Association for Paediatric Palliative Medicine

APPM Guidelines supported by Cochrane Response

Gastrointestinal Dystonia in children and young people with severe neurological impairment in the palliative care setting

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Introduction on purpose of APPM guidelines:

As the ability to offer complex care in out-of-hospital settings and multi-stepped innovative interventions and treatments increases, paediatric palliative medicine is presented with increasingly complex patient symptomology. The development of the APPM clinical guidelines seeks to address symptoms, topic by topic, offering robust evidence-based, peer-reviewed clinical guidance to clinicians working with children and their families to support symptom management, palliative and end of life care. APPM members identified key symptoms of concern and prioritised them according to clinical need.

Nomenclature:
'Children and CYP' refers to everyone under 18 years old. This includes neonates, infants, and young people when applicable.

'Parents or carers' refers to the people with parental responsibility for a child or young person. If the child or young person or their parents or carers (as appropriate) wish, other family members or people important to them should also be given information and be involved in discussions about care.

Target audience:
Health professionals caring for life-limited children including primary, secondary, tertiary and services and third sector providers.

Age range:
Neonates to children and young people up to 18 years of age. Those over 16 years may be managed using this or adult palliative care guidance.

APPM guidelines group membership:
The APPM guidelines group consisted of doctors in specialist, general and community paediatrics, a paediatric pharmacist, nurses from specialist hospice and hospital settings who all work with life limited children, alongside a patient service user and two parents.

PPI engagement:
The guidelines group wish to acknowledge the unwavering support and commitment of Amy-Claire Davies, Tim Gibb and Lizzie Griffiths who kept the child and young person at the heart of the guidance and ensured their voices were at the forefront of our considerations and recommendations.

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Supporting Evidence:
Supporting evidence for the development of this clinical guideline can be accessed from the APPM website.
Evidence includes:

- Methodology report
- Guideline process flow chart
- Protocol of a guideline: Cochrane Review
- Systematic review: Cochrane Review
- Evidence to Decision
- Conflict of interest forms

Management of Gastrointestinal Dystonia (GI Dystonia) in Children and Young people with Severe Neurological impairment (SNI) in a Palliative care setting

Scope of guidance for topic:

This guideline sets out recommendations for the management of children with Gastrointestinal Dystonia (GID) who may benefit from a palliative care approach. It concentrates on the management of the constellation of symptoms related to GID. These recommendations may also benefit children with other gastroenterological pathology. The recommendations should be seen as a 'toolkit' from which to draw options for assessment and management, that may be used individually or in combination to manage the child's presenting symptoms. Each child is unique and working with them and their carers to understand their lived experience is the cornerstone of assessment and management.

Population included:

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.

Population excluded:

1. Patients who are aged 19 years and over
2. CYP without severe neurological impairment
3. CYP with malignant bowel obstruction
4. Where symptoms do not have a temporal relationship with feeding and the gastrointestinal tract

Definitions:

Gastrointestinal Dystonia (GID) is defined in line with the definition from the appropriateness panel on the 'Definition, Investigation and Management of GID in Children and Young People with Neurodisability' 2019-2022). This panel included representation from the following:

- British Society of Paediatric Gastroenterology Hepatology and Nutrition (BSPGHAN)
- British Association of Paediatric Surgeons (BAPS)
- British Paediatric Neurology Association (BPNA)
- Association of Paediatric Palliative Medicine (APPM)

"Clinical manifestations of distress (pain behaviour, hypertonicity, retching, vomiting, vagal phenomenon, abdominal distension) attributable to the gastro-intestinal tract, directly and indirectly related to feeding and bowel habit, where confounding systems distress have been addressed or excluded."[1]

General principles

Children with Gastrointestinal Dystonia (GID) do not adhere to the traditional definitions of 'gastrointestinal failure' which include the following:

1. Reduction of functional gastrointestinal mass below that needed for digestion and absorption of fluid and nutrients for maintenance in adults and growth in children.
2. A child requiring greater than 50% of calories by the parenteral route for greater than or equal to 28 days post term corrected gestational age.[2]

However, due to their symptoms and distress, children with GID are unable to tolerate enteral feeding, leading to nutritional insufficiency and in some cases failure of enteral nutrition and death.[3]

Other important aspects of this definition:

Must include:

- Child has Severe Neurological Impairment (SNI) (GMFCS 4-5 or equivalent)[4]
- A temporal relationship with feeding and symptoms must be present (although this may lesson or cease in progressive disease)
- Feed intolerance that has reached the threshold for GID would include malnutrition primarily due to periods of feed cessation or reduction, or gastrointestinal symptoms being the greatest burden on quality of life (QoL) for the child and family.

May include:

- Common features of pain, distress, retching, autonomic activation and hypertonicity.
- Less common features such as temporal relationship with bowel habit, involuntary movements and hypersalivation.[1]

The presentation of GID is very variable and this is a heterogeneous group (table 1). Each child is an individual and there is often significant overlap of these phenotypes. Division into these phenotypes is to support assessment and rational management for clinicians working with these children.

Table 1: Example phenotypes that typify GID

Phenotype	Presentation
Pain predominant	•Intermittent feed intolerance with pain and distress which resolves or improves on feed reduction/cessation. •Feed intolerance associated with episodes of intercurrent illness. •Increased frequency and severity of episodes over time. •Autonomic symptoms which may mimic potential infection.
Upper GI predominant	•Intermittent nausea, retching or vomiting which resolves or improves on feed reduction or cessation. •Nausea, retching and vomiting cause distress/agitation and worsen dystonia. •Symptoms often exacerbated by episodes of intercurrent illness. •Increased frequency and severity of episodes over time. •May include pain, distention and be associated with autonomic activation.
Lower GI predominant	•Intermittent abdominal distention, severe pain and distress with temporal association to bowel habit. •Symptoms worse when bowels not opened for period of time. •Symptoms worse on defecation and passing wind. •Associated with marked, intermittent abdominal distension in absence of other signs of acute abdomen.

Communication

An overall clinical lead should be identified alongside a named lead should be identified from each team involved in the child's care. When the diagnosis of GID is considered, the clinical lead should organise a multidisciplinary team (MDT) meeting with all teams involved to facilitate a shared understanding of the child's condition, disease trajectory and holistic care needs.[5] For the child with unstable symptoms, consider regular MDT meetings to ensure clear communication and facilitate shared decision making.[5] Goals of care should be agreed between professionals and the carers. Goals may include nutritional intake, weight gain/maintenance, amelioration of symptoms, and/or ensuring comfort. Goals should be regularly monitored, reviewed and evaluated.[6]

Assessment

GID is an active diagnosis, not a diagnosis of exclusion and should be considered alongside other pathology.[1] MDT assessment should include joint interdisciplinary. Assessment and review of potential gastro-oesophageal reflux disease (GORD), constipation, dystonia and spasticity should be completed.[7] History, examination, and investigation (as appropriate) for other sources of pain is an essential component of the diagnosis and ongoing management of the child with GID. When assessing children with SNI for sources of pain and gastrointestinal symptoms, the benefit of conducting an investigation should be weighed against the discomfort and possible risks.[8]
Management of distressing symptoms should not be delayed if investigations are ongoing or until a diagnosis is reached.[8]

Initial considerations

The primary role of this guideline is to make recommendations for management of resistant symptoms, once initial diagnosis and early gastroenterological, surgical and neurological management has taken place. The authors are aware that not all patients have access to paediatric subspecialists therefore we have included a summary of some of the common suggested recommendations for management of GID by these teams here. This guidance is taken from consensus documents published or in the process of publication and are referenced accordingly.

1. Ensure accurate assessment of fluid and calorie requirements in children with GID.[7]

A realistic assessment of weight gain should be made as children with GID may not follow normal growth centiles. This is vital to provide optimal nutrition and avoid overfeeding.[9] Calorie requirement in children with SNI is commonly overestimated and overfeeding is a known contributor to feed intolerance.[7,10] Algorithms for calculating nutritional requirements can only provide an estimate, be prepared to modify feed intake in the light of actual growth and other nutritional markers.[7]

2. Ensure optimisation of enteral nutrition which may include modification of feeding regimen and or modification of feed composition.[1]

Examples include:

- Consideration of a 30% reduction in feed volume with monitoring of weight, symptoms and assessment of benefit over 2-4 weeks.[7]
- A dietician should determine the need for micronutrient and protein supplementation when feeds are decreased.
- Permissive undernutrition at any stage may be appropriate to minimise symptoms.[7]
- Trial of whey-based formulas.[9,10]
- Trial of hydrolysed or elemental formula even in the absence of food allergy.[10]
- Avoidance of hyperosmolar feeds.[10]
- Use of smaller more frequent bolus feeds e.g.
- Running bolus feeds at a rate of <15ml/kg body weight/feed.
- Trial of continuous gastric feeding running at a rate of less than 8ml/kg body weight/hr.
- Use of a combination of nocturnal continuous feeds with daytime bolus feeds in children with high-caloric needs or poor tolerance to volume.[7]

3. Trial of blended diet

Currently the evidence around the use of blended diet is inconclusive and dietetic advice should be sought[1]. There are some reports of tolerance of greater feed volume, reduction in pain, retching, vomiting, gastroesophageal reflux disease (GORD) and constipation with its use.[11] It may be effective for improving food intake for those with chronic diarrhoea and post fundoplication surgery.[12] It has substantial social benefits for the child and family and increases parental satisfaction.[13]

There is a potential risk of inadequate fluid or protein intake, low calorie density and contamination. A dietician should be involved in initiation of blended diet, monitoring of input/output recording and weight monitoring. If high calorie needs are present the addition of high calorie formula to blended diet has been recommended and is tolerated by some children.[11]

Blended diet may be considered in children from the usual age of weaning. Blended diet may be given in lieu of or alongside prescribed formula feeds. Blended diet may be given to children who are usually gastrostomy or jejunostomy fed.

It is recommended that the blended diet itself is administered as boluses and only through a gastrostomy or the gastric port of a gastro-jejunostomy. Blended diet should generally be considered prior to trial of post pyloric feeding.[1,14] Decisions should be made using a shared decision-making approach in partnership with the child's carers.

4. Ensure optimal management of GORD if present[9]

- First line treatment with a proton pump inhibitor[9]
- Trial of at least one prokinetic drug to promote oral/gastric feeding prior to consideration of jejunal tube feeding. Notably, the use of prokinetics should be reserved for uncontrolled GORD due to weak efficacy and side effects.[9]
- Failure of response to anti-reflux treatment should prompt a thorough diagnostic review. Complications of GID may be mis-attributed to GORD.[15]

5. Ensure optimal constipation management[9,16]

In addition to fluid and dietary measures, a combination of stimulant and softener is usually required in the first instance titrating to effect.[9]

- Movicol Paediatric Plain is an osmotic laxative with softening and bulking properties and may be considered first line if fluid volume is tolerated.[16]
- Senna or Sodium Picosulphate are stimulants which may be added/substituted if required.[16]
- Docusate sodium is a combination of softener and stimulant and may be combined with Senna or Sodium Picosulphate if further stool softening is required.[16]

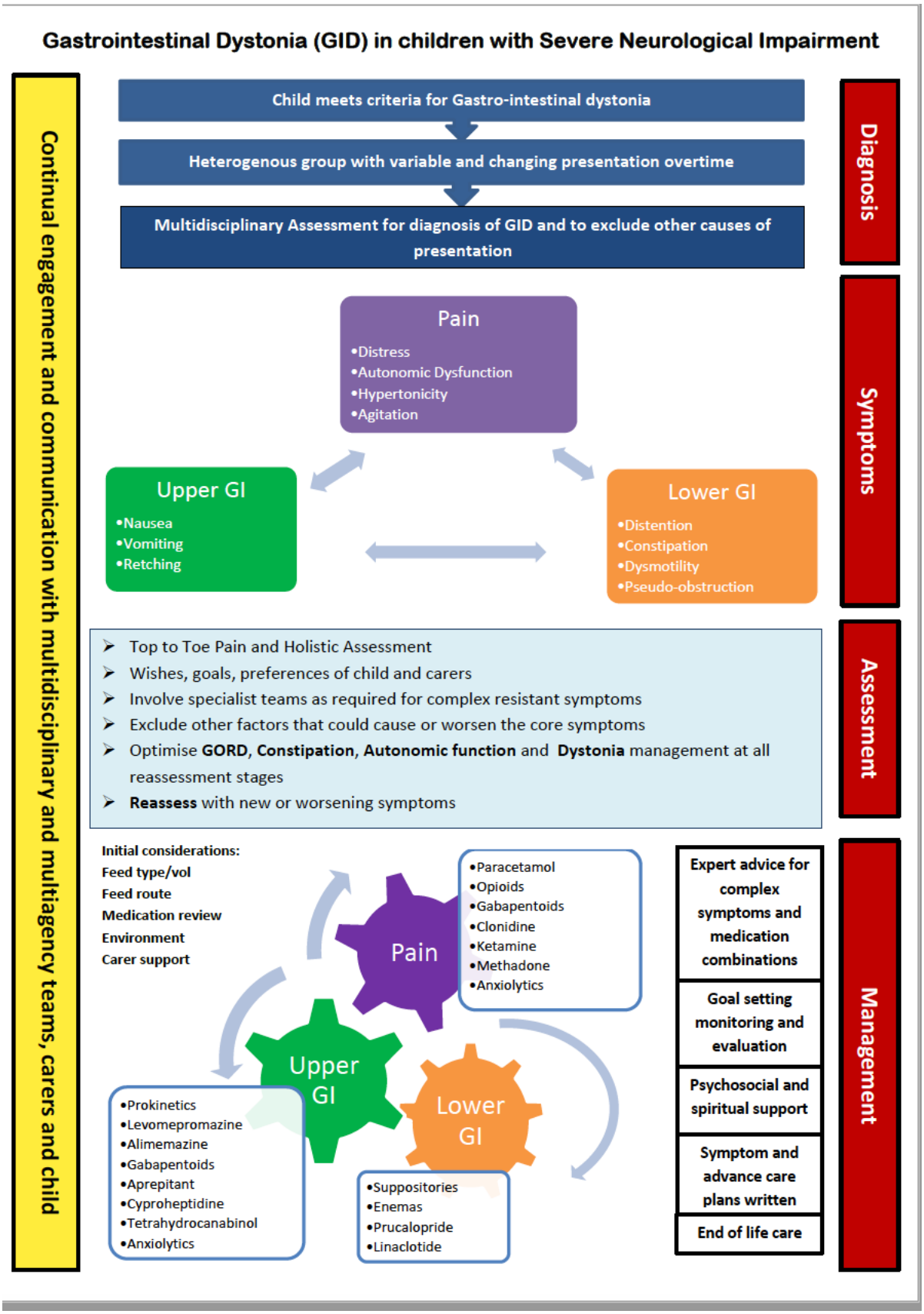
6. Gastrostomy tube venting to reduce gastric distension. Intermittent tube aspiration is an alternative.[7]

7. Consideration of post pyloric feeding in discussion with gastroenterology and surgical teams.[1,10,14]

8. Medication rationalisation

Polypharmacy is often necessary for management of the complex medical needs of the child with SNI. However, consider rationalisation of medications where possible, especially medications impacting on gastrointestinal motility.[10]

Management Algorithm



Non-pharmacological Management

Ensure optimisation of other health problems

- Children with chronic respiratory disease may experience recurrent cough which aggravates vomiting and feed intolerance.
- Treatment of spasticity and generalised dystonia may improve symptoms of GID.
- Secretions may drive coughing and vomiting.
- Autonomic activation may lead to gastrointestinal dysmotility, worsening GID symptoms. Conversely, gastrointestinal dysfunction may precipitate unwanted autonomic symptoms. Advice from a paediatric neurologist should be sought were dysautonomia is suspected to be a significant contributor to distressing symptoms.
- Sleep is important for the management of dystonia as well as pain tolerance. Sleep-wake patterns should be monitored and recorded in discussion with neurodisability, neurology or paediatric sleep teams.[6,7]
- Psychiatric conditions should be assessed and managed with advice from Child and Adolescent Mental Health Services (CAMHS).

Optimise environmental factors

- Liaise with carers and the wider multidisciplinary team (MDT) to identify appropriate methods of comfort for the child during distressing symptoms. These may include distraction, massage, music, containment by cuddling or in blankets, sensory stimulation or reduction of sensory input.
- Address environmental factors with particular emphasis on seeking a calm environment, free from strong aroma, with air moving (consider fan and/or open window).
- Nursing pad/nappy changes, mouth/skin care episodes may be clustered to avoid movement, especially rolling which can cause discomfort.
- Children who prefer sitting in a chair might benefit from a slightly reclined position to alleviate a 'full feeling' (reducing gastric compression).
- Children who predominantly lie down may require care on a slight incline to utilise gravity against reflux or alternative bed options. Safe sleeping guidance should be always followed, and advice sought if concerned.[8]

Management and support for caregiver anxiety and distress

Looking after a child with GID takes a significant toll on carers. The emotional impact of seeing the child in distress, alongside the uncertainties of the future and burden of care delivery can lead to anxiety, especially during episodes of symptom exacerbation. Active management of carer responses and emotion during episodes of distress is vital and can modify the expression and experience of pain in children with SNI.[8]

Empower caregivers by involving them in the development of treatment goals and management strategies.[8,15]Honest and open discussions should be had with caregivers to ensure professional and caregiver expectations are aligned in relation to trajectory of illness, feed volume, growth and symptom reduction goals.[15]Specialist teams should work collaboratively with the child's general practitioner and local general paediatric team to ensure consistent communication, management and support for children and caregivers.

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.[1,32-35]

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 benefits, harms and risks associated with continuing PN (as above for a time limited trial) or its withdrawal.[1] Consideration of the child's clinical trajectory, prognosis and preferred place of care should be actively discussed when considering initiation of PN.[1]

Pharmacological Management

General principals

Consider route of medication administration carefully where concerns regarding absorption exist.[6]
Initiate all medication on a trial basis with a clear review date for continuing, changing or stopping.[6]
Use of low dose, short acting analgesics and anxiolytics early in a symptom exacerbation may break the spiral of 'pain – anxiety – dystonia'. If management is delayed, much higher doses may be required to halt the episode when distress, pain and dystonia are more established.

Symptom Specific

These specific recommendations have been grouped according to the dominant symptoms seen in the child with GID. Most children have a combination of the symptoms described, and a combination of medications may be required. These recommendations should be seen as a 'toolkit' from which to draw options for symptom management. They may be used individually or in combination to manage the child's presenting symptoms.

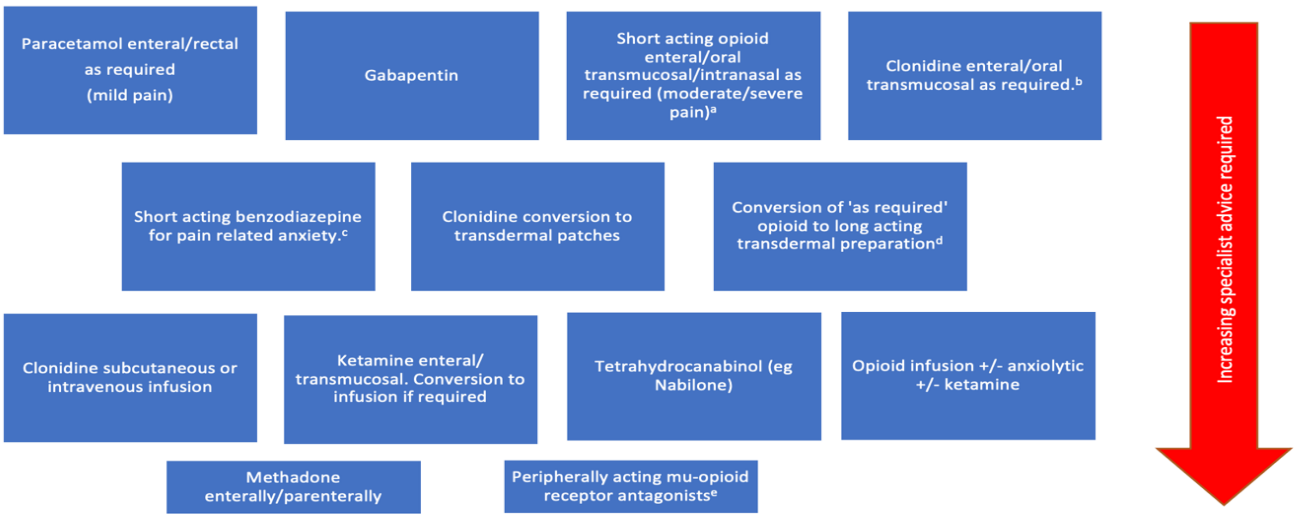
Pain/discomfort

Aim: -Reduce pain signal generation (e.g through reducing gastrointestinal distention, supporting regular peristalsis and gastric emptying)
-Managing visceral hypersensitivity and central pain
-Management of pain related anxiety and agitation

There is a selection of analgesic options (Figure 1). Ideally use non-opioids first. Surgical and interventional procedures (e.g., intrathecal baclofen, gastrointestinal surgery) have not been included here. Choice of analgesic will depend on existing medications, available routes of administration, and pattern of pain experienced. Clinicians should start with medications towards the top of the figure, moving down the analgesic options as pain becomes more severe, resistant, or complex. Clinicians should **only** prescribe within their confidence and competence and specialist advice should be sought from pharmacists and specialist palliative care teams where uncertainty exists or there are a number of potential drug interactions.[1,6-8,18-24]

In children who have a significant dystonic or autonomic activation component to their pain, gabapentin and clonidine may be more effective than long term use of regular opioids.[17] If treatment is ineffective consider whether there is an alternative cause of discomfort e.g., nausea.[15] Anxiety, agitation and distress must be managed alongside pain. The use of short acting benzodiazepines may be helpful for managing symptom exacerbations. If effective, longer acting agents may be used if anxiety is a persistent or recurring feature.[8,22]

Figure 1: Pain medication trials



a)e.g. morphine sulphate or oxycodone enterally, diamorphine or fentanyl transmucosally or intransal using pre-prepared or IV solution.
b)Transmucosal clonidine using IV solution.
c)e.g. morphine sulphate or oxycodone enterally, diamorphine or fentanyl oral transmucosally or intransal using pre-prepared or IV solution.
d)Transmucosal clonidine using IV solution.
e)e.g. Midazolam buccal or Lorazepam sublingual.
f)Opioids with less impact on gastrointestinal motility eg Buprenorphine and Fentanyl transdermally.
g)Opioid anagesics with peripharally acting antagonist to counter impact on gastrointestinal motility (Seek specialist advice)

Upper GI predominant symptoms (nausea, retching, vomiting)

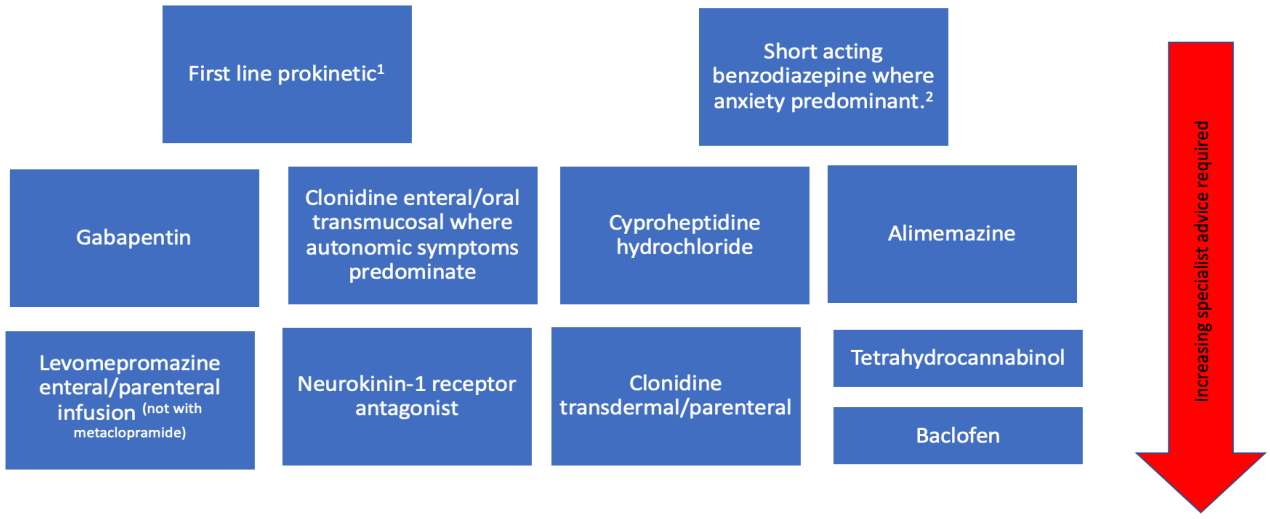
Nausea, vomiting and retching is usually related to a combination of factors:

- Central damage to the areas of the brain involved in the vomiting reflex (especially the area postrema and the extensive neuronal network in the brainstem referred to as the 'vomiting centre')
- Gastrointestinal dysmotility including delayed gastric emptying
- Inappropriate autonomic activation
- Visceral hypersensitivity
- Secretion clearance difficulties triggering repeated coughing or gagging may activate the vomiting reflex
- Emetogenic medications
- Pain
- Psychological factors[7,10]

Aim:

- Improve gastrointestinal motility
- Facilitate gastric emptying
- Increase quantity of feed retained
- Remove triggers to activation of the vomiting reflex
- Suppress activation of the vomiting reflex
- Manage neurological factors

Figure 2: Nausea, retching, vomiting trials



1. Examples include erythromycin, domperidone
2. Examples include midazolam buccally, lorazepam sublingually
3. Gabapentin is demonstrated to improve vomiting likely due to alteration of gastrointestinal motility, dysautonomia and management of visceral hyperalgesia.
4. Metoclopramide should be used for a short trial in over 1 year olds only or in the context of palliative care (unlicensed)

In consensus with experienced specialist palliative care teams and our peer reviewers from fields of neurology, surgery, dietetics and gastroenterology, a list of medication options for children with upper gastrointestinal symptoms has been developed (Figure 2). Surgical and interventional treatments (e.g., intrathecal baclofen, fundoplication) have not been included here.

Choice of agent will depend on existing medications, availability of routes of administration, and pattern of symptoms experienced. Clinicians should start with medications towards the top of the figure, moving down the options as symptoms become more severe, resistant, or complex. Clinicians should **only** prescribe within their confidence and competence and specialist advice should be sought from pharmacists and specialist palliative care teams where uncertainty exists or there are a number of potential drug interactions.[1,7,9,10,12,22-30]

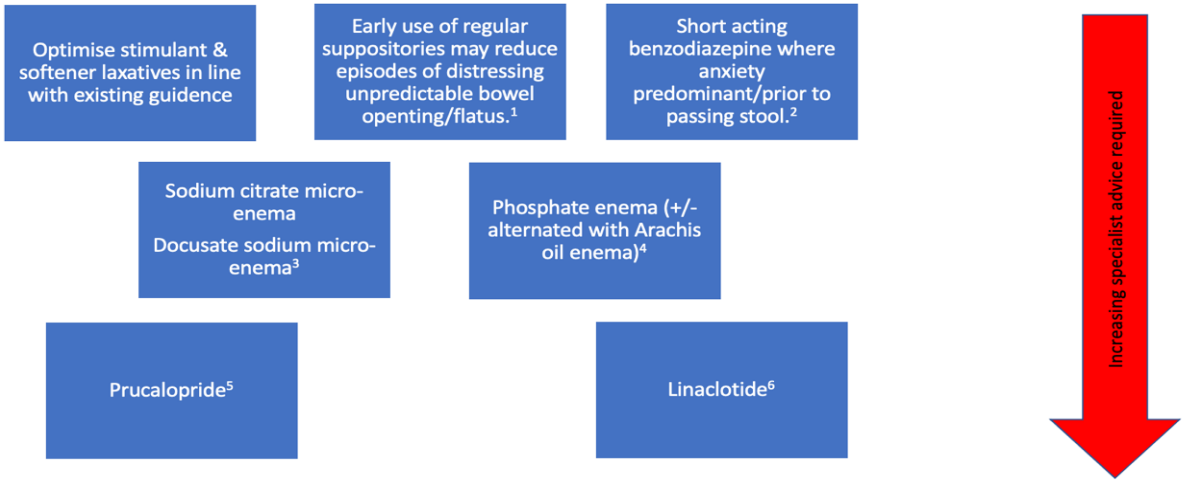
Lower GI predominant symptoms (bloating, obstructive symptoms, pain and distress on defecation or passing flatus)

Children with GID may present with prolonged periods of constipation, pain on defecation or passing flatus and symptoms similar to those seen in intestinal obstruction.

The following factors contribute to these symptoms:

- Chronic constipation leading to alterations in bowel compliance.
- Upper motor neurone damage leading to sphincter dysfunction.
- Autonomic dysfunction.
- Visceral hypersensitivity.
- Intestinal dysmotility.
- Medications leading to impaired peristalsis.[1]

Figure 3: Lower GI symptoms



1. Consider Glycerol (under 4 years) or Bisacodyl (over 4 years) suppositories daily or alternate days. Where distress on defecation is significant, use of a short acting benzodiazepine may be helpful prior to insertion.
2. Examples include buccal midazolam or sublingual lorazepam.
3. May be required in resistant cases or as needed if bowels not open in 48-72hrs for example.
4. May be appropriate for resistant impaction where arachis oil enema can be used to soften stool, instilled and left overnight. This may be followed by phosphate enema the following day.
5. Prucalopride is a 5HT4 receptor agonist with mostly lower GI prokinetic effects. For older children only under specialist advice.
6. Linaclotide is a Guanylate cyclase-c agonist increasing secretion of ions & water into GI tract. For older children only under specialist advice

There is a selection of medication options for children with lower GI symptoms of GID (Figure 3). Choice of agent will depend on existing medications, availability of routes of administration, and pattern of symptoms experienced. Clinicians should start with medications towards the top of the figure, moving down the options as symptoms become more severe, resistant, or complex. Clinicians should **only** prescribe within their confidence and competence and specialist advice should be sought from pharmacists and specialist palliative care teams where uncertainty exists or there are a number of potential drug interactions.[1,9,16,22,31]

Focus on optimising bowel emptying and softening stool alongside management of pain and distress associated with defecation, passing flatus, and bloating or distention.[6,16]

Wean medications which slows gastrointestinal motility if possible, weighing up benefit vs. harm as agents may be important for managing other symptoms e.g., anticholinergic medications, 5HT3 receptor antagonists, some opioids.[7]

If opioids are used for pain, consider preferentially using agents that minimise bowel impact (buprenorphine, fentanyl) or the addition of PAMORAs (partial-mu-opioid receptor antagonist).[18]

Liaise with gastroenterology and surgical colleagues for intractable symptoms where consideration of surgical decompression and stoma formation may appropriate.[15]

Bloating, flatulence

Management of bloating usually improves with optimisation of the symptoms discussed above and alterations to enteral feed described previously.

- Probiotics may be considered as a trial for 8-12 weeks whilst enteral feed is tolerated.
- Peppermint tea or prescribed peppermint water has been reported to be of benefit.
- If unpredictable painful flatulence occurs throughout the day, then once daily suppositories may help to reduce the frequency of these episodes.
- Liaison with a gastroenterology team regarding treatment for bacterial overgrowth and decontamination with antibiotics if not already considered.[1,15]

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 onset (Fentanyl/Diamorphine) or immediate release (Morphine/Oxycodone) analgesic alternating with a rapid acting benzodiazepine (e.g., buccal midazolam) may be required to manage both pain and anxiety components of distress episodes.
- Ensure at least an hour between doses of these immediate release medications, with close monitoring and access to advice.
- Seek specialist advice for more detailed management.[22,23]

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.

End of life care

As 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.[32-36]

As the child's condition deteriorates, goals of care shift more completely towards comfort and control of distressing symptoms.[1] It may be appropriate to slow down gastrointestinal motility as obstructive symptoms progress. This reduces symptom generation from stretch of the gastrointestinal tract. This approach would usually involve discontinuing prokinetic agents and placing gastric feeding tubes on drainage. Consider use of anticholinergic agents.[1]

Offer advice and support if carers wish to persist with 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.[1] Discussions around the discontinuation of feed or fluids at the end of life is often a highly emotive and distressing for carers and will require repeated discussion and consideration. A joint approach with an experienced dietician is recommended. It is usually not appropriate to initiate clinically assisted hydration and nutrition (PN, intravenous or subcutaneous fluids) at the end of life. Some patients experience uncomfortable oedema and increased respiratory secretions. It may limit choice of preferred place of care whilst dying.[1,32-35]

Careful attention must be paid to mouthcare, avoidance of skin pressure areas, maintaining skin integrity, management of oedema.[36] All decisions should be made using a shared decision-making approach in partnership with the child and family. [32-36]

The advance care planning process for survival and optimisation of hydration and nutrition and deterioration of the child's condition and death, is part of standard care for the child with GID.[1] Discuss with the child, where appropriate, and carers of the child about feeding and hydration at the end of life.[32-35] Children and their carers should have the opportunity to develop a clear written 'advance care plan' signed by the lead professional caring for the child, shared with the whole multidisciplinary team and reviewed regularly.[1,36-39] All decisions should be made using a shared decision-making approach in partnership with the child and family. [36-39]

Summary

This clinical guidance focuses on children with severe neurological impairment and palliative care needs presenting with GI dystonia. A consensus definition was developed across interdisciplinary health care professional groups. *"Clinical manifestations of distress (pain behaviour, hypertonicity, retching, vomiting, vagal phenomenon, abdominal distension) attributable to the gastro-intestinal tract, directly and indirectly related to feeding and bowel habit, where confounding systems distress have been addressed or excluded."* The initial management of GI dystonia requires optimisation of nutrition, feeding, constipation, gastro-oesophageal reflux and polypharmacy. Ongoing MDT engagement is vital due to the complexity of managing GI dystonia with the need to regularly revisit goals of care through the use of an inter- and multi-disciplinary teams. Non-pharmacological approaches including optimising the management of other health problems, reducing caregiver distress and considering environmental factors and decisions around clinically assisted nutrition and hydration. Pharmacological intervention targets the management of specific symptoms: pain, upper and lower GI predominant symptoms, bloating, agitation and generalised dystonia. Medication should be offered on a trial basis with assessment of clinical benefit. Choice will also depend on existing medications, availability of routes of administration, the drug profile, clinician experience and pattern of symptoms experienced. Goals of care should be agreed between professionals and the carers and may include nutritional intake, weight gain/maintenance, amelioration of symptoms, and/or ensuring comfort. Goals should be regularly monitored, reviewed and evaluated. When managing GI dystonia towards end of life, careful consideration regarding the benefit and harm of artificial nutrition and hydration should be discussed with the child's family. A focus should remain on ensuring effective management of GI dystonia symptoms.

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