Association for Paediatric Palliative Medicine

GI Dystonia:
Stage A: SCOPING AND SET UP PROCESS

Topic and/or Title of proposed guideline	GI dystonia, chronic intestinal failure, progressive or recurrent feed intolerance, and 'neurogenic gut' in the palliative care setting prioritising symptom experience with focus on optimising quality of life from the individual patient with severe neurological impairment and family's perspective.

Specialty area(s) to be addressed	1. Management of GI Dystonia (GID) related symptoms eg reflux, vomiting, nausea and pain (visceral and other types) 2. Management of nutrition and hydration in the face of GID in a palliative care context 3. Role and potential benefits of early engagement of palliative care support where the CYP is/has received optimal management from gastroenterological and other specialist services. 4. Parallel planning for intermittent gut failure 5. Multi-professional approach to GID including role of palliative care in ethical decision-making 6. Management of GID at end of life 7. Consideration of management at different developmental stages 8. Use of alternative routes of medication in GID
Background on the topic	BSPGHAN- developing guidance Look at adult experience
Clinical need for guideline	• Gap in evidence • Emerging issue • Acknowledged patient populations that would benefit • Anxiety about managing of GID- from professionals/families • Non-pharmacological guidance is lacking • Other the counter remedies used with no guidance/experience • Primary vs secondary gut failure issues eg Movement disorder • Artificial Nutrition and hydration and ethical decision-making
Describe the specific issues planning to address through key recommendations	Definitions Consistent approach Auditing practice (national) Other areas identified this as a need
Overall objective(s) of guidelines (scope)	1. Improvement in quality of life for patients 2. Symptom reduction 3. Identification of heralding or early warning signs of gut/intestinal failure/GID 4. Consideration for the impact on behavioural responses due chronic or persisting pain experience 5. Recognition the insidious and variable nature of the condition and the spectrum of severity in terms of symptoms 6. Improved responsiveness and approach to symptom management of the progressive and intermittent nature of the condition 7. Awareness of acute surgical complaints on chronic gut symptoms eg appendicitis, intussusception, complete obstruction secondary to adhesions 8. Improved symptom management approach to autonomic 9. Improvement in the management of symptoms secondary to poor or deteriorating nutrition 10. Standardising approach to care across UK and all health care settings 11. Minimising/reduce health care professional distress 12. Support desired place of care 13. Supporting a good death 14. CYP/Parental/Carer satisfaction experience 15. Transferability of care between care settings and maintaining choice 16. Recognise the liaison role of palliative care for complex symptom management in management of GID even when not at EOL. 17. To support risk/ benefit discussions about interventions and side effects with families including young people when able. 18. Approach to ethical and care management decision making about long term or intermittent PN 19. Maintaining weight or growth rates. 20. Reduction in skin markings, mouth and pressure sores 21. Information on alternative routes of medication

Specific Questions to be addressed	What pharmacological and non-pharmacological interventions are effective for the practical management of the effects of gut failure symptoms (GID dystonia) in infants, children and young people with severe neurological impairment and palliative care needs.
Current evidence existing guidelines? consensus expert opinion Include references	BSPGHAN APPM master formulary RCPCH approved 2014 -palliative care in neonatal (Mancini) Julie Hauer- American Association of paediatric- feed intolerance paper ANH guidance-journal pal med 2019 Paediatric chronic abdominal pain guidance Irritable bowel syndrome guidance
Target audience	Professionals caring for life-limited children including primary, secondary and tertiary services and third sector providers. CYP should ideally be cared for by MDT palliative care team. Funding and commissioning bodies. Infants, children and young people and those caring for them
Age range	1. Neonates 2. Children 3. Adolescents and young people (up to 19years)- over 16yrs may be managed by adult guidance
Population	CYP with life limiting conditions and benefiting from a palliative care approach. This might be defined by complexity, route of drug administration, place of care or phase of illness.
Excluded populations	1. CYP best managed by gastroenterology/ general paediatric teams who do not require palliative care input. 2. CYP with GI Dystonia who are not life limited 3. Age 19 years and over 4. Mechanical obstruction (partial or complete) eg tumour compression 5. Functional complete and partial bowel obstruction secondary to abdominal tumour 6. Those without severe neurological impairment
Clinical condition(s)	A CYP with severe neurological impairment and ▪ Chronic or intermittent Gut failure • Neurogenic gut • Chronic pseudo-obstruction • Gut intestinal dystonia • Intestinal motility disorder • Dysregulation peristalsis • Gut related pain • Visceral hyperalgesia • Feed intolerance • Autonomic dysfunction
Intervention(s)	<u>Pharmacological:</u> Omeprazole, lansoprazole, Ranitidine, Famotidine, Domperidone, Gaviscon. metoclopramide, erythromycin, levomepromazine, cyclizine, ondansetron, granestron, stemetil, nabilone, other cannabinoids, apprepritant, baclofen. gabapentin, pregabalin, amitriptyline, clonidine, SSRI- Fluoxetine, Duloxetine, diazepam, midazolam, lorazepam, clonazepam, clobazam, chloral hydrate. Opiates (morphine, fentanyl, oxycodone, dihydrocodiene and buprenorphine) methadone, ketamine. lactulose, Movicol, enaemas, ducosate, picosulfate, senna. Alimemazine, octreotide, Neostigmine, pyridostigmine, cyproheptadine, H. Pylori treatment. Over-counter remedies: Peppermint tea/oil PN/TPN, home TPN/PN, fluids IV/SC <u>Non-pharmacological:</u> Perastigmen treatment, Farrell bag, flatus tube, replogle tube, ng feeding, jej feeding, venting. hydrolysed formulaes, alterations of feeding regimen, blended diet, exclusion diets, feed thickeners, carobel. Psychological intervention, distraction therapy, music therapy, art therapy, play therapy, complementary therapies, acupuncture, hydrotherapy, reflexology, abdominal massage. <u>Environmental triggers:</u> place of care, access to tissue viability, bed and seating cushions, mattresses including airflow, oral care and hygiene, over feeding, formula osmolarity, feeding rate reduction. <u>Surgical/procedural:</u> botox, celiac plexus block, gastrostomy, jejunostomy, fundoplication, defunctioning colostomy, gut resection, transplant, PN, central line, midlines, PICC lines, Roux en y, stenting, dilatation

Comparison(s)	Placebo No treatment / usual care Cross comparison between any of the above (within group and between group) Combinations of the above – reducing triggers and pharmacological management. Routes of administration (same drug or same drug class)
Health care setting or context	UK, Hospital, home, hospice and community settings where skills and resources allow. Managed clinical network support may enable this.
Outcome(s)	1. Reduced frequency or intensity of gut related symptoms (pain, nausea, vomiting, retching, bloating, gastric losses, constipation, diarrhoea) 2. Reduced distress as experienced by child and family. 3. Supporting individualised family choice around most appropriate use of hydration and nutrition 4. Establishing new goals of care and accepting changes in care goals 5. Potential improvement in gut motility and/or improve feed tolerance 6. Care in place of choice. 7. Improved patient and family experience/ carer satisfaction. 8. Improved trust in healthcare support including perceived quality of care and quality of experience. 9. Reduction in presentation to acute care. 10. Minimise harm / side effects - eg. Hydration, nutrition status and weight maintenance 11. Minimising harm from PN eg line infections and liver failure 12. Minimising harm related to investigations and surgical interventions 13. Reduction of symptoms associated with poor nutrition eg skin markings, mood 14. Acceptability to patients / families and professionals. 15. Achieving a 'good' death as determined by patient and family. 16. Improving confidence and ability to participate in activities of daily living. 17. Identification of heralding or early warning signs of GID and symptom management guidance 18. Impact on behavioural responses due chronic or persisting pain experience 19. Spectrum of severity 20. Progressive and intermittent nature leads to challenges around approach to symptom management 21. Recognition of acute surgical complaints on chronic eg appendicitis, intussusception, complete obstruction secondary to adhesions 22. Approach to ethical and care management decision making about long term PN and no artificial nutrition including judgements about child's quality of life 23. Side effects and risks to interventions offered 24. Autonomic dysfunction and other associated side effects of gut failure eg bladder problems
Stakeholders	APPM Clinical guidelines group and topic specific group Parents and users <u>Before literature:</u> Gastroenterology BSPGHNA (Dr A Barclay and Dr S Protheroe) Neurology BPNA Paediatric BACD and BACCH Psychology -British society of psychology (no response) Dieticians -BDA- bpaediatric dietican (no response) Julie Hauer (Amercian paediatric guidance) (no response) <u>After literature review:</u> Wider APPM membership Gastroenterology BSPGHNA- ask about psychology input Neurology BPNA Psychology -British society of psychology Dieticians -paediatric dietican
Conflict of interest form	Completed No conflict of interest
Questions formulated	1) In infants, children and young people with palliative care needs experiencing GID, what pharmacological, non-pharmacological, procedures and surgical interventions are effective for: a) reducing the distress as experienced by child, their family and professionals b) reducing the frequency and intensity of gut related c) improving associated conditions and side effects d) reducing acute hospital admissions e) potentially improving gut motility and/or improving feed tolerance f) minimising harm, symptoms and side effects from poor nutritional and hydration states g) potentially reducing long term harm through early recognition and intervention h) the management of gut failure at end of life

	i) supporting a good death j) support desired place of care 2) What interventions or measures may be helpful in improving quality of life for patients who experience GID and/or carers? B. in empowering professionals- minimising health carer distress C. in standardising paediatric palliative care across UK and across all health care settings D. in ensuring family/carer satisfaction E. to enable transferability of care between care settings F. to optimise choice of care setting G. to support risk/ benefit discussions with families, including young people when able. 3) How do we define when GID management in collaboration with palliative care is recommended? a) recognising variable disease trajectory and severity b) identifying heralding or early warning signs 4) What is the approach to balancing the benefits and risks of offering interventions (including home PN) with consideration for: a) side effects of poor hydration and nutritional states b) side effects of PN (liver failure) and central access (sepsis, vascular access) c) symptom burden d) perceived benefit/burden to child's quality of life and carers
Literature review	20 years Child only to start, including adult, dependant on results. All study design – including single case reports, posters and abstracts from meetings.
Search strategies	Embase, MEDLINE, PsycINFO, CINAHL, Cochrane, CENTRAL, NICE, HDAS (health education England), Grey literature (abstracts, unpublished papers, posters)
Search words	Neonate, infant, children, young people, paediatric, Adolescent, minor, palliative, terminally ill, dying, terminal, hospice care, end of life care, life-limiting, quality of life, syringe driver, Gut intestinal dystonia, Chronic gut failure, intermittent intestinal failure, neurogenic gut, chronic pseudo-obstruction, feed-induced dystonia, Intestinal motility disorder, peristalsis dysregulation, gut pain, Visceral hyperalgesia, Feed intolerance, Autonomic dysfunction, gut-related pain, gut-related neuropathic pain, nausea, vomiting, retching, bloating, gastric losses, metoclopramide, erythromycin, levomepromazine, cyclizine, ondansetron, granestron, stemetil, nabilone, other cannabinoids, apprepritant, baclofen. gaviscon, gabapentin,-pregabalin, amitriptyline, clonidine, Fluoxetine, Duloxetine, diazepam, midazolam, lorazepam, clonazepam, clobazam, chloral hydrate, alimemazine, cryptoheptadine, cinnizi, Alimemazine, octreotide, apprepritant, Neostigmine, pyridostigmine, Opiates (opiates, morphine, fentanyl, oxycodone, dihydrocodiene and buprenorphine, ketamine, methadone Over-counter remedies: Peppermint tea/oil Perastigmen treatment, hydrolysed formulaes, alterations of feeding regimen, blended diet, abdominal massage, hydrolysed formulaes Farrell bag, flatus tube, replogle tube, formula osmolarity, feeding rate reduction, paraenteral nutrition

GI Dystonia: Stage B:

Guideline development process outline	Scope identified and PICO created (stage A) 1. Systematic review (stage B) 2. Expert opinion: draft developed against each scope identified using systematic review, where no evidence expert opinion used (stage B) 3. Delphi survey - where no consensus was met with expert opinion 4. Evidence linked to each statement
1.Systematic review	Completed by topic specific group and Cochrane Response Reports generated: 1.Systematic review Protocol (Cochrane Response) 2.Systematic review results
2.Expert opinion	Draft guidance created against each clinical statement Using systematic review but where no evidence expert opinion used Expert opinion process: -topic specific group created draft -wider clinical guidelines review (meeting and post-meeting written draft shared)
3.Delphi method	No Delphi survey needed since used combined resources with BSPGHAN
4.Evidence	Each stated guidance has evidence link and grade/rating of quality of evidence. Reports generated: Evidence to Decision table Cochrane Protocol and report
Guideline	Completed January 2023
Additional information	1. Methodology report 2. Guidelines process summary 3. Cochrane Response protocol for systematic review 4. Cochrane Response systematic review results 5. Evidence to Decision table 6. Clinical guidelines participants list 7. Conflict of interest forms (on request)
Funding	NHSE funding for completion of 3 topics and commencement of next 2 topics

Guideline process for GI Dystonia

RCPCH Standards for development of Clinical guidelines
Methodology report
Conflict of interest form

Stage A: Set up

Group

- 42 Clinical guidelines group members
- 2 parents; 1 patient
- 1 Cochrane Response
- 6 core topic group members
- Conflict of interest forms completed
- External stakeholders identified: Palliative care and BSPGHAN

Topic

- Identify a topic lead:
- Tim Warlow
- Methodology report completed for stage A including Clinical question(s), scope, aims,PICOs, search strategy developed by Clinical guideline group over 3 x 2hour virtual sessions

Review

- Reviewed by external Stakeholders: BSPGHAN and Paediatric Palliative care
- Call for evidence from stakeholders and group
- Process follow RCPCH Standards for development of clinical guidelines

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Stage B: Production

RCPCH Setting standards for Clinical guidelines
Summary of Systematic review (Cochrane Response report)
Evidence to Decision table

Systematic review

- Systematic review via PRISMA method with search strategy
- Summary of Systematic review report (see Cochrane Response report)
- Cross reference with BSPGHAN review

Cross referencing

- Several members of the core group including the lead joined the BSPGHAN group lookin at GI Dystonia
- Topic lead contributed to section of BSPGHAN GI Dystonia guidance

Expert opinion meeting:

- 18 attendees including 2 laypeople
- Systematic review presented
- Where there was minimal or no evidence for questions then expert consensus discussion for each question
- Clear consensus for each question with no need for Delphi survey

Draft guideline

- Short version developed by core topic group
- Linked evidence and recommendation by Evidence to Decision table
- Draft presented to guidelines group (one meeting)
- Internal Guideline group review (via email over 6 weeks)
- External review to Stakeholders: Paediatric Palliative care

Stage C: Completion

Guidance process completed (this document)

Revision

- Responses received from Paediatric Palliative care
- Core topic review responses

Dissemination

- APPM Website
- APPM Webinar 2023
- APPM study day 2022
- NHSE newsletter

Updating

- Review date: January 2026

GI Dystonia: Stage C: REVIEW AND PUBLICATION

Guidance final	Final draft sent out to stakeholders including APPM membership, specialist paediatric palliative care group and those involved in the scoping.
Economic impact of guidance	As discussed in evidence to decision table, availability and range of non-pharmacological interventions may be a significant issue in many clinical settings. Specialist palliative care expertise workforce across the systems remains very low.
Barriers to guidance stated	Concern that providing guidance could lead to individual clinician's working beyond their scope of practice. It is a clinician's responsibility to consider and understand their level experience when using the guidelines.
Audit recommendations	To be developed
Dissemination and publication plan	APPM website and webinar series NHSE newsletter
Review date agreed	January 2026