Hirschsprung Disease and Contrast Enema: Diagnostic Value of Simplified Contrast Enema and Twenty-Four-Hour-Delayed Abdominal Radiographs

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ABSTRACT

Hirschsprung disease is common in children and is generally diagnosed by contrast enema. The objective of our study was to determine whether radiation time can be decreased and an adequate diagnostic sensitivity and specificity can be obtained using a simplified contrast enema as compared to using a conventional contrast enema. Moreover, the study describes the diagnostic value of a 24-hour-delayed radiograph.

We reviewed the data for 198 consecutive children who had received a contrast enema between January 2009 and December 2010 in our hospital for suspicion of Hirschsprung disease. Of the 198 children, 18 were excluded because 24-hour-delayed abdominal radiographs were not available, they had previously undergone a gastrointestinal tract surgery, or they had previously received a contrast enema. We reviewed the radiographs taken during contrast enema examination of the remaining 180 children. The time taken for the procedure was recorded for 15 patients. Rectal suction biopsy was performed in 45 patients. The results obtained in all cases were analyzed.

The results of conventional and simplified contrast enema examinations showed markedly high correlation in all cases, except for 1 case. The sensitivity of the 24-hour-delayed radiograph ranged from 60.87% to 98.65% and its specificity ranged from 8.92% to 35.67%, depending on different diagnostic rules. The radiation time required for simplified contrast enema was approximately 33.87% of that required for conventional contrast enema.

Similar diagnostic values were obtained using both simplified and conventional contrast enema examinations; however, the radiation time required for simplified contrast examination was significantly lower. The 24-hour-delayed abdominal radiograph added to the diagnostic value; however, its specificity was very low.

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Constipation is a common problem in children, and approximately 10% of children with constipation have an organic disorder. Hirschsprung disease (HD) is a well-known organic disorder with an incidence of approximately 1 per 5000 live births. In general, there are 3 diagnostic tests for HD, that is, contrast enema, anorectal manometry, and rectal suction biopsy. Of these tests, contrast enema has the lowest sensitivity (65–80%) and specificity (66–100%), while rectal suction biopsy has the highest sensitivity (91–100%) and specificity (97–100%). To date, the gold standard for diagnosing HD is full-thickness biopsy [1, 2]; however, some scholars also regard rectal suction biopsy as the gold standard [3].

The generally accepted theory for HD is that the
proximal-to-distal migration of vagal neural crest cells is arrested. Therefore, the affected segment of the colon is the region that starts from where the vagal neural crest cells are not found and extends up to the end of the bowel. Skip-segment HD is controversial and extremely rare, with only 24 cases reported in the literature [4]. In approximately 90% of HD cases, the transition point is located in the rectosigmoid colon; this condition is known as “short-segment aganglionosis [5].” Conversely, in “long-segment aganglionosis,” the affected region extends from a more proximal segment of the bowel up to the anorectum.

In our hospital, the use of contrast enema for diagnosing HD is delayed until the colon is fully distended, after which, a 24-hour abdominal radiograph of the patient is taken. The 3 main objectives of our study are as follows: (1) Because the transition point of HD is usually located in the rectosigmoid colon and contrast enema is not considered as the gold standard for diagnosing HD, we investigated whether the diagnostic rate of contrast enema is acceptable if the procedure is terminated when the contrast material enters the descending colon and completely distends the rectosigmoid colon. (2) We also aimed to determine the reduction in radiation time if simplified contrast enema (SCE) was acceptable and used instead of conventional contrast enema (CCE). (3) This study also evaluated the diagnostic value of the 24-hour-delayed abdominal radiograph.

MATERIALS AND METHODS

Patients

In this retrospective study, we collected and reviewed the results of contrast enema examination of 198 consecutive children who were clinically suspected of HD between January 2009 and December 2010 in our hospital. Of these, 18 children were excluded from the study because 24-hour-delayed abdominal radiographs were not available, they had a history of gastrointestinal tract surgery, or they had previously received a contrast enema. Of the remaining 180 children, 85 were girls and 95 were boys (age range: 1 day–13 years; average age: 2.4 years). The time taken for the contrast enema was recorded for 15 patients. Rectal suction biopsy was performed in 45 patients. The biopsy site was selected considering the barium enema or manometry results so that the most possible problematic colonic tissue could be obtained.

Definitions

SCE procedure time (S-time) was defined as the duration between the time at which the first radiograph in the lateral position was taken and the time at which the radiograph showing the entry of the contrast material into the descending colon with complete distention of the rectosigmoid colon was taken. CCE procedure time (C-time) was defined as the duration between the time at which the first radiograph in the lateral position was taken and the time at which the radiograph showing complete colonic distension was taken. Contrast infusion was started immediately after the first radiograph in the lateral position was taken. In our study, the following 2 signs indicated positive contrast enema results: (1) a rectosigmoid ratio of less than 1 (reverse rectosigmoid index) (Fig. 1a) and (2) detection of a transition zone (Fig. 1b). CCE results (C-results) and SCE results (S-results) were considered positive when at least 1 of the 2 abovementioned signs was observed on the radiographs taken during C-time and S-time, respectively. The 24-hour-delayed abdominal radiograph results (24-h-results) were considered positive when the contrast material was retained in the bowel region proximal to the descending colon. However, the presence or absence of the contrast material in the appendix was not considered. Positive 24-h-results showing the contrast material in the descending colon were termed as 24-h-D-results and those showing the contrast material in the rectosigmoid colon were termed as 24-h-RS-results. The final diagnosis was considered positive when hypoganglionosis or aganglionosis was confirmed on the basis of the pathology reports of either rectal suction biopsy or intraoperative full-thickness biopsy. In contrast, the final diagnosis was considered negative when normal rectal ganglion cells were detected in the specimens examined or when a biopsy was not performed because HD was not highly suspected in the clinical follow-up.

Statistical Analysis

Three senior radiologists evaluated the C-results, S-results, 24-h-results, 24-h-D-results, and 24-h-RS-results. We compared the sensitivities and specificities of the S-results and C-results in the 180 cases examined in our study. Furthermore, we compared the S-time and C-time for 15 patients. The 24-h-results, 24-h-D-results, and 24-h-RS-results were analyzed and compared with the C-results and S-results by using the SPSS for Windows software (Version 17). A p-value of less than 0.05 was considered statistically significant (Student’s t test).

RESULTS

Of the 180 children examined in our study, 23 were proven to have aganglionosis or rectal hypoganglionosis at different levels (rectum: 20, sigmoid colon: 2, and splenic flexure: 1). Contrast enema examination showed that 2 of the remaining 157 children had malrotation, with the cecum located in the left side of the abdomen. In contrast, except for HD-related signs, contrast enema examination showed no specific findings in the other 155 children.

The correlation between the C-results and S-results in all the 180 patients was markedly high, and only 1 patient showed a difference between the C-results and S-results (Table 1). This patient had been diagnosed with long-segment...
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The sensitivities and specificities of the 24-h-result, 24-h-D-result, and 24-h-RS-result were 60.87% and 35.67%, 73.91% and 22.93%, and 95.65% and 8.92%, respectively. Except for the patient with long-segment aganglionosis at the level of the splenic flexure, 17 (77.27%) of the remaining 22 patients showed positive S-results and C-results, and 13 (59.09%), 16 (72.73%), and 21 (95.45%) patients showed positive 24-h-results, 24-h-D-results, and 24-h-RS-results, respectively. Furthermore, 2 (9.09%), 3 (13.64%), and 5 (22.73%) patients showed positive 24-h-results, 24-h-D-results, and 24-h-RS-results, respectively, but neither the C-results nor the S-results were positive for these patients. The additional diagnostic rates of the C-results and S-results are not significant (p = 0.08) with the 24-h-results but are significant with the 24-h-D-results (p = 0.04) and with the 24-h-RS-results (p = 0.01). However, on comparing the additional diagnostic rate of the 24-h-RS-results with the combined diagnostic rate of the C-results/S-results and 24-h-D-results, we found that the total diagnostic rate did not significantly increase (p = 0.08) (Table 2).

The radiation time needed for SCE was significantly lower than that needed for CCE (p < 0.001) (Fig. 2). The average radiation time needed for SCE was 127 seconds and that needed for CCE was 375 seconds. Thus, approximately only one-third (33.87%) of the radiation time needed for CCE was needed to perform SCE.

Table 1. Relationship between C-results and S-results

<table>
<thead>
<tr>
<th>S-Result * C-Result Crosstabulation</th>
<th>Total</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>C-Result</td>
<td></td>
</tr>
<tr>
<td>Negative</td>
<td>151</td>
</tr>
<tr>
<td>Positive</td>
<td>29</td>
</tr>
<tr>
<td>Total</td>
<td>180</td>
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DISCUSSION

HD is a well-documented disease in children and is a common cause of pediatric constipation. Contrast enema is 1 of the 3 most popular examination methods used for diagnosing HD, and it is also important for preoperative planning as it helps in detecting the transition zone [6]. It has been reported that contrast enema examination in children can be terminated when the transition zone is detected [5]. However, in non-HD cases or in cases where the transition zone is not detected in the colon, the examination is continued until the colon is completely filled with the contrast material. Furthermore, a 24-hour-delayed abdominal radiograph is also considered helpful in diagnosing HD [5].

In this study, the results of these 2 diagnostic methods were found to be almost the same in all cases, except in

![Figure 1](image-url)
1 case (Table 1). We think that the pathogenetic features of HD are responsible for this high correlation. First, the transition zone is usually located at the level of the rectosigmoid colon, which is examined by SCE. In addition, a proximal bowel segment with aganglionosis is rarely noted in a normal rectosigmoid colon.

The sole patient who showed different results for the 2 procedures was diagnosed with long-segment aganglionosis, with the transition zone at the level of the splenic flexure. In this case, radiographs taken during the S-time did not show a reverse rectosigmoid index or a transition zone. However, the visible segment of the distal colon appeared to be serrated (Fig. 1c), which is a typical feature of HD. In this situation, HD with a transition zone proximal to the serrated segment was highly suspected; therefore, it was rational to perform a contrast enema until the transition zone was detected.

There are several diagnostic indexes of contrast enema for HD, including transition zone detection, irregular contractions, rectosigmoid index, spasms, cobble-stone appearance of the mucosa, mucosal irregularity, serrations, and delayed evacuation. Among these indexes, only transition zone detection, rectosigmoid index, and delayed evacuation have sensitivities greater than 50%, but delayed evacuation has a sensitivity of 66% and a specificity of 20% [7]. In our study, the sensitivities and specificities of the 24-h-results and 24-h-D-results were similar to those of the delayed evacuation index. The other 2 indexes with

<table>
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<tr>
<th>Table 2. Diagnostic rates of S/C-results, 24-hour-delayed radiograph, and the combination of S/C-results and 24-hour-delayed radiograph for 22 cases of short-segment HD</th>
</tr>
</thead>
<tbody>
<tr>
<td>* p &lt; 0.05</td>
</tr>
<tr>
<td>---------------------------------------------------------------</td>
</tr>
<tr>
<td>S- / C-Result</td>
</tr>
<tr>
<td>24-h-Result</td>
</tr>
<tr>
<td>24-h-D-Result</td>
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<tr>
<td>24-h-RS-Result</td>
</tr>
<tr>
<td>S- / C-Result + 24-h-Result</td>
</tr>
<tr>
<td>S- / C-Result + 24-h-D-Result</td>
</tr>
<tr>
<td>S- / C-Result + 24-h-RS-Result</td>
</tr>
</tbody>
</table>

**Figure 2**

The average C-time and S-time values are 375 seconds and 127 seconds, respectively. A significant difference is noted between these values (p < 0.001).
high sensitivity and specificity were the transition zone (sensitivity: 65%, specificity: 60%) and rectosigmoid index (sensitivity: 77%, specificity: 65%) [7]. These 2 indexes were also largely included in SCE, except for the fact that a less common transition zone proximal to the rectosigmoid colon could not be detected (approximately 10%) [5]. This viewpoint supports the high correlation noted between the C-results and S-results.

A delayed radiograph is helpful for diagnosing HD in neonates, particularly in those with total colonic aganglionosis. However, it is not useful in older children with constipation [5]. Our study showed that a 24-hour-delayed radiograph obtained after contrast enema increases the diagnostic rate when retention of the contrast material in the bowel proximal to the rectosigmoid colon, except in the appendix, is considered as a positive result. However, the use of a 24-hour-delayed radiograph can be omitted because of its low specificity, availability of other examination procedures with much more reliable diagnostic values, and the fact that radiographs are not very helpful in preoperative planning. Moreover, during SCE, this diagnostic rule should be excluded unless it still proves to be useful in this kind of contrast enema, with low amount of contrast material.

In our study, the diagnostic value of SCE was sufficient to replace CCE with SCE for diagnosing HD. We think that SCE is more beneficial because it can lead to a significant decrease in the radiation time and its diagnostic value is almost the same as that of CCE. However, during SCE, whenever any distinctive feature of HD is observed in the segment distal to the transition zone, it is recommended to continue the contrast enema procedure until the transition zone is identified. Malrotation was incidentally found in 2 (1%) of the 180 cases evaluated in our study. Therefore, CCE, rather than SCE, is recommended in cases where colonic disorders other than HD are suspected.

To sum up, we present a flowchart that is based on the findings of this study (Fig. 3) and may be used to survey children suspected of having HD. We think SCE helps to considerably reduce unnecessary radiation time. However, this study has some limitations. It was difficult to control the total amount of contrast material infused in the colon in different patients, which might have influenced the results pertaining to the 24-hour-delayed radiograph. In addition, most of the patients did not undergo biopsy; therefore, there may be some undiagnosed cases. However, this does not affect the correlation between the C-results and S-results. Furthermore, the radiation time was calculated on the basis of the time recorded on the radiographs (procedure time) instead of on a real-time recorder (true radiation time). Although fluoroscopy may have been paused during the examination, these interruptions occupy only a small amount of the procedure time. This is because the level of the contrast material in the colon is continuously traced and patients are made to change their positions in order to improve the quality of the results for each segment of the colon. Therefore, true radiation time can be considered similar or at least positively correlated to the procedure time. This study was a retrospective study, and therefore, its results will not be as reliable as those of a prospective study. However, on the basis of our current findings, we believe that the topics investigated in this study warrant a well-designed prospective study in the future.

Figure 3. A flowchart that can be used in the diagnosis of HD shows that CCE is performed only when long-segment aganglionosis is suspected by SCE. However, except in the case of total colonic aganglionosis, contrast enema can be terminated when a transition zone is identified. On the basis of this flowchart, the radiation time can be reduced in HD or non-HD cases instead of opting to perform CCE initially. (HD: Hirschsprung disease, SCE: simplified contrast enema, CCE: conventional contrast enema)
CONCLUSION

The diagnostic values of SCE and CCE for diagnosing HD in children are quite similar. However, the radiation time for SCE is one-third that of CCE; this difference is statistically significant. A 24-hour-delayed abdominal radiograph adds to the diagnostic value but has very low specificity. However, if the 24-hour-delayed radiograph is to be used, the positive result should be more adequately defined as presence of the contrast material in any segment proximal to the rectosigmoid colon.

REFERENCE