Evidence to Decision (EtD) table: GI Dystonia (GID)

Question

What pharmacological and non-pharmacological interventions are effective for the practical management of the effects of gastrointestinal dystonia (GID) symptoms in infants, children and young people with palliative care needs.	
Background	Although gastrointestinal dystonia (GID) is a relatively new diagnosis its symptoms are well recognised as challenging to manage in paediatric palliative care with little guidance and evidence currently existing to support practice. Areas to be addressed: • Management of GID related symptoms eg reflux, vomiting, nausea and pain (visceral and other types) • Management of nutrition and hydration in the face of GID in a palliative care context • Role and potential benefits of early engagement of palliative care support or intervention where the CYP is/has received optimal management from gastroenterological and other specialist services. • Parallel planning for GID • Multi-professional approach to GID including considerations around PN and end of life care • Use of alternative routes of medication in GID
Objective	1. Improvement in quality of life for patients/carers 2. Reduction in gut and gut-related symptoms 3. Identification of heralding or early warning signs of GID and symptom management guidance 4. Consideration for the impact on behavioural responses due chronic or persisting pain experience 5. Recognition of 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. Improved symptom management approach to autonomic dysfunction and other associated symptoms of GID 8. Improvement in the management of symptoms secondary to poor or deteriorating nutrition 9. Standardising approach to care across UK and all health care settings 10. Minimising/reduce health care professional distress 11. Support desired place of care 12. Supporting a good death 13. Child/Parental/Carer satisfaction experience 14. Transferability of care between care settings and maintaining choice 15. Recognise the liaison role of specialist paediatric palliative care for complex symptom management of GID even when not at end of life. 16. To support risk/ benefit discussions about interventions and side effects with families including young people when able. 17. Approach to ethical and care management decision making in relation to long term or intermittent parenteral nutrition (PN) use and clinically assisted nutrition 18. Information on alternative routes of medication

Population	Child 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. Excluding: • Aged 19 years and over. • Without severe neurological impairment. • Malignant bowel obstruction. • Where symptoms do not have a temporal relationship with feeding and the gastrointestinal tract.
Intervention/ comparison	**Pharmacological:** Omeprazole, lansoprazole, Ranitidine, Famotidine, Domperidone, Gaviscon. metoclopramide, erythromycin, levomepromazine, cyclizine, ondansetron, granestron, stemetil, nabilone, other cannabinoids, appreprintant, 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, prucalopride, linaclotide Over-counter remedies: Peppermint tea/oil PN, home PN, fluids intravenously/subcutaneously. **Non-pharmacological**: Perastigmen treatment, Farrell bag, flatus tube, replogle tube, nasogastric feeding, jejeunal 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. **Environmental triggers**: 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. **Comparisons:** 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)

Main outcomes	• Reduced frequency or intensity of gut related symptoms • Reduced distress as experienced by child and family. • Supporting individualised family choice around most appropriate use of hydration and nutrition • Potential improvement in gut motility and/or improve feed tolerance • Care in place of choice. • Improved patient and family experience/ carer satisfaction. • Improved trust in healthcare support including perceived quality of care and quality of experience. • Reduction in presentation to acute care. • Minimise harm / side effects - eg. PN); investigations and surgical interventions • Acceptability to patients / families and professionals. • Achieving a 'good' death as determined by patient and family. • Improving confidence and ability to participate in activities of daily living. • Identification of heralding or early warning signs of GID and symptom management guidance • Impact on behavioural responses due chronic or persisting pain experience • Progressive and intermittent nature leads to challenges around approach to symptom management • Approach to ethical and care management decision making about long term PN and clinically assisted nutrition •
Setting	UK, Hospital, home, hospice and community settings where skills and resources allow. Supported by Managed clinical network for Children's Palliative Care.
Perspective	Professional working with children with life limited conditions, patients and carers and other health professionals with expertise in GID.

Assessment

	JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
PROBLEM	Is the problem a priority? ☐ No ☐ Probably no ☐ Probably yes x Yes ☐ Varies ☐ Don't know	No primary research evidence identified	'Gastrointestinal failure' was recognised in an APPM survey as 1 of 3 priority symptom topics that needed addressing to support clinical practice (APPM member survey 2019). Experts from the fields of Palliative Medicine, Surgery, Gastroenterology, Neurology have recognised the need for clear definitions and approach to management for these children, with discussions between national societies representing these groups during 2020. Since this time the constellation of symptoms commonly called 'gastrointestinal failure' has been more accurately defined by an appropriateness group for the British Society of Paediatric Gastroenterology, Heptaology and Nutrition (BSPGHAN),British Association of Paediatric Surgeons (BAPS), British Paediatric Neurology Association (BPNA) and Association of Paediatric Palliative Medicine (APPM). The new diagnostic term is 'Gastrointestinal dystonia' (GID)[1]. The symptoms of GID can be numerous and varied including pain; nausea, retching and vomiting; constipation; bloating; pain on defecation and these symptoms can occur at varying times through the childs illness trajectory including, but not exclusive to during the end-of-life phase. Additionally, throughout the childs disease course there can be ethically complex and emotionally challenging decisions to be made, including in relation to artificial nutrition and hydration. GID is a phenomenon not encountered outside of those with severe neurological impairment (SNI) and therefore, is rare, with no formal guidance outside of the paediatric setting. BSPGHAN have also recognised an urgent clinical need for guidance around management of GID and have produced a consensus document alongside representatives from the APPM, BPNA and BAPS. This document (awaiting publication) includes an approach to management. The organisations have worked closely together and as such this current palliative care guidance aims to 'dovetail' with the consensus document to give a complete guide to management of all aspects of care. Recommendations may also be of benefit to other children with similar gastrointestinal symptoms, benefitting from a palliative care approach but who do not fit the formal criteria for a diagnosis of GI dystonia.

DESIRABLE EFFECTS	How substantial are the desirable anticipated effects? ☐ Trivial ☐ Small ☐ Moderate ☐ Large ☒ Varies ☐ Don't know	**See Systematic Review Report here** **Pharmacological interventions :** • TPN may be indicated for a period of gastrointestinal rest prior to reintroduction of feeds in children with gastrointestinal failure and using this to enable gut rest can lead to resolution of symptoms (1 single case study)[2].	Whilst there was a significant paucity in primary interventional studies to base guideline evidence upon there were a number of smaller experimental studies, case series and case reviews. This is in addition to expert consensus documents from specialities working with these children. Recommendations around communication and assessment are 'best practice' recommendations and were agreed by the expert APPM panel writing the guideline consisting of palliative medicine, general paediatric and community paediatric specialists and parent representatives. They were reviewed and agreed by members of BSPGHAN, BPNA and an academic dietitian. Some of these recommendations are based on from Julie Hauer's work on assessment and management of pain in children with SNI[3,4]. The recommendations made in the 'initial management of GID including summary of established early management approaches' section includes both non-pharmacological interventions and non-specialist pharmacological interventions which are key to management of GID, especially in the early stages. These may reduce or eliminate the need for specialist pharmacological symptom management. In general, they should be used prior to specialist palliative medicine pharmacological recommendations. Recommendations are taken from previously published expert opinions and expert consensus opinions and there has also been additional discussion amongst the expert panel with some additional good practice recommendations added. These have been reviewed by external groups including BSPGHAN and BPNA. Expert consensus opinions included in consideration of writing these recommendations: • NICE Guidance Constipation[5]. • BSPGHAN consensus jejunal feeding guidance[6]. • BSPGHAN consensus management of feeding in children with SNI[7]. • Other published expert opinions including work by Julie Hauer on feed intolerance in children with SNI[8]. Specific recommendations made using this previous literature include: **Non-pharmacological interventions :** • Consider use of gastrostomy tube venting to reduce distention[7-9]. • Ensure accurate fluid and calorie assessment (and that there is not evidence of overfeeding)[7-90]. • Optimise enteral nutrition and consider modification of feeding regimen and feed composition. [7-90]. • Consider use of smaller more frequent bolus feeds[9] -Consider running bolus feeds at a rate of <15ml/kg/feed[8]. - Consider a trial of continuous gastric feeding with feeds at a rate of <8ml/kg/hr[8]. - Consider use of a combination of nocturnal continuous feeds with daytime bolus feeds in children with high-caloric needs or poor tolerance to volume[7].

			• Consider trial of blended diet[9,10]. • Consider post-pyloric feeding[6,9]. • Review medication for reduction and rationalisation[9]. • Optimise environmental factors (good practice recommendation by expert panel) • Manage caregiver anxiety and distress (good practice recommendation by expert panel). **Non-Specialist Pharmacological Interventions:** • Optimise Management of gastrooesophageal reflux disease (GORD)[6,7.] • Optimise Management of constipation[5,7] • Optimise management of other health problems[9] Due to the paucity of evidence for the management of GID specialist palliative medicine recommendations including specialist pharmacological interventions in 'symptom specific recommendations' are derived from a combination of evidence found by additional literature searching (not identified within the initial systematic review) and by recommendations made by the panel of expert professionals and patient/parent representitives based on established practice within the wider field of paediatric palliative medicine. This literature included some weak primary evidence in areas which have relevance to GID and review papers including expert opinion. The expert panel have recommended a 'toolkit' of clinically appropriate medications currently used within recognised expert practice which are appropraite to consider in managing the symptoms of GID. Rationale for each of the individual symptom 'toolkits' was dervied following discussions at length with the expert panel and in collaberation with BSPGHAN and BPNA. A 'toolkit' approach was used with recognition that there is not strong evidence for a specific, defined hierarchy in which medications should be trialled and the most appropriate approach is likely to be uniqie for each specific CYP dependant on their situation including medical and wider aspects. Rather than a specific heirachical approach to pharmacological management the toolkit recommends levels of medication trials. These recommendations have been developed based on the weak primary evidence available and consensus opinion within the expert APPM panel and have been discussed at length in collaboration with BSPGHAN and BPNA. They take into account evidence avaliable for medication use including both within the field of paediatric palliative mdicine more widely and in areas relavent to GID; commonly accepted practice within the field of paediatric palliaitve medicine; drug specifc information including the side effect profile and any risk of adverse effects (e.g. risk of dosing errors, side effects, consideration of difficulties and familiarity with route given, requirement for inpatient management/need for investigations prior to procedures, ease of administration, familiarity with drugs).

Those treatments with the best evidence; highest familiarity and lowest risk of adverse effects are included within the recommendations for preliminary pharmacological management moving through to those with the least evidence and familiarity and highest adverse risk profile in the later levels of recommendations.

These considered in more detail for each specific symptom:

<u>Pain:</u>
Current published literature included recommendations of:

- Gabapentin, pregabalin and tricyclic antidepressant trial for visceral hyperalgesia and central pain[8].
- Clonidine for pain perception during gastric and colonic distention[8].
- Gabapentin may be effective in managing pain and visceral hyperalgesia in children with SNI (1 single centre retrospective chart review n=42 and 2 single centre retrospective case series n=31; very-low certainty evidence)[11-13].
- Use of Nabilone for pain, nausea and vomiting in GID (commenced below previously described paediatric dosing and then incremented in 250ug doses to a dosage of <18kgs: 500 ug bd and 18-27kg 500 ug tds). Small case series (n=3); very low-certainty evidence[14].

Within the 'toolkit' there are 4 'levels' of suggested approaches.

Within the first line includes :

- paracetamol, a commonly used and recommended first line analgesic for both acute and chronic pain with minimal established side effects
- gabapentin for which evidence was found (as above) for use in children with SNI and symptoms suggestive of GID (but literature published prior to an established definition of GID), additionally this is known to be a safe medication commonly used within the field of paediatric palliative medicine which brings a familiarity by professionals
- short acting opioids by a number of routes – an established 'second line' analgesic approach for both acute and chronic pain and clonidine for which evidence/recommendation was found within the little literature we found and, again, a commonly used drug within the field of paediatric palliative medicine which has been shown to have a relatively 'safe' side effect profile.

The second 'level' of pain management strategies recommends:

- consideration of a benzodiazepine for anxiety and it is appropriate this is considered only after first line analgesia to address pain
- conversion of clonidine and opioids to long-acting transdermal preparations – this is standard practice within palliative medicine to convert to longer acting preparations if

shorter acting preparations are shown to be effective when an effective dose is established and without presence of any prohibitive side effects.

Third 'level' of pain management includes

- clonidine SC/IV infusion, ketamine, tetrahydrocannabinol and an opioid infusion – these are drugs which require specialist management where the risks of use may be higher than those within level 1 and 2 recommendations.
- Additionally these drugs and routes are less commonly used within standard practice and are likely to require supervision and expertise of a specialist team with prior experience.

Finally, level 4 includes

- methadone and peripherally acting mu-opioids receptor antagonists. These are drugs with higher potential risk and side-effect profile which are likely to require specialist expertise and supervision as they are uncommonly required within paediatric practice.

<u>Upper GI predominant symptoms (nausea, retching, vomiting):</u>

Current published literature included recommendations of:

- Alimemazine 0.25mg/kg TDS (max 2.5mg per dose) may be more effective than placebo for post-fundoplication retching. (Prospective double blind randomised crossover placebo-controlled study; very-low certainty evidence)[15].
- Cyproheptidine may be effective in improving feed tolerance in children with prematurity and brain injury post-NICU admission (1 retrospective chart review; n=39; very-low certainty evidence)[16].
- Cyproheptadine to improve feed tolerance, decrease emesis and retching post fundoplication[8].
- Consideration of trials of prokinetic agents (Domperidone and metoclopramide), Alimemazine, Cyproheptadine, Seratonin 5HT3 antagonist, Levomepromazine, Neurokinin receptor antagonist[6,9]
- Use of Nabilone for pain, nausea and vomiting in GID (commenced below previously described paediatric dosing and then incremented in 250ug doses to a dosage of <18kgs: 500 ug bd and 18-27kg 500 ug tds). Small case series (n=3); very low-certainty evidence[14].

Within the tool kit first line recommendations include a first line prokinetic (as per published guidance); a short acting benzodiazepine (considered within standard paediatric palliative medicine practice).

			Second line alimemazine and cyproheptadine (as per published guidance and weak evidence above); gabapentin and clonidine (as per standard paediatric palliative medicine practice and work of Julie Hauer). Third line metoclopramide, levomepromazine, NK-1 receptor antagonist and clonidine transdermal patches are all considered by the expert panel to be standard established practice within current paediatric palliative medicine practice. Finally, tetrahydrocannabinoil and baclofen have the lowest evidence for us in this group with increasing risk profile and need to use under supervision of an expert team but may provide some benefit when other measures have not been successful in controlling symptoms. <u>Lower GI predominant symptoms</u> (bloating, obstructive symptoms, pain and distress on defecation or passing flatus) : Current published literature included recommendations of: • Pyridostigmine may lead to improvement in bowel opening and reduction in vomiting and distention (single case study; very-low certainty evidence)[17]. This is not recommended within the guidance as not felt to be a helpful and common practice within the field of paediatric palliative medicine and evidence is very weak (from a single case study only). The toolkit approach includes medications widely recommended for managing constipation in children. Additionally further recommendations are made based on routine practice within children's palliative care as agreed by the expert APPM panel writing the guidance. Prucalopride and Linaclotide are recommended only within the final level as they are recommended to be use under specialist guidance only due to unfamiliarity in the paediatric population and have least established evidence for this group. <u>Clinically assisted nutrition and hydration:</u> Recommendations are based on evidence relavent to GID: • PN may be indicated for a period of gastrointestinal rest prior to reintroduction of feeds in children with gastrointestinal failure and using this to enable gut rest can lead to resolution of symptoms (1 retrospective case series)[2,18]. Other recommendations are made based on current accepted good paediatric palliative medicine practice and are consensus opinions of the APPM expert panel and have been formed in agreement with BSPHGAN and BPNA groups. End of life considerations and advance care planning recommendations have been written based on current accepted good paediatric palliative medicine practice and are consensus opinions of the APPM expert panel and have been formed in agreement with BSPHGAN and BPNA groups.

UNDESIRABLE EFFECTS	How substantial are the undesirable anticipated effects? ☐ Large ☐ Moderate ☒ Small ☐ Trivial ☐ Varies ☐ Don't know		Undesirable anticipated side effects of the recommended interventions are 1) Failure to adequately manage symptoms 2) Side effects of the pharmacological interventions recommended Each drug recommended within the guideline has its own specific cautions, pre-requisites and side effect profile which can be found within the British National Formulary for Children[19] and APPM master formulary 2020[20]. Medications should only be used within the skill and expertise of those prescribing and overseeing response. There are several drugs recommended within the guidance (including ketamine, methadone, tetrahydrocannabinol) which we would expect to need to be used under specialist supervision and guidance of those with expertise in paediatric palliative medicine. Several of the drugs recommended for symptom management (e.g. baclofen, clonidine, opioids) will slow the GI tract and these specific side effects need careful consideration in GID and weighing against the benefits in reduction of symptoms.
CERTAINTY OF EVIDENCE	What is the overall certainty of the evidence of effects? ☒ Very low ☐ Low ☐ Moderate ☐ High ☐ No included studies	**Non-pharmacological interventions** • No included studies for non-pharmacological interventions **Pharmacological interventions** • Very low certainty regarding the use of PN (short term to allow gut rest)[2]. • No included studied for other pharmacological interventions	Whilst the systematic review to inform this guideline was limited we have drawn from a number of consensus documents and indirect evidence in order to put those recommendations in the context of rigorous and holistic assessment.
VALUES	Is there important uncertainty about or variability in how much people value the main outcomes? ☐ Important ☒ Possibly ☐ Probably no ☐ No important		Most parents and children will value the main outcomes which aim to minimise the distress caused by GID. Input from parent representatives was central to the development of this guidence from inception, including in developing primary outcomes.

BALANCE OF EFFECTS	Does the balance between desirable and undesirable effects favor the intervention or the comparison? ☐ Favors the comparison ☐ Probably favors the comparison ☐ Does not favor either the intervention or the comparison ☒ Probably favors the intervention ☐ Favors the intervention ☒ Varies ☐ Don't know		Given the variety of symptoms in this presentation and the lack of primary evidence, in combination with the fact that most of the medications included would be being used in an 'off licence' capacity, we are unable to make definitive claims of effeteness. This patient group is very heterogenous in terms of their disease as well as existing medication used. Therefore use of this guidance depends on significant expertise of the medical teams involved. This guidance signposts to sources of advice and support and emphasises at various points where specialist advice should be sought and caution taken.
RESOURCES REQUIRED	How large are the resource requirements ? ☐ Large costs ☐ Moderate costs ☐ Negligible costs and savings ☐ Moderate savings ☐ Large savings ☐ Varies ☒ Don't know		Many of the interventions outlined in the guidance are already part of clinical practice, so there should not be significant cost implications in these recommendations. It is important to consider the cost of medications used, as well as any associated cost of human resources. Most of the pharmacological and non-pharmacological interventions used to manage GID are relatively inexpensive and use of this to improve symptom managment may also lead to cost savings (e.g. through reduced hospital admissions).

CERTAINTY OF EVIDENCE OF REQUIRED RESOURCES	What is the certainty of the evidence of resource requirements (costs)? ☒ Very low ☐ Low ☐ Moderate ☐ High ☒ No included studies	n/a	n/a
COST EFFECTIVENESS	Does the cost-effectiveness of the intervention favor the intervention or the comparison? ☐ Favors the comparison ☐ Probably favors the comparison ☐ Does not favor either the intervention or the comparison ☐ Probably favors the intervention ☐ Favors the intervention ☐ Varies ☒ No included studies	n/a	No formal health economic impact study was conducted
EQUITY	What would be the impact on health equity?		Children would ideally have equal access to good symptom management regardless of their choice of setting, and wherever they lived within the UK. Standarised guidance offers the ability to provide consistency across services that may serve different populations.

	☐ Reduced ☐ Probably reduced ☐ Probably no impact ☒ Probably increased ☐ Increased ☐ Varies ☐ Don't know		However, for more expensive interventions these may not be accessible to children and their families in all regions and across all care settings. Advice in this guidence will not contribute to the extensive inequalities that already exist in the UK health care system and may improve access to certain medications where specilist expertise does not exist. Many recommendataions are already in routine use in paediatrics for other conditions. When confidence is provided to generalists caring for these chlidren by this guidence it may support them in treating the sypmtoms of GID without those tertiary expertise.
ACCEPTABILITY	Is the intervention acceptable to key stakeholders? ☐ No ☐ Probably no ☒ Probably yes ☐ Yes ☐ Varies ☐ Don't know		The provision of a standarised consistent methodical approach to managing the symptoms experienced by GID would be acceptable to key stakeholders. The interventions recommended are already widespread in clinical practice and therefore, should be acceptable.
FEASIBILITY	Is the intervention feasible to implement? ☐ No ☐ Probably no ☒ Probably yes ☐ Yes ☐ Varies ☐ Don't know		Many of the interventions outlined in the guidance are already part of widespread clinical practice, so there should not be significant issues with implementation. However, there will be variation in the pharmacological management which may require some educational support to embed guidance into clinical practice.

Summary of judgements

	Judgement							Implications
PROBLEM	No ☐	Probably no ☐	Probably yes ☐	Yes ☒		Varies ☐	Don't know ☐	
DESIRABLE EFFECTS	Trivial ☐	Small ☐	Moderate ☐	Large ☐		Varies ☒	Don't know ☐	
UNDESIRABLE EFFECTS	Large ☐	Moderate ☐	Small ☒	Trivial ☐		Varies ☐	Don't know ☐	
CERTAINTY OF EVIDENCE	Very low ☒	Low ☐	Moderate ☐	High ☐			No included studies ☐	
VALUES	Important uncertainty or variability ☒	Possibly important uncertainty or variability ☒	Probably no important uncertainty or variability ☐	No important uncertainty or variability ☐				
BALANCE OF EFFECTS	Favors the comparison ☐	Probably favors the comparison ☐	Does not favor either the intervention or the comparison ☐	Probably favors the intervention ☐	Favors the intervention ☐	Varies ☒	Don't know ☐	
RESOURCES REQUIRED	Large costs ☐	Moderate costs ☐	Negligible costs and savings ☐	Moderate savings ☐	Large savings ☐	Varies ☐	Don't know ☒	
CERTAINTY OF EVIDENCE OF REQUIRED RESOURCES	Very low ☐	Low ☐	Moderate ☐	High ☐			No included studies ☒	
COST EFFECTIVENESS	Favors the comparison ☐	Probably favors comparison ☐	Does not favor either ☐	Probably favors intervention ☐	Favors the intervention ☐	Varies ☐	No included studies ☒	
EQUITY	Reduced ☐	Probably reduced ☐	Probably no impact ☐	Probably increased ☒	Increased ☐	Varies ☐	Don't know ☐	
ACCEPTABILITY	No ☐	Probably no ☐	Probably yes ☐	Yes ☒		Varies ☐	Don't know ☐	
FEASIBILITY	No ☐	Probably no ☐	Probably yes ☐	Yes ☒		Varies ☐	Don't know ☐	

Recommendations

TYPE OF RECOMMENDATION	Strong recommendation against the option ☐	Conditional recommendation against the option ☐	Conditional recommendation for either the option or the comparison ☒	Conditional recommendation for the option ☐	Strong recommendation for the option ☐

RECOMMENDATION

Make a clear diagnosis of GID in line with the nationally recognised definition. This should be considered alongside other pathology.

General Principals and Communication

- An overall lead clinician and named lead from each team should be identified from each team involved in the childs care.
- When the diagnosis of GID is considered an initial MDT should be arranged and consideration given to regular MDTs
- Goals of care should be agreed between professionals and the family, monitored and regularly reviewed.

Assessment

- GID is an active diagnosis, not a diagnosis of exclusion and should be considered alongside other pathology.
- MDT assessment should include joint working between Nutrition team; Surgical team; Neurology/neurodisability; Specialist Paediatric Palliative Care; secondary care teams and general practitioners.
- Complete a thorough assessment and review of Gastro-oesophageal reflux disease, constipation, dystonia and spasticity.
- Thorough history, examination, and investigation for other sources of pain is an essential component of the diagnosis and ongoing management of the child with GID.
- Children with SNI should be investigated appropriately for sources of pain and GI symptoms . Management of symptoms should not be delayed until investigations are completed.

Initial Management of children with GID

- Ensure optimal Management of GORD
- Ensure optimal constipation management
- Ensure accurate fluid and calorie assessment
- Ensure optimisation of enteral nutrition which may include optimisation of feeding regimen and/or feeding composition
- Consider trial of blended died
- Gastrostomy tube venting may reduce GI distendion
- Consideration of post-pyloric feeding
- Medication reducation and rationalisation
- Ensure optimisation of other health problems
- Optimise environmental factors
- Management and support for caregiver anxiety and distress

Pharmacological Management- General Principals

- Route of administration of medication must be considered carefully where concerns regarding absorption exist.
- Trials of medication should be conducted using a rigorous approach
- Use of low dose, short acting analgesics and anxiolytics early may break the spiral of ‘pain – anxiety – dystonia’.

Symptom Specific Recommendations

Upper GI predominant symptoms (nausea, vomiting and retching)

- Prokinetic firstline
- Short acting benzodiazepine where anxiety predominant
- Gabapentin has been demonstrated to improve vomiting likely due to alteration of GI motility, dysanutonomia and visceral hyperalgesia
- Metaclopramide can be used for a short trial in over 1year old or in palliative care context.
- Other medication can be considered including: Clonidie, Cyproheptidine, Alimemazine, Leveomepromazine, Neurokinin-1-receptors, Baclofen, Tetrahydrocannabinol (specialist input may be required)

Lower GI predominant symptoms

- Consider Glycerol or Bisacodyl suppositories and possibly enaemas may be required
- Optimise stimulant and softener laxatives
- Short acting benzodiazepine where anxiety predominant/prior to passing stool
- Prucalopride and Linaclotide may be considered under specialist advice

Pain

- Standard stepwise approach to pain management starting with Paracetamol
- Consider non-enteral routes eg transdermal , where appropriate, may be useful to avoid enteral absorption concernc
- Additional medication to consider includes Clonidine, Ketamine, Tetrahydrocanabinol, methadone, PAMORAs

Bloating, flatulence

- Management of bloating usually improves on optimisation of the symptoms and alterations to enteral feed
- Probiotics may be considered as a trial for 8-12 weeks whilst enteral feed is tolerated.
- Peppermint tea or oil can be of benefit according to some families.
- Use of regular suppositories for managing constipation, may improve the regulation of flatulence causing discomfort.

Agitation and anxiety:

- Management of agitation and anxiety is vital as these play a key role in distress expressed by children with GID.
- A combination of comfort measures, a rapid acting analgesic (eg buccal diamorphine) alternating with a rapid acting benzodiazepine (eg buccal midazolam) may be required to manage both pain and anxiety components of distress episodes.
- Seek specialist advice for more detailed management.

Dystonia predominant presentations:

- There is significant cross over between medications used for pain and dystonia and where both exist, these should be considered early in distress management.

Use of clinically assisted nutrition and hydration:

- Where initiation of parenteral nutrition (PN) is considered by the MDT and carers to be in the best interest of the child, it should be offered initially with a clear time limited trial. There must be clear, agreed, written goals of treatment with a strong likelihood that PN will contribute to achieving those goals. A clear written plan for monitoring effect, burden, benefit and risks must be in place.

	• If PN is started during an acute illness, a MDT meeting should be arranged with professionals and carers as soon as practically possible to discuss the best interests of the child, including the benefits, harms and risks associated with continuing PN (as above for a time limited trial) or its withdrawal. • Consideration of the CYP's clinical trajectory, prognosis and preferred place of care should be actively discussed when considering initiation of PN. **End of Life Considerations** • As a the child with GID approaches the end of life, reduced food and fluid requirements are part of the natural dying process. The desire for hydration and nutrition also diminishes during the dying phase. This can make assessment of benefits and harms of continuing to provide assisted nutrition and hydration in this stage challenging for families and professionals. • As the child's condition deteriorates towards the end-of-life, goals of care shift more completely towards comfort and control of distressing symptoms. • It may be appropriate to slow down gastrointestinal motility as obstructive symptoms progress. This reduces symptom generation from stretch of the GI tract. This approach would usually involve discontinuing prokinetic agents and placing gastric feeding tubes on drainage. Consider use of anticholinergic agents. • Offer advice and support if carers wish to persist with oral or enteral feeding and it brings pleasure to the child, even if considered a risk or ineffective so long as this does not cause the child distress. • Discuss early any concern that the child may be unable to tolerate any enteral feed or fluids as this is often a highly emotive & distressing time and will require repeated discussion and consideration. • It is usually not appropriate to initiate clinically assisted hydration and nutrition at the end of life. This includes PN and intravenous/subcutaneous fluids. This may lead to oedema, increased respiratory secretions and distress, and limit choice of preferred place of care whilst dying. • Careful attention to mouthcare, avoidance of skin pressure areas, maintaining skin integrity, management of oedema. **With respect to advance care planning for child with GID:** • The advance care planning process for a) optimisation of hydration and nutrition and b) deterioration of the child's condition and death is part of standard care for the CYP with GID (Consensus agreement BSPGHAN, BPNA and the APPM). • Discuss with the CYP, where appropriate, and carers of the child about feeding and hydration at the end of life. • CYP and their carers should have the opportunity to develop a clear written 'advance care plan' signed by the lead professional caring for the CYP, shared with the whole multidisciplinary team and reviewed regularly.
SUBGROUP CONSIDERATIONS	The guideline focuses on the management of those CYP with a formal diagnosis of GID but recommendations may be relevant to others with symptoms emerging from the gastrointestinal tract.
IMPLEMENTATION CONSIDERATIONS	• Are there any limitations/barriers when caring for a child at home (vs hospital/clinic setting)? • Access to 24/7 specialist support for complex medications and other complex decision making (in all care settings).
MONITORING AND EVALUATION	Review in 3 years, January 2026
RESEARCH PRIORITIES	Prospective case series of management

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