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# Point-of-Care Ultraschall zur Untersuchung des transrektalen Durchmessers bei Kindern mit kolorektalen Erkrankungen: Betrachtung von Bowel Management im Gesamtkonzept der ganzheitlichen, standardisierten und patientenzentrierten Versorgung von Kindern mit Morbus Hirschsprung

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# 1 Synopsis

## 1.1 Einleitung

Morbus Hirschsprung ist eine seltene angeborene Fehlbildung des Darms, bei der die Ganglienzellen in der Darmwand fehlen. Morbus Hirschsprung betrifft etwa 1 von 5000 Lebendgeborenen weltweit. In Deutschland werden jährlich 150 bis 200 Kinder mit Morbus Hirschsprung geboren. Die Versorgung erfolgt in Deutschland bisher nicht zentralisiert (1).

Diese Fehlbildung führt zu einer chronischen Motilitätsstörung des Darms, die einem Darmverschluss ähnelt. Klinische Symptome treten meist im ersten Lebensjahr auf, können aber auch erst bei älteren Kindern offensichtlich werden (2).

Neben der fachgerechten Durchführung der korrigierenden Operation benötigen die Patienten eine individuell angepasste Langzeitnachsorge nach einem standardisierten Schema, einschließlich Bowel Management, um soziale Kontinenz zu erreichen (3,4).

Während der fehlgebildete aganglionäre Darm mit mehreren gut etablierten chirurgischen Techniken sicher entfernt werden kann, wird zunehmend deutlich, dass ein erheblicher Anteil der Patienten mit Morbus Hirschsprung trotz technisch adäquater Durchführung der Operation im Langzeitverlauf medizinische Probleme und Einschränkungen der Lebensqualität erleiden (5,6).

Eine Beeinträchtigung der Lebensqualität wird durch persistierende Stuhlentleerungsstörungen und Stuhlschmieren verursacht. Diese Symptome können durch adäquates Bowel Management verbessert werden (7–10). Bowel Management ermöglicht durch auf den Patienten angepasste supportive Therapien wie Stuhlmodulation mit Laxanzien, Irrigation[1] und Suppositorien eine regelmäßige gewollte kontrollierte Stuhlentleerung. Das Ziel von Bowel Management ist zwischen den Stuhl Entleerungen im Sinne von sozialer Kontinenz sauber zu sein (7-10).

International wird empfohlen, das Bowel Management mit Röntgenuntersuchungen zu begleiten (7,11–13). Bereits 2016 wurde über Fehlinterpretationen von Stuhlretention in Röntgenuntersuchungen berichtet (14). Die fehlende Evidenz für die

1 Unter rektaler Irrigation wird die Dekompression des Darms mit Hilfe eines Katheters und Spülflüssigkeit verstanden.

Nutzung von Röntgen zur Diagnose einer Obstipation wurde kürzlich (2024) in einem systematischen Review adressiert (15).
Die sonographische Bestimmung des transrektalen Durchmessers wurde für die funktionelle Obstipation bereits beschrieben (15-19), jedoch wurden Kinder mit einer kolorektalen Erkrankung wie Morbus Hirschsprung oder Analatresie systematisch anhand der Ausschlusskriterien aus diesen Arbeiten ausgeschlossen (8,16).
Bisher ist unklar, ob die Messung des sonographischen transrektalen Durchmessers bei Kindern mit kolorektalen Erkrankungen genutzt werden kann, um eine Stuhlretention zu visualisieren. Es gibt bisher keine Untersuchung zu einer sonographischen Beurteilung des “Neorektums” in dieser Patientengruppe und dementsprechend wie dieses nach der Operation bildmorphologisch aussieht (16).
Unsere Arbeitsgruppe hat erstmalig den transrektalen Durchmesser (TRD) bei Kindern mit kolorektalen Erkrankungen, insbesondere Morbus Hirschsprung, untersucht, um a) eine Stuhlretention zu bestimmen und b) ein Langzeit-Monitoring zu ermöglichen. Die sonographische Untersuchung durch KlinikerInnen wurde als Point-of-Care Untersuchung prospektiv erfasst und für Kinder mit kolorektalen Erkrankungen validiert (8,16).
Der Einfluss von Ernährung auf die abdominellen Beschwerden, Darmfunktion und Bowel Management wurde durch unsere Arbeitsgruppe in einer „Patient Reported Outcome Measurement“ (PROM) Umfrage adressiert (17).

Der kumulativen Habilitationsschrift liegen die Arbeiten mit den folgenden Fragestellungen zugrunde:

a) Beschreibung und Evaluation von Bowel Management zur Vorbereitung, perioperativen Begleitung und postoperativen Nachsorge bei Kindern mit Morbus Hirschsprung; Lindert, 2024 (3)

b) Kann die strahlenfreie Durchführung der transrektalen Sonographie für Patienten mit kolorektalen Erkrankungen wie M. Hirschsprung genutzt werden?; Erkel, 2024 & Lindert, 2024 (8,16)

c) Point-of-Care Ultraschall in der Kinderchirurgie; Lindert 2023, Naidoo, 2024 (18,19)

d) Ernährung und Einfluss auf abdominelle Symptome und Bowel Management bei Kindern mit M. Hirschsprung; Lindert, 2024 (17)

e) Standardisierte Erfassung: Continence Score bei Kindern mit M. Hirschsprung; Lindert, 2024 (20)

## 1.2 Bowel Management bei Kindern mit M. Hirschsprung

Unsere Arbeitsgruppe beschreibt klinische Behandlungsempfehlungen für das standardisierte präoperative, perioperative und postoperative Bowel Management bei Kindern mit Morbus Hirschsprung. Unsere Studie evaluiert das institutionelle perioperative Bowel Management Protokoll anhand der klinikeigenen Kohorte. Es wurden Patienten mit einer primären Durchzugsoperation eingeschlossen (3).

Für die präoperative Phase hebt unsere Studie, analog der ERNICA Guideline, die Bedeutung der transanalen Irrigation zur effektiven Darmdekompression und Vorbereitung auf die Operation hervor (1,3). Unsere Studie betont die Relevanz eines effektiven Bowel Managements, um Komplikationen wie eine Enterokolitis zu vermeiden. Die Eltern werden hierbei durch das medizinische Personal geschult, die rektale Irrigation täglich selbständig zu Hause durchzuführen (2,3,21). Wie auch andere Arbeitsgruppen empfehlen wir entsprechend der ERNICA Empfehlung, die Eltern zur selbständigen rektalen Irrigation anzuleiten. Eine japanische Arbeitsgruppe berichtet hingegen, einen rektalen Irrigationskatheter dauerhaft für die Zeit der präoperativen Irrigation zu fixieren (22).
Die Durchführung der rektalen Irrigation durch die Eltern kann Einfluss auf die Familiendynamik haben (23,24). Die Unterstützung der Familien durch geschultes Personal wie Fachpflegekräfte (25,26) oder Selbsthilfeorganisationen beziehungsweise Patientenvertretungen (27) hat sich als hilfreich herausgestellt.
Trotz der Empfehlung zur Vorbereitung auf die korrigierende Durchzugsoperation mit transanaler Irrigation ohne Stomaanlage geben 30% der europäischen KinderchirurgInnen in einer Umfrage an, eine Stomaanlage als initiale Therapie durchzuführen (28).
Unsere Arbeit zeigt unter anderem, dass die transanale Irrigation in der frühen postoperativen Phase sicher genutzt werden kann (3,29).

Es ist nicht ungewöhnlich, auch nach einer korrekt durchgeführten Operation Symptome von Stuhlentleerungsstörungen und Stuhlschmieren vorzufinden (5,11,30–32). Miteinbeziehung von Bowel Management in eine Standardnachsorge unterstützt das Ziel der sozialen Kontinenz.
Das Leben mit einer chronischen Erkrankung wie der Hirschsprung Erkrankung kann langfristige Auswirkungen auf die psychosoziale Gesundheit der betroffenen Kinder und ihrer Familien haben (24,33). Daher ist eine patientenzentrierte, umfassende Betreuung entscheidend, um Bewältigungsmechanismen zu fördern (3,24,25,25).

## 1.3 Evaluation und Validierung der sonographischen Bestimmung des transrektalen Durchmessers

### 1.3.1 Transrektale Ultraschallmessung bei Kindern mit kolorektalen Erkrankungen

Es ist bekannt, dass Kinder mit einer kolorektalen Erkrankung eine Langzeitbetreuung unter Einbeziehung von Bowel Management zum Erreichen der sozialen Kontinenz benötigen (7,8,11,21,34).
Insbesondere US- amerikanische Gruppen empfehlen die Evaluation der Stuhllast mit Röntgendarstellungen (7,12,13,34).
Die sonographische Darstellung der Stuhlretention mit dem transrektalen Durchmesser ist für die funktionelle Obstipation bereits beschrieben (35–38) und kürzlich durch ein Review (2024) als strahlenarme Diagnostik empfohlen worden (15).
Unsere Arbeitsgruppe entwickelte das Ultraschallprotokoll von Kijin (39) unter Rektum Kinder Sonographie (ReKiSo) für Kinder mit kolorektalen Erkrankungen weiter und beschreibt erstmalig die Anwendung von transrektalem Ultraschall im Rahmen von Bowel Management für diese Patientengruppe (8,16).

Bisher haben die vorhandenen Studien systematisch Kinder mit kolorektalen Erkrankungen ausgeschlossen (35–38). Somit gibt es keine Beschreibung der Machbarkeit und Validierung des sonographischen transrektalen Durchmessers (TRD) bei Kindern mit kolorektalen Erkrankungen. Der erste Teil dieser Proof-of-

Concept Studie konzentriert sich auf die Evaluation des sonographischen transrektalen Durchmessers (TRD) bei Kindern mit kolorektalen Erkrankungen (n=302) (16). In einer prospektiven Studie von über 300 Kindern konnten wir die von Klijn (39) (2004) beschriebene Point-of-Care Ultraschalluntersuchung für Kinder mit kolorektalen Erkrankungen validieren (16).

Wir konnten die Machbarkeit einer routinemäßigen Durchführung der sonographischen Bestimmung des transrektalen Durchmessers durch KlinikerInnen in der regulären Sprechstunde zeigen. Dies ist die erste Beschreibung einer sonographischen Visualisierung des Neorektums bei Kindern mit kolorektalen Erkrankungen. Darüber hinaus wird erstmalig die Bestimmung des transrektalen Durchmessers bei Kindern mit anorektalen Malformationen, Hirschsprung Erkrankung und anderen kolorektalen Erkrankungen wie neurogene Darmentleerung Störung (beispielsweise bei Spina bifida oder Kaudalen Regressionssyndrom u.a.) in der Langzeitnachsorge zum Monitoring von Bowel Management beschrieben (8,16).

Unsere Ergebnisse zeigen einen signifikanten Zusammenhang zwischen dem für die funktionelle Obstipation in anderen Studien verwendeten transrektalen Durchmesser mit einem Wert von über 3 cm und Symptomen der Stuhlretention. Wir konnten zeigen, dass symptomfreie Kinder mit anorektaler Malformation, Hirschsprung Erkrankung oder neurogener Darmentleerungsstörung ähnliche TRD-Werte aufweisen wie Kinder der Kontrollgruppe ohne kolorektale Erkrankung (8,16). Ein Wert über 3 cm zeigte eine Stuhlretention bei Kindern mit kolorektalen Erkrankungen ebenso wie bei Kindern mit funktioneller Obstipation an (16,35–38). Unsere gewählten Cut-off Werte von 3 cm ergaben eine Sensitivität von 95,89% und eine Spezifität von 72,84% (16).

Zusätzlich kann die sonographische Untersuchung als Point-of-Care Untersuchung eingebettet in die Vorstellung in der Klinik direkt durch die behandelnden KlinikerInnen durchgeführt werden und somit eine schnelle Visualisierung sowohl für die ÄrztInnen als auch die Familie ermöglichen.

### 1.3.2 Transrektale Ultraschallmessung bei Kindern mit M. Hirschsprung

Bei bis zu 80 % der Kinder mit Hirschsprung Erkrankung treten postoperativ Stuhlentleerungsstörungen oder Obstipation auf (2,4,5,30). Wenn anatomische oder mechanische Ursachen ausgeschlossen sind und keine Reoperation indiziert ist, benötigen diese Kinder ein standardisiertes individuelles Bowel Management. Bowel Management zur Förderung der sozialen Kontinenz sollte in die langfristige Nachsorge integriert sein (7,8,13,20,34,40,41). Das Therapieziel ist das Erreichen von sozialer Kontinenz zur Ermöglichung der Teilhabe am sozialen Leben. Durch soziale Kontinenz können negative Reaktionen der Menschen in der Umwelt vermieden werden. Soziale Kontinenz kann erschwert werden durch Probleme wie Stuhlschmieren und Verstopfung (6-8). Zudem sollten Probleme wie aufgetriebenes Abdomen und das Auftreten einer Hirschsprung-assoziierten Enterokolitis verhindert beziehungsweise frühzeitig erkannt werden (7-9,14,44–48).

Zur Beurteilung der Stuhlretention bei kolorektalen Erkrankungen wie der Hirschsprung Erkrankung werden Röntgenaufnahmen und radiologische Kolon-Kontrasteinläufe durch andere Arbeitsgruppen empfohlen (11–13,48). Diese Methoden setzen die Kinder jedoch ionisierender Strahlung aus. Potenzielle Langzeitfolgen einschließlich eines erhöhten Malignom Risikos können auftreten (49,50). Da Kinder mit kolorektalen Erkrankungen häufig wiederholt evaluiert werden, untersuchte unsere Arbeitsgruppe die Möglichkeit einer strahlenfreien Methode zur Bestimmung der Stuhlfüllung und zum Monitoring des Bowel Managements (8,16).

Im Rahmen der korrigierenden Operation wird das native Rektum entfernt, wobei bisher die sonographische Visualisierung des durchgezogenen Darms im Sinne des Neorektum unbekannt war (8). Es gab bisher keine Studie zur Messung des sonographischen transrektalen Durchmessers bei Kindern mit Hirschsprung Erkrankung.

Wir führten eine prospektive Studie unter Einschluss von 193 Kindern (im Alter von 3 bis 16 Jahren, darunter 60 Kinder mit Morbus Hirschsprung, 51 mit funktioneller Verstopfung und 51 gesunde Kontrollpersonen) durch. In dieser Arbeit wurden Kinder mit einer totalen Kolonaganglionose ausgeschlossen. Wir konnten zeigen, dass Kinder mit Verstopfungs- oder Stuhlschmierproblemen sowohl mit Hirschsprung Erkrankung wie auch funktioneller Verstopfung signifikant erhöhte sonographische

TRD-Werte haben. Ein TRD von mehr als 3 cm korrelierte, stark mit Verstopfung und Stuhllast, während ein TRD unter 3 cm eine normale Darmentleerung anzeigte (8).
Wir konnten zeigen, dass die sonographische Messung des TRD nicht nur als einzelne Untersuchung die Stuhlfüllung anzeigt, sondern ebenso geeignet ist, das Bowel Management bei Kindern mit Hirschsprung Erkrankung zu begleiten und den Therapieerfolg zu visualisieren (8). Ein erhöhter TRD normalisierte sich durch adäquates Bowel Management auch bei Kindern mit Hirschsprung Erkrankung, wie bereits bei Kindern ohne Hirschsprung Erkrankung mit funktioneller Obstipation bekannt ist (8,36).
Diese Ergebnisse zeigen, dass der sonographische TRD eine verlässliche Methode zur Beurteilung der Stuhlfüllung und zur Überwachung des Bowel Managements bei Kindern mit Hirschsprung Krankheit ist. Eine Untersuchung mittels Ultraschall bietet daher eine schnelle, nicht-invasive Beurteilung ohne Strahlenbelastung direkt in der ambulanten Sprechstunde.

## 1.4 Point-of-Care (PoC) Ultraschall in der Kinderchirurgie

Point-of-Care Untersuchungen ermöglichen eine direkte Beantwortung einer Fragestellung am Patientenbett. Die Anwendung von Point-of-Care Ultraschalluntersuchungen (POCUS) durch nicht-radiologische ÄrztInnen ist eine gezielte Untersuchung, die eine spezifische medizinische Frage mittels Ultraschall beantwortet (18,51,52). Ultraschall ist bei Kindern eine bevorzugte Bildgebungsmethode, da dieser eine hohe Sensitivität bietet und keine ionisierende Strahlung verwendet (53–56). Viele chirurgische Erkrankungen bei Kindern können mit Ultraschall diagnostiziert werden (53,57).
Geschulte KlinikerInnen können beispielsweise bei Indikationen wie Appendizitis und Invagination POCUS mit hoher Zuverlässigkeit durchführen, was zu einer verkürzten Diagnosezeit für die PatientInnen führt (53,57–61). Vermittlung von POCUS-Fähigkeiten an ChirurgInnen zur Diagnose häufiger kinderchirurgischer Erkrankungen kann nicht nur die Zeit bis zur Diagnose und Behandlung verkürzen, sondern auch die Kosten der Versorgung und damit den Zugang zur Versorgung verbessern (19).
Es existieren derzeit bereits einige POCUS Protokolle für kinderchirurgische Erkrankungen wie beispielsweise Appendizitis, Darminvagination und hypertrophe

Pylorusstenose. Diese diagnostischen Anwendungen sind erfolgreich in die klinische Routine integriert (58,59,62,63). In der pädiatrischen Notfallmedizin wird das Potenzial von POCUS zunehmend anerkannt, da es eine globale Verbesserung der Versorgung pädiatrischer PatientInnen ermöglicht. Die Anwendung von Ultraschall durch KlinikerInnen kann chirurgische Prozeduren wie beispielsweise Abszessentlastung, Fremdkörperentfernung, Drainageneinlage gezielt und sicher ermöglichen (64). Die konkrete Anwendung durch KinderchirurgInnen in Europa (18) und global ist jedoch bislang unklar (19).

Unsere Umfragen zeigen, dass die Mehrheit der sich an der Studie beteiligten KinderchirurgInnen (90%; 159/176) an einer formalen POCUS Schulung teilnehmen würde, wenn ein POCUS Programm angeboten werden würde. Obwohl ein beträchtlicher Anteil der ChirurgInnen gewisse Ultraschallkenntnisse angibt, haben die meisten ihre Fähigkeiten informell entwickelt. Bei denjenigen, die eine Schulung (formell oder informell) absolviert haben, setzt die Mehrheit ihre Fähigkeiten routinemäßig in der Praxis ein (18,19,52).

In den Umfragen werden als Hürden und Hindernisse vor allem die Überlastung lokaler Radiologieabteilungen und der Mangel an geschultem Personal (18,19) angegeben.

In Ländern mit niedrigem und mittlerem Einkommen (LMIC) stellt zudem der Kostenfaktor für Ultraschalluntersuchungen eine erhebliche zusätzliche Hürde dar. In vielen dieser Einrichtungen ist der Zugang zu medizinischer Versorgung nur gegen Vorauszahlung möglich, was bestimmte Untersuchungen für ärmere Familien unerschwinglich macht (19,65).

Die Stärkung von Trainingsmodulen und POCUS für kinderchirurgische Erkrankungen könnten die Patientenversorgung erleichtern und die Diagnostik beschleunigen (18,19, 52).

Unsere Untersuchungen zur Point-of-Care Untersuchung der sonographischen Bestimmung des transrektalen Durchmessers ergänzen die bestehenden POCUS Anwendungen. Es wird erstmals eine standardisierte POCUS Methode für die Evaluation von Stuhlbelastung bei kolorektalen Erkrankungen validieren (8,16).

Die Teilnehmenden sowohl der europäischen als auch der globalen Umfrage äußerten großes Interesse daran, diese strahlenfreie Methode zur Diagnostik von Stuhlretention zu erlernen (18,19).

## 1.5 Zusammenhang zwischen Ernährung und Darmfunktion bei M. Hirschsprung

Viele Familien von Kindern mit Hirschsprung Erkrankung berichten von einem Zusammenhang zwischen bestimmten Ernährungsbestandteilen, abdominellen Beschwerden und beeinträchtigter Darmfunktion (17,66). Bis zu 80% der Kinder mit M. Hirschsprung berichten über Verstopfung und Stuhlentleerungsstörung (5,8,21,30,67-70). Derzeit gibt es keine spezifischen Ernährungsempfehlungen für Patienten mit einer Hirschsprung Erkrankung (17).

Erstmals haben Untersuchungen von Telborn et al. in einem schwedischen Kollektiv (13 PatientInnen) die Einflüsse spezifischer Nahrungsmittel auf abdominelle Symptome sowie die Anwendung von Eliminationsdiäten erfragt (71,72).

Unsere Arbeitsgruppe führte eine multizentrische, internationale Patientenbefragung (Patient reported Outcome Measurement (PROM)) in sieben europäischen Ländern (Deutschland, Großbritannien, Italien, Niederlande, Österreich, Polen, Schweden) durch.

Der von unserer Arbeitsgruppe entwickelte und validierte Fragebogen wurde in den jeweiligen Landessprachen (deutsch, englisch, niederländisch, polnisch, schwedisch und italienisch) zur Verfügung gestellt und über die lokalen Selbsthilfegruppen an die Familien gesendet. Insgesamt konnten wir die Antworten von 563 PatientInnen in der bis dato teilnehmerstärksten und einzigen internationalen Studie mit einbeziehen. Wir befragten die PatientInnen beziehungsweise deren Eltern direkt zur Darmfunktion, zu abdominellen Beschwerden, Bowel Management und beobachteten Zusammenhänge zwischen einzelnen Lebensmitteln (17).

33% der Befragten unseres multinationalen Patientenkollektivs gaben die Länge des aganglionären Anteils als kurz (nur das Rektosigmoid betreffend), 45% als lang und 21% als totale Kolonaganglionose mit in 10% relevanter Dünndarmbeteiligung an. Insgesamt gaben 90% der Befragten an, eine gemischte Ernährung zu sich zu nehmen. 61% der Patienten/ Familien gaben an, dass die Diagnose von Morbus Hirschsprung ihre Ernährungsgewohnheiten verändert habe, und 77% konnten spezifische Lebensmittel oder Zutaten identifiziert, die ihre Symptome beeinflussten. Beide Aspekte traten signifikant häufiger bei PatientInnen mit totaler Kolonaganglionose auf (17). 80% der Befragten gaben an, den Einfluss der verschiedenen Lebensmittel auf die Darmfunktion selbst ohne Beratung von

ÄrztInnen oder Ernährungsberatern herausgefunden zu haben. Eine ballaststoffreiche Ernährung wurde von 24 % der Teilnehmenden und eine ballaststoffarme Ernährung von 18% der Teilnehmenden befolgt (17). Ein Großteil der PatientInnen identifizierte spezifische Lebensmittel, die ihre Symptome verschlimmerten, hierunter waren Hülsenfrüchte (54%), Zucker (48%), Früchte (39%) sowie Milchprodukte (30%). Bei der Analyse der berichteten unerwünschten Darmbeschwerden war Durchfall die häufigste Symptomatik, die insbesondere von Milchprodukten (51%) und raffiniertem Haushaltszucker (36%) ausgelöst wurde (17).
Vergleichsweise weniger Patienten berichteten analog den Ergebnissen der mononationalen Studie in Schweden über Symptome im Zusammenhang mit Eiern (9 %), Weizen/ Gluten (12%) und Soja (10%) (17,66,72).
Unsere Studie untersucht ein für Betroffene und Familien wichtiges Thema. Familien betonen, dass sie einen relevanten Einfluss der Ernährung und Nahrungsmittel auf die Darmfunktion ihres Kindes beobachten (17,66).
Darüber hinaus verwendeten 31 % der befragten Patienten Probiotika (17). Probiotika wurden insbesondere bei langstreckigem Morbus Hirschsprung verwendet. Besonders Patienten mit langstreckiger Hirschsprung Erkrankung berichteten häufiger von negativen Auswirkungen durch Lebensmittel. Probiotika scheinen eine Rolle bei der Verbesserung dieser Symptome zu spielen (17,73); unsere Befragung konnte diesen Effekt besonders bei Patienten mit vollständig entferntem Dickdarm bestätigen (17).
Der Stellenwert der Ernährung sowie des Mikrobioms bei der Entwicklung beziehungsweise Prävention einer Hirschsprung assoziierten Enterokolitis (HAEC) ist bisher noch nicht hinreichend verstanden (74,75).
Ebenfalls ist die Pathogenese der HAEC noch wenig verstanden, jedoch scheinen eine gestörte Darmmotilität, Dysbiose sowie eine beeinträchtigte Schleimhautabwehr und Darmbarrierefunktion eine wesentliche Rolle zu spielen (74,76,77).
Zusammengefasst zeigen wir in unserer Befragung entsprechend der Ergebnisse anderer Arbeitsgruppen, dass die Ernährung bei Patienten mit Morbus Hirschsprung die Darmfunktion und das Bowel Management sowie abdominelle Symptome beeinflusst und eine zentrale Rolle zur Mitigierung von adversen Symptomen einnimmt (17,66,72,78). Die Ergebnisse dieser multizentrischen Studie in sieben europäischen Ländern unterstreicht die Bedeutung einer multidisziplinären

Langzeitbetreuung, die auch eine Ernährungsberatung einbezieht, um eine ausgewogene Ernährung sicher zu stellen und Symptome zu reduzieren (17,78).

## 1.6 Erfassung der Kontinenz bei Kindern mit M. Hirschsprung: Vor- und Nachteile der Nutzung eines Kontinenzscores

Fortschritte in chirurgischen Techniken und perioperativer Versorgung haben die kurz- und mittelfristigen postoperativen Ergebnisse von Patienten mit Morbus Hirschsprung verbessert. Die Langzeitergebnisse treten immer mehr ins Interesse der behandelnden KlinikerInnen (4,5,31,79–82).

Es wird angenommen, dass die langfristigen Ergebnisse für die Mehrheit der Patienten trotz der bekannten Probleme mit Verstopfung und/ oder Stuhlinkontinenz gut sind (5,30,67,83–86).

Es wird beobachtet, dass sich die funktionellen Ergebnisse verbessern, wenn die Patienten älter werden, insbesondere nach Erreichen der Adoleszenz wird eine Besserung beschrieben (85).

Es gibt bisher keine allgemein anerkannten Leitlinien für postoperative Bowel Management Programme und auch keine einheitlich definierten Nachsorgeleitlinien oder Empfehlungen zur einheitlichen Erfassung der Darmfunktion. Hierdurch wird die Interpretation der berichteten Ergebnisse erschwert (20,79,81,83).

Zur Verbesserung der Evaluierung des Langzeit Outcome verschiedener Gruppen und auch besseren Nachvollziehbarkeit im Langzeitmonitoring individueller Patienten sollte eine standardisierte Erfassung und Beschreibung der Parameter wie Kontinenz und Bowel Management sowie im Rahmen der Transition auch Sexualität und Fertilität mit einbezogen werden (87,88). Wünschenswert wäre eine einheitliche Erfassung der Darmfunktion, welche bisher durch unterschiedliche Scores wie zum Beispiel den Rintala Score, Krykenbeck Score und andere Scores erfasst werden (20,87,89).

Bisher werden solche Scores meist durch die KlinikerInnen erfasst, jedoch ist die routinemäßige Erfassung in den Sprechstunden durch begrenzte Ressourcen in der klinischen Routine nicht immer zuverlässig durchführbar. Dementsprechend bieten Patienten Reported Outcomes (PRO) Erfassungen eine sinnvolle Ergänzung (80).

Zusammen im internationalen europäischen OASIS-Holistic Care in Hirschsprung Disease Netzwerk haben wir die vorhandenen Scores zur Kontinenzerfassung

analysiert und in der Expertengruppe sowie mit Vertretern der verschiedenen teilnehmenden Patientenorganisationen (SOMA Deutschland: Selbsthilfeorganisation für Betroffene von Morbus Hirschsprung und Anorektale Fehlbildungen, Amhori Italien: Italian Association of Hirschsprung's Disease A.Mor.Hi, Niederlande: Vereniging Ziekte van Hirschsprung) diskutiert.

Wir stellten fest, dass die aktuell vorhandenen Scores alle für die anorektalen Malformationen entwickelt wurden und es bisher keinen M. Hirschsprung spezifischen Erfassungsscore für die Darmfunktion gibt (20,89).

Idealerweise wird ein umfassender validierter Score benötigt, um die Darmfunktion bei PatientInnen mit M. Hirschsprung zuverlässig zu bewerten und im Langzeitverlauf zu untersuchen (20). Ein regelmäßiges longitudinales Monitoring der Darmfunktion ist entscheidend, da diese sich physiologisch im Verlauf der kindlichen Entwicklung verändert (5). Viele Studien nutzen selbst definierte nicht validierte Scores (20). Insgesamt sind wir in diesem paneuropäischen Netzwerk übereingekommen, dass sich der Rintala Score derzeit als bereits validierter Score zum Monitoring der Darmfunktion und des Bowel Managements bei Kindern mit M. Hirschsprung anbietet. (20)

Wir empfehlen daher als OASIS-Gruppe, den Rintala Score als geeignetes Instrument zur Bewertung der Darmfunktion bei HD-PatientInnen, da er einfach zu nutzen und gut validiert ist (20). Automatisierte Datenerfassungsinstrumente sollten für eine allumfassender Erfassung im longitudinalen Monitoring zur Verbesserung der kontinuierlichen, objektiven Bewertung entwickelt werden (20,80).

## 1.7 Zusammenfassung und Ausblick

Unsere Studien sind Teil eines patientenzentrierten Ansatzes zur Behandlung von Kindern mit kolorektalen Erkrankungen, insbesondere der Hirschsprung Erkrankung (3,29).

Die bisherigen Studien zeigen vielversprechende Ergebnisse; so unter anderem, dass die POCUS Bestimmung des transrektalen Durchmessers bei PatientInnen mit kolorektalen Erkrankungen eingesetzt werden kann (8,16).

Wir zeigen eine gute Korrelation zwischen den Ultraschallbefunden und klinischen Symptomen der Stuhlretention sowie eine gute bildmorphologische Korrelation des

transrektalen Durchmessers zum Monitoring von Bowel Management bei kolorektalen Erkrankungen wie insbesondere Morbus Hirschsprung (8,16).
Darüber hinaus liefert die multinationale multizentrische direkte Patientenbefragung wertvolle Erkenntnisse über die Darmfunktion und den Einfluss von Ernährungsbestandteilen auf die Darmfunktion bei Kindern mit Hirschsprung Erkrankung (17). Diese Informationen unterstützen uns als KlinikerInnen, um Familien gezielt und fundiert zu beraten.
Die Einbeziehung der Bowel Function sowie die Visualisierung des transrektalen Durchmessers mit Hilfe der Point-of-Care Untersuchung und die standardisierte Erfassung eines Kontinenz Scores - am besten durch die Patienten selbst – unterstützen eine ganzheitliche Langzeitbetreuung (3,8,16,17,).

## 1.8 Abkürzungen und Anmerkung

| | |
|---|---|
| HD | Hirschsprung Disease = Hirschsprung Erkrankung = M. Hirschsprung |
| HAEC | Hirschsprung assozierte Enterokolitiis |
| POCUS | Point-of-Care Ultraschall |
| PROM | Patient Reported Outcome Measurement |
| SOMA | Selbsthilfeorganisation für Betroffene von Morbus Hirschsprung und anorektale Fehlbildungen |
| TRD | Transrektaler Durchmesser |
| TCA | Totale Kolonaganglionose |

In dieser Arbeit wurde bewusst für ÄrztInnen und KlinikerInnen gegendert, um die Realität in den Kliniken abzubilden.
Bei Patienten sind selbstverständlich immer alle Geschlechter gemeint; da die Kinder mit Hirschsprung bekannterweise überwiegend Jungen sind, wurde hier aufgrund der besseren Lesbarkeit auf das Gendern verzichtet

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# 2 Zugrundeliegende Arbeiten

## 2.1 Bowel Management in Hirschsprung Disease - Pre-, Peri- and Postoperative Care for Primary Pull-Through.

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*children*

MDPI

*Article*

# Bowel Management in Hirschsprung Disease—Pre-, Peri- and Postoperative Care for Primary Pull-Through

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**Abstract:** (1) Background: Bowel management contributes throughout the pathway of care for children with Hirschsprung. Preoperative bowel management prepares the child and family for the pull-through surgery. Perioperative bowel management supports early recovery and tailored bowel management in the follow-up supports the achievement of social continence. (2) Methods: We conducted a cross-sectional assessment of our institutional bowel management program to illustrate the pre-, peri- and postoperative bowel management strategies. (3) Results: A total of 31 children underwent primary pull-through, 23 without a stoma and 8 with a stoma, at a median age of 9 months. All children without a stoma were prepared for surgery by using rectal irrigations. Children with a stoma were prepared for surgery with a transfer of stoma effluent. Transanal irrigation supported early recovery. (4) Conclusions: Bowel management is a key pillar of the management of children with Hirschsprung disease. Incorporating bowel management in the pathway of care facilitates primary pull-through and supports perioperative recovery.

**Keywords:** bowel management; Hirschsprung disease; rectal washout; transanal irrigation; preparation for surgery; distal outlet obstruction; fecal incontinence



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### 1. Introduction

Hirschsprung disease (HD) is a congenital malformation of the intestine in which the intestinal wall lacks ganglion cells. This results in a chronic intestinal motility disorder, equivalent to an intestinal obstruction. HD is a rare disease with an incidence of 1 in 2500 live births. A total of 90% of patients with HD are diagnosed in the neonatal/early infant period. The majority (approximately 80%) of cases involve the rectosigmoid [1].

The main treatment goal for HD is to achieve social continence. In addition to specialized surgery, patients require preparation for reconstructive surgery and a standardized but individualized long-term follow-up with bowel management. Thus, bowel management accompanies HD patients from the neonatal period through surgical reconstruction and right through to long-term follow-up. Living with a chronic disease such as HD can have a long-term impact on the psycho-social health of the affected children and their families [2,3]. Therefore, patient-centered, comprehensive care is important for the promotion of coping mechanisms.

To avoid obstructive symptoms and prevent unnecessary episodes of Hirschsprung-associated enterocolitis (HAEC), effective and adequate decompression must be achieved before and after reconstructive surgery. Approximately 30% of HD patients will experience at least one episode of HAEC. Note, however, that there is currently no consent on the exact definition of HAEC [1,4].

According to the ERNICA guideline, patients should receive rectal irrigations 1–3 times a day to decompress the bowel until the definitive pull-through operation is performed, which can be achieved in 75% of patients [1]. A survey of European clinicians found that 30% would create an ostomy awaiting pull-through surgery [5].

The purpose of this study is to describe the standard protocol of a specialized institution and present a descriptive study of patients over two years to review the pre-, peri- and postoperative strategies in patients with standard HD. We describe patient characteristics at presentation, anatomical features, preparation for surgery and our institutional pathway for early perioperative recovery and postoperative rehabilitation.

## 2. Materials and Methods

We conducted a cross-sectional chart review of all patients with HD—excluding total colonic aganglionosis and redo pull-through—who underwent reconstruction between 22 January and 23 December at our center. Ethical approval was obtained from the University of Rostock (A2022-0187 and A2024-0018).

Standard preparation consisted of ensuring adequate histopathological diagnosis, determining the length of the aganglionic bowel segment using a colonic contrast enema, and assessing the patient's nutritional status. In preparation for pull-through surgery, we used the Rostock Irrigation Protocol described below. Rectal irrigation facilitated adequate decompression, intending to avoid an ostomy.

If a stoma was present, obtaining catheter tolerance and transferring fresh stool into the distal limp prepared the distal bowel and perineum for pull-through.

We advised parents to use a soft silicone Foley catheter to accustom the child to rectal tube insertion. Our preoperative bowel management protocol is shown in Table 1.

**Table 1.** Bowel management protocol at the Colorectal Center Rostock—preparation for pull-through surgery in Hirschsprung disease.

| | No Stoma | Stoma Present |
|---|---|---|
| **Perianal Skin Protection** | Regularly rectal irrigation; bring the perineum in contact with stool. | Transfer fresh stoma effluent to distal limp (at least for a minimum of 4 weeks prior to pull-through). |
| **Perianal Hygiene** | Shower after defecation; use water to rinse, e.g., HappyPo ©.<br>No wiping. | |
| **Dietary Advice** | Ideally keep on breastmilk till pull-through. | Ideally keep on breastmilk till pull-through.<br>If older, follow a low-fiber diet. |
| **Acquisition of Tolerance to Catheter** | Acquired with ongoing rectal irrigation. | Insert rectal tube intermittently. |
| **Microbiome** | Probiotics only if not breastfed. | (a) Probiotics if not breastfeeding;<br>(b) Transfer fresh stoma effluent to distal limp. |
| **Antibiotics** | Not routinely. | Not routinely. |

### *2.1. Immediate Preoperative Bowel Management*

We aim for an effective preoperative washout before the pull-through surgeries. Subsequently, the washout the day before surgery was conducted by our medical staff to secure adequate decompression.

### *2.2. Pull-Through Surgery Operative Approach—Swenson Type*

The operative technique used for all children is a laparoscopic-controlled transanal pull-through, leaving 1–2 cm of muscle.

### *2.3. Postoperative Bowel Management*

Enhanced recovery after surgery (ERAS) is applied to all patients. This includes rapid feeding, good pain relief, and early mobilization. Routine transanal irrigation is used in the event of abdominal distension and is continued for the rehabilitation period.

We actively avoid routine anal dilatations. Three months after the pull-through surgery, we perform one digital examination to palpate the anastomosis in the outpatient clinic. The institutional protocol is shown in Table 2.

The definition used in this study for **Hirschsprung-associated enterocolitis (HAEC)** is the Frykman modification of the Pastor score, with a cut-off value of 4 [1,4]. The HAEC score contains information on patient history, physical and radiological examination, and laboratory results, with a maximum value of 20 [1,4].

### *2.4. Perianal Skin Condition*

We used the definition visualized in Figure 1 to provide an objective way of assessing the perianal condition using a standardized grading system. Standardization facilitates communication between nurses and doctors and between families and the medical team.

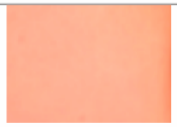
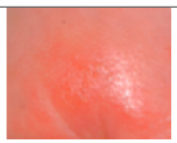
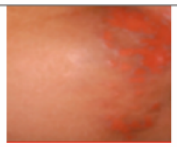
| | | Description |
|---|---|---|
| **A normal** |  | **Skin intact**<br>**No erythema** |
| **B mild** |  | **Skin intact**<br>**Slight Erythema** |
| **C moderate** |  | **Some small skin breakdown**<br>**Erythema** |
| **D severe** |  | **Skin Breakdown of larger areas**<br>**Erythema**<br>**Moist lesion** |

**Figure 1.** Perineal skin condition. Adapted from the Nappy Rash Protocol, Great Ormond Street Hospital, London.

### *2.5. Rostock Irrigation Protocol Used for Washouts*

*Material*: We use a very soft silicone Foley catheter (12–16 French depending on age and size), a 60 cc syringe and two bowls. Vaseline is used to lubricate the catheter.

*Washout fluid: hand-warm* tap water is safe for all ages in a country like Germany, with well-controlled tap water also suggested by clinicians in Europe and the USA [6].

*Volume*: We do not use calculations to assess the correct amount of water and there is no upper limit as described by de la Torre [7]. The endpoint of successful irrigation is reached when the water flowing back is transparent and the child's abdomen is very flat, almost below the level of the thorax.

*Frequency*: usually 1–2 irrigations per day will allow for sufficient decompression and an additional irrigation should be performed if necessary.

*Technique*: The Foley catheter is initially inserted 5–10 cm to allow for the decompression of gas and stool. To facilitate catheter advancement, it is sometimes helpful to introduce water during insertion. The syringe is disconnected, and stool and gas will passively flow back. A small amount of water (20 mL aliquots) is injected and the catheter is then disconnected again to allow the stool and gas to flow back. Sometimes, stool can be aspirated. Next, the catheter should be moved forward, i.e., more proximally. This helps

to evacuate trapped air and feces. Subsequent bowel massage helps to ensure effective decompression and reassures the child. This is repeated until the backflow is clear and the waste pot is filled with the amount of water installed.

*Position*: Newborn babies usually tolerate rectal irrigation in the supine position. Older children may prefer to sit, in which case we insert the catheter, close the nappy, and irrigate in a sitting position.

### *2.6. Bowel Management Protocol after Pull-Through for Hirschsprung Disease*

Table 2 shows the bowel management protocol after successful surgery.

**Table 2.** Bowel management protocol at Colorectal Center Rostock—after pull-through for Hirschsprung disease.

| | All | Too Much Stool | Perianal Skin Excoriation | Enterocolitis | Constipation |
|---|---|---|---|---|---|
| Perianal Skin Protection | | -Barrier cream<br>-Increase TAI | -Ilex® barrier cream<br>-Increase TAI | | |
| Perianal Hygiene | Showering after defecation, using water to rinse, e.g., HappyPo ©, and no wiping | | | | |
| Dietary Advice | Low-fiber diet | Stool thickener, early carrot, and psyllium husks | Early carrot | | Low-fiber diet |
| Transanal Irrigation (TAI) | If no spontaneous defecation, ideally advised in postoperative period | Increase irrigation to 2–3 times daily | Increase irrigation to 2–3 times daily | Increase irrigation to 2–3 times daily | If no spontaneous defecation, at least once daily |
| Microbiome | Probiotics | | | | |
| Antibiotics | Only in case of enterocolitis | | | Metronidazole oral | |
| Laxatives | | | | | Macrogol-based laxatives |

### *2.7. Data Management and Statistics*

The data were retrieved from electronic medical records, clinical reviews and discharge letters and entered into an Excel sheet. After data cleaning, the data were transferred to SPSS, and an analysis was performed with SPSS 26.0. We analyzed the data for the total group and the two subgroups (A no stoma; B stoma). The results were expressed as the mean and standard deviation or the median and range. The analysis of the groups was performed using t-test or chi-square depending on the variables. A linear-by-linear association model was applied for nominal contingency chi-square tests. The Fischer exact test was used for categorical variables. A $p$-value of <0.05 was considered significant.

## 3. Results

A total of 132 children with HD were seen in our colorectal clinic during the study period. In total, 47 children with any form of HD underwent pull-through surgery during the 2-year study period. We excluded 15 children with total colonic HD who underwent pull-through surgery during the study period. We excluded one child with redo pull-through surgery in standard HD. Subsequently, only children with standard HD and primary pull-through ($n$ = 31) were included in our institutional pathway analysis. We describe the pre-, intra- and postoperative data.

Our group consisted of 26 boys and 5 girls. The median age at pull-through surgery was 9 months. In total, 19 (59.3%) had already had a sufficient external rectal biopsy at the time of presentation to our center. A total of 12 (38.7%) children presented with an insufficient biopsy or no biopsy yet, so we performed a rectal biopsy. All children had a

diagnostic rectal biopsy confirming aganglionosis. All children had a contrast enema to determine the length of the aganglionic segment.

Table 3 shows the characteristics of the children with HD for the whole cohort and distinguished between children with no stoma and with an existing stoma. There was no significant difference in the affected length of aganglionosis. However, we note a significant difference in the anatomical level of HD. In total, 23/31 (75%) had no stoma at the time of consultation and all were using rectal washouts to decompress the bowel. A total of 8/31 (25%) children had a stoma, of whom 4 (50%) had a colostomy and 4 (50%) had an ileostomy.

**Table 3.** Characteristics at presentation.

| | **All *n* = 31** | **Group A No Stoma *n* = 23** | **Group B Prior Stoma *n* = 8** | ***p*-Value** |
|---|---|---|---|---|
| Sex (*n*) | Male: 26<br>Female: 5 | 20<br>4 | 7<br>1 | ns |
| Additional anomalies (*n*) | No: 26<br>Yes: 5<br>Trisomie 21: *n* = 3<br>If other, which:<br>Cardiac: 2<br>Duodenal Atresia: 1 | 21<br>2<br>1<br><br>2 | 5<br>3<br>2<br><br>1 | ns<br><br>ns |
| Age at contrast (months) | Mean: 8.3<br>Median: 6.5<br>Range: 1–26 | 8.8<br>6.5<br>1–26 | 6.8<br>6<br>1–13 | ns |
| Transition zone on contrast (*n*)<br>Type 1<br>Type 2<br>Type 3<br>Type 4 | <br>3<br>22<br>5<br>0 | <br>3<br>18<br>2<br>0 | <br>0<br>4<br>3<br>0 | <br><br>$p = 0.039$ |
| Aganglionic lengths on contrast (cm) | Mean: 8.7<br>Range: 5–15 | 8.1 ± 2.5<br>5–12 | 12.3 ± 2.5<br>10–15 | $p = 0.07$ |
| Type of stoma | | | Ileostomy: 4/8<br>Colostomy: 4/8 | |
| Age at rectal biopsy (months) | Mean: 8.2<br>Range: 1–26 | 8.8<br>1–26 | 2.5<br>1–4 | ns |

Transition zone on contrast: Type 1 = rectum, Type 2 = rectosigmoid, Type 3 = colon descendens to left flexure, and Type 4 = up to colon ascendens. ns—non significant.

Table 4 shows the perioperative patient data for all children undergoing pull-through during the study period. We noted 6.5% involved complications, with one unplanned stoma in each of the groups. One patient with no prior stoma developed a fever after discharge and was taken to another hospital where a stoma was performed due to a potential wound-healing problem at the rectal anastomosis. Another patient with a prior stoma and pull-through with reversal of the colostomy deteriorated 5 days after surgery, with signs of systemic infection. On explorative laparotomy, the colostomy reversal was unremarkable and there was a potential wound-healing disturbance at the rectal anastomosis dorsally.

Early postoperative bowel management was followed by a structured postoperative follow-up and the monitoring of bowel function, perineal skin condition, overall growth development and episodes of enterocolitis. We saw our children 7–10 days, 6 weeks and 3 months after discharge and then every 6 months thereafter, usually extending to an annual follow-up once toilet training was achieved. Follow-up was carried out both in person and by means of telemedicine. This is especially useful if the child is doing well and the family lives far away. In addition, we provided a low-threshold, easy way for the families to communicate in case they have any questions.

**Table 4.** Perioperative bowel management.

| | **All *n* = 31** | **Group A No Prior Stoma *n* = 23** | **Group B Prior Stoma *n* = 8** | ***p*-Value** |
|---|---|---|---|---|
| Age at pull-through (months) (*n* = 36) | Median: 9.5<br>Mean: 15.4<br>Range: 4–97 | 9<br>15.9 ± 19.8<br>4–97 | 9<br>11.8 ± 7.8<br>5–27 | ns<br>ns<br>ns |
| Weight at pull-through (kg and percentile) (*n* = 36) | Mean: 9.4, P33<br>Range: 7–12, P0.5–94 | 9.6, P32<br>7–12 | 8.9, P31<br>7.2–11.6 | ns |
| Operative time of pull-through (h) primary pull-through | Mean: 2:41<br>Range: 1:26–4:27 | 2:24<br>1:26–4:27 | 2:50<br>2:37–3:57 | ns |
| Primary pull-through (*n*) | 31 | 23 | 8 | |
| Rectal irrigation prior to pull-through (*n*) | 23 | 23 | 0 | |
| Stool transfer to distal limp (*n*) | 8 | Not applicable | 8 | |
| Training catheter tolerance (*n*) | 8 | Not applicable | 8 | |
| Length of resected segment during surgery (cm) | Mean: 13.8<br>Range: 5–28 | 12.8<br>5–25 | 15.8<br>7–28 | ns |
| Protective stoma after pull-through (*n*) | 0 | 0 | 0 | |
| Unplanned stoma within 30 days (*n*) | 2 | 1 | 1 | ns |
| Clear fluids on operative day (*n*) | 31 | 23 | 8 | |
| Slow establishment of enteral intake; no full enteral feeds after 48 h (*n*) | 5 | 1 | 4 | $p < 0.001$ |
| Time to first spontaneous defecation (*n*) | 24 within 24 h<br>7 within 24–48 h | 22<br>1 | 3<br>5 | |
| Time to rectal irrigation (*n*) | Performed on day 1 post-op | | 5 | |
| 30-day complication occurrence (*n*) | 2 | 1 | 1 | ns |
| Rectal dilatation (*n*) | 0 | 0 | 0 | |

ns—non significant.

The follow-up is visualized in Figure 2. We did not have a significantly ($p = 0.04$) higher rate of nappy rash 3 months after pull-through in children who had a stoma before pull-through.

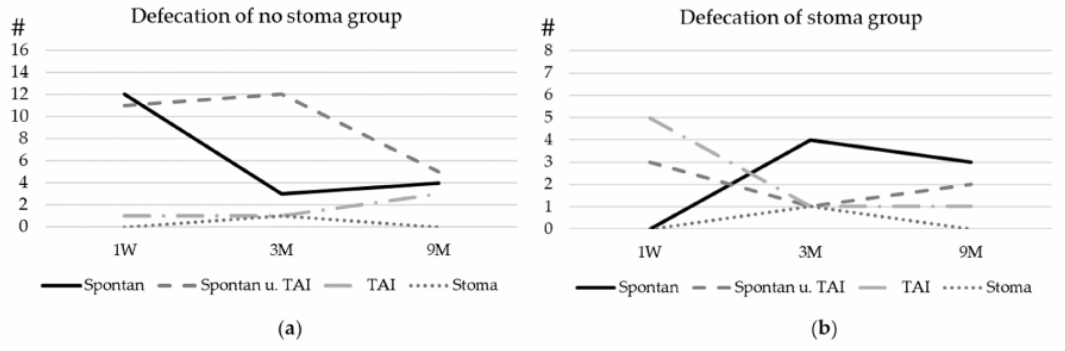



**Figure 2.** *Cont.*

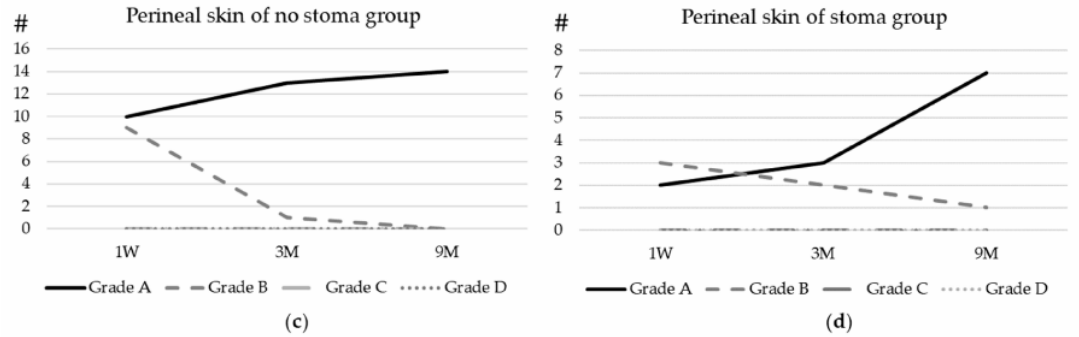



**Figure 2.** Postoperative bowel function and bowel management: the development of defecation of patients with (**a**) no stoma before surgery and (**b**) a stoma before surgery. (**c**) The development of the perineal skin condition in patients with no prior stoma and (**d**) the development of the perineal skin condition in patients with a prior stoma. For the graduation of the skin, please refer to Figure 1. Spontan = spontaneous defecation, TAI = transanal irrigation, and Stoma = defecation through stoma. Y-axe # Number of patients.

## 4. Discussion

Preoperative preparation paves the way for intraoperative surgical reconstruction in primary pull-through surgery for HD. The ability to achieve effective decompression through the use of washouts can help to avoid the need for a stoma. The pediatric surgical community has moved to avoid stomas and aims to create washouts whenever possible in preparation for pull-through surgery [8]. Avoiding a stoma facilitates a technically easier pull-through for the surgeon, with a shorter operative time and faster recovery. Furthermore, stoma-related problems described in up to 30% of children with a stoma with a background of Hirschsprung [9] are avoided. More importantly, recovery in terms of regaining enteral intake and bowel function is significantly faster, as demonstrated by comparing the two groups with and without a stoma in our cohort treated in the same colorectal unit by the same team.

An intact dentate line, anal canal and anal sphincter are essential for continence. Structured postoperative bowel management supports postoperative recovery and the attainment of social continence [10].

### *4.1. Child without a Stoma—Preoperative Bowel Management to Prepare for Surgery: PREHABILITATION*

Ideally, daily washouts by the family will allow for adequate decompression and prevent enterocolitis. In addition, effective washouts will relieve the obstruction and keep the colon decompressed. An adequate washout helps reduce an initially dilated prestenotic bowel segment and facilitates smooth anastomosis [7].

We emphasize the importance of educating the family to perform an adequate and effective washout. Not only should we aim to avoid an unnecessary procedure, but we also aim to see the smoother recovery of children who do not have a stoma.

### *4.2. Child with a Stoma: Preoperative Situation*

Stool is evacuated through the stoma until pull-through surgery, leaving the aganglionic and adjacent bowel uninvolved in the circulation, i.e., subsequently unused mucosa.

Children with a preoperative stoma are more likely to develop a perianal rash/skin excoriation despite thorough preparation with stool transfer to the distal limb. In total, 40% of children with a stoma report having a perianal rash 3 months after pull-through surgery, compared with only 7% of children who did not have a stoma before pull-through surgery.

Perianal excoriation can be managed through adequate perineal care, bowel management and the use of various barrier creams [1].

Even with this thorough preparation, children with a stoma pass stool later than their counterparts without a stoma and are significantly slower to achieve full enteral nutrition ($p < 0.001$).

### *4.3. Perioperative Bowel Management*

Rapid recovery using an ERAS protocol has significantly changed the perioperative and early postoperative management of children undergoing abdominal surgery [11,12]. In particular, the transanal approach avoids intra-abdominal dissection and early feeding is recommended [13]. In our cohort, all children had their first enteral feed on the day of surgery regardless of a previous stoma; however, we noted a significantly faster full enteral intake in children without a previous stoma ($p < 0.001$).

Bearing in mind that pain and stress can affect bowel motility, which is also disturbed by the technical act of surgery, both can lead to temporary postoperative bowel paralysis.

In total, 78% (N = 25) of children in our cohort passed stool within the first 24 h and the remaining within the second day.

Immediately after surgery, some children have difficulty passing air, and rectal irrigation can relieve symptoms of bloating and abdominal distension.

Similarly, Zhang et al. show that rectal irrigation not only reduces abdominal distension but also prevents postoperative HAEC [14]; we did not see any HAEC in our direct postoperative observation. Controversially, Beltmann et al. describe a likely mortality due to perforation when early postoperative irrigation is used; in a cohort of 106 pull-through operations within a 15-year period in which 22% had postoperative complications anyway [15].

Unpublished data from our department show that children tolerate rectal irrigation well if introduced smoothly in a child-friendly, relaxed environment before surgery. In order to support the adaptation and recovery of the pull-through, we use early transanal irrigation as physiotherapy for the bowel.

### *4.4. Postoperative Bowel Management: REHABILITATION*

(a) Developing spontaneous bowel movement

We see an overall good potential for spontaneous bowel movement in the very early recovery period, with 52% of all children passing stool spontaneously, but after 3 months, 90% need at least supportive rectal irrigation to pass stool effectively and can be weaned off irrigation over time. By 9 months, 44% are passing stool spontaneously and this figure is now increasing steadily. At 2-year follow-up, the majority defecates spontaneously.

This highlights the importance of incorporating postoperative transanal irrigation into routine bowel physiotherapy. Systematic irrigation not only reduces the risk of enterocolitis but also helps children to eventually become socially clean.

In older children, we progress to active transanal irrigation as needed. We use hydrosonographic ultrasound to estimate the amount of water needed. We monitor the success of the bowel management program by assessing fecal load through ultrasound [16]; both sonographic techniques have the advantage of sparing ionizing radiation, which is often described in other bowel management programs [17].

(b) Overcoming perianal rash

It is not uncommon to have frequent bowel movements with small portions during the early recovery period. These low-volume bowel movements are partly caused by the removal of the obstructive pressure and partly by the perioperative antibiotics, which affect the consistency of the stool. Too many stools, especially small ones, cause perianal rash. In children with a stoma, 3 months after pull-through, we note significantly ($p = 0.04$) more cases of nappy rash. In these cases, bowel management with transanal irrigation can reduce the frequency of defecation and thus improve the nappy rash.

(c) Overcoming outlet obstruction

Rehabilitation is a well-accepted concept in musculoskeletal surgery or surgery for other functional impairments. The pull-through bowel not only needs to find its new position but also needs to learn to function as a neorectum. The healthy ganglionic bowel representing the prestenotic dilatation will become the neorectum once anastomosed to the remaining small native rectum, overcoming a size mismatch. The pull-through bowel will take over its function and will regain an adequate peristalsis, relieved from the functional obstruction caused by the aganglionic segment.

Outlet obstruction characterized by the persistent absence of the rectoanal inhibitory reflex (RAIR) can be a functional symptom after an otherwise successful pull-through. However, mechanical complications such as twist, stricture or persistent aganglionosis should be ruled out and, if present, appropriately treated [17–20].

We observe, as do others, that maturing children learn to overcome the obstruction of a non-relaxing internal sphincter and grow out of the outlet obstruction that is presented as smearing and sometimes mistaken for fecal incontinence [13,21,22]. However, Fosby et al. reports no improvement with age in a Norwegian cohort [23].

Other groups have successfully used Botox to relieve this well-known temporary outlet obstruction, which can be repeated if necessary [24]. However, in our experience, using transanal irrigation as active physiotherapy trains the distal bowel.

### *4.5. Education and Support for Families*

Families can be overwhelmed by bowel management, especially when faced with a new diagnosis of Hirschsprung [2,3]. Educating parents and involving them as early as possible in the management of rectal irrigation alongside the medical team builds parental confidence [1,10]. Families benefit from being able to raise their questions and concerns at any time with a low threshold. E-mail, telephone and video calls can be used as a supplement, especially for families living further away [2].

Looking at the experience of affected families, a Dutch nationwide study reveals that if the family is familiar with Hirschsprung, not only does the likelihood of ongoing bowel management increase but also the families are more likely to use transanal irrigation as their preferred method of bowel management in both rectosigmoid and long-segment Hirschsprung. This familiar coping positively influences the patients coping [25].

### *4.6. Strength and Limitations*

We describe our standardized bowel management protocol by analyzing a cohort treated by the same team within a short time period. This means that there are no confounders by the overall medical advancements. Monitoring data in a clinical environment is challenging, even if the patients are followed prospectively, as sometimes not all information is sufficiently recorded.

## 5. Conclusions

The success of pull-through surgery is supported by preoperative bowel management and structured follow-up, with bowel management as physiotherapy.

Avoiding a stoma in the first place not only avoids surgery but also allows for a faster recovery after pull-through surgery. If a stoma is present, careful preparation will help to prepare the distal bowel, although postoperative recovery of bowel function will take longer than in patients without a stoma, despite the best preparation.

Rectal irrigation is helpful in the postoperative period to facilitate a smooth recovery and to support postoperative recovery, serving as physiotherapy for the bowel.

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J.L. and F.S.; supervision, S.M.; project administration, J.L.; funding acquisition, J.L. All authors have read and agreed to the published version of the manuscript.

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**Institutional Review Board Statement:** This study was conducted in accordance with the Declaration of Helsinki and approved by the Ethics Committee of University Rostock (A 2023-0123 on 7 July 2023).

**Informed Consent Statement:** Patient consent was waived as routine clinical data and usual clinical practice were monitored as part of an audit.

**Data Availability Statement:** The data presented in this study are available on request from the corresponding author. The data is recorded in our Rostock Colorectal Hirschsprung patient registry due to privacy or ethical restrictions.

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## Abbreviations

| | |
|---|---|
| ERAS | Enhanced recovery after surgery |
| HAEC | Hirschsprung-associated enterocolitis |
| HD | Hirschsprung disease |
| ns | Not significant |
| RAIR | Rectoanal inhibitory reflex |
| TAI | Transanal irrigation |

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## 2.2 Assessing fecal load with ultrasound in children with colorectal pathology: ReKiSo study.

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ORIGINAL ARTICLE

# Assessing fecal load with ultrasound in children with colorectal pathology: ReKiSo study

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**Abstract**

**Purpose** To evaluate bowel management for children with colorectal pathology by measuring transverse rectal diameter (TRD) and assessing fecal load with transabdominal rectal ultrasound (TRU).

**Methods** Prospective case–control study of children receiving bowel management (BM) between 04/2023 and 04/2024 was done. There was inclusion of patients with Hirschsprung disease (HD), anorectal malformation (ARM) and functional constipation (FC). Patients with other congenital or neurological conditions were excluded. Control group consisted of inpatients and outpatients without abdominal complaints. FC was diagnosed according to ROM-IV-criteria. For HD and ARM, we followed a list of symptoms. To assess fecal load, we visualized the TRD using the Klijn (Klijn et al. in J Urol 172:1986–1988, 2004) method. The bladder was moderately full. The fecal load was assessed retrograde from the rectum. Follow-up was at 1/3/6 months. Secondary data were collected from medical records. Sample size calculated a priori and follow-up group with new gathered data.

**Results** $p$ value for TRD in all groups significant with $p < 0.05$ and in grouped follow-up.

**Conclusion** Ultrasound is a useful tool for assessing fecal load and helps diagnose constipation and monitor BM. Irrespective of colorectal pathology, a cut-off of 3 cm seems to discriminate between children without constipation/overload symptoms and asymptomatic patients. We present a radiation-free method for monitoring bowel management.

**Keywords** Point-of-care ultrasound · Constipation · Transabdominal ultrasound · Transrectal diameter · Bowel management · Pediatric colorectal disease · Fecal load

## Introduction

Anorectal malformation (ARM) and Hirschsprung disease (HD) are rare congenital colorectal disorders. Despite surgical treatment in the form of posterior/anterior sagittal anorectoplasty (PSARP/ASARP) or pull-through surgery, many of these patients suffer from constipation, stool soiling and problems of defecation [1]. The follow-up concentrates on bowel management (BM) to achieve social cleanness and continence [2, 3]. During BM, the fecal load of the colon is monitored by daily abdominal x-rays for a week [4, 5]. The long-term effects of repetitive exposure in children to radiation are described by Linet [6], and Wall [7] estimated the highest risk for any kind of cancer in the age group < 10 years using the Monte Carlo method, which is the primary target group for this kind of intervention and follow-up. Overall, the smaller height, slight reduction and increased scattered radiation result in a higher absorbed doses [6].

In addition, functional constipation (FC) is the main cause of constipation and a common reason for admission to the emergency department [8]. For diagnosis or evaluation of fecal load abdominal, X-rays are used in 70–77.5% of the cases, although the utility is considered low [9], lacks medical evidence [10] and ESPGHAN/NASPGHAN has not included X-ray in their recommendation [11]. Furthermore, the benefit is questionable due to low sensitivity, specificity, diagnostic accuracy and simply subjective assessment. There is no standardized evaluation and scores by Barr, Leech or stool loading show low interobserver reproducibility [12, 13]. In particular, stool loading correlates poorly with the symptoms of constipation and even an unremarkable X-ray

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does not represent a normal finding or the exclusion of a serious disease [14, 15]. In fact, the most common missed diagnoses include acute appendicitis and intussusception [12, 16]. Otherwise, a readmission leads to overdiagnosis [16]. This should be put into consideration because patients with constipation show more readmissions and patients with ARM or HD need a prolonged BM [12].

Recent research presents abdominal ultrasound examination as an alternative diagnostic tool for FC and evaluation of treatment. In comparison to abdominal radiographs, this is an accurate modality and would avoid X-rays despite established scores [17] or charting [18]. Ultrasound is non-invasive and has no adverse effect. This simple procedure should make a digital rectal examination (DRU) obsolete [19, 20]. Furthermore, not inconsiderable costs for unjustified X-ray requests can also be saved in these cases [17] and the principle of ALARA (as low as reasonably achievable) satisfied. Unfortunately, there are no data regarding ARM or HD and those patients are consequently excluded in the current research. The aim of the ReKiSo Study (German: Rektum Kinder Sonographie) is to provide new data for these patient groups using established sonographic methods.

## Methods and patients

### Literature research and data extraction

Before setting up the study design, we performed literature research on scientific articles exploring the utility of ultrasound in children with FC. We used several keywords (i.e., transabdominal ultrasound, transrectal diameter, constipation) on pubmed.gov and found 15 articles to put into consideration. Klijn [21] described first a new method of abdominal ultrasound as a diagnostic tool for constipation in children with dysfunctional voiding by measuring the transrectal diameter (TRD) in the transverse plane. The following scientific research adapts Klijn's method and faces new questions (i.e., position of probe, bladder filling, influence of treatment). In general, the inclusion of patients follows the ROM-criteria or an equivalent list of symptoms for chronic constipation. All studies excluded children with ARM or HD. Our key findings are summarized in Table 1 [17, 19, 21–33] and the complete table is attached to the appendix.

The researchers present data of TRD which was significantly higher ($p < 0.05$) in the case group compared to the control group for each study except in the age group < 1 year [33] and < 3 years [31]. Three studies [17, 22, 27] calculated a cut-off for constipation at 2.7–3.8 cm. The sensitivity varied between 71 and 100% and specificity between 71 and 97% [17, 25, 27, 32].

To evaluate TRD in case and control groups, we focused on seven studies with the specification of a mean value [17, 19, 21, 23–25, 29]. To assume the reduction of TRD in the follow-up, we orientated on five studies [17, 19, 25, 29, 30]. For estimation of the TRD in the case (ARM, HD and FC) and control groups, the median (3.16 cm) between mean TRDcase = 4.0674 cm and TRDcontrol = 2.259 cm of the reviewed studies was calculated. This median exceeded the 95% confidence interval of the mean difference = 1.82 cm (1.07–2.56). Thus, there is no overlap of the case and control groups. We concluded a minimum mean TRDcase of > 3.16 cm contrary to < 3.16 cm in the control groups as well for the case groups in the follow-up. The mean reduction of TRD in the follow-up was 1.26 cm and exceeded the calculated limiting value of 3.16 cm. Following the findings of Gatzinsky [33], TRD should not be useful for children < 1 year old.

### Study design

ReKiSo is a prospective case–control study at the Clinic for Paediatric Surgery of the Rostock University Medical School (UMR) enrolling 302 children over a period of one year (04/2023—04/2024). We included 155 patients with ARM, HD or FC. Patients with other congenital anomaly affecting bowel function (ileal atresia, gastroschisis, omphalocele, cloacal exstrophy) or neurological condition (cerebral palsy, spina bifida, tethered cord) were excluded in the present analysis, but they were also monitored and will be reported separately. The control group consisted of children without a gastrointestinal pathology and without clinical signs of constipation, which were either hospitalized children or consulting the outpatients department for other reasons (trauma, urological or other pathology). FC was diagnosed by ROM-IV criteria [34] (see Table 2). For children with HD and ARM, we followed a list of symptoms as shown in Table 2. Secondary data (age, weight, height, comorbidity, surgery, transanal irrigation, clinical classification) were collected from medical records. The consultation in the special pediatric outpatient clinic for colorectal diseases included a careful anamnesis, clinical examination and finally the abdominal ultrasound scan. To our knowledge, we were the first to present data about TRD of children with ARM or HD. Therefore, the process for in- and exclusion is displayed in the following Figs. 1 and 2.

### Ultrasound

We used the method of Klijn [21] by placing a curved array of 3.5 MHz (Toshiba Aplio 300, Toshiba Medical Systems GmbH, Germany, Neuss) above the symphysis and measured the largest TRD at a downward angle of at least 15 degrees from the transverse plane after distinguishing sigma from rectum. In constipated children, the rectum conducted as an adynamic structure

**Table 1** Key findings literature research

| Author, year | Number of study participants | TRD in cm | *p* value |
|---|---|---|---|
| Klijn et al. [21] | $N=49$ | $mean_{case}$: 4.9 (SD 1.101)<br>$mean_{control}$: 2.1 (SD 0.64) | $p<0.001$ |
| Singh et al. [22] | $N=177$ | $median_{case}$: 3.4 (2.1–7.0 with IQR 1.0)<br>$median_{control}$: 2.4 (1.3–4.2 with IQR 0.72) | $p<0.001$ |
| Bijoś et al. [23] | $N=225$ | Results for all subgroups by age:<br>$mean_{case}$: $4.3060 \pm 0.968$<br>$mean_{control}$: $3.183 \pm 0.824$ | $p<0.001$ |
| Joenssons et al. [19] | $N=51$ | Results pre-treatment:<br>$mean_{case}$: $3.96 \pm 0.82$<br>$mean_{control}$: $2.14 \pm 0.6$ | $p<0.001$ |
| Di Pace et al. [24] | $N=270$ | $mean_{case}$: $3.9958 \pm 0.6906$<br>$mean_{control}$: $1.0 \pm 0.8319$ | $p<0.0005$ |
| Karaman et al. [25] | $N=66$ | Results pre-treatment:<br>$mean_{case}$: $3.42 \pm 1.04$ (full bladder)<br>$mean_{control}$: $2.12 \pm 0.65$ (full bladder) | $p<0.001$ |
| Modin et al. [26] | $N=28$ | $mean_{case}$: 3.5<br>$mean_{control}$: 1.9 (SD 0.3) | –/– |
| Hatori et al. [27] | $N=100$ | $median_{case}$: 3.53<br>$median_{control}$: 2.0 | $p<0.0001$ |
| Doninger et al. [17] | $N=50$ | $mean_{case}$: $4.3 \pm 1.35$ (IQR = 1.52)<br>$mean_{control}$: $2.85 \pm 1.16$ (IQR = 1.63) | Statistically significant |
| Momeni et al. [28] | $N=76$ | $mean_{case}$: $3.172 \pm 0.963$<br>$mean_{control}$: $1.985 \pm 0.437$ | $p<0.001$ |
| Pop et al. [29] | N = 65 | Results for all subgroups by age<br>$mean_{case}$: $3.59 \pm 1.41$<br>$mean_{control}$: $2.42 \pm 0.71$ | $p<0.05$ |
| Imanzadeh et al. [30] | $N=154$ | Results pre-treatment:<br>$mean_{case}$: $3.879 \pm 1.017$ | $p<0.001$ |
| Doğan et al. [31] | $N=304$ | $mean_{case}$: subgroups by age<br>$mean_{control}$: subgroups by age | $p=0.04$ (group 3.1–6 years),<br>$p=0.003$ (group 6.1–12 years) |
| Hamdy et al. [32] | $N=100$ | $median_{case}$: 3.55 (3.2–4)<br>$median_{control}$: 2.3 (1.8–2.5) | –/– |
| Gatzinsky et al. [33] | $N=110$ | $mean_{case}$: subgroups by age<br>$mean_{control}$: subgroups by age | Not statistically significant |

**Table 2** ROM-IV-criteria and list of symptoms

| ROM-IV-criteria for FC | List of symptoms for ARM and HD |
|---|---|
| - Min. 2 criteria for children > 4 years, duration > 1 month: ≤ 2 defecation/week, ≥ 1 period of incontinence/week, excessive stool retention, painful or hard bowel movements, large fecal mass in the rectum, large diameter stools that can obstruct the toilet<br>- Both criteria for children < 4 years: ≥ 1 period of incontinence /week and large diameter stools that can obstruct the toilet only after development of cleanliness | - Constipation<br>- Fecal incontinence<br>- Painful defecation<br>- Extensive defecation<br>- Fecalith/filled bowel loop palpable<br>- Stool in DRU<br>- Enlarged/protruding abdomen<br>- Meteorism |

and physiologically the ampulla recti as well the neorectum was empty. Thus, it was not necessary to determine TRD several times. The bladder was moderately full and acted as an acoustic window. We assessed fecal load retrograde starting at the rectum. Figs. 3 and 4 show typical measurements of TRD for asymptomatic and constipated patients. The follow-up was performed after 1/3/6 months, but readmission was possible at any time.

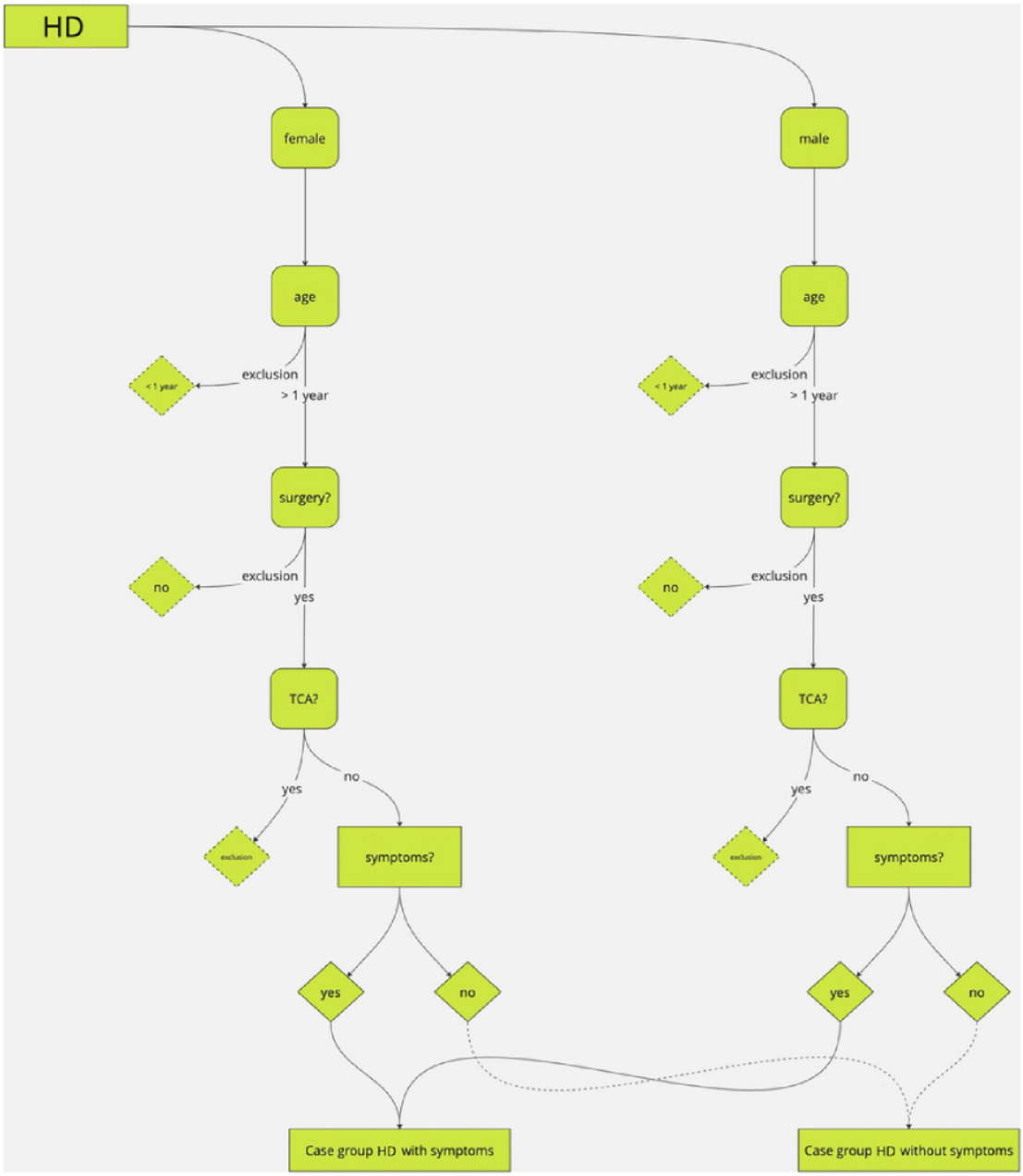


**Fig. 1** Process of in-/exclusion

Several patients were monitored with tele-medicine until reaching the clinical outcome (absence of symptoms) due to long distance. We treat children from all over Germany and neighboring countries in the patient cohort, but we lost final ultrasound measurements in their follow-up. The ultrasound scans were performed by one consultant and one medical student under her supervision.

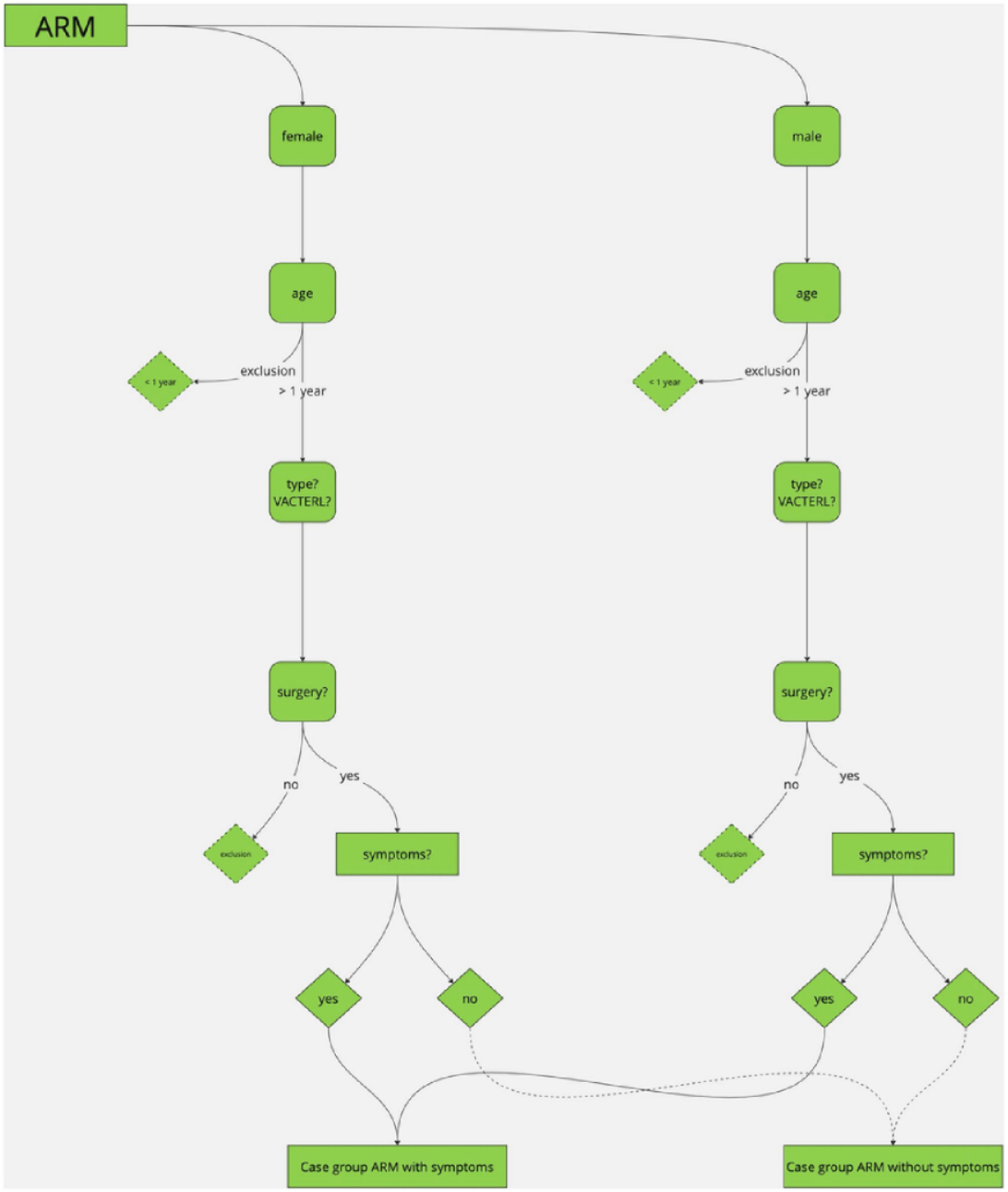


**Fig. 2** Process of in-/exclusion

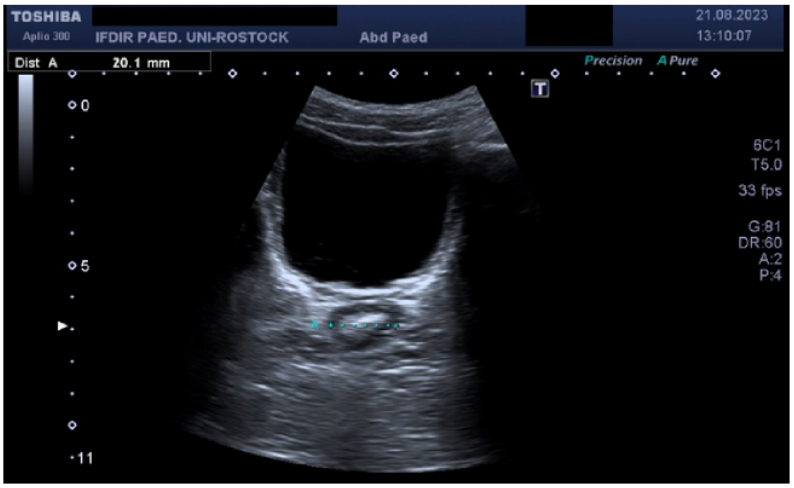


**Fig. 3** Asymptomatic patient with oval shaped and empty rectum

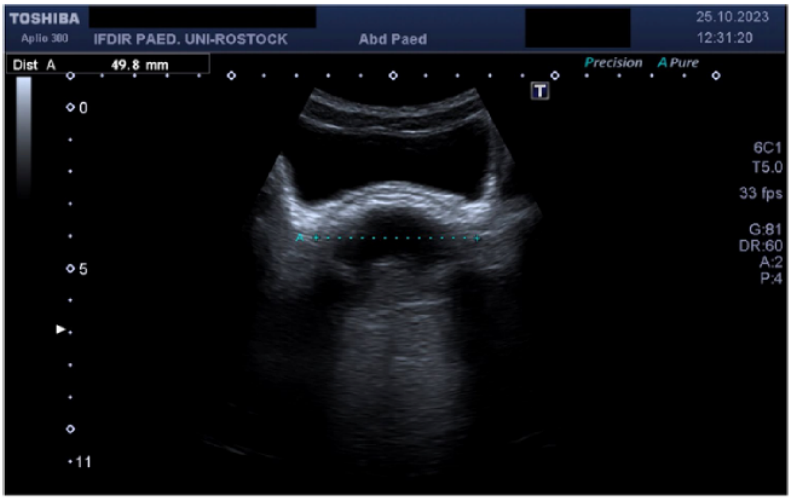


**Fig. 4** Constipated patient with hyperechoic crescent and posterior shadowing

## Statistics

Data management in our clinical patient registry on excel used pseudonymized acronyms. Statistical analyses were performed with SigmaPlot 13.0 (Inpixon GmbH, Germany, Düsseldorf) for descriptive statistics and verification of normal distribution by Shapiro–Wilk test. The *t*-test was used for normal distributed continuous values and otherwise the Mann–Whitney-*U*-Test. $p$ value $< 0.05$ was statistically significant. Cut-offs, sensitivity and specificity were calculated using receiver operating characteristic. Non-linear correlation was computed for correlation in secondary data. To determine the number of cases in each group, G*Power 3.1.9.6 (Faul, Erdfelder, Lang & Buchner, 2007) [35] was operated for power analysis a priori with setting $\alpha = 0.05$ and effect size $d = 0.80$. The sample size was a minimum of $n = 42$ for each case and control group. In the follow-up, a total sample size of $n = 8$ were calculated using the new gathered data. For this purpose, ARM and HD were assembled as a coherent group. For evaluation of the follow-up, the paired *t*-test was used.

## Ethical approval

This study was approved by the ethic committee for the Medical School of Rostock (A 2023-0066; 18.04.2023)

and conducted in conformity to the Declaration of Helsinki. Informed consent was obtained from parents or legal guardians.

## Results

For one year, we performed ultrasound scans on 302 children and Fig. 5 shows the composition of groups considering the process of in- or exclusion. Patient's characteristics are summarized in Table 3. In the first stage of the study, we investigated particularly the utility of ultrasound for children with colorectal pathology. We found significantly increased TRD for children with ARM or HD due to constipation, fecal soiling and associated symptoms. These findings conducted similar to published data of the literature research based on children with FC following ROME-IV-criteria.

The control group was used for comparison to all case groups and faced problems considering demographic or physical characteristics. Generally, sex in patients with HD showed a ratio of 1:4 while ARM was balanced. There was no significant difference considering sex only in HD and the control group ($p = 0.353$). On the other hand, age was not statistically different for ARM and FC compared to the control group ($p = 0.067$ and $p = 0.53$) fading the sex difference. Only in FC height and weight were not statistically significant ($p = 0.595$ and $p = 0.933$).

The fecal load was assessed retrograde from the rectum. The ampulla recti was empty in all patients of the control group. Although patients in the case groups reported any symptoms, there was stool present in other parts of the colon ranging from 8.82 to 18.75%. The fecal load increased respectively presenting symptoms according to Table 2 for these groups from 71.43 to 91.43%. The correlation between TRD and fecal load was high and statistically significant

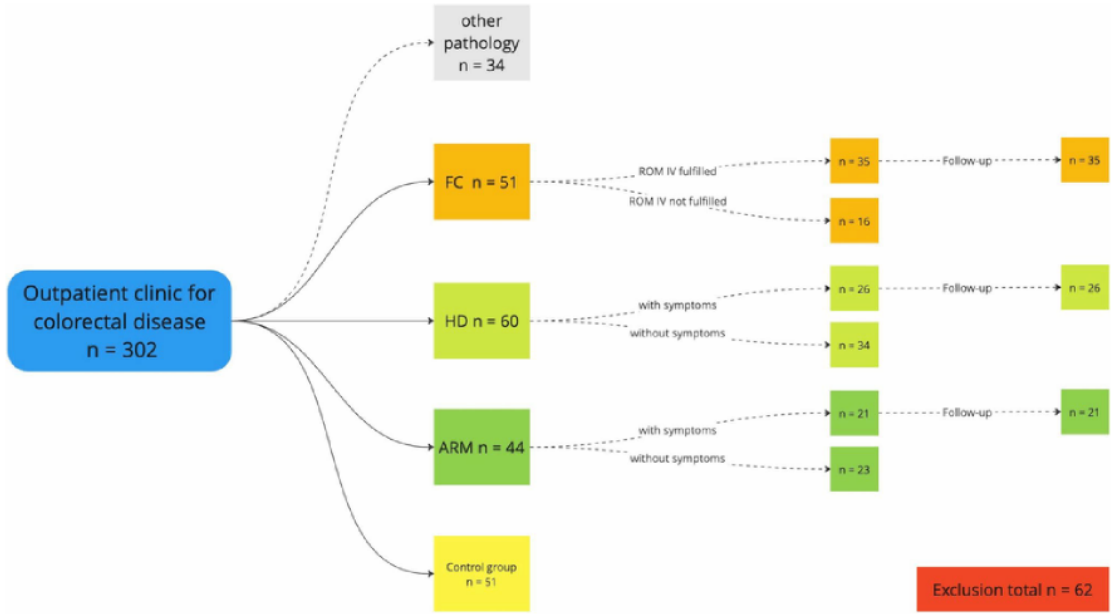


**Fig. 5** Overview of included patients

**Table 3** Patient's characteristics

| Group | Control group | HD | ARM | FC |
|---|---|---|---|---|
| Number | 51 | 60 | 44 | 51 |
| Sex (female:male) | 15:36 | 13:47 | 24:20 | 26:25 |
| Age (years, mean, SD) | 7 (3.72) | 5 (3.95) | 6 (3.28) | 7 (3.76) |
| Height (cm, mean, SD) | 123.23 (24.73) | 105.00 (26.44) | 107.23 (21.5) | 118.85 (26.43) |
| Weight (kg, mean, SD) | 27.84 (14.18) | 18.85 (11.47) | 18.27 (6.54) | 29.55 (19.10) |

**Table 4** Sensitivity, specificity and cut-off in case groups

| Group | MH | ARM | FC |
|---|---|---|---|
| Sensitivity | 97.60% | 95.65% | 100% |
| Specificity | 68.00% | 57.14% | 85.71% |
| Cuf-off (cm) | 2.975 | 3.095 | 2.96 |

($p < 0.0001$). The correlation between demographic and physical characteristics was overall not statistically significant. The receiver operating characteristic was used for calculating cut-offs, sensitivity and specificity in each case group. The computed values are shown in Table 4.

The second stage of the study focused on the follow-up with BM and 82 children with symptoms or FC were included. The treatment covers laxatives, enemas or transanal irrigation and patients were evaluated after 1/3/6 months. The clinical endpoint was the absence of any signs of constipation according to ROM-IV-criteria or our list of symptoms (22 patients), see Table 2. The following Table 5 summarizes both stages of the study and the measured values are shown in Fig. 6.

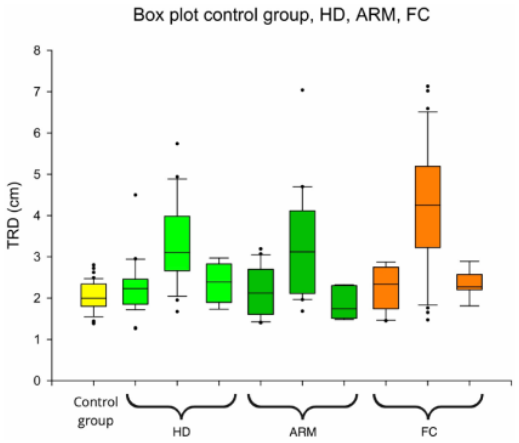


**Fig. 6** Box plot control group, case groups in the sequence of 'no symptoms, symptoms and follow-up' for HD, ARM and FC

## Exclusion

During the survey, 34 patients with constipation and fecal soiling or associated symptoms were admitted in the pediatric outpatient clinic for colorectal diseases and ultrasound scans were performed. These patients had other congenital anomaly affecting bowel function (esophageal and ileal atresia, gastroschisis, omphalocele) or neurological condition (cerebral palsy, spina bifida, tethered cord). They were excluded in the present analysis, but they were additionally monitored and will be reported separately. Additionally, 62 patients of the case groups and control groups were finally excluded. Detailed exclusion is commented on below.

MH: In general, children after surgical treatment were included (pull-through surgery), but those with TCA were excluded in this analysis due to the total resection of the colon and inconsistent data. Following ROM-IV-criteria, patient groups younger or older than 4 years were investigated and finally patients younger than 1 year were excluded due to insufficient data.

ARM: In general, children after surgical treatment were included (PSARP/ASARP), but those without surgical treatment or special forms of ARM (i.e., atypical form, cloacal exstrophy) were excluded due to limited sample size. Following ROM-IV-criteria, patient groups younger or older than 4 years were investigated and finally patients younger than 1 year were excluded due to insufficient data.

FC: The main inclusion criterion was ROM-IV and some patients received surgical interventions of the abdomen in the past (i.e., appendectomy, rectal biopsy). Following ROM-IV-criteria, patient groups younger or older

**Table 5** Summary

| Group | Number of participants | $Mean_{TRD}$ in cm | $p$ value |
|---|---|---|---|
| Control | $N=51$ | 2.049 (SD 0.368) | –/– |
| ARM | $N=44$ | - Without symptoms: 2.169 (SD 0.592)<br>- With symptoms: 3.308 (SD 1.304)<br>- Follow-up: 1.874 (SD 0.405) | - $p<0.001$*<br>- Follow-up $p=0.011$* |
| HD | $N=60$ | - Without symptoms: 2.314 (SD 0.573)<br>- With symptoms: 3.348 (SD 1.006)<br>- Follow-up: 2.490 (SD 0.514) | - $p<0.001$*<br>- Follow-up $p=0.0382$** |
| FC | $N=51$ | - ROM-IV not fulfilled: 2.310 (SD 0.505)<br>- ROM-IV fulfilled: 4.357 (SD 1.382)<br>- Follow-up: 2.476 (SD 0.362) | - $p<0.001$*<br>- Follow-up $p<0.001$* |

* = Mann–Whitney $U$ test, ** = $t$-test

than 4 years were investigated and finally patients younger than 1 year were excluded due to insufficient data.

## Discussion

Patients with ARM or HD often present symptoms of constipation, stool soiling and problems of defecation during long-term follow-up. Sufficient BM is essential in the follow-up to achieve social cleanliness. Monitoring BM for these patient groups widely focused on abdominal X-rays [4, 5]. While ultrasound imaging for patients with FC was developed for diagnosing and the evaluation of BM in current research, these patients with colorectal pathology were excluded consistently.

The first part of the study focused on the utility of TRD to discriminate asymptomatic patients from patients with constipation, fecal incontinent or associated symptoms in ARM and HD. The current analysis of the study presented statistically significant data of TRD according to present data of FC and exceeded their cut-offs. The calculated cut-offs were similar to those of FC. Fecal load increased and was found retrograde in the colon. The sensitivity of abdominal ultrasound for ARM and HD was equivalent to FC. Specificity was slightly lower compared to data of FC because there was no score used and the study followed a list of symptoms. Additionally, patients with ARM reported more stool soiling while patients with HD were more likely to be constipated. However, the collective sensitivity of 95.89% and specificity of 72.84 for ARM, HD and FC were equivalent to values in the current research. The calculated cut-off for ARM and HD was 2.975 cm, but we suggest a practical cut-off of 3 cm for patients older 1 year.

During follow-up, TRD for ARM and HD was statistically significant lower as endpoints reached and conducted comparable to patients with FC. The options of treatment were identical for all case groups and FC was also scanned with abdominal ultrasound to exclude environmental effects. Thus, ultrasound imaging is an easy, non-invasive and not harmful tool for monitoring BM in patients with ARM and HD.

### Limits

The control group was used for three case groups with statistical differences. Although TRD was not affected, two individual control groups for each ARM and HD may be helpful to investigate further correlation or influence of secondary data. Due to tele-medicine, ARM and HD were grouped to reach the calculated sample size in the follow-up, but TRD was already statistically significant lower.

## Appendix

For access of the complete table for the literature research (Table 1), please contact the corresponding author.

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**Author contributions** Study conception and design: JL, DE. Data acquisition: DE, JL, SM. Data analysis and interpretation: DE, JL. Drafting of the manuscript: DE, JL. Figures and tables: DE. Critical revision: JL, SM.

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**Data availability** For request of the research data, please contact the corresponding author.

## Declarations

**Conflict of interest** The authors declare there is no conflict of interest.



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## 2.3 Is the Transrectal Diameter (TRD) Suitable for Assessing Faecal Load and Monitoring Bowel Management in Children with Hirschsprung Disease - ReKiSo Study: Prospective Study.

Lindert J, Erkel D, Schulze F, Hofer M, Rzepka E, Märzheuser S. Children. 2024. 11(8):921.

*children*

MDPI

*Article*

# Is the Transrectal Diameter (TRD) Suitable for Assessing Faecal Loads and Monitoring Bowel Management in Children with Hirschsprung Disease—ReKiSo Study: Prospective Study

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**Abstract:** Background: Constipation and outlet obstruction may persist after successful pull-through in Hirschsprung Disease (HD). The radiographic assessment of the faecal load is widely used but exposes the child to radiation. This study aims to evaluate whether the transrectal diameter (TRD) assessed with ultrasound correlates with symptoms of faecal load and whether the TRD normalises when symptoms disappear. Method: Children with HD after pullthrough and functional constipation presenting to our colorectal clinic between 4/23 and 4/24 were assessed for symptoms of constipation, smearing and outlet obstruction, as well as healthy controls. Ultrasound measurement of the TRD was conducted. Bowel management was initiated according to our institutional pathway using Peristeen© irrigation after an orthograde disimpaction regime. Results: A total of 193 children underwent TRD assessment. Of 60 children with HD, 26 (43.3%) presented with obstructive symptoms, and 34 (56.7%) were asymptomatic. In asymptomatic patients with HD, the mean TRD of 2.26 cm (SD 0.61) was significantly ($p < 0.001$) lower than in HD with symptoms, with a mean TRD of 3.35 cm (SD 1.03). Individuals without colorectal pathology had a mean TRD of 2.04 cm (SD 0.37), and children with functional constipation and symptoms showed a mean TRD of 4.36 cm (SD 1.32). The mean TRD after symptom resolution was 2.37 cm. Conclusions: Children with HD without obstructive symptoms have a TRD < 3 cm, as do controls. The transrectal diameter allows the clinician to sonographically assess the faecal load in children with HD at the bedside without radiation. The TRD is useful for monitoring a bowel management program in children with HD.

**Keywords:** Hirschsprung Disease; bowel management; rectal ultrasound; clinician ultrasound



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### 1. Introduction

The long-term follow-up of children with Hirschsprung Disease to assess the development of bowel function is part of the holistic management of Hirschsprung Disease [1,2]. Constipation and outlet obstruction can persist even after technically sound ganglionic pullthrough. Outlet obstruction and constipation are reported in up to 80% of patients with Hirschsprung Disease (HD) [3–5]. Several groups report that the length of the aganglionic segment does not determine the probability of persistent symptoms [5]. Children are followed routinely, and bowel management is initiated if necessary. Anatomical causes of post-pullthrough problems need to be excluded during a systematic workup [6,7] and treated, if present. We distinguish children with constipation due to outlet obstruction as a consequence of sphincter achalasia or withholding from children with anatomical causes. Postoperative obstacles such as twisted pullthrough, residual aganglionic segments, large Duhamel's pouches or other pouches need to be assessed with clinical examination, ultrasound and, if necessary, contrast enema, repeated biopsy and examination under anaesthesia [6,7].

Soiling in children is most likely due to outlet obstruction; damage to the anal canal should also be assessed by clinical examination [6,7]. Recurrent Hirschsprung's enterocolitis can occur, especially if there are anatomical problems [6,7]. In some children, bowel function problems persist despite a technically sound pullthrough. Defecation disorder is a feature of Hirschsprung's pathophysiology that needs to be addressed through holistic long-term follow-up. To define faecal impaction, thorough clinical examination and history are essential. To assess the severity, visualisation of the faecal impaction with imaging is crucial.

The radiographic assessment of the stool burden is widely used in both colorectal disease and functional constipation, but it exposes the child to radiation [8–10]. Radiographic assessment on plain films is usually made by clinical interpretation [9]. Previously, three different scoring systems have been proposed to assess and evaluate the amount of faecal burden on a plain abdominal radiograph [11]. Assessment on X-ray was first described by Barr in 1979 [12], then by Blethyn in 1995 [13] and then by Leech in 1999 [14]. None of these methods are used routinely in clinical practice. Just recently, a standardised approach to defining different degrees of faecal load and the success of bowel management has been proposed [15]. This approach uses repeated X-ray imaging over several days (5–10) of accumulation of high exposure to radiation [15,16].

Patients with HD require multiple assessments with X-ray during the diagnostic pathway. This exposes the child and, in particular, the pelvic organs such as the ovaries and testes to repeated ionising radiation.

To avoid further radiation exposure, this study aims to measure the transrectal diameter (TRD) to assess the faecal load with ultrasound. Children with HD and children without bowel pathology were investigated. In contrast to previous studies, which excluded children with colorectal disease, we focused on patients with colorectal disease like Hirschsprung Disease [17–19]. This study aims to assess whether the transrectal diameter in children with HD correlates with symptoms of stool burden and whether it normalises when symptoms disappear.

## 2. Materials and Methods

Children with HD presenting to our colorectal clinic between 4/23 and 4/24 after pullthrough surgery were clinically assessed for symptoms of constipation, stool smearing and outlet obstruction. The treating clinicians routinely performed ultrasound measurements of the TRD (Figure 1). If necessary, bowel management was initiated according to our institutional pathway. Peristeen© irrigation was started after orthograde disimpaction. The required irrigation volume was determined by hydrosonography [20]. If transanal irrigation was indicated, the child was trained to be comfortable with rectal catheter insertion, and each child underwent oral disimpaction just before the hydrosonography.

Families were given the opportunity to familiarise themselves with the irrigation system and were supported by a bowel management nurse and a colorectal case manager. Once the irrigation was started, we routinely followed up with the family after 4 weeks, usually with a telephone follow-up and 3 months later with a regular physical follow-up.

We included children with standard Hirschsprung ($n = 60$) and excluded children with Total Aganglionosis ($n = 31$). We included children with functional constipation (FC, $n = 51$). We included healthy controls without any abdominal pathology ($n = 51$). In this analysis, we excluded other colorectal pathologies such as anal atresia and neurogenic conditions. We excluded children below 1 year of age.

### *2.1. Definition of the Level of Hirschprung Disease*

We use the following classification: Type 1—only Rectum, Type 2—Rectosigmoid, Type 3—up to Colon descendens, Type 4—up to Colon ascendens, Type 5—TCA with a maximum of 20 cm of small bowel involvement.

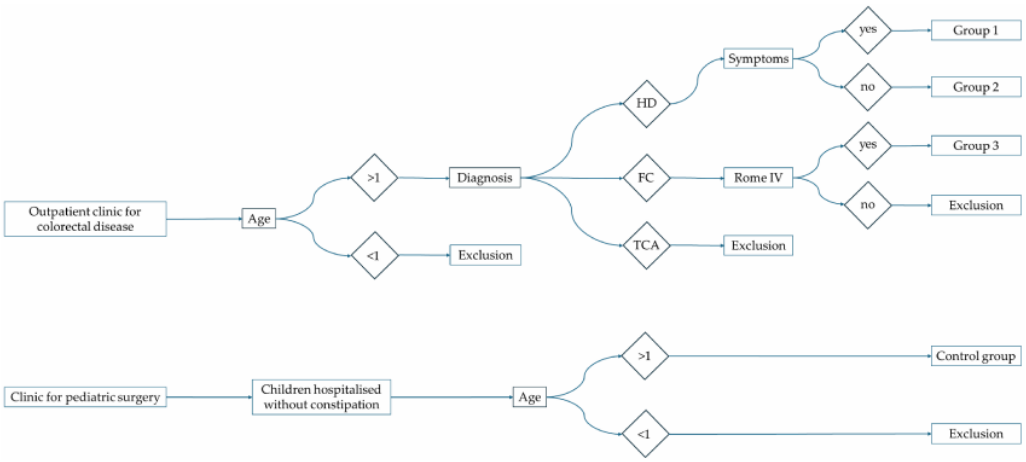


**Figure 1.** Overview of Patient inclusion and selection.

### 2.2. Definition of Symptoms in Children with Hirschsprung Disease

For all children, we assess if their underwear remains clean and if accidental loss appears.

We aim to use the Rintala Bowel Function [1,21] score once the child is toilet-trained for stool to assess bowel function longitudinally.

The Rintala Bowel Function score used to evaluate children with colorectal pathologies assesses the following items: ability to hold back defecation, feels/reports the urge to defecate, frequency of defecation, soiling/staining, accidents/poo in underwear, constipation and social problems.

### 2.3. Definition of Symptoms in Children with Functional Constipation

For functional constipation, we additionally use the Rome IV criteria [22]. Stool consistency was reported according to Bristol Stool chart.

### 2.4. Transrectal Ultrasound Assessment

We used the method of Klijn [23] by placing a curved array of 3.5 MHz (Toshiba Aplio 300, Toshiba Medical Systems GmbH, Neuss, Germany) above the symphysis and measured the largest TRD at a downward angle of 15 degrees from the transverse plane; see Figure 2.

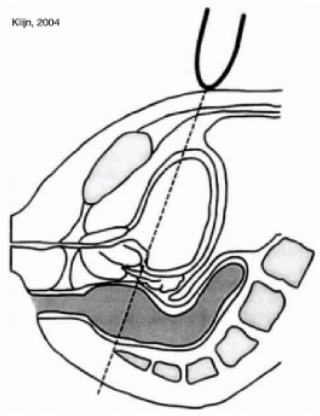


**Figure 2.** Schematic Graphic; TRD according to Klijn. Reprinted with permission from [23].

Figure 3 displays typical ultrasound images with a normal TRD (Figure 3a,c) and feacal load (Figure 3b,d) in children with Hirschsprung Disease (Figure 3a,b), healthy controls (Figure 3c) and those with functional constipation (Figure 3d).

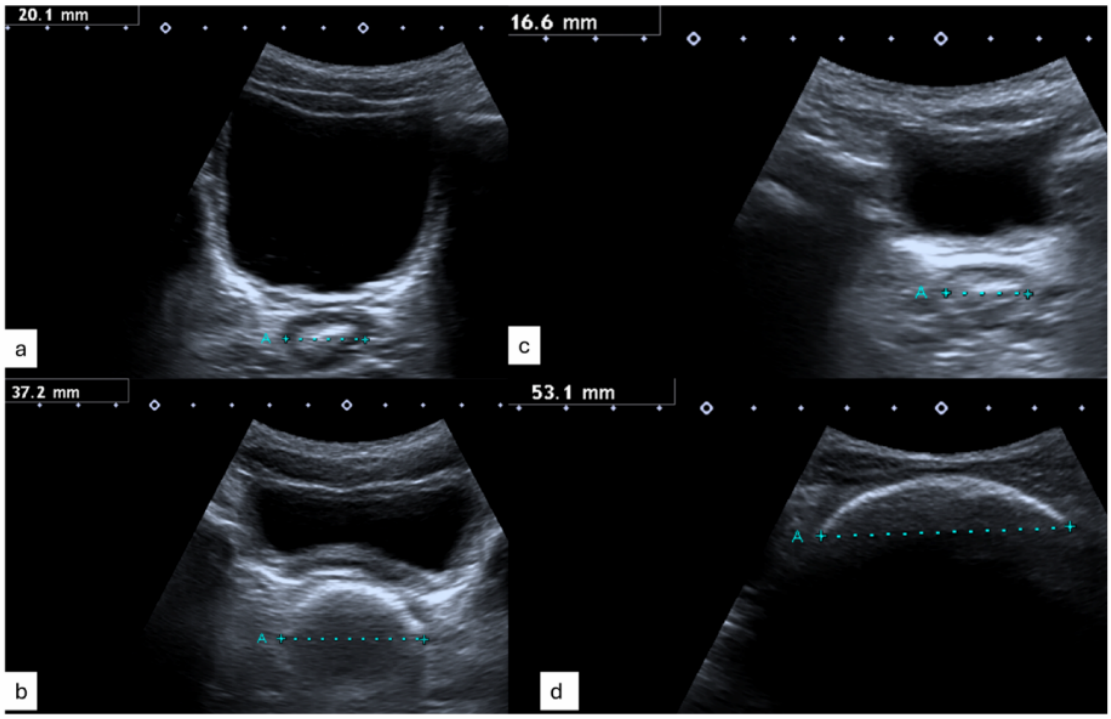


**Figure 3.** Transrectal Diameter and Picture Constipation of (**a**) a child with HD and normal bowel function, (**b**) a child with HD and obstructive symptoms, (**c**) a healthy control and (**d**) a child with functional constipation and symptoms.

## 3. Results

We included 162 children with sonographic assessment of the transrectal diameter. There were a total of 60 children with standard Hirschsprung Disease, of whom 26 (43.3%) presented with outlet obstruction/constipation symptoms and 34 (56.7%) presented without any symptoms. We scanned 51 controls without any symptoms and 51 children with functional constipation.

In this cohort we have follow-up data with a mean of 44.3 months after pullthrough for the children with HD (76% males, 24% females). In total, 34 (56.7%) children had no obstructive symptoms, and 26 (43.3%) had symptoms at the index assessment. The patient characteristics are shown in Table 1.

A total of 162 children underwent a sonographic evaluation of TRD. A total of 60 children had HD, of whom 26 (43.3%) had symptoms of outlet obstruction/constipation and 34 (56.7%) had no symptoms. We scanned 51 controls without symptoms and 51 children with functional constipation. In asymptomatic patients with HD, the mean TRD of 2.26 cm (SD 0.61) was significantly ($p < 0.001$) lower than in HD with symptoms, with a mean TRD of 3.35 cm (SD 1.03); see Figure 4.

**Table 1.** Patient characteristics.

| | **Hirschsprung with Symptoms** ***N* = 26** | **Hirschsprung without Symptoms** ***N* = 34** | **Control without Symptoms** ***N* = 51** | **Functional Constipation** ***N* = 51** | ***p*-Value** |
|---|---|---|---|---|---|
| Gender (female, male) | 7:19 (27%:73%) | 6:28 (18%:72%) | 36:15 (71%:29%) | 26:25 (51%:49%) | |
| Age at pullthrough (months, mean) | 13 | 23 | - | - | |
| Age at rectal ultrasound (years, mean) | 4.85 | 5.23 | 7 | 7 | - HD with vs. without symptoms: $p = 0.457$<br>- HD with symptoms vs. Control: $p = 0.032$<br>- HD with symptoms vs. FC: $p = 0.067$ |
| Months after pullthrough (mean, min.–max.) | 45.2 (8–161) | 38.42 (0–191) | - | - | |
| Mean transrectal diameter TRD at first scan (cm) | 3.45, SD 1.01 | 2.31, SD 0.57 | 2.05, SD 0.37 | 4.36, SD 1.39 | - TRD HD with symptoms vs. without symptoms: $p < 0.001$<br>- TRD HD without symptoms vs. CG: $p = 0.02$<br>- TRD HD with symptoms vs. FC: $p = 0.001$<br>- TRD HD follow-up vs. FC follow-up: $p = 0.874$ |
| Symptoms Yes/No | Yes | No | - | Yes 26 (51%)<br>No 34 (49%) | |
| Symptoms<br>- soiling<br>- stool accidents<br>- distended abdomen | - soiling: 14 (54%)<br>- distended abdomen: 6 (23%)<br>- constipation: 12 (46%) | - | - | - soiling: 17 (65%)<br>- distended abdomen: 7 (27%)<br>- constipation (94%)<br>- faecaloma: 15 (58%) | |
| Months last follow-up (mean) | 4.6 | | - | 3.3 | |
| Transrectal diameter TRD last follow-up (mean) | 2.49, SD 0.5 | | | 2.48, SD 0.36 | |

Individuals ($N = 34$) without colorectal pathology had a mean TRD of 2.04 (SD 0.37), and children with functional constipation and symptoms had a mean TRD of 4.36 cm (SD 1.32). The TRD was significantly larger when children presented with symptoms regardless of the cause; interestingly, children with functional constipation presented with a significantly ($p = 0.001$) larger TRD when comparing HD with symptoms and FC.

Age-appropriate bowel management was initiated using transanal irrigation with a Foley catheter or Peristeen©. The mean time to the resolution of obstructive symptoms was 3.25 months, and the mean TRD after the resolution of symptoms was 2.37 cm.

The irrigation volume was individually determined by hydrosonography and varied between 200 and 800 mL, highlighting the importance of individual ultrasound assessment in each case.

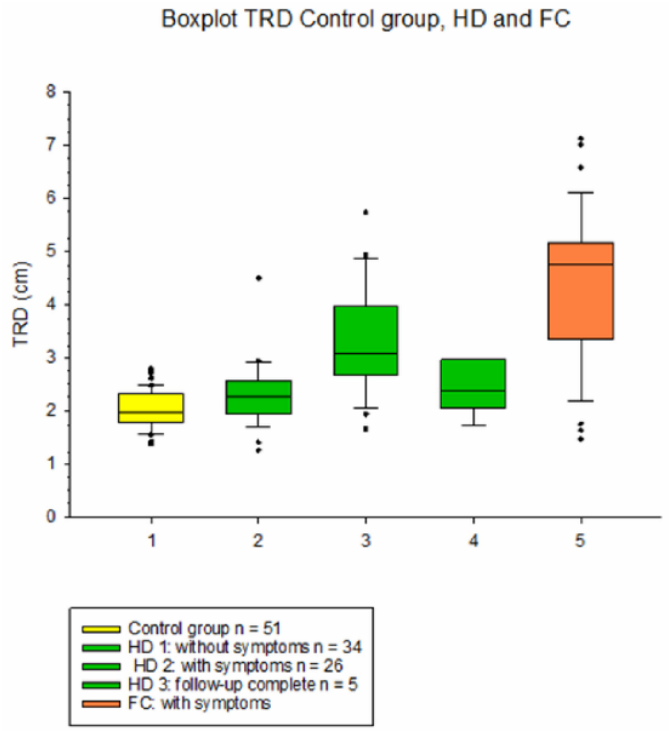


**Figure 4.** Boxplots TRD in control children (yellow), children with Hirschsprung without outlet obstruction/constipation (HD1), children with HD and outlet obstruction/constipation (HD2) and with successful bowel management (HD3) and children with functional constipation with symptoms (orange).

## 4. Discussion

We report on the prospective evaluation of 60 children with standard Hirschsprung Disease at a mean of 44.3 months after pullthrough compared to 51 children with functional constipation in our colorectal clinic. The sonographic transrectal diameter (TRD) was compared with that of 51 healthy controls. In our cohort, 26 children with HD (43.3%) presented with outlet obstruction/constipation and a mean transrectal diameter of 3.35 cm with symptoms. The main symptom was soiling in 54% and constipation in 46% of children with symptoms. When symptoms were resolved with tailored bowel management, these children had a significantly ($p = 0.001$) smaller mean TRD of 2.49 cm.

We performed routine transrectal ultrasound assessments in all children with Hirschsprung Disease as well as controls without any colorectal pathology [24].

A cut-off value of 3 cm in diameter has been validated for children without colorectal pathology to differentiate between constipation and regular defecation [17–19].

In our patients, we have confirmed that this cut-off reliably distinguishes (HD with symptoms vs. without symptoms: $p < 0.001$) patients with Hirschsprung Disease and impaired bowel function from symptom-free HD children.

The pulled through bowel takes over the function of the resected rectum. A filling greater than 3 cm indicates an increased faecal load, which is often associated with impaired bowel function.

### *4.1. Bowel Management in Hirschsprung*

With our individualised yet standardised bowel management program, the transrectal diameter on ultrasound diminished and was lower (⌀2.49 cm) than the proposed cut-off when symptoms disappeared at a mean follow-up of 3.25 months.

The bowel management is initiated with ultrasound control. For toddlers, we opt for passive irrigation using a Silicon Foley and syringe and teach the parents to perform those washouts.

Usually around 3–4 years, we introduce active irrigation with Peristeen© Special to our practice is that we determine the amount of irrigation fluid using ultrasound. Other groups empirically judge the amount or would use a contrast enema [15].

This bedside ultrasound by us enables the families and child to directly observe the irrigation process and engages the patient actively.

Likewise, other groups note that the understanding and cooperation of the patient and parents is crucial for the success of transanal irrigation regardless of the underlying cause [20,25,26].

We empower children to participate early and note that from around 8 years, many can conduct their irrigations independently [27].

Other groups routinely use Botox when outlet obstruction persists, which relieves the Sphincter Achalasia. The effect usually is present for 3–6 months, and the administration may be repeated, but anaesthesia is needed for the intrasphincterial administration. However, transanal irrigation supplements the effect in Botox and should ideally be performed at the same time.

### *4.2. Bowel Management in Chronic Constipation*

Our data on 51 children with functional constipation showed that the sonographic monitoring of TRD supports the implementation of a tailored bowel management programme. The main presenting symptoms were constipation (94%) and soiling (65%). With a success rate of 90% with transanal irrigation (either Peristeen© or Quofora Irrisedeo Mini Go®), we note a higher likelihood of becoming clean compared to others describing the antegrade continence enema (ACE) route, with a success rate of 31.5% [28]. Children are clean on clinical symptoms, and the TRD decreases significantly from abnormal (⌀4.36 cm) to normal (⌀2.48 cm) when symptoms disappear.

Supervised training and sustained follow-up are key features to achieving the best possible outcome with irrigation in children with functional or organic bowel dysfunction [25]. In both Hirschsprung Disease, due to persistent sphincter achalasia caused by impaired anorectal inhibitory reflex, and functional constipation, usually associated with some degree of withholding behaviour, it may be particularly important to use transanal irrigation rather than antegrade irrigation to overcome the outlet obstruction by direct stimulation [29]. Over time, families and especially patients benefit greatly from learning self-management strategies and becoming independent in their bowel management. Self-management has been described as the key element in multidisciplinary disease management strategies in chronic disease care [27].

## 5. Conclusions

This prospective longitudinal assessment of the transrectal diameter on clinician ultrasounds in children with Hirschsprung and controls without colorectal pathology enables the detailed measurement and definition of a transrectal diameter in children with Hirschsprung Disease for the first time.

Children with HD without obstructive symptoms have a transrectal diameter < 3 cm, as do controls. The transrectal diameter allows the clinician quickly to sonographically assess the faecal load in children with HD at the bedside without radiation. The TRD is useful for monitoring a bowel management program in children with HD.

**Author Contributions:** Conceptualisation, J.L. and D.E.; methodology, J.L., D.E., F.S. and S.M.; software, J.L., D.E. and F.S.; validation, J.L., D.E., M.H., E.R., F.S. and S.M.; formal analysis, J.L., D.E. and F.S.; investigation, J.L., D.E., M.H., E.R. and F.S.; resources, J.L.; data curation, J.L., D.E. and F.S.; writing—original draft preparation, J.L. and D.E.; writing—review and editing, D.E., F.S. and M.H.; S.M.; visualisation, D.E. and F.S.; supervision, J.L. and S.M.; project administration, J.L. All authors have read and agreed to the published version of the manuscript.

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**Institutional Review Board Statement:** The study was conducted in accordance with the Declaration of Helsinki and approved by the Ethics Committee of University Rostock (protocol code of A2023-0066 and date of 6 April 2023.

**Informed Consent Statement:** Informed consent was obtained from all subjects involved in the study.

**Data Availability Statement:** The data presented in this study are available on request from the corresponding author. The data are recorded in an anonymous institutional database due to privacy or ethical restrictions.

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**Conflicts of Interest:** The authors declare no conflicts of interest.

## Abbreviations

| | |
|---|---|
| HD | Hirschsprung Disease |
| TRD | Transrectal diameter |
| FC | Functional constipation |
| ACE | Antegrade Continence Enema |

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## 2.4 Point of Care Ultrasound in Pediatric Surgery across the European Region - European Pediatric Surgery Association Endorsed Survey.

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Original Article

# Point of Care Ultrasound in Pediatric Surgery across the European Region—European Pediatric Surgery Association Endorsed Survey

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Eur J Pediatr Surg

**Abstract**

**Introduction** Our purpose was to assess the state of training, clinical practice, and barriers to use point-of-care ultrasound (POCUS) in pediatric surgery in Europe.

**Methods** An electronic survey was disseminated among European pediatric surgeons utilizing the European Pediatric Surgery Association network and other existing networks.

**Results** There were 186 respondents from 27 European countries and 7 non-European countries. In most countries (86.6%; *N* = 161), the initial ultrasound for acute admissions is performed by radiologists, with 1 to 6 hours turnover in 62.9% (*N* = 117) of urgent cases. Ultrasound by pediatric surgeons (point-of-care ultrasound/POCUS) is performed by 48.4% (*N* = 90) of respondents, with 29% (*N* = 54) using it at least once per week. The most common indications for POCUS include abdominal focused abdominal sonography in trauma (53.8%; *N* = 100), diagnosis of appendicitis (41.9%; *N* = 78), and intussusception (44.6%; *N* = 84). In malrotation-volvulus, 28.5% (*N* = 53) used ultrasound for its diagnosis, while 27.5% (*N* = 51) would not see an indication here. Training in POCUS occurred informally for 55.4% (*N* = 103) of participants, while 31.2% (*N* = 58) attended formal training courses. Almost all respondents wanted to attain further POCUS training (89.3%; *N* = 166), only 7% (*N* = 13) did not think this would be useful. For 73.1% (*N* = 136), POCUS is not currently part of the pediatric surgery training curriculum in their country. Perceived barriers to POCUS use include a lack of training opportunities (26.3% [*N* = 49]) and a paucity of portable ultrasound machines (17.8% [*N* = 33]).

**Conclusion** There is a wide spectrum of POCUS use in pediatric surgery across Europe. For those surgeons who practice POCUS, it is most used for the diagnosis of abdominal conditions. There are differing views among clinicians concerning the most useful applications of POCUS. The extent to which ultrasound is taught during pediatric surgery training differs substantially across European curricula.

**Keywords**

- point of care ultrasound
- clinician ultrasound pediatric surgery
- EUPSA survey ultrasound
- pediatric surgery ultrasound

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## Introduction

Ultrasound is usually the preferred imaging mode for children, given its high sensitivity and avoidance of exposure to ionizing radiation. Many surgical conditions affecting children are easily diagnosable with ultrasound. In more recent years, the development of affordable, portable handheld ultrasound devices has helped to expand the use of ultrasound beyond the radiology department. This use of ultrasound by nonradiologist clinicians at the patient bedside is termed point-of-care ultrasound (POCUS). POCUS is a focused examination, aimed to answer a very specific medical question relevant to immediate patient diagnosis and management (e.g., "is there free fluid in the abdomen, yes or no?").[1] In pediatric emergency medicine the potential of POCUS is increasingly recognized, and it has been credited with being able to provide "global improvement of pediatric patient care."[2,3]

Currently, the use of POCUS among pediatric surgeons in the European region seems to be poorly defined. To better understand the current landscape and obtain data on the use of POCUS in pediatric surgery across Europe, we designed and distributed a survey regionally.

## Materials and Methods

Following approval by the EUPSA (European Pediatric Surgery Association) Network Office, a validated questionnaire on the current use, training, and experience of clinician-led ultrasound (POCUS) by pediatric surgeons within the European region was distributed. The survey was launched at the EUPSA Annual Congress in Tel Aviv, Israel, 2022. The period of survey was 10 months. The survey was also disseminated via the EUPSA Homepage and spread via social media.

We defined POCUS (a.k.a. clinician-led ultrasound) as the use of ultrasound technology by a clinician to assess the patient at the bedside, to aid clinical decision-making. It is practiced by clinicians (i.e., surgeons, intensivists, and emergency physicians) and not trained radiologists/ultrasonographers.

The survey was structured in four sections:

- Personal information about the respondent
- Personal experience of POCUS in current practice of respondent
- POCUS training
- Ultrasound use at the institution of the respondent

The collected data were entered into an anonymized database and analyzed using SPSS. Descriptive statistics was performed. All respondents gave their consent for publication of the survey findings.

## Results

### Personal Information about the Respondent

In total, 186 participants ($N = 186$) from 27 countries in the European region (►Fig. 1) and a further 7 non-European countries answered the survey (see ►Fig. 1). In 96.4% cases (180/186), the respondent's current country of work was the same as in their country of training. In total, 35.5% (66/186) were EUPSA members (see ►Fig. 1).

Respondents mainly worked in tertiary hospitals as fully qualified surgeons in pediatric surgery. Respondents had a wide range of experience, with 37% ($N = 67$) of the surgeons at the beginning of training (within 5 years) and 19.9% ($N = 37$) with over 20 years of experience. For more detailed information on the background of respondents see ►Table 1.

### Personal Experience of Point-Of-Care Ultrasound in Current Practice of Respondent

In total, 18.3% (34/186) of respondents had never heard of the term "POCUS" prior to the survey. 46.8% ($N = 87/186$) already used POCUS in their routine clinical practice. Of these 87 surgeons that use ultrasound, 43.7% ($N = 38/87$) use this skill almost every time they are on duty, 37.9% ($N = 33/87$) practice it at least once per week, and 20.7% ($N = 18/87$) at least once per month.

Self-perceived level of competency as stated by the participants of the survey for selected ultrasound examination skills to state basic knowledge (*blue*), confident to perform (*red*), confident to perform and able to train others (*yellow*), unable to perform and respondent does not see a role for ultrasound assessment (*orange*), and respondent cannot perform but would like to learn (*green*; see ►Fig. 2).

Basic knowledge (*blue*) in the inquired ultrasound examinations stated generally a quarter of the respondents with the peak competence level in abdominal focused abdominal sonography in trauma (FAST) in 19.9% ($N = 37/186$) and pleural effusion in 20.4% ($N = 38/186$; see ►Fig. 2).

Confident to perform a selected ultrasound applications (*red*) had a range between 4.8% (9/186) for spinal ultrasound and a maximum of 18.3% (34/186) for ultrasound-assisted guided drainage of an abscess and ultrasound-guided vascular access and diagnosis of intussusception. The highest level of confidence and able to train others (*yellow*) were stated for bladder capacity assessment in 20.9% (39/186), abdominal FAST in 18.8% (35/186), and ultrasound-guided vascular access in 18.3% (34/186; see ►Fig. 2).

In the category "cannot perform and would like to learn" (*green*), the three key applications respondents wanted to learn were pyloric stenosis 29.0% (54/186), diagnosis of intussusception 27.4% (51/186), and diagnosis of malrotation/volvulus 26.3% (49/186). The lowest level was given for bone fractures in 15% (28/186), soft-tissue injuries and hip ultrasound both in 17.2% (32/186; see ►Fig. 2).

The answers for the category "cannot perform and do not see a role for POCUS" (*orange*) showed that 62/186 did not see an indication for malrotation/volvulus, 60/186 for undescended testis, and 86/186 for bone fractures (see ►Fig. 2). Furthermore, our respondents also used POCUS for other clinical conditions not listed in the survey and to aid procedures (see ►Table 2).

## Point-of-Care Ultrasound Training

Only 20.4% (38/186) of respondents indicated that POCUS was included in their pediatric surgery training. In the

 

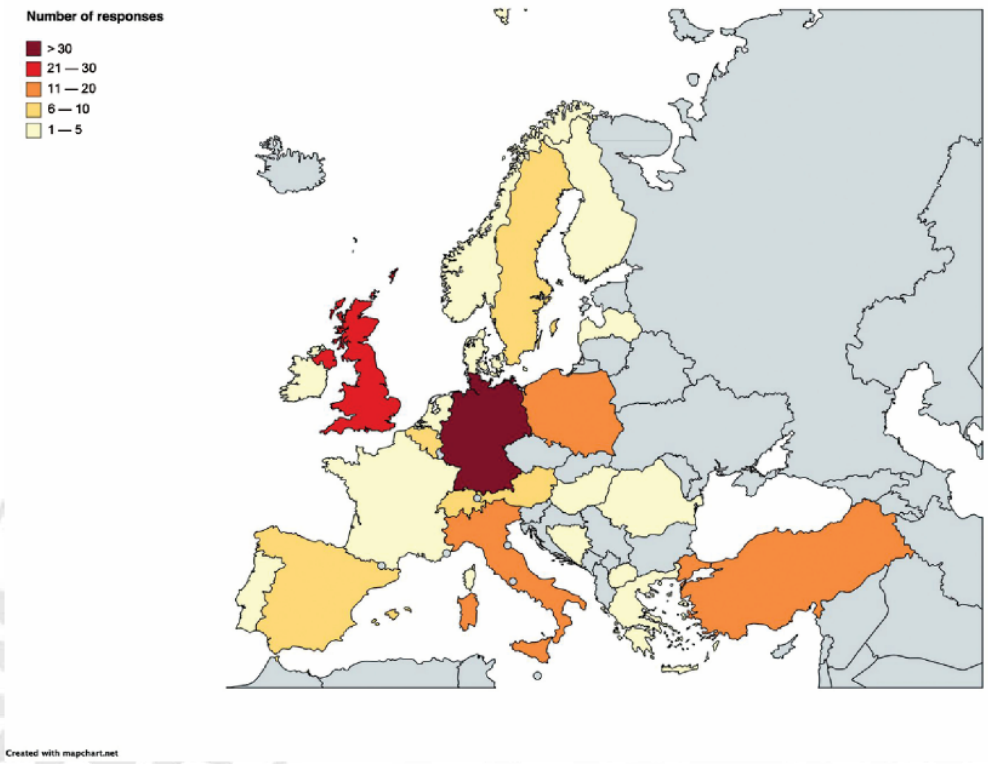


**Fig. 1** Geographic background of respondents—current country of work.

**Table 1** Background of respondents—who took part

| | |
|---|---|
| Level of hospital | Tertiary 92% $N=171$<br>Secondary 6% $N=11$<br>Primary 2.2% $N=4$ |
| Qualification | Qualified pediatric surgeon 62.4% $N=115$<br>Resident/in training 38.2% $N=71$ |
| Specialty | Pediatric surgery 97.8% $N=182$<br>Pediatric trauma/ortho 7.5% $N=14$<br>Pediatric emergency medicine 7% $N=13$ |
| Working experience in pediatric surgery | 20 y 19.4% $N=36$<br>11–20 y 20.4% $N=38$<br>6–10 y 23.7% $N=44$<br>2–5 y 26.9 $N$ 50<br>< 2 y 9.1% $N=17$ |
| Gender | Female 53.2% $N=99$<br>Male 44.6% $N=83$<br>Prefer not to say 2.2% $N=4$ |

following countries surveyed, POCUS was part of pediatric surgery training: Austria, Croatia, Germany, and Ukraine.

Most respondents (73.1% [$N=136$]) had not been trained in POCUS use.

POCUS was not formally included in the training in Belgium, Bosnia, Denmark, Finland, France, Greece, Hungary, Ireland, Italy, Latvia, the Netherlands, North Macedonia, Norway, Serbia, Spain, Switzerland, Turkey, and Portugal.

Of the respondents who performed POCUS, 55.4% ($N=103/186$) learned the skill informally (i.e., from a colleague), 31.2% ($N=58/186$) attended a formal training course, 12.9% ($N=24/186$) learned during training in a different specialty, and 5.9% ($N=11/186$) were taught POCUS during medical school.

In terms of the overall perceived level of POCUS competency among respondents, 31.2% ($N=58/186$) stated they had basic competence, 18.3% ($N=34/186$) felt competent enough to teach others, and 11.3% ($N=21/186$) declared they did not feel competent (see ►Fig. 3).

## Ultrasound Use at the Institution of the Respondent

We asked participants about access to ultrasound performed by radiologists and/or ultrasonographers at their workplace (multiple answers possible). In most institutions (86.6% [$N=161/186$]), ultrasound is performed by a radiologist. In total, 31.2% of ultrasound is also performed ($N=58$) by a pediatric surgeon, 16.7% ($N=31$) by an ultrasonographer/technician, 21% ($N=39$) by an emergency physician, and in 12.3% ($N=23$) by any other doctor.

 

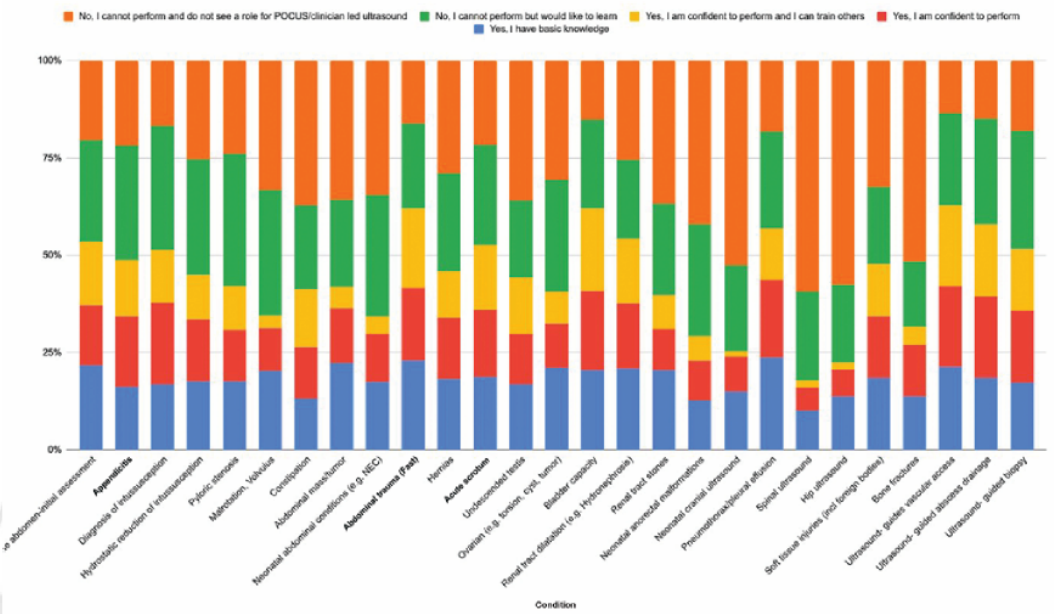


**Fig. 2** Conditions POCUS is used to diagnose by respondents and their self-perceived level of competency.

**Table 2** Further indications where ultrasound is used by pediatric surgeons

| Further gastrointestinal use | Musculoskeletal | CNS questions |
|---|---|---|
| Endorectal ultrasound | Muscle lesion | Intracranial pressure nervus opticus |
| Perineal ultrasound | Joint ultrasound | Cranial and abdominal ultrasound in children with VP shunt |
| Fluid status IVC | Ultrasound guided suprapubic | |
| Gall bladder | | |
| Bowel management | | |
| Intraoperative assessment biliary tract | Lymphangioma | |
| Intraoperative transplant patency particularly liver | | |

Abbreviations: IVC, inferior vena cava; VP, ventriculoperitoneal.

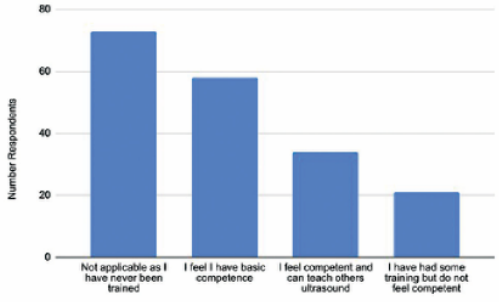


**Fig. 3** Self-perceived general level of POCUS competency among pediatric surgeons.

In total, 53.2% ($N = 99$) of respondents claim to routinely request an ultrasound from the radiology department and 31.7% ($N = 59$) request an ultrasound from the radiology department almost every day. In terms of the time-to-ultrasound being performed from the time-of-request, in 28% ($N = 52/186$) of cases, an ultrasound scan could be attained from the radiology department within 1 hour. For 62.9% ($N = 117/186$), the scan took approximately 1 to 6 hours; for 3.8% ($N = 7/186$), 6 to 12 hours; for 2.7% ($N = 5/186$), 12 to 24 hours, and for 1.1% ($N = 2/186$) of cases, there was no guarantee of the scan being performed once requested.

Access to departmental ultrasound scans out of hours was also assessed in the survey. For 40.9% ($N = 76/186$) of respondents, radiologists/ultrasonographers performed ultrasound outside working hours. However, for 43.5% ($N = 81/186$), this

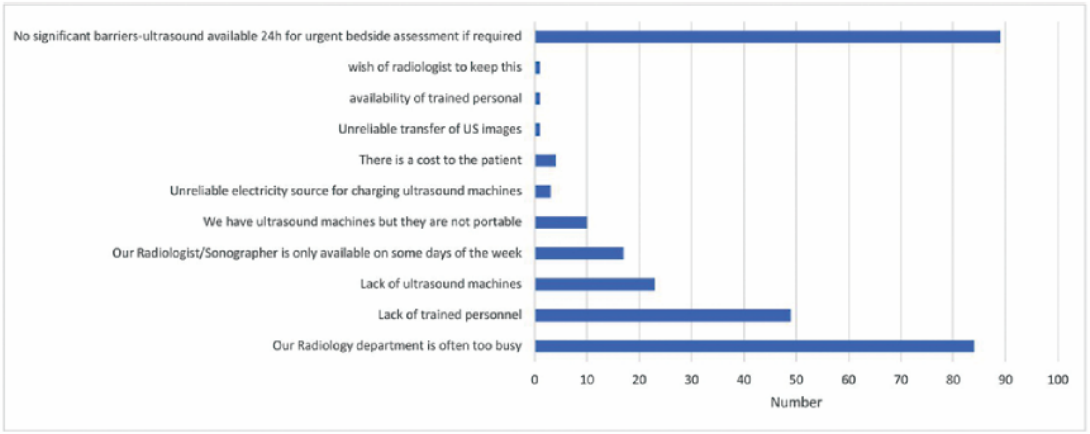


**Fig. 4** Barriers to access ultrasound.

was only in the case of an emergency and the radiologist/ultrasonographer was off-site. For 14.5% ($N = 27/186$), there was no ultrasound available outside or normal working hours.

Respondents were asked to identify barriers to accessing ultrasound services at their institution and multiple statements were possible. The main perceived barrier to attaining a radiology department ultrasound was the department being too busy to accommodate the demand, stated in 45.2% (84/186). Other barriers identified included a lack of trained personal in 26.3% (49/186), and a lack of accessible ultrasound devices in 12.3% (23/186), see ►Fig. 4.

Participants were also given the opportunity to write free script feedback on their views on the role of POCUS in pediatric surgery. ►Table 3 summarizes some of the free responses. Given the opportunity for free answers, we found a variability of answers asking to include ultrasound in training and enable clinicians to get real-time visualization but also respondents expressed their concerns that this would overwhelm the workload and ultrasound should remain in the hand of radiologist, see ►Table 3.

## Discussion

Our survey is the first assessment of the current practices and views European Pediatric Surgeons have regarding the value of POCUS to our specialty. The results of this survey demonstrate that there is a wide variation in the degree to which pediatric surgeons are skilled in POCUS and apply it in their clinical practice across the region.

The possibility to use ultrasonography at the bedside by using mobile or hand-held devices particularly benefits pediatric patients by avoiding transfer and changing environments.[4–7]

### Personal Information about Respondent

We have captured the practice of 186 pediatric surgeons representing all levels of work experience working in 34 countries: 27 in the European region and 7 outside Europe. Thirty-five percent, 66/186 were EUPSA members. 53.2% (99/186) were female reflecting a balanced surgical workforce and the reality of pediatric surgery. The survey reached interest across the level of seniority, and the respondents were equally distributed to work experience.

### Personal Experience of Point-Of-Care Ultrasound in Current Practice of the Respondent

Surprisingly, 18.3% ($N = 34/186$) of respondents had not heard of the term "POCUS" prior to the survey, despite the relatively widespread integration of pocus into contemporary medical practice across multiple specialties. However, those pediatric surgeons who did have POCUS skills used it regularly in their clinical practice, to enable real-time diagnosis of a wide range of pediatric surgical conditions.[4] This demonstrates that POCUS adds value to the daily practice of surgeons who have the skill. Of those with POCUS skills, they mainly used it to assess children presenting with an acute abdomen. Clinician ultrasound is mostly used to assess and facilitate the diagnostic workup of intussuception, appendicitis, or FAST-ultrasound (focused assessment with sonography in trauma) scan in the setting of pediatric trauma.

This is in keeping with the literature, where POCUS has been described in several institutions as a valuable means of providing timely diagnosis of intussusception, in order to avoid delays in treatment and reduce the risk of bowel necrosis.[8–10] Several studies have demonstrated 100% sensitivity in detecting intussusception when POCUS is performed by non-radiology clinicians.[8–11] 44.5% ($N = 83/186$) of respondents had self-perceived competence (from basic to being confident enough to train others) in diagnosing intussusception with POCUS. About 27.4% ($N = 51/186$) would like to learn POCUS for this purpose. 14.5% ($N = 27/186$) did not see value in their learning to diagnose intussusception with POCUS.

### Point-of-Care Ultrasound Training

Regarding training options for pediatric surgeons to learn POCUS, our survey highlighted the lack of POCUS training within surgical curricula across Europe. Only 20.4% (38/186) stated that clinician-led ultrasound was part of their pediatric surgery curriculum. For those participants who did have POCUS skills,

 

**Table 3** Free comments on clinician ultrasound use

| |
|---|
| Regular rotation to radiology department |
| We should all be trained in ultrasound (US) as sometimes a radiologist is not available. US by surgeon is specific to the points the surgeons need. |
| I think a POCUS course with a combination of formal training and scans that are reviewed by an accredited instructor and this training could be valuable for surgical trainees. |
| In my experience, even highly qualified, specialist pediatric radiologists can find it difficult to perform USS and always be definitive. Conversely, I've seen nonradiologists perform POCUS and get things done in a very wrong way, resulting in the detriment of the patient. We have specialist radiologists for a reason. We should focus on having more of them/ access to them OOH if it's needed, and we should NOT be encouraging "hobby-ists" who think they can do it. Pediatric surgeons already do/look after too much! Why are we adding to our professional burdens. |
| İt would be great to have training in USG-guided interventional procedures, especially percutaneous drainage of abscess or fluid collections, biopsies, and PTC procedures. However, the legal issues should be investigated, and legal ground should be developed. |
| POCUS course should be introduced in EUPSA Annual Congresses ×2 |
| US is a real-time application; it is necessary for the radiologist himself to do it. However, most cases requiring US are emergency cases such as acute abdomen, testicular torsion, and trauma, and they often occur at work hours; we, pediatric surgeons, are on duty 24 h a day and 7 d a week, and radiologists should likewise be included in the seizure system. Please do not fall into this trap of radiologists |
| Clinical application of POCUS may probably be (1) country- or language-dependent and (hence) (2) remuneration-driven (if in a country remuneration for US is only reimbursed to radiologists… only radiologists will perform US, as other specialists cannot receive remuneration for the act of performing US) |
| Respondents think further research should include |
| Demonstration of safety and establishment of a protocol for training and implementation |
| FAST POCUS and US-guided intervention |
| Vascular access |
| Acute illness that includes immediate decision and time-sensitive flowchart (i.e., testicular torsion, ovarian torsion, chest, and abdominal trauma |

Abbreviations: ECHO, echocardiography; EUSPA, the European Paediatric Surgeons' Association; FAST, focused abdominal sonography in trauma; FICE Focused Intensive Care Echocardiography; OOH, out of hours; POCUS, point-of-care ultrasound; PTC, percutaneous transhepatic cholangiogram; USG, ultrasonography; USS, ultrasound scans.

most of them had attained these through self-teaching or informal mentorship. We note that different countries have different approaches and only participants from Germany, Spain, Turkey, Croatia, and Ukraine state ultrasound practice is part of their pediatric surgery training program. This is contrasted with the United States where clinician-led ultrasound (POCUS) is increasingly incorporated in a multitude of training programs, particularly in pediatric emergency medicine.[1–5] The literature that evaluates POCUS use in surgery mainly focuses on surgeons performing FAST scans, ultrasound-assisted vascular access, soft tissue assessment, regional anaesthesia, focused echocardiography, and ultrasound guidance for other interventional procedures.[2,12] Trainees report increased self-efficacy and confidence even after short course lengths, that is, a total of 14 hours.[2]

Unfortunately, there is no consensus in the literature as to what constitutes minimum education requirements and minimum competency standards for POCUS training in surgical applications. Including a lack of clarity as to the accepted minimum number of scans that need to be performed to achieve a safe level of clinical accuracy.[12] It is clear, however, that once training is received, the practitioner needs to continually use the skill to master the procedural skill of ultrasound. Many POCUS training models, therefore, emphasize the importance of mentor-led training schemes and refresher courses. Training can also be augmented by high-fidelity simulators and virtual-reality simulators which are increasing in both popularity and affordability.[13] However, we do notice that for most applications a quarter of our respondents' state to be confident not only to perform the ultrasound examinations but also able to teach others.

### Ultrasound Use at the Institution of the Respondent

Our findings regarding the general demand for ultrasound scans by pediatric surgeons confirm the critical importance of this imaging modality to our specialty and the universal use of ultrasound to assess children. All respondents regularly requested ultrasound in their routine practice and the majority of these were performed by radiologists/ultrasonographers.

The barriers to timely ultrasound reported by our respondents are similar to those reported in other high- and low-income countries, that is, pressure on local radiology departments and lack of trained personnel.[14] POCUS provides a potential solution to some of these issues, as it allows a trained clinician to make a rapid real-time diagnosis at the patient's bedsides directly informing patient management. This form of "task-shifting" could help to relieve some of the demand on local radiology services, where "simple"

 

ultrasound scans are required and allow the radiology department to focus on in-depth ultrasound questions we as clinicians cannot answer with POCUS. An additional issue is the lack of radiology-performed ultrasound availability outside working hours, where 14.5% of respondents ($N = 27/186$) had no ultrasound access out of hours. Performing POCUS for selected indications in this context could help clinicians provide the same standard of care for patients 24 hours a day.[4]

One of the concerns about the use of POCUS relates to the reliability and accuracy of clinical-led ultrasound scans. However, several studies have shown that for basic indications such as appendicitis and intussusception, trained clinicians are able to perform ultrasound with reliable accuracy, leading to a reduction in time to diagnosis for the patient.[2,4,5,7,9,11,15,16]

The individual free-text responses from participants in the survey do highlight some conflicting views as to whether pediatric surgeons should formally incorporate POCUS into their practice. Looking across the Atlantic, precedent has been set to incorporate clinician-led ultrasound into undergraduate curricula and many different residency training programs. There is also mass support from radiologists who encourage the uptake of ultrasound skills by clinicians and support POCUS training.[17] Overall, there is growing evidence promoting POCUS in the assessment of acutely unwell children to facilitate prompt diagnosis and appropriate targeted treatment.[6,8,17,18]

## Limitations

This study has several limitations, including that that this survey asks clinicians about their training and practice, and therefore, reflects only the view of individual participants. Although we had many responses ($N = 186$), the proportional overall representation from each of the 34 countries was low and therefore we acknowledge that our results more reflect the general trend in pediatric surgery in Europe, but we cannot reliably read in a country-level data. There might be a degree selection bias as is the case with most surveys, respondents tend to be people with an interest in the topic at hand. However, our response rate is similar to previous surveys endorsed by the EUPSA network office (i.e., EUPSA Appendix Survey 169 respondents, EUPSA Survey Management Primary Spontaneous Pneumothorax 131 respondents, and EUPSA Survey Management Hirschsprung 294 members).[19]

## Conclusion

This is the first survey to evaluate POCUS use by pediatric surgeons in Europe, investigating the current state of ultrasound accessibility and training in the European region. Our data reflects a very heterogeneous landscape. We do, however, demonstrate that many pediatric surgeons have already incorporated POCUS into their practice and use it for a broad range of indications and are confident enough to teach others. There is a strong demand for formal POCUS training courses/curricula among pediatric surgeons across Europe. Evidence-based training modules and clinician-led ultrasound protocols for pediatric surgery conditions would help to facilitate training and set minimum standards for clinicians' accuracy and confidence levels. Overall, incorporating POCUS in the training and daily routine of pediatric surgeons has the potential to enhance the speed of diagnosis and reduce the time to definitive care for children with surgical disease.

### Conflict of Interest

None declared.

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## 2.5 Global survey on point-of-care ultrasound (Pocus) use in child surgery.

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ORIGINAL ARTICLE

# Global survey on point-of-care ultrasound (pocus) use in child surgery

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**Abstract**

**Purpose** To undertake a global assessment of existing ultrasound practices, barriers to access, point-of-care ultrasound (POCUS) training pathways, and the perceived clinical utility of POCUS in Child Surgery.

**Methods** An electronic survey was disseminated via the GICS (Global Initiative of Children's Surgery) network. 247 anonymized responses from 48 countries were collated. 71.3% (176/247) worked in child surgery.

**Results** Ultrasound was critical to practice with 84% (147/176) of requesting one daily or multiple times per week. Only 10% (17/176) could access emergency ultrasound < 1 h from request. The main barrier was a lack of trained personnel. HIC surgeons were more likely to have ultrasound training (24/29; 82.8%) compared with LMICs (74/147; 50.3%) (p = .001319; CI 95%). Self-perceived POCUS competence was associated with regularity of POCUS use ($p < 0.001$; CI 95%). Those who already practice POCUS most commonly use it for trauma, intussusception, and ultrasound-guided procedures. Majority (90%; 159/176) of child surgeons would attend formal POCUS training if available.

**Conclusions** Ultrasound is critically important in children's surgery globally, however, many surgeons experience barriers to timely access. There is a strong interest in learning POCUS for relevant pediatric surgical applications. Further research is needed to evaluate the best methods of training, accreditation, and governance.

**Keywords** POCUS · Point-of-care ultrasound · Pediatric surgery · Bedside ultrasound · Clinical ultrasound

## Introduction

Point-of-care ultrasound (POCUS) or 'clinician-led' ultrasound refers to the use of ultrasound by non-radiologist clinicians at the patient's bedside. Although for many years ultrasound has been the domain of radiologists and specialist sonographers, several critical developments have occurred that have altered the clinical landscape. Technological advances have facilitated the development of cheaper and increasingly smaller handheld devices that interface wirelessly with most screens [1]. The user-friendliness of ultrasound devices in part has contributed to the rapid expansion of POCUS into almost all areas of clinical medicine [2–7]. The specialists who have led the way include Emergency Physicians and Intensivists who have developed POCUS curricula, courses, and accredited training programs for their respective fields [8–10]. The value of POCUS to Anesthesia through facilitating vascular access, cardiovascular monitoring, and guiding regional anesthesia has also been demonstrated [8, 11]. Similarly, acute gynecological presentations and obstetric care have utilized POCUS in their diagnostic

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algorithms and have a large body of evidence to support its use [5, 12].

The field of Pediatric Surgery could stand to benefit from this technology, given the large proportion of pathologies amenable to ultrasound diagnosis, the conducive body habitus of children, and the importance of avoiding exposure to ionizing radiation. In emergency medicine in particular, many studies have shown equivalent diagnostic accuracy when emergency physicians have been trained to diagnose appendicitis, intussusception, and pyloric stenosis [13–15]. While there is evidence of some pediatric surgeons practicing POCUS in Europe [6], and sporadic reports from other regions [14, 16–19]; overall there is a paucity of published data regarding current POCUS practices in pediatric surgery worldwide.

While the majority of POCUS published literature exists in high-income countries (HICs), theoretically low-resourced healthcare settings also stand to benefit from the 'task shifting' and 'capacity-building' advantages POCUS offers. A particular challenge in LMICs is a critical lack of radiologists. Of 26 HICs evaluated, the average number of radiologists is 100 per million population [20–24]. Compared with studies evaluating countries in Africa, where the estimated number of radiologists is between 6.5 and 12 per million [20–24]. The practice of pediatric radiology is especially neglected, with only 4 of 54 countries in Africa offering sub-specialization in pediatric radiology (Ethiopia, Nigeria, South Africa, Tunisia) [25].

Using just one example, intussusception is a common emergency pediatric surgical condition for which there is a significant disparity in outcomes between high and low-income countries [26]. In LMICs, the mortality rate is greater than 10%, compared with < 0.2% in HICs [26]. The diagnosis of intussusception is time-critical and hinges on ultrasound use. Ultrasound can also be used to guide hydrostatic or pneumatic reduction in the treatment of intussusception. Focusing on even this single condition, there is the potential to help improve diagnostic and referral pathways for children with intussusception in LMICs through more ubiquitous access to ultrasound skills for clinicians.

Given the lack of data regarding POCUS use in children's surgery, we developed a global survey to evaluate existing practices, barriers to ultrasound access, training pathways, and the perceived utility of POCUS in our field.

## Methods

Following approval by the Global Initiative of Children's Surgery (GICS) an electronic survey was disseminated via the GICS network and associated social media webpages in English language. Components of the survey were developed by pediatric surgeons with POCUS expertise. The term 'POCUS' was defined as the use of ultrasound by a (non-radiologist/non-sonographer) clinician in the assessment or treatment of a patient.

The survey assessed 4 areas:

- **Domain 1: Demographics**—information about the respondent's role and country of practice.
- **Domain 2: Ultrasound Services**—information regarding ultrasound services, infrastructure, and barriers to access at the respondent's institution.
- **Domain 3: POCUS Training** – respondent's experience of POCUS training (or lack thereof) and utilization of POCUS skills in their current clinical practice.
- **Domain 4: POCUS Applications** – assessment of the respondent's views on the clinical relevance of potential POCUS uses ('applications').

Twenty two potential POCUS applications were selected based on common pediatric conditions amenable to ultrasound diagnosis and the current use of POCUS in children's healthcare as described in the literature. The survey was closed after 12 months. All responses were anonymous, and all respondents gave consent for publication of the survey findings.

There were 247 responses to the survey. These were collated in a secure electronic database. Of the 247 responders, 176 ([71.3%) were child surgeons; the remaining 71 worked in medical specialties. 'Child surgeons' were defined as those who identified as having specialized in 'Pediatric Surgery' or were from a different surgical specialty (i.e. general surgery, orthopedics, etc.) but routinely cared for children. Responses were compared between LMICs and HICs. Descriptive statistics were performed and chi-squared tests for statistical significance were used for categorical data. Statistical significance is indicated if $p < 0.05$ with a 95% confidence interval (CI).

## Results

### Domain 1—demographics

A total of 247 responses from 48 countries were received (additional data on a number of responses per country are given in Online Resource 1). These represented the continents of Africa, Asia, Europe, North and South America (Fig. 1). Countries were grouped by income level according to their official classification in the *World Bank List of Economies* [27] (Fig. 2). Most respondents were from LMICS 85.4% (211/247), with 14.6% (36/247) responses from HICs. Of the 176/247 child surgeons ($n = 176$), there was an almost equal split between consultant/attending level doctors 52.8% (93/176) and trainees/clinical officers

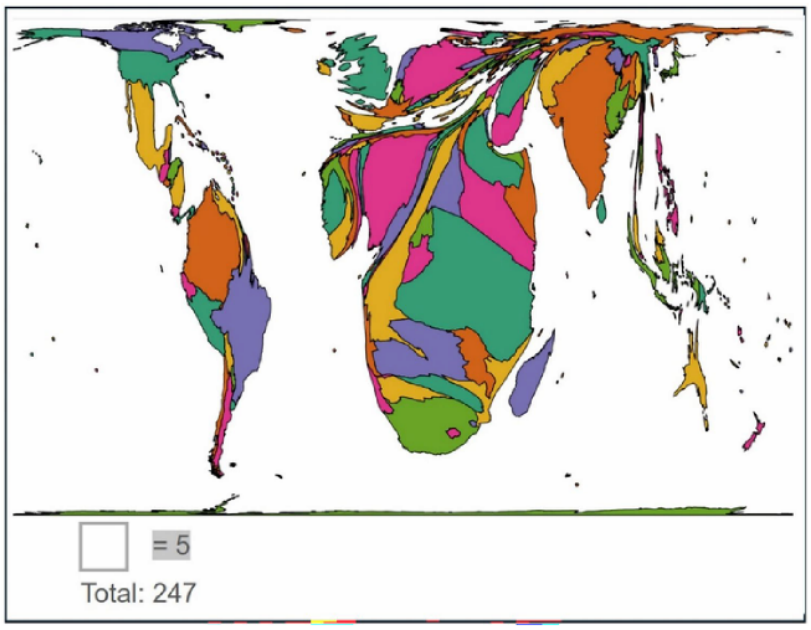
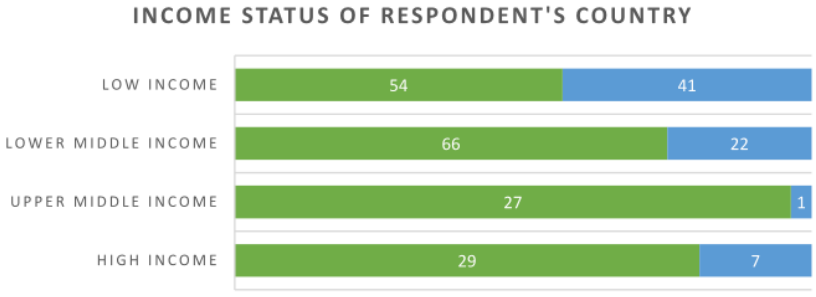



**Fig. 1** Cartogram showing the proportion of survey responses from different countries (Go-Cart program used to create cartogram)




| | High income | Upper middle income | Lower middle income | Low income |
|---|---|---|---|---|
| Child Surgeons (n=176) | 29 | 27 | 66 | 54 |
| Non Surgeons (n=71) | 7 | 1 | 22 | 41 |

**Fig. 2** Number of responses according to the income level of the country as defined by the Work Bank

47.2% (83/176). Child surgeons predominantly worked in tertiary level institutions 83% (145/176); compared with secondary level 16% (28/176); and only 1% (3/176) from primary level healthcare facilities.

Surgeons ($n = 176$) were mainly specialists in Pediatric Surgery 63% (110/176) or General Surgery 29% (30/176). The remaining responses were from 5 other surgical specialties (Fig. 3). All respondents routinely assessed or operated on children.

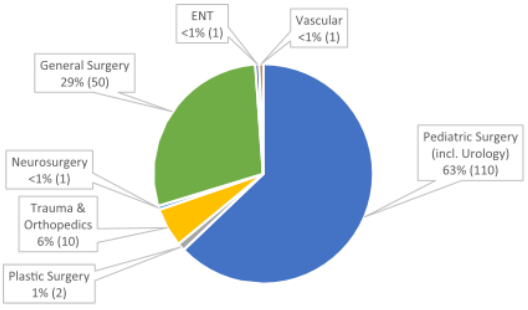



**Fig. 3** Breakdown of Child Surgeons specialties ($n = 176$)

## Domain 2 – ultrasound services

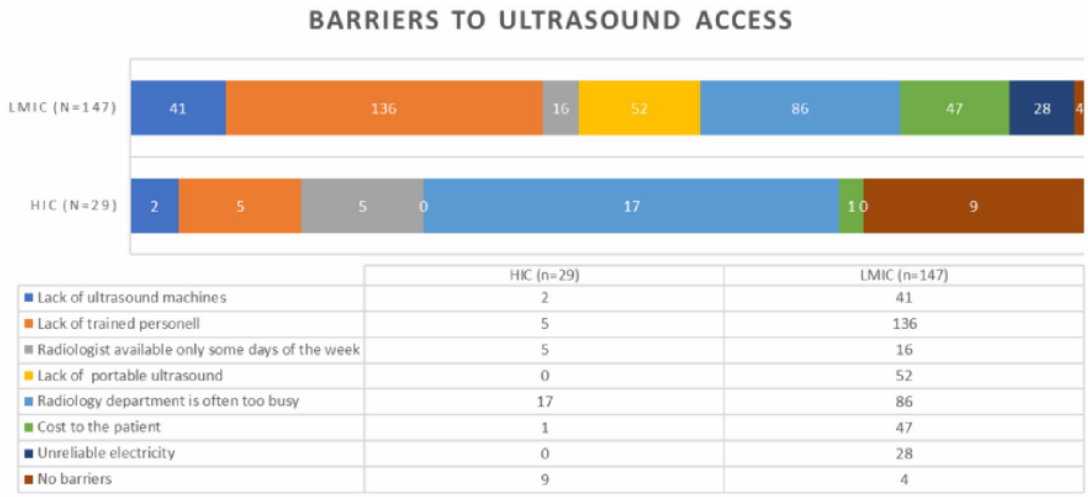
We evaluated the availability of clinical investigations and ultrasound practices at the respondent's hospital (Table 1). All had blood tests and ultrasound devices available at their institution (176/176). Cross-sectional imaging was available in all HIC settings (29/29) but only available in 65% (96/147) of LMICs. In terms of 'normal' ultrasound practices, in most respondent's institutions ultrasound was typically performed by radiologists (or specialist sonographers) 64% (112/176). However, in almost a quarter of responses 24% (43/176), clinicians were the primary ultrasound operator, not radiologists. An equal mix of radiologists and clinician-led ultrasound was found in 11% (20/176). One respondent had access to ultrasound devices but no trained personnel to perform the skill. There was no statistically significant difference between LMICs and HICs in terms of who the principal ultrasound user was ($P > 0.05$; CI 95%).

In the clinical practice of child surgeons, ultrasound was a commonly used investigation, with 84% (147/176) requesting an ultrasound for a child either daily or multiple times per week—with no significant difference between LMIC and HIC groups ($p > 0.05$; CI 95%). In terms of 'out of hours' access to radiology-performed ultrasound, 19% (34/176) had no access, while 63% (111/176) could access these services but only in an emergency. We assessed the time for an emergency ultrasound from the request for an acute indication. In only 10% (17/176) of cases would emergency ultrasound be reliably performed < 1 h from request. For 40% (58/147) of LMIC respondents, an emergency ultrasound took more than 6 h to be performed; with 14% of these beyond 24 h from request (Table 1). No significant differences between LMIC and HIC were found across time intervals ($p > 0.05$).

We also investigated barriers to ultrasound access that child surgeons encountered at their hospital (Fig. 4). A lack of ultrasound-trained personnel and the workload of the radiology department were the commonest issues. In LMICs compared with HICs the following barriers were found to be statistically significant (CI 95%) – lack of machines ($p = 0.0162$); lack of trained personnel ($p = 0.0001$); lack of portable machines ($p = 0.0001$); cost to the patient ($p = 0.0016$); reliability of electricity source ($p = 0.0016$). HICS were more likely to have no barriers ($p = 0.0001$).

**Table 1** Ultrasound infrastructure and access at the respondent's institution

| | Total ($n = 176$) | HIC ($n = 29$) | LMIC ($n = 147$) |
|---|---|---|---|
| Available Investigations | | | |
| Blood tests | 100% (176) | 100% (29) | 100% (147) |
| Radiographs | 100% (176) | 100% (29) | 100% (147) |
| Ultrasound device/s | 100% (176) | 100% (29) | 100% (147) |
| Cross-sectional imaging | 71% (125) | 100% (29) | 65% (96) |
| Who normally performs ultrasound? | | | |
| Radiologist/sonographer only | 64% (112) | 48% (14) | 67% (98) |
| Non-radiologist (doctor or clinical officer) only | 24% (43) | 7% (2) | 12% (18) |
| Mix of both | 11% (20) | 45% (13) | 20% (30) |
| No ultrasound users available | < 1% (1) | 0% (0) | < 1% (1) |
| How often do you request an ultrasound? | | | |
| Almost daily | 52% (91) | 55% (16) | 51% (75) |
| Multiple times per week | 32% (56) | 24% (7) | 33% (49) |
| Multiple times per month | 9% (15) | 3% (1) | 10% (14) |
| Rarely/never | 7% (12) | 7% (2) | 4% (6) |
| N/A | < 1% (1) | 7% (2) | 1% (2) |
| No response | < 1% (1) | 3% (1) | 0% (0) |
| Time to ultrasound from request? *(Radiology Department) | | | |
| < 1 h | 10% (17) | 10% (3) | 10% (14) |
| 1–6 h | 52% (92) | 59% (17) | 51% (75) |
| 6–12 h | 13% (23) | 10% (3) | 14% (20) |
| 12–24 h | 9% (16) | 10% (3) | 9% (13) |
| > 24 h | 10% (17) | 0% (0) | 12% (17) |
| No guarantee it will be done | 5% (8) | 3% (1) | 5% (7) |
| No radiology department | < 1% (1) | 0% (0) | < 1% (1) |
| No response | 1% (2) | 7% (2) | 0% (0) |




| | HIC (n=29) | LMIC (n=147) |
|---|---|---|
| Lack of ultrasound machines | 2 | 41 |
| Lack of trained personell | 5 | 136 |
| Radiologist available only some days of the week | 5 | 16 |
| Lack of portable ultrasound | 0 | 52 |
| Radiology department is often too busy | 17 | 86 |
| Cost to the patient | 1 | 47 |
| Unreliable electricity | 0 | 28 |
| No barriers | 9 | 4 |

**Fig. 4** Barriers to ultrasound access described by respondent

**Table 2** Child surgeons (n = 176) experience of POCUS training, self-perceived competency, and clinical

| POCUS Training | |
|---|---|
| Have you ever had any ultrasound training? ($n=176$) | |
| No, I have never been trained | 44% (78) |
| Yes, I attended a training course | 27% (48) |
| Yes, I received informal training from colleagues | 28% (50) |
| If you have received training, how competent do you feel using ultrasound to assess patients? ($n=98$) | |
| I do not feel competent | 32% (31) |
| I feel I have basic competence | 46% (45) |
| I feel competent and can teach others | 18% (18) |
| Non-response | 4% (4) |
| If you have received training, how often do you personally perform ultrasound on your patients? ($n=98$) | |
| Daily | 26% (25) |
| At least once per week | 21% (21) |
| At least once per month | 24% (23) |
| Never | 25% (24) |
| Non-response | 5% (5) |

## Domain 3—ultrasound training

We asked respondents about their experience of ultrasound training, competence levels, and use of POCUS in their clinical management of children (Table 2). Approximately half of all respondents had received some kind of informal or formal ultrasound training (55%; 98/176). HIC surgeons were statistically more likely to be trained 24/29 (82.8%) compared with LMIC surgeons 74/147 (50.3%) ($p=0.001319$; CI 95%). Of those who had received training ($n=98$), most 46% (45/98) felt they had basic ultrasound competence; and almost half 47% (46/98) used POCUS daily or weekly in their practice. Self-perceived competence was associated with where the child surgeon used POCUS regularly in their clinical practice ($p<0.001$; CI 95%). There were high levels of interest in attending a POCUS training course designed specifically for child surgeons, with 90% (159/176) saying they would be interested in attending such a course; 4% (7/176) had no interest; and 6% were unsure—no difference between LMIC and HIC groups ($p>0.05$; CI 95%).

## Domain 4—POCUS applications

Respondents were asked to identify which of 22 potential POCUS applications they either already used POCUS for, would be interested in learning to use POCUS for, or had no interest in (Fig. 5). The top five most common uses of POCUS amongst child surgeons ($n=176$) were FAST (focused assessment with sonography for trauma), intussusception, ultrasound-guided procedures, hydronephrosis, and pneumothorax detection. However, the most common applications respondents wanted to learn POCUS for were malrotation, pyloric stenosis, acute scrotum, appendicitis, and assessing the neonatal abdomen. The least useful applications included fractures, cranial ultrasound, constipation, hernias, and foreign bodies/soft tissue injury. Overall, there were high rates of interest in learning all of the 22 potential applications for POCUS, with a nadir of 57% (101/176) for fractures.

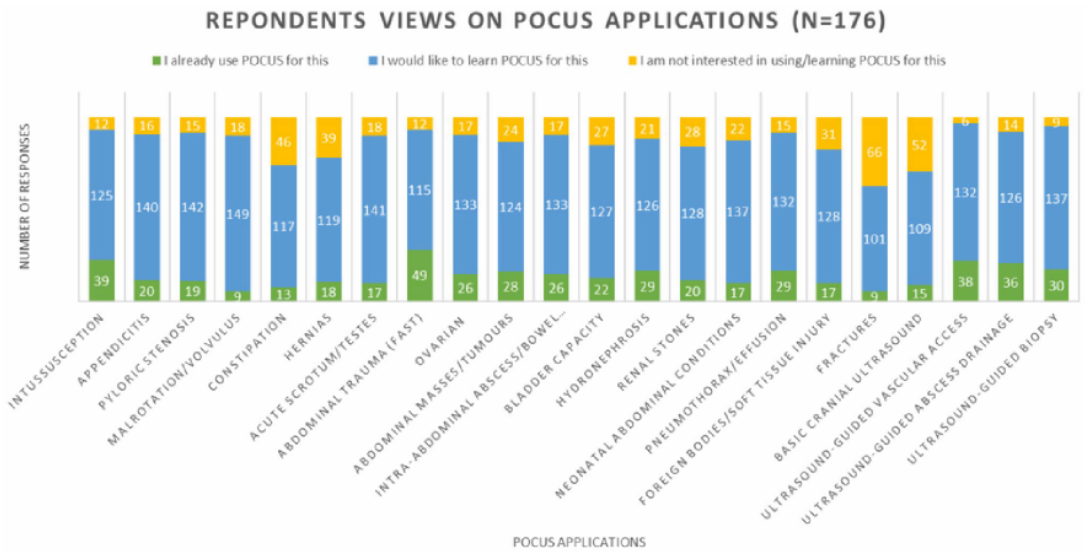



**Fig. 5** Response regarding applications of pocus, evaluating if respondents already used POCUS for this purpose or were interested or not interested in being trained to use POCUS for a particular condition/procedure

## Discussion

Within the pediatric global health community, there is increasing recognition of the magnitude of the burden of surgical disease particularly in LMICs where almost half the population are children [26, 28]. The Lancet Commission on Global Surgery 2015 has been instrumental in defining this problem and highlights issues such as lack of diagnostics as contributory factors, and imploring solutions that are interdisciplinary, innovative, and technology-enabled [28]. Additionally, one of the biggest challenges LMICs face in healthcare provision is the low ratio of doctors per capita compared with HICs [29]. To help address this, the global health community has endorsed 'task shifting' in many clinical areas [30–32]. Given the critical lack of pediatric radiologists in many low-income settings [20], there is a strong rationale for 'task-shifting' some of the most basic ultrasound applications to doctors and surgeons. This is the first study to attempt to delineate global practices and the views of child surgeons on the value of POCUS to children's surgical care.

### Ultrasound use in child surgery

Our findings describe current norms and experiences of child surgeons concerning access to ultrasound services at their institution and the use of POCUS. We demonstrate that ultrasound in pediatric surgical care is critical in all settings, with child surgeons heavily reliant on this mode of imaging in their daily clinical practice (Table 1). Across both HIC and LMIC settings, the primary user of ultrasound (64%) was typically a radiologist or sonographer, as would be traditional. However, in almost a quarter of institutions surveyed, it was the norm for ultrasound to be clinician-led (POCUS) (Table 1). While the reason for this is unclear, it could reflect the general rise in POCUS use across the medical community in recent decades and an evolving status quo [33].

### Barriers

We also evaluated how easy it was for surgeons in their place of work to access traditional ultrasound services, with the majority (93%; 154/167) reporting some difficulty. The main barrier reported was not a lack of devices but rather a lack of ultrasound-skilled personnel (Fig. 4). Secondary to this was the workload pressure on local radiology departments (Fig. 4). Only 10% of respondents could reliably access emergency ultrasound within an hour from the time of request (Table 1). Our findings, therefore, demonstrate that the demand for ultrasound services in child surgery appears to outstrip the supply of trained ultrasound users. In LMIC institutions a significant additional barrier included the cost of ultrasound scans to families. In many institutions access to care requires payment 'upfront' which may render certain investigations inaccessible to poorer families. Equipping surgeons with

POCUS skills to diagnose simple common pediatric conditions could help not only reduce the time to diagnosis and treatment of children but also improve the affordability of care.

### Cultural sensitivity

We asked respondents to describe (in free text) any other issues they experienced with POCUS use in their setting. A HIC respondent described that their radiology department was "unwilling for anyone else to do ultrasound". In some regions (i.e. Europe/North America), such issues have been formally addressed through directives and guidance from Radiology associations that have published their endorsement of POCUS and encourage radiologists to support doctors who which to integrate it into their practice [34, 35]. A further consideration highlighted by our survey is region-specific cultural barriers to clinician-led ultrasound. A respondent working in India described that the regulation of ultrasound was stringent because of concerns over prenatal sex selection [36, 37]. In their setting, it was not advisable for surgeons outside of large training institutes to venture into ultrasound practice for this reason. It is important, therefore, that POCUS initiatives developed for surgeons must be contextually appropriate and sensitive to local culture. Ideally, a collaborative approach should be taken in developing POCUS programs, through proactive engagement with local radiology services and relevant governance bodies [9, 10, 34, 38, 39].

### Training

Our study indicates that most child surgeons (90%;159/176) would engage in formal POCUS training if a program with relevant content was made available. While a considerable proportion of surgeons already have some degree of ultrasound training, most have developed their skills informally. For those with training (formal or informal), the majority continued to practice their skills on a routine basis. However, we found that their perception of competence was associated with how frequently they practiced POCUS. Those who felt competent were more likely to use it regularly compared with those who did not. This finding is in keeping with published literature on POCUS training that emphasizes the importance of longitudinal training programs which over time build both competence and confidence in learners. This can be achieved through mentor-led learning and the use of simulation technology [40–42]. The latter, however, could be inaccessible to LMIC learners due to the large expense of current simulators. To better support LMIC POCUS learners there is a need to develop low-cost simulation technology.

### POCUS Applications

Our evaluation of clinical applications of POCUS found that surgeons who already practice POCUS most commonly use it for trauma (FAST), intussusception, and ultrasound-guided procedures. This is consistent with published reports of the most common protocolized uses of POCUS relevant to our specialty [43–46]. While respondents had high rates of interest in learning all 22 potential applications, the most valued were the diagnosis of malrotation/volvulus, pyloric stenosis, and acute scrotum. The accepted 'gold standard' for diagnosis of malrotation is fluoroscopy, with the role of ultrasound still under debate [47]. The interest shown in learning POCUS to diagnose malrotation (with or without volvulus) may represent a lack of access to fluoroscopy services in many respondents' institutions. However, the evidence for the role of ultrasound is growing. The largest published meta-analysis of the diagnostic accuracy of ultrasound in malrotation/volvulus indicates the superiority of ultrasound which has a sensitivity of 94% (range 89% 97%; 95% CI) and specificity of 100% (range 97%-100%; 95% CI); compared with fluoroscopy sensitivity 91% (range 84%-96%; 95% CI) and specificity 94% (range 72% -99%; 95% CI) [47]. However, it does not account for the individual ultrasound user's accrued experience and expertise which could impact diagnostic accuracy. The interest in learning POCUS for pyloric stenosis and the acute scrotum may be due to the frequency with which pediatric surgeons manage these conditions, where diagnostic uncertainty may lead to delayed care or negative operative findings. A curriculum that aims to accredit child surgeons with POCUS competencies should aim to distinguish between 'basic' and 'advanced' applications so that they are feasible and safe to teach.

## Limitations

We acknowledge several limitations to this study. As with many surveys, there is likely to be a degree of selection bias given respondents, may be those who already have an interest in POCUS. The survey was circulated in the English language, which could have deterred non-English speakers from responding. However, of the 48 countries represented only 17 of these have English as an official language: with less than half 42% [103/247] of respondents working in an English-speaking country. Due to the low proportional response rate per country (range 1–17 responses per country), we cannot infer country-wide views on POCUS, our data instead likely reflects individual clinician views albeit within the area of child surgery.

## Conclusion

Our findings demonstrate the critical importance of ultrasound as a diagnostic tool in children's surgery globally. Despite this, many child surgeons experience significant barriers to timely access to local ultrasound services for their patients. While approximately half of the respondents had some form of ultrasound training, the continued use of their skills was associated with self-perceived competence. Overall, we found high levels of interest amongst child surgeons in learning POCUS for relevant pediatric surgical applications. Training child surgeons in basic applications of POCUS could serve to improve access to diagnostics and reduce time to definitive care for children. Central to this intervention is linking this skill (POCUS) to the surgical care provider, thereby reinforcing the crucial link between the imaging and clinical findings. Further research is needed to evaluate the best training methods, mentorship, accreditation, and governance mechanisms for child surgeons learning POCUS.

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**Author contribution** GN—conceptualization, survey tool development, methodology, data analysis & write-up. MS—survey tool development, survey dissemination, review & edit of the manuscript. AJ—survey tool development, survey dissemination, review & edit of the manuscript. AH—supervision of the work including review & edit of the manuscript. KL— supervision of the work including survey tool development, survey dissemination, & contribution to the manuscript. JL—conceptualization, survey tool development, methodology, survey dissemination, data analysis, write up, review & edit of manuscript.

**Data availability** No datasets were generated or analysed during the current study.

## Declarations

**Conflict of interest** The authors declare no competing interests.



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## 2.6 Influence of Diet on Bowel Function and Abdominal Symptoms in Children and Adolescents with Hirschsprung Disease - A Multinational Patient-Reported Outcome Survey.

Lindert J, Day H, de Andres Crespo M, Amerstorfer E, Alexander S, Backes M, de Filippo C, Golebiewski A, Midrio P, Mohideen M, Modrzyk A, Lemli A, Rassouli-Kirchmeier R, Pfaff-Jongman M, Staszkiewicz K, Telborn L, Stenström P, Holström K, Kohl M, Curry J, Loukogeorgakis S, Davidson JR. Children. 2024. 11(9):1118.

*children*

MDPI

*Article*

# Influence of Diet on Bowel Function and Abdominal Symptoms in Children and Adolescents with Hirschsprung Disease—A Multinational Patient-Reported Outcome Survey

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**Abstract: Introduction:** This study aimed to understand the influence of diet and nutrition items on gastrointestinal symptoms in patients with Hirschsprung Disease (HD). **Method:** An online questionnaire was created to obtain patient-reported outcomes using the multinational Holistic Care in Hirschsprung Disease Network. This was distributed in Dutch, English, German, Italian, Polish, and Swedish via patient associations. Information on demographics, the extension of disease, current diet, and the influence of food ingredients on bowel function were obtained. **Results:** In total, 563 questionnaires were answered by parents or patients themselves. The length of the aganglionic segment was short in 33%, long in 45%, total colonic aganglionosis (TCA) in 11%, and involved the small intestine in 10%. Overall, 90% reported following a mixed diet, and 31% reported taking

probiotics, with twice as many patients taking probiotics in the TCA group compared to standard HD. Mealtimes and behaviours around eating were affected by 61%, while 77% had established food items that worsened symptoms, and of these, 80% stated that they had worked these items out themselves. A high-fibre diet was followed by 24% and 18% a low-fibre diet. Symptoms were reported, particularly from dairy in 30%, fruits in 39%, pulses in 54%, and sugar in 48%. **Conclusions:** This first multinational survey on diet and bowel function in HD reports an association between certain dietary items with gastrointestinal symptoms. This study can support an improved understanding of the interaction between food items and bowel function in children with HD. We suggest a multidisciplinary approach to balance dietary exclusions and support adequate growth, preventing nutrition deficiencies and enhancing quality of life.

**Keywords:** Hirschsprung disease; nutrition; food items; probiotic use; patient-reported outcome of Hirschsprung disease; bowel functions related to Hirschsprung

## 1. Introduction

Hirschsprung Disease (HD) involves a congenital absence of enteric ganglion cells, resulting in functional distal bowel obstruction. Surgical management involves the resection of the affected aganglionic segment and pull-through of the healthy bowel, for which there are several well-established techniques with similar long-term outcomes [1]. Even after optimal surgical management, many patients may suffer long-term problems with defecation, mainly soiling and constipation, as well as Hirschsprung-associated Enterocolitis (HAEC) [2–5]. These bowel symptoms need to be addressed during follow-up to limit their impact on the quality of life (QoL) for patients and their families [2,4,5]. A recent study of parents of children with HD demonstrated a strong influence of diet on bowel function, with up to 70% of patients reporting diet-related symptoms [3,6]. Furthermore, the latest ERNICA guidelines recommend attention to dietary modifications as an integral part of the postoperative pathway of care [7]. Specialist information on nutrition and bowel function were also among the key items identified by parents [8].

Nutritional intake during childhood determines physical, cognitive, and emotional development and is integral to social interactions. Adequate nutrition is also critical for proper neurodevelopment and the establishment of a fully functional immune system [9]. This is particularly important for patients with HD, where ongoing problems with bowel management can affect the time for regular socialising and lead to increased school absenteeism and poor academic performance [10]. These symptoms undoubtedly affect the QoL of patients and their parents.

In individuals with chronic constipation who do not have HD, medications such as stool softeners are generally the first-line recommendation [11]. Nevertheless, dietary measures, such as increasing fluid intake, adjusting fibre intake and avoiding cow's milk, are common practices in the management of paediatric constipation [12]. These recommendations are often replicated for the treatment of HD. To our knowledge, no international study has been conducted to evaluate whether specific dietary adjustments may affect bowel function in patients with HD.

This study uses patient-reported outcomes to describe the influence of diet on bowel functions and symptoms in children and adults with HD, as reported by patients/carers, using a multilingual, international questionnaire.

## 2. Materials and Methods

### *2.1. Study Design and Design of Online Questionnaire*

An online questionnaire on dietary practice, food items, and bowel function was developed in English by the multinational OASIS Holistic Care in HD Network in preparation for the OASIS Holistic Care in Hirschsprung Disease Summer Symposium 2023. The questionnaire was developed by an international stakeholder group, including surgeons,

physicians, dietitians and patient representatives, in a series of virtual consensus meetings. The questionnaire was translated into Dutch (R R-K, MB), German (EA, JL), Italian (CdF, PM), Polish (AM), and Swedish (LT, PS) by native speakers with a medical background and programmed into REDcap (JD, MC). The survey was disseminated through patient associations via newsletters, internet homepages, and social media groups in Austria, Germany, Italy, Netherlands, Poland, the United Kingdom, and Sweden (AL, SA, CdF, KS, MP-J, JD, MM). The questionnaire was open between July and October 2023, and patients could choose to complete the questionnaire in any language (e.g., anyone could complete it in English, etc.). The full questionnaire is available as Supplementary Material.

### *2.2. Definitions Used in the Questionnaire*

We defined the level of disease as follows for this questionnaire:

Short disease (involving just the rectum), long disease (involving more than the rectum but not the whole colon), total colonic aganglionosis (the whole colon), and small intestine (the whole colon as well as a length of the small bowel). Throughout the manuscript, comparisons are made between patients with and without their colon in situ (i.e., rectosigmoid + extended segment vs. total colonic aganglionosis with/without small intestinal involvement).

The questionnaire included questions on 10 specific food groups to investigate effects on bowel function; specifically, the following 10 items were enquired after: 1, I pass stools more often | 2, My stools are more liquid | 3, I pass stools less often | 4, My stools are harder | 5, I have more problems with soiling (staining in the underwear) | 6, I have more accidents with stools | 7, I have more bloating | 8, I have more cramping pains | 9, I have more issues with flatulence | 10, No symptoms | 11, Other (please specify).

The informants were parents of children with HD or HD patients themselves to obtain patient-reported outcomes (PROs). Information on demographics, disease severity, current diet, and the influence of food components on bowel function was answered by the families directly. Patients who had toilet-trained children and were not managing an enterostomy were asked to complete the Rintala Bowel Function Score (BFS).

### *2.3. Statistical Analysis*

The answers were captured on REDcap and subsequently analysed using SPSS 26.0 and Prism 9.0 (GraphPad).

Data are presented either as percentages ($n$(%)) or as the median with an associated interquartile range (median [IQR]). Appropriate statistical tests were performed for categorical (Chi-square) or continuous (Mann–Whitney U) variables. Patients with incomplete questionnaires were included as long as the disease characteristics were present; therefore, throughout the manuscript, denominators for different aspects of the survey differ.

### *2.4. Ethics*

The study was approved as a multinational survey study by the Ethics Committee of the University of Rostock in July 2023 (July/23: A 2023-0123). Participants answered anonymously, and no identification could be tracked.

## 3. Results

### *3.1. Characteristics of Patients Participating in the OASIS Nutrition Survey*

Overall, 563 patients from seven countries responded to the questionnaire (Table 1). Almost 1/3 of respondents answered in German (Germany/Austria), while just over one quarter answered in Italian. While 70–80% of a typical HD population would be expected to have short-segment disease—these accounted for just 33% of our study cohort—a more severe phenotype was suggested in those who were active in patient support groups. Approximately one in eleven patients managed with an enterostomy.

**Table 1.** Patient demographics (*n* (%) or median [IQR]).

| | Overall | German | English | Italian | Netherlands | Polish | Swedish |
|---|---|---|---|---|---|---|---|
| Number | 563 | 181 | 29 | 145 | 101 | 80 | 27 |
| Age, y Med (IQR) | 6 (3–11) | 6 (3–10) | 5 (2–13) | 7 (3–11) | 10 (4–20) | 4 (2–7) | 6 (3–10) |
| Segment, *n* (%) | | | | | | | |
| *Rectosigmoid* | 188 (33) | 48 (27) | 14 (48) | 66 (46) | 34 (34) | 20 (25) | 6 (22) |
| *Long segment* | 252 (45) | 92 (51) | 9 (31) | 47 (32) | 51 (50) | 38 (48) | 15 (56) |
| *Total colonic* | 63 (11) | 17 (9) | 4 (14) | 47 (32) | 7 (7) | 38 (48) | 3 (11) |
| *Small intestine* | 59 (10) | 23 (13) | 2 (7) | 15 (10) | 9 (9) | 17 (21) | 3 (11) |
| *Not declared* | 1 (0.2) | 1 (0.5) | 0 | 0 | 0 | 0 | 0 |
| Stoma, *n* (%), completed y/*n* | 50/562 (8.9) | 12/181 (6.6) | 1/29 (3.4) | 5/144 (3.5) | 4/101 (3.9) | 23/80 (23.8) | 5/27 (18.5) |
| Toilet-trained (without a stoma, completed y/*n*) | 276/447 (61.7) | 84/150 (56%) | 12/23 (52.2) | 97/126 (76.9) | 44/73 (60.3) | 33/54 (61.1) | 7/21 (33.3) |
| ACE, *n*(%), completed y/*n* | 16/56 (28.6) | 3/180 (1.7) | 2/29 (6.9) | 3/144 (2.1) | 4/101 (3.9) | 2/79 (2.5) | 2/27 (7.4) |
| TAI, *n* (%), completed y/*n* | 129/561 (22.9) | 50/181 (27.6) | 5/29 (17.2) | 21/144 (14.5) | 34/101 (33.6) | 6/80 (7.5) | 13/27 (48.1) |
| Tube-feeding, *n* (%), completed y/*n* | 11/561 (1.9) | 3/181 (1.6) | 0/29 | 2/143 (1.4) | 4/101 (3.9) | 2/80 (2.5) | 0/27 |
| Parenteral feeding, *n* (%), completed y/*n* | 5/561 (0.9) | 0 | 0 | 1/143 (0.7) | 0 | 4/80 (5%) | 0 |

ACE: antegrade continence enema, TAI: transanal irrigation.

Bowel function outcome was assessed in patients who were toilet-trained and did not have an enterostomy ($n = 290$). Table 2 depicts the results with completed scores by segment length. We noted, surprisingly, that in this cohort, patients with shorter segment HD did not have superior functional outcomes when assessed with the Rintala Bowel Function Score (BFS; overall BFS out of 20—colon in situ vs. removed: 15(12–17) vs. 15(13–17); Mann–Whitney U: $p = 0.311$). It is important also to notice that only around half of the patients were assessed with the BFS; shorter segment patients were more likely to not have been toilet trained yet (154/420 (37%) vs. 18/114 (16%); $p < 0.0001$) and longer segment patients were not assessed as they were more likely to have stoma (27/114 (23%) vs. 24/420 (6%); $p < 0.0001$).

### *3.2. Dietary Characteristics Were Assessed in Children over the Age of 2 Years*

#### 3.2.1. Overall Diet

Ninety percent of respondents (449/499) followed a mixed diet, 2% stated they were vegetarian/vegan, and 7% followed a special diet with specific exclusions (for example, dairy, gluten, FODMAP, or sugars; see Table 3). While approximately half of the respondents did not pay specific attention to their dietary fibre intake, patients with a shorter segment were much more likely to opt for a high-fibre diet, while patients with total colonic disease tended to opt for lower fibre intake ($p < 0.0001$ for both).

**Table 2.** Patient-reported bowel function according to the level of Hirschsprung Disease.

| | | Hirschsprung Disease—Colon Partially In Situ | | Hirschsprung Disease—Colon Fully Removed | |
|---|---|---|---|---|---|
| | | **Short Disease (*n* = 107)** | **Long Disease (*n* = 119)** | **TCA (*n* = 33)** | **Small Intestine (*n* = 31)** |
| **Overall Score (Median [IQR])** | | 14 [12–17] | 15 [12–17] | 14 [12–16] | 15 [13–17] |
| **Are you aware of the feeling when you need to pass stool?** | Always | 32 (30%) | 41 (34%) | 17 (52%) | 16 (52%) |
| | Most of the time | 41 (38%) | 40 (34%) | 11 (33%) | 9 (29%) |
| | Often uncertain | 24 (22%) | 30 (25%) | 3 (9%) | 4 (13%) |
| | No awareness | 10 (9%) | 8 (7%) | 2 (6%) | 2 (6%) |
| **Are you able to hold back when you need to pass stools?** | Always | 50 (47%) | 61 (51%) | 15 (45%) | 20 (65%) |
| | Occasional problems, less than once per week | 36 (34%) | 35 (29%) | 12 (36%) | 8 (26%) |
| | Problems holding in stool every week | 12 (11%) | 12 (10%) | 4 (12%) | 2 (6%) |
| | No control over bowels, problems every day | 9 (8%) | 11 (9%) | 2 (6%) | 1 (3%) |
| **How often do you pass stool?** | Less than once every 2 days | 7 (7%) | 8 (7%) | 0 | 0 |
| | Every 2 days | 16 (15%) | 12 (10%) | 0 | 3 (10%) |
| | Once per day | 55 (51%) | 40 (34%) | 1 (3%) | 2 (6%) |
| | Twice per day | 16 (15%) | 31 (26%) | 4 (12%) | 1 (3%) |
| | More than twice per day | 13 (12%) | 28 (24%) | 28 (85%) | 25 (81%) |
| **How often do you have issues with soiling or staining of the underwear?** | Never have issues with soiling | 22 (21%) | 26 (22%) | 6 (18%) | 11 (35%) |
| | Soiling less than once a week, only rarely needing a change in underwear | 43 (40%) | 44 (37%) | 14 (42%) | 10 (32%) |
| | Soiling every week, often requiring a change in underwear | 29 (27%) | 29 (24%) | 7 (21%) | 6 (19%) |
| | Soiling all the time, using protective aids | 13 (12%) | 20 (17%) | 6 (18%) | 4 (13%) |
| **How often do you have accidents with stools in the underwear?** | Never | 45 (42%) | 52 (44%) | 11 (33%) | 21 (68%) |
| | Rarely, less than once per week | 37 (35%) | 44 (37%) | 15 (45%) | 6 (19%) |
| | Weekly, wearing protective aids | 12 (11%) | 10 (8%) | 2 (6%) | 2 (6%) |
| | Daily, wearing protective aids day and night | 13 (12%) | 13 (11%) | 5 (15%) | 2 (6%) |
| **Do you suffer from constipation?** | No | 60 (56%) | 69 (58%) | 29 (88%) | 20 (65%) |
| | Constipation managed with diet | 17 (16%) | 15 (13%) | 2 (6%) | 4 (13%) |
| | Constipation managed with medication | 20 (19%) | 21 (18%) | 2 (6%) | 6 (19%) |
| | Constipation managed with enemas | 10 (9%) | 14 (12%) | 0 | 1 (3%) |
| **What is the social impact of your bowel function?** | No impact on social life | 40 (37%) | 46 (39%) | 6 (18%) | 8 (26%) |
| | Some impact | 48 (45%) | 54 (45%) | 21 (64%) | 16 (52%) |
| | Problems that limit social activities | 12 (11%) | 15 (13%) | 5 (15%) | 2 (6%) |
| | Major social or psychological issues related to bowel function | 7 (7%) | 4 (3%) | 1 (3%) | 5 (16%) |

Overall, 341/560 (61%) of patients/families reported that their HD diagnosis had altered the way they chose to eat, and 433/562 (77%) had identified specific food items or ingredients that affected their symptoms, both of which were more prevalent in patients with TCA and small bowel aganglionosis ($p < 0.001$ for both); 104 (24%) concluded this following a discussion with a healthcare professional, most commonly their surgeon ($n = 72$) or a dietitian ($n = 53$). In total, 80% (345/433) had self-identified problematic foods.

**Table 3.** Dietary modification in patients ≥ 2 years (note that an age filter ≥ 2 years and age/segment blanks were removed).

| | *n* = 499 | Hirschsprung Disease—Colon Partially In Situ | | Hirschsprung Disease—Colon Fully Removed | | |
|---|---|---|---|---|---|---|
| | | **Short Disease (*n* = 167)** | **Long Disease (*n* = 219)** | **TCA (*n* = 62)** | **Small Intestine (*n* = 51)** | ***p*-Value (Chi-Square)** |
| General diet | Mixed diet | 150 (90%) | 198 (90%) | 55 (89%) | 43 (84%) | 0.300 |
| | Pescetarian | 3 (2%) | 1 (0.5%) | 0 | 0 | - |
| | Vegetarian | 2 (1%) | 5 (2%) | 0 | 2 (4%) | - |
| | Vegan | 1 (1%) | 1 (0.5%) | 0 | 0 | - |
| | Special/exclusion | 11 (7%) | 14 (6%) | 7 (11%) | 5 (10%) | 0.239 |
| | Complete tube feeding | 0 | 1 | 0 | 0 | - |
| | *n*/A (nil enteral) | 0 | 0 | 0 | 1 | - |
| Dietary fibre | Deliberately high | 59 (35%) | 66 (30%) | 7 (11%) | 8 (16%) | <0.0001 |
| | Neither | 81 (49%) | 115 (53%) | 32 (52%) | 27 (53%) | |
| | Deliberately low | 27 (16%) | 38 (17%) | 23 (37%) | 16 (31%) | <0.0001 |
| Probiotic use | | 46/187 (25%) | 70/252 (28%) | 33/63 (52%) | 24/60 (40%) | <0.0001 |
| Hirschsprung affects eating and mealtimes | | 94/186 (51%) | 151/252 (60%) | 49/61 (80%) | 46/60 (77%) | <0.0001 |
| Specific ingredients/items affect symptoms | | 115/186 (62%) | 172/252 (68%) | 46/61 (75%) | 49/60 (82%) | 0.008 |

Probiotics were used by 31% of patients (177/562), and the rate of use was higher in patients with total colonic aganglionosis (TCA) and TCA with small intestinal disease (46%) compared to those with part of the colon in situ (26%; $p < 0.0001$); see Table 3. Among patients who knew the composition of probiotics, 64% reported taking multi-strain products such as Omniflora®: *Lactobacillus gasseri* and *Bifidobacterium longum*, VSL3®: *S. thermophilus* BT01, *B. breve* BB02, *B. animalis* subsp. lactis BL03, *B. animalis* subsp. lactis BI04, *L. acidophilus* BA05, *L. plantarum* BP06, *L. paracasei* BP07, *L. delbrueckii* subsp. bulgaricus BD08 or Vivomixx®: * *Streptococcus thermophilus* DSM24731®, *Bifidobacterium breve* DSM24732®, *Bifidobacterium longum* DSM24736®, *Bifidobacterium infantis* DSM24737®, *Lactobacillus acidophilus* DSM24735®, *Lactobacillus plantarum* DSM24730®, *Lactobacillus paracasei* DSM24733®, and *Lactobacillus delbrueckii* ssp. *bulgaricus* DSM24734® DSM24734®. Interestingly, although most patients took multi-strain products, among Italian patients, most patients opted to take *Lactobacillus monoculture* products, such as *L. rhamnosus* and *L. reuteri*.

#### 3.2.2. Food-Related Dietary Symptoms

We asked enclosed questions on these 10 separate food items, and the results are displayed in Table 4. Symptoms were more commonly seen in patients without their colon in situ for all except eggs and high-sugar-containing foods. High-sugar foods were related to decreased stool frequency—(25/474) 5%; harder consistency—(41/474) 9%; increased soiling—(58/474) 12%; and more faecal accidents—(21/474) 4%. The different symptoms relating to respective food items are displayed in Table 4, while the most frequent symptoms associated with the three most problematic foods (pulses, high-sugar foods, and fruit) are shown in Table 5. A comparative overview of the proportion of patients reporting any symptoms according to food items is shown graphically in Figure 1.

**Table 4.** Food items causing gastro-intestinal symptoms in relation to the type of Hirschsprung Disease. Patients who had not tried a particular food item were not included.

| Food Item | Total | Short Disease | Long Disease | TCA | Small Intestine | *p*-Value (Chi-Square) |
|---|---|---|---|---|---|---|
| Dairy (cows milk), *n* (%) | 152/509 (30) | 39/172 (23) | 17/219 (8) | 22/59 (37) | 25/53 (47) | <0.0001 |
| Eggs, *n* (%) | 42/488 (9) | 12/164 (7) | 17/219 (8) | 2/55 (4) | 11/52 (21) | 0.170 |
| Wheat, *n* (%) | 61/511 (12) | 13/171 (8) | 25/226 (11) | 13/37 (23) | 10/57 (18) | 0.0004 |
| Soya, *n* (%) | 29/286 (10) | 3/95 (3) | 13/127 (10) | 9/28 (32) | 4/36 (11) | 0.0043 |
| Pulses (beans, peas, lentils), *n* (%) | 220/409 (54) | 63/141 (45) | 101/188 (54) | 30/45 (67) | 26/35 (74) | 0.0012 |
| Alliums (onions, garlic), *n* (%) | 90/418 (22) | 19/132 (14) | 37/191 (19) | 13/51 (25) | 21/44 (48) | 0.0003 |
| Chilli, *n* (%) | 69/212 (33) | 14/63 (22) | 28/99 (28) | 10/21 (48) | 17/29 (59) | 0.0005 |
| Fruits (Top 3: apples, grapes, bananas), *n* (%) | 208/531 (39) | 54/175 (31) | 92/244 (38) | 32/62 (52) | 30/51 (59) | 0.0001 |
| High-sugar foods, *n* (%) | 227/474 (48) | 69/161 (43) | 104/211 (49) | 29/56 (52) | 25/46 (54) | 0.265 |
| Artificial sweeteners, *n* (%) | 39/209 (19) | 10/70 (14) | 18/90 (20) | 5/22 (23) | 6/27 (22) | 0.0063 |

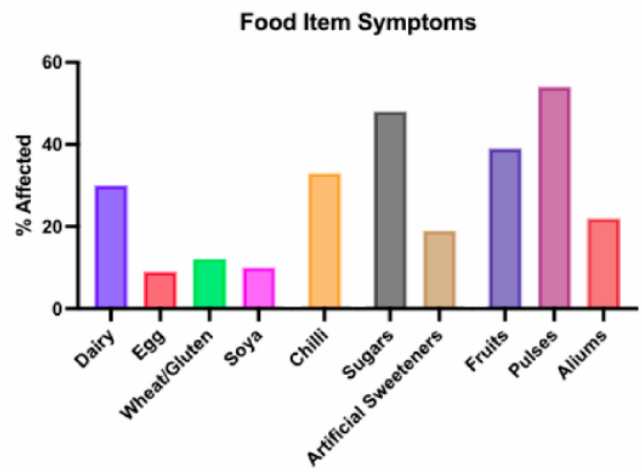


**Figure 1.** Overall onset of adverse bowel symptoms noted by patients and their families according to food items.

**Table 5.** Food items and evoked symptoms.

| Food Item (Number Affected) | Symptom (Top 3) | Prevalence of Symptoms (%) |
|---|---|---|
| Pulses ($n$ = 220) | Flatulence | 52 |
| | Bloating | 50 |
| | Cramping | 30 |
| High-sugar foods ($n$ = 227) | Bloating | 37 |
| | More liquid stools | 34 |
| | Increased frequency | 33 |
| Fruits (apples, grapes, bananas, $n$ = 208) | More liquid | 41 |
| | Increased frequency | 34 |
| | Bloating | 26 |

## 4. Discussion

This multinational questionnaire was answered by over 500 patients ($n$ = 563) from seven European countries: the largest patient-reported study on dietary aspects of gastrointestinal symptoms in HD. Overall, 3 in 5 patients reported that HD affected the items they chose to eat, while three-quarters established food items that worsened symptoms, and, of these, 80% stated that they had worked these items out themselves without any support from healthcare professionals.

This questionnaire addresses an important issue for families, which was specifically highlighted in qualitative focus group discussions, where families stated the key influence that diet has on their child's bowel function [3]. There are currently no dietary recommendations for patients, particularly children, diagnosed with HD. Specific recommendations for those with increased colonic transit include a constipating diet with bulking agents. In general, constipation, or, more likely, outlet obstruction, is reported to affect up to 80% of children with HD [13,14]. Healthy young children would open their bowel once daily with soft stool and usually take a low fibre diet [15].

### *4.1. Functional Outcomes and Impact of Specific Food Items*

The patient cohort was assessed using Rintala BFS to make some objective measures of functional status. Interestingly, by dividing the cohort by age and segment length, we could see that the documented population is not representative of the overall HD population. Specifically, only one-third of patients had rectosigmoid disease (compared to the 80% expected [6]), and the patients with rectosigmoid disease did not have better bowel outcomes than those with longer segments, as has been reported elsewhere [2]. However, in spite of worse-than-expected function in patients with short segments, they experienced a much-reduced impact from the consumption of various food items, took fewer probiotics, and fewer patients declared that HD affected how they chose to eat.

Diet has also been reported by parents to be the dominant influence on their child's bowel function [6]. Among our cohort, gastrointestinal symptoms were most commonly noted in relation to pulses (54%), sugars (48%), fruits (39%), and dairy (30%). Analysing the type of adverse bowel symptoms that were reported, the most common was diarrhoea, which was the most prevalent for dairy (51%) and sugars (36%).

Comparatively fewer patients reported symptoms for egg (9%), wheat/gluten (12%), and soya (10%), which is in line with the results from a previous study in a Swedish setting [6].

Foods high in refined sugar provoked symptoms in 48% of patients compared to 19% for artificial sweeteners. A similar pattern has been shown in patients with inflammatory bowel disease (IBD), where 23% reported "Sugary/sweet food" to worsen their gastrointestinal symptoms, while only 4% specified artificial sweeteners to trigger their symptoms [16,17]. However, research into the effect of artificial sweeteners on gastrointestinal symptomatology is scarce and has produced conflicting results [17,18]. Studies in irritable bowel syndrome and inflammatory bowel disease suggest an impact secondary

to changes in gastrointestinal motility and intestinal microbiota, especially induced by polyols [17,18].

Pulses were the food item most likely to cause gastrointestinal symptoms, mainly due to bloating and flatulence. Foods rich in fermentable residues, such as pulses, have also been suggested to increase intestinal gas production in healthy children [19]. However, in a study on healthy children, only 24% reported any gastrointestinal symptoms [19] compared to over 50% of children with HD in this study—suggesting an additional disease-specific effect. Previous research has shown that fruits, foods high in fat content, grains including bread and cereal, vegetables, and dairy are commonly excluded to reduce gastrointestinal symptoms [20]. Speculatively, the difference in symptom generation may be related to differences in intestinal microbiota or motility reactions to food between healthy and HD patients [21–23].

A low Fermentable Oligosaccharides, Disaccharides, Monosaccharides, and Polyols (FODMAPs) diet has gained popularity for the treatment of functional abdominal pain and other functional abdominal symptoms, including bloating. This diet results in a reduction in fermentable carbohydrates (FODMAPs). Many of the foods excluded by the participants in this study were high in FODMAPs, including wheat, legumes, dairy products, fruits, alliums, and some artificial sweeteners. Wheat, legumes, and alliums are classified as fructans and galacto-oligosaccharies (GOSs). These FODMAPs are fermentable by the microbiota in the colon [24,25] and, due to the limited available colon in HD, may further exacerbate symptoms. A diet high in FODMAPs may be important to consider in patients with ileostomies, as it has been shown to increase fluid output and decrease absorption [26].

The reported findings of interaction between food items and bowel function can help in counselling families to ameliorate adverse dietary symptoms; however, we believe that appropriate and supervised exclusion should be considered to enable varied nutritional intake to encourage appropriate growth and development. As there are no guidelines for specific dietary advice for people with HD, the results show that many patients and carers may modify their diet in response to symptoms perceived to be associated with specific foods. Reducing fructose and lactose in patients with HD has previously been shown to reduce symptoms of faecal incontinence [23]. Similar results were seen in our study's families, who reported reducing intake because of loose stools.

Overall, this work suggests that there are clear food-related symptoms that occur in a large percentage of patients with HD. This raises the possibility that soiling and accidents that are noted in children may be, in fact, related to dietary intake and may trigger investigations post-pull-through, such as GI endoscopy and rectal biopsy. Dietary changes should be made under the guidance of healthcare professionals, such as dietitians, who can make personalised recommendations based on an individual's specific needs and symptoms. Each person with HD may have a unique response to foods, so tailoring dietary changes is essential to optimise symptom management and overall health. Symptoms and dietary changes have been shown to increase stress and feelings of exhaustion [3]. Research in this area may provide further insight into effective dietary strategies for people with HD, which may help patients feel more supported in their condition.

### *4.2. Microbiome and Probiotics*

In addition to dietary impacts on symptoms, this study also surveyed the use of probiotics taken by patients. In this study, 35% of the respondents used probiotics with Bifidobacteria and Lactobacillus. We further noted that patients with total colonic HD were twice as likely to use probiotics compared to patients who still had part of their colon in situ. The microbiome plays a crucial role in digestion, and it is influenced by geographical food composition and altered by the removal of a certain length of the colon or entire colon and perioperative and intermittent antibiotic use. Significant differences in the gut microbiota between healthy individuals and children with functional constipation have been described [27]. The microbiome is potentially linked to intestinal motility. Further research showed that probiotics not only significantly diminished the incidence but also

decreased the severity of HAEC [28]. However, the mechanisms by which pre-, pro-, and symbiotics may reduce inflammation and their particular role in HD are not fully understood [28–30].

### *4.3. Differences Due to the Length of Hirschpsrung Disease Segment*

We note, in this study, that patients with short-segment disease reported symptoms from specific food items less frequently than those with TCA. This was the case for all food items surveyed, with the exception of egg (which was generally low across all segment lengths) and refined sugars (around 50% across different levels of disease). Also, patients with TCA reported a higher rate of impact on their social life for the BFS, which may, in part, be due to the significantly higher dietary impact reported here. Knowledge of the resected part and remaining bowel is crucial for the medical team to anticipate bowel function and best advise parents. Often, the level of disease is judged by the removed segment of the bowel, although we suggest to rather focus on the remaining segments, which determine the function. Unfortunately, there is not yet a universally agreed consensus on categories for the length of the HD segment, and thus, a comparison remains challenging.

### *4.4. Holistic Nutrition Counselling and Specialist Dietician Contribution*

In this patient-reported study, we demonstrate that in the majority of instances, patients and families had established the effects of food items themselves, often making changes and exclusions to their diet in the absence of any professional supervision. Our research highlights foods that potentially exacerbate or lead to negative bowel function symptoms; however, clearly, it cannot be determined whether exclusion could alleviate these symptoms and to what extent. We also need to carefully consider the nutritional needs of the growing child, so dietary changes, particularly exclusions, would benefit from appropriate assessment by dieticians. The role of a dietitian in the multidisciplinary Hirschsprung Disease team may include nutritional advice as well as the clinical observation and monitoring of dietary changes; see Figure 2.

| |
|---|
| Anthropometry monitoring. |
| Review of biochemical markers related to nutrition (electrolytes, iron, Vit B12, albumin, bilirubin, and trace elements + fat-soluble vitamins). |
| Clinical information on bowel function is reported and standardised using a bowel function assessment tool. |
| Detailed nutritional review, including a 24 h diet recall, restricted or excluded foods, and any nutritional supplements given. |
| Support families to achieve participation in age-related activities (kindergarden/school meals and school trips). |

**Figure 2.** Checklist role of dietician.

Malnutrition has been recognised in children with Hirschsprung Disease, and this was particularly the case if surgery was performed after the age of 3 years [8]. At present, we cannot reliably attribute poor nutritional status to failure to thrive due to pre-existing Hirschsprung's Disease or to dietary factors.

### *4.5. Strengths and Limitations*

This is the largest cohort of patient-reported data on diet and bowel function in patients with HD. Information was collected across different age groups from seven European countries with a heterogeneous cultural dietary profile. Patient representatives were involved throughout, which has been shown to improve the quality and relevance of research [31]. We did not collect any information about our participants' medical history (i.e., reasons for stoma formation, the timing of pull-through) and solely relied on patient/caregiver-declared information (i.e., segment length). We were not able to compare

different pull-through techniques. Our study is further limited by two distinct biases: firstly, outreach through patient associations may only reach involved families—potentially emphasising an access bias known to exist across demographic differences. Secondly, we anticipate a reporting bias from those patients with more adverse symptoms (themselves more likely to be involved in patient support organisations); this is suggested by the under-representation of patients with short-segment disease and comparatively worse BFS scores in these patients than might be expected.

## 5. Conclusions

This multinational survey provides important insights into the experience of food-related clinical symptoms in patients with HD. Eggs, wheat, and soya were not reported to evoke adverse symptoms more than expected, while pulses, sugars, and fruits seem to result in adverse bowel functions in a large proportion of patients. Overall, the majority of responders reported that their diet influences bowel function and tried to limit this impact by selecting or avoiding specific food items on their own. Food counselling, involving dietitians to ensure appropriate dietary intake to support development and growth, should be delivered with the awareness that there is still very limited evidence highlighting specific foods and their impact on bowel function in patients with HD.

**Supplementary Materials:** The following supporting information can be downloaded at https://www.mdpi.com/article/10.3390/children11091118/s1. Table S1: Nutrition questionnaire, English version.

**Author Contributions:** Conceptualisation, J.L., H.D. and J.R.D.; methodology, J.L. and J.R.D.; software, J.R.D. and M.d.A.C.; validation, J.L., J.R.D., E.A., M.B., P.M., R.R.-K. and A.M.; formal analysis, J.R.D.; investigation, J.L., H.D., M.d.A.C., J.R.D., E.A., M.B., S.A., A.G., M.P.-J., K.S., M.M., K.H., C.d.F., R.R.-K., A.L. and K.S.; resources, J.L. and J.R.D.; data curation, J.R.D.; writing—original draft preparation, J.L., H.D. and J.R.D.; writing—review and editing, L.T., M.d.A.C., P.S., S.A., A.L., R.R.-K., M.K., S.L. and J.C.; visualisation, M.d.A.C. and J.R.D.; project administration, J.L. and J.R.D.; funding acquisition, J.L. All authors have read and agreed to the published version of the manuscript.

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**Institutional Review Board Statement:** The study was conducted in accordance with the Declaration of Helsinki and approved by the Ethics Committee of University Rostock (A 2023-0123 on 7 June 2023).

**Informed Consent Statement:** Implied consent to submit anonymised information on bowel function and diet was obtained from all subjects involved in this study.

**Data Availability Statement:** The data presented in this study are available on request from the corresponding authors. The data are recorded in an anonymous Redcap Database due to privacy or ethical restrictions.

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## Abbreviations

| | |
|---|---|
| ACE | Antegrade continence enema |
| BFS | Bowel Function Score |
| FODMAP | Fermentable Oligosaccharides, Disaccharides, Monosaccharides and Polyols |
| HD | Hirschsprung Disease |
| IBD | Inflammatory bowel disease |

| | |
|---|---|
| QoL | Quality of life |
| TAI | Transanal irrigation |
| TCA | Total colonic aganglionosis |

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## 2.7 OASIS (Holistic Care in Hirschsprung Disease) Network Position Paper: Bowel Function Score in Longterm Follow-up for Children with Hirschsprung Disease.

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*children*

MDPI

*Article*

# Bowel Function Score in Long-Term Follow-Up for Children with Hirschsprung Disease: OASIS-Holistic Care in Hirschsprung Disease Network Position Paper

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**Abstract:** Introduction: The assessment of bowel function in patients with Hirschsprung disease (HD) remains controversial, as several different bowel function scores are used in the literature and are therefore not suitable for reliable comparison. Conference Section: The OASIS-Holistic Care in Hirschsprung Disease Network Group addressed this issue and evaluated the most commonly used scores for their utility in HD patients. Scoping Review and Expert Group Consensus were performed. It was agreed that the ideal score for patients with Hirschsprung disease should be a comprehensive, validated score that allows reliable assessment of bowel function and continence, depicts changes according to management and natural history, and quantifies baseline parameters. This score should also enable comparisons of bowel function in patients born with HD worldwide. Concluding Remarks: The OASIS-Holistic Care in Hirschsprung Disease Network Group agreed that this ideal score does not yet exist and is therefore a future goal of the group. Meanwhile, clinicians should use a score for comparable, standardized, objective assessment, and the Rintala Score is suggested. Future developments may also lead to easily accessible patient-reported outcome measures and semi-automated dashboards that allow automated data extraction from electronic health records.

**Keywords:** Hirschsprung disease; continence score; scoring postoperative results; incontinence; bowel function; patient-reported outcome measurements (PROM)



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## 1. Introduction

Hirschsprung disease (HD) affects approximately 1 in 5000 live births, of whom 80–85% have HD confined to the rectosigmoid and 20–25% of patients have HD extending upwards to varying lengths [1]. HD is characterized by the absence of enteric ganglion cells in the affected bowel, resulting in chronic functional bowel obstruction. Most children are diagnosed with HD within their first year of life [1]. Corrective surgical strategies aim to resect the abnormally innervated bowel and connect the healthy, adequately innervated bowel to the distal rectum without leaving the affected bowel in situ [1–3].

Despite technically sound reconstruction in experienced hands, long-term functional impairment is common after pull-through surgery [3–6]. Bowel function can deteriorate significantly at any age, and longitudinal monitoring is recommended. Impaired bowel function is known to alter psychosocial functioning and quality of life (QoL), announcing

the need for dedicated bowel management to mitigate these sequelae and enable children to be continent [7–9]. Therefore, highly specialized follow-up is required, and eventually, transition to adult services is recommended [1,10,11].

Besides increasing awareness of the need for a long-term follow-up of HD, there is no agreement on a standardized way of evaluating bowel function in the long-term follow-up, yet. The abundance of different classifications and scoring systems creates challenges in comparing the functional results in the literature [12].

A total of 84 different questionnaires exist to assess bowel function in colorectal diseases. Only 8 of them are validated, 15 are established and broadly used, and 61 are self-designed [13]. These questionnaires are mostly developed and mainly intended to be used in patients with an anorectal malformation (ARM) [12].

Although HD and ARM are two different entities with different reconstructive surgical procedures, these two pediatric colorectal conditions are usually combined in analyses of bowel function utilizing continence scores and bowel function outcome measurements [12].

Interestingly, the ERNICA (European Reference Network for rare Inherited and Congenital (digestive and gastrointestinal) Anomalies) guidelines on rectosigmoid HD do not discuss the use of bowel function scores nor does it recommend a certain score that should be used to assess bowel function [1].

There is currently no consensus on the most effective way to evaluate bowel function in long-term follow-up. Multiple bowel function scoring systems exist, yet their applicability and consistency in HD are varied. The purpose of this article is to provide an overview of appropriate tools for monitoring bowel function in patients with HD and to highlight the holistic, patient-centered care practices advocated by the OASIS-Holistic Care in Hirschsprung Disease Network Group.

## 2. Conference Section

The OASIS-Holistic Care in Hirschsprung Disease Network Group prepared a session dedicated to "Continence Scores" to discuss this topic at the international "OASIS-Holistic Care in Hirschsprung Disease Symposium" in Warnemünde, Germany, in July 2023.

The working group "Continence Score of the OASIS-Holistic Care in Hirschsprung Disease Network Group" prepared the content of the session and discussion, which is reflected in this manuscript. The information was obtained through a scoping review of bowel function scores and other colorectal scores, which was presented and discussed in the session dedicated to Continence Scores. This session also included and further evaluated the perspectives of patient associations and affected families. A total of 98 participants from 11 countries shared experiences and discussed holistic aspects of the care of Hirschsprung disease. Attendees came from Germany, Netherlands, Poland, Italy, United Kingdom, Austria, Spain, Sweden, Norway, Switzerland, and France. A total of 85 pediatric surgeons, 8 representatives from patient associations, and 15 researchers (some of them clinicians as well) were present. A model for real-time data collection and assessment of standardized data outcomes using a pediatric bowel function and patient quality of life dashboard was presented [14]. Finally, overall considerations for achieving consensus among pediatric colorectal surgeons were discussed. The scores were discussed, and the opinions of patient representatives and patients were heard. A voting system was used, and when the majority agreed, the view was accepted for this statement.

### *2.1. Overview Bowel Function Assessment Tools Used in Hirschsprung Disease*

The OASIS-Holistic Care in Hirschsprung Disease Network Group evaluated the following 8 bowel function scores during the Continence Score session for the applicability in HD during the Network meeting (Table 1). The following 8 scores were selected by the expert group preparing session after a review of the available scores.

**Table 1.** Bowel function scores commonly used in Hirschsprung disease.

| | Holschneider | Rintala | Wingspread | Krickenbeck | Baylor Continence Scale | Fecal Incontinence Index (FII) | Pediatric Incontinence and Constipation Score | Groningen Pediatric Defecation and Fecal Continence Questionnaire | Bowel Diary | ARM-Net |
|---|---|---|---|---|---|---|---|---|---|---|
| First used | | 1995 | 1986 | 2005 | 2007 | | | | | |
| Filled out by whom | parents | parents | | parents | patients | patients | | | parents | clinician |
| Validated | | | | | yes | yes Brazil | | | | no |
| Number of items | 7 | 7 | 4 | 3 | 23 | 8 | 13 | 75–88 | 4 | |
| Includes type of ARM | no | no | | no | | | | | no | |
| Includes type of Hirschsprung | no | no | no | no | no | no | no | no | no | |
| Incontinence | yes | yes | yes | yes | yes | yes | | yes | Yes | yes |
| Constipation | yes | yes | yes | yes | yes | yes | | yes | Yes | yes |
| Urinary problems | no | no | no | | yes | | | yes | no | yes |
| Anal sensation | yes | no | no | no | | | | yes | no | |
| Social problems | no | yes | no | no | yes | | | yes | no | |
| Includes results with bowel management | | | yes | yes | | | | yes | yes | yes |

### 2.2. Criteria for Bowel Function Assessment Tools Applicable in Patients with HD

A bowel function assessment tool for HD needs to address the following items:

- *Quantify baseline variables*

  Objective quantification may allow different team members to picture the situation and share their professional assessment [14,15].

- *Track changes over time*

  Bowel function and continence physiology change over the normal childhood developmental phases, which need to be reflected during assessment [16,17].

- *Validity*

  A validated assessment tool should ideally be used. Validation is conducted in each language the tool is used and should include culturally sensitive translation and a validity concept that refers to the comprehensibility and relevance of items [15,18].

- *Reliability*

  The tool should reflect the real bowel function, as experienced by the patients and their families. Reliability is given when the score produces the same or similar results in patients when repeatedly administered [14].

- *Assess continence with and without bowel management*

  Many children with HD require bowel management and may achieve social cleanness [8,16]. Bowel management approaches vary according to age and also clinician and family preference; hence, a variety of suppositories, enemas, rectal washouts, and transanal irrigation are used to relieve symptoms. The ideal bowel function assessment tool should incorporate the reality achieved with bowel management. It should also assess the degree of fecal incontinence by distinguishing between mild soiling and more severe fecal incontinence. It should differentiate between the different causes of soiling and/or fecal incontinence, such as critical reduction in sphincter tone, reflex incontinence due to an injured or removed anal canal, and outlet obstruction causing overflow incontinence (pseudo incontinence). Clinicians are interested in understanding both the real baseline bowel function to support and individualize the bowel management concept, with the ultimate aim of achieving continence or social continence in the latter case, as well as the achieved real-life continence [16,17]. It is of equal importance to assess the bowel function with bowel management as well. Thus, a score for patients with HD should provide insight into the patient's bowel function and enable changes to be tracked with bowel management.

- *Specific groups/comorbidity*

  Common comorbidities in HD patients, such as trisomy 21, are associated with neurodevelopmental delay and other co-factors, e.g., muscular hypotonia, which may influence the ability to achieve continence as well.
  The criteria for bowel function assessment were established by the group of authors as the minimal requirement for a clinical score, following a review of existing scoring systems. The group reached full consensus.

### 2.3. OASIS Group Recommendation to Use the Rintala Bowel Function Score

The OASIS group recommends adopting the Rintala Bowel Function Score as the standard tool for assessing bowel function in HD. This score is widely recognized and consists of seven items that assess key domains such as defecation frequency, incontinence, constipation, and social problems. Its brevity and comprehensive nature make it a practical tool for clinical use. Importantly, it has been validated in Scandinavian countries and has been used extensively to evaluate both HD and anorectal malformation (ARM).

### 2.4. Real-Time Data Collection

The peculiarity of the assessment of intestinal function is characterized by the physiological changes with normal child development. A retrospective evaluation of electronic health records often lacks information, and the existing records are often subjective.

Using a standardized score may improve the comparability of the reported bowel function outcome. Combined with automated extraction from electronic records, monitoring may become easier. However, the information input relies on the meticulous dedication of a healthcare professional who is documenting a bowel function score [14].

Therefore, patient-reported outcome measures (PROMs) empower the patient and overcome the clinician factor. A semi-automated dashboard has the further potential of enabling automated data extraction from electronic health records [14].

## 3. Concluding Remarks

The OASIS group acknowledges the availability of a variety of bowel function scores, including both validated and non-validated scores. The participants agree that to compare results, a bowel function score should be used as a reference. The group points out that currently, the available literature on long-term outcomes is difficult to interpret due to the heterogeneity of the scores used. Clinicians reflect that it is challenging to perform bowel function assessments due to limited time during outpatient clinic appointments. Clinicians and patient representatives are also unsure if patients provide consistent answers when questioned by their surgeons.

Standardized bowel function questionnaires have been developed to provide a holistic view of patients' bowel function and, at the same time, predict patient outcomes. Validated scores allow for objective comparison between patient groups and their longitudinal follow-up.

Considering the abundance of mostly self-designed scores used in pediatric colorectal disease, we question the need for a specific score for patients with Hirschsprung disease (HD). In the literature, patients with anorectal malformation (ARM) and HD are often pooled and discussed together [12], despite representing two distinct entities with disease-related specificities. A core set for the assessment of urinary and fecal incontinence was under development at the Zürich University of Applied Sciences, Switzerland, the ICF Incontinence Assessment Form (ICF-IAF), but it has not been completed yet. Unfortunately, it could not be brought to completion because of a lack of funding [19–21].

Children with HD, per pathophysiology, have an intact continence organ but may suffer from persistent outlet obstruction and overflow pseudo incontinence, which may present with soiling that may be misdiagnosed as incontinence [16]. Indeed, this outlet obstruction needs to be adequately addressed in the functional assessment and follow-up of patients with HD, ideally reflected in an HD-tailored continence score. However, it should be noted that surgical damage to the sphincters or squamous epithelium of the anal canal can also cause chronic fecal incontinence [4,16,22].

According to the OASIS-Holistic Care in Hirschsprung Disease Network Group, a bowel function score that can be used in patients with HD should quantify baseline variables, allow changes to be tracked over time, be valid and reliable, and assess continence with and without bowel management. To ensure compliance, the ideal score should include the above key features, but remain short and comprehensive.

Using PROMs may further help to understand outcomes that matter to our patients [22]. This is echoed by patient representatives at the OASIS-Holistic Care in Hirschsprung Disease Symposium, who stated that it is ultimately more important to patients that they experience a deviation from their normal peers than to be informed of their actual score.

Another general tool that can be used in follow-up to obtain a broad overview of bowel habits is a bowel diary [14]. By self-documenting daily bowel habits, including information on medications such as laxatives or stool softeners, as well as describing stool consistency according to the Bristol score, stool frequency, and the incidence of soiling or fecal accidents over a two-week period, the treating physician can obtain important information to guide

treatment and also evaluate treatment effects in an objective way [15,23,24]. However, a bowel diary cannot be used as a tool to compare patient groups and can therefore only be used as a patient-centered diagnostic tool for care and follow-up. Longitudinal follow-up is important because bowel function can change throughout life [5].

Ideally, restorative surgery is performed in infancy when patients' bowel function can be compared with that of healthy peers, and children are not yet physiologically toilet trained [1,2]. There is, of course, inter-individual variation in the timing of toilet training. In general, up to 53% of patients with HD have obstructive symptoms, up to 37% have Hirschsprung-associated enterocolitis (HAEC), and up to 45% have fecal incontinence symptoms that can change over time [3,5,22,25,26]. Monitoring bowel function over time allows the clinician to understand the evolution of bowel problems associated with HD and may also serve as a motivator for the patient and family [14]. As mentioned in the literature, it is commonly observed that the pathognomonic persistent outlet obstruction usually disappears once the patient reaches adolescence. Systematic monitoring using a consistent set of measurements can help to improve our understanding of bowel function in HD patients in relation to disease severity, surgical technique, and patient age, which will ultimately enable clinicians to manage and counsel their HD patients and parents in a disease-tailored manner in the future [5,6,27].

As there is no consensus among pediatric surgeons on scores to assess bowel function, a large number of different scores can be found in the literature. According to the aforementioned criteria for a bowel function assessment tool applicable to patients with HD, the bowel function score established by Rintala and Lindahl [28] is the most commonly used continence score. After an extensive discussion of the different scores described in the literature, the "Continence Score of the OASIS-Holistic Care in Hirschsprung Disease Network Group" considered Rintala and Lindahl's bowel function score to be the most appropriate to assess bowel function in patients with HD. It is an established 7-item scoring system with domains of sensation and urge to defecate, ability to hold defecation, frequency of defecation, fecal continence, constipation, and social problems related to bowel function, with a maximum score of 20. It was originally developed for children with ARM and compared with an age-matched healthy population in Finland [28–30]. Since then, it has also been used worldwide to assess bowel function in patients with HD and has become the most popular and widely used bowel function score for HD in the last decade [3,5,10,27,31–64].

Scores have consistently correlated closely with clinical outcomes, and it is therefore considered a validated score in Scandinavian countries [28,33,35]. However, despite its worldwide use to assess bowel function in patients with ARM and HD, its degree of validation is not entirely clear, as cultural adaptation and validation concepts have not been applied elsewhere. The advantage of this questionnaire is that it is short, comprehensive, and observer-independent, as it can be completed by the child or their parents without the need for a clinical examination.

Consistent use of a single score will ultimately lead to a reliable comparison of bowel function obtained from HD patient groups. In addition, consistent use of the same score will allow assessment of bowel function over time as individuals are followed, which will allow adaptation and optimization of treatment and thus contribute to the associated quality of life. Longitudinal monitoring is particularly important in patients with HD, as the bowel function is known to change during growth; up to 53% of HD patients experience obstructive symptoms despite corrective surgery, which has been described as a cause of Hirschsprung-associated enterocolitis (HAEC), which has been reported to occur in up to 37% of patients post-operatively, while 45% of patients even present with fecal incontinence [3,5,22,25,26]. Recent results have shown that bowel function does not necessarily improve over time and that adolescent and adult HD patients still have to cope with problems such as soiling, constipation, or even incontinence [6,65].

Therefore, targeted and evidence-based follow-up is paramount to assess bowel function and meet the long-term care needs of this patient cohort, which ultimately contributes to each patient's quality of life.

Future developments will undoubtedly lead to PROMs that can be implemented in a structured long-term follow-up via apps where patients can report directly on their bowel function. This could be introduced as an automatic reminder, e.g., during the transition process. Self-reported outcome parameters have the advantage of being unbiased and reflecting the actual perception of the respondent. With the use of technology and artificial intelligence, this may be the optimal assessment tool to obtain a bowel function score in the future.

*Limitation*

Our work contributes to the ongoing discussion on the most effective ways to evaluate and report outcomes in pediatric surgery, not only for assessing the current situation but also for gathering evidence by pooling patient data. However, this manuscript is limited to describing specific items and discussion points and reflects the collective opinion of the author group.

## 4. Key Message by OASIS-Holistic Care in Hirschsprung Group

The implementation of a standardized tool like the Bowel Function and Continence Scoring Questionnaire across clinicians treating patients with Hirschsprung disease (HD) is crucial for several reasons:

1. Consistency in Reporting: Uniform use of a single tool will lead to standardized reporting of bowel function outcomes, ensuring that patient data are comparable across different centers and clinical practices. This can enhance the quality of research and patient care, enabling the identification of trends and best practices.
2. Longitudinal Monitoring: Regular use of a scoring system will allow for continuous monitoring of each patient's bowel function over time. Clinicians will be able to detect patterns of improvement, stabilization, or deterioration in symptoms as the patient grows, facilitating more personalized and proactive management strategies.
3. Integration with Patient-Reported Outcome Measures (PROMs): Incorporating the scoring tool alongside other PROMs enables clinicians to have a more holistic view of the patient's experience. Patients and caregivers can contribute their perspectives on symptoms and quality of life, giving clinicians a fuller picture of the effectiveness of treatments.
4. Guiding Care and Follow-up: By consistently using these tools, clinicians will have objective data to guide decision-making in care pathways. Bowel function and continence scores can inform the need for interventions, adjustments in treatment plans, or further assessments, leading to more targeted and effective management.
   The focus on improving bowel function-related symptoms, especially as patients progress through growth milestones, will be essential for optimizing long-term outcomes in HD management.

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## Abbreviations

| | |
|---|---|
| ARM | Anorectal malformation |
| ERNICA | European Reference Network for rare Inherited and Congenital (digestive and gastrointestinal) Anomalies |
| HAEC | Hirschsprung-associated enterocolitis |
| HD | Hirschsprung disease |
| ICF | International Classification of Functioning, Disability and Health |
| PROM | Patient-reported outcome measures |
| QoL | Quality of life |

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# 3 Publikationen Judith Lindert

| | **Originalarbeiten mit Impact Faktor** | |
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| | | **Impact Faktor** |
| O25 | **Lindert J,** Hoel AT, Schmiedecke E, Curry J, Loukogeorgakis S, Amerstorfer E. OASIS (Holistic Care in Hirschsprung Disease) Network Position Paper: Bowel Function Score in Longterm Follow-up for Children with Hirschsprung Disease. Children. 2024. 11(11):1284. | **2,6** |
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| O22 | **Lindert J**, Day H, de Andres Crespo M, Amerstorfer E, Alexander S, Backes M, de Filippo C, Golebiewski A, Midrio P, Mohideen M, Modrzyk A, Lemli A, Rassouli-Kirchmeier R, Pfaff-Jongman M, Staszkiewicz K, Telborn L, Stenström P, Holström K, Kohl M, Curry J, Loukogeorgakis S, Davidson JR. Influence of Diet on Bowel Function and Abdominal Symptoms in Children and Adolescents with Hirschsprung Disease - A Multinational Patient-Reported Outcome Survey. Children. 2024. 11(9):1118. | **2,0** |
| O21 | Märzheuser S, Schulze F, **Lindert J**. Surgical Strategies in Total Colonic Aganglionosis: Primary Pullthrough-Pathway of Care. Children. 2024.11(8):911. | **2,0** |
| O20 | **Lindert J**, Erkel D, Schulze F, Hofer M, Rzepka E, Märzheuser S. Is the Transrectal Diameter (TRD) Suitable for Assessing Faecal Loads and Monitoring Bowel Management in Children with Hirschsprung Disease - ReKiSo Study: Prospective Study. Children. 2024. 11(8):921. | **2,0** |
| O19 | Erkel D, Märzheuser S, **Lindert J**. Assessing fecal load with ultrasound in children with colorectal pathology: ReKiSo study. Paediatric Surgery International. 2024. 40(1):202. | **1,8** |
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| O16 | **Lindert J**, Straube T, Larsen B, Siebert J, Liodaki E, Tafazzoli-Lari K, Wünsch L. An Optical Tomography-Based Score to Assess Pediatric Hand Burns. Eur BurnJ. 2024. 5:155–165. | **1,0** |
| O15 | Bbaale D, Mohr C, **Lindert J**, Allorto N, Mabanza T, Katabogama JB, Chamania S, Elrod R, Boettcher M, Elrod J. Barriers and prospects for skin grafting in burn treatment across African countries. Burns. 2024. 50(5):1150-1159. | **3,2** |
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| O12 | **Lindert J**, Bbaale D, Mohr C, Chamania S, Bandyopadhyay S, Boettcher J, Katabogama JB, Alliance BW, Elrod J. State of burns management in Africa: challenges and solutions - A survey of burn care in Africa. Burns. 2023. 49(5):1028-1038. | **2,7** |
| O11 | **Lindert J**, Sharma D, Cotton M. Minding the Ps and Qs in global surgery outreach programmes. Tropical Doctor. 2023. 53(1):1-3. | **0,4** |
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| O5 | **Lindert J**, Tafazzoli-Lari K, Tüshaus L, Larsen B, Bacia A, Bouteleux M, Adler T, Dalicho V, Vasileidos V, Kisch T, Stang F, Welzel J, Wünsch L. Optical coherence tomography provides an optical biopsy of burn wounds in children-a pilot study. J Biomed Opt. 2018. (10):1-6. | **3,6** |
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| K17 | Hageman IC, Trajanovska M, King SK, van der Steeg HJ, Morandi A, Amerstorfer EE, de Blaauw I, van Rooij IA; **ARM-Net Consortium.** Anorectal Malformation Patients in Australia and Europe: Different Location, Same Problem? A Retrospective Comparative Registry-Based Study. J Pediatr Surg. 2024. 59(12):161879. | **2,4** |
| K16 | National Institute for Health and Care Research (NIHR) Global Health Research Unit on Global Surgery. Global access to technologies to support safe and effective inguinal hernia surgery: prospective, international cohort study. Br J Surg. 2024. 111(7): 164. | **8,6** |
| K15 | NIHR Global Health Research Unit on Global Surgery, **STARSurg Collaborative**. A prognostic model for use before elective surgery to estimate the risk of postoperative pulmonary complications (GSU-Pulmonary Score): a development and validation study in three international cohorts. Lancet Digit Health. 2024. 6(7):e507-e519. | **23,8** |
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| K10 | GlobalSurg Collaborative and NIHR Global Health Research Unit on Global Surgery. Effects of hospital facilities on patient outcomes after cancer surgery: an international, prospective, observational study. Lancet Glob Health. 2022. 10(7):e1003-e1011. | **19,9** |
| K9 | Global Health Research Group on Children's Non-Communicable Diseases Collaborative**.** Impact of the COVID-19 pandemic on patients with paediatric cancer in low-income, middle-income and high-income countries: a multicentre, international, observational cohort study<br>BMJ OPEN. 2022. 12(4):e054690. | **2,9** |
| K8 | COVIDSurg Collaborative, GlobalSurg Collaborative. SARS-CoV-2 infection and venous thromboembolism after surgery: an international prospective cohort study. Anaesthesia. 2022. 77(1):28-39. | **2,8** |
| K7 | Spielberger B, Pfeil J, Assaad K, von Both U, Janda A, Kitz C, Kobbe R, Kunze M, Lindert J, Ritz N, Trapp S, Hufnagel M. Infektiologische Versorgung minderjähriger Flüchtlinge am Beispiel der Ukraine: Vorgehen in Deutschland gemäß der S1-Leitlinine (AWMF-Register Nr. 048-017) [Care of infectious diseases in underage refugees exemplified by Ukraine]. Monatsschr Kinderheilkd. 2022. 170(12):1103-1112. | **0,4** |
| K6 | Peter N, Bandyopadhyay S, Lakhoo K. Global Health Research Group on Children's Non-Communicable Diseases Collaborative. Impact of the COVID-19 pandemic on paediatric patients with cancer in low-income, middle-income and high-income countries: protocol for a multicentre, international, observational cohort study. BMJ Open. 2021. 11(6):e045679. | **2,9** |
| K5 | COVIDSurg Collaborative, GlobalSurg Collaborative. Effects of pre-operative isolation on postoperative pulmonary complications after elective surgery: an international prospective cohort study. Anaesthesia. 2021. 76(11):1454-1464. | **6,9** |
| K4 | Global PaedSurg Research Collaboration. Mortality from gastrointestinal congenital anomalies at 264 hospitals in 74 low-income, middle-income, and high-income countries: a multicentre, international, prospective cohort study. Lancet. 2021. 398(10297):325-339. | **98,4** |

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| K3 | COVIDSurg Collaborative, GlobalSurg Collaborative. SARS-CoV-2 vaccination modelling for safe surgery to save lives: data from an international prospective cohort study. Br J Surg. 2021. 108(9):1056-1063. | **8,6** |
| K2 | COVIDSurg Collaborative, GlobalSurg Collaborative. Timing of surgery following SARS-CoV-2 infection: an international prospective cohort study. Anaesthesia. 2021. 76(6):748-758. | **6,9** |
| K1 | GlobalSurg Collaborative and National Institute for Health Research Global Research Unit on Global Surgery. Global variation in postoperative mortality and complications after cancer surgery: a multicentre, prospective cohort study in 82 countries. Lancet. 2021. 397(10272):387-397. | **98,4** |
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| | **Sonstige Originalarbeiten, Übersichtsartikel, Korrespondenz ohne Impact** | |
| S6 | Steinle J, **Lindert J**, Mothes H. Globale Chirurgie in Lübeck – Das Global Surgery Symposium der DTC. Passion Chirurgie. 2020. 10(1):04_02. | |
| S5 | **Lindert J**, Kotsias-Konopelska S, Urlaubsbekanntschaft mit Gifttieren. Kinder-und Jugendmedizin. 2019. 19(02):107-116. | |
| S4 | Mothes H, **Lindert J**, Borsche A. Zum Für und Wider chirurgischer Projekte. Passion Chirurgie.2018. 18(07) :8-11. | |
| S3 | Rau C, **Lindert J**, Kotsias-Konopelska S, Kobbe R. Aerogen übertragene Infektionskrankheiten bei Kindern auf Reisen.Kinder-und Jugendmedizin. 2018. 18 (6) :422-426. | |
| S2 | Kotsias-Konopelska S & **Lindert J**. Reisen mit Kindern Teil II : Zeitverschiebung, Reisen in großen Höhenlagen, heißes/feuchtes und kaltes Klima, Rad- und Autoreisen. Kinder- und Jugendmedizin. 2018. 18 (5) :322-327. | |
| S1 | Kotsias-Konopelska S & **Lindert J**. Reisen mit Kindern Teil I : Sonnenschutz. Kinder- und Jugendmedizin. 2018 .18(5) :317-321. | |
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| | **Buchbeiträge** | |
| B2 | In Hrsg. Schubert K, Özbay J, Tinnemann P. Hier&Dort, Einblicke in die Globale Gesundheit / Here and There: Insights in Global Health 2009 | |
| B1 | Stark M, Wacker J, **Lindert J**. Die Durchführung des Kaiserschnitts in armen und reichen Ländern (Misgav-Ladach-Sectio, „der sanfte Kaiserschnitt“). Hrsg : Wacker J, Rothe C, En-Nosse M. Globale Frauengesundheit. ISBN: 978-3-662-66080-5 | |

# 4 Danksagung

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# 5 Erklärung

Hiermit erkläre ich, **Judith Lindert**, dass

- durch mich weder früher noch gleichzeitig ein Habilitationsverfahren angemeldet oder durchgeführt wurde.
- mir die Bestimmungen der Habilitationsordnung bekannt sind und von mir anerkannt werden.
- dass ich die Habilitationsschrift selbständig und ohne fremde Hilfe verfasst habe. Ich habe keine anderen als die angegebenen Quellen und Hilfsmittel verwendet.
- die beschriebenen Ergebnisse selbst gewonnen wurden sowie die verwendeten Hilfsmittel und Zusammenarbeit mit anderen WissenschaftlerInnen und mit technischen Hilfskräften sowie die verwendete Literatur vollständig in der Habilitationsschrift angegeben wurde.