Title:	**GUIDELINE FOR THE MANAGEMENT OF SUSPECTED HIRSCHSPRUNG'S ASSOCIATED ENTEROCOLITIS (HAEC) IN PAEDIATRIC PATIENTS**
Version:	**2**
Supersedes:	Version 1
Application:	The guideline is intended for use by any hospital team caring for infants, children and young people under 16 years age across the Paediatric Critical Care Network in the North-West (England) & North Wales region.

Originated /Modified By: **Designation:**	Kathryn O'Shea Consultant Paediatric Surgeon Royal Manchester Children's Hospital
Reviewed by:	North-West (England) and North Wales Paediatric Transport Service (NWTS) North-West & North Wales Surgery in Children ODN North-West & North Wales Paediatric Critical Care ODN North-West & North Wales Children's Major Trauma ODN Sophina Mahmood, Paediatric Pharmacist, PCC ODN & RMCH
Ratified by:	North-West & North Wales Surgery in Children ODN
Date of Ratification:	SiC Oversight: Date: 13th December 2024
Ratified by:	RMCH (Host Trust): - Paediatric Medicines Management Committee (MMC) - Paediatric Policies & Guidelines Committee
Date of Ratification:	11th April 2025

Issue / Circulation Date:	19th May 2025
Circulated by:	NW PCC SiC LTV ODN
Dissemination and Implementation:	Via NW PCC ODN / NW SiC ODN
Date placed on the websites (NWTS / PCC, SiC & LTV ODN) + MFT intranet	19th May 2025

Planned Review Date:	3 years
Responsibility of:	Clinical lead North-West (England) & North Wales Surgery in Children ODN & NWTS guideline lead consultant and nurse.

Minor amendment (if applicable) notified to:	
Date notified:	

EqIA Registration Number:	**2025-110**

1. Detail of Procedural Document

Guidelines for the management of suspected Hirschsprung's Associated Enterocolitis in paediatric patients.

2. Equality Impact Assessment.

EqIA registration Number for RMCH:	**2025-110**

3. Consultation, Approval and Ratification Process

This guideline was developed with input from:

- North-West (England) & North Wales Surgery in Children Network
- Children's Major Trauma Network, North-West & North Wales
- North-West (England) and North Wales Paediatric Transport Service (NWTS).
- North-West (England) and North Wales Paediatric Critical Care Operational Delivery Network
- Representatives from the District General Hospitals within networks above.

All comments received have been reviewed and appropriate amendments incorporated.

For ratification process for network guidelines see appendix 1.

4. Disclaimer

These clinical guidelines represent the views of the North-West (England) and North Wales Paediatric Transport Service (NWTS) and North-West (England) and North Wales Paediatric Networks mentioned above. They have been produced after careful consideration of available evidence in conjunction with clinical expertise and experience.

It is intended that trusts within the Network will adopt this guideline and educational resource after review and ratification (including equality impact assessment) through their own clinical governance structures.

The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient.

Clinical advice is always available from NWTS on a case-by-case basis.

Please feel free to **contact NWTS (01925 853 550)** regarding these documents if there are any queries.

Suspect Hirschsprung's Associated Enterocolitis (HAEC) IF:

Background of Hirschsprung's disease

Fever

Abdominal distention

Constipation/Diarrhoea

→ NO

↓

Consider alternative diagnosis (see acute abdomen guideline).
Surgical input as deemed appropriate by local team.

↓ YES

Urgent Paediatric Consultant review +/- Anaesthetic review

↓

ANY SYMPTOMS TO SUGGEST SEVERE HAEC?

-Lethargy
- CRT > 3s
-Severe Tachycardia (see chart page 6)
- Any hypotension
- Severe abdominal distention compromising respiration
- Temperature > 38.4℃

→ YES

Management of suspected Severe HAEC

Aim $SpO_2 \geq 94\%$
Consider I&V (if necessary)
IO/IV access x 2
NGT on free drainage/NBM
Urgent blood gas + lactate
FBC/U&E/LFT/Clotting/Calcium/CRP/Amylase/G&S
Blood cultures
Abdominal X-ray
Fluid bolus (10-20mL/kg):
(Plasmalyte 148 or Hartmann's solution)
IV tazocin & metronidazole
Start IV maintenance e.g. Plasmalyte 148 + 5% glucose
Consider early inotropic support
Rectal washout if trained member of staff available

↓

TIME CRITICAL?

Contact the on call surgical registrar at AHCH/RMCH via switchboard ASAP to establish urgency of transfer (regardless of bed state)

Rectal washout ONLY if trained member of staff available

↓ YES

Local team to arrange CATEGORY 2 TRANSFER +/- NWTS advice on stabilisation and transfer (see page 5)

RMCH on call surgical registrar (switchboard): 0161 276 1234

NO →

Local hospital/Tertiary teams to agree management plan +/- NWTS advice re: stabilisation +/- transfer

AHCH on call surgical registrar (switchboard): 0151 228 4811

For drug calculations use: https://www.nwts.nhs.uk/emergency-drug-guides

KEY POINTS

Any child with suspected Hirschsprung's Associated Enterocolitis (HAEC) should be referred to the tertiary paediatric surgical team.

Hirschsprung's associated enterocolitis (HAEC) can mimic simple gastroenteritis. Any child presenting unwell with a background of Hirschsprung's Disease (H.D) should undergo a thorough clinical assessment.

Severe HAEC carries a significant associated mortality. Resuscitation, rectal irrigations, and antibiotic administration should be instigated immediately. This is applicable to a child of any age with a diagnosis of HD pre or post definitive surgery, with or without a stoma.

BACKGROUND

Hirschsprung's disease (H.D) is characterised by aganglionosis of a variable segment of the bowel. The lack of ganglion cells means the affected bowel is constricted, leading to a functional bowel obstruction. Nine out of ten babies with H.D. present in the neonatal period, it is more common in babies with trisomy 21. Prior to their definitive operation or following their operation, children with H.D are at risk of developing Hirschsprung's associated enterocolitis (HAEC). This is the leading cause of morbidity and mortality in children with H.D. Symptoms of HAEC include abdominal distention, change in bowel habits (constipation, diarrhoea, offensive stools), rectal bleeding, vomiting, and lethargy. Any child presenting unwell with a background of H.D should undergo a thorough clinical assessment, in line with this guidance. Any child with suspected Hirschsprung's associated enterocolitis should be discussed with the tertiary paediatric surgical team.

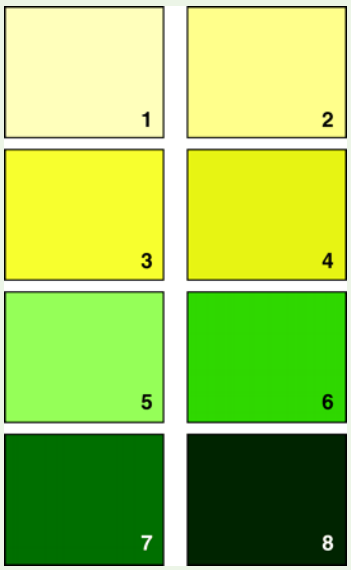
RISK FACTORS FOR SEVERE DISEASE

Awaiting definitive surgery
Previous enterocolitis
Underlying medical condition—in particular T21

BEFORE contacting tertiary paediatric surgical team ASK ABOUT:

Vomiting: What colour is the vomit?
(bile-stained vomit = 7 & 8 indicates significant obstruction and mandates an NG tube)
(https://www.bmj.com/content/332/7554/1363)
Any change in bowel habit? Yes / No
Reduced frequency of opening bowels? Yes/no
Diarrhoea: liquid stool/liquid stool with blood
Foul-smelling stool: Yes/No
Rectal bleeding without diarrhoea: Yes/No
Explosive flatus: Yes/No
Lethargy: Yes/No

Tool for assessment of vomit



EXAMINE THE CHILD FOR:

Prolonged CRT: <2s/3-4/>4
Tachycardia: green / amber / red (see page 6)
Hypotension: Yes/No (see page 6)
Temperature: apyrexial/Low grade (up to 38.3)/High grade (38.4 and above)
Abdominal distention: Yes/No

INVESTIGATIONS:

Blood gas within 30 minutes of referral. FBC, U&E,CRP
Abdominal Xray

RECTAL WASHOUTS

Rectal washouts are a life saving measure in Hirschsprung's associated enterocolitis.
If a trained member of staff is available these should be included as part of resuscitation and stabilisation.
Parents are often trained and may be able to perform a washout if provided with the correct equipment.
Rectal irrigation tube size: 2-4kg = 8ch. 4-6kg = 10ch. >6kg =16ch.
Use 20mL/kg of warmed 0.9% NaCl (this is a starting volume, the bowel should be irrigated until clear)
Ensure instilled fluids drains: irrigation fluid left dwelling in the bowel could worsen the clinical situation
https://www.alderhey.nhs.uk/wp-content/uploads/2023/06/Rectal-Washouts-Leaflet-PIAG-315.pdf

RESPONSIBILITIES OF LOCAL TEAM: TIME CRITICAL TRANSFERS

Stabilise child

Intubate and ventilate child if required

Treat circulatory instability (discuss with NWTS for advice)

Contact Consultant Surgeon at tertiary centre (telephone numbers below)

Discuss need for Imaging with Consultant Surgeon at tertiary centre

Identify appropriate transfer team (eg experienced anaesthetist + appropriate nurse/ODP) using STOPP risk assessment (see below)

Contact NWAS via 0345 140 0144 and ask for "**Category 2**" ambulance

https://www.nwas.nhs.uk/services/professionals/emergency-ambulance/

OR contact Welsh Ambulance Service via healthcare professional line (or 999) & request a time critical transfer following local policy

Arrange transfer of all images via PACS

Refer to safeguarding team if appropriate

Undertake transfer within 1 hour of agreed need for transfer to tertiary centre

AMBULANCE REQUEST FOR TIME CRITICAL TRANSFER VIA NORTH WEST AMBULANCE SERVICE (NWAS) = CATEGORY 2

- **Category 2 response time = 18-minute** mean response time and should be used if child or young person is due to have intervention, ie surgery, on arrival at tertiary centre.
- **Category 3:** for any urgent transfers via emergency department for further surgical assessment / review

https://www.nwas.nhs.uk/services/professionals/emergency-ambulance/

For further information on organising inter-hospital transfers with NWAS. The code assigned is based on clinical presentation and not location of patient. All requests must be placed on the basis of clinical need.

AMBULANCE REQUEST FOR TIME CRITICAL TRANSFER VIA WELSH AMBULANCE SERVICE: follow local policy

IF ANY DELAY IS ANTICIPATED eg ambulance NOT arrived within 20 minutes: **escalate** the call to the senior clinician on duty for either NWAS or Welsh ambulance service

TEAM COMPOSITION:

- Always use **STOPP tool** (**https://nwts.nhs.uk/guidelines**) for all paediatric transfers
- Complete a risk assessment prior to any transfer; if any delay repeat just prior to transfer
- Joint decision should be made by paediatric & anaesthetic consultants on team composition
- Any potential airway concerns require an anaesthetic escort

NWTS: 08000 84 83 82 NWTS will….

Contact the Consultant Surgeon at tertiary centre if not already done so by the local team via conference call

Advise local team on stabilisation & transfer if required

For drug calculations use: https://www.nwts.nhs.uk/emergency-drug-guides

RESOURCES: Quick reference guide for National PEWS

TARGETS for managing any critically sick child			
ALL AGES	SpO_2 ≥ 94%	ET CO_2: 4-4.5 kPa	Glucose: ≥ 3 mmol/L

CAUTION pulse oximetry values may over-read especially in those with dark skin, masking occult hypoxaemia. Occult hypoxaemia is defined as low arterial oxygen < 88% when compared with pulse oximetry (SpO_2) ≥ 92%.

AGE	TARGET MEAN BP	AGE	TARGET MEAN BP
0-11 Months	45-55	5-12 Years	60
1-4 Years	55-60	>13 Years	60-65

Respiratory Rate (Score up to 4)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
0-11 months	0-10	11-20	21-20	31-49	50-59	60-69	≥70
1-4 years	0-10	11-20		21-39	40-49	50-59	≥ 60
5-12 years	0-10	11-15	16-20	21-24	25-39	40-49	≥ 50
>13 years	0-10		11-15	16-24	25-29	30-39	≥ 40

ALL AGES Score	Respiratory Distress (Score up to 4)
0 = none	None
1 = mild	Nasal flaring, subcostal recession
2 = moderate	Tracheal tug, intercostal recession, inspiratory or expiratory noises
4 = severe	Supraclavicular recession, grunting, exhaustion, impending respiratory arrest

ALL AGES Score	Oxygen Saturations (Score up to 4)
0	95-100%
2	92-94%
4	≤ 91%

ALL AGES Score	Oxygen Requirement (Score up to 4) - ALL AGES
0	Room Air
2	0.01 up to 4 litres/min
4	4 or more litres/min **NB** High flow humidified NC oxygen, NIV CPAP or BiPAP score 4 (irrespective of oxygen requirement)

Heart Rate (Score up to 4)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
0-11 Months	0-80	81-90	91-110	111-149	150-169	170-179	≥ 180
1-4 Years	0-60	61-70	71-90	91-139	140-149	150-169	≥ 170
5-12 Years	0-60	61-70	71-80	80-119	120-139	140-159	≥ 160
>13 Years	0-50	51-60	61-70	71-99	100-119	120-129	≥ 130

Blood Pressure Systolic (Score up to 4)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
0-11 Months	0-50	51-60	61-70	71-89	90-99	100-109	≥ 110
1-4 Years	0-50	51-60	61-80	81-99	100-119	120-129	≥ 130
5-12 Years	0-70	71-80	81-90	91-109	110-119	120-129	≥ 130
>13 Years	0-80	81-90	91-100	101-119	120-129	130-139	≥ 140

Capillary Refill Time (CRT) (Score up to 2)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
All Ages		≥ 3 secs		<3 secs		≥ 3	

Guidelines for the management of suspected Hirschsprung's Associated Enterocolitis in paediatric patients

NPEWS: CHECK IF YOUR PATIENT HAS ANY ADDITIONAL RISK FACTORS		
RISK FACTOR	**THINK!**	
☐ Baseline vital signs outside normal reference ranges	Always score relevant PEWS value even if this is normal for the patient eg cyanotic heart disease	Vital sign: ☐ Patient's normal value: ☐
☐ Tracheostomy / Airway Risk	Do you need additional help in an airway emergency? Needs review by local anaesthetics & ENT teams. Consider d/w NWTS early	
☐ Invasive/Non-invasive ventilation/high flow	Check oxygen requirement on additional respiratory support. Remember High Flow/BiPAP & CPAP score max 4 on oxygen delivery	
☐ Neutropenic/immunocompromised	Sepsis recognition & escalation has a lower threshold	
☐ <40 weeks corrected gestational age	Sepsis recognition & escalation has a lower threshold (beware hypothermia)	
☐ Neurological condition (ie meningitis, seizures)	Remember: check pupil response if anything other than ALERT on AVPU	
☐ Neurodiversity or Learning Disability	Be aware of the range of responses to pain & physiological changes	

NPEWS ESCALATION LEVEL	ACTIONS	MEDICAL REVIEW	OBSERVATIONS / PLAN
E0 – no concerns **Score = 0**	None	Not required	Continue current observations
E1 – Increased monitoring **Score = 1- 4**	Inform Nurse-in-Charge Consider medical review (ST3+ or equivalent) Ensure feedback to parents	As required Discuss with Nurse-in-Charge whether medical review required	Reassess within 60 mins & document ongoing plan
E2 – Needs clinical review (within 30 mins) **Score = 5-8**	Review by Nurse-in-Charge Ensure feedback to parents	Within 30 mins Review by ST3+ or equivalent Discuss stabilisation plan with consultant	Reassess within 30 mins & document ongoing plan Continuous SpO_2 monitoring
E3 – Needs rapid review (within 15 mins) **Score = 9-12**	Immediate review by Nurse-in-charge Discuss medical plan with consultant Senior feedback to parents	Within 15 mins Alert to ST3+ or equivalent Stabilisation plan to be agreed after review by consultant Consider NWTS referral after consultant review	Reassess every 30 mins Continuous monitoring SpO_2, RR, & ECG Record full GCS if change in AVPU
E4 – Needs emergency review (immediate) **Score > 12**	Immediate review by Nurse-in-Charge Consider immediate 2222 call for immediate emergency medical response Inform paeds consultant Senior feedback to parents	Immediate Alert to ST3+ or equivalent Consultant review ASAP Anaesthetic review Consider NWTS referral after appropriate initial interventions	Reassess every 15 mins Continuous SpO_2, ECG, & RR Record full GCS if change in AVPU

NB Escalation levels can also be selected and triggered if parent or carer expresses concern that their child needs increased monitoring +/- clinical review despite PEWS, OR parent or nursing gut instinct irrespective of score.

Medical Plan for Stabilisation:

Structured plan must be documented including:

1. Specific actions to be taken
2. Expected outcome
3. Outcome deadline / in timeframe
4. Escalation if outcome not met by deadline / in timeframe

GUIDELINES FOR < 16 YEARS: ***www.nwts.nhs.uk/clinicalguidelines***

Emergency Drug Guides (wt based) via NWTS website home page - for intubation drugs / sedation regime / inotropes …

https://www.nwts.nhs.uk/emergency-drug-guides

Safe Transfer of Paediatric Patients (STOPP) tool which includes risk assessment prior to transfer, and checklists

NWTS LocSIPPS / Checklists includes sizes of ETT, suction, NGT, CVL & arterial lines

Guidelines include intubation and difficult airway, sepsis, insertion of intraosseous line, collapsed neonate or infant, management of under 16 years outside PCC level 3 unit, and transfer

EDUCATION: ***www.nwts.nhs.uk/education-website***

Includes recordings of NWTS education eg time critical transfers, surgical abdomen etc

Login details for NWTS education site are available from your nursing, AHP and medical paediatric critical care operational delivery network links

OR via email: info@nwts.nhs.uk

FOR DRUG DOSES:

British National Formulary for Children

Emergency Drug Guide via https://www.nwts.nhs.uk/emergency-drug-guides

RECEIVING TERTIARY CENTRE FOR TIME CRITICAL ACUTE ABDOMEN PATIENTS

REFERRING HOSPITAL	General Surgery	Neonatal Surgery	Major Trauma	Preferred PICU
Aintree	AHCH	AHCH	AHCH	AHCH
Arrowe Park	AHCH	AHCH	AHCH	AHCH
Bangor	AHCH	AHCH	AHCH	AHCH
Furness General	RMCH	RMCH	AHCH	RMCH
Blackpool Victoria	RMCH	RMCH	AHCH	RMCH
Royal Blackburn	RMCH	RMCH	RMCH	RMCH
Royal Bolton	RMCH	RMCH	RMCH	RMCH
Burnley General	RMCH	RMCH	RMCH	RMCH
Countess of Chester	AHCH	AHCH	AHCH	AHCH
Chorley	RMCH	RMCH	RMCH	RMCH
Fairfield General (Bury)	RMCH	RMCH	RMCH	RMCH
Glan Clwyd	AHCH	AHCH	AHCH	AHCH
Royal Lancaster Infirmary	RMCH	RMCH	AHCH	RMCH
Royal Liverpool	AHCH	AHCH	AHCH	AHCH
Leighton (Crewe)	AHCH	AHCH	AHCH	AHCH
Macclesfield General	RMCH	AHCH	RMCH	RMCH
Nobles, Isle of Man	AHCH	AHCH	AHCH	AHCH
North Manchester General	RMCH	RMCH	RMCH	RMCH
Royal Oldham	RMCH	RMCH	RMCH	RMCH
Royal Preston	RMCH	RMCH	RMCH	RMCH
Southport and Ormskirk	AHCH	AHCH	AHCH	AHCH
Salford	RMCH	RMCH	RMCH	RMCH
Stepping Hill	RMCH	RMCH	RMCH	RMCH
Tameside	RMCH	RMCH	RMCH	RMCH
Trafford General	RMCH	RMCH	RMCH	RMCH
Warrington	AHCH	AHCH	AHCH	AHCH
Whiston	AHCH	AHCH	AHCH	AHCH
Wigan	RMCH	RMCH	RMCH	RMCH
Wrexham Maelor	AHCH	AHCH	AHCH	AHCH
Wythenshawe	RMCH	RMCH	RMCH	RMCH

REFERENCES

STOPP transfer document ***www.nwts.nhs.uk/clinicalguidelines***

Pastor AC, Osman F, Teitelbaum DH, Caty MG, Langer JC. Development of a standardized definition for Hirschsprung's-associated enterocolitis: a Delphi analysis. J Pediatr Surg. 2009 Jan;44(1):251-6. doi: 10.1016/j.jpedsurg.2008.10.052. PMID: 19159752.

Gosain A, Frykman PK, Cowles RA, Horton J, Levitt M, Rothstein DH, Langer JC, Goldstein AM; American Pediatric Surgical Association Hirschsprung Disease Interest Group. Guidelines for the diagnosis and management of Hirschsprung-associated enterocolitis. Pediatr Surg Int. 2017 May;33(5):517-521. doi: 10.1007/s00383-017-4065-8. Epub 2017 Feb 2. PMID: 28154902; PMCID: PMC5395325.

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Walker G M, Neilson A, Young D, Raine P A M. Colour of bile vomiting in intestinal obstruction in the newborn: questionnaire study *BMJ 2006; 332 :1363 doi:10.1136/bmj.38859.614352.55*

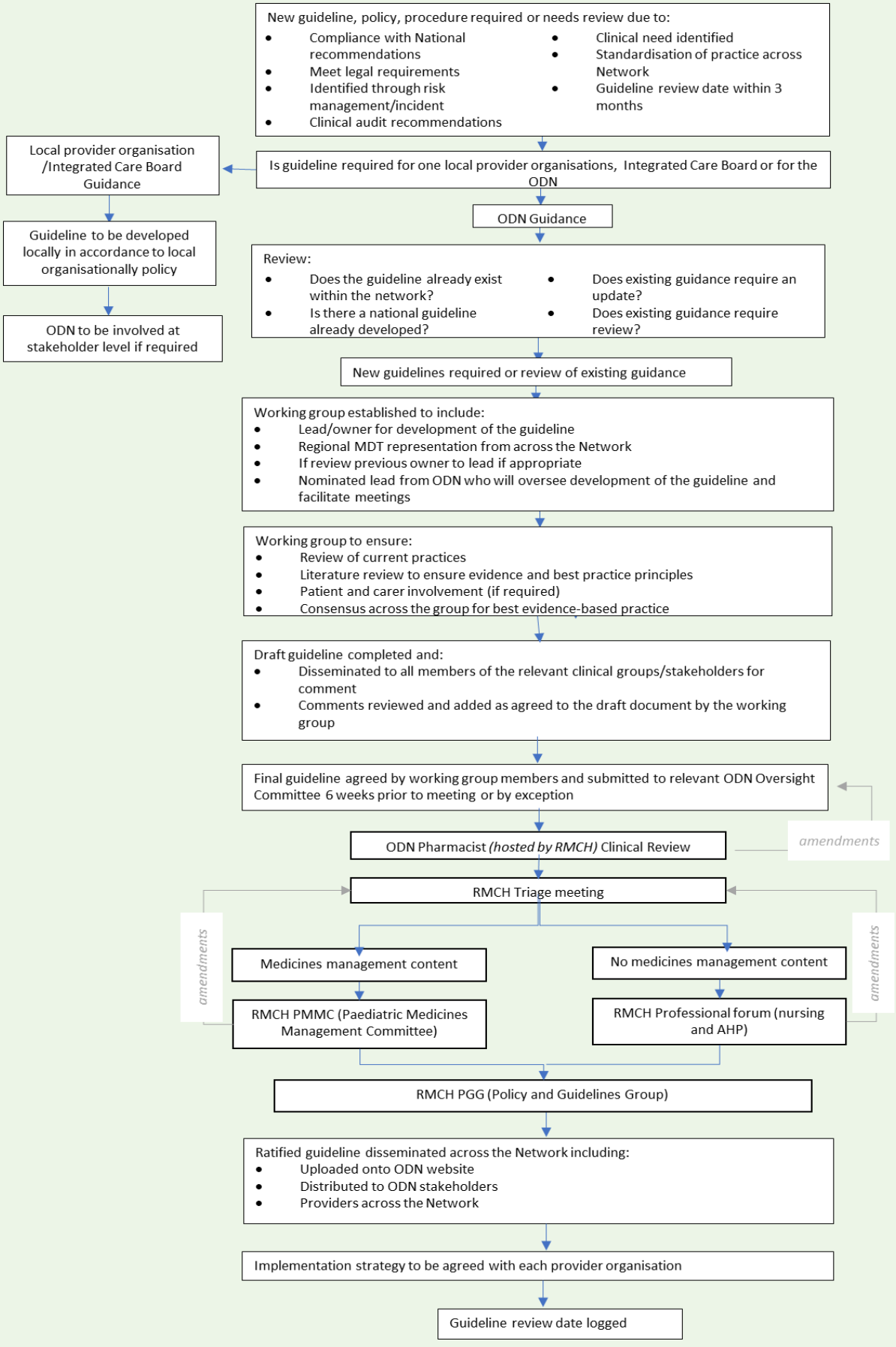
Bokova E, Prasade N, Janumpally S, Rosen JM, Lim IIP, Levitt MA, Rentea RM. State of the Art Bowel Management for Pediatric Colorectal Problems: Hirschsprung Disease. Children (Basel). 2023 Aug 20;10(8):1418. doi: 10.3390/children10081418. PMID: 37628417; PMCID: PMC10453740.

GUIDELINES REFERENCED

Royal Melbourne Children's Hospital: Hirschsprung's Associated Enterocolitis https://www.rch.org.au/clinicalguide/guideline_index/Hirschsprung_associated_enterocolitis_HAEC/

Alder Hey Children's Hospital: Rectal Washouts https://www.alderhey.nhs.uk/wp-content/uploads/2023/06/Rectal-Washouts-Leaflet-PIAG-315.pdf

RATIFICATION PROCESS



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For the most up to date version of this guideline please visit PCC / SiC / LTV ODN
https://northwestchildrensodnhub.nhs.uk/ or
NWTS website https://www.nwts.nhs.uk/clinicalguidelines/regionalguidelines-a-z