

## Validity of Transition Zone in Contrast Enema to Detect the Level of Aganglionic Segment in Patients with Hirschsprung's Disease

Huda Hameed Hameedy<sup>1</sup>, Shatha Abdulameer Alkawaz<sup>2</sup>

### ABSTRACT:

#### BACKGROUND:

Hirschsprung's disease is a functional intestinal obstruction resulting from congenital absence of ganglion cells. The water-soluble contrast enema is the radiologic examination that can be used to assist in diagnosis the level of aganglionosis. A "transition zone" between a narrow distal aganglionic bowel segment and grossly distended proximal ganglionated bowel is considered the most accurate radiologic feature.

#### OBJECTIVE:

The aim of the study is to determine the validity of contrast enema in detecting the level of aganglionic segment in patients with HD

#### MATERIALS & METHOD:

Over 2 years period (2018-2020), 46 patients were diagnosed as HD, all of them had preoperative contrast enema findings regarding the level of aganglionic bowel & were compared with operative & pathology reports. All contrast enema film reviewed by the same radiologist.

#### RESULTS:

This study showed that 40 of 46 (86%) HD's patients had the same level of TZ on contrast enema & histopathology. In 34 of 40 (85%) of patients with rectosigmoid HD, the RTZ and pathological level of aganglionic bowel were concordant. The concordance rate between radiographic & pathologic TZ in-patient with splenic HD was 100%

#### CONCLUSION:

The level of TZ on contrast enema can be affected by the presence of fecal impaction & when there is no adequate dilatation of a ganglionic segment in young infants.

**KEYWORDS:** Hirschsprung's disease, Transition zone, Contrast Enema, aganglionic.

<sup>1</sup> Ministry of Health, Shrine Karbala Health Directorate, Al-Hassan Al-Mujtaba Teaching Hospital.

<sup>2</sup> Consultant Pediatric Surgeon, Baghdad University, College of Medicine., Baghdad, Iraq

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### INTRODUCTION:

Hirschsprung's disease, also known as "congenital megacolon," is characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the intestine. The first known description of this condition was by ancient Hindu surgeons in the Shushruta Samheta, and the first descriptions in the modern medical literature were from the 17th century<sup>(1)</sup>. In 1887, Harald Hirschsprung, a pediatrician from Copenhagen, described two cases of the condition that ultimately bore his name. At that time, most children with congenital megacolon died from malnutrition and enterocolitis. As the underlying pathologic basis of the disease was unknown, surgeons removed the massively dilated proximal bowel and created

a colostomy. Attempts at reanastomosis were uniformly unsuccessful<sup>(2)</sup>

In H.D, the underlying pathophysiological feature is functional obstruction caused by a narrowed colon that hinders the propagation of peristaltic waves due to the absence of parasympathetic intrinsic ganglion cell<sup>(3)</sup>. The appropriate diagnostic approach may vary, depending on the age of the patients and the presenting clinical pictures. After a careful history and physical examination, the diagnostic steps may include radiographic studies, anorectal manometry, and rectal biopsy.<sup>(4)</sup>

Hirschsprung disease on contrast enema diagnosed by transition zone (a calibre change between

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a narrowed or normal-size distal aganglionic segment and a dilated proximal ganglionic bowel), Serrations(irregular contractions of the wall of the aganglionic segment), spasm and/or mucosal irregularity, Retained contrast agent on delayed radiographs& theRectosigmoid ratio<sup>(5)</sup>. The definitive diagnosis of HD rests on histological review of rectal biopsy. The biopsy is taken at least 2 cm above the dentate line on the posterior wall to minimize the risk of perforation. Carefully examine biopsy specimens for the presence or absence of ganglion cells in the submucous plexus (suction rectal biopsy) or myenteric plexuses (transanal wedge resection), Calretinin is helpful in ruling out HD by staining the ganglion cells and intrinsic nerve tissue. <sup>(6)</sup>

The aim of the study is to determine the validity of transition zone on contrast enema in detecting the level of aganglionic segment in patients with HD

### PATIENT AND METHOD:

The study include 46 patients have been conducted in this study, the patients age ranges from 2 months to 32 months and 37 patients were boys and 9 were girls.All patients were known to have rectal biopsy-confirmed HD that show absence of ganglion cell and nerve hypertrophy preoperatively. Wholly patients with positive transition zone on contrast enema were included in this study. As well, patients had pull through procedures at our pediatric surgery department.

Furthermore, the whole patients received one CE at our institution before operation. C.E was water-soluble contrast slowly injected into colon via non-inflated ballon catheter until total colon was clearly visible. All contrast enema reports provided by the same pediatric radiologist. Review of the pathology and surgical notes for the extent of aganglionic bowel. This was determined by the site that a full-thickness biopsy showed normal innervated bowel.

Data collecting form in this study were included the Age, Gender, History of delayed passage of meconium, Contrast Enema, Rectal Biopsy.

### Study design and setting

This is a prospective study conducted at pediatric surgery department in children welfare teaching hospital – medical city / Baghdad – Iraq in a 24 months period from 1<sup>st</sup> October 2018 to 1<sup>st</sup> October 2020.

### RESULTS:

This study enrolled a 46 pediatric patient, who attended pediatric surgery department at pediatric welfare teaching hospital- medical city, With HD & diagnosed as H.D by preoperative C.E and rectal biopsy.

The age range from 2-32months with mean  $\pm$ SD (1.04  $\pm$ 7.06), At time of H.D diagnosis 11 of 46(23.9%) patients were younger than 3 months, 35 Of 46 (76.1%) from 3 months up to 32 months.

Table 1: Show age distribution of H.D patients.

		gender		Total
		Male	Female	
age	Less than 3 months	9(19.6%)	2(4.3%)	11(23.9%)
	Older than3 months	28(60.9)	7(15.2%)	35(76.1%)
Total		37(80.5%)	9(19.5%)	46(100%)

This study included 37(80.4%) male ,& 9(19.6%) were female( figure 1)

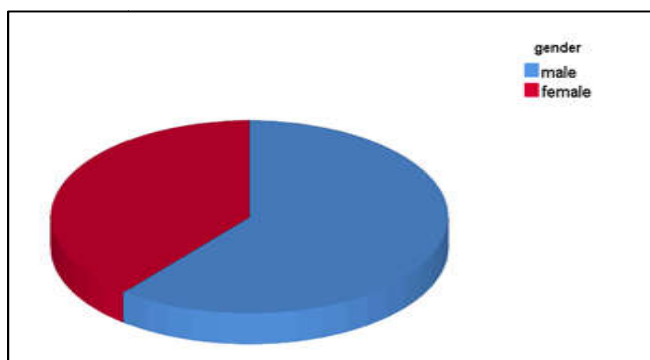
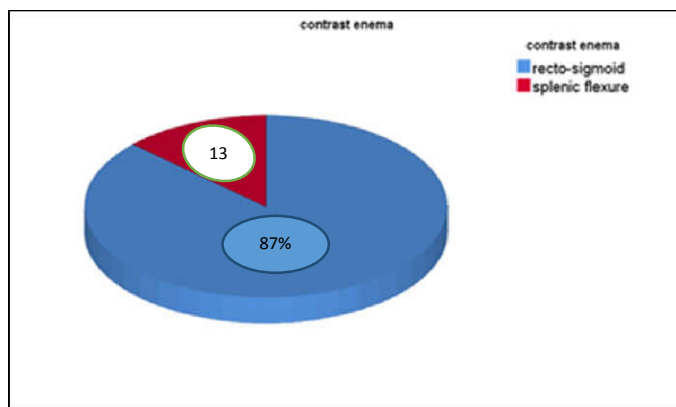


Figure 1: Gender distribution of participant.

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On C.E. 40 of 46(87%) patients were rectosegmoid level H.D on & 6 of 46(13%)patients were splenic level H.D ( figure 2)



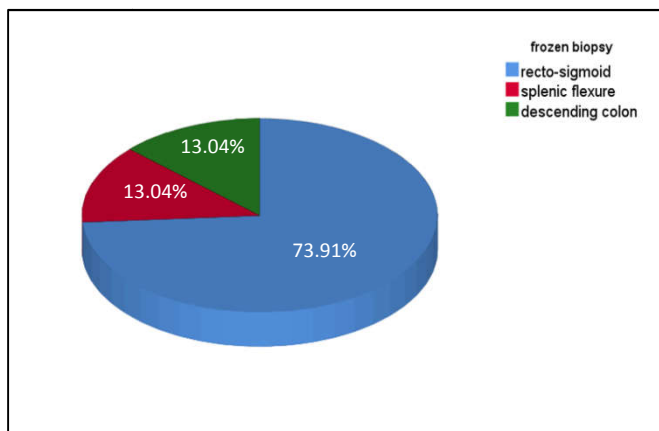
**Figure 2:** Shows the level of an aganglionic segments on C.E.

On C.E 32 of 37 (86.5%) boys were rectosegmoid level H.D & 5 of 37(13.5%) boys were splenic H.D, 8 of 9 (88.9%) girls were rectosegmoid H.D and 1of 9(11.1) were splenic level H.D.

**Table 2: Contrast enema results according to gender.**

		Gender		Total
		male	female	
contrast enema	recto-sigmoid	32(69.5%%)	8(17.4%)	40( 86.9%)
	splenic flexure	5(10.9%)	1 (2.2%)	6(13.1%)
Total		37(80.4%)	9(19.6%)	46 (100%)

Frozen biopsy approach showed that 34 (73.91%) have descending colon, All the explained data have recto-sigmoid aganglionic segment, presented in table and figure 3 6 (13.04%) have splenic flexure and 6, (13.04%)



**Figure 3:** Level of aganglionic segments on frozen biopsy.

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- The overall concordance rate of contrast enema and frozen biopsy in detecting the level of aganglionosis of H.D was 86% .
- The overall disconcordace rate was 14% between C.E and frozen biopsy
- Patients with RTZ in rectosegmoid 34 of 40(concordance rate 85%) had matching the level of aganglionosis, wheres 6 of 40 (15% disconcordance rate) had aganglionosis level at descending colon.
- 4 of 46(8%) of patient with rectosegmoid H.D were younger than 3 months &, the RTZ on C.E didn't match level of aganglionosis on frozen biopsy.
- 2 of 46(4%) of patients with rectosegmoid H.D were older than 3 months didn't correctly matching the level of aganglionosis.
- 6 patients with RTZ in splenic flexure had matching the level of aganglionosis (concordance rate 100%), older than 3 months.

The strength of association and prediction ability measured using lambda test, the two diagnostic measures are depended on each other, and performing contrast enema study will reduce incorrect predication by 83.3%. Kappa test was 0.61. h

**Table 3: Correlation of contrast enema and frozen biopsy.**

		Frozen biopsy			total	P value	Lambda test	Kappa test
		rectosegmoid	Splenic flexure	Descending colon				
Contrast enema	rectosegmoid	34	0	6	40	0.00*	0.883 P= 0.01*	0.617 P= 0.00*
	Splenic flexure	0			6			
total		34	6	6	46			

\*Chi-square test is significant if P value  $\leq 0.05\%$

The sensitivit100%, specificity50%, positive predictive value85%, negative predictive value1% of contrast enema that detect recto-sigmoid segment.

### DISCUSSION:

In this study we used contrast enema to evaluate the level of aganglionic segment in order to compare it with frozen section biopsy.

In our study male to female ratio was 4:1<sup>(7)</sup>.

In this study, we found the overall concordance rate between RTZ and pathological results was 86% which was nearly similar to Chen x et al<sup>(8)</sup> 86.4% , while Proctor et al<sup>(9)</sup> was 89% & Jamieson et al<sup>(10)</sup> was 90.6% as our study and Chen x et al<sup>(8)</sup> C.E film was reviewed by one radiologist, while the other studies Proctor et al<sup>(9)</sup> & Jamieson et al<sup>(10)</sup>

C.E film was reviewed by more than one radiologist , multiple readers minimize the interpretation variability & bias<sup>(11)</sup>

The overall disconcordance rate in our study was 6 of 46 (13%) occurred 5 of 46 (11%) of disconcordance rate in infant younger than 3 months age with rectosegmoid H.D ( mean age 2.4 months ) , with 1 of 46(2%) of disconcordance in patients with age from 3 months up to 12 months with no disconcordance in patients older than 12 months , while the over all of disconcordance in

Proctor et al<sup>(9)</sup> was 21% ,11% of disconcordance rate below one month age & 8% disconcordance in age from 1 month-12 months with no disconcordance between C.E & pathological level of aganglionosis in older than 12 months, Chen et al<sup>(8)</sup> 31% of disconcordance in patients younger than 3 months & 14% of disconcordance in older than 3 months , higher disconcordance rate in younger infant as younger infant with H.D have a shorter course of disease compared with older children possible reason that colonic dilatation needs time to develop before it can detected on contrast enema and thus might not be visible in first 3 months of life chenx et al<sup>(8)</sup> & the RTZ detected radiologically based on caliber change between normally innervated colon and aganglionic segment.<sup>(9)</sup>

In rectosegmoid group concordance rate was 85%, which was nearly similar to Chen x et al<sup>(8)</sup> concordance rate with rectosegmoid group was 88.5% & proctor et al<sup>(9)</sup> 89% concordance rate.

In our study , the concordance rate of splenic group was 100% with no disconcordance , while disconcordance rate for long segment H.D proctor et al<sup>(9)</sup> was 11% , as in our study patients with H.D with mean age 8 months ( 3 months -9 months),while Proctor et al<sup>(9)</sup> patients with long H.D were younger than one month with there was

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no fecal impaction on contrast enema as faecal impaction filling proximal ganglionic segment with retained stool make RTZ appear more distal.<sup>(9)</sup> Kappa test was 0.617 ( p 0.05) , with moderate agreement between C.E & histopathology as with Chen x et al<sup>(8)</sup> was 0.692 ,in contrast to Haikal et al<sup>(12)</sup> was 0.198 slight agreement

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