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**ORIGINAL ARTICLE**

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## **How Useful is Contrast Enema in the Diagnosis of Hirschsprung's Disease? Five-year Experience from a Local Referral Centre**

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### **ABSTRACT**

**Objective:** To review the diagnostic performance of contrast enema examinations for the investigation of Hirschsprung's disease, determine the usefulness of delayed 24-hour X-rays, and validate water-soluble contrast enemas for the investigation for patients with suspected Hirschsprung's disease.

**Methods:** This was a retrospective study of all neonates and infants referred for radiological evaluation of symptoms and signs suspicious of Hirschsprung's disease in Queen Mary Hospital, Hong Kong from January 2007 to December 2011. Patients were included in this study when all radiographs were available and Hirschsprung's disease had been confirmed or excluded by rectal biopsy or by a combination of enema results, manometry, and close clinical follow-up. Radiographs and reports were reviewed with a checklist of radiological criteria described in the literature, including transitional zone, rectosigmoid index (maximum width of rectum divided by maximum width of sigmoid; abnormal if  $<1$ ), irregular contractions, mucosal irregularity, spasm, serrations and retention of contrast agent on delayed radiographs. The sensitivity, specificity, positive and negative predictive values of each of these criteria were calculated and considered in the context of recommendations described in the literature.

**Results:** A total of 136 such patients were evaluated during the study period, of whom 86 had barium enemas and 57 had water-soluble contrast enemas. Among all the patients reviewed, the sensitivity, specificity, positive and negative predictive values for all enema examinations (excluding delayed films) in the diagnosis of Hirschsprung's disease were 69%, 89%, 44% and 96%, respectively. While the corresponding values of all enema examinations (including delayed films) were 100%, 78%, 36% and 100%, respectively. Similar results were noted in patients having barium enemas and water-soluble contrast enema examinations. For patients having barium enemas or water-soluble contrast enemas, with the inclusion of delayed films there was a significant increase in sensitivity compared to sensitivity derived without recourse to delayed films, whilst there was only a slight reduction in specificity.

**Conclusion:** Recourse to water-soluble contrast enemas with the addition of a 24-hour delayed film was useful in the diagnosis of Hirschsprung's disease.

**Key Words:** Contrast media; Diagnostic tests, routine; Enema; Hirschsprung disease

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## 中文摘要

### 灌腸診斷先天性巨結腸症有用嗎？一所本地轉介中心的五年經驗

黃慧妍、曾承峰、林慧文

**目的：**探討灌腸檢查對先天性巨結腸症的診斷性能和24小時延遲X光片的診斷效用，並驗證水溶性對比劑灌腸對先天性巨結腸症疑似病例的研究。

**方法：**本研究回顧2007年1月至2011年12月期間具有疑似先天性巨結腸症狀及指徵而被轉介至瑪麗醫院作放射性檢查的新生兒和嬰兒病例。病人被納入本研究時，均已有X光片，其先天性巨結腸症已由直腸活檢或灌腸結合測壓及臨床監察的方法證實或排除。根據文獻描述的放射學標準目錄而回顧X光片及其報告，包括過渡區、直腸乙狀結腸指數（直腸的最寬徑除以乙狀結腸的最寬徑， $<1$ 為異常）、不規則宮縮、粘膜不規則、痙攣、鋸齒狀和延遲X光片上對比劑滯留。為每項標準分別計算其敏感性、特異性，以及陽性和陰性預測值，並與文獻上的數據作比較。

**結果：**136例中，有86例進行鋇劑灌腸，57例進行水溶性對比劑灌腸。回顧病例中，灌腸檢查（不包括延遲X光片）對先天性巨結腸症診斷的敏感性、特異性，以及陽性和陰性預測值分別為69%、89%、44%和96%。把延遲X光片結果納入統計之內，相應值為100%、78%、36%和100%。接受鋇劑灌腸和水溶性對比劑灌腸檢查的兩組患者均有類似的結果。同樣地兩組患者中，與不包括延遲X光片的數據比較，包括延遲X光片的數據顯示其敏感性明顯上升，而特異性則輕微下降。

**結論：**水溶性對比劑灌腸檢查結合24小時延遲X光片對診斷先天性巨結腸症有用。

## INTRODUCTION

Hirschsprung's disease (HD) is a congenital lower intestinal obstruction caused by absence of ganglion cells in the submucosal and intramuscular plexuses of gastrointestinal tract.<sup>1</sup> Currently, in many centres a contrast enema is the first examination requested to evaluate neonates with distal intestinal obstruction or patients with abnormal bowel habits. Together with the clinical and radiological findings, the clinician decides whether manometry, rectal biopsy, or both, are required to confirm or exclude HD.<sup>1</sup>

The aims of our study were to: (1) review the diagnostic performance of contrast enema examinations for the investigation of HD, (2) determine the usefulness of delayed 24-hour abdominal X-rays (AXRs), and (3) validate low-osmolality water-soluble contrast enemas (WSCEs) for the investigation for patients with suspected HD.

## METHODS

This was a retrospective study of all neonates and infants referred for radiological evaluation of symptoms and signs suspicious of HD in Queen Mary Hospital, Hong Kong from January 2007 to December 2011. Both

barium and low-osmolality WSCEs (using omnipaque 300 or visipaque 270) were performed in our hospital and evaluated separately. Findings from the enemas and delayed radiographs as well as the medical records of these neonates and infants were reviewed. Patients were included in this study only when all radiographs were available, and HD had either been confirmed or excluded by rectal biopsy or a combination of enema results, manometry, and close clinical follow-up. Two patients were excluded from the study as they had defaulted follow-up and hence had no rectal biopsy.

Radiographs and reports were reviewed using a checklist of radiological criteria described in the literature,<sup>1-4</sup> including transition zone, rectosigmoid index (maximum width of rectum divided by maximum width of sigmoid; abnormal if  $<1$ ), irregular contractions, mucosal irregularity, spasm, serrations and retention of contrast agent on delayed radiographs. Delayed evacuation was defined as presence of any residual contrast in the ascending colon on 24-hour delayed abdominal X-rays. The occurrences of various criteria from the literature were recorded. The sensitivity, specificity, positive and negative predictive values of each criterion were calculated and contrasted with recommendations for the

diagnosis in the literature.

Examples of a typical transition zone (Figures 1 and 2), abnormal rectosigmoid index (<1) (Figure 3) and delayed evacuation (Figure 4) are shown.

## RESULTS

In all, 136 patients were evaluated during the study period, of which 75 (55%) were males and 61 (45%) were females. Their mean age was 43 (range, 0-350) days; 78 (57%) were neonates. Of the 136 patients, 86 had a barium enema (BE) and 57 had a WSCE. Of the latter patients, seven had two WSCE examinations performed to evaluate symptoms — five underwent a repeat WSCE as the first examination was incomplete (contrast not having reached the caecum), while two patients with persistent symptoms after the first examination had a repeat examination owing to uncertainty about the diagnosis.

In 43 of our study patients, final diagnoses were reached as listed in Table 1. In the remainder, no significant pathology was identified after imaging, rectal biopsy, and follow-up, and were treated as functional bowel disorder problems. The frequency of abnormal enema findings is shown in Figure 5. In all, 16 patients had

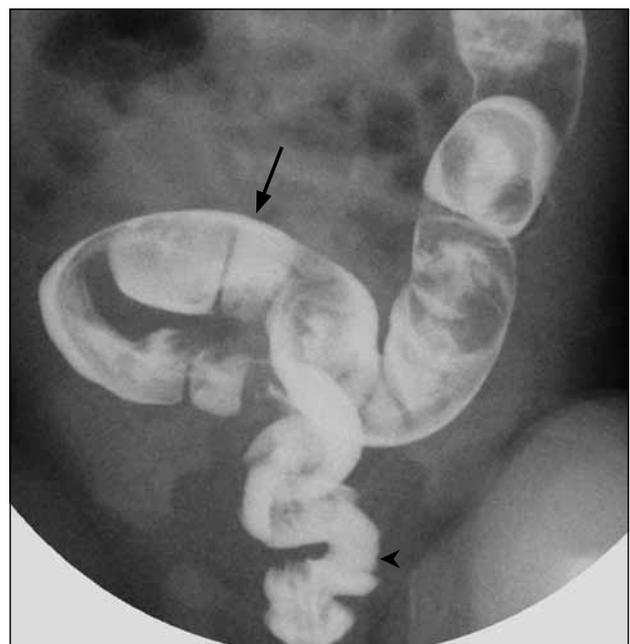
HD, all of whom had either abnormal enema findings or delayed evacuation. Results were categorised into three groups: all patients undergoing enema (BE and WSCE) examinations, those undergoing BEs, and those



**Figure 2.** A frontal radiograph of contrast enema of another patient with Hirschsprung's disease shows the transition zone at the rectum (arrow).



**Figure 1.** A frontal radiograph of water-soluble contrast enema of a neonate with Hirschsprung's disease shows transition zone at the rectosigmoid junction (arrow).



**Figure 3.** A radiograph of contrast enema shows an abnormal rectosigmoid index (<1), and dilated sigmoid colon (arrow) and narrow rectum (arrowhead).

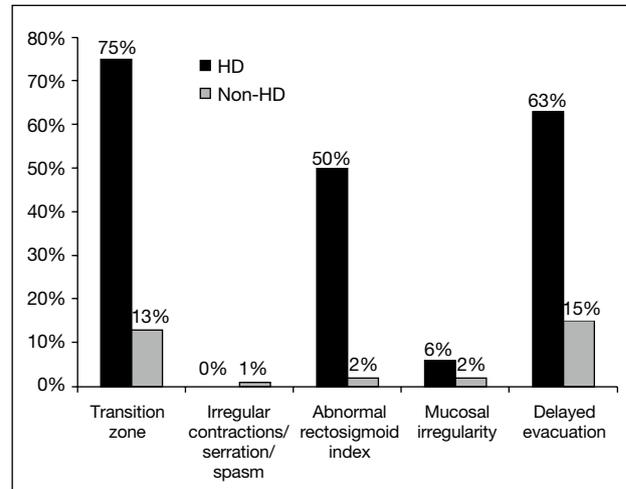


**Figure 4.** A delayed 24-hour abdominal X-ray shows residual contrast in the ascending colon (arrow) and terminal ileum (arrowhead), suggestive of delayed evacuation.

**Table 1.** Final diagnoses in 43 patients with bowel symptoms.

Diagnosis	No. (%) of patients
Hirschsprung's disease	16 (11.8)
Neonate	13 (9.6)
Infant	3 (2.2)
Small bowel atresia	10 (7.4)
Meconium plug syndrome	5 (3.7)
Stricture	3 (2.2)
Meconium ileus	2 (1.5)
Intussusception	2 (1.5)
Midgut volvulus	2 (1.5)
Small bowel obstruction	2 (1.5)
Colitis	1 (0.7)

undergoing WSCEs. The sensitivities, specificities, positive and negative predictive values of these three groups were compared (Table 2). Comparison was also made between patients having a delayed 24-hour AXR with those not having this delayed X-ray. Table 3 shows the sensitivities and specificities of the radiological criteria for the combined group, patients undergoing BEs, and those having WSCEs. In our study, water-soluble contrast / thin barium was injected manually using a Foley catheter via the anus. The endpoint of the examination was the caecum. Among all the cases, five BEs were incomplete (2 up to hepatic flexure, 2 up to the splenic flexure, and 1 to the descending colon). Of these five patients, one was due to a colonic splenic flexure stricture (consistent with BE findings) in whom no definite aetiology was identified; in the four other



**Figure 5.** Frequency of abnormal enema findings in our study (for both barium enema and water-soluble contrast enema). Abbreviation: HD = Hirschsprung's disease.

patients, no significant abnormality was identified after follow-up and rectal biopsy. Notably, WSCEs were incomplete in 11 patients — five up to the splenic flexure, three up to mid-transverse colon, and three up to the hepatic flexure. Regarding these 11 patients, one each had jejunal atresia, duodenal atresia, meconium ileus, intussusception, and ileal atresia, whilst two had midgut volvulus, two had meconium plug syndrome, and two had no significant abnormality identified after follow-up and rectal biopsy.

## Overall Results

Of the 16 cases of HD, short-segment disease involving the rectosigmoid colon was the most common presentation and affected 13 (81.3%) of the patients, 2 (12.5%) had long segment disease and 1 (6.3%) had total colonic aganglionosis. Five (31.3%) of the cases involved only the rectum, while 8 (50.0%) involved the rectosigmoid colon, 1 (6.3%) each had involvement up to the descending colon, the transverse colon, and the whole colon (total colonic aganglionosis). These findings are consistent with those reported in the literature.<sup>5</sup>

The overall sensitivities, specificities, and positive and negative predictive values of all enema examinations and of radiological criteria used in our study were also comparable to those reported in the literature.<sup>1,3,4,6</sup>

## Delayed 24-Hour Abdominal X-ray

With the inclusion of delayed 24-hour AXR in the

**Table 2.** Overall sensitivity, specificity, and positive and negative predictive values of diagnosing Hirschsprung's disease.

Findings	All enemas (BE and WSCE)				BE				WSCE			
	% Sensitivity	% Specificity	PPV (%)	NPV (%)	% Sensitivity	% Specificity	PPV (%)	NPV (%)	% Sensitivity	% Specificity	PPV (%)	NPV (%)
Exclude delayed 24-hour AXR	69	89	44	96	75	82	41	95	50	98	67	96
Include delayed 24-hour AXR	100	78	36	100	100	66	32	100	100	94	57	100

Abbreviations: AXR = abdominal X-ray; BE = barium enema; NPV = negative predictive value; PPV = positive predictive value; WSCE = water-soluble contrast enema.

**Table 3.** Sensitivity, specificity, and positive and negative predictive values of radiological criteria for diagnosing Hirschsprung's disease.

Findings	All enemas (BE and WSCE)				BE				WSCE			
	% Sensitivity	% Specificity	PPV (%)	NPV (%)	% Sensitivity	% Specificity	PPV (%)	NPV (%)	% Sensitivity	% Specificity	PPV (%)	NPV (%)
Transition zone	75	87	43	97	75	81	39	95	75	96	60	98
Irregular contraction	0	99	0	89	0	100	0	86	0	98	0	93
Spasm / serration	0	100	0	89	0	100	0	86	0	100	0	93
Abnormal rectosigmoid index	50	98	73	94	42	99	83	91	75	96	60	98
Mucosal irregularity	6	98	25	89	8	97	33	87	0	98	0	96
Delayed evacuation	63	85	35	95	67	77	32	93	50	96	50	96

Abbreviations: AXR = abdominal X-ray; BE = barium enema; NPV = negative predictive value; PPV = positive predictive value; WSCE = water-soluble contrast enema.

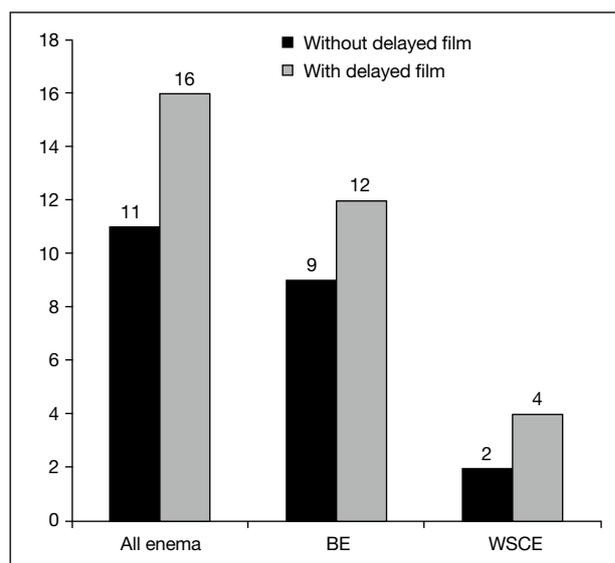
diagnosis of HD, sensitivity increased from 69% to 100%, while specificity was only slightly reduced from 89% to 78%. Similar results were obtained in patients undergoing BE and WSCE examinations. Number of patients for whom the diagnosis of HD changed from negative to positive after review of delayed 24-hour AXR are shown in Figure 6. In five patients with delayed evacuation (2 WSCE and 3BE), the diagnosis of HD changed from negative to positive after reviewing 24-hour delayed AXR, and were finally confirmed histologically. In all, 14 BE cases with delayed evacuation finally turned out to be normal, whilst two WSCE cases with delayed evacuation finally turned out to be normal. Among all the patients with HD, two had short-affected segments and no delayed evacuation; they only had a transition zone. While all three with long affected segments (including one with total colonic aganglionosis) had delayed evacuation.

### Water-soluble Contrast Enema Versus Barium Enema

Sensitivity in patients undergoing WSCE without a delayed 24-hour AXR was only 50%, compared to 75% in those having a BE. However, with the inclusion of delayed 24-hour AXR, the sensitivity for both types of enema increased to 100%. Specificity was comparable in both groups of patients and in the combined group.

### DISCUSSION

We showed that recourse to a delayed 24-hour AXR significantly improved sensitivity for the diagnosis of HD to 100% for patients having BEs or WSCEs.



**Figure 6.** Patients diagnosed as Hirschsprung's disease before and after review of delayed 24-hour abdominal X-ray. Abbreviations: BE = barium enema; WSCE = water-soluble contrast enema.

Any residual contrast in the ascending colon on a delayed 24-hour AXR is the critical cut-off. The literature dwells on residual contrast in the proximal to sigmoid colon as the cut-off point,<sup>1</sup> and based on this criterion our sensitivity for diagnosis of HD was 63%, which is slightly lower than the figure of 66% listed in literature,<sup>1</sup> but with a much higher specificity of 85%, compared to 20% reported in the literature.<sup>1</sup> The significant increase in specificity in our study may have been due to the larger sample size compared with those in previously described studies.<sup>1</sup>

As there were few cases with delayed evacuation and a final diagnosis of no abnormality, we believe that delayed evacuation has to be interpreted together with other radiological findings to make the diagnosis of HD.

In our series, although all the cases with long-segment HD had delayed evacuation, the number was too small to arrive at any conclusion on whether short or long aganglionic segments affect contrast evacuation from the ascending colon differently.

We believe that delayed 24-hour AXR is useful in the diagnosis of HD, and is applicable to patients undergoing BE or WSCE examinations. We recommend using the ascending colon as the cut-off point for retention of contrast, as it significantly improves sensitivity and reduces specificity only slightly. We therefore believe that for patients with a normal enema examination and no delayed evacuation on the delayed 24-hour AXR, invasive investigation such as rectal biopsy may not be necessary and can be avoided. A further prospective study with a larger sample size may help to consolidate this conclusion.

Of all the radiological diagnostic signs to detect HD, transition zone is the most sensitive, while other ancillary signs (irregular contraction, spasm, serration, and mucosal irregularity) help to confer high specificity. However, these are not frequently detected on enema examinations and notably, we did not detect any instance of spasm or serration, and only three cases with mucosal irregularity. The latter three patients also had other radiological signs (transition zone/delayed evacuation/microcolon) that suggest the diagnosis of HD or other pathology. Thus, if these ancillary signs are detected during the examinations, other features of HD should be carefully looked for.

We showed that the above-mentioned radiological

diagnostic signs can be readily detected in both BEs and WSCEs and that with the inclusion of a delayed 24-hour AXR, the sensitivity, specificity, and positive and negative predictive values of WSCE were also comparable. Colonic perforation is reported to occur in 3% to 4% of HD patients,<sup>7,8</sup> in which case WSCE prevents barium spillage into the peritoneal cavity and provides a larger margin of safety. Probably it is therefore justified as the diagnostic radiological examination for HD.

One limitation of this study was that it was retrospective and entailed retrospective review of images. Thus, it was difficult when it came to evaluating irregular contractions and spasm (two of the radiological criteria of diagnosing HD). A second limitation was the small sample size and the limited number of patients who actually had HD. A further prospective study with a larger sample size may help to address these limitations.

## CONCLUSION

We have shown that delayed 24-hour AXR using ascending colon as the cut-off is very useful in improving sensitivity for the diagnosis of HD in patients having BEs and WSCEs, in which case the performance of types of examination becomes comparable. We recommend WSCE as the diagnostic evaluation of choice for suspected HD, as it minimises the risk of barium spillage into peritoneum in cases of colonic perforation.

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