The Accuracy of Transitional Zone and Aganglionic Segment in Hirschsprung Disease

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Background: *The goal of surgical management for Hirschsprung disease is resection of the affected aganglionic bowel. Determining the level of aganglionosis is very important for transanal endorectal-pullthrough.*
Objective: *The aim of this study was to determine the accuracy of intra-operative finding transitional zone compared to the radiologic transitional zone and the histological extent of aganglionic bowel.*
Material and Method: *Twenty-six children with underlying Hirschsprung disease, operated at the pediatric surgery unit, Siriraj Hospital, during the period from August 2012 to September 2015 were included in this prospective study. Patients with colostomy and those underwent Redo pullthrough were excluded. Pre-operative contrast studies were identified for radiologic transitional zone in standard anatomic sectors (rectum, recto-sigmoid, descending, splenic flexor, transverse, hepatic flexor, ascending colon). The operative transitional zone was identified and full thickness colonic tissue samples were sent for histological examination from transitional zone and every 2 centimeters proximal to transitional zone until the ganglionosis was found. The radiologic transitional zones were correlated to operative findings and compared to the pathological extent of aganglionic bowel.*
Results: *Twenty-one cases of short segment and 5 cases of long segment Hirschsprung disease showed correspondence of radiologic and operative transitional zone. Only 4 cases were proved by frozen section that the operative transitional zone was the level of ganglionosis, whereas the other 22 found that the level of ganglionosis were more proximal. All of the true ganglionic segments were located within 10 centimeters proximal to the operative transitional zone.*
Conclusion: *The operative transitional zone consistently correlated with radiologic information while the true ganglionic segment would be located within 10 centimeters proximally. The study result benefits in predicting the location of ganglionic segment pre-operation and during the operation.*

Keywords: *Hirschsprung disease, Transitional zone, Aganglionic segment*

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Hirschsprung disease is a developmental disorder characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine. Because these cells are responsible for normal peristalsis, patients with Hirschsprung disease present with functional intestinal obstruction at the level of aganglionosis.

For the children with a clinical picture suggesting distal bowel obstruction, the first step in the diagnostic pathway is water-soluble contrast enema[1-4]. The pathognomonic finding of Hirschsprung disease on contrast enema is a transition zone between the normal and aganglionic bowel[5]. The accuracy of contrast enema in making the diagnosis of Hirschsprung disease has been examined previously. In 1984, Rosenfield et al found the radiologic transitional zone to match the level of aganglionosis identified at the time of surgery in all 27 patients compared[1]. However, several investigators have reported cases in which the radiological and pathological transitional zones differ[6]. In 1991, Grahame H et al studied the usefulness of barium enema in the evaluation of the extent of aganglionosis. They found that the level of transitional zones being accurately identified in only 8 out of 13 cases (53% error rate in determining the level of transition[7]). In 2012, Muller CO et al described the correlation between the level of radiologic transitional zone on contrast enema and that of histological confirmed aganglionosis remains low[3].

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contrast enema is used for the diagnosis and evaluation of the extent of Hirschsprung disease, in preparation for the definitive surgery: transanal pullthrough for recto-sigmoid disease and laparotomy assisted for long-segment diseases requiring more proximal colonic mobilization. Transanal approach offers excellent cosmetic and minimizes postoperative pain[8-10]. The choice of operative technique is based in part on the anticipated level of aganglionosis as determined by the location of transitional zone on preoperative contrast enema. The aim of the presented study was to determine the accuracy of intra-operative finding transitional zone compared to the radiologic transitional zone and the histological extent of aganglionic bowel.

Material and Method

Forty-seven children with underlying Hirschsprung disease were treated at pediatric surgery unit, Siriraj Hospital, for definitive surgery during the period from August 2012 to September 2015. Patients with colostomy and those who underwent Redo pullthrough were excluded; altogether 26 patients were included in this prospective study. All patients had clinical of Hirschsprung disease and underwent preoperative contrast enema to evaluate the level of aganglionosis.

There were 18 boys and 8 girls, ages ranged from 1 month to 12 years old at the time of diagnosis, thirteen patients presented in the first month of life.

All pre-operative contrast studies were identified for radiologic transitional zone in standard anatomic sectors (rectum, recto-sigmoid, descending, splenic flexor, transverse, hepatic flexor, ascending colon). For each patient, the operative transitional zone was grossly identified, and full thickness colonic tissue samples sent for pathological examination (frozen section) starting at transitional zone and every 2 centimeters proximally until the ganglion cells were found. The radiologic transitional zone was correlated to operative finding and compared to the histological confirmed level of aganglionic bowel.

Ethical approval

This study received the approval of Siriraj Institutional Review Board.

Statistical analysis

The radiologic and operative transitional zone was compared in terms of accuracy. The distance from operative transitional zone to the level of histological confirmed ganglionosis was analyzed using mean value as the average (n = 26).

Results

The group of patients included 18 boys and 8 girls. The operation was performed using transanal endorectal pullthrough in 17 patients. Eight patients underwent laparotomy assisted transanal endorectal pullthrough, 6 had long segment disease and 2 with operative transitional zone in recto-sigmoid but the level of ganglionosis was further proximal approximately 8 and 10 centimeters respectively (Table 1).

Out of 26 patients, 12 patients had transitional zone at upper rectum, 9 patients at recto-sigmoid and 5 patients had long segment disease. Concordance between radiologic transitional zone and operative transitional zone occurred in all patients (Table 2).

Of the 26 patients, only 4 cases were proved by frozen section that the operative transitional zone was the level of ganglionosis. The other 22 cases found that the level of ganglionosis were more proximal. All of them confined within 10 centimeters from the operative transitional zone (Table 3). The mean distance between the operative transitional zones to the level of ganglionosis was 4 centimeters.

Discussion

Radiologic transitional zone occurs at the transition from proximal dilated bowel to the narrow aganglionic distal bowel. The accuracy of contrast enema in making diagnosis of Hirschsprung disease has been examined previously. Although the presence of a radiologic transitional zone on contrast enema may aid in the diagnosis of Hirschsprung disease, the correlation between the location of the radiologic transitional zone and the histological level of aganglionosis has not been well studied. Our data showed correspondence of radiologic transitional zone and operative transitional zone. Conversely the majority of cases found the level of ganglionosis to be more proximal to the operative transitional zone. The mean distance was 4 centimeters and 10 centimeters from transitional zone was the highest of ganglionic level.

The goal in performing the definitive surgical management for Hirschsprung disease is to remove the aganglionic bowel and reconstruct the intestinal tract by bringing the normally innervated bowel to the anus. Transanal endorectal pullthrough is the definite operation for Hirschsprung disease in Siriraj Hospital. Transanal approach offers excellent cosmetic but it can be done only for the recto-sigmoid lesion. The decision

Table 1. Patient demographic data

Variables	n = 26 n (%)
Sex	
Male	18 (68%)
Female	8 (31%)
Age at operation	
Younger than 1 month	13 (50%)
1 to 6 months	7 (27%)
7 to 12 months	-
Older than 1 year	6 (23%)
Manifestation	
Delay pass meconium	8 (27.5%)
Constipation	10 (34.5%)
Abdominal distension	11 (38%)
Surgery	
Transanal pullthrough	17 (65%)
Laparotomy-assisted	8 (31%)
Laparoscopic-assisted	1 (4%)

Table 2. Radiologic and intraoperative transitional zone

Radiologic	Intraoperative	No.	%
Upper rectum	Upper rectum	12	46
Rectosigmoid	Rectosigmoid	9	35
Descending colon	Descending colon	-	-
Splenic flexor	Splenic flexor	5	19
Transverse colon	Transverse colon	-	-
Ascending colon	Ascending colon	-	-

Table 3. Ganglionic level from transitional zone

Ganglionic level	No.	%
At transitional zone	4	15
2 centimeter	4	15
4 centimeter	10	39
6 centimeter	5	19
8 centimeter	2	8
10 centimeter	1	4

making between transanal approach only or laparotomy assisted depend on the level of aganglionosis. The results from this study raise awareness in determining the innervated bowel intra-operatively, which histology may not correspond with radiologic transitional zone identified pre-operatively. The lesion that cannot be reached by transanal approach only should be operated through the abdominal cavity first, to prevent non benefit forceful traction of anal canal. To prevent retaining aganglionic bowel segments, it is safe to say that the bowel at least 10 centimeters more proximal to the transitional zone is likely to be innervated and a suitable segment for the pullthrough procedure. Accordingly, a frozen section is required whenever in doubt.

Conclusion

The operative transitional zone consistently correlated with radiologic information while the true ganglionic segment would be located within 10 centimeters proximally. This result may benefit in predicting the location of ganglionic segment pre-operation and during the operation.

What is already known on this topic?

The pathognomonic finding of Hirschsprung disease on contrast enema is a transition zone between the normal and aganglionic bowel. Several investigators have reported that correlation between the level of radiologic transitional zone on contrast enema and that of histological confirmed aganglionosis remains low. The exact distance of ganglionosis from the transitional zone is unknown.

What this study adds?

The results from this study raise awareness in determining the innervated bowel intra-operatively, which histology may not correspond with radiologic transitional zone identified pre-operatively. The true ganglionic segment was located within 10 centimeters proximally. To prevent retaining aganglionic bowel segments, it is safe to say that the bowel around 4 to 10 centimeters more proximal to the transitional zone is likely to be innervated and a suitable segment for the pullthrough procedure.

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Potential conflicts of interest

None.

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การศึกษาเพื่อดูความแม่นยำระหว่างระดับของรอยคอดกับระดับของปมประสาทบนลำไส้ในผู้ป่วยที่วินิจฉัยลำไส้ใหญ่โป่งพองแต่กำเนิด

มงคล เลาหเพ็ญแสง, สุคนธา ตรีฤทธิวิไล

ภูมิหลัง: เป้าหมายในการรักษาโรคลำไส้โป่งพองแต่กำเนิดคือการตัดส่วนลำไส้ที่ไม่มีปมประสาทออกทั้งหมด ดังนั้นการประเมินความยาวของลำไส้ที่ไม่มีปมประสาทนั้นจึงมีความสำคัญ การศึกษานี้จัดทำขึ้นเพื่อดูความแม่นยำ ระหว่างตำแหน่งของลำไส้ที่ไม่มีปมประสาท โดยดูจากรอยคอดของภาพถ่ายสวนสารทึบรังสีเทียบกับตำแหน่ง ของลำไส้ที่พบปมประสาทในระหว่างทำการผ่าตัด

วัสดุและวิธีการ: ทำการศึกษาในผู้ป่วยเด็กที่เข้ารับการผ่าตัดเนื่องจากเป็นลำไส้โป่งพองแต่กำเนิดที่โรงพยาบาลศิริราช ตั้งแต่ เดือนสิงหาคม พ.ศ. 2555 ถึง เดือนกันยายน พ.ศ. 2558 โดยผู้ป่วยเด็กที่ยกลำไส้มาถ่ายทางหน้าท้องหรือเคยได้รับผ่าตัดลำไส้เพื่อรักษาภาวะลำไส้โป่งพองแต่กำเนิดมาแล้วจะไม่นำมารวมในการศึกษาครั้งนี้ รวมผู้ป่วยทั้งสิ้น 26 ราย ผู้ป่วยทุกรายจะได้รับการส่งตรวจภาพถ่ายสวนสารทึบรังสีและบันทึกตำแหน่งรอยคอดตามตำแหน่งกายวิภาคของลำไส้ จากนั้นในระหว่างการผ่าตัดจะทำการตรวจดูลำไส้ในระหว่างผ่าตัดและบันทึกตำแหน่งรอยคอดและทำการตัดชิ้นเนื้อส่งตรวจทางพยาธิวิทยาเพื่อหาปมประสาทที่ตำแหน่งรอยคอดและระยะทุก ๆ 2 เซนติเมตร เหนือจากรอยคอดขึ้นไปจนกว่าจะพบตำแหน่งที่มีปมประสาท

ผลการศึกษา: ผู้ป่วยทุกรายที่เข้าร่วมการศึกษาพบว่ามีตำแหน่งของรอยคอดภาพถ่ายสวนสารทึบรังสีและลำไส้ในระหว่างการผ่าตัดในตำแหน่งเดียวกัน และพบว่ามีผู้ป่วยเพียง 4 ราย ที่ตำแหน่งรอยคอดนั้นเป็นตำแหน่งที่เริ่ม มีปมประสาทจริงจากผลตรวจชิ้นเนื้อในขณะที่ผู้ป่วย 22 รายนั้นพบว่าตำแหน่งที่เริ่มมีปมประสาทจะอยู่สูงขึ้นไปภายในระยะห่าง 10 เซนติเมตร จากตำแหน่งรอยคอดโดยระยะห่างเฉลี่ยอยู่ที่เหนือจากรอยคอดขึ้นไป 4 เซนติเมตร

สรุป: จากการศึกษานี้พบว่ารอยคอดจากภาพถ่ายสวนสารทึบรังสีกับลำไส้ที่พบในระหว่างผ่าตัดนั้นเป็นตำแหน่งเดียวกัน ส่วนตำแหน่งที่ลำไส้เริ่มมีปมประสาทนั้นจะอยู่สูงขึ้นไปภายในระยะห่าง 10 เซนติเมตร เหนือรอยคอด