CHAPTER 21

PALLIATIVE CARE

21.1 SYMPTOM CONTROL

Z51.5

DESCRIPTION

The World Health Organisation (WHO) defines paediatric palliative care as the active total care of the child's body, mind and spirit that also involves giving support to the family. Palliative care should begin when illness is diagnosed and continue regardless of whether or not a child receives treatment directed at the disease. Healthcare providers must evaluate and alleviate a child's physical, psychological, and social distress. Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited. It can be provided in tertiary care facilities, in community health centres and even in children's homes.

A key component to relieving suffering is the management of distressing symptoms that include both pain and non-pain symptoms (e.g. nausea, anxiety, etc.). There are certain key principles that should be applied when managing these symptoms, i.e.:

» Determine and treat underlying causes of the symptom, including non-physical causes.
» Relieve the symptom without creating new symptoms or unwanted side effects.
» Consider different types of interventions: drug and non-drug interventions.
» Consider whether the treatment is of benefit to the individual patient.

PRINCIPLES FOR THE SAFE AND EFFECTIVE PRESCRIBING OF MEDICINES IN PAEDIATRIC PALLIATIVE CARE

When prescribing medicines for children in need of palliative care the following needs to be considered:

» Children with advanced illness may have organ dysfunction (esp. renal and liver) and/or be malnourished which may alter their drug handling capabilities, often necessitating the use of lower doses of medications or increased dosing intervals.
» Children with complex medical conditions may already be on several medications to manage their underlying condition – always consider the dangers of polypharmacy and drug interactions.
» Although growing, the evidence base for prescribing in paediatric palliative care is still limited given the ethical challenges of doing research

in this population. For this reason, several medications commonly used in paediatric palliative care are prescribed 'off-licence'. The benefit of prescribing to relieve suffering needs to be weighed against potential harm or drug side effects.

» As children enter the terminal stages of their illness, they may lose their ability to swallow medications or gaining intravenous access may be increasingly difficult. Palliative care practitioners have devised several alternative routes of drug administration to decrease unnecessary procedural pain and enable families to care for their child at home. These should only be utilized by practitioners who have been trained in their use (see section 21.3: End of life and Terminal care).

This chapter provides an approach to the management of several non-pain symptoms commonly encountered in paediatric palliative care. Also see Chapter 20: Pain.

Where these symptoms are already addressed in other chapters, a referral to the relevant sections has been added. It is important to note, however, that when an underlying disease cannot be cured or controlled, these chronic distressing symptoms may need to be managed using medications that would not ordinarily be used in children with acute symptoms that usually resolve with treatment of the underlying disease. Where symptoms are persistent, cause distress, impact on sleep and ability to function resulting in poor quality of life, they need to be addressed through the use of both pharmacological and non-pharmacological means.

21.1.1 GASTRO-INTESTINAL SYMPTOMS

21.1.1.1 ODYNOPHAGIA

R13

DESCRIPTION

Pain that arises from the oropharynx and/or pain with swallowing. Not only does it lead to irritability and discomfort but can affect intake of feeds and medication. May be an unrecognised source of pain in neurologically impaired non-verbal children.

GENERAL AND SUPPORTIVE MEASURES

Perform a thorough examination to determine and treat the underlying cause where possible:

» Uncomplicated gingivitis (see Chapter 2: Alimentary Tract, section 2.1.1).
» Necrotising periodontitis (see Chapter 2: Alimentary Tract, section 2.1.3).
» Aphthous ulcers (see Chapter 2: Alimentary Tract, section 2.1.5).
» Herpes gingivostomatitis (see Chapter 2: Alimentary Tract, section 2.1.6).
» Stevens-Johnson Syndrome (see Chapter 5: Dermatology, section 5.2.2).
» Candidiasis oral (See Chapter 8: Infective/Infectious Diseases, section 8.6).
» Mucositis (secondary to chemo or radiotherapy).

Xerostomia (dry mouth) is also common in palliative care patients and can cause significant discomfort. This is especially prevalent in children who are not taking in orally, are being fed via NGT or feeding gastrostomy, cannot close their mouths and/or are receiving anti-cholinergic medications. Xerostomia is common in children with prolonged hospitalisation with poor attention to oral hygiene. In severe cases the tongue may stick to the roof of the mouth causing pain and bleeding when pried away.

Prevent xerostomia with good oral nursing care and brushing of teeth with a soft toothbrush and fluoride containing toothpaste.

Place ice-chips in mouth to prevent xerostomia and keep lips moist with white soft paraffin.

In children who are feeding orally but where pain is preventing sufficient oral intake, rehydration and/or temporary feeding via naso-gastric tube may be needed (if appropriate: see section 21.3: End of life and Terminal care) while oral lesions heal.

Avoid hot, spicy or acidic food and carbonated drinks in older children still able to swallow and change to soft or puréed diet until condition improves.

Drink through a straw to bypass the mouth.

MEDICATION AND TREATMENT

- Regular washes (2–4 times a day) with chlorhexidine 0.2% mouthwash.
- Paracetamol oral (or per NGT) or PR 15 mg/kg/dose 6 hourly.
- In severe cases of mucositis, oral or even IV morphine may be necessary. (See Chapter 22: Pain Control.)

21.1.1.2 NAUSEA AND VOMITING

R11

DESCRIPTION

Nausea is an unpleasant sensation vaguely referred to the epigastrium and abdomen that often but not always culminates in vomiting. Vomiting is the forcible expulsion of the contents of the stomach through the mouth. It should be distinguished from passive regurgitation and reflux.

In acute illnesses, these presenting symptoms usually resolve with the treatment of the underlying disease and treatment is not indicated. In chronic conditions, especially where there is no cure (e.g. chronic renal failure), this symptom may persist and warrant targeted drug treatment. Untreated nausea and vomiting may contribute to anorexia and weight loss which can accelerate disease progression.

GENERAL AND SUPPORTIVE MEASURES

» Avoid foods/smells that aggravate nausea and vomiting, especially spicy, very sweet or fatty foods.
» Offer foods that are bland and dry until symptoms improve.
» Be careful of wearing strong-smelling perfumes around patients.
» Provide psychological support as anxiety and emotional distress can aggravate nausea and vomiting.
» Maintain hydration by giving small amounts or oral rehydration solution.
» Review medications that may be causing nausea and vomiting, discontinue or change where possible and/or space these out across the day if necessary.
» Nausea is a possible early side effect of opioids that can be managed with short-term anti-emetics. Tolerance usually develops to this side effect within 5–7 days.

MEDICATION AND TREATMENT

Targeted anti-emetic therapy is best to treat the specific underlying cause.

Knowledge of the neuronal pathways, sites of action and receptors involved in nausea and vomiting as well as the sites of action of the anti-emetics are essential to provide targeted therapy.

Targeted anti-emetic therapy in palliative care patients:

Cause	Site	Anti-emetic of choice
Anxiety, fear	Cortex	Lorazepam
Chemotherapy or medications (esp. opioids) or Metabolic (increased urea, calcium)	Chemoreceptor trigger zone	Ondansetron Metoclopramide Haloperidol

Cause	Site	Anti-emetic of choice
Vomiting centre	Viscera (GIT obstruction) Raised intracranial pressure	Hyoscine butylbromide Betamethasone
Obstruction, spasm	Gastric outlet	Metoclopramide
GI inflammation	Oesophagus: GORD Stomach: gastritis	Proton pump inhibitor: e.g.: omeprazole

- Lorazepam, oral:
  - Child < 2 years: 25 mcg/kg 8–12 hourly.
  - Child 2–5 years: 500 mcg 8–12 hourly.
  - Child 6–10 years: 750 mcg 8 hourly.
  - Child 11–14 years: 1 mg 8 hourly.
  - Child > 15 years: 1–2 mg 8 hourly.

- Ondansetron:
  - Oral, 0.1–0.2 mg/kg 12 hourly.

  If oral route cannot be used:
  - Ondansetron, IV, 0.1 mg/kg immediately and every 8–12 hours, thereafter, given over 2–5 minutes. Maximum: 4 mg/day.

- Metoclopramide, oral or IV:
  - Neonates: 100 mcg/kg 6–8 hourly.
  - 1–11 months (up to 10 kg): 100 mcg/kg 12 hourly. Maximum: 1000 mcg/dose (1 mg/dose).
  - 1–18 years: 100–150 mcg/kg 8 hourly. Maximum: 10 mg/dose.

Use with caution as extrapyramidal side effects may occur (especially at higher doses).

- Haloperidol, oral:
  - 1 month to 11 years: 0.01–0.1 mg/kg 12–24 hourly.
  - 12–17 years: 1.5 mg at night, increase to 1.5 mg 12 hourly. Maximum: 5 mg 12 hourly.

- Hyoscine butylbromide, oral, IM or IV:
  - 1 month to 4 years: 300–500 mcg/kg 6–8 hourly. Maximum: 5000 mcg/dose (5 mg/dose).
  - 5–11 years: 5–10 mg 6–8 hourly.
  - 12–17 years: 10–20 mg 6–8 hourly.

- Betamethasone, oral:
  - Child < 1 year: 250 mcg 8 hourly. Maximum: 1000 mcg (1 mg) 8 hourly.
  - Child 1–5 years: 1 mg 8 hourly increased to a maximum of 2 mg 8 hourly.

- Child 6–11 years: 2 mg 8 hourly up to 4 mg 8 hourly.
- Child 12–17 years: 4 mg 8 hourly.

Better to give as intermittent short courses (5 days) rather than for protracted periods of time.
Need to wean if given for > 2 weeks, over a number of weeks.

21.1.1.3 INTRACTABLE DIARRHOEA

R19.7

DESCRIPTION

See guidelines for Persistent and Chronic Diarrhoea (including Non-infectious) in Chapter 2: Alimentary Tract, sections 2.2.5 and 2.2.6 respectively.

Although uncommon, intractable diarrhoea not amenable to any treatment may be encountered in paediatric palliative care. Examples of causes include:

» Short bowel syndrome in neonatal survivors of NEC (< 50 cm, no ileo-caecal valve).
» Severe burns.
» Unmanaged HIV.
» Uncontrolled inflammatory bowel disease.
» Degenerative leiomyopathy.
» Graft vs Host disease.
» Bowel failure.

GENERAL AND SUPPORTIVE MEASURES

» Whilst many of these children may be initially treated at tertiary level, they may be down referred to district or regional hospitals, especially if ongoing treatment is futile.
» Address the underlying cause in as much as this is possible.
» Feed for comfort for as long as possible and whilst child is hungry but do not prolong dying if pre-terminal and not candidates for TPN, see section 21.3: End of life and Terminal care.
» Pay close attention to perineal and peri-anal excoriation that can cause much discomfort.
» Manage abdominal cramps.

MEDICINE TREATMENT

- Loperamide: 0.1 mg/kg 6 hourly increasing up to 2 mg/kg/day.

For acute hypocalcaemia associated with tetany:

- Calcium gluconate:
  - Loading intravenous bolus: 10% calcium gluconate 0.5 mL/kg (0.11 mmol/kg) to a maximum of 20 mL over 10 minutes (maximum rate 0.5 mmol/minute) followed by a continuous intravenous infusion over 24 hours of 0.5–1.0 mmol/kg (maximum 8.8 mmol).

Spasmodic abdominal pain:

- Hyoscine butylbromide, oral, IM or IV:
  - 1 month to 4 years: 300–500 mcg/kg 6–8 hourly. Maximum: 5000 mcg/dose (5 mg/dose).
  - 5–11 years: 5–10 mg 6–8 hourly.
  - 12–17 years: 10–20 mg 6–8 hourly.

REFERRAL

» Refer for trial of TPN in cases where underlying disease is under control.

LoE III[1]

21.1.1.4 CONSTIPATION

K59.0

DESCRIPTION

The infrequent passage of hard stools. See Chapter 2: Alimentary Tract, section 2.2.2. Constipation/Faecal loading.

In palliative care, the most frequent causes are drug related (especially opioids), pain, immobility and neurological impairment (e.g. cerebral palsy).

GENERAL AND SUPPORTIVE MEASURES

» Review medications causing constipation.
» Ensure adequate intake of fluids and fibre when dietary measures are appropriate.
» Mobilize where possible.
» Consider abdominal massage in bed-bound patients: massage in small clockwise circles in the periumbilical area for small bowel and in U-shaped pattern following the direction of stool in colon.

MEDICINE TREATMENT

See Chapter 2: Alimentary Tract, section 2.2.2: Constipation/Faecal loading.

Laxatives should be used prophylactically in all older children receiving morphine. This is not usually needed in infants and younger children.

In bed-bound patients, sometimes disimpaction is needed with the help of a glycerine suppository or enema. **Do not use the rectal route in paediatric oncology patients with neutropenia or thrombocytopenia.**

Be wary of peristalsis-inducing laxatives in children, especially those with neurological conditions: this may cause cramping pain and irritability.

- Lactulose**,** oral, 2.5–10 mL 12 hourly.

OR

- Sorbitol:
  - Children 2–11 years: 2 mL/kg, oral, (70% solution) once.
  - Children 12 years and older: 30–150 mL, oral, (70% solution) once.

Severe constipation in patients unable to swallow:

- Glycerine (glycerol) suppositories:
  - 2 to less than 6 years: 0.891 mL/1.26 g suppository when necessary.
  - 6 years and older: 1.698 mL/2.4 g suppository when necessary.

OR

- Phosphate enema (sodium phosphate 6 g, sodium biphosphate 16 g/100 mL):
  - 2–5 years: 32 mL.
  - 5–11 years: 64 mL.
  - Repeat once, if necessary.

OR

- Polyethylene glycol 59 g/L solution with sodium sulphate and electrolytes, oral/naso-gastric tube, 10–25 mL/kg/hour until clear fluid is passed rectally.

Note: No additional ingredient should be added to the solution, e.g. flavourings or sugar containing cold drinks.

21.1.2 RESPIRATORY SYMPTOMS

21.1.2.1 DYSPNOEA

R06.0

DESCRIPTION

A subjective feeling of breathlessness, when breathing becomes unpleasant, difficult and tiresome.

GENERAL AND SUPPORTIVE MEASURES

» Find the position in which the patient is most comfortable: upright, semi-fowlers or prone. In patients with large effusions: position with the bad lung down to allow for maximum aeration of the good lung.

» Treat the underlying cause where appropriate:
  > Pneumonia is a common end of life event in many palliative care patients, with recurrent infections. Discuss with family when it may be appropriate to withhold or withdraw antibiotics.
  > Consider burden versus benefit of repeat drainage of malignant effusions that re-accumulate.
  > Blood transfusions for anaemia, especially at the end of life, have limited benefit.
  > Diuresis for patients with heart or renal failure may provide transient relief.

» Provide supplemental oxygen if beneficial in patients with end-stage disease. Manage the patient and not the saturation monitor. If dyspnoea

is not relieved, then discontinue oxygen especially if this keeps patients in hospital.

» Reduce anxiety by addressing psychosocial factors, e.g. parental separation.
» Non-pharmacological interventions such as mindfulness, relaxation techniques, music and controlled breathing exercises can be beneficial.
» Blowing cool air onto the face with a fan or open window may also help relieve dyspnoea.
» Warm the air in the room using a humidifier.
» Keep mouth and lips moist in open mouth breathers.
» Manage secretions:
  > If thick, try loosen with saline nebulisation.
  > Physiotherapy and postural drainage where appropriate.
  > Suction as needed but avoid excessive suctioning.
  > See medication for excessive secretions.

MEDICINE TREATMENT

Morphine

Many fear using morphine for dyspnoea because of its potential to suppress respiration. If used at dyspnoea doses and titrated upwards against dyspnoea and/or pain, this is not a concern and should not be withheld even if the child is not for ventilation.

- Morphine, oral [Immediate release morphine (liquid)].
  - Dyspnoea starting dose for opioid naïve patients is 50% of the pain dose.
  - For patients already on morphine (e.g. for pain) increase their current dose by 30–50%.
  - For moderate dyspnoea, oral:
    - If 0–1 month: 25 mcg/kg/dose 6 hourly.
    - If > 1–12 months: 50 mcg/kg/dose 4 hourly.
    - If > 12 months: 100–200 mcg/kg/dose 4 hourly.
  - For severe dyspnoea, IV or SC:
    - If 0–1 month: 0.01 mg/kg/dose immediately.
    - If > 1–12 months: 0.025 mg/kg/dose immediately.
    - If > 12 months: 0.05–0.1 mg/kg/dose immediately.
    - Repeat in 25 minutes if given IV or 30 minutes if given SC and then start continuous IV or SC infusion.
    - IV/SC infusion starting dose: 10 mcg/kg/hour and titrate upwards in increments of 30 to 50% against dyspnoea.

Midazolam

Route	Dose
Oral	• 125–250 mcg/kg [maximum: 20 000 mcg (20 mg)] as a single dose.
Buccal/ Intranasal	• 6 months to 9 years: 50–150 mcg/kg [maximum: 5 mg (5000 mcg)] as a single dose. • 10 to 17 years: 1.5–3.5 mg as a single dose.
IV or SC injection	• 1 month to 5 years: Initially 6.25–25 mcg/kg, to be administered over 2–3 minutes; dose can be increased if necessary in small steps to maximum total dose of 3000 mcg/dose (3 mg/dose). • 6–11 years: Initially 6.25–25 mcg/kg, to be administered over 2–3 minutes; dose can be increased if necessary in small steps to a maximum total of 3750 mcg (3.75 mg). • 12–17 years: Initially 6.25–25 mcg/kg, to be administered over 2–3 minutes; dose can be increased if necessary in small steps to a maximum of 5000 mcg (5 mg).
IV or SC infusion	• 250–1500 mcg/kg/24 hours as starting dose titrating upwards against symptoms and sedation. (Equates to: 10–60 mg/kg/hour.)

For excessive secretions

- Atropine ophthalmic solution 1%:
  - Starting dose (all ages): 1 drop, sublingual every 6 hours.
  - Increase to 2 drops, sublingual every 6 hours.
  - Stop if mouth becomes too dry.
  - **Note**: Following systemic absorption, mydriasis may occur.
  - Ensure this agent is discontinued when evaluating patients for brain death with brain stem testing.

- Hyoscine butylbromide, SC/IV:
  - 1 month to 4 years: 300–500 mcg/kg (maximum: 5 mg/dose) 6–8 hourly.
  - 4–11 years: 5–10 mg 6–8 hourly.
  - 12–17 years: 10–20 mg 6–8 hourly.
  - Continuous SC infusion:
    - 1 month to 4 years: 1.5 mg/kg/24 hours (maximum: 15 mg/day).
    - 5–11 years: 30 mg over 24 hours.
    - 12–17 years: Up to 60–80 mg over 24 hours.
    - Higher doses may be needed: doses used in adults range from 10–120 mg/24 hours (maximum: 300 mg/day).

21.1.2.2 CHRONIC COUGH

R05

DESCRIPTION

Involuntary cough lasting more than 3 weeks caused by chronic stimulation of cough receptors impairing sleep, communication and feeding.

GENERAL AND SUPPORTIVE MEASURES

» Determine and treat the underlying cause where possible.
» Exclude undiagnosed GORD and bronchospasm.
» If caused by ACE inhibitor, consider changing meds.
» Try simple linctus to soothe throat.
» Alternatively, a homemade solution of hot water with a squeeze of lemon and teaspoon of honey.

MEDICATION AND TREATMENT

- Severe and disturbing cough that impacts negatively on sleep and quality of life warrants suppression.
- Codeine is no longer recommended given issues with metabolism (non-metabolizers or ultra-rapid metabolizers).
- Use morphine at dyspnoea doses (see Dyspnoea, section 21.1.2.1).
- If already on morphine for pain, increase dose by 30–50%.

21.1.3 NEUROPSYCHIATRIC SYMPTOMS

21.1.3.1 ANXIETY

F41.9

DESCRIPTION

Anxiety is common in children with life-threatening and life-limiting illnesses and often exacerbates other symptoms (e.g. dyspnoea and insomnia).

Anxiety in this population may be:
1. Transitory – often triggered by certain situations or procedures.
2. Formal generalized anxiety disorder.
3. ‘Death anxiety.’

GENERAL AND SUPPORTIVE TREATMENT

» Anxiety is often triggered by other co-existent symptoms such as breathlessness and unaddressed pain.
» Manage procedure related pain and anxiety (see Chapter 20: Pain).
» Avoid separation from parents or caregivers where possible.
» Communicate openly with children with life-threatening and life-limiting illnesses: especially those that are dying.
» Use cognitive behavioural therapy to desensitize to anxiety provoking situations (e.g. needle phobia).

» Provide opportunities for self-expression and stress release.
» Teach children coping strategies.
» Address and treat insomnia.

MEDICINE TREATMENT

Transitory anxiety
Use short-acting benzodiazepines for procedure related anxiety or panic attacks. (See Chapter 20: Pain, section 20.1.2: Procedural sedation and analgesia.)
However; increase utilisation of non-pharmacological strategies to manage procedural pain to decrease recurrent experience of sedation that becomes disorientating and removes sense of control.
Short-term use of hypnotics for anxiety-induced insomnia may be useful but avoid longer-term use as this disrupts sleep-architecture.

Formal Generalized Anxiety Disorder
See Chapter 14: Paediatric Psychiatry, section 14.5.1: Generalised anxiety disorder.

- Fluoxetine, oral, 0.5 mg/kg/day.
  - Dose range: 20–40 mg daily.
  - Recommended average dose: 20 mg/day.
  - Consider citalopram if fluoxetine is not tolerated or is ineffective.

- Citalopram, oral, 0.4 mg/kg/day.
  - Dose range: 5–40 mg daily.
  - Recommended average dose: 10–20 mg/day.

Death anxiety
Best managed with open communication, however, with fear of sleeping, short-term use of benzodiazepines at night may be warranted.

- Benzodiazepine, e.g.:
  - Diazepam, oral, 8 hourly.
    - If > 2–12 years: 2–3 mg.
    - If > 12–18 years: 2–10 mg.

21.1.3.2 DEPRESSION

F32-34

DESCRIPTION

Low mood in children and adolescents who become aware of their impending death is often congruent and appropriate and does not always warrant treatment. However, around 10 to 30% of chronically ill children are depressed. DSM 5 criteria instruct clinicians to exclude “symptoms that are

clearly due to a general medical condition" BUT this may lead to an under diagnosis of depression in this population.

GENERAL AND SUPPORTIVE TREATMENT

» Exclude and treat unaddressed pain.
» Communicate openly about diagnosis and prognosis.
» Address commonly occurring non-illness stressors (educational, familial, social and financial).
» Provide supportive counselling: opportunities for open communication about fears and worries.
» Cognitive behavioural therapy plus peer group support.
» Provide creative outlets: music, art and writing.
» Legacy work (memory making) in children nearing end of life.

MEDICATION AND TREATMENT

Mild or congruent depression does not always need to be medicated and is best addressed with non-pharmacological measures as above.
Moderate to severe depression may need to be treated. Medication should never be prescribed alone without counselling interventions.
SSRIs are generally the mainstay of treatment of depression in this population.

See Chapter 14: Paediatric Psychiatry, section 14.4.1: Depression in childhood and adolescence.

Although fluoxetine is usually first-line treatment for depression in children without other medical illnesses, citalopram may be safer in palliative care patients as it has fewer interactions (esp. involving cytochrome p450 enzyme) with other drugs (including chemotherapy). Citalopram also has the added benefit of being helpful if there is co-existent anxiety. Note that it takes 4 to 6 weeks for SSRIs to achieve therapeutic effect.

- Citalopram, oral, 0.4 mg/kg/day.
  - Dose range: 5–40 mg daily.
  - Recommended average dose: 10–20 mg/day.

21.1.3.3 DYSTONIA/MUSCLE SPASMS/SPASTICITY

DESCRIPTION

Dystonia

A movement disorder caused by diseases of the basal ganglia, in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements as well as abnormal posturing that is often painful.
Dystonia can be a primary disorder (e.g. brain tumours, metabolic conditions, neurodegenerative disorders and demyelinating diseases or, secondary to

diseases (e.g. TB meningitis) and injury (e.g. hypoxic or traumatic brain injury) of the CNS. Acute dystonia may also be drug induced.

Muscle spasms and Spasticity

Spasticity is velocity-dependent increased resistance to stretch observed in upper motor neuron damage. The increased tone can cause muscle injury resulting in pain secondary to inflammation. Intermittent muscle spasms may also occur secondary to pain, acute illness, positioning or constipation.

GENERAL AND SUPPORTIVE TREATMENT

» Prevent secondary trauma that can be caused by the child hurting herself during dystonic movements – this can be achieved by padding cot sides or nursing on the floor.
» Do not restrain the child with dystonia.
» A weighted blanket may help.
» If feeding is problematic and there is a risk for aspiration, consider naso-gastric or feeding gastrostomy.
» Physiotherapy and massage.
» A warm bath may help unlock spasms.
» Supportive counselling for parents and caregivers as this can be very distressing for them, especially if it is difficult to control.
» Identify and treat triggers for muscle spasticity, especially pain.
» Involve physiotherapists and occupational therapists for exercises, splints and positioning to prevent contractures.

MEDICATION AND TREATMENT

If drug dystonia suspected, discontinue offending medication.

- Diazepam, oral, 12 hourly.
  - Child 1–11 months: Initial dose of 250 mcg/kg twice a day.
  - Child 1–4 years: Initial dose of 2.5 mg twice a day.
  - Child 5–11 years: Initial dose of 5 mg twice a day.
  - Child 12–17 years: Initial dose of 10 mg twice a day. Maximum total daily dose of 40 mg.

For pain:

- Ibuprofen, oral, 5–10 mg/kg 8 hourly.

If severe, use morphine short-term:

- Morphine, oral [Immediate release morphine (liquid).]
  - Starting dose:
    - If 0–1 month: 50 mcg/kg/dose 6 hourly.
    - If > 1–12 months: 100 mcg/kg/dose 4 hourly.
    - If > 12 months: 200–400 mcg/kg/dose 4 hourly.

Neuropathic pain and/or neuro-irritability is common in ‘evolving cerebral palsy’, refer to tertiary level for consideration of a gabapentinoid.

REFERRAL

» To Paediatric Neurology where dystonia is difficult to manage to consider use of other dystonia medications (such as carbidopa/levodopa) and to pain specialists for unresolved neuro-irritability (to consider gabapentin). Also see Chapter 20: Pain Control.

21.1.3.4 INTRACTABLE SEIZURES

G41

For status epilepticus, see Chapter 13: Central Nervous System, section 13.3.

DESCRIPTION

Intractable seizures in palliative care refer to seizures that do not respond to treatment and often occur at the end of life in children with severe CNS pathology who are not candidates for ventilation. These may range from repetitive self-limiting seizures that do not cause distress to constant fitting not responsive to treatment. Causes include brain tumours, neuro-degenerative diseases, inborn errors of metabolism, severe congenital structural malformations including migrational disorders, and brain damage secondary to trauma or hypoxia.

GENERAL AND SUPPORTIVE TREATMENT

» Counselling support for parents, caregivers and professional care-giving staff.
» Reassurance if seizures are brief and not causing any obvious distress.
» Prevent secondary trauma by padding the side of the bed/cot or nursing on the floor.
» Do not restrain the patient as it may make seizures worse.
» Nurse on the side to minimize aspiration.
» If at end of life, stop feeds and follow guidance as per section 21.3: End of life and Terminal care.

MEDICINE TREATMENT

These children have often already been treated as per status epilepticus protocols but have not responded to standard treatment.
In children at the end of life or with poor prognosis, where admission to ICU for thiopentone infusion is not recommended, the mainstay of treatment is midazolam infusion to provide terminal sedation. (Consider subcutaneous infusion if no IV access is available.)

- Midazolam by SC or IV infusion over 24 hours for seizure control at end of life:
  - Neonate to 18 years: Initial dose: 1–3 mg/kg/24 hours increasing up to 7 mg/kg/24 hours (maximum 60 mg/24 hours or 150 mg/24 hours in specialist units for patients with refractory epilepsy).

- Morphine SC or IV infusion at 10–20 mcg/kg/hour titrated upwards in 30% increments against pain.

REFERRAL

» Seek specialist advice and consider addition of other agents.

21.1.4 DERMATOLOGICAL SYMPTOMS

21.1.4.1 PRURITUS

R29.9

DESCRIPTION

An unpleasant cutaneous sensation that provokes the desire to scratch. Can interfere with sleep and activities of daily living. Scratch marks may lead to skin excoriation and secondary infections. May be difficult to control with incurable conditions. Commonly seen in cholestasis, renal failure, burns, Hodgkin's lymphoma and occasionally as a side effect of opioids.

GENERAL AND SUPPORTIVE CARE

» Keep nails short and cover fingers with mittens but do not restrain.
» Keep skin moist with aqueous cream and emollients.

MEDICINE TREATMENT

Treatment depends on the underlying cause:

Cause	Mechanisms	Treatment
Opioid related	Stimulation of mu opioid receptors. NOT histamine mediated.	• Opioid switch if possible. • Ondansetron
Uraemia (Chronic renal failure)	Several factors including dry skin, uraemic toxin accumulation, neuropathy.	• Non-sedating antihistamines have NOT been shown to be effective. • Refer to tertiary level for consideration of a gabapentinoid.
Cholestasis	Bile acids in skin.	• Phenobarbitone
Burn wounds	Post burn during healing phase: caused by peripheral sensitisation.	• Refer to tertiary level for consideration of a gabapentinoid. • Sedating antihistamines. • Non-sedating antihistamines (cetirizine). • Ondansetron

LoE II[2]

Corticosteroid creams should not be used for generalized pruritus unless associated with inflammatory conditions (e.g. atopic eczema) as they will result in skin thinning and damage.

REFERRAL

» Severe pruritus not responding to standard treatment.

21.1.4.2 MALODOROUS FUNGATING WOUNDS/TUMOURS

DESCRIPTION

Non-healing fungating tumours that are often secondarily infected and smelly causing social ostracization and distress to child and family. Examples include exophytic retinoblastoma, infected bedsores, rhabdomyosarcoma, osteosarcoma or Kaposi's sarcoma.

GENERAL AND SUPPORTIVE MEASURES

» Supportive counselling.
» Set realistic goals: may not include wound healing but could include odour eradication.
» Regular wound cleaning and dressing changes.
» Adequate ventilation.
» Disguise smell by placing a bowl of vanilla essence in the room, burn incense or place kitty litter under the bed to absorb smell.
» Air-fresheners and perfumes do not work.
» Change bedding and clothing regularly.

MEDICINE TREATMENT

- Provide good procedural pain management (see Chapter 20: Pain Control) and use distraction/relaxation techniques before and during dressing changes.
- Irrigate wounds with warmed normal saline. Gentle debridement with gloved hand, not sharp instruments.
- Consider formal surgical debridement in a patient who still has some life expectancy.
- Topical metronidazole:
  - Irrigation and cleaning of wound: 2 L of saline combined with 13 crushed metronidazole 400 mg tablets (2 L 0.9% sodium chloride: 5200 mg metronidazole).
  - Metronidazole crushed tablet topical: Crushed metronidazole tablet 400 mg per 35 cm^2 area twice daily to ameliorate malodour.
- Activated charcoal dressings also help to absorb odours.
- For wound pain consider using topical anaesthetics such as lidocaine/prilocaine.

21.2 PAEDIATRIC PALLIATIVE CARE EMERGENCIES

21.2.1 MUCOSAL BLEEDS

DESCRIPTION

Massive bleeds at the end of life, although rare, can occur in several conditions and include:

» Haematemesis (e.g. bleeding varices, fulminant liver failure).
» Epistaxis (in haematological malignancies).
» Haemoptysis (rare in children).
» Tumour related erosion of blood vessels (less common in children).

GENERAL AND SUPPORTIVE CARE

» If a major bleed is anticipated it is best to be prepared, warn the family about the possibility that this may happen and ensure that they have what they need to manage an emergency: consider providing emergency home packs.
» Major bleeds are often preceded by smaller bleeds.
» In haematological causes associated with thrombocytopenia, platelet transfusions may be used to prevent bleeds up to a point where these no longer work, have to be given frequently or are in short supply. Repeat red blood cell transfusions at the end of life are costly, may not improve quality of life and could postpone dying.
» Have aprons and gloves ready.
» Use dark towels or green surgical sheets to reduce the visual impact of the blood.

MEDICINE TREATMENT

See Chapter 17: Ear, Nose and Throat, section 17.4: Epistaxis.
See Chapter 2: Alimentary Tract, section 2.3.3.1: Bleeding oesophageal varices.

For small mucosal bleeds:

- Topical treatment: crushed tranexamic acid tablets or gauze soaked in 100 mg/mL solution for injection.

Bleeding wounds:

- Use topical adrenalin 1:1000 mL on gauze and apply directly to the wound.

In massive bleeds associated with hypotensive shock, consciousness is lost and family/caregiver distress becomes the focus of attention.
With slower but large distressing bleeds occurring at the end of life, rapid sedation with benzodiazepines may be required using available routes including IV, SC and rectal (see terminal sedation under end of life care).

21.2.2 SPINAL CORD COMPRESSION

DESCRIPTION

Loss of neurological function in limbs (usually lower) caused by spinal compression from tumour or other disease process. Should be managed as an emergency in an attempt to delay complete loss of function for as long as possible.

Caused in children by intramedullary or intradural metastases or extradural compression (e.g. vertebral collapse).

Early signs:

» Change in nature of long-standing pain.
» Neuropathic pain radiating down legs.
» Positive Lhermitte's sign: electric shock-like pain on neck flexion.

Late signs:

» Limb weakness, sensory deficits – level detected, decreased to absent reflexes, paraplegia.

GENERAL AND SUPPORTIVE MEASURES

» Early detection in high-risk patients.
» Once established manage as per paraplegia with careful attention to pressure support as well as bowel and bladder care (intermittent catheterisation).

MEDICINE TREATMENT

Commence high dose dexamethasone:

- Dexamethasone, IV:
  - Loading dose of 1–2 mg/kg followed by 0.25–0.5 mg/kg every 6 hours has been suggested.

REFERRAL

» Palliative radiotherapy.

21.2.3 RESPIRATORY PANIC

DESCRIPTION

Sudden onset of severe respiratory distress sometimes experienced with an end of life event such as pulmonary embolus, pneumothorax, pulmonary oedema, SVC syndrome, upper airway obstruction, and bleed into CNS tumour.

GENERAL AND SUPPORTIVE CARE

» Remain calm and provide reassurance.

» Position in semi-fowlers position.
» Gentle suctioning.
» Supplemental oxygen.

MEDICINE TREATMENT

If no IV line, use buccal midazolam and morphine:

- Midazolam: 500 mcg/kg and morphine 100 mcg/kg repeating every 10–30 minutes as needed until child settles or subcutaneous infusion is commenced.

21.3 END OF LIFE AND TERMINAL CARE

Z51.1

DESCRIPTION

The end of life phase is defined as that stage where it is recognized that the child's health is in a state of steady decline, whereas the terminal stage is the last few hours of life.

Although prognostication is more difficult in children compared to adults especially when it comes to non-malignant conditions, the following may serve as indicators that prognosis is becoming more limited:

» Increasing frequency or duration of hospitalisations.
» More severe disease-related complications with downward drifting baseline.
» Decreasing response to disease modifying treatments.
» Decreased duration of benefits of transfusions (especially when these benefits are shorter than the expected lifespan of the blood product).
» Increasing fatigue and prolonged periods of sleep.
» Anorexia and cachexia.
» Decreasing urine and stool output.
» Child less interactive, becoming increasingly withdrawn and less interested in surroundings.
» Deteriorating physiological parameters.
» Increasing anxiety or agitation or in some children increasing peace and serenity and actively seeking out of opportunities for legacy making.

GENERAL AND SUPPORTIVE MEASURES

» Advance care planning:
  > Is a process of discussions between families and healthcare providers about preferences for care (including place), treatments and goals in the context of the child's current and anticipated future goals.
  > Should include information on resuscitation status and documentation of decisions on limitations of interventions.
  > Should outline plans for anticipated complications/end of life events

before they arise.

» Symptoms often intensify at the end of life and treatment should be continued and intensified as needed. Alternative routes for administering medications should be sought in children who lose the capacity to swallow: these include the buccal and rectal routes, naso-gastric tubes, transdermal patches and the subcutaneous route.
» Parents, caregivers and siblings should be actively counselled and prepared for the child's death.
» Try limit hospital admissions or reduce the duration of hospital stays where family have the capacity to care for their child at home.

21.3.1 TERMINAL CARE

Indications for inpatient hospital or hospice inpatient terminal care:

» Hypoxia and respiratory distress where oxygen therapy provides relief and not already available at home.
» IV/naso-gastric fluid requirements or medication administration needed to relieve suffering.
» Carer/s unable to cope at home.
» Symptom control needing to be intensified or provided intravenously.

Feeds and fluids at the end of life:

» Anorexia and refusal of feeds/fluids in dying patients is a normal phenomenon and not an indication for naso-gastric feeds or intravenous fluids as these may prolong dying.
» Encourage the family to 'feed for comfort only' and reassure them that the dying child is not hungry.
» Decreasing fluid intake is helpful for the dying process: decreases excessive secretions, urination, pulmonary and cerebral oedema.
» Ketosis is also beneficial in that is suppresses appetite and also aids release of endorphins (natural pain killers).

Investigations at the end of life:

» Investigations should be kept to a minimum and only done if it is believed that doing these will shorten the duration of hospital stay or in some way contribute to the child's comfort.
»

MEDICINE TREATMENT

Antibiotics at the end of life:

- Oral antibiotic therapy may be started, where it is thought that a course of antibiotics could shorten the duration of discomfort or hospital stay.
- Non-treatment of a terminal pneumonia (a common end of life event) is an acceptable palliative care practice.

Medication at the end of life:

- Stop all unnecessary medications (e.g. multivitamins, TB treatment, ART) that are not contributing towards symptom control and adding to pill burden in an actively dying child.
- Continue medications that if discontinued could cause distressing symptoms (e.g. calcium, anti-failure medications).
- Consider use of alternative routes of administration of symptom control if not able to take orally.

Management of terminal agitation or restlessness:

» Exclude unmanaged pain.
» Exclude urinary retention.
» Check pressure sites for bedsores.

- Midazolam
  - By IV/SC infusion: 250–1500 mcg/kg/24 hours as starting dose, titrating upwards against symptoms and sedation, 10–60 mcg/kg/hour.

REFERRAL

Discuss with a specialist:

» Children with symptoms not described here.
» Children not responding to management.

References

[1] Loperamide dose: South African Medicines Formulary (SAMF), 12th Edition. Division of Pharmacology, Faculty of Health Sciences, University of Cape Town. 2016. Association for Paediatric Palliative Medicine Formulary available from URL.
https://www.appm.org.uk/_webedit/uploaded-files/All%20Files/Event%20Resources/2020%20APPM%20Master%20Formulary%202020%20protected.pdf

[2] A comparative analysis of cetirizine, gabapentin and their combination in the relief of post-burn pruritus. Burns Volume 37, Issue 2, 2011, Pages 203-207.