

13th ASEAN SOCIETY OF PAEDIATRIC SURGEONS CONGRESS

ORAL PRESENTATION
REFERENCE NUMBER : 90

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TITLE

ASSESSMENT OF CHILDREN WITH
HIRSCHSPRUNGS DISEASE POST DEFINITIVE
SURGERY – ANORECTAL MANOMETRY AND
FUNCTIONAL OUTCOME

Introduction

- Clinical Research
- Prospective Cross-Sectional Study
- Duration :1st January 2017 – 31st March 2018
(13 months)
- Scope : Hirschsprungs Disease (HD)

- Hirschsprungs disease(HD) is a common intestinal obstruction among neonates, and children.
- Description as early as 1691 by Fredycius Ruysch, Amsterdam.
- In 1886, Sir Harald Hirschsprung presented case series with description of ‘congenital megacolon’ evidenced by pathological specimens
- In 1946 , Dr Swenson ,revolutionized the management of HD. Introduced the first definitive surgery for HD.

Sir Harald Hirschsprung (1830-1916)



Investigation

- Abdominal radiograph
- Barium Enema – 1948 , by Dr Swenson, Neyhauser and Picket ; delineate area of spasm in rectum and rectosigmoid
- Histopathology specimen – suggested by Dr Swenson. Dobbins and Bill, developed a suction rectal biopsy
- Rectoanal inhibitory reflex(RAIR) – 1877 Gowers and confirmed by Denny Robertson 1935

RAIR

- Transient relaxation of the internal anal sphincter in response to rectal distention.
- Loss of the rectoanal inhibitory response is interpreted as being consistent with Hirschsprungs disease.

Literature Review

- Carlos Z et al ; 89% persistent absence of RAIR
– poor surgical outcomes ; enterocolitis, incontinence and constipation
- Mishalany HG et al ; 10% normal RAIR, no relation with incontinence
- S.Ladi-Seyedian et al ; RAIR present 37% after surgery

Rationale

- Constipation and soiling - potential problems after surgeries.
- Assessment are done post op - DRE, and questions on symptoms – subjective.
- Anorectal manometry – more objective assessment
- PICSS as standardized questionnaires to correlates with the findings

Objectives

- **Primary objective**

1. To assess Anal Resting Pressure (ARP) and rectoanal inhibitory reflex (RAIR) for children with Hirschsprung disease post definitive surgery.

- **Secondary objectives**

1. To compare the anorectal manometry results of anal resting pressure (ARP) and RAIR for children with Hirschsprung disease, based on different types of definitive procedures (i.e. Duhamel and Non Duhamel procedure.)

2. To correlate the results of anorectal manometry with functional outcome; constipation and incontinence by using the Paediatric Incontinence and Constipation Scoring System (PICSS)

Methodology

- Prospective cross-sectional study
- Settings – University Malaya Medical Center and University Malaya Specialist Centre.

Inclusion and exclusion criteria

- **Inclusion criteria:**
 - Histology confirmation – resection margin, ganglionic
 - 6 months post-op – No complications
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- **Exclusion criteria:**
 - HD children with ongoing post-operative complications.



Stage 1

- Ethics committee approval by Medical Research Ethics Committee UMMC -(Ethics number: 2017621 – 5354)
- Recruitment via phone call, or followup clinic visit
- Selection ; inclusion and exclusion criteria



Stage 2

- PICSS questionnaires
- Date set for anorectal manometry



Stage 3

- Anorectal manometry performed
- Data analysis

Sample size estimation

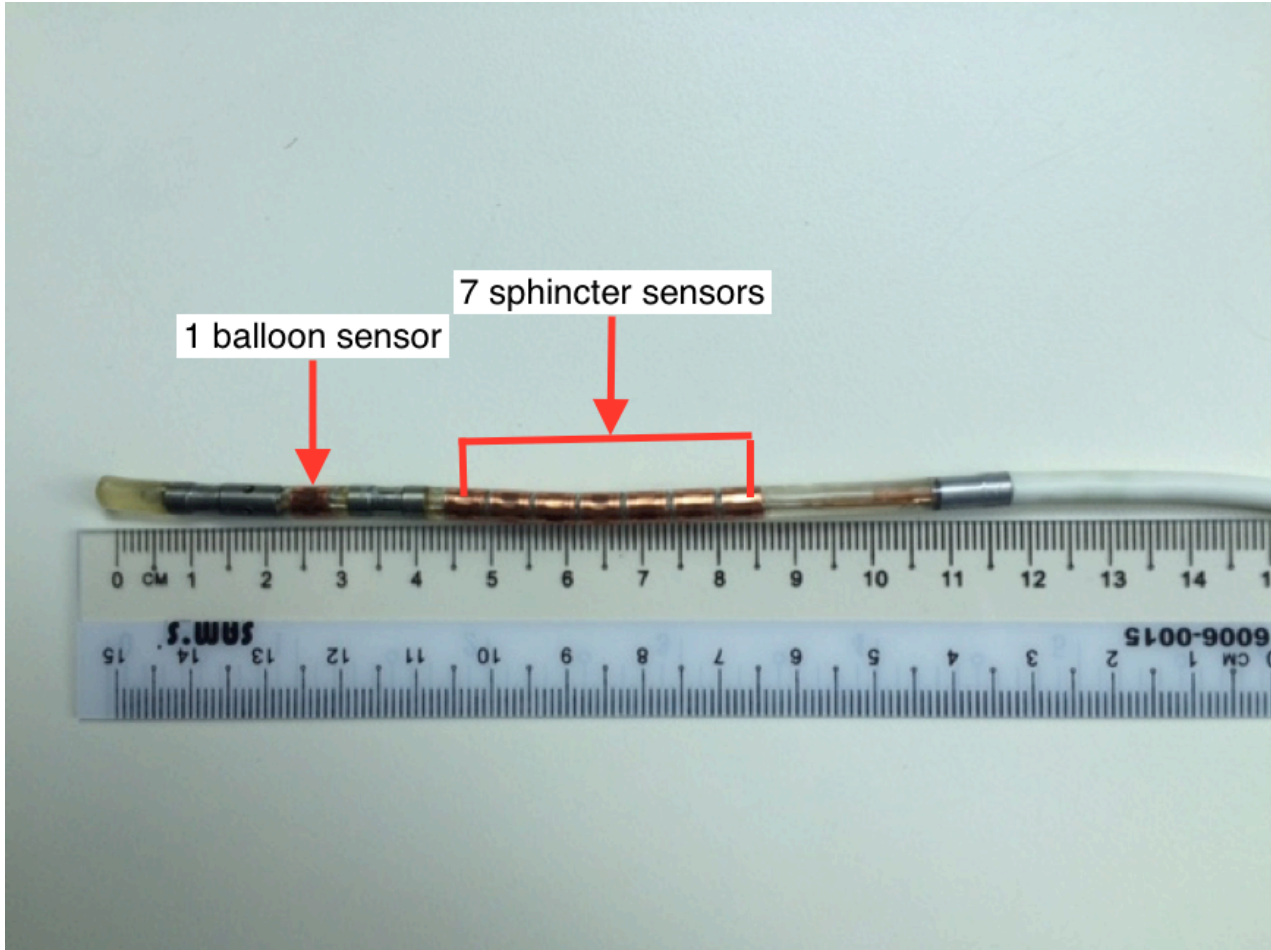
- Based on 2 proportions formula
- Estimated sample size = 30
- Summary of the manual calculations for sample size estimation based on two proportions formula
-
- $$N = \frac{P_1(1-P_1) + P_2(1-P_2)}{(P_1-P_2)^2} \times (z_{\alpha} + z_{\beta})^2$$
-
- N = sample size estimation
- P_1 (literature review) = proportion of abnormal BFS among abnormal resting pressure 0.64 (chung et al,2015) ²⁰
- P_2 (expert opinion) = proportion if normal PICSS among abnormal resting pressure = 0.2 (estimated)
-
- **$Z_{\alpha} = 1.96$ for $\alpha = 0.05$ (two tailed)**
- **$Z_{\beta} = 0.84$ for power of study = 80%**
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- $$N = \frac{0.64(1-0.64) + 0.2(1-0.2)}{(0.64 - 0.2)^2} \times (1.96 + 0.84)^2$$
-
- **N = 15 per group**
- **Total N = 30 (for 2 groups ; normal and abnormal)**

Measures outcomes

- PICSS - children who score below their age-specific lower limit (95 % confidence interval (CI) scores) are considered to have incomplete continence or constipated.
- Data were presented as **mean \pm standard deviation** for numerical variables while frequency (%) for categorical variables.
- **Fisher's Exact tests** are used to determine the significance of RAIR and ARP in correlation with the PICSS, comparing groups of different surgeries.
- **One-way Anova** was used to demonstrate the significance of RAIR and ARP.
- **Descriptive analysis** was used to calculate the constipation and incontinence mean scoring, and mean anal rectal pressure.
- **Independent t test** was used to compare mean anal rectal pressure between the Duhamel and Non Duhamel corrective surgery groups.

Materials

- Anorectal manometry
- High resolution, 8 channel anorectal catheter that consist of 7 sphincter sensors and 1 balloon sensor.
- It measures 4.2mm circumferentially, with 6 mm spacing between sensors.
- 8 pressure-sensing elements that detect pressure a length of 1mm in each 8 radially dispersed sectors.
- balloon (MSS– 3598) 150mls air



1 balloon sensor

7 sphincter sensors

- Normal anal resting pressure for children aged 1 month old to 1 year old is at 30-50mmHg, and for children above 1 year old is at 40-60mmHg .
- RAIR is considered 'present' if 2 successive rectal distensions produced pressure fall more than 5 mmHg below the low point of fluctuating waves. RAIR is considered absent if no pressure fall of > 5mmHg occurs or if a pressure fall occur, but not below the low point of the fluctuating waves

Paediatric Incontinence and Constipation Scoring

Paediatric Incontinence/Constipation Score	Incontinence			Constipation		
Does your child wear diapers/nappies during the day? Always Sometimes No	0	2.5	5	-	-	-
Does your child wear diapers/nappies during the night? Always Sometimes No	0	2.5	5	-	-	-
How often does your child open its bowels? Several times a day Once daily Less often	0	2.5	5	4	2	0
What does the stool usually look like? Watery Variable Thick	0	2	4	1	0.5	0
Can your child control the urge to open its bowels? Yes always Sometimes No	5	2.5	0	-	-	-
Can your child tell the difference between stool and air in the bowels? Yes always Sometimes No	4	2	0	1	0.5	0
Does your child regularly soil its underclothes by involuntarily passing small amounts of stool? Yes always Sometimes No	-	-	-	0	1	2
Does your child have trouble opening its bowels completely (incomplete emptying)? Yes always Sometimes No	-	-	-	0	1.5	3
Does your child feel pain when opening its bowels? Yes always Sometimes No	2	1	0	0	2	4
Does your child have to press hard to empty its bowels? Yes Normal No	2	1	0	0	2	4
Does your child have a lot of wind? Yes always Sometimes Never	-	-	-	0	1	2
Does your child suffer from constipation? Yes always Sometimes Never	-	-	-	0	2	4
Does your child have pains in the tummy? Yes always Sometimes Never	-	-	-	0	2	4
Total score:						

- Total of 82 patients was traced from year 2000- 2016 in UMMC.
- 37 responded , and 33 consented for the study

Result

Number of patients

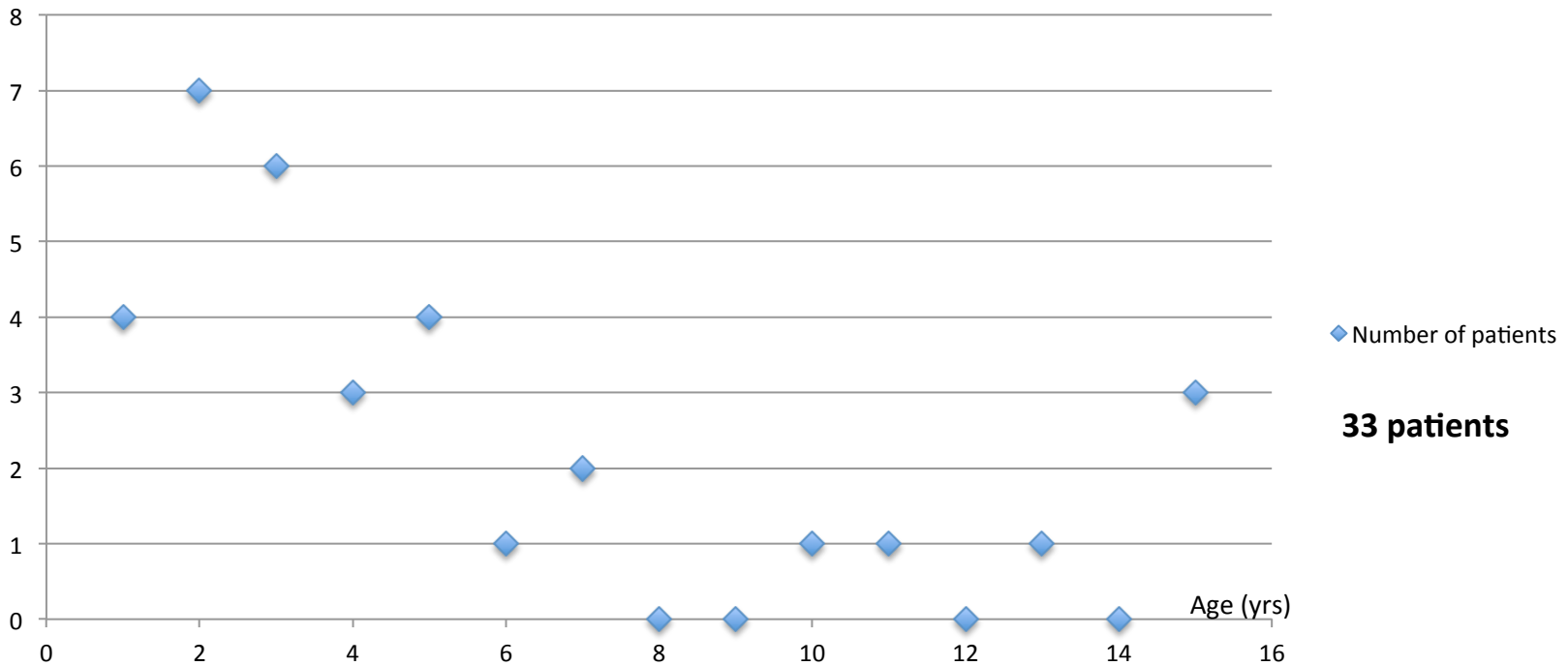


Chart 3.1 : Age and number of patients recruited

Variable	Frequency (%)	Mean ± SD
Age	10 months – 15 years old 2 months	5.0 ± 4.3
Gender	Male	28 (84.4)
	Female	5 (15.6)
Level of aganglionosis	Long segment HD (Sigmoid, Descending, Transverse and Ascending colon)	12 (36.4)
	Short (Rectosigmoid)	12 (36.4)
	Total Colonic Aganglionosis with ileal involvement	9 (27.2)
Approach	Open	21 (62.5)
	Laparoscopic	12 (37.5)
Type of operation	Swenson (open)	9 (27.3)
	Duhammel (open – 2) (Laparoscopic – 7)	9(27.3)
	Soave(open)	3 (9.1)
	Transanal pullthrough Open (2) Laparoscopic (10)	12 (36.3)
	Syndromic association	Trisomy 21

RAIR and ARP in HD children after corrective surgery

RAIR	Frequency (%)
Absent	20 (60.6)
Presence	13 (39.4)

ARP	Frequency (%)
Normal	24 (72.8)
Abnormal	9 (27.2)

Result of RAIR in respective of surgical types

Types of Surgery	RAIR present	RAIR absent	Total
Duhamel	1 (12.5%)	8 (87.5%)	9
Swenson	2(22.2%)	7(77.3%)	9
Soave	1(33.3%)	2(66.7%)	3
Transanal pullthrough (TAPT)	9(75%)	3(25%)	12

Result of ARP in respective of surgical types

Types of surgery	ARP normal	ARP abnormal	Total
Duhamel	6 (66.7%)	3(33.3)	9
Swenson	7(77.8%)	2(22.2%)	9
Soave	0(0%)	3(100%)	3
Transanal pullthrough (TAPT)	10(83.3%)	2(16.7%)	12

RAIR in HD children after Duhamel and Non-Duhamel surgeries

	RAIR present	RAIR absent	P value
Duhamel	1(11%)	8(89%)	<0.041
Non Duhamel	12 (50%)	12(50%)	

	Duhammel (n=8) Mean±SD	Non-duhammel (n=24) Mean±SD	Mean difference 95% CI	p-value
ARP	55.9 ± 23.9	56.6 ± 28.2	-0.71 (-23.04, 21.99)	0.950

Mean Anal Resting Pressure in HD children after Duhamel, Soave, Swenson and Transanal pullthrough surgery

Types of Surgeries (n)	ARP (mmHg)
Duhammel (8)	55.9 ± 23.9
Soave (3)	101.7 ± 45.4
Swenson (9)	53.6 ± 18.7
TAPT (12)	47.6 ± 19.4

Correlation Mean Anal Resting Pressure (ARP) with the PICSS result

Mean ARP (mmHg)	PICSS (n)			P - value
	Normal	Incontinence	Constipation	
Normal	47.4 ±14.9(16)	39.3±14.1 (6)	37.5±2.1 (2)	0.967
Abnormal	65.1±15.3 (5)	11.2±59.4 (2)	77.0±8.5 (2)	

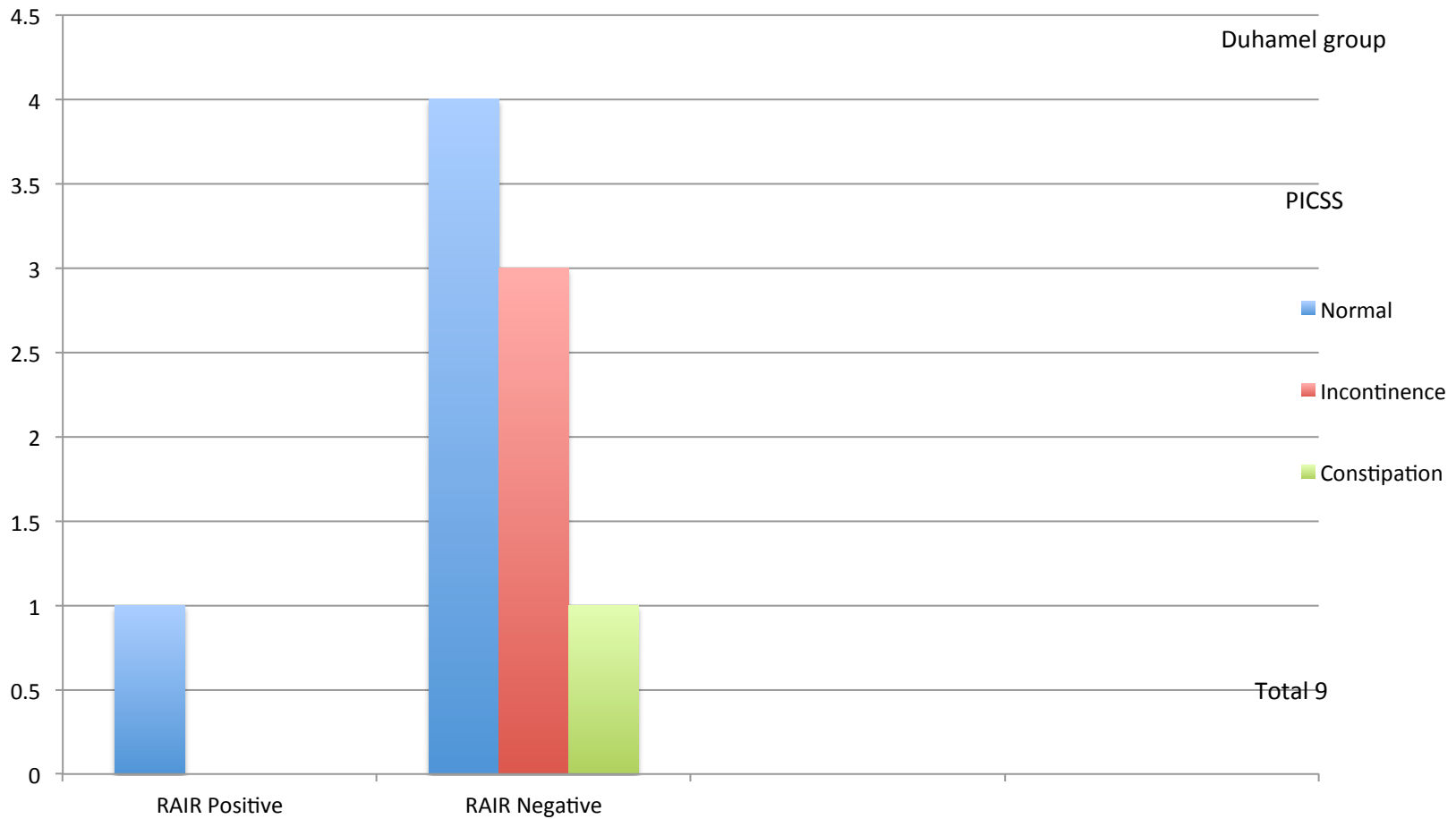
Correlation of RAIR and PICSS result.

PICSS	RAIR		P value
	Absent	Present	
Normal	11 (52.6%)	10 (76.9%)	0.191
Incontinence	5 (26.3%)	3 (23.1 %)	
Constipation	4 (21.1%)	0	

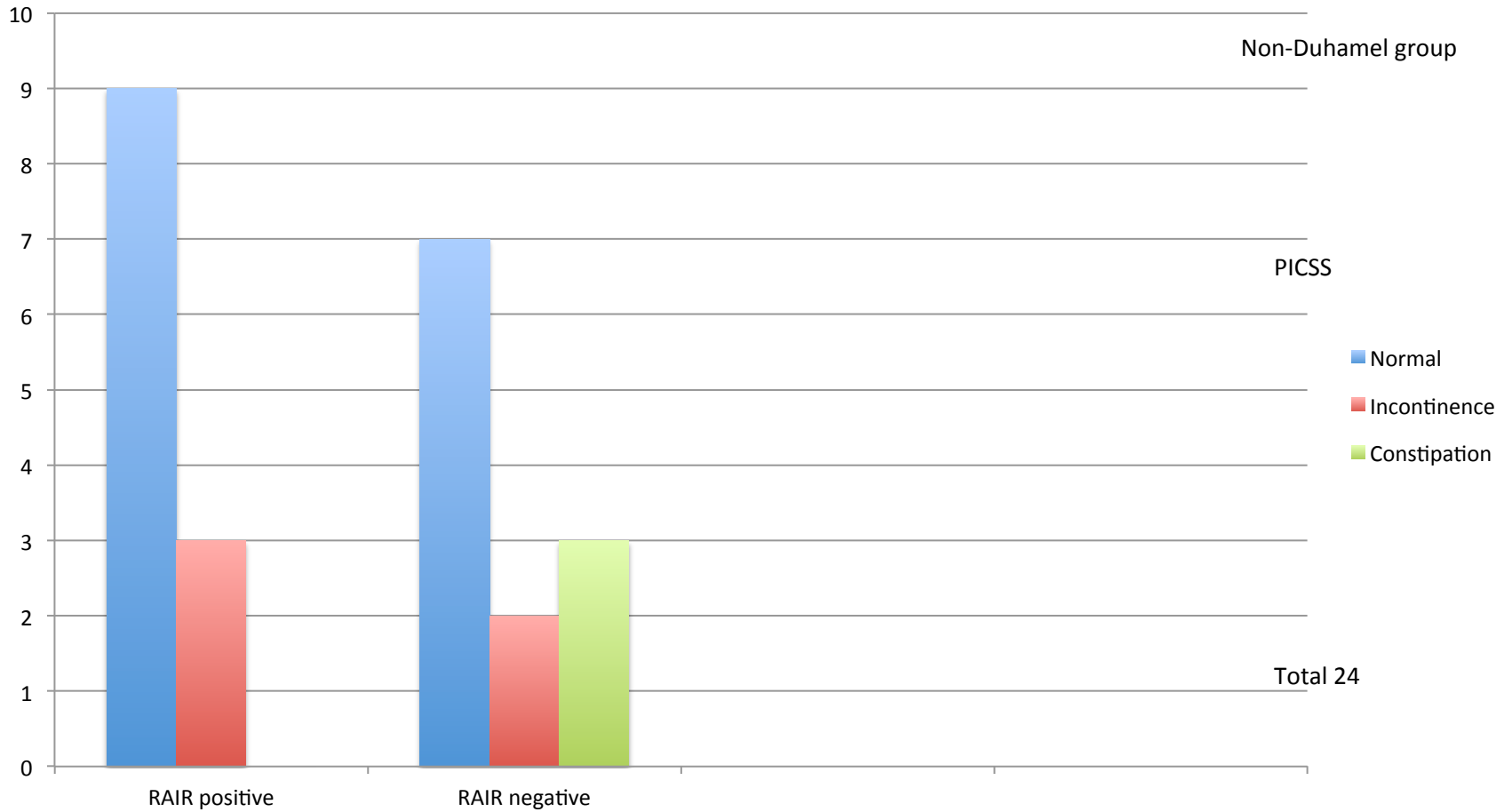
Comparing transanal pullthrough and Soave, with Duhamel and Swenson surgery with the RAIR

RAIR	Surgery		p-value
	Duham +swen(%)	Soave+TAPT(%)	
absent	15 (83.3)	5 (33.3)	0.005
present	3 (16.7)	10 (66.7)	

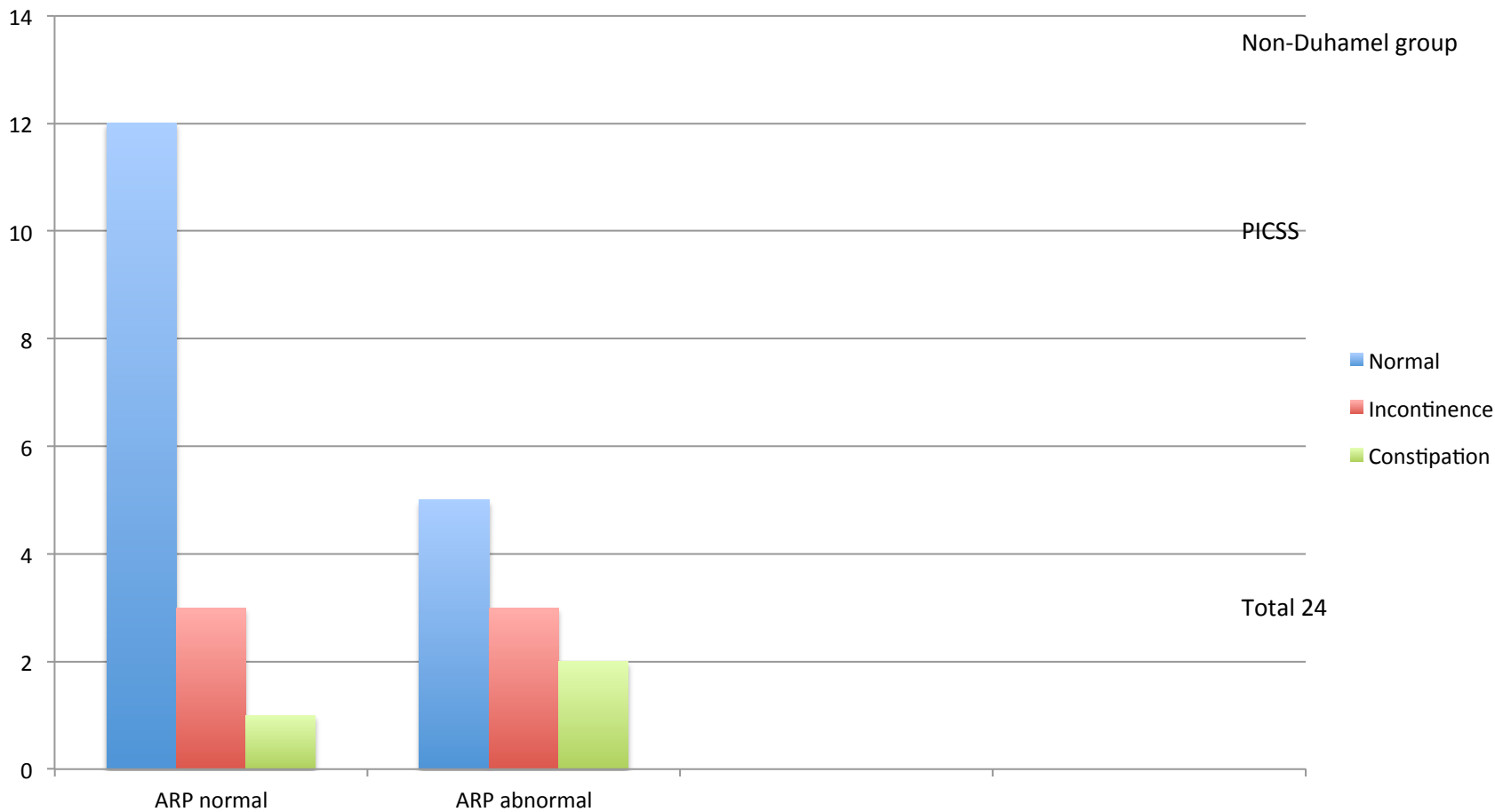
Distribution of HD children who underwent Duhamel surgery with their RAIR and PICSS result



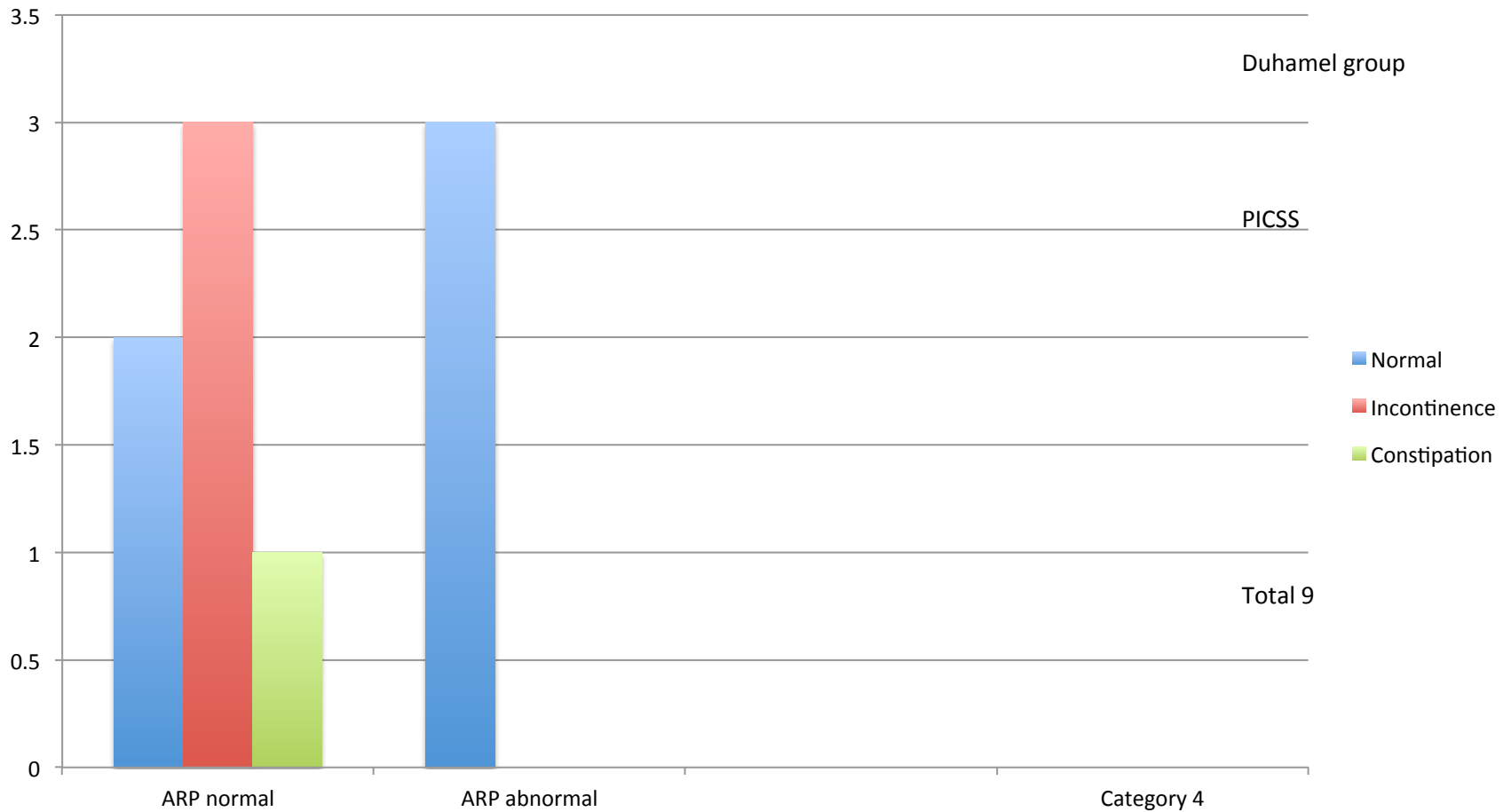
Distribution of HD children who underwent Non Duhamel Surgery with their RAIR and PICSS result



Distribution of HD children who underwent Non Duhamel surgery with their ARP and PICSS result



Distribution of HD children who underwent Duhamel surgery with their ARP and PICSS result



Discussion

- Significant difference of the RAIR result, comparing HD children who underwent Duhamel surgery and Non Duhamel surgery.
- This can be attributed by the preservation of more achalactic internal sphincter and residual aganglionic rectal pouch which was above retrorectum pullthrough bowel compared to other types of surgery

Discussion

- In Duhamel surgery and Swenson surgery, risk of pelvic injury, and nerve erigentes are higher during the transanal dissection and mobilization of the bowel, compared to transanal pullthrough, in which the aganglionic segment at the muscular cuff in the rectum are left untouched.
- Thus, this may explain the significant difference of the RAIR result when comparing Duhamel and Swenson, to Transanal pullthrough and Soave(p value = 0.005).

- Soave surgery has the highest mean anal resting pressure 101.7 ± 45.4 mmHg, relatively much higher compared to other groups of corrective surgical methods.
- This result may be explained by the seromuscular tunnel of rectum through which the ganglionic colon is pulled through after denuding the rectal mucosa.

- The current study result showed that, RAIR doesn't significantly influence the functional outcome on my PICSS result (p value = 0.191)

- This study showed that with the presence of RAIR post HD surgery, it will ensure good bowel continence, while the absence of RAIR may not result in a definite poor bowel continence

Limitations to this study

- The various techniques of corrective surgeries for HD in my centre were performed by 4 different consultants. Their respective assessment could be partially influenced by some slight different surgical techniques, despite the same principle of operation.

Conclusion

- The demographic data in my study are mostly comparable to other international studies.
- Even though RAIR is important in bowel emptying, the absence of RAIR post surgery does not always result in constipation and incontinence.
- Despite the limitation this studies, transanal pullthrough has demonstrated to have a better outcome in comparison to other procedures performed for HD children at our center.
- Anorectal manometry and PICSS can be recommended as an adjunct to assess the trend of continence for children with Hirschsprung disease, after definitive surgery.

- End

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