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ULTRASOUND

Diagnosis of Hirschsprung disease by hydrocolonic sonography in children

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Abstract

Objectives To compare hydrocolonic sonography with histopathology for diagnosing children with symptoms highly suggestive of Hirschsprung disease (HD).

Methods In this prospective study, patients presenting refractory constipation highly suggestive of HD underwent hydrocolonic sonography with retrograde infusion of saline into the colon. The dilated segments, narrowed segments, luminal diameter ratio, transition zone (TZ), thickening, and blood perfusion of the upstream bowel were evaluated. The sensitivity and specificity of combined and single parameters were determined in comparison with biopsy.

Results One hundred and three children were included in this study; 49 were confirmed to have HD. The luminal diameter ratio showed superiority over other parameters. An area under the curve (AUC) of 0.969 (95% confidence interval [CI]: 0.936–1.000) and a cutoff value of 1.51 were established by receiver operating characteristic (ROC) curve analysis of the luminal diameter ratio (sensitivity: 89.8%; specificity: 96.3%). By combining the luminal diameter ratio as the major criterion with two minor criteria, hydrocolonic sonography showed the same sensitivity (91.8%) and better specificity (96.3% vs 87%) than contrast enema, but this difference was not statistically significant ($p = 0.063$). Consistency analysis showed a kappa value of 0.825 ($p < 0.001$), indicating excellent agreement between hydrocolonic sonography and contrast enema.

Conclusions Hydrocolonic sonography is a valuable diagnostic tool with both high sensitivity and specificity for HD diagnosis, allowing morphological and vascular assessments of the colon, and correlates well with contrast enema. In the appropriate setting, hydrocolonic sonography may be an alternative screening method for HD in a large group of children with constipation.

Key Points

• *Hydrocolonic sonography is a simple, well-tolerated diagnostic tool with both high sensitivity and specificity for HD diagnosis.*

• *Hydrocolonic sonography allows morphological and vascular assessments of the colon, and correlates well with contrast enema.*

• *Hydrocolonic sonography is a possible alternative modality for paediatric patients highly suggestive of HD.*

Keywords Ultrasonography · Hirschsprung disease · Child · Colon

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Abbreviations

HD	Hirschsprung disease
HSP	Henoch-Schonlein purpura
RSI	Rectosigmoid index
TZ	Transition zone

Introduction

Hirschsprung disease (HD) is a common congenital disorder with a global incidence of approximately 1 in 5000 in newborns [1–4]. HD is characterised by an absence of ganglion cells of the submucosa and myenteric nerve plexus in the distal bowel (usually localised to the rectosigmoid region), which causes the affected bowel to lose its peristaltic ability and leads to dilation of the upstream colon, resulting in refractory constipation. HD is classified according to the length of the aganglionic section into short-segment (confined to the rectosigmoid region), long-segment (extending beyond the rectosigmoid region), total colonic (entire colon and terminal ileum), and total intestinal (most of the intestines are involved) HD [5]. An operation is recommended once the diagnosis has been established [6].

The diagnosis is based on symptoms, contrast enema, rectal suction biopsy, and full-thickness rectal biopsy. Contrast enema is the imaging modality of choice for HD, with sensitivity ranging from 76 to 88.9% and specificity from 84.2 to 98% [7–9]. With regard to contrast enema, patients are exposed to radiation; therefore, parents may have safety concerns. Rectal biopsy, despite being radiation-free, is invasive, sometimes requires anaesthesia, and is not widely accepted by parents. Over 80% of patients who undergo biopsies have normal ganglionic biopsies [10, 11]. In our centre, the negative biopsy rate is 74%. Therefore, there is a clear need for a noninvasive, safe, and highly sensitive diagnostic method to increase the positive rate of rectal biopsy. Ultrasound (US), although noninvasive and radiation-free, has limitations for the evaluation of HD: diffuse intraluminal gas precludes the detailed evaluation of the colon wall. Second, the rectosigmoid region is the most commonly involved region in HD, and identification of this region by trans-abdominal US is challenging due to its deep location. Third, colonic dilatation is an essential clue for HD diagnosis, and this pathology may be overlooked after laxative therapy or enema, which are commonly used therapies for HD. Some common drawbacks of US include a limited field of view and operator dependence. Since the late 1980s [12], retrograde instillation of water or other contrast agents into the colon, also known as hydrocolonic sonography, has been proposed as an alternative to contrast enema and colonoscopy for diagnosing colonic tumours, polyps, and inflammatory diseases [13]. It overcomes the drawbacks mentioned above by replacing gas, distending the intestinal lumen, and facilitating visualisation of the entire colon and rectum. This made us wonder if it would be possible to use this method in the evaluation of HD. To our knowledge, there have been no reports in the literature regarding the accuracy of hydrocolonic sonography for HD diagnosis. In this context, the aim of our study was to prospectively evaluate the sonographic features of HD by hydrocolonic sonography and to compare these features with contrast enema and histopathologic findings.

Materials and methods

This was a prospective study from February 2019 to May 2020 at a tertiary centre. The study was approved by the Institutional Review Board of our hospital. Information regarding the procedures, risks and benefits related to hydrocolonic sonography, contrast enema, and rectal biopsy was given to the guardians of each child and written informed consent was obtained.

Inclusion and exclusion criteria

Inclusion criteria: (a) patients aged 1 month to 18 years; (b) children who presented with severe constipation (voluntary bowel movement less than twice per week) refractory to dietary modifications and medical management in 1 to 3 months, which was highly suggestive of HD, with or without a history of delayed passage of meconium. Exclusion criteria: (a) individuals who previously underwent a contrast enema or a biopsy; (b) patients with mechanical factors inducing constipation such as anorectal malformation and post-necrotising enterocolitis stenosis; (c) patients who had a history of bowel surgery; and (d) individuals with serious concomitant disease.

US and enema examination

Hydrocolonic sonography was performed before contrast enema and biopsy. The US study was conducted on an EPIQ 5 system (Philips Healthcare) equipped with curved (1–5 MHz) and linear (3–12 MHz) probes and a Logiq E9 system (GE Healthcare) equipped with curved (1–5 MHz) and linear (6–9 MHz) probes. All US examinations were performed on either prepared or unprepared colons by a paediatric radiologist (Y.M.) with 8 years of paediatric US experience. The patients were placed in the supine position. After routine examination of the abdomen by conventional US (including scans of the stomach, duodenum, small intestine, and colorectum), retrograde instillation of warm saline (35–37 °C, 50–500 mL) was injected by a small calibre catheter (diameter: 6.7 mm) placed in the rectum with its tip 5–10 cm from the anus. Continuous trans-abdominal scanning from the rectum to the caecum started immediately at the time of saline instillation (Fig. 1). If narrowing or dilatation of the bowel loops was detected, saline was injected until the upstream normal bowel was identified. If there was no dilatation or narrowing, the saline needed to reach the caecum. The catheter was withdrawn after trans-abdominal scanning. The transducer was then placed longitudinally on the perineum with the patients in the lithotomy position to provide a midsagittal image of the rectum (Fig. 1).

Image analysis

US images were initially interpreted by the same radiologist (Y.M.) during the scanning process. After the US scanning, another radiologist (Z.J.Z., with 6 years of paediatric US experience), who was blinded to the results of the former radiologist, reviewed the images and video recordings. In case of discrepancy, images were jointly reviewed by both radiologists and a consensus was reached. The US signs of HD were evaluated, including the following items: (a) dilated segment, which was defined as a lumen diameter greater than that of other sections of the colon; (b) narrowed segment, which was defined as relative bowel loop narrowing compared with other sections of the colon; (c) the luminal diameter ratio was calculated using the diameter of the most dilated bowel divided by the diameter of the narrowed segment or the adjacent downstream segment (if the narrowed segment was not identified); measurements were taken three times on longitudinal sections for each patient, and the data are expressed as the mean values; (d) TZ, which was identified as an abrupt calibre change from a dilated bowel to a narrowed or relatively normal segment; (e) thickening of the upstream bowel, considered present when the TZ and/or dilated proximal bowel wall measured 1.9 mm or more [14]; (f) hypervascularity in the upstream bowel, defined as an increased vascular signal of the TZ and/or dilated proximal bowel based on the Limberg semiquantitative method for Crohn's disease [15, 16]. The measurement of the calibre change and US signs of HD are shown in Figs. 2 and 3.

Other clinical information

Contrast enema was performed after the saline had been expelled (within 7 days after hydrocolonic sonography). A radiologist (Y.C.Y., with 17 years of paediatric radiology experience) and a surgeon (Q.L., with 14 years of clinical and research experience in HD), both of whom had no knowledge of the results of the US, assessed the images independently. Diagnoses were considered positive for HD when the TZ (an abrupt calibre change as defined above) or

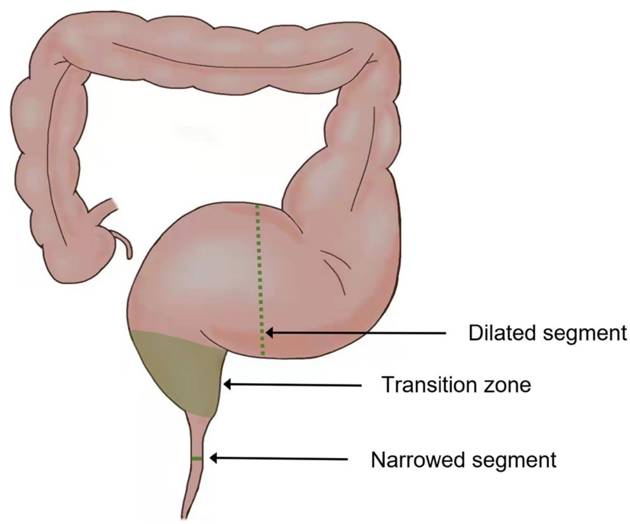

Fig. 2 Illustration of the measurement of the calibre change in HD

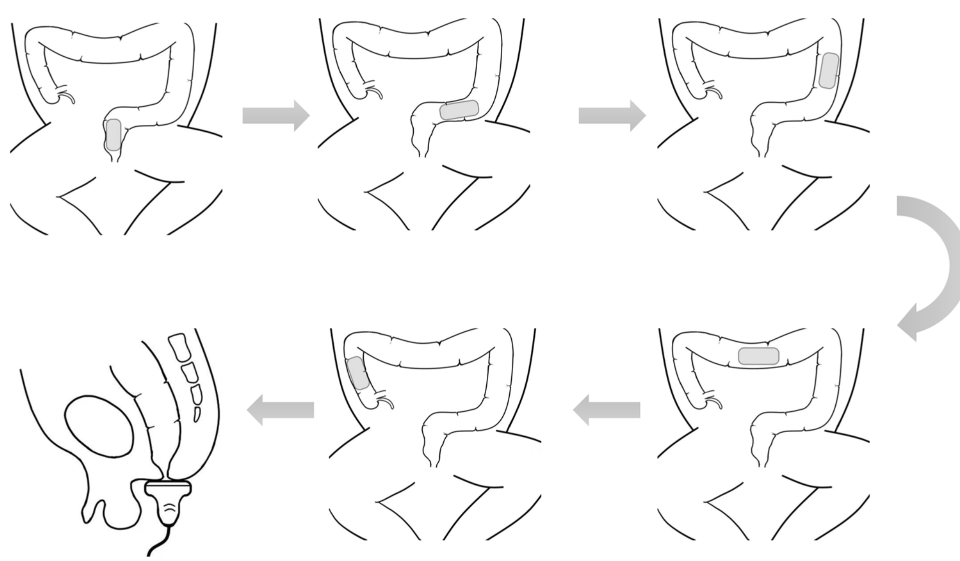

Fig. 1 US scanning of the colon. Continuous trans-abdominal scanning from the rectum to caecum in an anticlockwise direction. After withdrawing the catheter from the rectum, the transducer is placed longitudinally on the perineum with the patient in the lithotomy position to provide a midsagittal image of the rectum

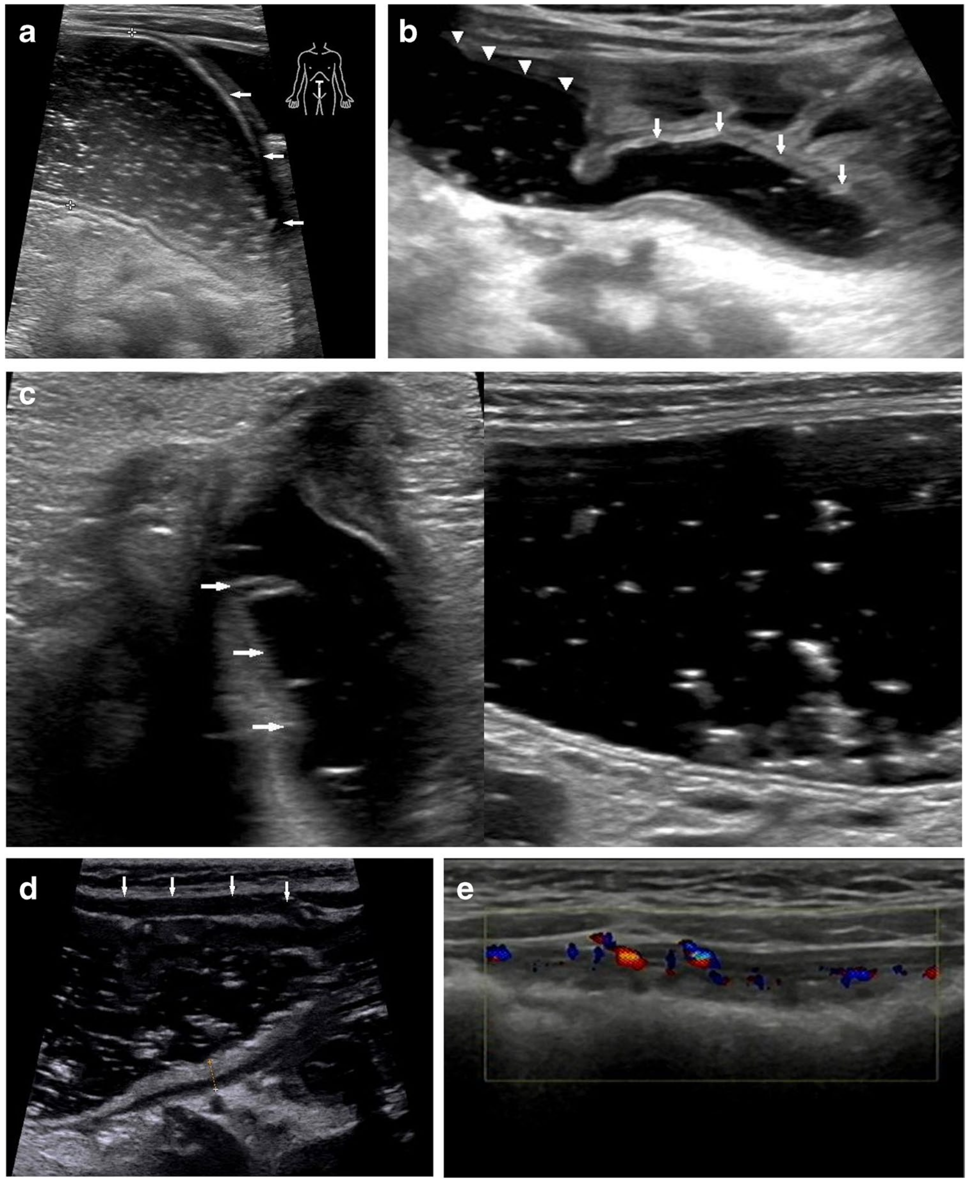

Fig. 3 Signs of HD on hydrocolonic sonography. **a** Longitudinal sonogram of a 3-month-old infant shows a dilated sigmoid colon (crosses) and a TZ in the rectosigmoid colon (arrows), regarded as the site of abrupt calibre change from the dilated bowel to a nondilated segment. **b** Longitudinal US section of the sigmoid colon in a 3-month-old infant shows the narrowed and rigid distal sigmoid (arrows) and dilated upstream colon (arrowheads). **c** The image on the left shows the longitudinal plane of the rectum on perineal ultrasound in a 3-month-old infant. The rectum was relatively narrow in comparison with the dilated sigmoid colon on the right side. **d** Longitudinal sonogram of a 2-month-old infant shows the funnel-shaped TZ in the rectosigmoid junction (arrows). Note that thickening of the bowel wall (crosses) is demonstrated. **e** Longitudinal colour Doppler ultrasound image of a 2-year-old boy depicts increased vascularity within the thickened TZ bowel wall

abnormal rectosigmoid index (RSI) (calculated using the widest diameter of the rectum divided by the widest diameter of the sigmoid, normal RSI > 1) was identified by both the radiologist and the surgeon, and disagreement was resolved by consensus. All the patients underwent rectal biopsy for histologic examination of the presence or absence of ganglionic cells after both US and contrast enema evaluation. Suction biopsy was recommended for patients aged 6 months or younger, and full-thickness biopsy under general anaesthesia was recommended for patients older than 6 months based on the clinical routine of our centre. If the contrast enema was negative and the biopsy was positive, a second biopsy was performed in case of false-positive results of the first biopsy. An operation was performed if no ganglionic cells were found. Non-HD patients were offered conservative therapies, including anal dilation, enemas, and laxatives.

Statistical analysis

Statistical analysis was performed using IBM SPSS statistics 20 (IBM). To test differences between groups, Student's *t* test or the Mann–Whitney *U* test was used for continuous variables, and the chi-squared test was used for categorical variables. A receiver operating characteristic (ROC) curve was constructed to determine the cutoff value of the luminal diameter ratio with the best sensitivity and specificity to predict HD. The sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) of each US parameter were determined. Agreement between contrast enema and hydrocolonic sonography was calculated using Cohen's kappa. A *p* value of less than 0.05 was considered to indicate a statistically significant difference.

Results

Patient's characteristics

Between February 2019 and May 2020, 139 consecutive children met the inclusion criteria; however, 33 patients were subsequently excluded for contrast enema performed at another hospital, 2 were excluded because of final diagnoses of post-necrotising enterocolitis stenosis, and 1 was excluded for a final diagnosis of the Currarino syndrome. The final analysis was performed on 103 patients (median age, 6 months; interquartile range, 3–20 months). There were 41 (39.8%) girls and 62 (60.2%) boys.

According to the rectal biopsy, the absence of ganglion cells was found in 49 patients. In 4 of these, no abnormal findings of HD were observed on contrast enema (two of them had abnormal hydrocolonic sonography); therefore, a repeat biopsy was subsequently performed, with aganglionosis confirmed in all 4 cases. Thus, 49 patients were diagnosed with HD (43 cases were short-segment disease and 6 were long-segment disease) and 54 cases were ruled out (Fig. 4). There was no significant difference regarding age in the HD and non-HD groups (Mann–Whitney test; $U=276.000$; $p=0.756$). The HD group had a greater proportion of males (73.5% vs 48.1%, $p=0.009$).

Ultrasound and contrast enema findings

No significant side effects were noted during or after the hydrocolonic sonography procedure. The US characteristics of HD are summarised in Table 1. The subjective judgement of the dilated segment achieved the highest sensitivity (95.9%) and moderate specificity (83.3%). The ROC curve for the luminal diameter ratio is shown in Fig. 5. The area under the curve (AUC) was 0.969 (95% CI: 0.936–1.000), indicating a high diagnostic accuracy. A cutoff value of 1.51 was established by ROC analysis, yielding a sensitivity of 89.8%, a specificity of 96.3%, a PPV of 95.7%, and an NPV of 91.2%. The TZ, narrowed segment, and thickening of the upstream bowel achieved high specificity (96.3%, 98.1%, and 98.1%) with moderate sensitivity (81.6%, 71.4%, and 69.4%). Hypervascularity of the upstream bowel had the best specificity of 100% but a poor sensitivity of 38.8%. The US characteristics of HD are shown in Fig. 6.

We included the luminal diameter ratio as the major criterion and the other four parameters (TZ, narrowed segment, thickening of the upstream bowel, and hypervascularity of the upstream bowel) as minor criteria with the intention of elevating the sensitivity. Ultrasound findings were considered positive for HD if the luminal diameter ratio was larger than 1.51 or at least 2 of these minor criteria were present. Then, we obtained a sensitivity of 91.8% and a specificity of 96.3%. When compared with contrast enema, the combined method of US had the same sensitivity (91.8%) and better specificity (96.3% vs 87%), but this difference was not statistically significant ($p=0.063$). Consistency analysis showed a kappa value of 0.825 ($p<0.001$), indicating excellent agreement between hydrocolonic sonography and contrast enema. Contrast enema findings of HD are summarised in Table 2. The comparison of diagnostic performances

Fig. 4 Rectal biopsy results of the 103 patients

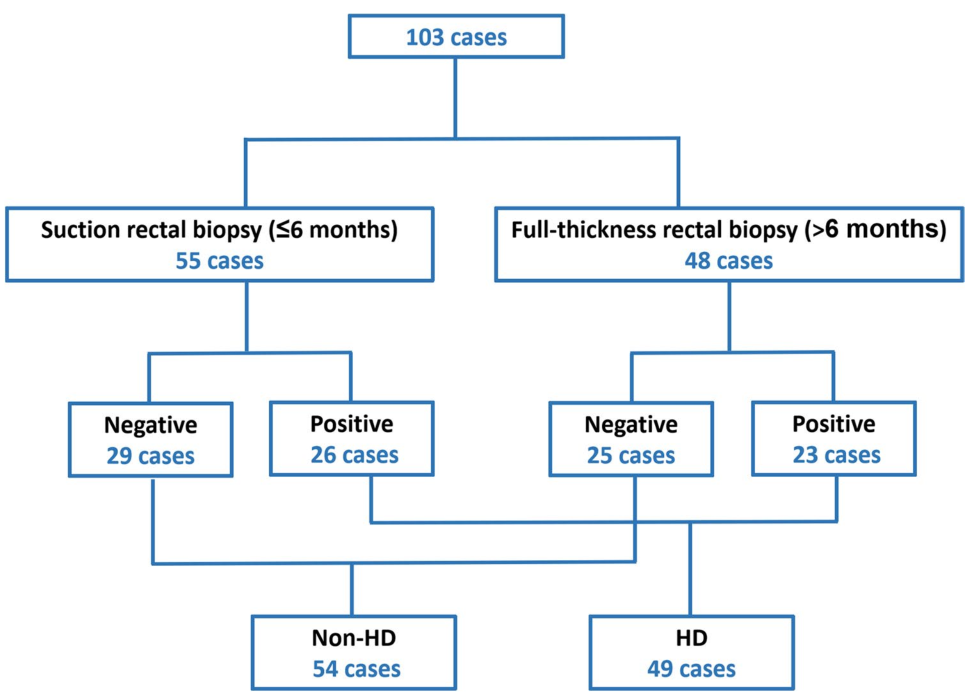

Table 1 The hydrocolonic sonography characteristics of HD. Data in parentheses are 95% confidence intervals, and data in brackets are numerators and denominators. *TZ*, transitional zone; *PPV*, positive predictive value; *NPV*, negative predictive value; *CI*, confidence interval

	χ^2	*p* value	Kappa	Sensitivity %	Specificity %	PPV %	NPV %
Dilatation	65.04	< 0.001	0.787	95.9 (84.9–99.3) [47/49]	83.3 (70.2–91.6) [45/54]	83.9 (71.2–91.9) [47/56]	95.7 (84.3–99.3) [45/47]
Narrowing	54.7	< 0.001	0.704	71.4 (56.5–83.0) [35/49]	98.1 (88.8–99.9) [53/54]	97.2 (83.8–99.9) [35/36]	79.1 (67.1–87.7) [53/67]
Luminal diameter ratio	77.04	< 0.001	0.863	89.8 (77.0–96.2) [44/49]	96.3 (86.2–99.4) [52/54]	95.7 (84.0–99.2) [44/46]	91.2 (80.0–96.7) [52/57]
TZ	64.6	< 0.001	0.784	81.6 (67.5–90.8) [40/49]	96.3 (86.2–99.4) [52/54]	95.2 (82.6–99.2) [40/42]	85.2 (73.3–92.6) [52/61]
Thickening of the upstream bowel	52.23	< 0.001	0.684	69.4 (54.4–81.3) [34/49]	98.1 (88.8–99.9) [53/54]	97.1 (83.4–99.9) [34/35]	77.9 (65.9–86.7) [53/68]
Hypervascularity of the upstream bowel	25.68	< 0.001	0.399	38.8 (25.5–53.8) [19/49]	100.0 (91.7–100.0) [54/54]	100.0 (79.1–100.0) [19/19]	64.3 (53.0–74.2) [54/84]
Combined method of hydrocolonic sonography	80.43	< 0.001	0.883	91.8 (79.5–97.4) [45/49]	96.3 (86.2–99.4) [52/54]	95.7 (84.3–99.3) [45/47]	92.9 (81.9–97.7) [52/56]

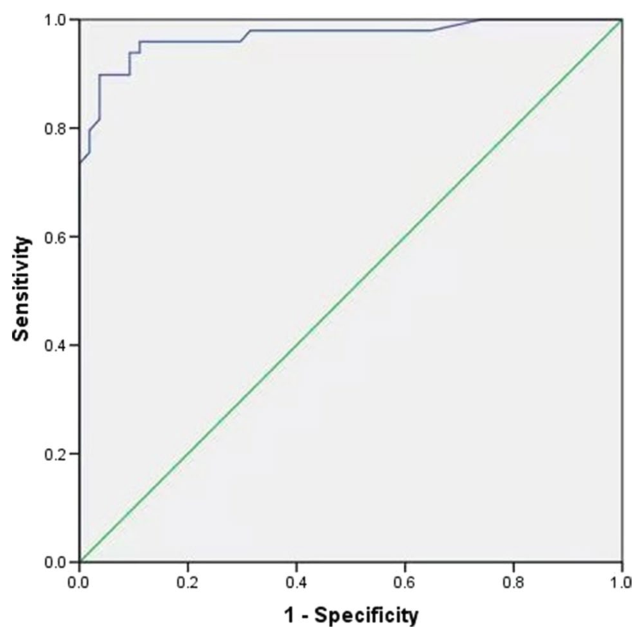

Fig. 5 Receiver operating characteristic (ROC) curve analysis of the luminal diameter ratio for predicting HD

between hydrocolonic sonography and contrast enema is shown in Table 3. Inversion of the RSI was detected only in 2/6 of the long-segment HDs, while all 6 cases had an abnormal luminal diameter ratio; the TZ was also detected by both US and contrast enema in all of them.

In four of the HD patients, there was a false impression of a normal hydrocolonic sonograph. All of them were less than 1 year of age (2 to 7 months). Two of them were detected by contrast enema, and the other two were also negative on contrast enema. Two subjects with increased luminal diameter ratios of 1.60 and 1.67, respectively, both presenting TZ on US and both of whom had a positive RSI and TZ on contrast enema, were erroneously considered to have HD. The following biopsy demonstrated the presence of ganglion cells, and conservative therapies were recommended.

Clinical outcomes

Forty-nine patients underwent the laparoscopic-assisted endorectal pull-through procedures. These non-HD cases showed improvement with conventional conservative therapy during follow-up of 1–6 months except one 6-year-old girl who did not show any improvement after 6 months, during which recurrent faecal impaction occurred. Repeated rectal biopsy showed the presence of ganglion cell as before. This patient received laparoscopic-assisted endorectal pull-through procedure considering possible negative influences of constipation on both quality of life and physical development, and no constipation developed after surgery.

Discussion

Our analysis of 103 patients with suspected HD provided a new method to detect the morphological changes associated with HD. As identification of the distal rectum (the site commonly involved in HD) by trans-abdominal US is challenging due to its deep location, we used a transperineal approach to obtain sonographic images of this region. This is the second novel aspect of our study. By combining the luminal diameter ratio and two minor criteria, we obtained high sensitivity and specificity of 91.8% and 96.3%, respectively.

Hydrocolonic sonography has been reported to be effective in the detection of neoplastic lesions and inflammatory

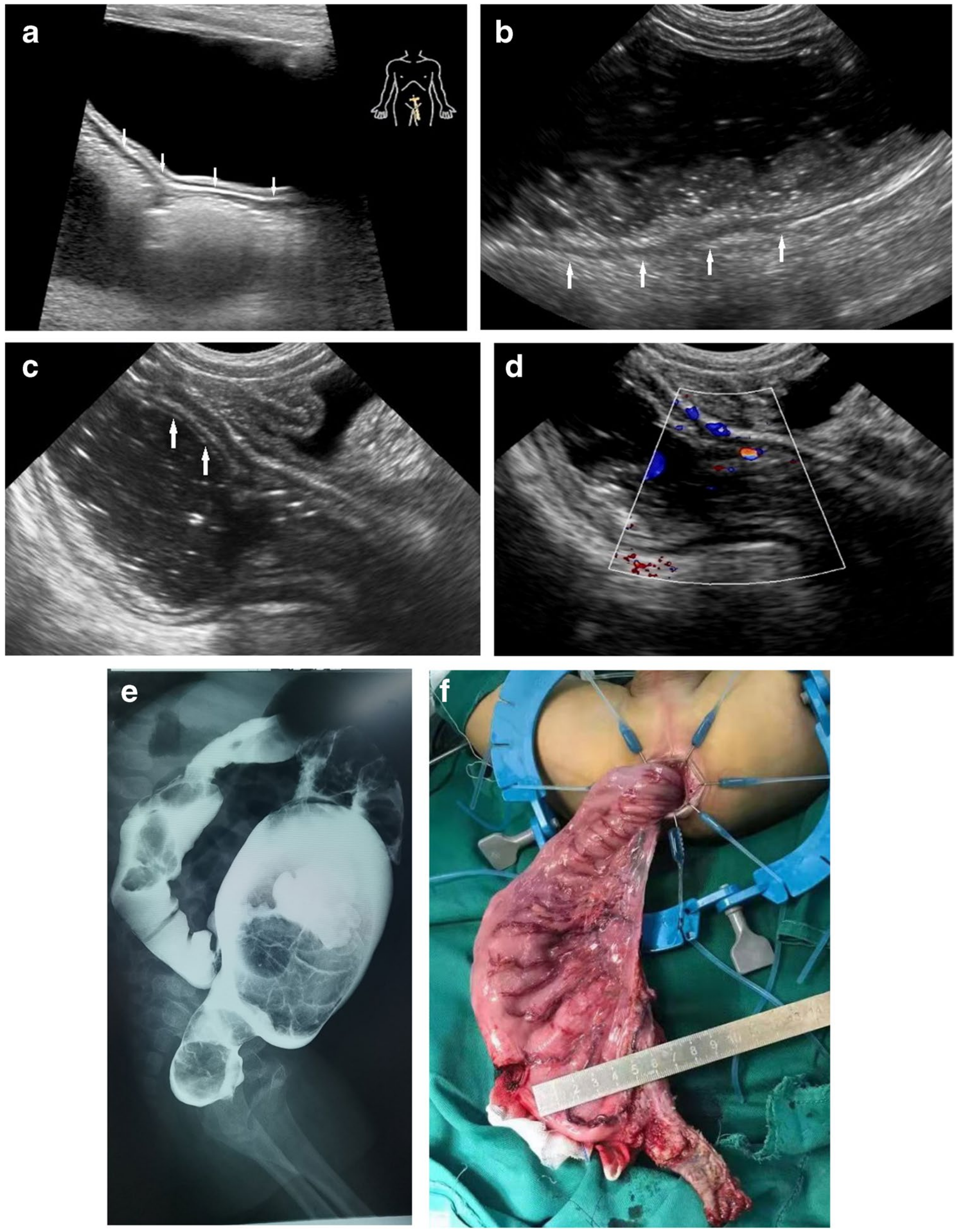

Fig. 6 A 3-year-old boy with refractory constipation. **a** This conventional US image shows a bright reflective surface (arrows) created by intraluminal gas in the rectosigmoid region that obscures the underlying anatomy. **b** A funnel-shaped TZ is clearly depicted in the longitudinal sonogram of the rectosigmoid region on hydrocolonic sonography. Retained faecal particles can be detected because bowel preparation was not mandatory in our study. **c** The bowel wall thickness (arrows) is shown in the lower part of the TZ on longitudinal sonogram. **d** This longitudinal colour Doppler ultrasound image depicts increased vascularity within the thickened bowel wall. **e** True lateral projection on contrast enema shows the TZ in the rectosigmoid colon and dilated upstream bowel. **f** The dilated colon, narrowed segment, and intervening TZ were observed in the operative photograph

Table 2 The contrast enema characteristics of HD. Data in parentheses are 95% confidence intervals, and data in brackets are numerators and denominators. *RSI*, rectosigmoid index; *TZ*, transitional zone; *PPV*, positive predictive value; *NPV*, negative predictive value; *CI*, confidence interval

	χ^2	*p* value	Kappa	Sensitivity %	Specificity %	PPV %	NPV %
RSI	54.5	< 0.001	0.727	83.7 (69.8–92.2) [41/49]	88.9 (76.7–95.4) [48/54]	87.2 (73.6–94.7) [41/47]	85.7 (73.2–93.2) [48/56]
TZ	67.1	< 0.001	0.805	85.7 (72.1–93.6) [42/49]	94.4 (83.7–98.6) [51/54]	93.3 (80.7–98.3) [42/45]	87.9 (76.1–94.6) [51/58]
Final results of contrast enema	63.930	< 0.001	0.787	91.8 (79.5–97.4) [45/49]	87.0 (74.5–94.2) [47/54]	86.5 (73.6–94.0) [45/52]	92.2 (80.3–97.5) [47/51]

diseases. One prospective study performed in 90 subjects showed that hydrocolonic sonography had 100% sensitivity and 81.8% specificity for identifying patients with active inflammatory bowel disease [17]. For the detection of colonic neoplastic lesions in adults, although a high specificity of 98 to 100% has been observed, the sensitivity

Table 3 Comparison of diagnostic performances between hydrocolonic sonography and contrast enema in HD. Data in parentheses are 95% confidence intervals, and data in brackets are numerators and denominators. *RSI*, rectosigmoid index; *TZ*, transitional zone; *PPV*, positive predictive value; *NPV*, negative predictive value; *CI*, confidence interval

	Sensitivity %	*p* value	Specificity %	*p* value	PPV %	*p* value	NPV %	*p* value
Ratio								
Luminal diameter ratio	89.8 (77.0–96.2) [44/49]	0.51	96.3 (86.2–99.4) [52/54]	0.13	95.7 (84.0–99.2) [44/46]	0.28	91.2 (80.0–96.7) [52/57]	0.36
RSI	83.7 (69.8–92.2) [41/49]		88.9 (76.7–95.4) [48/54]		87.2 (73.6–94.7) [41/47]		85.7 (73.2–93.2) [48/56]	
TZ								
TZ on hydrocolonic sonography	81.6 (67.5–90.8) [40/49]	0.77	96.3 (86.2–99.4) [52/54]	1.00	95.2 (82.6–99.2) [40/42]	1.00	85.2 (73.3–92.6) [52/61]	0.67
TZ on contrast enema	85.7 (72.1–93.6) [42/49]		94.4 (83.7–98.6) [51/54]		93.3 (80.7–98.3) [42/45]		87.9 (76.1–94.6) [51/58]	
Final results								
Combined method of hydrocolonic sonography	91.8 (79.5–97.4) [45/49]	1.00	96.3 (86.2–99.4) [52/54]	0.06	95.7 (84.3–99.3) [45/47]	0.22	92.9 (81.9–97.7) [52/56]	1.00
Final results of contrast enema	91.8 (79.5–97.4) [45/49]		87.0 (74.5–94.2) [47/54]		86.5 (73.6–94.0) [45/52]		92.2 (80.3–97.5) [47/51]	

of hydrocolonic sonography varied from 24.2 to 97.5%, depending on the study design, disease stage, site, and size of the lesion [13, 18–20]. On the other hand, in children with rectal bleeding, hydrocolonic sonography exhibited better performance for the detection of colorectal polyps, with increased sensitivity from 37% by conventional US to 89% [21], although the sensitivity of conventional US in this study was lower than that in a previous report (47%) [22]. Assessing the pathological changes of HD by hydrocolonic sonography is not that challenging compared with assessing neoplastic and inflammatory disease. Bowel preparation, such as fasting or laxative intake, was not mandatory in our study because retained stool, which could mask polyps and tumours of the colon, would not influence the depiction of morphological features in HD, such as luminal dilation and narrowing.

In HD patients, the qualitative assessment of colonic dilatation by hydrocolonic sonography based on personal experience is ill-defined. Furthermore, there was a clear relationship between age, height, and the rectal diameter, and no generally accepted definition for colonic dilation is currently available. Thus, we used the ratio instead of absolute values to standardise colonic size for age and height. The results indicated that the luminal diameter ratio achieved the highest diagnostic value. It had an increased specificity of 96.3% compared with 83.3% for the subjective assessment but a slightly decreased sensitivity (from 95.9 to 89.8%). Notably, the luminal diameter ratio differs from the widely used radiographic parameter RSI. RSI has a reported sensitivity of 75–90% and specificity of 83.3–95% [9, 23, 24]. RSI achieved a sensitivity of 83.7% and specificity of 88.9% in the present study, which were in keeping with previously published data. Radiographic RSI is an effective tool for evaluating rectosigmoid HD but is not a reliable item used in long-segment disease [23, 25]. One study showed that RSI was negative in all 3 long-segment HDs [23]. In our study, RSI inversion was detected in only 2/6 of the long-segment HDs, while all 6 cases had an abnormal luminal diameter ratio and TZ (on hydrocolonic sonography and contrast enema). The luminal diameter ratio provided information on both the distended proximal colon and the aganglionic segment, which may enhance the diagnostic potential of hydrocolonic sonography for both rectosigmoid and long-segment HD.

An abundant vascular network could be seen on the outer surface of the TZ and/or dilated segment during surgery. Pathologically, the proliferation of blood vessels is a common finding as well. We supposed that increased vascularity might be detected on US. However, it appeared in only 38.8% of them, probably due to the technical limitations of US. Intravenous contrast-enhanced ultrasound might have the potential to assist in the visualisation of blood flow and improve the sensitivity. Hypervascularity was uniquely detected in the HD group, which means that the presence of hypervascularity of the upstream bowel may lead to a diagnosis of HD, whereas the absence of this sign does not reliably rule out HD. US allows the assessment of intestinal wall thickness and stratification—similar to hypervascularity such information cannot be provided by contrast enema. Assessment of upstream bowel wall thickening is an essential component of the examination in HD because colonic wall hypertrophy might reflect severe chronic obstruction and probably results in aggressive resection of the proximal dilated segment. In the initial stage of the disease, the bowel wall thickness might be too subtle to be detected by ultrasound in the current form. This may partly explain the relatively low sensitivity (69.4%) of this sign.

Comprehensive assessment of four minor criteria with the luminal diameter ratio provided information on the lumen, wall morphology, and blood perfusion of HD, and thereby increased the diagnostic accuracy to levels higher than those obtained by using each indicator alone. The diagnostic performance of this combined method was comparable to that of contrast enema, and the specificity was numerically higher than that of contrast enema, although without statistical significance. Hydrocolonic sonography failed to identify 4 cases of HD. All of these patients were less than 1 year old. In young infants, the pathological change is more difficult to demonstrate, and a rectal biopsy is mandatory in such circumstances.

Limitations of this study should be discussed. Because of safety considerations, neonates were not included in our study. The presentation and diagnosis of HD in the neonatal period are very different from those in older children [26, 27]. The false-negative rate of radiological studies is relatively high in the newborn period [23]. Further investigation is needed to determine the diagnostic performance of hydrocolonic sonography in neonates. Second, bowel wall hypertrophy and hyperaemia have also been described in many other conditions, including inflammatory bowel diseases and gastrointestinal involvement of Henoch-Schonlein purpura (HSP). We did not include such patients, and the role of hydrocolonic sonography in differentiating those conditions was not addressed, as this was not the aim of our study. Third, although the second reader reviewed the US images and video recordings, hydrocolonic sonography examination was performed by only one person, and observer bias cannot be avoided.

In conclusion, in our analysis of 103 patients with HD, we first introduced five sonographic parameters based on hydrocolonic sonography to assess the structural and vascularity changes caused by HD. The combination of those criteria showed good performance in differentiating HD from other causes of chronic constipation and had excellent congruence with contrast enema. In the appropriate setting, hydrocolonic sonography may be an alternative screening method for HD in a large group of children with constipation.

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Declarations

Guarantor The scientific guarantor of this publication is Qi Li.

Conflict of interest The authors of this manuscript declare no relationships with any companies whose products or services may be related to the subject matter of the article.

Statistics and biometry No complex statistical methods were necessary for this paper.

Informed consent Written informed consent was obtained from the guardians of each patient (child) in this study.

Ethical approval Institutional Review Board approval was obtained.

Methodology • prospective

• diagnostic or prognostic study
• performed at one institution

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