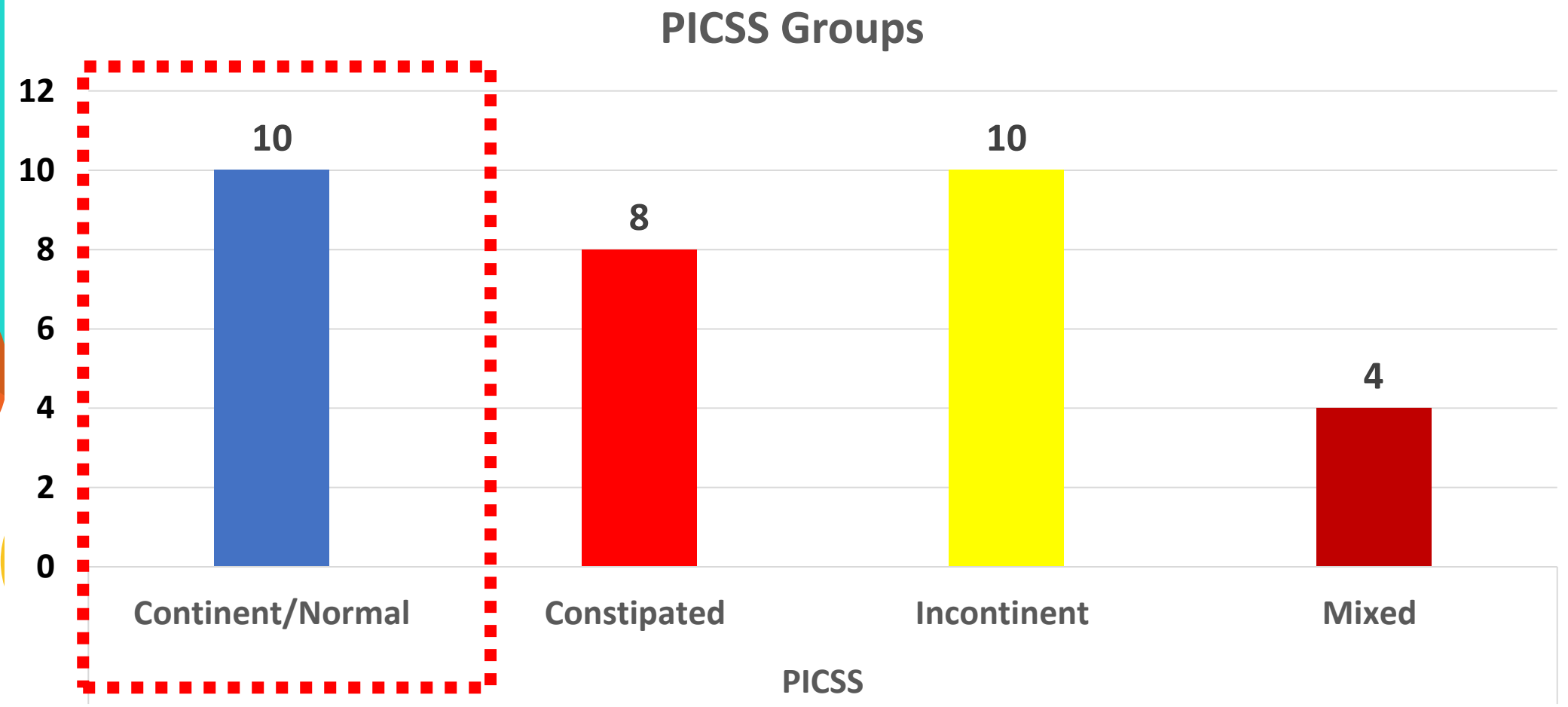


DEFINITIVE SURGERY PERFORMED

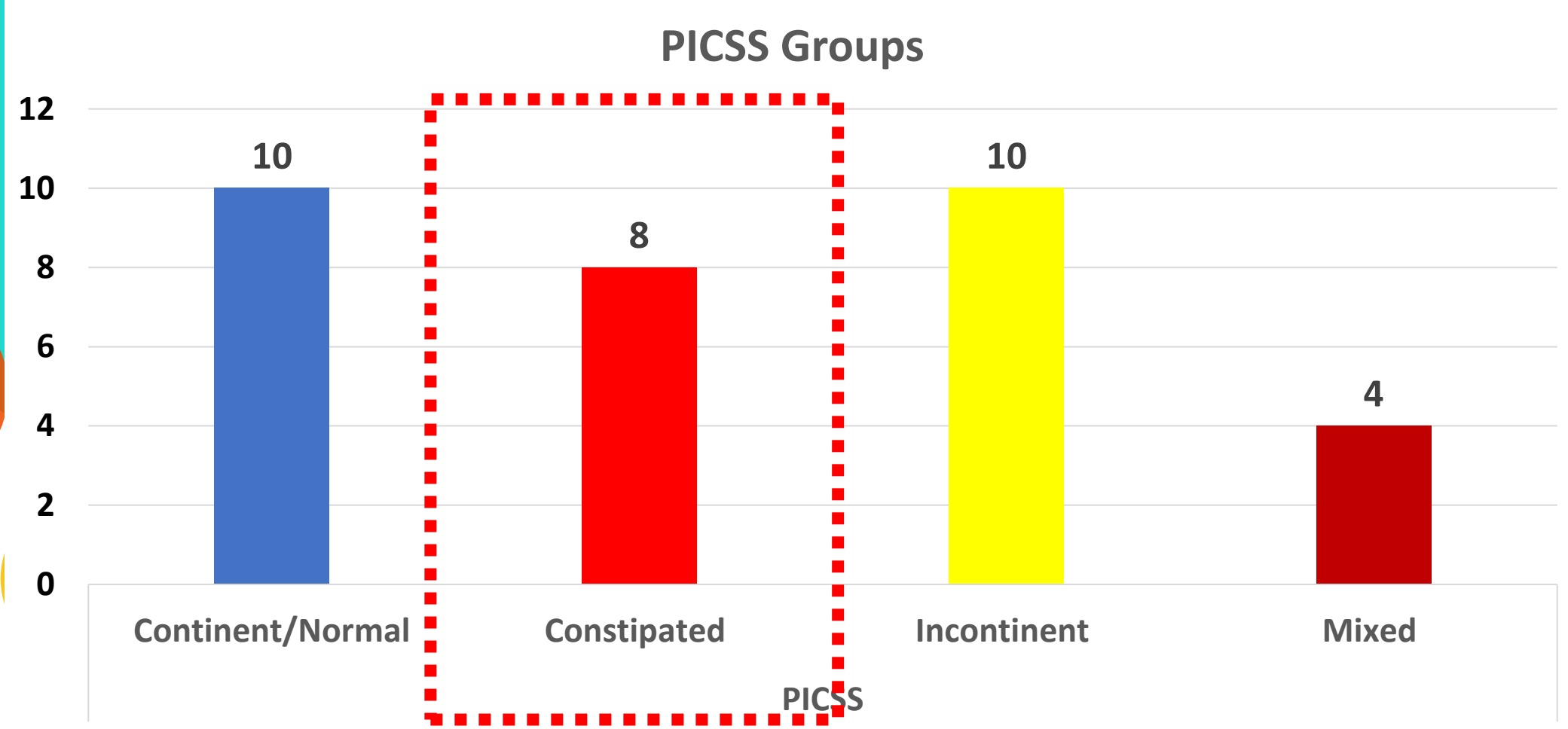


Resected bowel histology





	Group	Number	%
PICSS	Continent/Normal	10	31.3%
	Constipated	8	25%
	Incontinent	10	31.3%
	Mixed	4	12.5%



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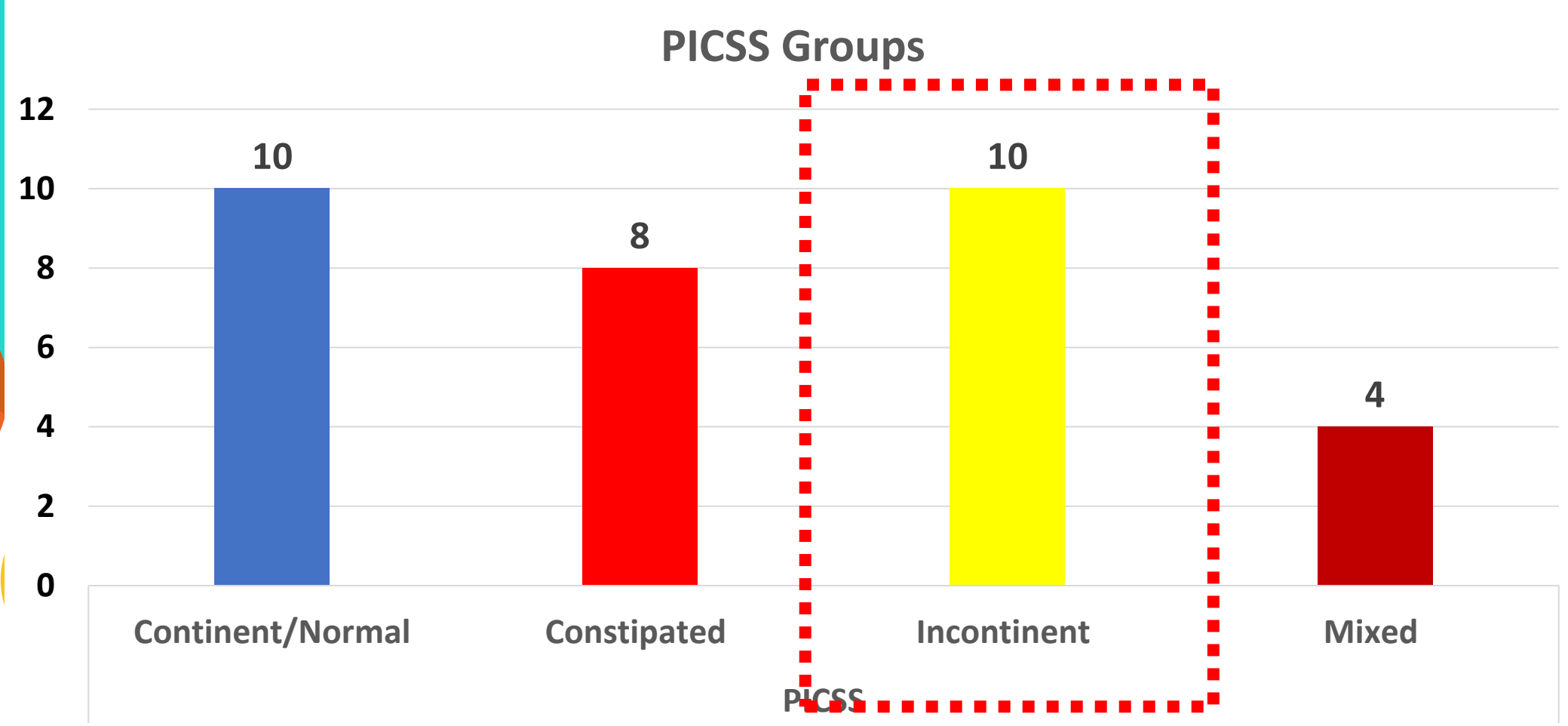

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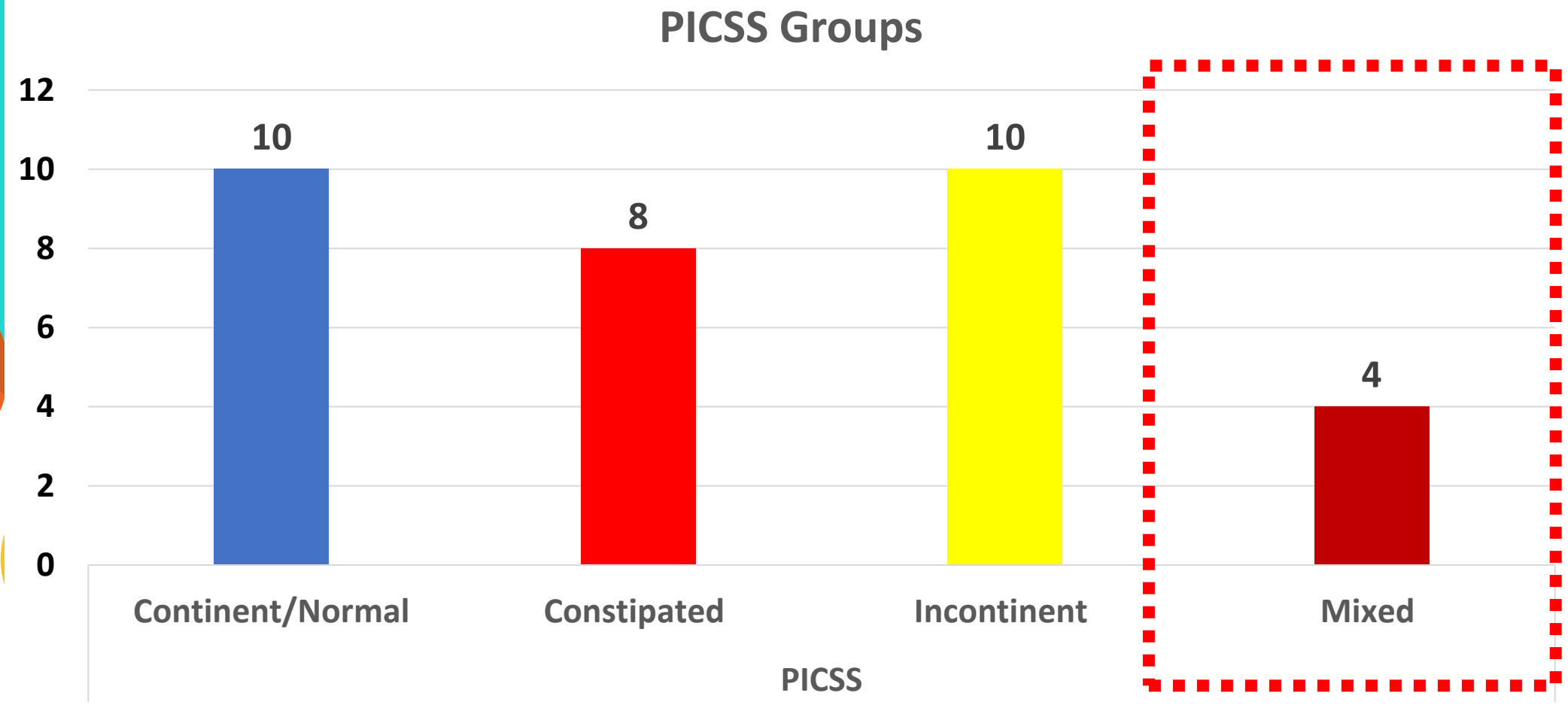

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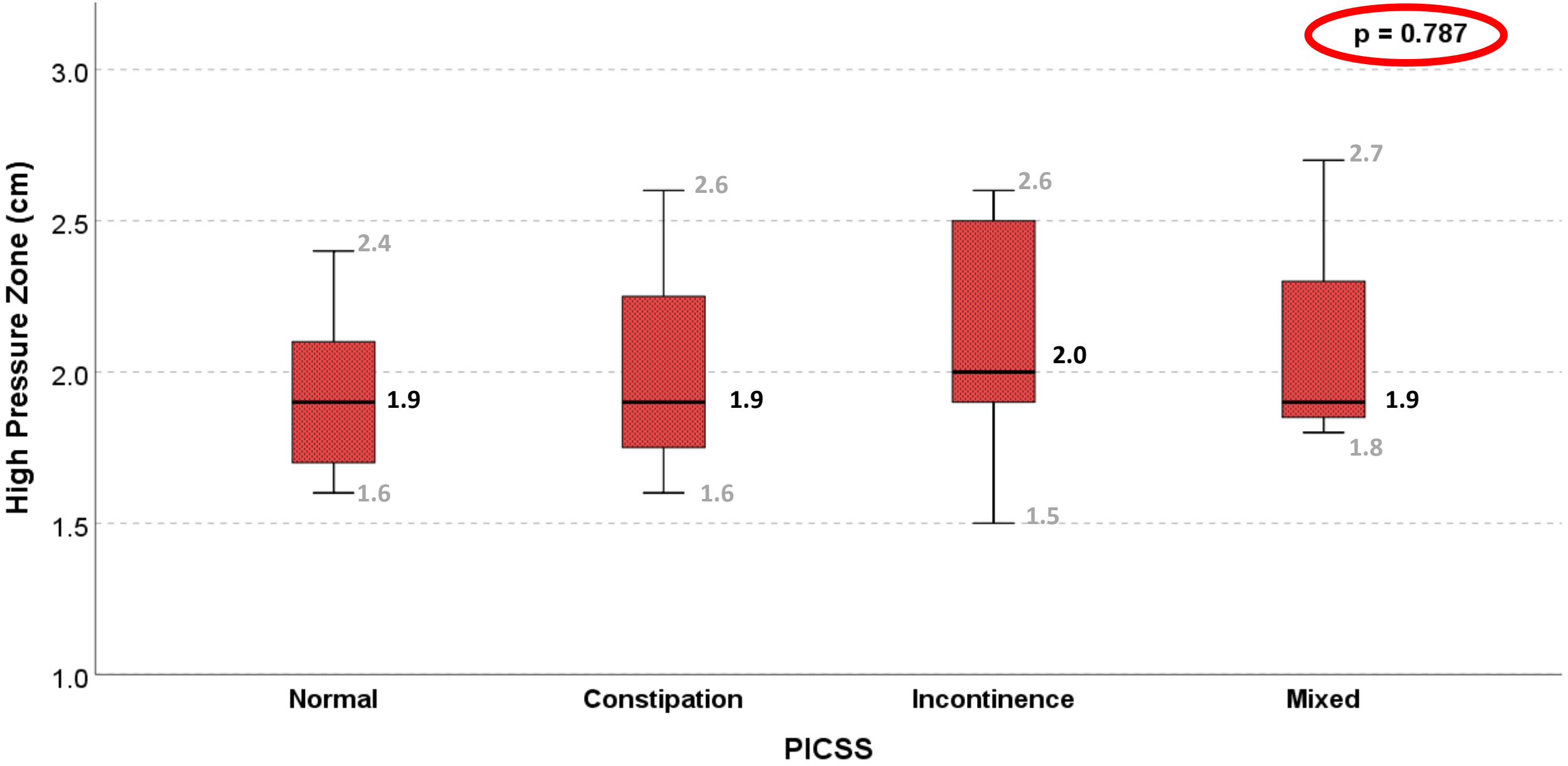
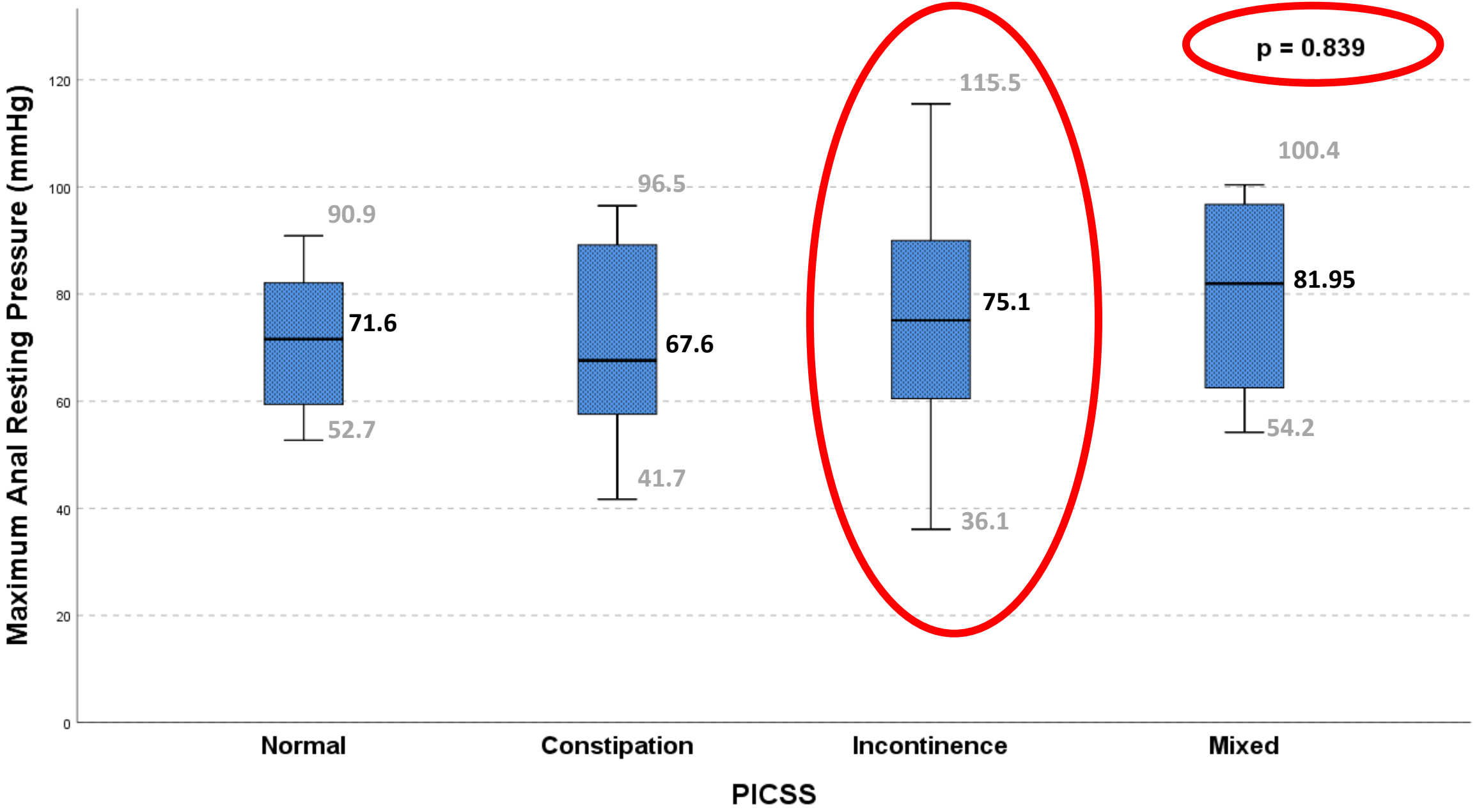
Anorectal manometry profiles		Results
Anal resting pressures	High pressure zone (cm)	**1.9 (1.5-2.7) ***
	Maximum anal resting pressure (mmHg)	**70.85 (36.10-115.50) ***
	Mean anal resting pressure (mmHg)	**62.60 (30.80-106.90) ***

*Median (Range)

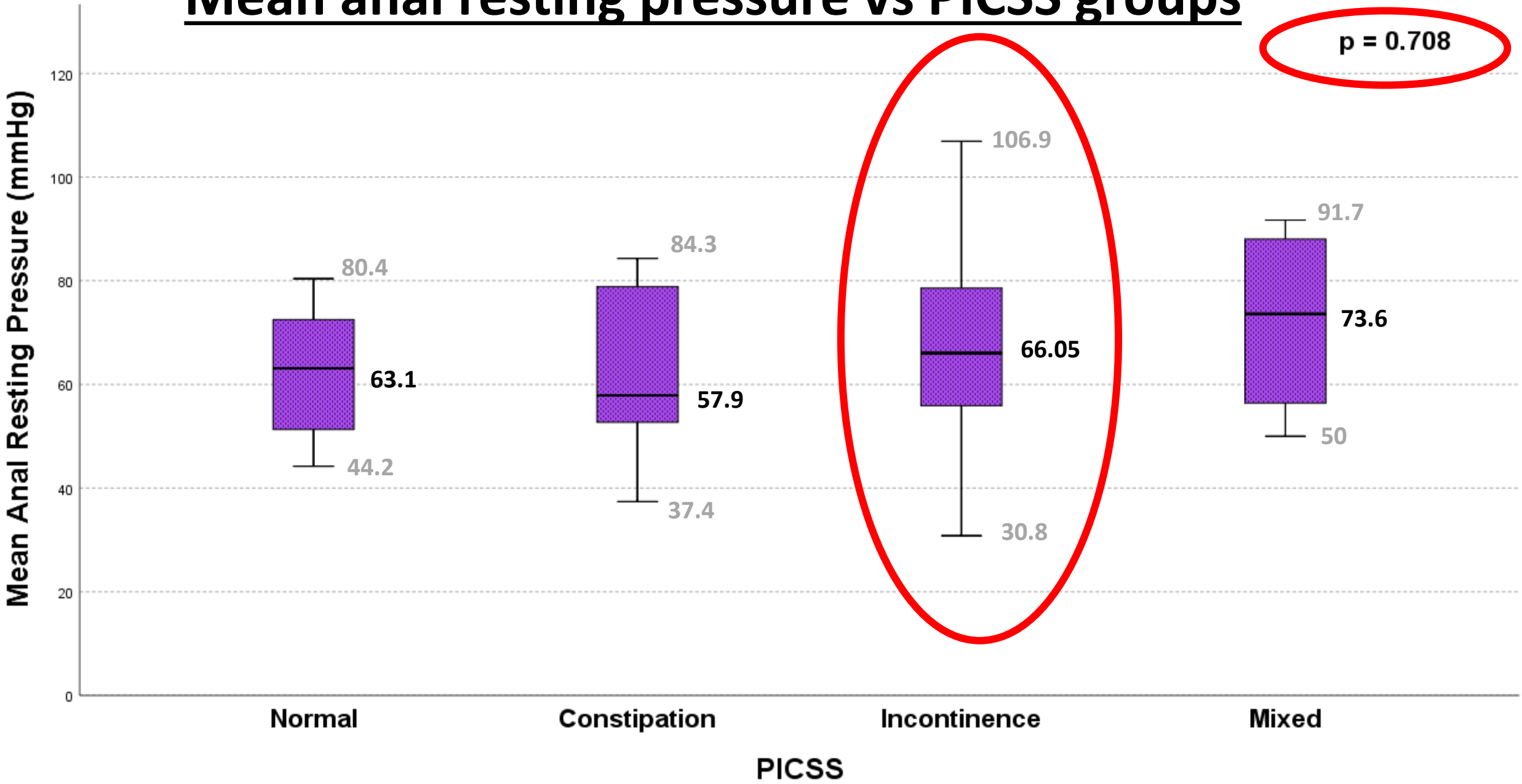
High-pressure zone (HPZ) vs PICSS groups



Maximum anal resting pressure vs PICSS groups

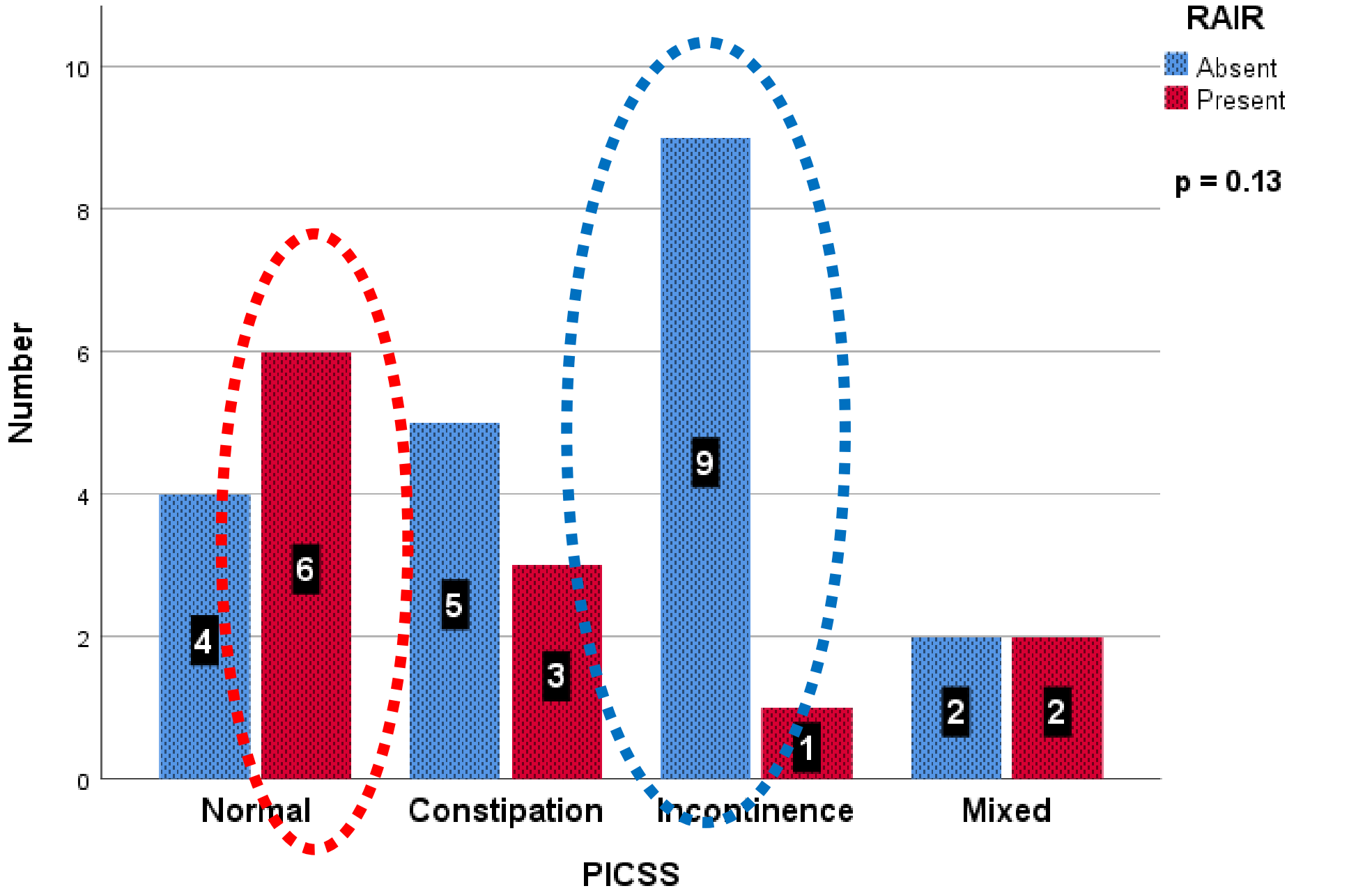


Mean anal resting pressure vs PICSS groups



Anorectal manometry profiles		Results
RAIR	Present	**12 (37.5%) #**
	Absent	**20 (62.5%) #**
RAIR balloon	Volume (cc)	**30 (10-60) ***

Recto-anal inhibitory reflex (RAIR) vs PICSS groups



Discussion

- Our study shows that majority patients with Hirschsprung disease exhibit bowel dysfunction
  - **69% have some form of bowel dysfunction**

- Higher than reported literature 5-48%

Discussion

- Biggest challenge in paediatric anorectal manometry is the **lack of standardized normative values**.

- British Society of Paediatric Gastroenterology, Hepatology and Nutrition – Motility work group (**BSPGHAN-MWG)**
  - Suggest adopting **normative values** published by Banusiuk et al. [1]
  - Largest series (61 normal children)

1. Banasiuk M, Banaszkiewicz A, Dziekiewicz M, Załęski A, Albrecht P. Values From Three-dimensional High-resolution Anorectal Manometry Analysis of Children Without Lower Gastrointestinal Symptoms. Clin Gastroenterol Hepatol. 2016 Jul;14(7):993-1000.e3. doi: 10.1016/j.cgh.2016.01.008. Epub 2016 Jan 25. PMID: 26820403.

Discussion

- In our study, overall anal resting pressure results seem to **trend lower** than normative values reported Banusiuk et al. [1]

- However, our findings **correspond to other studies** on anorectal manometry for post surgery HD patients. [2-4]

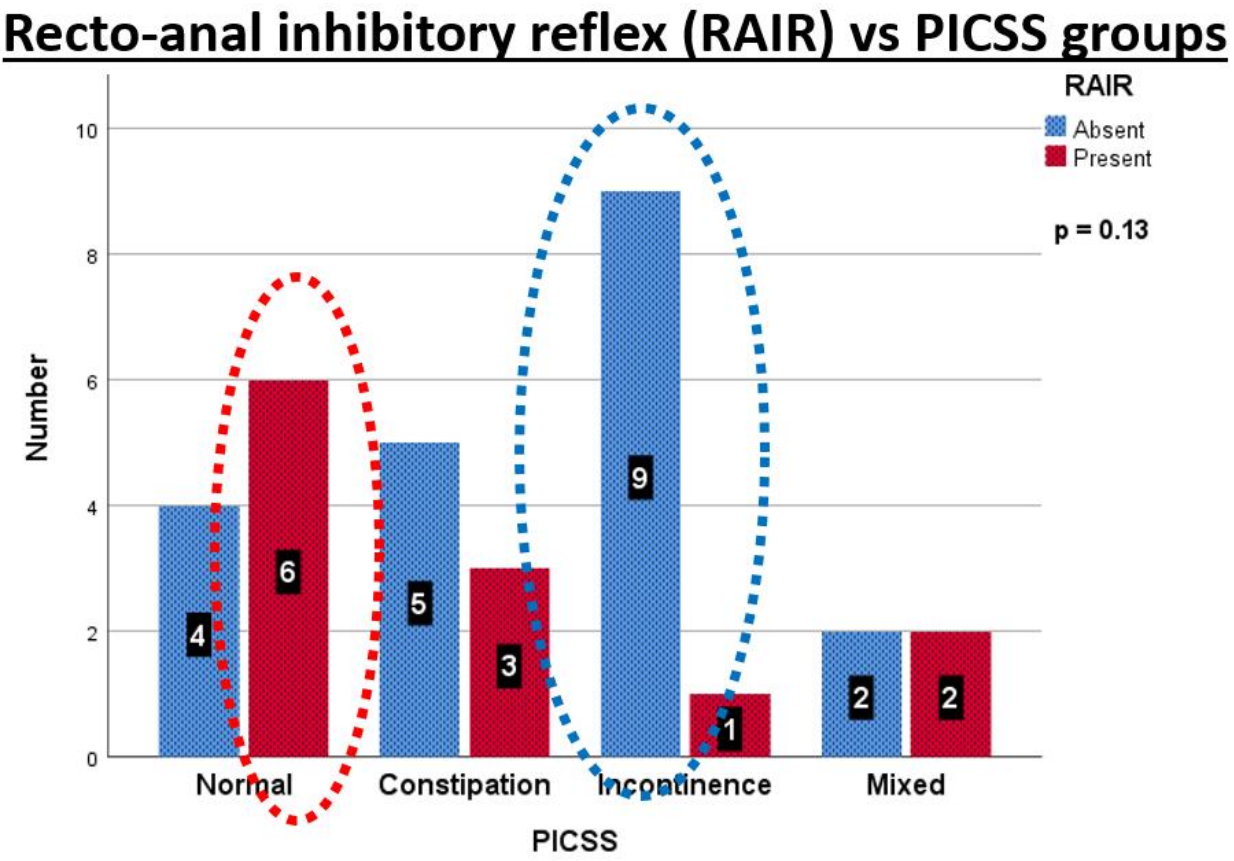
1. Banasiuk M, Banaszkiewicz A, Dziekiewicz M, Załęski A, Albrecht P. Values From Three-dimensional High-resolution Anorectal Manometry Analysis of Children Without Lower Gastrointestinal Symptoms. Clin Gastroenterol Hepatol. 2016 Jul;14(7):993-1000.e3. doi: 10.1016/j.cgh.2016.01.008. Epub 2016 Jan 25. PMID: 26820403.
2. Gad El-Hak, N. A., M. M. El-Hemaly, E. H. Negm, E. A. El-Hanafy, M. H. Abdel Messeh and H. H. Abdel Bary (2010). "Functional outcome after Swenson's operation for Hirshsprung's disease." Saudi J Gastroenterol **16**(1): 30-34.
3. Demirbag, S., T. Tiryaki and T. Purtuloglu (2013). "Importance of anorectal manometry after definitive surgery for Hirschsprung's disease in children." **10**(1): 1-4.
4. Tran, V. Q., T. Mahler, P. Bontems, D. Q. Truong, A. Robert, P. Goyens and H. Steyaert (2018). "Interest of Anorectal Manometry During Long-term Follow-up of Patients Operated on for Hirschsprung's Disease." J Neurogastroenterol Motil **24**(1): 70-78.

Discussion

- Lower anal resting pressure may reflect an **internal sphincter dysfunction inherent to HD** or **iatrogenic sphincter damage** secondary inadvertent harmful stretching during pullthrough operation for HD. [1-2]

1. Bjornland K, Diseth TH, Emblem R. Long-term functional, manometric, and endosonographic evaluation of patients operated upon with the Duhamel technique. Pediatr Surg Int 1998;13:24-8.
2. Heikkinen M, Rintala R, Luukkonen P. Long-term anal sphincter performance after surgery for Hirschsprung's disease. J Pediatr Surg 1997;32:1443-6.

Discussion



- **Absent RAIR trended higher in incontinent group**; while **presence of RAIR trended higher in the normal group**.
  - NO significant association noted (PICSS vs RAIR)
- A larger group of patients may produce statistically significant values.

Limitations

1. **Sample size**
   i. COVID-19 pandemic
   ii. Parental refusal for asymptomatic child
   iii. Perception of anorectal manometry

2. **Surgeon factors**

Conclusion

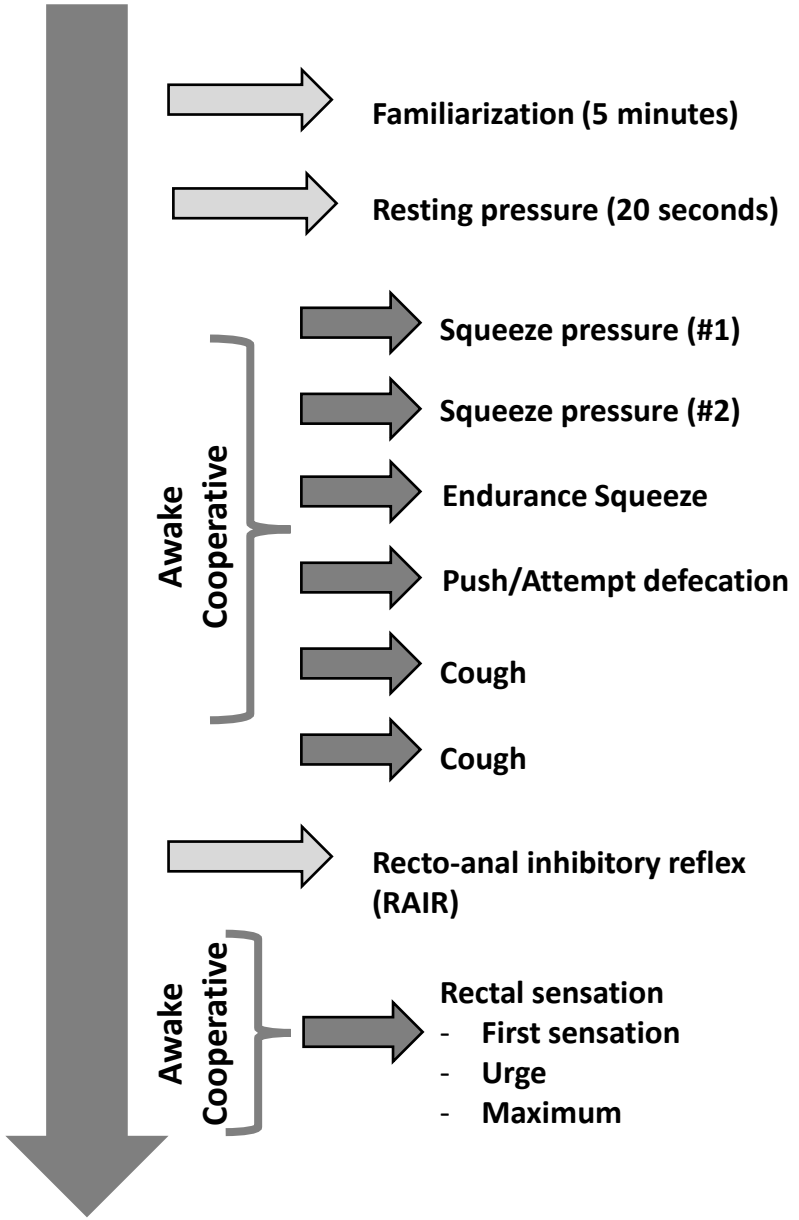
In our study,

- Majority of patients post HD surgery have **bowel dysfunction**
- Overall ARP **trended lower** in HD patients post surgery.
- Recto-anal inhibitory reflex **(RAIR) was present in 37.5%** HD patients post surgery.
- We **did not find** any correlation between bowel dysfunction (as assessed by **PICSS)** with either **anal resting pressure** (mean and maximum ARP), **high-pressure zone**, or **recto-anal inhibitory reflex.**

*But our study is limited by small sample size.

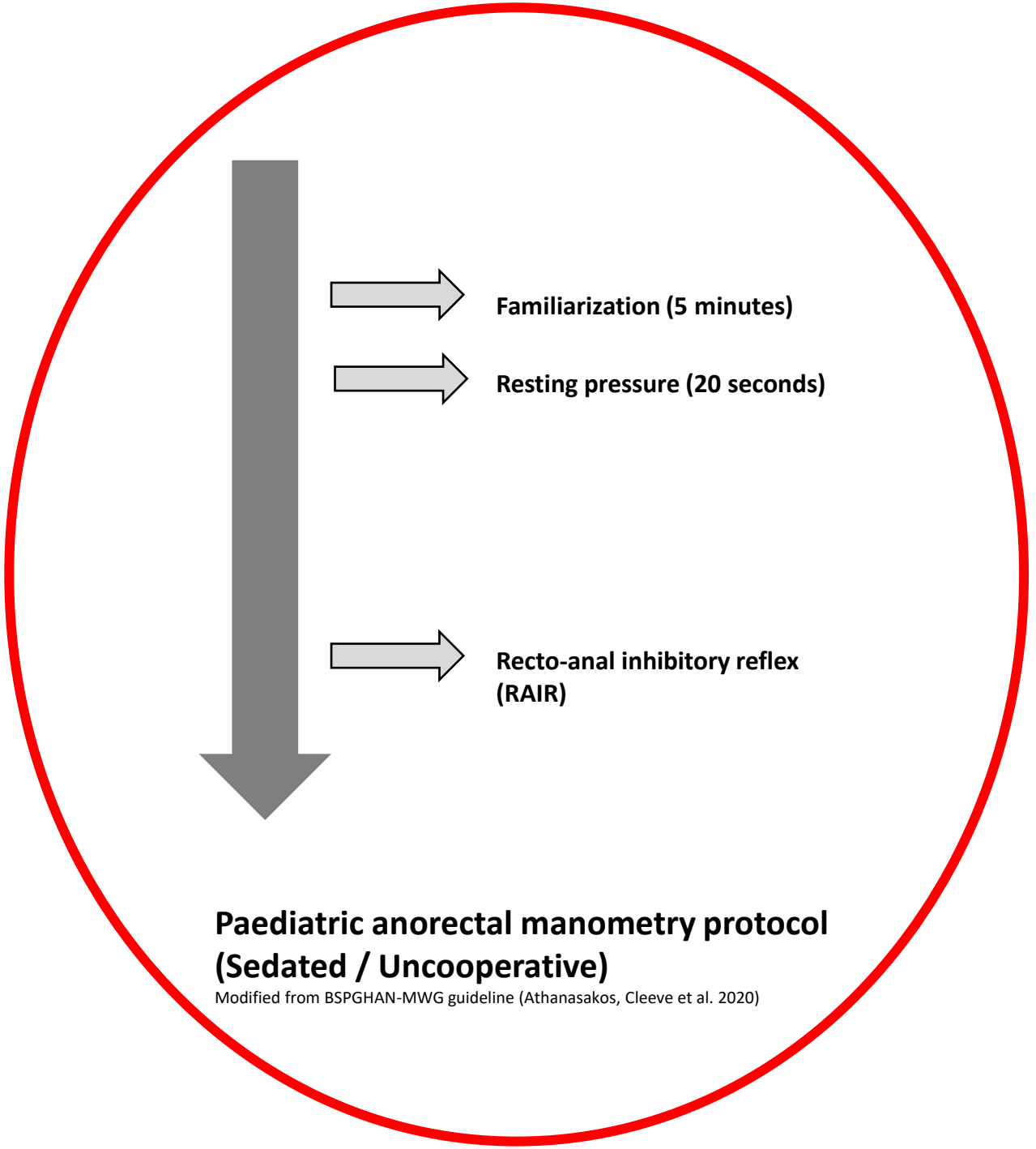
Thank you

EXTRA SLIDES



Paediatric anorectal manometry protocol (Awake/ Cooperative)

Modified from BSPGHAN-MWG guideline (Athanasakos, Cleeve et al. 2020)



Paediatric anorectal manometry protocol (Sedated / Uncooperative)

Modified from BSPGHAN-MWG guideline (Athanasakos, Cleeve et al. 2020)

Group variable	1-5 years Mean (+/- SD)	5-8 years Mean (+/- SD)	9-12 years Mean (+/- SD)	12-18 years Mean (+/- SD)
Maximum anal resting pressure (mmHg)	115 (28)	104 (20)	112 (17)	110 (22)
Mean anal resting pressure (mmHg)	94 (24)	86 (15)	94 (15)	96 (19)
Maximum squeeze pressure (mmHg)	201 (60)	206 (40)	206 (59)	229 (65)
Length of HPZ (cm)	2.2 (0.5)	2.4 (0.4)	2.9 (0.6)	3.1 (0.7)
RAIR (cm3)	13.3 (7.5)	11.1 (3.2)	13.7 (5.9)	18.6 (15.1)

Normative values for paediatric anorectal manometry

Banasiuk M, Banaszkiewicz A, Dziekiewicz M, Załęski A, Albrecht P. Values From Three-dimensional High-resolution Anorectal Manometry Analysis of Children Without Lower Gastrointestinal Symptoms. Clin Gastroenterol Hepatol. 2016 Jul;14(7):993-1000.e3. doi: 10.1016/j.cgh.2016.01.008. Epub 2016 Jan 25. PMID: 26820403.

	n	Surgery	System	Anal resting pressure (Mean +/- SD) mmHg			RAIR
				Incontinent	Continent	Constipation	n (%)
(Gad El-Hak et al., 2010)	52	Swenson	Water perfused catheter; conventional polygraph	31.7 (12.1)	58.1 (15.1)	-	5 (10%)
(Demirbag, Tiryaki, & Purtuloglu, 2013)	18	Modified Duhamel	Water perfused catheter; conventional polygraph	-	36 (8.8)	40.45 (8.09)	4 (22%)
(Tran et al., 2018)	19	Soave	Water-perfused catheter; conventional polygraph	47 (12)	63 (11)	-	8 (42.1%)

1. Gad El-Hak, N. A., M. M. El-Hemaly, E. H. Negm, E. A. El-Hanafy, M. H. Abdel Messeh and H. H. Abdel Bary (2010). "Functional outcome after Swenson's operation for Hirshsprung's disease." Saudi J Gastroenterol **16**(1): 30-34.
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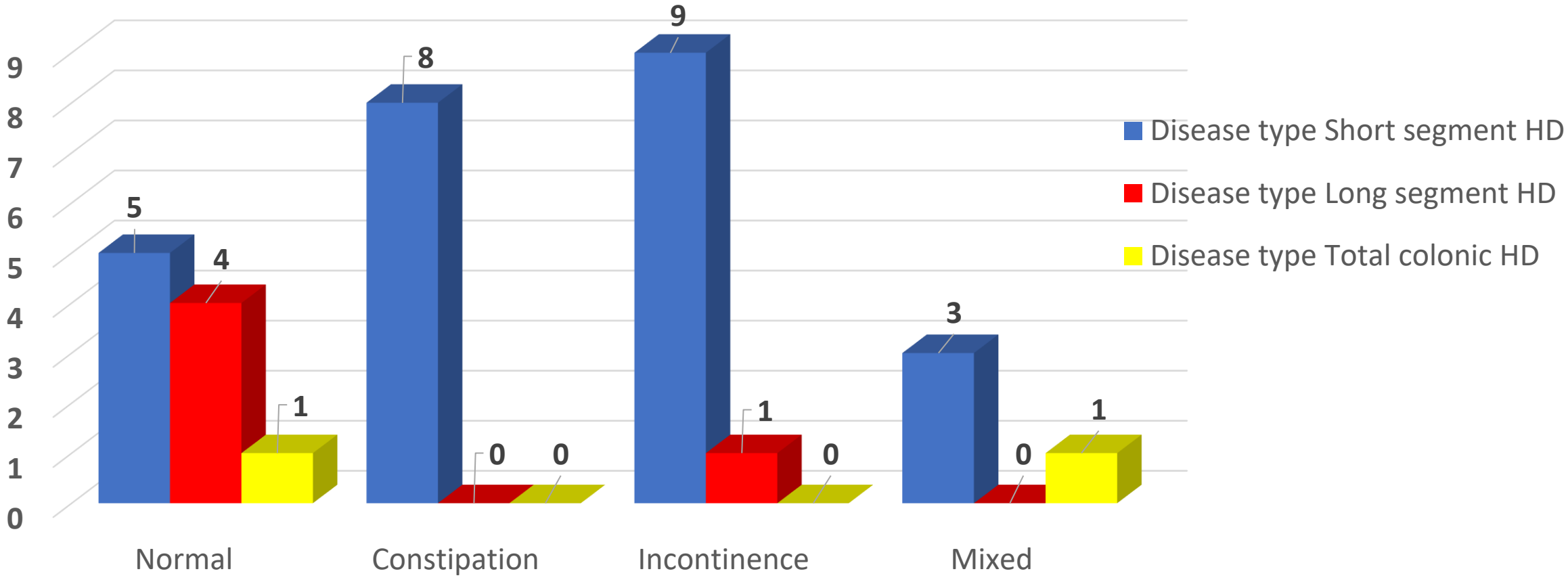
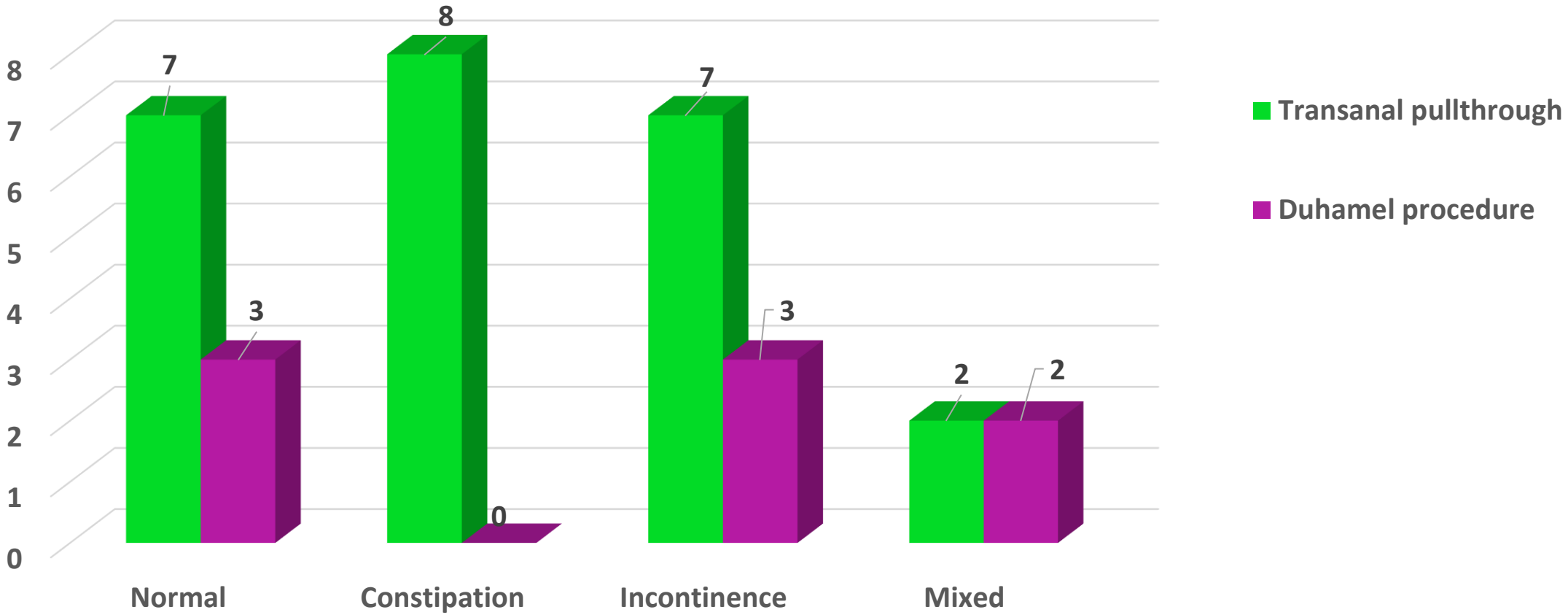
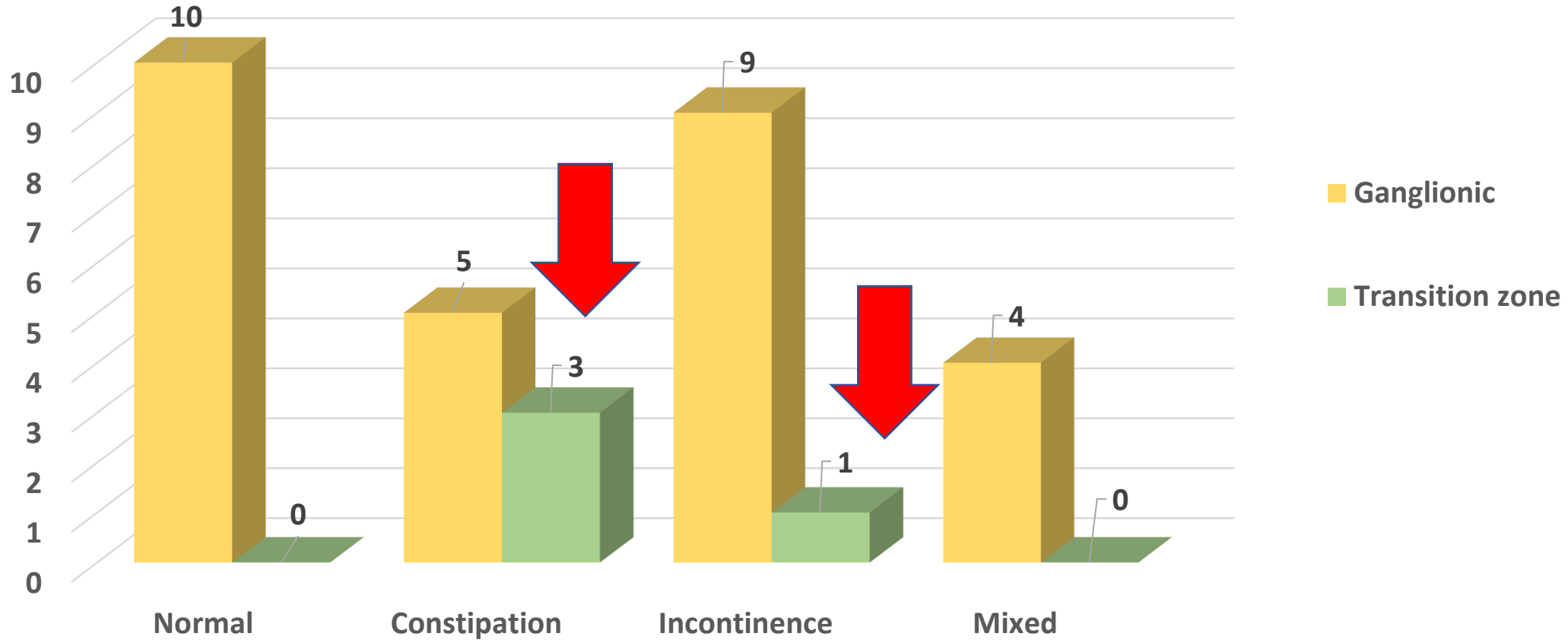
3. Tran, V. Q., T. Mahler, P. Bontems, D. Q. Truong, A. Robert, P. Goyens and H. Steyaert (2018). "Interest of Anorectal Manometry During Long-term Follow-up of Patients Operated on for Hirschsprung's Disease." J Neurogastroenterol Motil **24**(1): 70-78.



		PICSS				p-value
		Normal (n=10)	Constipation (n=8)	Incontinence (n=10)	Mixed (n=4)	
Disease type	Short segment HD	5	8	9	3	p = 0.083
	Long segment HD	4	0	1	0	
	Total colonic HD	1	0	0	1	

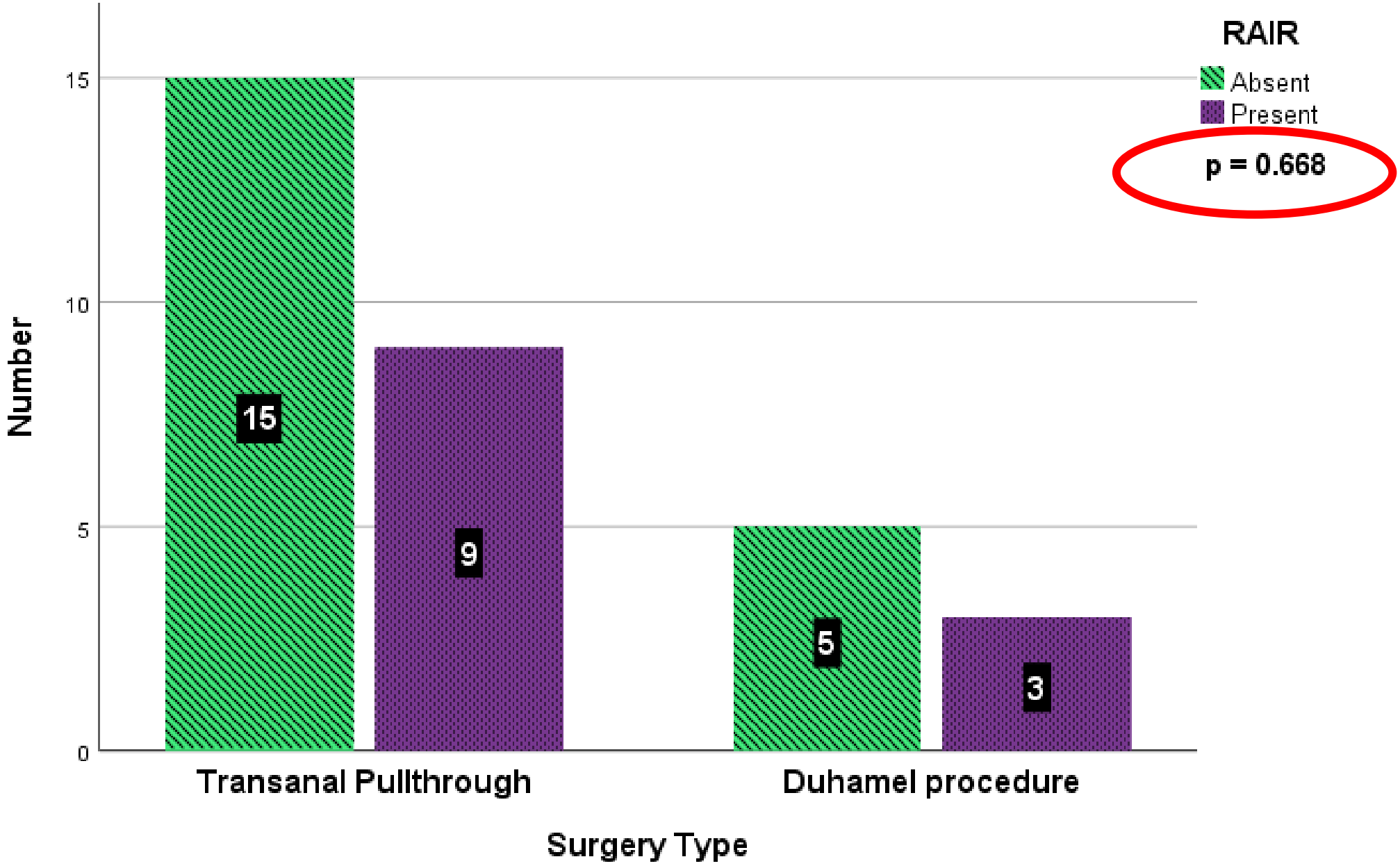


		PICSS				p value
		Normal (n=10)	Constipation (n=8)	Incontinence (n=10)	Mixed (n=4)	
Types of surgery	Transanal pullthrough	7	8	7	2	p = 0.234
	Duhamel procedure	3	0	3	2	



		PICSS				p value
		Normal (N=10)	Constipation (n=8)	Incontinence (n=10)	Mixed (n=4)	
Pullthrough bowel histology	Ganglionic	10	5	9	4	p = 0.085
	Transition zone	0	3	1	0	

Recto-anal inhibitory reflex (RAIR) vs Surgery type



Recto-anal inhibitory reflex (RAIR) vs bowel histology