

MANAGEMENT OF COMMON SYMPTOMS AND PROBLEMS IN PAEDIATRIC PALLIATIVE CARE

CLINICAL GUIDELINES



FORWARD:

“To cure sometimes, to relieve often, to comfort always”

Hippocrates,

It is with great joy that I write this forward to the long awaited first edition of a separate Paediatric Palliative Care Section to the Hospice and Palliative Care Associations Clinical guidelines.

Whilst we recognise that children are unique and should not be treated “like small adults” we need also to acknowledge that the general principles and philosophies of palliative care for children are shared with those of adult palliative care. The WHO definition talks of children’s palliative care as being unique but also closely related to adult care.

Unfortunately fears around providing palliative care for children (particularly wrt drug doses) as well as the perceived high emotional burden of caring for them has led to many children not receiving the care that they need. This has resulted in many children dying in less than ideal circumstances often in busy acute care hospital facilities or without support at home.

It is the hope of the Bigshoes Foundation’s hospital based paediatric palliative care teams and the Hospice Palliative Care Association of South Africa that these guidelines will increase the confidence of all health care professionals in all settings (from the tertiary hospital to the child’s own home) to provide a continuum of quality palliative care to children in need. These guidelines were put together by Bigshoes’ teams, edited by myself and reviewed by Dr Khaliah Johnson. Dr Johnson is a Paediatrician from the USA who has just completed her fellowship in Palliative care at the Children’s Hospital of Philadelphia (CHOP). It was interesting to hear from Khaliah that even in the well-resourced USA there are insufficient numbers of child-focused hospices to meet the palliative care needs of children and that adult hospices in the States are increasingly taking on the challenge.

South Africa has a shortage of Paediatricians and also paediatric trained nurses especially in more rural areas and the general practitioner and nurse has a vital role to play in the provision of palliative care for children.

A child psychologist in Johannesburg when being introduced to palliative care for children for the first time said:

“Because these children are living with life limiting and life threatening illnesses, bad things are going to happen to them whether we are ready or not. If we can do but any small thing to help make their (child and family) journey a little easier, we have done a great thing”.

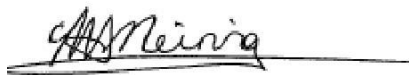
I would like to encourage all of those providing palliative care to adults to extend their provision of services to children and all those professionals skilled in caring for children to increase their skills in providing palliative care to their own patients in partnership with the hospice and home-based care networks.

As a well know African proverb says about journeys: "If you want to go quickly, go alone. If you want to go far, go together."

The distribution of these paediatric guidelines for the first time at the Hospice and Palliative Care Association's 25th anniversary conference is exciting as in keeping with the theme of the conference (celebrating partnerships), these guidelines celebrate the result of past partnerships and the promise of new ones.

Further help along the can be sourced from the South African Children's Palliative Care Network -an initiative of the South African Children's Palliative Care Alliance between the Hospice Palliative Care Association of South Africa, the Bigshoes Foundation and the International Children's Palliative Care Network (ICPCN). Further web-based resources for children's palliative care can also be found at www.baobabppc.org.za

I would like to sincerely thank everyone who contributed to the writing of these guidelines and trust that they will be helpful to relieve the suffering of some of the estimated 830 000 children in South Africa who may be in need of palliative care.



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Disclaimer:

In composing these guidelines, the authors have been diligent in their efforts to verify its content through the date of its publication (August 2012). However, it is important to note that drug dosing, uses, side-effects, and contra-indications change over time. All clinicians prescribing from these guidelines should take the time to ensure that drug information is consistent with the manufacturer's current recommendations. Prescribers must also use their own discretion in advising routes of administration that may be common practice in palliative care, but unlicensed by the manufacturer.

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PAEDIATRIC PALLIATIVE CARE DEFINED

The World Health Organisation (WHO) states that although paediatric palliative care is closely related to adult palliative care, it has its own special unique characteristics. WHO's definition of palliative care is appropriate for children with life threatening and life limiting chronic illnesses.

WHO defines Paediatric Palliative Care as the active total care of the child's body, mind and spirit, and also involves giving support to the family. It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease (Fig 1). Health 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, healthcare providers with varied areas of expertise, and 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 the child's own home. Other definitions include a focus on the enhancement of quality of life, the provision of respite and bereavement support.

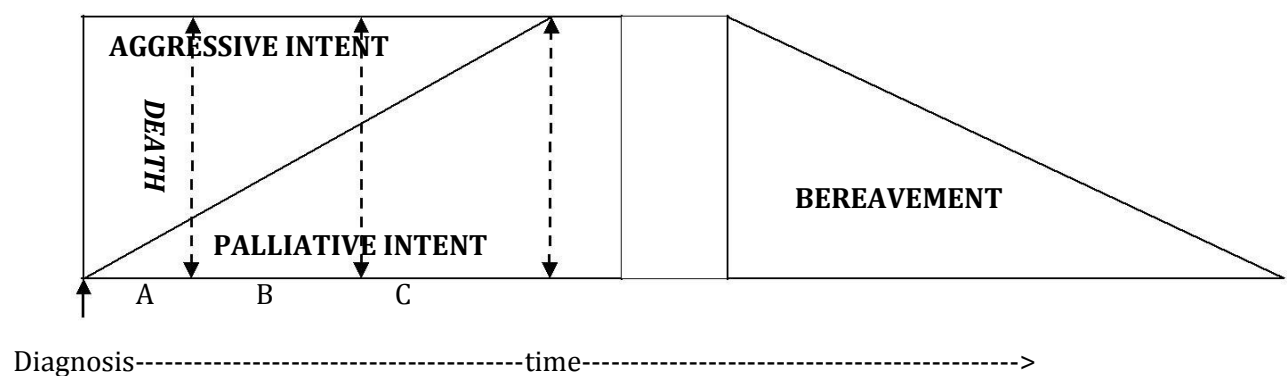


Fig. 1. Modern integrated palliative care services model (Frager 1997).

The integrated model of palliative care proposes that palliative care be introduced from the time of diagnosis. As disease advances (points A, B, C) the amount of aggressive cure-focused treatment decreases and the amount of palliative-focused treatment increases.

Understanding the disease trajectories for different conditions helps to guide decision making in terms of balancing 'active treatment' with 'palliative care'. Together for Short Lives (UK) has recognized four categories of children who would benefit from a palliative care approach. These categories are defined largely by their disease trajectories (Table 1).

Table 1. Categories of life-limiting conditions

- Group 1:** Life-threatening conditions for which curative treatments may be feasible but can fail, e.g. cancer (acute lymphoblastic leukaemia), reversible organ failure.
- Group 2:** Conditions where premature death is inevitable, where there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities, e.g. HIV/AIDS on anti-retrovirals, cystic fibrosis.
- Group 3:** Progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years, e.g. many syndromes, inborn errors of metabolism, muscular dystrophy.
- Group 4:** Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death, e.g. severe cerebral palsy, spinal cord insult.

Similarities and differences between adult (APC) and paediatric palliative care (PPC).

Similarities	Differences
Both focus on improving quality of life by relieving distressing symptoms. Specific attention is given to relieving and preventing suffering at the end of life.	Death is “not a normal process” for most children. This presents unique challenges in the psychosocial and bereavement care of children and families facing serious illness.
Both provide support to families in addition to the patient, including bereavement support.	Children are not small adults and professionals caring for them should be well versed in aspects of paediatric care. (including care of families of paediatric patients and especially child siblings).
Palliative care involves assisting with medical decision making and defining goals of care throughout illness and end of life.	Medical decisions in young children are usually made by their carers whereas the adult patient may make his/her own decisions (unless he/she is unable to).
Both recommend a multi-disciplinary approach, including physicians, nurses, social workers, and chaplains.	The pediatric palliative care multi-disciplinary team should ideally also involve other child-caring professions including teachers and play therapists (where available).

UNIQUE CHARACTERISTICS OF PPC:

The “paediatric population” covers a wide spectrum of ages (birth to 18 years). At each stage of childhood there are different developmental needs and capabilities. Children have distinct educational needs and perceptions of illness, death and dying that are influenced by their developmental level and cultural traditions.

Diseases of childhood are complex and diverse in a way that differs from adult illness. The spectrum of diseases includes congenital anomalies, genetic syndromes, inborn errors of metabolism, and other rare conditions not seen in adults. Often children with these conditions do not survive into adulthood.

Symptoms can present differently in children and symptom assessment can be more challenging, especially in pre-verbal children. Medications and dosages are more complex in children, as dosing is largely weight-based and side effects may differ.

Paediatric palliative care can be more emotionally draining on family and staff than adult palliative care.

References:

1. WHO definition of Paediatric Palliative Care. Available from URL:
<http://www.who.int/cancer/palliative/definition/en/> (accessed July 2011).
2. Association for Children with Life Threatening Illnesses (ACT) Definition of Palliative Care for Children. Available from URL
<http://www.act.org.uk/page.asp?section=59§ionTitle=What+is+children's+palliative+care> (accessed July 2011).

BASIC PRINCIPLES OF SYMPTOM CONTROL IN CHILDREN

“The essence of palliative care is the relief of suffering.”

Dr Derek Doyle

Symptom control is the essence of the clinical practice of palliative care. Even when a child's underlying disease cannot be cured, there is “always something that can be done.” The child and his/her families' distress can be relieved by focusing on assessing and managing the symptoms that are contributing to distress. Decreasing suffering in this way can improve quality of life for the child and provide comfort for the family.

Whilst pain is a major symptom present in many diseases (both acute and chronic), “symptoms other than pain” are equally as important and can cause just as much distress.

What is a symptom?

- A symptom is defined as “any sensation or change in bodily function that is experienced by a patient and is associated with a particular disease”.

What are ‘symptoms other than pain’?

- This is a commonly used grouping in palliative care for symptoms besides pain associated with diseases in all of the major organ systems. For example, cough and dyspnoea are grouped under respiratory symptoms.
- Remember that symptoms are subjective and are reflective of the patient's experience of the disease.
- Symptoms should not be confused with signs that are actual physical indicators (mostly visible, sometimes palpable or percussible) of the presence of disease.

CHALLENGES TO SYMPTOM CONTROL IN CHILDREN:

- **Challenges related to the child:**
 - Pre-verbal and non verbal children (e.g., mentally retarded children) are not able to report verbally on their experience of a symptom. This requires health care workers to use other indicators to evaluate distress that may be present as a result of underlying pathology.
- **Challenges related to the child's caregiver:**
 - Caregivers may both under or over report on a symptom.
 - Caregivers may assign significance to a symptom based on their association or understanding of that symptom that does not necessarily reflect the true situation.

- In the case of children looked after by multiple caregivers subtle or early symptoms may be missed or overlooked.
- **Challenges related to the health care worker:**
 - Health care workers may be more focused on the disease that the child has rather than the suffering that it is causing and dismiss symptoms as insignificant when they are in fact causing considerable distress.
 - Health care workers may be reluctant to use symptom control drugs for fear of their side effects.
 - Child health trained practitioners spend a lot of time learning that treatment of acute self limiting symptoms is not warranted and may even be dangerous (e.g., giving immodium with acute gastro or cough suppressants for an acute upper or lower respiratory tract infection) and they may apply this principle even to chronic distressing symptoms.
- **Challenges related to drugs necessary for symptom control**
 - Symptom control drugs can have side effects.
 - Many are not licensed for use in children (esp under 2 years) and are sometimes used “off-code” in palliative care.
 - Suitable paediatric formulations (eg syrups) of many symptom control drugs are not available.

KEY PRINCIPLES IN SYMPTOM CONTROL:

Irrespective of what the symptom is there are certain key principles that can be applied to the management of any distressing symptom. These are:

KEY PRINCIPLES OF SYMPTOM CONTROL:

1. Determine and treat the underlying cause of the symptom including non-physical causes.
2. Relieve the symptom without creating new symptoms or unwanted side effects.
3. Consider different types of interventions: drug and non-drug interventions.
4. Consider whether the treatment is of benefit to the individual patient (weigh the advantages and disadvantages of therapy).

GENERAL PRINCIPLES OF PAIN MANAGEMENT IN CHILDREN

DEFINITION OF PAIN:

Pain is an unpleasant sensory or emotional experience associated with actual or potential tissue damage, or described in terms of such damage".

Merskey: International Association for the study of pain.¹

- As pain is a subjective experience the gold standard of assessment is self-report. Unfortunately many children are unable to talk about their pain, either because they are pre-verbal or impaired by disability or symptoms.
- Even though pre-verbal or non-verbal children may not be able to talk about their pain, they are still able to communicate in a "unique and effective manner through their bio-behavioural responses".
- Most human beings (especially mothers) have an instinctive ability to "pick up" on pain signals (usually facial gestures, tone of cry, behavioural responses) in their infants. This has a survival advantage in that it mobilizes care-giver behaviour that meets the infant's physical and emotional needs.

ASSESSMENT OF PAIN IN CHILDREN:

- Globally pain in children is inadequately assessed and undertreated.
- Pain is so prevalent that some recommend it become the fifth vital sign measured alongside BP, pulse, temperature and respiratory rate.
- Although it may be more difficult to assess pain in children than adults, common sense should prevail: **if the injury, illness or procedure would cause pain in an adult it will be painful to a child.**

QUESTT principle to assess paediatric pain:

- **Q**uestion the child and parent/caregiver
- **U**se pain rating scales
- **E**valuate behaviour, physical findings, and physiologic changes
- **S**ecure the parent's/care giver's involvement
- **T**ake the cause of the pain into account
- **T**ake action and evaluate results

Questioning a child/parent on pain:

- **P:** Precipitating / Palliating / Provoking factors
- **Q:** Quality / Quantity
- **R:** Region / Radiation / Related factors
- **S:** Severity
- **T:** Time course

Using pain rating scales:

- These are useful for establishing a baseline and for measuring response to treatment.
- Different scales have been developed for different ages and levels of development in both non-verbal and verbal children.

1. NEONATAL PAIN RATING SCALE:

**Instructions for use*

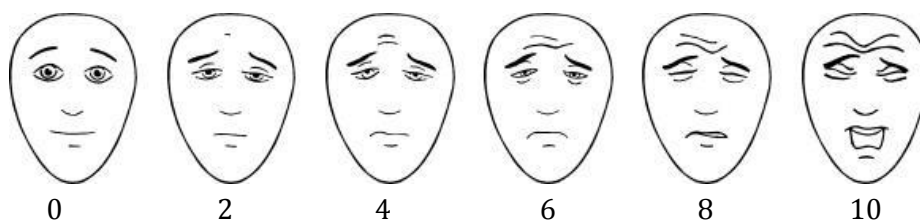
Facial expression		Score
0- Relaxed muscles	Restful face, neutral expression	
1- Grimace	Tight facial muscles, furrowed brow, quivering chin, tight jaw	
Cry		
0 -No cry	Quiet, not crying	
1-Whimper	Mild moaning, intermittent	
2-Vigorous cry	Loud scream, rising, shrill, continuous	
	Note: silent cry may be scored if baby is intubated and ventilated as evidenced by obvious mouth and facial movements	
Breathing pattern		
0-Relaxed	Usual pattern for infant	
1-Change in breathing	Labored, irregular, faster than usual, gagging, breath holding	
Arms		
0-Relaxed	No muscle rigidity, occasional random movements of arms	
1-flexed, extended	Tense, straight arms; rigid and/or rapid extension/flexion of arms	
Legs		
0-Relaxed	No muscle rigidity, occasional random movements of legs	
1-flexed, extended	Tense, straight legs; rigid and/or rapid extension/flexion of legs	
State of arousal		
0-Sleeping/awake	Quiet, peaceful or alert, random leg movement	
1-Fussy	Alert/restless and thrashing	

2. FLACC SCORE: For non-verbal children or children < 3 years

Use like an Apgar score: evaluating each item and arriving at a total score/10.

Item	0	1	2
Face	No particular expression or smile	Occasional grimace or frown, withdrawn disinterested	Frequent to constant frown, clenched jaw, quivering chin
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking, or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting back and forth, tense	Arched, rigid or jerking
Cry	No cry (awake or asleep)	Moans or whimpers, occasional complaint	Crying steadily, screams or sobs, frequent complaints
Consolability	Content, relaxed, no need to console	Reassured by occasional touching, hugging or "talking to", distractible	Difficult to console or comfort

3. REVISED FACES PAIN SCALE:



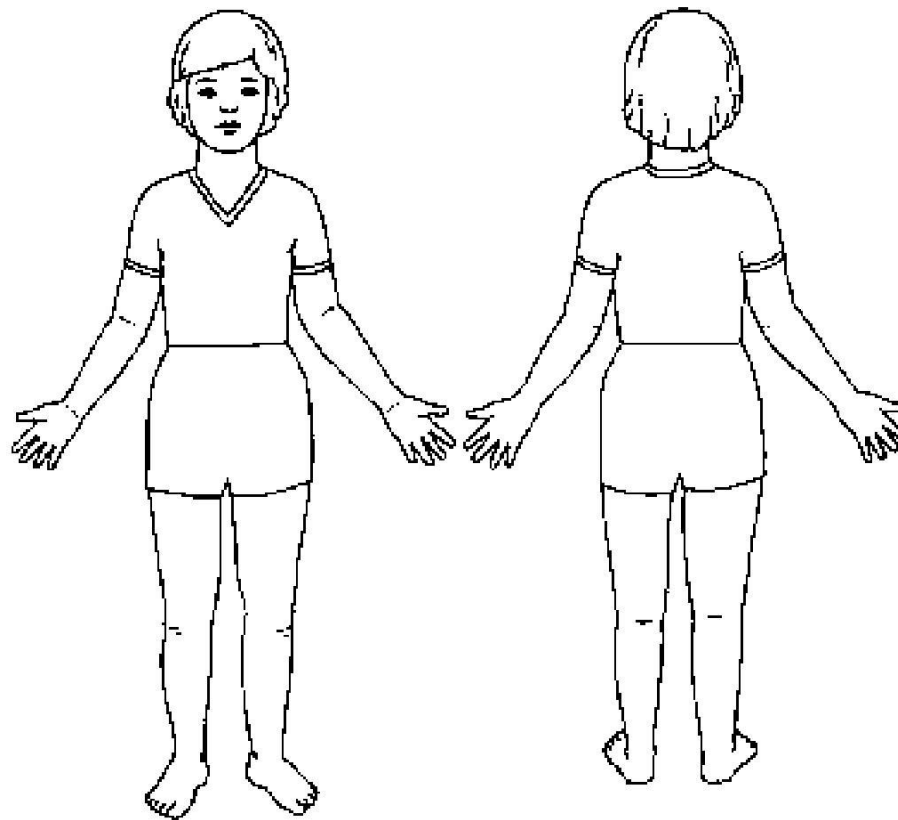
- Use in children over 4 years
- Ask them to point to the face that best depicts their level of pain

4. Numeric/word pain scale:

										I
0	1	2	3	4	5	6	7	8	9	10
No pain		Little pain		Medium pain		Large pain				Worst possible pain

- Use in children over 7 years
- Ask them to choose the number and word description that best reveals their level of pain

5. Eland body tool:



NO PAIN



MILD PAIN



MODERATE PAIN



SEVERE PAIN

Instructions: get child to assign colours to no pain (eg green), Little pain (eg yellow), Moderate pain (eg orange) and severe pain (eg red).

Ask them to colour in the bodies using the different colours to depict different levels of pain in different areas.

Evaluate behaviour and physiologic changes:

PAIN RELATED BEHAVIOURS AND PHYSICAL FINDINGS IN CHILDREN

Age	Behavioural indicators of pain in children of different ages	
	ACUTE PAIN:	CHRONIC PAIN:
Newborns	<ul style="list-style-type: none"> • Crying and moaning • Muscle rigidity • Flexion and flailing • Irritability • Guarding • Reflex withdrawal to painful procedures 	<ul style="list-style-type: none"> • Apathy • Lack of interest in surrounding • Irritability • Changes in sleeping and eating patterns
Toddlers and pre-schoolers	<ul style="list-style-type: none"> • Crying, screaming and vocalizing hurt • Facial expression: eye squeeze, brow bulge, open mouth, taut tongue, chin quivering, grimacing • Thrashing of arms and legs • Pushes away, withdraws limbs • Clings to parent/caregiver • Restless and irritable 	<ul style="list-style-type: none"> • Moody • unruly behaviour • decreased play • change in appetite • change in sleep pattern
School-aged child	<ul style="list-style-type: none"> • Crying • Muscle rigidity, clenched fists, white knuckles, clenched teeth, closed eyes • Stalling techniques when anticipating a painful procedure 	<ul style="list-style-type: none"> • school refusal • declining school performance • decreased socialisation • anxiety and depression • disturbed sleep
Adolescents	<ul style="list-style-type: none"> • Verbalisation of pain • Muscle tension • Loss of appetite • Insomnia or hypersomnia 	<ul style="list-style-type: none"> • depression and anxiety • low self esteem • decreased socialisation • drug/alcohol abuse • insomnia/hypersomnia

- Physiological responses to pain include increased pulse, raised BP, diaphoresis (perfuse sweating), pallor or flushing, decreased oxygen saturation, dilated pupils, increased tone, rapid shallow respiration and hyperglycaemia.
- Adaptation however occurs with ongoing pain and physiological manifestations may be absent in chronic pain.

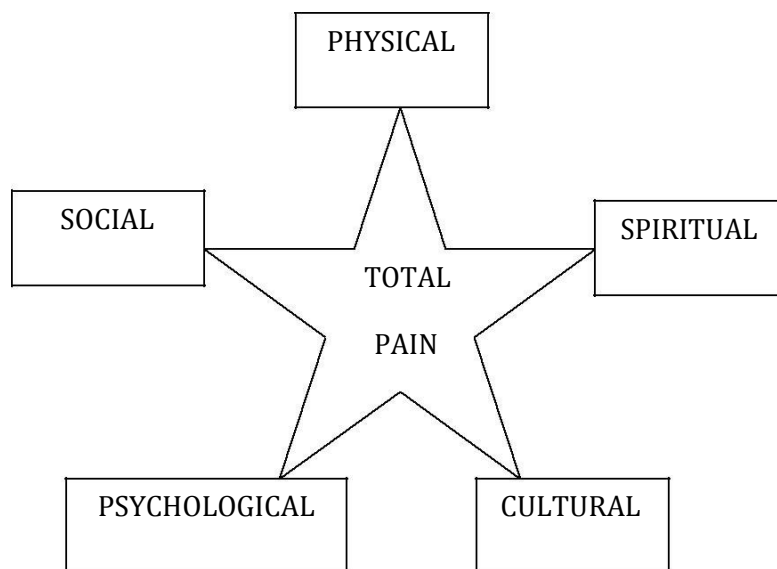
Securing the parent/caregivers involvement:

- Listen to mothers, fathers and carers; they know their child best and are more tuned to subtle changes in behavior.
- Include carers in decision making regarding symptoms management approaches. They often know what works best to comfort the child.

Take the cause of pain into account:

DETERMINING THE UNDERLYING CAUSE:

- Determining the type of pain helps to determine its treatment.
- Pain can be classified according to:
 - *Duration* eg: acute or chronic
 - *Underlying mechanism*: Nociceptive (visceral + somatic), neuropathic, sympathetic, psychogenic
 - *Situation*: incident pain, procedural pain, breakthrough pain
- *Pain is a "total experience" it is not just physical, it has psychological, spiritual, cultural and social components: this is known as "total pain."*



ACUTE PAIN	CHRONIC PAIN
Usually due to a definable acute injury or illness	Results from a chronic pathological process
Has a definite onset and its duration is limited and predictable	Has a gradual or ill-defined onset, continues unabated and may become progressively more severe; persists longer than the expected healing time for the injury or illness in question
Is accompanied by anxiety and clinical signs of sympathetic over-activity	The patient appears depressed or withdrawn -there may be no sympathetic over-activity and patients are frequently labelled as "not appearing to be in pain".
Treatment is directed at the acute illness or injury causing pain, with the short term use of analgesics	Treatment is directed at the underlying disease where possible, regular use of analgesics to relieve pain and prevent recurrence as well as psychological supportive care

SITUATIONAL PAIN:

- **Incident pain** occurs only in certain circumstances, such as after a particular movement or on standing. It should be regarded as chronic pain but, as it is intermittent, it is better managed with local measures where possible.
- **Breakthrough pain** is a transitory exacerbation of pain that occurs on a background of otherwise stable and controlled pain.
- **Procedural pain** is pain related to procedures /interventions. This is especially important in children with chronic illnesses (HIV and malignancies) and is an important cause of anxiety in children that can be prevented.

Characteristic	Nociceptive	Neuropathic	Sympathetic	Psychogenic
Cause	Caused by stimulation of sensory receptors by trauma or disease Normal neural pathways	Caused by an abnormal reaction to stimuli or spontaneous discharge by damaged nerves (neuropathy)	Caused by damage to sympathetic nerves	Usually in patient with evidence of psychopathology . Also called ' neuropathic pain of central origin ' Organic causes excluded
Characteristic	Somatic pain Superficial (cutaneous) arising from skin, subcutaneous tissues or mucous membranes; sharp and well localised pain Deep arising from muscles, tendons, joints, etc.; more diffuse and dull Visceral pain from organs, dull and poorly localised pain; often referred to cutaneous sites; may be associated with autonomic responses (e.g. sweating, nausea)	Burning pain (dysaesthesia) Shooting pain (lancinating) Aching sensation relieved by pressure applied to the affected area Allodynia non- painful stimuli are perceived as being painful	Burning pain and increased sensitivity (hyperaesthesia) Localised vasomotor instability (erythema, pallor, oedema) Trophic changes	Examples - Headache Muscle spasms Back pain Abdominal pain
Treatment	Non-opioids + opioids	Less responsive to opioids and non-opioids; often more responsible to adjuvant analgesics	Less responsive to opioids and non-opioids; responds well to regional sympathetic block	Psychotherapy Anti-depressants Non-narcotic pain killers

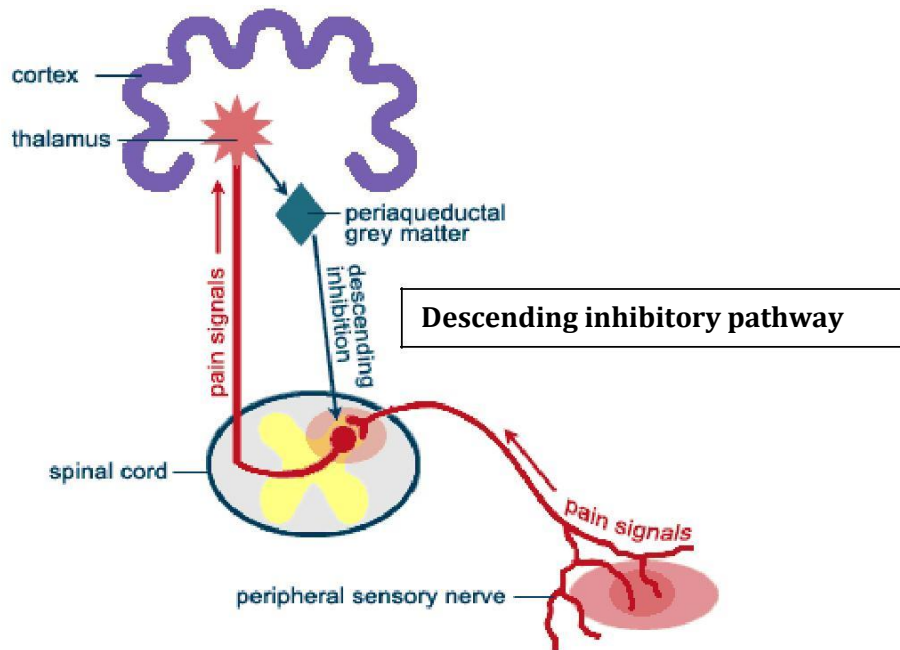
Take action and evaluate results:

- Assess pain → develop a treatment plan → reassess (use pain rating scales) → revise treatment plan
- Pain diaries are also helpful for constant re-evaluation in children with chronic pain.

**When in doubt as to whether pain is present or not, rather err on the side of treating.
There is seldom harm in a trial of analgesia.**

NON-DRUG (*non-pharmacologic*) MEASURES USED TO MANAGE PAIN:

- Non-drug measures can be used for both acute and chronic pain
- Distraction (blowing bubbles, counting) is commonly used during procedures (acute pain)
- Non drug measures are especially important in helping children to cope with chronic pain
- Non-drug measures work by increasing stimulation of the descending inhibitory pathway which decreases the amount of afferent signals received by the brain



- Non-drug measures are divided into counter-irritation and psychological methods:

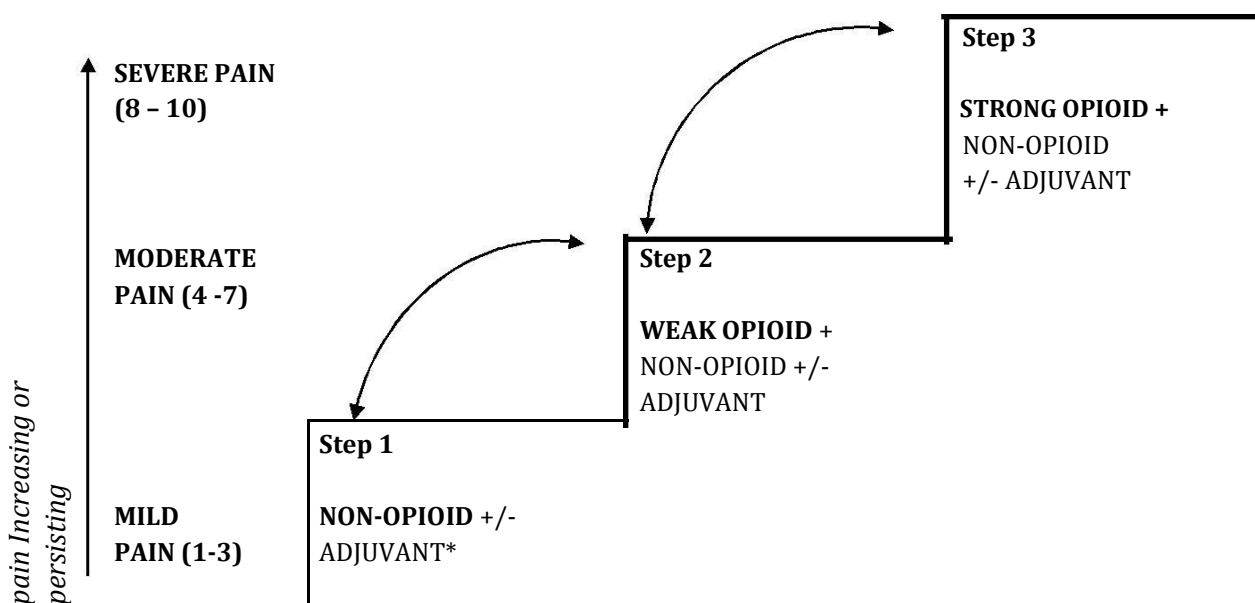
Counter-irritation	Psychological methods
Heat	Distraction
Cold	Imagery
Touch/Massage	Relaxation
<i>Transcutaneous electrical nerve stimulation (TENS)</i>	<i>Cognitive-behavioural therapy</i>
<i>Acupuncture</i>	Music therapy
Vibration	<i>Biofeedback</i>
Aromatherapy	Hypnosis

DRUG (pharmacologic) MEASURES TO MANAGE PAIN

- The broad principles of analgesic use in children (WHO):
 - By the clock (regular rather than prn dosing)
 - By the correct route for the type of pain (preferably oral, avoid IM injections)
 - By the child (individualize treatment)
 - By the WHO pain ladder

The correct use of the correct analgesic will relieve most pain in children.

THE WHO PAIN LADDER:



ADJUVANTS: Adjuvant analgesics are defined as drugs with a primary indication other than pain that have analgesic properties in some painful conditions.

Commonly used drugs in the ladder:

NON-OPIOIDS	WEAK OPIOID	STRONG OPIOID	ADJUVANTS
Paracetamol NSAID's (Ibuprofen, Diclofenac)	Codeine phosphate Tilidine (Valoron) Tramadol	Morphine MST Methadone Fentanyl	Prednisone Carbamazepine Amitryptaline Gabapentin

Important principles:

- It is preferable to use “pure drugs” rather than fixed combinations in children as these are easier to titrate. Increasing doses of fixed combination drugs has been associated with inadvertent Paracetamol overdose.
- Aspirin should not be used in children under 12 years because of its association with Reye syndrome (fatty infiltration of the liver and encephalopathy associated with a high mortality rate).
- International paediatric pain experts are now recommending that codeine no longer be used as the weak opioid of choice in children for pain. Bio-availability of codeine is very variable (15 – 80%) and up to 36% of children show inefficient conversion of codeine to morphine (the active metabolite of codeine). Low dose morphine can be used as an alternative to codeine phosphate where there are no alternative weak opioids.
- Tramadol has recently been added to the EDL for hospitals and primary health care clinics for use in adults in SA. Use in children is limited by unavailability of syrup formulation. Syrup can be prepared from crushed tablets as in the table to follow.
- **IM Pethidine is not recommended in children.**
- Weak and strong opioids should never be combined as they are competitive agonists at the same receptors. Combining a weak opioid with a strong opioid decreases the efficacy of the strong opioid. Stop the weak opioid before starting the strong opioid.

DOSES OF COMMONLY USED ANALGESICS IN CHILDREN:

Drug	Dose	Notes
Paracetamol	Oral: Loading dose: 20mg/kg po Then 10-15mg/kg/dose po 6 hourly Rectal: 30mg/kg stat then 20mg/kg every 4 – 6hours	Decrease dose and increase dosing interval to 8hourly in hepatic and renal impairment
Ibuprofen	5mg – 10mg/kg po 6-8hourly Max 30mg/kg/day in 3-4 divided doses Juvenile Rheumatoid Arthritis: 60mg/kg in 4 – 6 divided doses.	Give with food; avoid with asthma, low platelets, peptic ulcer disease and renal dysfunction
Codeine phosphate	0.5-1mg/kg 6hourly	No longer recommended See notes above
Tramadol	< 50kg: 1-2mg/kg/dose 4 -6hrly (max 8mg/kg/day) >50kg: 50 – 100mg 4 – 6hrly (max 400mg/day)	Syrup 5mg/ml can be made by crushing 6X 50mg tabs and adding to 30ml ora plus + 30ml ora-sweet solution
Tilidine (Valoron)	1mg/kg/dose po 6 hourly = 1 drop per 2.5 kg body weight	Contra-indicated in head injuries with raised ICP, asthma

Morphine sulphate	Oral: < 1 month: 1-12 months: 0.1mg/kg/dose po 4 hourly >12 months: 0.2-0.4mg/kg/dose po 4 hourly	Oral morphine can be constituted in different strengths:
Short acting	Intravenous injection: 0-3 months: 0.025mg/kg/dose 6 hourly. 3-6 months: 0.05mg/kg/dose 6 hourly 6-12 months: 0.1mg/kg/dose 4 hourly >12 months: 0.1 – 0.2mg/kg/dose 4 hourly	Oral: 5mg/5ml 10mg/5ml 20mg/5ml 100mg/5ml IV: 10mg/ml 15mg/5ml

HOW TO USE MORPHINE IN CHILDREN:

- Many myths and fear surround the use of Morphine. However, If used correctly morphine is a very versatile drug with no ceiling effect (a maximum dose above which no further analgesic effect is obtainable). It can be increased incrementally to provide more analgesic effect.
- There is a tendency for lay persons to associate morphine with “terminal care” and the “beginning of the end”. Although morphine is an excellent drug for terminal care it can be used at any stage of a disease to control pain and can be withdrawn if the patient no longer needs it.
- Parents worry that their child may become addicted to Morphine. True addiction does not develop in the management of true pain.
- Doctors worry about causing respiratory depression. If used correctly respiratory depression is rare. Pain itself is the physiological antagonist to the respiratory depressant effect of Morphine.
- **How to prescribe oral Morphine for pain:**
- Start with an oral starting dose of Morphine Sulphate syrup (short acting Morphine) as outlined in the preceding table. In infants the 5mg/5ml concentration is convenient as small doses (e.g., 0.2mg) can be given using an insulin syringe (1mg = 1ml).
- Morphine has a half life (if given orally) of 2-3 hours and should be prescribed 4 hourly. In patients with delayed clearance (newborns, hepatic and renal dysfunction) it can be prescribed 6 – 8 hourly.
- As per the WHO pain guidelines Morphine should be given regularly (by the clock) and not prn. Regular dosing controls pain better and ultimately results in lower doses than if given prn.
- If the patient experiences pain before the next dose (breakthrough pain) an extra “breakthrough dose” (BTD) of Morphine may be given. A BTD is 50 - 100% of the regular dose. Morphine takes up to 30 minutes to have effect so a BTD should not be given before 30 minutes.
- There are two ways to increase Morphine as required for pain:

1. Increase the regular dose by 30 – 50% of the previous dose if pain is not controlled.
 - Eg: a patient receiving 5mg of morphine 4 hourly could have his morphine increased to 6.5mg (+30%) to 7.5mg (+50%) 4 hourly.
2. Add up all BTM given in 24 hours and divide this by 6 and add to the following day's 4 hourly regular doses.
 - Remember also to increase the BTM as the regular dose is being increased.
 - Eg.: a patient receiving 5mg of morphine 4hourly received 4x breakthrough doses of 2.5mg of Morphine in 24 hours.
 - Total BTM received in 24 hrs = $4 \times 2.5 \text{ mg} = 10\text{mg}$
 - $10\text{mg} \div 6 = 1.67$
 - $5 + 1.67 = 6.67\text{mg}$ round up to 7mg
 - New dose: 7mg po 4hourly with 3.5 to 7mg for breakthrough pain.
- After pain is controlled with 4 hourly short acting morphine, it can be converted to sustained release long acting morphine (MST) that is given 12 hourly for greater convenience. To determine the dose of MST add up all the doses given in 24 hours and divide by 2. This is the number of milligrams of MST to prescribe.
- MST comes in 10mg, 30mg, 60mg and 100mg tablets that should not be crushed. Short acting syrup should still be prescribed for patients on MST for breakthrough pain.
- **Side-effects of morphine:**
 - Include **sedation** (resolves within 2-3 days), **nausea and vomiting**, **constipation**, **pruritis**, **urinary retention (uncommon)** and **respiratory depression**.
 - Tolerance develops in a few days to most side effects with the exception of constipation. Constipation can be prevented by the prophylactic use of laxatives (such as Lactulose, Sorbitol, Senna).
 - Haloperidol (Serenace): 1-4mg/day po in 2/3 divided doses or 25 – 50ug/kg/24 hrs subcut or Metoclopramide (Maxalon): 0.15 – 0.3mg/kg QID PO/IV/SC/PR are good drug choices for opioid associated nausea and vomiting.
 - Patients with urinary retention may need to be catheterized.
 - Pruritis is **not** related to histamine release and is best treated with ultra-low dose Naloxone (0.25ug/kg/hr) or opioid switch.
 - Consider opioid switch (use of another opioid eg Fentanyl, Methadone etc) if side effects are intolerable.
 - Always wean Morphine (decrease by 1/3 every 3 days) if it has been given for more than 10 -14 days to prevent opioid withdrawal symptoms.
 - If prescribing Morphine as an out-patient always complete the scheduled drug form and ensure that the script is written in both numbers and words and that the total amount (in millilitres or number of MST tablets) required for 2-4weeks is calculated and written in numbers and words.

ADJUVANTS AND OTHER ANALGESICS:

Drug	Dose	Notes
Amitriptyline	2 – 12 years: 0.1 – 0.2mg/kg at night. Increase to max 1-2mg/kg at night over 1 – 2 weeks. 12-18 years: 10 – 25mg po at night, increase to 75mg max.	Do ECG before using. Contraindicated in patients with prolonged QT interval. Pain dose is lower than for depression. Takes up to 3 weeks for effect. No longer considered first line if other adjuvants are available.
Carbamazepine	5-20mg/kg/day in 2 or 3 divided doses, increase gradually to avoid side effects.	Induces cytochrome P450. Drug interactions with ARVs. Can cause pancytopenia.
Gabapentin	2-12 years: 3 – 5mg/kg/dose. Start nocte, then BD then TDS. Increase to 10 – 20mg/kg/dose. 12-18 years: 300mg on day 1, 300mg BD on day 2, 300mg tds on day 3. Max 1200mg TDS.	Avoid sudden withdrawal Do not use in patients with history of psychiatric illness.
Corticosteroids	Prednisone 1-2mg/kg/day Dexamethasone: 0.1 – 1.5 mg/kg (max 10mg) increased to 0.1- 0.25mg/kg/dose BD for 14 days	Useful for Neuropathic pain, bone pain, IRIS.
Clonidine	1-4ug/kg/dose 6 – 12 hourly	Caution in renal failure, vascular disease, depression.
Diazepam	1-6 years: 1mg/day in divided doses (2-3) 6-14 years: 2 – 10mg/day in divided doses (2-3)	Used for associated anxiety.
Hyoscine butylbromide (Buscopan)	1 month – 2 years: 0.5mg/kg/dose PO 8 hourly 2 – 5 years: 5mg po 8 hourly 6 – 12 years: 10mg po 8 hourly	For colicky abdominal pain. Can cause nausea, dry mouth and constipation.

ETHICAL CONSIDERATIONS:

- When titrating doses of opioids in patients who are nearing end of life, one should escalate dosing as needed, with the goal of maximally controlling the child's pain and preventing suffering. By the doctrine of double effect, it is permissible to accept the risk of possible respiratory depression or apnoea in this setting, knowing that it means bringing about the desired effect of the patient's comfort.
- There is a notable difference between **opioid dependence** and **opioid tolerance**.



Tolerance is a physiologic process where the body adjusts to a medication that is frequently present, usually requiring higher doses of the same medication over time to achieve the same effect. It is a common occurrence in individuals taking high doses of opioids for extended periods, but does not predict any relationship to misuse or addiction.

- Physiologic dependence is characterised by unpleasant withdrawal symptoms that occur if opioids are abruptly discontinued. The withdrawal symptoms for opiates include severe dysphoria, sweating, nausea, rhinorrhea, depression, severe fatigue, vomiting and pain. Slowly reducing the intake of opioids over days and weeks will reduce or eliminate the withdrawal symptoms (see recommendations above).
- Psychological dependence is characterized by an individual's inability to stop using opioids even when objectively it is in his or her best interest to do so. The core concept of the WHO definition of “drug dependence” requires the presence of a strong desire or a sense of compulsion to take the drug. Studies show that most opioid dependent patients suffer from at least one severe psychiatric comorbidity.
- Most children who are on long-standing opioids for pain management will develop tolerance, and physiologic dependence will be observed if opioids are rapidly stopped. However, most of these children **DO NOT** develop psychological dependence. Signs of psychological dependence in the paediatric patient should prompt immediate psychiatric referral and evaluation.
- It is unethical to refrain from treating a child’s pain because of concerns that the child will become addicted or is developing an addiction. In children in whom psychological dependence or addiction is a concern, a structured, closely regulated pain management plan should be formulated, ideally in consult with psychology/psychiatry.

MANAGING DIFFICULT PAIN IN CHILDREN

NEUROPATHIC PAIN

DEFINITION:

Neuropathic pain is pain caused by an **abnormal reaction** to stimuli or spontaneous discharge by **damaged nerves (neuropathy)**.

Neuropathic pain may arise from the **peripheral** nervous system (eg tumour invasion of nerves) or **central** nervous systems (e.g. CVA, CNS tumour).

ASSESSMENT:

- A complete evaluation of motor, sensory, cerebellar, cranial nerve, reflex, cognitive and emotional functioning is important.
- **The following features** are suggestive of neuropathic pain:
 - **Characteristic descriptions:** burning, shooting, electrical, pins and needles.
 - **Associated neurological deficit** : weakness, loss of sensation.

- **Allodynia:** When harmless stimuli like light stroking elicit excruciating pain (in the absence of other skin problems or injury), this is highly suggestive of neuropathic pain.
 - **Hyperalgesia:** This refers to a situation when a patient has a decreased threshold to pain. Hyperalgesia to cold occurs more frequently than hyperalgesia to warmth. The distribution is generally not restricted to particular dermatomes as in adults and has a glove and stocking distribution.
 - **Focal autonomic or trophic changes:** smaller skin hairs, muscle atrophy, skin temp etc.
- Look for possible underlying causes (including medication history) of neuropathic pain.
 - **Nerve conduction studies:** These may give some insight into the location and type of nerve injury. However, the use of invasive electromyogram (EMG) may not be acceptable to children or available in resource constrained settings.

DETERMINING THE UNDERLYING CAUSE:

- **CNS:** post stroke, brain tumours
- **Infections:** HIV, Post herpetic neuropathy, meningitis
- **Nerve infiltration:** tumours
- **Vitamin deficiencies:** folate, B12 (more common in adults)
- **Metabolic:** Diabetes Mellitus, Fabry's disease
- **Drug related/toxins:** INH, ARVs (Stavudine, Efavirenz), Chemo: Vincristine, Metronidazole, heavy metal poisoning
- **Surgical causes:** trauma, damage to nerves during surgery
- **Other:** Guillian-Barre Syndrome, Phantom Limb (post amputation), complex regional pain syndrome, erythromyalgia

NON-DRUG TREATMENT MEASURES:

- Therapeutic touch or Reiki
- Breathing exercises, guided imagery, hypnosis
- Pressure bandages or stockings
- Counter irritation: light massage

DRUG TREATMENT MEASURES:

- **Use WHO ladder:** opioids are indicated in neuropathic pain even though it is classically described as pain that is not “opioid-sensitive.” Opioids are particularly useful in neuropathic pain while allowing time for adjuvants to work. Opioid rotation may be considered for difficult neuropathic pain (particularly Fentanyl and Methadone).
- **Adjuvants** are usually the mainstay of neuropathic pain management and help to spare opioids and their associated side effects. *See dosages in table “Adjuvants and Other Analgesics”*
- **Amitriptyline** commonly takes up to several weeks to work, is not always effective (esp. in HIV associated neuropathy) In addition, it generally has an unfavourable side effect profile (*see table “Adjuvants and Other Analgesics”*).
- **Gabapentin (Neurontin):** Effect may be evident in 24- 48 hrs, although may take longer. Gabapentin has fewer side effects than amitriptyline and more favourable metabolism. Dose needs to be adjusted down in patients with renal compromise. Dosing is generally started at a low end and increased gradually to avoid CNS side effects (particularly ataxia, dizziness). When stopping Gabapentin, patients should be titrated off over 1 week.
- **Pregabalin (Lyrica):** Use in children is not as well described as Gabapentin. Equivalence: 6mg Gabapentin = 1mg Lyrica. May be useful to switch to Lyrica if pain is not controlled with Gabapentin.
- **Corticosteroids:** can be useful for nerve compression as well as to decrease swelling around CNS tumours.
- **Clonidine:** Can provide additional anxiolysis and analgesia.
- **Other drugs (seek specialist guidance):** Methadone, Ketamine, Lidocaine patches, Capsaicin.

BONE PAIN

Pain arising from stimulation of pain sensitive structures (periosteum , endosteum Haversian cannals) in one or more bones.

ASSESSMENT:

Characteristics of bone pain:

- Aching to sharp, severe pain
- Generally more pronounced with movement
- Point tenderness common

DETERMINING THE UNDERLYING CAUSE:

- Fractures: traumatic and non-traumatic (pathological fractures)
- Bone tumours:
 - Primary bone tumours (e.g.- osteosarcoma)
 - Secondary bone tumours (bony metastases)
- Bone Marrow Infiltration (leukaemia)
- Vasculopathy or vascular crisis (sickle cell anaemia)
- Hemorrhagic event (joint bleeds seen in haemophilia)
- Infection (osteomyelitis)
- Inflammation (Autoimmune arthritis, e.g.- rheumatoid arthritis)

Mechanisms of bone pain:

- Mechanical: micro-fractures, tumour invasion
- Nociceptor stimulation by cytokines (released by inflammatory cells)
- Stretching of periosteum
- Nerve entrapment/invasion (incl Haversian canals)
- Secondary muscle spasm (therefore often mixed Nociceptive and neuropathic pain)

NON-DRUG MANAGEMENT:

• While many of the non-pharmacologic approaches to neuropathic pain can also be used for bone pain, (therapeutic touch, Reiki, guided imagery), another important component of non-pharmacologic management of bone pain is exercise and physical therapy (PT). Strengthening muscles gradually helps to build bone density and to offset stress on bones that can contribute

to worsening pain. In addition, exercise and physical therapy help to enhance mobility and function, which in turn help patients to feel better about themselves and their quality of life. Patients who suffer from bone pain should have an exercise or physiotherapy regimen as part of their overall care plan. At a minimum they should be performing active or passive range of motion exercises, depending on their functional status and ability to engage in activity. As needed or break through doses of pain medication can be administered prior to exercise or physio to enhance the patient's ability to tolerate activity and enjoy its benefits.

DRUG MANAGEMENT:

- Drugs that are useful in treatment of bony pain include:
 - NSAIDs
 - Opioids
 - Adjuvant pain meds (particularly Dexamethasone and Gabapentin)
 - Bisphosphonates (used to prevent breakdown of bone; helpful in the treatment of osteopaenia, osteoporosis, and Paget's disease)
 - Calcitonin (a synthetic hormone, Calcitonin can be used to treat osteoporosis, osteopaenia, and other benign bone conditions such as Paget's disease. Normally it is given as an injection with a pain medication, such as Morphine)

ADDITIONAL MEDICAL MANAGEMENT:

- In the case of certain bone cancers (either cancers that started primarily in the bone or those that are metastatic to the bone), radiation therapy may be an invaluable option in the management of bony pain. However, its utility would depend on availability, cancer type, and the patient's history of response to radiation therapy. Ideally, an oncologist or radiation oncologist should be consulted to advise accordingly.

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RESPIRATORY SYMPTOMS

DYSPNOEA

DEFINITION:

The word dyspnoea comes from the Greek “dys” meaning bad or difficult and “pnoea” relating to breathing.

Dyspnoea is a subjective feeling of breathlessness. Breathing becomes unpleasant and tiresome.

ASSESSMENT:

- **Children with dyspnoea may feel like...**

- They are gasping or panting
- They're drowning or suffocating
- Their chest is tight
- They are aware of every breath
- They have a headache or feel light headed
- They are Frightened

- **Look for signs of respiratory distress:**

Figure 2: Infant, Poor First Impression
Sick infant with poor muscle tone, nasal flaring, retractions

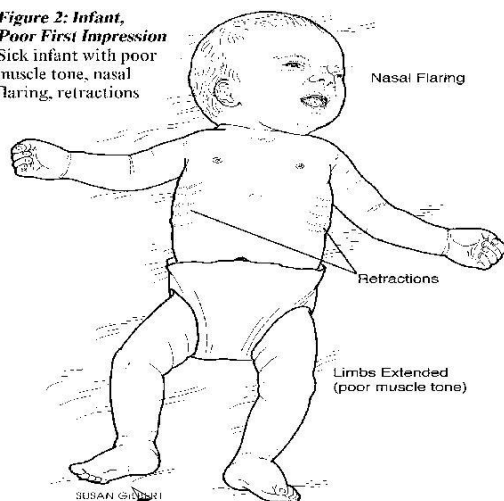
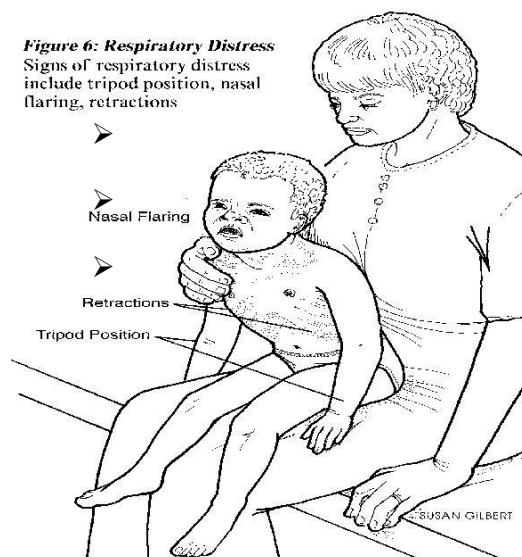


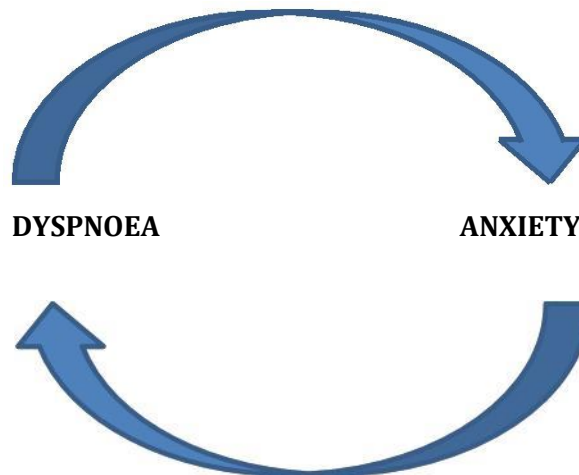
Figure 6: Respiratory Distress
Signs of respiratory distress include tripod position, nasal flaring, retractions



Not all children with respiratory distress feel dyspnoeic.

- Physiologic parameters of respiratory compromise do not always correlate with the degree of dyspnoea (for example, children with chronic hypoxia may be used to low oxygen saturations, while children with acute hypoxia may feel dyspnoeic even with oxygen saturations at the low end of normal).

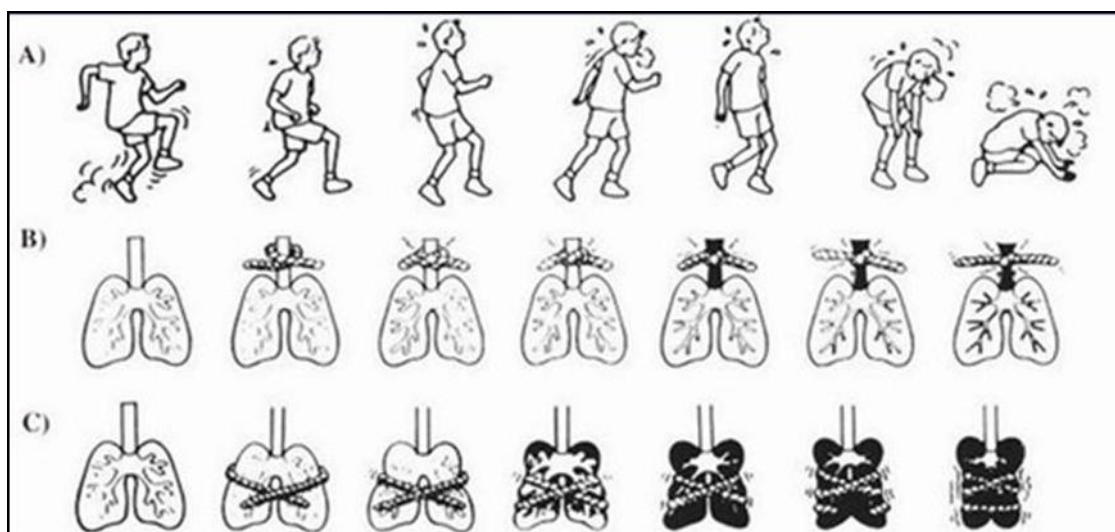
- Good functional indicators of dyspnoea are difficulty talking or feeding. Hypoxia may be associated with restlessness.
- Other symptoms that may be associated with breathlessness include anxiety, insomnia and chest pain.
- Anxiety often accompanies dyspnoea and may aggravate the feeling of breathlessness which can become a vicious circle if not addressed timeously:



- In children > 6 years the Dalhousie Dyspnoea scale may be used:

This is a 7 item pictorial self-report of 3 different aspects of breathlessness, namely effort intolerance, throat constriction and chest tightness. The scale has been used in children with cystic fibrosis and asthma.

DALHOUSIE DYSPNOEA SCALE:



DETERMINING AND TREATING THE UNDERLYING CAUSE:

UNDERLYING CAUSE:	TREATMENT:
Hypoxia	Oxygen therapy
Infection	Antibiotics, Bactrim for PCP, Anti-Tb drugs for TB
Pneumothorax, pleural effusion	Chest drain Note: malignant effusions are likely to recur and repeat drainage may be futile and pose considerable burden
Cardiac failure	Anti-failure treatment: diuretics, Digoxin, etc.
Anaemia	Haematinics, blood transfusion May help early on in the disease but may not be beneficial at the end of life and requires hospitalisation
Metabolic acidosis	Depending on cause: fluids, Na HCO ₃ (Sodium Bicarbonate), Insulin
Pulmonary Embolism	Anti-coagulation
Bronchospasm	Bronchodilator therapy
Hyperventilation	Breathing techniques, relaxation exercises, breathing into brown paper bag
Anxiety	Psychotherapy, relaxation techniques, benzodiazepines
Raised intra-cranial pressure	Dexamethasone

NON-DRUG MANAGEMENT:

- In verbal children, explore psychological influences on breathlessness and provide supportive counselling when appropriate
- Relaxation techniques and breathing exercises
- Correct positioning: semi-Fowler's or high Fowler's position for congestive heart failure, position with unaffected lung up and affected lung down in pleural effusion
- Gentle suctioning of oropharynx or nasopharynx with bulb syringe or mechanical suction
- Electric fan: cold stimuli in the distribution of the trigeminal nerve directly suppresses the vagal nerve and decreases respiratory rate
- Complementary therapies such as music and aromatherapy
- Hypnosis

DRUG MANAGEMENT:

The choice of treatment depends on the stage of disease, how actively the patient is still being managed and the availability of resources:

- Antibiotics may be useful to manage pneumonia as an underlying cause of dyspnoea if this is consistent with the patient and family's goals of care and stage of disease.
- In a patient where a decision is made to withhold antibiotics (especially with recurrent pneumonia associated with the end of life, i.e., in the case of recurrent aspiration events in a patient with severe irreversible neurological disease) Paracetamol or Ibuprofen can be given to manage pyrexia associated with dyspnoea.
- Oxygen: is not universally helpful for dyspnoea. Forcing therapy, especially at the end of life, is not helpful. One should focus on the patient's symptoms and whether or not the patient states he or she feels better or appears more comfortable with supplemental oxygen. Avoid focusing on the oxygen saturation monitor.
- Saline Nebulizer treatments can be used to help loosen secretions and to moisten the airways.
- Bronchodilators may be helpful if signs of bronchospasm are present.

- Hyoscine butylbromide (buscopan) helps to dry up secretions. It is especially useful in pneumonia at the end of life if antibiotics are withheld. Do not use in conditions where thickening secretions may worsen dyspnoea, such as cystic fibrosis and bronchiectasis.
- Diazepam or Lorazepam (benzodiazepines) may be used to address the anxiety component of dyspnoea if warranted. One must attempt to avoid dose stacking because of risk for respiratory depression.
- Clonidine is a good option for dyspnoea treatment as it provides both analgesia and anxiolysis.
- Midazolam (buccal, IV, or subcut) may be used especially for sudden severe onset breathlessness (e.g., pneumothorax, PE, or terminal dyspnoea).

The use of morphine to manage dyspnoea:

- Morphine can be used at “dyspnoea doses” to manage breathlessness. Dyspnoea doses are $\frac{1}{2}$ to $\frac{1}{3}$ of the normal oral dose for pain. It is important to note that if a patient has both pain and dyspnoea, Morphine should be dosed for treatment of the pain, and the benefit of dyspnoea management will still be attained.
- Morphine helps relieve dyspnoea by:
 - Decreasing the sensitivity of respiratory centre to CO₂: decreases awareness of breathlessness
 - Causes vasodilatation which in heart failure patients decreases the load on the heart
 - Having a sedative effect that reduces anxiety

ETHICAL CONSIDERATIONS:

- At dyspnoea doses, morphine does not suppress breathing. The doctrine of double effect may be relevant at higher doses (see ethics in pain section). Always weigh the burden of Morphine use vs. benefit.

COUGH

DEFINITION:

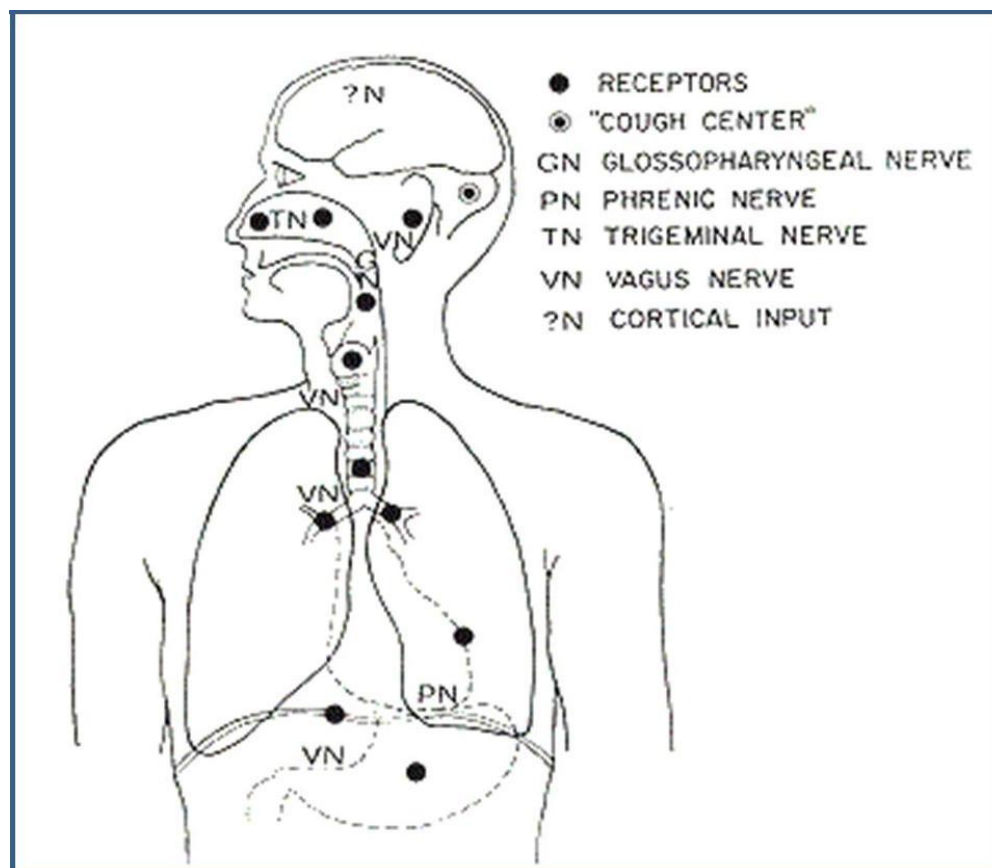
Cough is a sudden and often repetitively occurring reflex that helps to clear the large breathing passages from secretions, irritants, foreign particles and bacteria. Frequent coughing usually indicates the presence of disease. It becomes problematic and worthy of symptoms management when it interferes with sleep, rest, eating, and social functioning.

ACUTE: sudden onset, short duration

CHRONIC: > 2 weeks (usually suggests more “sinister” causes)

Coughing occurs through the stimulation of cough receptors that are present mostly within the respiratory centre but also outside of it:

Distribution of cough receptors:



ASSESSMENT OF COUGH:

If a patient presents with a cough, clarify the following on history^{viii}:

- Duration – days, weeks, months or years
- Course – constant, worsening, intermittent, paroxysmal (short, frequent, stereotyped), diurnal variation (variation day to night)
- Triggers – environmental allergens, irritants, swallowing, position
- Dry or productive (nature and volume of sputum) – clear/ white/grey (muroid), yellow/green (purulent) or bloody (haemoptysis)
- Other associated symptoms – shortness of breath, dyspnoea, chest pain, wheeze/tightness, loss of weight, fever, sweating

On examination, listen to the type of cough and look for associated physical findings:

- General – distress, fever, sweating, loss of weight, cyanosis, clubbing
- Respiratory – tachpnoea, bradypnoea, apnea, hyperinflation, hypoinflation, dullness, wheezing, rhonci, crackles, rales or pleural rub.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

The character of cough in children may help to determine its origin. Children with palliative care needs may develop any inter-current respiratory and upper airway problems that normal children develop, while some may be more prone to developing pathology in the respiratory system.

Type of cough:	Origin and/or likely cause:
Barking cough, associated stridor	Larynx / Croup, laryngitis
Dry + painful	Trachea, pleura/ tracheitis, pleuritis (pneumonia or pleural effusion)
Dry + irritant	Nasopharynx, laryngopharynx / post nasal drip, med side effect

Wet + productive	Bronchi + lung parenchyma / bronchitis, pneumonia
Paroxysmal	Whooping cough, asthma, cystic fibrosis
Nocturnal cough	Reflux, heart failure, asthma
Cough on awakening	Bronchiectasis, cystic fibrosis
Associated wheezing	Bronchiolitis, bronchitis, asthma, tumour
Haemoptysis	TB, PE, Bronchus Ca, left heart failure, tumour
Episodic clustering	Subclinical seizure activity

NON-DRUG MANAGEMENT:

- Air humidification (even a simple bowl of water in front of a heater, or a warm mist from the shower room)
- Physiotherapy
- Management of secretions: suctioning, saline nebulisation
- Safe home remedies include:
 - Tea with sugar
 - Hot water + 1 tsp honey + squeeze of lemon (recommended by the Integrated management of Childhood illnesses (IMCI))
 - Ample fluids to soothe a dry throat
- Unsafe home remedies include:
 - *Steaming and poultices*
 - Vicks Vapo-rub (may cause epistaxis and is considered unsafe in young children)

DRUG MANAGEMENT:

- **To treat cough or not to treat cough?**
 - Severe cough paroxysms
 - Cough interfering with feeding
 - Cough interfering with sleep
 - Cough leading to exhaustion
- } Warrants treatment (see med table below)

- **Cough suppressants** are generally not indicated in acute cough (except pertussis). They are often ineffective in children and may even be harmful. Some cough mixtures contain Atropine, alcohol and high doses of anti-histamines that are harmful, especially for young children (< 6 years of age).
- **Codeine** was once viewed as the gold standard in cough suppressants. Some recent placebo-controlled trials have found however that it may be no better than placebo for some etiologies including acute cough in children. It therefore is not recommended for cough in children.
- **Excessive secretions:** Excessive secretions may be coupled with cough, but children who have excessive secretions originating from the oral cavity (in the case of impaired swallowing) may not have cough as an on-going symptom. Diseases associated with excessive secretions include:
 - Cystic fibrosis
 - Pneumonia
 - Left heart failure
 - Bronchiectasis
 - Neuro-muscular diseases (eg myopathies): unable to swallow secretions
 - Cerebral Palsy
 - Common in pre-terminal stages of any disease (depressed LOC)

Medical management of the underlying cause of secretions is of primary importance, but symptom management can be further optimized with supportive measures, such as chest physiotherapy and postural drainage. Over-aggressive suctioning should be avoided as this can worsen secretions. In specialized units/hospitals, Botox injections may be available to children with neuro-muscular diseases. Anti-cholinergic agents (including Buscopan, Glycopyrolate, Scopolamine and Sublingual Atropine) can also be very useful (see med table below).

DOSES OF COMMONLY USED DRUGS IN RESPIRATORY SYMPTOMS:

Drug	Dose	Category and Uses	Notes
Salbutamol	Aerosol Inhaler: 2 puffs Q 4-6H PRN Nebulization: <1 yr: 0.05-0.15 mg/kg/dose Q 4 H PRN 1-5 yr: 1.25-2.5 mg/dose Q 4-6H PRN 5-12 yr: 2.5 mg/dose Q 4-6 H PRN >12 yr: 2.5-5 mg/dose Q 4-8 H PRN	B2 agonist; Bronchodilation	*Use of oral form is discouraged due to increased side effects and decreased efficacy compared to inhaled forms. Possible side effects include tachycardia, palpitations, tremor, insomnia, HA, nervousness, and nausea.
Ipratropium	Aerosol Inhaler: <12 yr: 1-2 puffs Q 6 H; max dose: 12 puffs/24H >12 yr: 2-3 puffs Q 6 H; max dose: 12 puffs/24H Nebulizer: Infant: 125-250 mcg/dose Q 8H Child: 250 mcg/dose Q 6-8 H >12 yr: 250-500 mcg/dose Q 6-8H	Anticholinergic; useful to decrease lower respiratory secretions	Contraindicated in soy or peanut allergy (for inhaler) and atropine hypersensitivity. May cause anxiety, dizziness, HA, GI discomfort.
Furosemide	IM/IV: Neonate: 0.5-1 mg/kg/dose Q 8-24H; max dose: 2 mg/kg/dose Infant and child: 1-2 mg/kg/dose Q 6-12 H Adult: 20-40 mg/24hr ÷ Q 6-12 H PO: Neonate: 1-4 mg/kg/dose once daily-BID Infant and child: 2 mg/kg/dose Q 6-8H; max dose: 6 mg/dose Adult: 20-80 mg/dose Q 6-12 hr; max dose: 600 mg/24H	Diuretic; may help with dyspnoea associated with pulmonary oedema, and increased secretions associated with CHF	Enteral bioavailability in neonates is poor.
N-Acetylcystine (Mucomyst)	Infant: 1-2 ml of 20% solution (diluted in equal volume of water to equal 10%) inhaled TID-QID Child: 3-5 ml of 20% solution (diluted in equal volume of water to equal 10%) inhaled TID-QID Adolescent: 5-10 ml of 10 or 20% solution inhaled TID-QID	Mucolytic (helps to reduce severe cases of congestion or secretions)	Use with caution in asthma. Ideal to administer after bronchodilator use and following postural drainage. May induce bronchospasm, stomatitis, drowsiness, rhinorrhea, nausea, vomiting, and haemoptysis.
Diphenhydramine	1-2 mg/kg/dose Q 6 hours PRN; max dose is 50 mg/dose, 300 mg/24H	Antihistamine; useful for allergy-associated congestion	Contraindicated with MAO inhibitor use, acute asthma attacks, GI or urinary

			obstruction. Use with caution in infants and young children. DO NOT use in neonates due to potential CNS effects. Adjust dose in renal failure.
Hyoscine butylbromide (Buscopan)	IM/IV: 1 mo-4 yr: 300-500 mcg/kg TID-QID; max dose 5 mg 5-12 yr: 5-10mg TID-QID 12 yrs-adult: 10-20 mg TID-QID PO: 1 mo-2 yrs: 300-500 mcg/kg TID-QID; max dose 5 mg 2-5 yrs: 5 mg TID-QID 5-12 yrs: 10 mg TID-QID 12 yrs-adult: 10 -20 mg TID-QID	Anti-cholinergic; useful in secretion management	Does not cross the blood brain barrier, and therefore does not cause drowsiness. IV version is not licensed for use in children < 6 yrs.
Glycopyrolate (Robinul)	IM/IV: Child: 0.004-0.01 mg/kg/dose Q 4-8H Adult: 0.1-0.2 mg/dose Q 4-8H; max dose 0.2 mg/dose or 0.8 mg/24H PO: Child: 0.04-0.1 mg/kg/dose Q 4-8H Adult: 1-2 mg/dose BID-TID	Anti-cholinergic; useful to decrease oral secretions as well as lower respiratory tract secretions	Use with caution in hepatic and renal disease, asthma, and urinary retention. Anti-cholinergic properties may be potentiated if given with other drugs with anti-cholinergic properties.

References:

1. Ainslie, Gilian. Assessment of cough. AJOL info 2009; 27 (2): 68-71.
2. Arcara K, Tscudy M, ed. The Harriet Lane handbook: a manual for pediatric house officers. 19th ed. St. Louis: Mosby; 2012.

GASTRO-INTESTINAL SYMPTOMS

ODYNOPHAGIA

DEFINITION:

Odyno = pain; Phagia = Gr. phagein = to eat. Refers specifically to pain within the oropharynx and/or pain with swallowing.

Oral pain occurs relatively commonly in the palliative care setting. Children in particular may be severely impacted by oral pain. Not only does it lead to irritability and discomfort, but impaired oral intake may also result in dehydration and loss of weight.

ASSESSMENT:

- Inquire about what seems to evoke the pain:
 - having anything in the mouth at all?
 - Specifically swallowing?
 - Or both?
- Inquire about where exactly the pain seem to be occurring:
 - In the mouth?
 - In the throat?
 - In the upper chest (heart burn)?
- On physical exam, pay close attention to the following:
 - The appearance of the oral cavity- Are there notable lesions on the tongue, buccal mucosa, or gingiva?
 - The condition of the dentition- is the child teething or erupting wisdom teeth? Are there caries evident? Is there purulent drainage from any of the teeth, or tenderness over the maxilla or mandible? Are the gingiva bleeding?
 - Is the breath malodorous?

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Cause	Treatment	Notes
Oral Candidiasis (Candida Albicans)	-Nystatin (Mycostatin) 1ml po QID (30 mins after feed) for 7 to 14 days -Miconazole (Daktarin) oral gel 4-6 hourly for 7 – 14 days (after meals) -Gentian violet if above meds are not available -If refractory or severe or oesophageal: Fluconazole- 6 mg/kg stat then 3mg/kg daily for 21 days -NSAIDS and/or Paracetamol for pain; Sucralfate may be useful in oesophageal candidiasis	-May be accompanied by napkin dermatitis -May form thick membranes (membranous candidiasis) -White spots that are difficult to scrape away (distinguish from milk feeds) and can bleed on contact. -May be confined to tongue and buccal mucosa or extend to the pharynx, larynx, or oesophagus (oesophageal candidiasis is an AIDS defining illness) -Suspect oesophageal involvement in child with hoarse voice, stridor, excessive drooling, dysphagia
Herpes stomatitis	-Acyclovir: > 2 yrs: 400 mg PO Q 8H for 5 days < 2 yrs: 200mg PO Q 8H hourly for 5 days IV is indicated if child is unable to take PO, or if disseminated -For superimposed bacterial infections: add amoxicillin and flucloxacillin -NSAIDs, Paracetamol, and/or opioids for pain •Beclometasone dipropionate (becotide) 50 – 100mcg sprayed twice daily on the oral mucosa may help manage oral ulceration (unlicensed use)	-May occur as: Acute primary infection Recurrent infection Chronic infection -Painful ulcer, typically 4-5mm in diameter on tongue, lips and buccal mucosa; may bleed, often -Interferes with eating, commonly associated with hypersalivation
Mucositis (chemotherapy related)	-Magic mouthwash: 100ml mucaïne, 10 ml lignocaine (1%), 30ml mycostatin suspension and 15 - 30mg of morphine. Use to gargle with and spit out. -Opioids	-Associated with severe oral pain, inability to feed, dehydration -May be complicated by oral herpes and candidiasis
Peridontal disease, caries	-Referral for dental care, if available -Oral antibiotics with anaerobic	-Anaerobic bacteria play an important role in dental infections; their overgrowth can be controlled

	coverage (eg metronidazole) may be appropriate in peridental abscess -NSAIDS and/or Paracetamol for pain	with regular mouth care - Halitosis and gingival bleeding may suggest periodontal disease or dental infection
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NON-DRUG MEASURES:

- Avoid hot, acidic or spicy foods
- Suck ice, lollies, Vit C
- Regular mouth care (brushing teeth or using mouth swabs BD as appropriate)
- Use mouth rinses (such as Saline, Chlorhexidine, Sodium Bicarbonate, and Lignocaine) daily as needed
- Hydrating lips with Vaseline and Paraffin can contribute greatly to oral comfort

DRUG MEASURES:

- Locally available teething gels (such as teejel or bonjela) that do not contain Benzocaine, (which has in rare cases been associated with Methaemoglobaemia) may provide symptomatic relief.

NAUSEA AND VOMITING

DEFINITION:

NAUSEA: an unpleasant sensation vaguely referred to the epigastrium and abdomen and often but not always culminating in vomiting.

VOMITING: the forcible expulsion of the contents of the stomach through the mouth.

ASSESSMENT:

In babies and young children, vomiting needs to be distinguished from **regurgitation**. Regurgitation is the passage of refluxed gastric contents into the oropharynx which is often effortless (ie does not involve the contraction of abdominal muscles). Regurgitation is a common feature of gastro-oesophageal reflux (GOR) that is pathologic when associated with poor weight gain, oesophagitis, respiratory symptoms and other life threatening events (aspiration). Gastro-oesophageal reflux disease is common in children with palliative care needs (esp neurological conditions).

Assessment of the vomiting child includes:

- Taking of full history of the vomiting, timing, frequency, food and drug intake
- Looking for possible underlying causes of the vomiting (eg raised intracranial pressure, surgical causes, infections etc)
- Looking for effects of vomiting (eg dehydration, aspiration etc).

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Symptom /sign/treatment

Possible cause

Blood stained vomiting (haematemesis)

Oesophagitis, swallowed blood, peptic ulcer, oesophageal varices

Coffee-ground vomitus

Upper GI bleeding, stress ulceration, pre-terminal event, DIC

Bile stained vomiting

Upper GIT obstruction

Undigested milk/food	Gastric outlet obstruction
Associated diarrhoea	Gastro-enteritis
Projectile vomiting	Raised ICP, Pyloric stenosis
Abdominal distension, increased bowel sounds	Bowel obstruction
Abdominal distension, decreased bowel sounds	Ileus, electrolyte disturbances
Fever, dysuria, frequency, rigors	UTI, Pyelonephritis
Tender right upper quadrant,	Hepatitis
jaundice Guarding and rebound	Surgical causes: appendicitis,
tenderness Oliguria, oedema	pancreatitis Renal failure
Chemotherapy, radiotherapy	Toxicity, radiation enteritis
Bulging fontanelles, blurred disc, hypertension, bradycardia	Raised ICP, hydrocephalus, space occupying lesion, intracranial bleed
Photophobia, meningismus (neck stiffness) Smell of ketones, coma	Meningitis
Associated headaches, blurred vision	DKA, other metabolic disorders
	Migraines

Drugs that commonly cause nausea and vomiting:

Narcotics	Anti-helminthics	SSRIs	Tramadol	Potassium salts
Chemotherapeutics	Oestrogens	Bisphosphonates	Antibiotics	Digoxin
Iron salts				

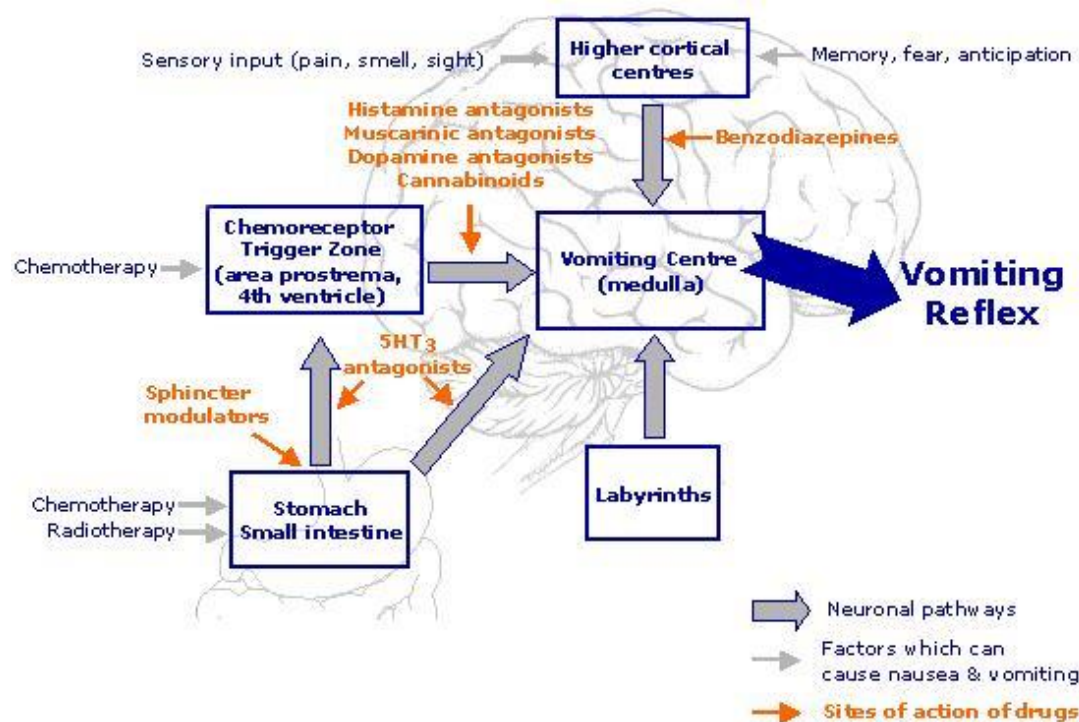
NON-DRUG MEASURES:

- Avoid foods/smells that aggravate nausea and vomiting especially spicy, very sweet or fatty foods
- Offer foods that are bland and dry (eg toast)
- Maintain hydration by giving small amounts of oral rehydration solution: may require cup and spoon or syringe feeding
- Try avoid letting a very thirsty baby suck too much air which leads to more vomiting
- May require IV or NGT fluids if rehydration indicated
- Rinse mouth and brush teeth after vomiting

- Split medication if vomiting associated with meds

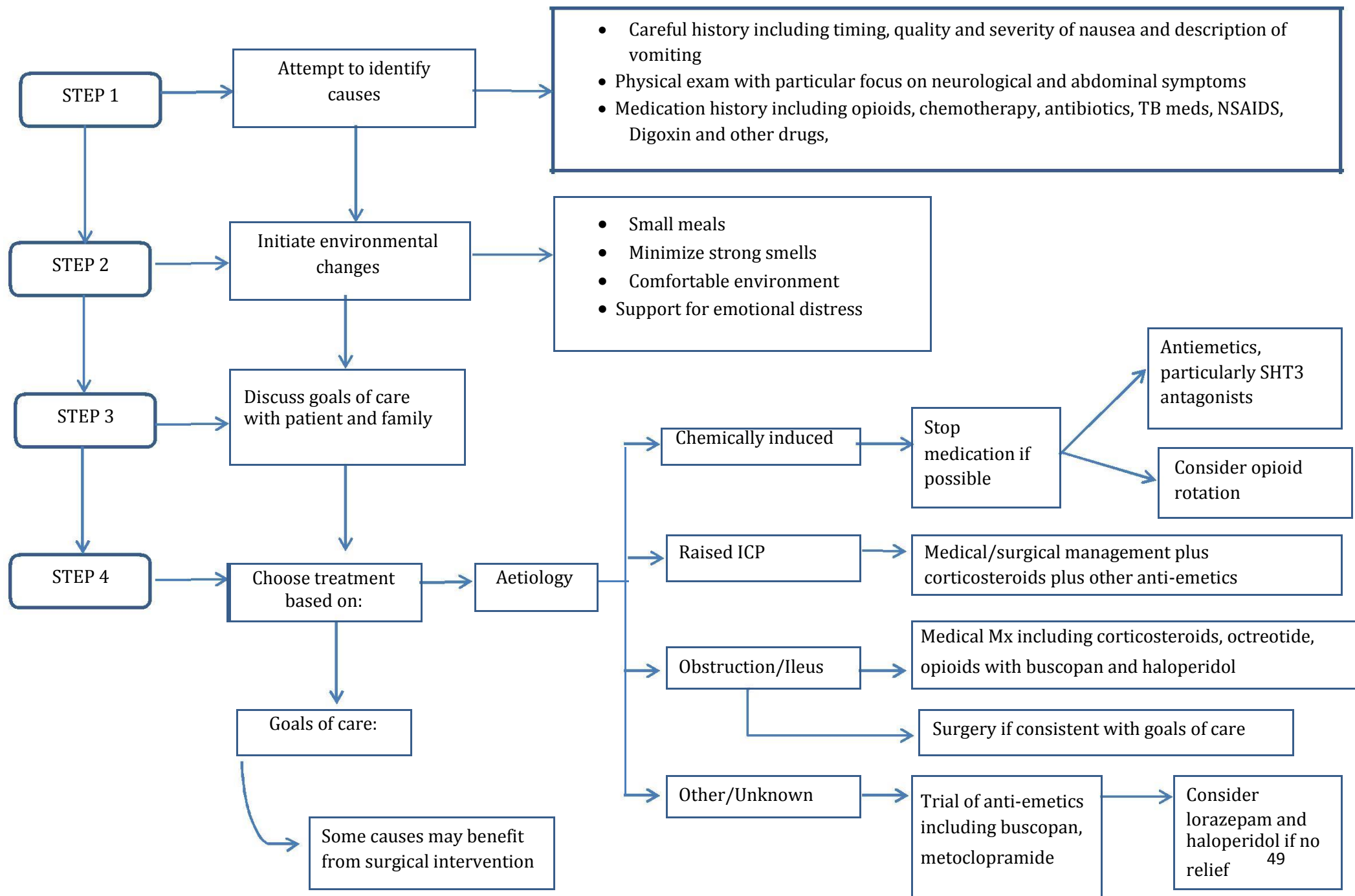
DRUG MEASURES:

- Knowledge of the neuronal pathways, sites of action and receptors involved in nausea and vomiting as well as the sites of action of anti-emetics are essential to correctly manage nausea and vomiting in the palliative care setting.
- Although health care professionals are more familiar with not treating vomiting associated with reversible causes, treatment of nausea and vomiting in the palliative care setting (particularly if chronic and associated with distress) is warranted. Side effects of anti-emetics need to be balanced against benefits and the most appropriate medication chosen according to the most likely pathology.



Classes of most commonly used anti-emetics:

Benzodiazepines	Histamine/muscarine antagonists	Dopamine antagonists	5 HT3 antagonists
Lorazepam	Promethazine	Metoclopramide	Metoclopramide
	Cyclizine	Haloperidol	Odansetron
		Domperidone	Granisetron



DOSES OF COMMONLY USED ANTI-EMETICS IN CHILDREN:

Drug	Dose	Category and Uses	Notes
Metoclopramide	PO, IV, or IM: Neonate: 100 mcg/kg Q 6-8H 1 mo- 1yr (wt up to 10 kg): 100 mcg/kg BD, ma 1 mg/dose 1-3 yrs (wt up to 10-14 kg): 1 mg BD-TD 3-5 yrs (wt up to 15-19 kg): 2 mg BD-TD 5-9 yrs (wt up to 20-29 kg): 2.5 mg TD 9-10 yrs (wt up to 30-60 kg): 5 mg TD 15yrs-adult (wt over 60 kg): 10 mg TD	Dopamine antagonist, 5HT-3 antagonist; antiemetic, prokinetic	Not licensed to be used as prokinetic in neonates. Adverse effects include restlessness, drowsiness, and focal dystonia . *The risk for extrapyramidal side effects are increased in patients < 20 yrs.
Ondansetron	PO/IV 1-18 yr: 0.1-0.15 mg/kg/dose Q 8-12H. Max single dose 4 mg. (Also comes in useful dispersable tablet: Zofran Zydis: 4mg and 8mg)	5HT-3 receptor antagonist; powerful anti-emetic	Used commonly for chemotherapy induced nausea/vomiting. Not licensed for use in children <2. Causes constipation.
Granisetron	IV: 10 – 40ug/kg (max 3mg) diluted in 20 – 30mls over 5 mins	5HT-3 receptor antagonist; powerful anti-emetic	Off licence < 2 years
Lorazepam	PO: <2 yrs: 25 mcg/kg BD-TD 2-5 yrs: 0.5 mg BD-TD 6-10 yrs: 0.75 mg TD 11-14 yrs: 1 mg TD 15-18 yrs: 1-2 mg TD Sublingual: All ages: 25 mcg/kg as single dose. Increase to 50 mcg/kg if necessary. Adult: 500 mcg-1 mg/dose as single dose, repeat as needed.	Benzodiazepine; useful for nausea and anxiety associated with frequent nausea and vomiting	May cause drowsiness and respiratory depression in large doses. Caution in renal and hepatic failure.
Haloperidol	PO: 12-18 yrs: 1 mg at night, increasing to 1 mg BD. Max 5 mg BD. IV: 12-18 yrs: Initial dose of 1 mg/24hrs. Increase as needed to maximum of 5 mg/24 hrs.	Dopamine antagonist; useful in metabolic causes of nausea/vomiting	*Use with caution in patients with QT-prolonging conditions. EKG monitoring advised if giving IV. Not licensed for use in children with nausea/vomiting.

DIARRHOEA

DEFINITION:

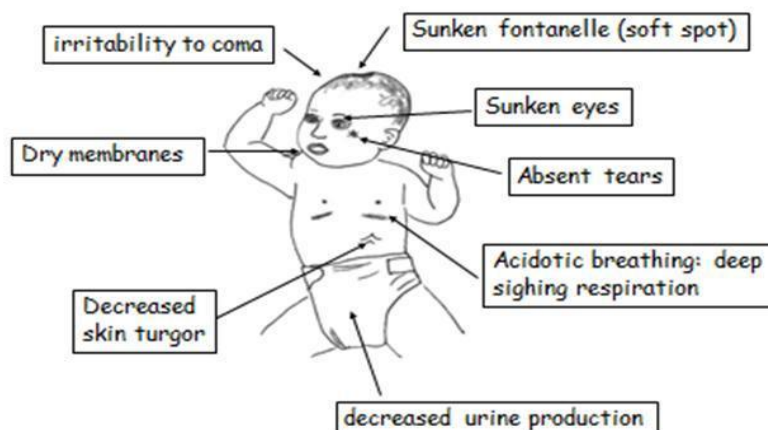
- Diarrhoea = passage of abnormally loose stools more frequently than normal.

OTHER CONCEPTS:

- Breastfed infants may pass stool with each feed
- Infant (bottle fed) > 7 stools/day
- Toddler and older child: > 3 stools per day
- Gastro-enteritis : infection of the gastrointestinal tract resulting in diarrhoea and vomiting.
- Acute Diarrhoea: sudden onset: 3-7 days
- Persistent Diarrhoea: more than 14 days, dehydrating
- Chronic diarrhoea: more than 14 days not dehydrating

ASSESSMENT OF INFANTS AND CHILDREN WITH DIARRHOEA:

- It is important to delineate the time course of the diarrhoea (acute vs. persistent/chronic), as this will help to distinguish the aetiology.
- On history, inquire about recent infectious exposures (have other people in the household or other children at school been similarly affected with diarrhoea?) and medication changes (recent antibiotic use is a common cause of diarrhoea).
- In the child with diarrhoea, look for the following findings on exam to assess for concurrent dehydration:



DETERMINE AND TREAT THE UNDERLYING CAUSE:

- Consider laboratory evaluation depending on child's presentation and availability of testing. Studies to possibly obtain include a complete blood count, electrolytes, haem-occult stool test, urine culture, stool culture (if child is febrile and stools are bloody), and microscopy.
- The aetiology of diarrhoea can be classified as one of the following:
 - **Inflammatory** (commonly infectious)
 - **Malabsorptive with secretory mechanism** (e.g., cholera)
 - **Malabsorptive with osmotic mechanism** (commonly seen in underlying chronic disease)
 - **Exudative** (inflammatory bowel diseases, such as Chron's and ulcerative colitis)
 - **Motility related** (hypermotility disorders, such as hyperthyroidism)
- Several underlying diseases can be implicated in diarrheal illness including:
 - HIV
 - Malignancies + para-neoplastic syndromes
 - Cystic Fibrosis
 - Coeliac disease
 - Inborn errors of metabolism
 - Nephrotic syndrome
 - Short-bowel syndrome secondary to resection (such as is seen in nectrotizing enterocolitis, or NEC)
- Cultural practices (such as enemas, herbal medications, and natural laxatives) must be considered in determining the underlying cause of diarrhoea. It is important to explore this possibility during the assessment phase while obtaining a history.
- Medications/therapies commonly associated with diarrhoea include:

Anti-biotics

Laxatives

Anti-retrovirals

Chemotherapy

Radio-therapy

NON-DRUG MEASURES:

- In cases of diarrhoea accompanied by dehydration, ORS (oral rehydration solutions) should be utilized as the primary source of rehydration.
 - A simple formulation for ORS is: 1 litre water (from clean or pure water source)
+ 8 tsp sugar + 1/2 tsp salt
 - Commercially available preparations include Gastrolyte and Rehydrate

➤ Cereal based ORS is recommended by the WHO for malnourished children

- DO NOT STOP MILK FEEDS in children with diarrheal illness except when rehydrating with ORS. Breast-milk contains many immunologic beneficial properties from which children with diarrheal illness can benefit.
- Early refeeding decreases stool output, improves nutrition and is comforting to the child
- Temporary lactose intolerance may develop in diarrheal illness due to temporary loss of the intestinal brush border due to inflammation. This may necessitate the short term use of soya based milks. Yoghurt and Maas may also be used.

DRUG MEASURES:

Doses of commonly used drugs in diarrhoea:

Drug	Dose	Category and Uses	Notes
Vitamin A	0-5 mo: 50 000 IU po single dose 6-11 mo: 100 000IU po single dose 1-5 yr: 200 000 IU po single dose	Supplementation	Supplementation in children 6- 60 mo reduces diarrhoeal disease mortality by 33%.
Zinc	0-6 mo: 2mg po od 6-12 mo: 3mg po od 1-3 yr: 3mg po od 4-8 yr: 5mg po od 9-13 yr: 8mg po od Males 14-18 yr: 11mg od po Females 14-18 yr: 9mg po od	Supplementation	Zn supplementation reduces the duration of HIV associated diarrhoea. Zn is recommended by WHO as an adjunct to ORS in acute diarrhoea in children.
Loperamide	1 mo-1yr: 0.1 mg/kg BD 30 minutes before feeds. Increase as necessary to max of 2 mg/kg/day in divided doses. 1-12 yrs: 0.1 mg/kg TD-QD. Increase as necessary up to max of 1.25 mg/kg/day in divided doses (max 16 mg/day). >12 yrs: 2 mg BD-QD. Increase as necessary to max of 16 mg/day in divided doses.	Opioid and Mu-receptor agonist; useful in diarrhoea associated with hypermotility and of non-infectious aetiology	Does not act centrally (targets myenteric receptors in the large intestine). Not licensed for use in children with chronic diarrhoea.
Lactobacillus	The strength of lactobacillus products is usually indicated by the number of living organisms per capsule. Typical doses range from 1 to 10 billion living organisms taken daily in 3-4 divided doses.	A normal flora bacteria commonly found in GI and urinary tracts thought to promote overgrowth of harmful bacteria. Also found in some yogurts and dietary supplements.	Use with caution in children who are immunosuppressed. No known interactions with other foods or drugs.

Additional notes:

- Antibiotic use should be considered in cases of bloody diarrhoea.
- Chronic diarrhoea is common in HIV infected children. In combination with other infections it can lead to significant damage to the bowel wall and associated malnutrition if not treated. Some points to consider include:
 - Ensure that the child is on ARVs, and consider how the ARV regimen might be contributing to diarrhoea (consider drug rotation drugs may be exacerbating the diarrhoea).
 - Cryptosporidium may be an associated infection, and is often difficult to treat (children might have to undergo more than one course of treatment).
 - Specialized formulas may be warranted if malabsorption can be documented. Such formulas include Isomil, AL110, and Alfare, and should be used under the supervision of a dietician.
 - Bowie's Cocktail/Regimen (Cholestyramine, Gentamycin, and Isomil) should be considered in formulating a treatment regimen.
- Opioids can be considered for symptoms management of chronic diarrhoea in older children because of their constipating side effects. All underlying infections should be treated before starting opioids for this purpose and it should not be used for acute diarrhoea.

CONSTIPATION

DEFINITION:

Infrequent or difficult evacuation of faeces that are usually hard.

ASSESSMENT OF CONSTIPATION:

- **On history, determine:**
 - - How long has stooling regularly been a problem?
 - How does the family define constipation (what are the frequency, consistency, and size of stools?)
 - Is there any associated pain or bleeding with stooling?
 - Has the child exhibited any other of the following symptoms?
 - Abdominal pain
 - Soiling of underwear with stool
 - Stool withholding behaviour
 - Abdominal distension
 - Nausea/vomiting

- Weight loss or poor weight gain
- What is the child's diet like (what does he or she typically eat)? What is his or her average daily fluid intake?
- Are there any psychosocial issues of note (recent stressors, history of depression, etc)?
- What are on-going medical issues/diagnoses that can be contributing?
- **On physical exam observe:**
 - External anal and perineum. Look for anal fissures, tears, or abscesses. A rectal exam should also be performed to determine anal tone, rectal size, and amount and consistency of stool in the rectal vault.
 - Children's general body habitus. Is their physical appearance consistent with failure to thrive?
 - Abdominal exam. Are the bowel sounds absent? Are they present, but hypo- or hyperactive? Is their abdominal distension and/or tenderness? Is there a palpable mass/masses?

DETERMINING AND TREATING THE UNDERLYING CAUSE:

DISEASE RELATED

Cerebral palsy
 Cystic fibrosis
 Neuropathic conditions
 Spina bifida (MMC)
 Hirschprung's disease
 Prune belly syndrome
 Gastroschisis
 Anatomic malformations (anal stenosis)
 Intra-abdominal or pelvic masses

Tx RELATED

Opiate therapy
 Chemotherapy
 Anti-depressants

FUNCTIONAL

Developmental
 Situational
 Depression
 Dehydration
 Low fibre diet

- Functional causes of constipation are the most common causes of constipation in the well paediatric population. Do not forget the impact that developmental disorders (cognitive deficits), situational circumstances (such as excessive discipline or abuse), and mood (depression) can have in chronically ill children.
- Always take a drug history. Opioid use for pain is one of the most preventable common causes of constipation in the palliative care patient.

NON-DRUG MEASURES:

- Dietary adjustments are often the first approach to managing constipation in children. Fibre can be increased in the diet by increasing intake of whole-grains, fruits, and vegetables. In infants > 2 months of age, prune juice or 100% apple juice can be given once a day as needed.
- Ensuring for adequate hydration is important in preventing constipation. Monitor urine output closely and increase formula/breast-milk intake in children <1yrs, and free water intake in children >1 yrs accordingly.
- A regular activity/exercise regimen is important in the maintenance of bowel motility and function and therefore in the prevention of constipation.
- In children whom have had aggressive regimens for constipation management including both non-drug and drug measures, yet still struggle with severe constipation or impaction, manual disimpaction (potentially under general anaesthesia) may be necessary.

DRUG MEASURES:

- Children on standing or long-term opioid regimens should ALWAYS be routinely placed on a stool-softener. Sennosides once daily are the regimen of choice for the prevention of opioid-induced constipation. Doses of sennosides can be increased in size and frequency if constipation worsens with increasing opioid dosing, and additional agents can also be added to the patients medication regimen (see table below).
- For the treatment of constipation, the agents typically used include bulk forming agents, stool softeners, lactulose, glycerine suppositories. A systematic approach to constipation treatment may include the following:
 - Drug 1: Lactulose
 - Drug 2: Add Senna or change to Docusate (ignore this step if child is on an opioid and already on a standing Sennoside)
 - Drug 3: Glycerine suppository
 - Drug 4: Phosphate enema (Fleet's) or Mineral oil enema
 - Last resort: manual removal (possible GA)

DOSES OF COMMONLY USED LAXATIVES IN CHILDREN:

Drug	Dose	Category/class	Notes
Senna	1 mo-2 yrs: 0.5 ml/kg (max 2.5 ml) PO daily 2-6 yrs: 2.5-5ml PO daily 6-12 yrs: 5-10 ml PO daily, or 1-2 tabs PO at night 12yrs-adult: 10-20 ml daily,	Stimulant laxative.	Onset of action is 8-12 hours. Dose can be exceeded for cases of severe constipation. Drug of choice for opioid induced constipation. Avoid in children with

	or 2-4 tables at night		existing abdominal pain.
Lactulose	1 mo-1yr: 2.5 ml/dose PO daily-TDS 1-5yrs: 5ml/dose PO daily-TDS 5-10yrs: 10 ml/dose PO daily-TD 10yrs-adult: 15 ml/dose PO daily-TDS	Osmotic laxative.	Can take 36-48 hrs for peak onset of action. Relatively ineffective in severe opioid induced constipation (needs to be paired with a stimulant). Poor taste may make it difficult to tolerate in children.
Polyethylene Glycol	0.5-1.5 G/Kg PO daily, not to exceed 17 G/day.	Osmotic laxative.	

References:

1. Arcara K, Tscudy M, ed. The Harriet Lane handbook: a manual for pediatric house officers. 19th ed. St. Louis: Mosby; 2012.
2. The Association of Pediatric Palliative Medicine Master Formulary. 2012.

NEURO-PSYCHIATRIC SYMPTOMS:

SEIZURES

DEFINITION:

A transient symptom of abnormal, excessive or synchronous neuronal activity in the brain. Outwardly, this can appear as wild as thrashing movement (tonic-clonic seizure) or as mild as a brief loss of awareness (absence seizure).

Status epilepticus is defined as a prolonged or recurrent seizure lasting greater than 30 minutes without the patient regaining consciousness.

ASSESSMENT:

- On history, it is important to determine:
 - Is this the child's first seizure?
 - Is there a family history of seizure disorder or neurologic problems?
 - If the seizures have occurred before, what things typically cause them?
 - Have there been any recent adjustments to seizure medications? Have any other new medications been added?
- On exam, a full-neurologic assessment should be performed.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

- The aetiology of seizure can be divided into epileptic and non-epileptic causes.

Epileptic causes	Non-epileptic causes
Epilepsy syndromes (neonatal seizures, infantile spasms, benign rolandic epilepsy, juvenile myoclonic epilepsy, Lenox-Gastaut syndrome, etc.)	Febrile illness
Cerebral palsy	Electrolyte imbalance (particularly hyponatraemia and hypo
Neuro-degenerative disorders	Infection (e.g., encephalitis, meningitis)
In-born errors of metabolism	Hypertension
Intra-cranial haemorrhage/trauma	Uraemia
CNS tumours or abscesses	Hypoglycaemia
CVA	Hypoxemia
Hydrocephalus	Hypocapnoea
	Medication induced (Opiates, Tri-cyclic anti-depressants, Neuroleptics)

- Febrile seizures are most commonly seen in children between 6 months and 6 years of age. They are non-focal, motor seizures that typically last no longer than 10 minutes and generally lead to rapid recovery. They are thought to occur not because of infection, but rather because of a sudden rise in temperature that can occur with fever. Despite popular belief, febrile seizures are not associated with brain injury, developmental delay, or significantly increased risk of epilepsy syndromes in later childhood or adulthood. Antipyretics are important in the management of febrile seizure in that they can help to decrease fever and increase the child's comfort level. However, it is not necessary to put children on around-the-clock dosing of antipyretics during febrile illness in attempts of preventing febrile illness.

NON-DRUG MEASURES:

- FIRST AID FOR SEIZURES (prevent patient's injury by):
 - Place a pillow under the head
 - Remove restrictive clothing
 - Do not try to restrain movement
 - Turn the patient on his or her side to prevent aspiration
 - Do not insert object into the mouth
 - Administer oxygen where possible (place on a non-rebreather face-mask)
 - Check blood glucose and give glucose if necessary
- Following a seizure a patient may enter a post-ictal state, in which they are very sleepy and less responsive than usual. This stage may last minutes to several hours, depending on the duration and nature of the seizure. Continue to keep the patient in a safe and comfortable position while they are in the post-ictal state.
- In patients with epileptic syndromes that are complex and very refractory to conventional therapy, alternative therapies can be considered under a neurologist's guidance. Examples of such therapies include supratherapeutic monotherapy, placement of a vagal nerve stimulator, ketogenic diets, removal of epileptogenic area, and anterior temporal lobectomy.

DRUG MEASURES:

- The pharmacologic management of epileptic seizures is beyond the scope of these clinical guidelines. The underlying causes of non-epileptic causes of seizures other than febrile seizure should be medically managed accordingly.
- Children with known seizure disorders or in whom it is anticipated that seizures may develop should have rescue doses of **Diazepam PR** (per-rectum) readily available.

Lorazepam can also be given sublingually in case of emergency. Medical staff and family members should consider giving emergency doses at the following doses for seizures associated with change in consciousness lasting longer than 10 minutes:

Diazepam	Lorazepam
<p>Neonate: 1.25-2.5 mg PR repeated once after 10 min. if necessary</p> <p>1 mo-2 yrs: 5 mg PR repeated once after 10 min. if necessary</p> <p>2-12 yrs: 5-10 mg PR repeated once Q 10 min. if necessary</p> <p>12 yrs-adult: 10 mg PR repeated once Q 10 min. if necessary</p> <p>Alternative: 0.3mg/kg/dose</p>	<p>0.1 mg/kg/dose of IV version administered PR up to a max of 2 mg/dose. May be repeated once after 10 min. if necessary.</p> <p>Sublingual Lorazepam is also available as 1mg tablets.</p>

- Patients will often complain of muscle soreness or cramping after a prolonged seizure. This can be treated with analgesics on level 1 of the WHO pain ladder, or Benzodiazepines if already prescribed for the patient.

MUSCLE SPASM AND SPASTICITY:

Definition:

Spasm: A sudden, involuntary contraction of a muscle, often associated with pain and limitations in function.

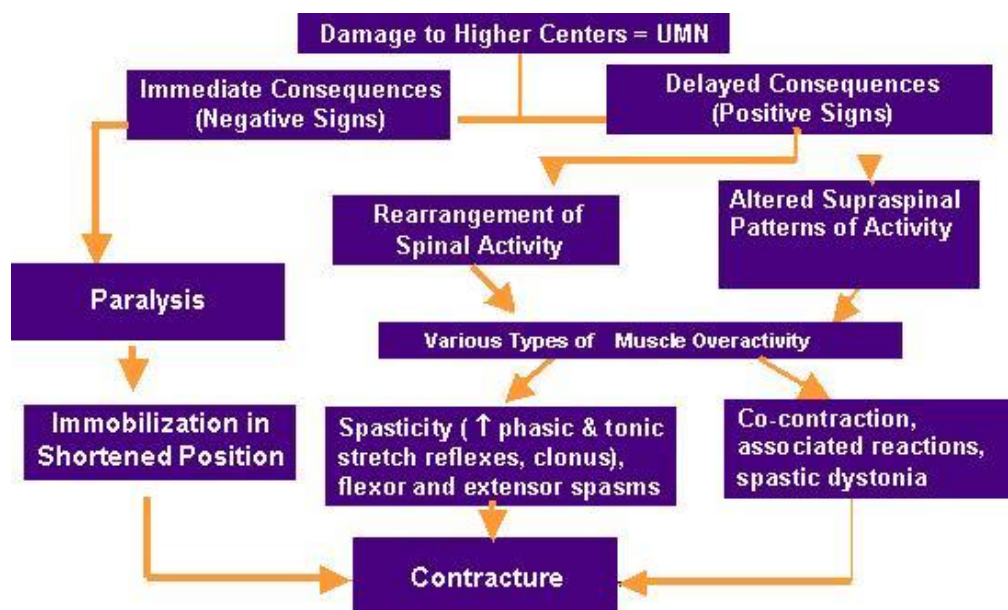
Spasticity: defined as velocity dependent resistance to stretch, where an absence of inhibition results in excessive contraction of the muscles, ultimately leading to hyperreflexia (overly flexed joints). It mostly occurs in disorders of the central nervous system (CNS) impacting the upper motor neuron in the form of a lesion.

Assessment:

- Because spasticity has important implications for the comfort and mobility of patients, it is important to have an approach to assessment. This also provides a means by which response to treatment can be measured/evaluated.

- To assess spasticity clinically (without the use of special devices) and to verify the velocity-dependency, the intensity of the muscle tone elicited at very slow and at rapid passive joint movement is compared and graded. It can also be quantified by measuring the joint angle at which the increase in muscle tone is encountered in a fast stretch and comparing it with the joint angle in a slow passive range of motion (ROM). There are a number of clinical tools that have been developed to comprehensively and systematically assess spasticity, including the Tardieu scale of spasticity.

DETERMINING AND TREATING THE UNDERLYING CAUSE:



- **CAUSES OF SPASTICITY IN CHILDREN:**
 - Cerebral palsy (birth asphyxia, prematurity)
 - Post traumatic brain injury
 - Near drowning
 - Post meningitis inc TBM
 - HIV encephalopathy
 - Malignancies
 - Spinal cord injuries/tumours
 - Strokes
 - Neuro-degenerative diseases (Tay-Sachs, Retts etc)
- **GOALS IN MANAGEMENT** (treating the underlying cause is not usually an option):
 - Reduce pain
 - Improve functionality where possible and necessary (but not at the expense of pain)
 - Improve quality of life
 - Improve social acceptability
 - Pharmacologic interventions/measures:

NON-DRUG MEASURES:

- Regular physical therapy and occupational therapy are important components of the management of spasticity. If the child cannot have access to formal rehabilitative therapy because of limited resources, basic range of motion exercises can be taught the care-takers.
- Massage and Reiki can be very healing, therapeutic forms of symptoms management in spastic patients, particularly those with associated pain.
- Additional interventions to consider depending on availability include Botox injections to affected joints/muscles, rhizotomy (a neurosurgical procedure that destroys problematic nerve roots in the spinal cord), and tendon lengthening.

DRUG MEASURES:

- NSAIDS are extremely effective analgesic agents in the management of pain associated with muscle spasms and spasticity.
- Additional medications used in the management of spasticity can be found in the table below.

Drug	Dose	Category	Notes
Diazepam	PO: 1-12mo: 250 mcg/kg BD 1-5 yrs: 2.5 mg BD 5-12 yrs: 5 mg BD 12-18 yrs: 10 mg BD; max total dose 40 mg	Benzodiazepine	May cause sleepiness when dose is first initiated. Use with caution in patients with liver disease. Clonazepam may also be used as an alternative, and has the benefits of being a longer-acting agent.
Baclofen	PO: 1-10 yrs: 0.75-2 mg/kg daily or 2.5 mg QD increased gradually according to age to maintenance: 1-2 yrs: 10-20 mg daily divided BD-TD 2-6 yrs: 20-30 mg daily divided BD-TD 6-10 yrs: 30-60 mg daily divided TD 10-18 yrs: 5 mg TD increased gradually; max 2.5 mg/kg	GABA receptor agonist	Use with caution, as may precipitate seizures. Ensure that existing seizures are under control before initiating.

DYSTONIA:

DEFINITION:

A neurological movement disorder, in which sustained muscle contractions cause twisting and repetitive movements or abnormal postures.

This is a common symptom in Children's Palliative Care as the spectrum of diseases seen include many neurodegenerative, and neurometabolic conditions.

ASSESSMENT:

Because dystonia is often associated with global impairment, assessment may be difficult. Dystonia may not be easy to differentiate from persistent focal seizures and the two often co-exist. Usually EEG is not helpful either, as with global impairment many children will have a grossly abnormal EEG.

The impact of dystonia may include:

- Reduced/absent mobility and control of movement
- Profound fatigue
- Muscle contractures resulting in bone/joint deformity
- Excessive secretions and drooling

These children often experience a painful vicious cycle as the spasms/dystonic symptoms are easily triggered and in turn cause pain. On obtaining a history, remember to ask about common triggers (which often include a full bladder, constipation, noise, fear and sudden touch).

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Causes:

- Damage to the brain (basal ganglia, thalamus and cerebellum), encephalopathy post-infection, birth hypoxia and cerebral bleeding, spinal cord lesion (upper motor neurons)
- Drugs (Haloperidol and Metoclopramide due to Extra-pyramidal side effects)
- Idiopathic

Management:

- Counselling and Explanation

Dystonia can be a distressing symptom for the child and the family. Adequate counselling to both the child, in an age appropriate manner, and the family must be offered. Families need to understand the triggers for their child and how to avoid them wherever possible. An explanation of the dynamic nature of the symptom needs to be explained; many children require more than one drug to manage the symptoms with a full range of non-pharmacological treatments.

- Involvement of the multidisciplinary team (physical and occupational therapist, social worker for accessing resources to enhance function/mobility, chaplain for spiritual support)
- Treat the cause
 - Stop causative/contributing drugs
 - Check for constipation/urinary retention and address if an issue
 - Ensure patient is in a routine, familiar environment as often as possible
 - Treat other causes of pain (such as acid reflux which is common in children with neurodegenerative conditions)
 - Control epilepsy

NON-DRUG MEASURES:

- Gentle handling, warm baths, massage, physio (which can be taught to caregiver)

DRUG MEASURES:

- Pharmacological treatments are only effective in 50% of dystonia and may make things worse overall; a reduction in tone may leave the child unable to sit/stand.
- One must weigh the risks and benefits of side effects, such as sedation and drooling.
- Baclofen and/or Diazepam are first line. Titrate the dose according to response.

ANXIETY

DEFINITION:

Feeling of apprehension caused by anticipation of danger, which may be internal or external.

Anxiety is an unpleasant emotion that may vary in severity from a mildly worried state to a state of uncontrolled panic.

Normal anxiety needs to be distinguished from pathological anxiety. Anxiety can be a normal accompaniment of growth and change, exposure to a new situation, etc. For example, in children, it is developmentally normal to develop “stranger anxiety” in late infancy and early toddlerhood, which they ultimately outgrow.

Pathological anxiety is an inappropriate response to a perceived threat, either in that the response is abnormally intense or lasts abnormally long.

ASSESSMENT:

Anxiety may aggravate other symptoms encountered in paediatric palliative care (eg dyspnoea, pain, diarrhoea).

Anxiety is often co-morbid with depression.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Always consider other organic underlying conditions that may “look like anxiety” eg: electrolyte disturbances, hyperthyroidism, hypoxia, pheochromocytoma, arrhythmias etc. as well as possible side effects of drugs (including illicit substances).

Procedural anxiety/ anticipatory pain are important considerations in the palliative care setting and especially where children with chronic illnesses have been exposed to recurrent painful procedures/intervention.

NON-DRUG MEASURES:

Psycho-therapy

Behaviour therapy

Relaxation techniques

Distraction

DRUG MEASURES:

Short term consider use of Lorazepam or Clonidine (1- 4ug/kg/dose 6 – 12 hourly)

With chronic or generalized anxiety it is best to consult a child psychiatrist.

DEPRESSION

DEFINITION:

A mental state of depressed mood, characterized by feelings of sadness, despair and discouragement.

Relevant statistics:

Incidence amongst pre-schoolers: 0.3 – 0.9%

Incidence amongst school going children: 2%

Incidence amongst adolescents: 5%

Incidence amongst chronically ill children: 20%

Incidence amongst chronically ill adolescents: 40%

It is estimated that suicide is attempted by 28% of chronically ill adolescents. Symptoms of depression are extremely common in children with chronic diseases, and are especially common in diseases where insight and intellect are maintained in the presence of deteriorating health.

ASSESSMENT:

Depression can be difficult to assess in the paediatric palliative care patient, as many of the “vegetative features” are similar to disease manifestations (anorexia, insomnia, etc.).

Infants and preschoolers are not able to express feelings of sadness in language. Therefore, depressive symptoms must be interpreted from behavioral changes, including apathy, withdrawal from caregivers, delay or regression of developmental milestones and failure to thrive that has no organic cause^{IX}.

Classic symptoms of depression in paediatric patients:

Young children:

Presents more often with more “positive symptoms”-

- Somatic complaints (abdominal pain, headache)
- Psychomotor agitation, restlessness
- Mood congruent hallucinations

Adolescents:

Presents more often with more “negative symptoms”-

- Apathy

- Hopelessness
- Psychomotor retardation
- Delusions
- Nihilism

In older patients/adolescents, standardized tools can be used to screen patients for depression.

Psycho-social stressors should always be assessed in the depressed child (e.g., is there financial strain in the family/household? Is there any concern for history of abuse?).

Consider the benefit of laboratory tests to rule out medical causes of depression, such as a complete blood count, liver function tests, electrolytes and renal function tests, thyroid function tests, and EEG to rule-out seizures.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Conditions Associated with Depression That May Apply to Children

Infections

Infectious mononucleosis

Human immunodeficiency virus infection

Neurologic disorders

Epilepsy

Post-concussion

Endocrine

Diabetes

Hyperthyroidism

Hypothyroidism

Addison's disease

Medications

Barbiturates

Benzodiazepines

Corticosteroids

Cimetidine (Tagamet)

Aminophylline

Anticonvulsants

Others

Alcohol abuse

Drug abuse and withdrawal

Oral contraceptives

Electrolyte abnormality

Hypokalemia

Hyponatremia

Anemia

Wilson's disease

NON-DRUG MEASURES:

Plan for hospitalisation if the child appears to be suicidal. Ideally, hospitalization should take place in a location where support in evaluation and treatment could be obtained from a psychiatric specialist (even if it is by phone consultation).

A combination of therapy and pharmacologic agents is thought to be the gold standard in the treatment of depression. This form of treatment is indicated in those patients in which depression is impairing functioning. Psychotherapy should always be considered in depressed patients who can access this type of care (recognizing that its availability will be limited in many locations).

Distraction, therapeutic touch, Reiki, and meditation are all non-pharmacologic approaches to treatment that may help to restore “normality” for depressed children.

DRUG MEASURES:

Commonly used anti-depressants in children include the selective serotonin reuptake inhibitors (SSRIs), given their generally lower side-effect profile. Tri-cyclic anti-depressants may also be used, but must be used with caution because of their potential side-effects (particularly prolonged QT syndrome; an ECG should always be obtained prior to initiating TCA therapy).

The potential for drug interactions should be taken into consideration when deciding on anti-depressant therapy in paediatric palliative care patients, who are often on multiple drugs at any given time.

There is some evidence that SSRIs may increase suicidal thoughts and/or behaviours in paediatric patients following the initiation of therapy. Therefore, patients should be monitored closely during the first two months that SSRIs are initiated, ideally with weekly visits or phone assessments for the first month.

One should always consider how uncontrolled pain may be contributing to symptoms in a depressed patient. Optimize pain control regimens of those patients with uncontrolled pain and symptoms of depression.

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HAEMATOLOGICAL PROBLEMS

BLEEDING/HEMORRHAGE:

DEFINITION:

Bleeding, otherwise known as haemorrhage, is the loss of blood from the circulatory system. Bleeding can occur internally, or inside the body, or externally, in which loss is through a naturally occurring body orifice, or through a break in the skin.

Desanguination refers to a massive blood loss.

Haemorrhage in which >15% of blood loss is experienced will typically lead to changes in the patient's vital signs, including tachycardia and possibly hypotension. However, in patients with palliative care needs, whom are already ill and compromised, these changes might be seen with less blood loss.

ASSESSMENT:

- Perform a comprehensive review of the patient's history of present illness and past medical history.
- Identify family history of bleeding/bleeding disorders.
- Determine the location(s) of the bleeding and the frequency of the bleeding episodes.
- What typically makes the bleeding episodes stop?
- Does the patient have any other associated symptoms with the bleeding? (e.g., palpitations, anxiety, pain)
- On physical exam, take note of vital signs. Hypotension and tachycardia might be an indication of significant blood loss and/or anaemia.
- Is there any physical evidence of bleeding on exam (e.g., pallor, old or crusted blood in nasal orifices, bleeding gums, bruising, petechiae, purpura)? Is there splenomegaly (which might be a cause of platelet sequestration)?
- Depending on the goals of care and the possibility of transfusion as a therapeutic option, it may be appropriate to check blood counts to determine if the patient has severely low platelet and RBC counts.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

The paediatric palliative care patient might experience bleeding is a possibility in the following situations:

Disorders of low platelets	Other causes of bleeding
Thrombocytopenia secondary to malignancy	Liver failure
Drug-induced thrombocytopenia	Bleeding disorder/clotting disorder (e.g., Haemophilia, Von-Willenbrand's disease)
Ideopathic thrombocytopenia (ITP)	Anti-coagulation (Warfarin or Heparin therapy)
HIV-associated thrombocytopenia	Tumour invasion into a blood vessel
Hepatitis-C associated thrombocytopenia	
Congenital thrombocytopenia	

NON-DRUG MEASURES:

If reversible and in earlier phases of the disease, attempts may be made to treat the bleed aggressively. Children with thrombocytpaenia who are at risk for bleeding, and who have a reasonable quality of life, can benefit from regular platelet transfusions. However, in the terminal phase of disease, aggressive management is usually inappropriate. If bleeding is a possibility, a sensitive and gentle pre-emptive discussion with the child's parents is helpful to warn the parents that this may occur. Red or dark towels are always suggested to be on hand so as to minimize the shock and fear created by the sight of a lot of blood.

Relaxation techniques, such as breathing exercises and guided-imagery can be used in children who have associated anxiety.

First Aid for Nosebleed

The following procedure is recommended by the American Academy of Otolaryngology—Head and Neck Surgery for minor nosebleeds:

1. Sit with your head leaning forward.
2. Pinch the nostrils shut, using your thumb and forefinger in such a way that The nasal septum (the nose's midsection) is being gently squeezed.
3. Hold for 15 uninterrupted minutes, breathing through your mouth.
4. At the same time, apply cold compresses (such as ice in a soft cloth) to the area around the nose.
5. For the next 24 hours, make sure your head is elevated above the level of your heart.
6. Also, wait 24 hours before blowing your nose, lifting heavy objects, or exercising strenuously.

Other self-care tips after the nose has stopped bleeding:

- Do things to keep the nostrils moist such as:
 - ✧ Use a cool-mist vaporizer or humidifier in your bedroom, especially in the winter.
 - ✧ Put a dab of petroleum jelly inside the nostril.
- Don't pick or rub the nose.

DRUG MEASURES:

Non-Acute, Small bleeds:

- Oral tranexemic acid: Child 1 month–18 years: 15–25mg/kg (max. 1.5g) 2–3 times daily (Cautions: to reduce dose in renal failure; not for use in children with haematuria because of the risk of clot retention)

- Tranexemic acid by intravenous injection over at least 10 minutes:

Child 1 month -18 years: 10mg/kg (max 1g) 2-3 times a day

- Tranexemic acid by continuous intravenous infusion:

Child 1 month -18 years: 45mg/kg over 24 hours.

- Tranexemic acid Topical treatment:

Apply gauze soaked in 100mg/ml injection solution to affected area.

- **Merical nasal tampon or pack nose with gauze soaked in Adrenalin**

Menorrhagia:

Child 12-18 years: Tranexemic acid 1g 3-4 times daily for up to 4 days; maximum 4g daily (initiate when menstruation has started). Vaginal bleeding can respond to oral progestogen

Liver dysfunction and other coagulation abnormalities:

- Liver dysfunction with coagulation abnormalities can be treated with

Vitamin K both orally (prevention) or by injection (acute bleed). Vitamin K oral, IM or slow IV bolus over 15-30 mins at 300mcg/kg daily for children 1 month to 12 years, or 10mg daily for children over 12 years.

Bleeding gums:

- Oral tranexemic acid: Child 1 month–18 years: 15–25mg/kg (max. 1.5g) 2–3 times daily
- Tranexemic acid Mouthwash 5% solution:

Child 6-18 years: 5-10mL 4 times a day for 2 days. Not to be swallowed.

OR use undiluted IV preparation and apply to bleeding point OR dilute 1:1 as a mouthwash

- Absorbable haemostatic agents such as Gelfoam or Gelfilm.

Bleeding wounds:

- Topical Adrenaline 1:1000 on gauze and directly to the wound (may not be practical approach if the wound is very large)

Acute and Catastrophic Bleeds:

Preparation for such a situation allows easier management of the situation, should it arise. This includes putting practical measures into place to relieve the distress of the child should it occur. An anxiolytic such as midazolam or diazepam is useful and should be readily available.

Remember that the subcutaneous route may not be well enough perfused in this situation, so buccal Midazolam or rectal Diazepam may be used instead.

- Midazolam Buccal doses for acute anxiety 100mcg/kg as a single dose (max dose 5mg)
- Diazepam by rectum (rectal solution):
 - Neonate: 1.25–2.5mg repeated once after 10min if necessary
 - Child 1 month–2 years: 5mg repeated once after 10min if necessary
 - Child 2–12 years: 5–10mg repeated once after 10min if necessary
 - Child 12–18 years: 10mg-20mg repeated once after 10min if necessary

Morphine can also be used but the bleed may not actually be painful. The doctrine of double effect may apply here.

NEUTROPAENIC SEPSIS:

DEFINITION:

Neutropaenia= A blood disorder characterized by an abnormally low number of neutrophils. Neutrophils comprise 50-70% of circulating white blood cells and are the main form of defence against bacterial infections.

Sepsis= A potentially fatal condition characterized by whole-body inflammation caused by infection.

Neutropaenia is typically classified according to **absolute neutrophil count (ANC)** as follows:

- Mild neutropaenia ($1000 \leq \text{ANC} < 1500$) — minimal risk of infection
- Moderate neutropaenia ($500 \leq \text{ANC} < 1000$) — moderate risk of infection
- Severe neutropaenia ($\text{ANC} < 500$) — severe risk of infection.

Of palliative care patients to consider, children who have received chemotherapy in the last 5-14 days are at risk for neutropaenia.

A Neutrophil count $< 500/\text{mm}^3$ or a fever of $> 38.5^\circ\text{C}$ are classically indications for starting empiric antibiotics after taking blood cultures and other cultures as appropriate.

Patients with a neutrophil count $< 1000/\text{mm}^3$ have an increased risk of bacterial infection, usually from the skin, URTI and GIT flora.

ASSESSMENT:

- Take a history and examine the patient for potential sites of infection:
 - URTI, Dental sepsis, oral ulcers, skin lesions, port and line sites, anal fissures, LRTI, GIT infections, embolic phenomena of septicaemia. Signs of infection may be minimal, so vigilance is required. Listen to the parent's description of behaviour.
- Consider the patient's goals of care and whether or not they are for curative treatment. If the focus of care is palliation and optimizing comfort while allowing death to occur naturally, IV antibiotics for neutropaenia may not be an appropriate approach to management. The option of not treating sepsis with IV antibiotics should ideally be discussed with patients who have a poor prognosis early on in the care planning.

- Explore what other symptoms are associated with the patient's illness (e.g., malaise, fatigue, rigors, nausea, dyspnoea, etc).
- Discuss each case with the Oncology Consultant who manages the child. Arrange urgent admission as appropriate. Get vascular access appropriately in a sterile manner if treatment of infection is a goal.
- ALWAYS do Blood Cultures and get an FBC and differential cell count when planning to treat.
- If indicated, send Urine for MC&S, Sputum for MC&S, do a CXR, send for stool MC&S and get a cross-matched blood sample. A Lumbar Puncture is usually contra-indicated as there is often associated thrombocytopenia.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Additional causes of neutropaenia include:

NON-DRUG MEASURES:

- ALWAYS talk to the child's parents and inform them of the possible concerns. Involve the child as much as is possible and appropriate.
- The child's comfort can be improved with cold compresses if complaining of/experiencing discomfort from fever. Avoid immersing a child in an ice-bath in the setting of fever.

DRUG MEASURES:

- Remember the role of anti-pyretics in maximizing comfort in patients with neutropaenia sepsis.
- In end-of-life patients with neutropaenia and sepsis that will not be treated with antibiotics, opioids and anxiolytics may need to be given to optimize comfort.
- In cases of infection in palliative care patients who have low grade fevers or higher neutrophil counts and who have evidence for a viral infection or a well localised site of infection, oral antibiotics may be appropriate and sufficient for treatment. However, these patients need to be carefully monitored over the next 24 hours.

- Persistent fevers may indicate the need for IV antibiotics. If uncertain, starting IV antibiotics may be safer, as they can be discontinued and changed to oral antibiotics by the Oncologist.

FIRST LINE Anti-biotics used in combination:

Piptazobactam 90mg/kg 6 hourly to a maximum of 2.5g 6 hourly

Amikacin Under 12 years - 25mg/kg daily to a maximum of 600mg

Over 12 years - 15mg/kg daily to a maximum of 900mg

SECOND LINE antibiotics (and in Suspected MRSA infection) :

Vancomycin 10mg/kg 6 hourly

THIRD LINE (and in suspected fungal infection):

Amphotericin B

Test dose of 1mg followed by starting dose of 0.5mg/kg. Then 1mg/kg alternate daily dose for empirical therapy OR 1mg/kg daily dose for proven fungal infection.

IF PROVEN ESBL:

Meropenem 30-40mg/kg 8 hourly

HYPERCALCAEMIA

DEFINITION:

Hypercalcaemia is a raised level of corrected calcium in the blood.

Total plasma calcium is the combination of free, ionised calcium and protein-bound calcium. If the albumin level is low, protein bound calcium is low. This may mask a high concentration of free, ionised calcium. Calcium is, therefore, 'corrected' for albumin level.

ASSESSMENT:

- Take a comprehensive history to assess for symptoms of hypercalcaemia. Symptoms usually start from irritation of the CNS and GIT systems.
- CNS symptoms include: headache, malaise, behavioural changes, irritability, proximal muscle weakness and confusion.
- GIT symptoms include: ileus with constipation, cramping, loss of appetite and nausea and vomiting. Bone pain, pruritis and conjunctivitis may occur. Ongoing hypercalcaemia even in the lower range may cause nephrogenic diabetes insipidus, renal failure and pancreatitis.
- Findings on examination relate to the cause and symptoms.
- Investigation: in a neonate, check the corrected calcium with serum protein and phosphate and PTH levels. May need to check sodium, potassium and magnesium. In older children, the history and examination are most important, and the same laboratory test may be needed.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Of all paediatric palliative care conditions, malignancies are those in which the occurrence of hypercalcaemia may be most notable. Even so, the incidence of hypercalcaemia in malignancy in children is far lower than in adults. Malignancies can include leukaemias, Non-Hodgkin lymphoma, Hodgkin lymphoma, Ewing sarcoma, neuroblastoma, Langerhans cell histiocytosis, and rhabdomyosarcoma.

Other causes of hypercalcaemia include endocrine disorders, genetic disorders, subcutaneous fat necrosis in the neonate, Multiple Endocrine Neoplasia (MEN), thyrotoxicosis, increased calcium intake, and chronic kidney disease.

NON-DRUG MEASURES:

Educate the family and patient about the symptoms typically seen with hypercalcaemia (“moans, groans, bones, and stones”).

Consider non-pharmacologic approaches for pain, nausea, and constipation described in prior sections of this manual.

DRUG MEASURES:

Treatment is aimed at treating the cause and managing the hypercalcaemia. The initial management is to address the dehydration related to the hypercalcaemia.

Use of isotonic sodium chloride solution intravenously is typically an appropriate choice. Loop diuretics (eg. Lasix) inhibit calcium reabsorption by the renal tubules so are useful. Watch electrolytes during diuresis.

IV bisphosphonates decrease calcium over 2-4 days (with effect seen in laboratory from 4 – 7 days). Studies indicate that Pamidronate (IV and oral) and Etidronate (oral) are safe to use in children, Pamidronate at a dose of 1-1.5mg/kg intravenously. IV Bisphosphonates should only be given after fluid rehydration and if closely monitoring renal function. If needed, consult with endocrinologists, oncologists, and/or nephrologists.

The pain and CNS symptoms associated with hypercalcaemia will largely improve as the hypercalcaemia is treated. However, as needed analgesics can be used to assist with treatment of bone pain, and appropriate pharmacologic measures can be taken to treat nausea and constipation as appropriate.

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CONSTITUTIONAL SYMPTOMS (and their associated syndromes)

DEFINITION:

A symptom indicating that a disease or disorder is affecting the entire body. Examples of constitutional symptoms include:

ANOREXIA: Absence or loss of appetite for food

ASTHENIA: Fatigue (physical and mental), easy tiring, general weakness

CACHEXIA: Profound weight loss due to catabolic state that combines weight loss, lipolysis, loss of muscle and visceral protein, anorexia, chronic nausea, and weakness

The pathogenesis of the syndromes associated with constitutional symptoms differ between disease entities.

Understanding these differences assists in the management.

The constitutional symptoms are especially common in children because of their high metabolic requirements especially during periods of rapid growth.

Constitutional symptoms cause considerable distress for family and friends. Feeding a child is a natural nurturing instinct and children's lack of interest in eating because of anorexia can be confusing and frustrating for family members.

Even though patients do not always experience hunger, families fear that they are "starving to death" or "wasting away".

ASSESSMENT:

In addition to assessing for symptoms that can often contribute to distress and impaired quality of life in children with constitutional illness (such as pain, nausea/vomiting, diarrhoea, depression, etc.), one should also take note of weight and other anthropometric measurements that can help to categorize the child's degree of nutritional impairment.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Three syndromes commonly associated with constitutional symptoms include: AIDS wasting disease, Kwashiorkor, and Marasmus. Constitutional symptoms can also be seen in any complex, chronic illness that significantly increases a child's metabolic demand (e.g., cancer, end-stage renal disease, disseminated TB).

AIDS wasting disease (Slims disease) is the involuntary loss of more than 10% of body weight, plus more than 30 days of either diarrhoea, or weakness or fever (AIDS Infonet January 10, 2005)

Due to the high prevalence of malnutrition in South Africa, all persons caring for children should be well trained in assessment and management of malnutrition that may complicate the clinical

presentation and course of many patients who also have chronic illnesses in need of palliative care.

WELLCOME CLASSIFICATION OF MALNUTRITION:

Expected weight for age (EWFA)	Oedema	No Oedema
> 60% EWFA	Kwashiorkor	UWFA
<60% EWFA	Marasmic-kwashiorkor	Marasmus



Management of malnutrition:

It is beyond the scope of these guidelines to address the management of malnutrition.

Readers should refer to local South African guidelines and should work alongside local nutritional rehabilitation programmes and feeding schemes.

NON-DRUG MEASURES FOR CONSTITUTIONAL SYMPTOMS:

In children with incurable illness who are approaching end of life, it is important to weigh the benefits and burdens of artificial nutrition and hydration (eg., nasogastric feedings and IV fluids). In these cases, artificial nutrition and hydration can actually contribute to worsening of symptoms that will impact on the patient's comfort, such as respiratory secretions, nausea/vomiting and abdominal distension, and oedema (in the lungs and peripheral tissues).

Educate families about the fluctuations that can be observed in the appetite of children with constitutional symptoms, which will help them to understand that this can be a normal component of the underlying disease process.

DRUG MEASURES:

When accessible, appetite stimulants, such as Megestrol Acetate (Megace) have been used to enhance the nutritional status and quality of life of children with life-threatening illness (including cancer, HIV/AIDS, end-stage renal disease, and cystic fibrosis). These medications should be considered if the child is expressing a desire to eat more (yet is generally disinterested in eating secondary to anorexia), and in children with anorexia who are experiencing co-morbidities secondary to poor nutritional status (e.g., more frequent infections, skin-breakdown, etc). If starting a patient on such a medication, it is important to ensure that the family has adequate resources to supply food to the child as he or she desires it.

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DERMATOLOGICAL PROBLEMS IN PEDIATRIC PALLIATIVE CARE

BEDSORES DECUBITUS ULCERS:

DEFINITION:

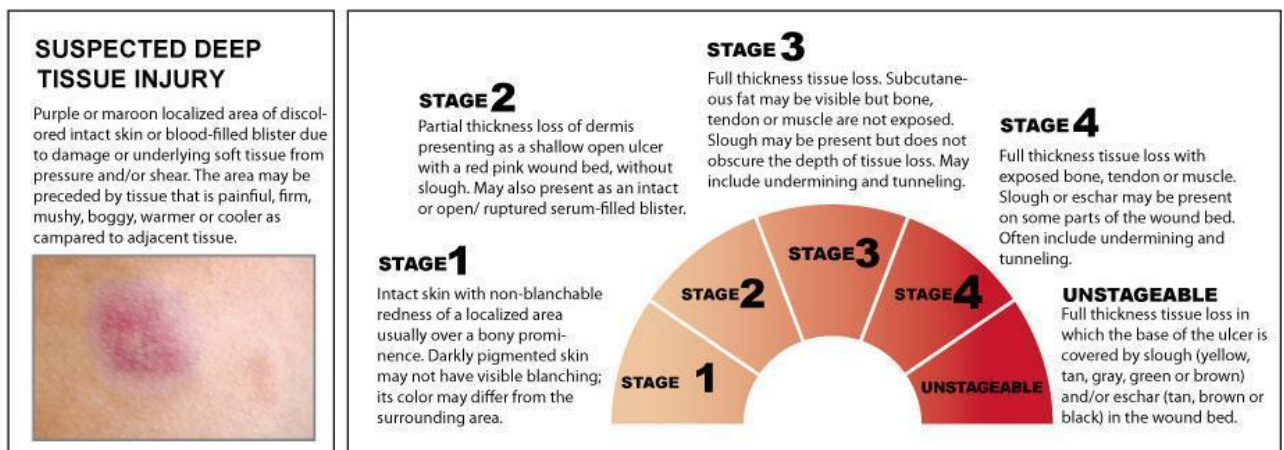
Lesions caused by unrelieved pressure on soft tissues which diminishes or completely obstructs the blood flow to the superficial tissues. Also known as pressure sores.

Bedsore are uncommon in infants, given their frequent movement and light weight. They are more common in older children and adolescents, especially when immobile, obese, and/or neuromuscularly impaired.

ASSESSMENT:

Location- In hospitalized children, pressure sores are likely to develop around nasal cannulae, IV drip sites, and the sites of monitoring probes (especially in newborns and prems). In older and heavier bed-bound children, they are likely to develop in dependent areas (including the sacrum, buttocks, elbows, heels).

Pressure ulcers can be described by stages as depicted below. This can be helpful in monitoring progress of treatment.



Determine what the family may be doing at home to treat bed sores (e.g., traditional therapies, topical medications, etc.). At times, such treatments may in fact be exacerbating the bed sores and should therefore be discontinued.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

Factors contributing to the development of pressure sores:

Poor nutrition

Inability to recognise or react to skin discomfort (eg: neurological damage, sedation)

Incontinence

Nappies

Impaired healing processes: anaemia, immunodeficiency,
malnutrition Treatment related: chemotherapy and radiotherapy

Hypoxia

Obesity

NON-DRUG MEASURES:

Prevention is better than cure! Consider the following care practices in patients prone to bed sores:

- Regular turning
- Water beds, profiled foam, donuts, air mattresses (static or alternating pressure), sheep skins (choice according to availability and child's risk for developing pressure sores)
- Nutritional support including a multi-vitamin supplement
- Regular bathing to prevent stagnation of secretions and other irritating substances

Bedsore that are stage 2 or greater should be packed and/or dressed accordingly, with a plan for regular dressing changes. Consult with nursing staff with experience in wound care to formulate an adequate management plan.

DRUG MEASURES:

Remember that bed sores can be quite painful! Pain can be managed with level 1-2 analgesics initially, and scaled up if necessary.

OTHER DERMATOLOGIC CONDITIONS THAT MAY BE SEEN IN PEDIATRIC PALLIATIVE CARE PATIENTS:

- Herpes Zoster (Shingles)
- Severe chicken pox
- Stevens Johnson Syndrome
- Burns
- Eczema
- Ichthyosis
- Scalded skin syndrome
- Rashes related to ART

- Graft versus host disease
- Dermatitis secondary to chemo and radiotherapy complications

In each of these conditions, treatment of the underlying problem must be considered. Antibiotic treatment may be warranted in the case of bacterial super-infection. In addition, symptomatic management of pain, pruritis should always be addressed (both with non-drug and drug approaches).

The psychosocial impact of physical disfigurement from severe dermatologic conditions can be profound. Affected children and their families can become isolated as result, and may need the additional support of a social worker, chaplain, or spiritual advisor to help them cope.

LYMPHOEDEMA:

DEFINITION:

A condition of localized fluid retention and tissue swelling caused by a compromised lymphatic system.

Secondary Lymphoedema can occur in children, as a result of cancer or cancer-related treatment, infection or a surgical procedure. It is not common.

There is usually swelling of a limb or an extremity. The genitalia may be included.

ASSESSMENT:

Diagnosis is by history and clinical examination. Lymphoscintigraphy can confirm or exclude it. An MRI can also assist.

Possible complications include skin changes, nail changes, pain and difficulty with daily activities or play. Children can have difficulty wearing shoes. Infection can occur in the form of lymphangitis or cellulitis.

The child's psychological status should be assessed as well, as deformity associated with lymph oedema can be distressing and socially isolating.

TREATMENT MEASURES:

Non-surgical treatment:

Lifestyle – Avoiding trauma and diligent hygiene and skin care is very important. Prevention of worsening is difficult, but parents can be encouraged to minimize trauma to the underarm and groin areas. Elevation of the affected limb is difficult to enforce, but it may help. Specific exercises are thought to promote lymphatic drainage. The goal should be for the child to work

up to a level of exercise that helps with overall fitness and assists with joint and limb strain, but that does not make the lymph oedema worse. Remember that exercise increases the work of the lymphatic system and so should be performed cautiously.

Static compression – Supportive stockings and elastic custom garments worn throughout the day to reduce the size of the affected area can slowly lead to an improvement in symptoms. Daily massage regimens taught by trained medical professionals may also be part of the treatment regime.

Pneumatic compression – Mechanical pneumatic compression pumps can be used overnight to reduce the child's limb size and improve symptoms.

Treatment of infection if it occurs should be prompt and with appropriate antibiotics and bed rest.

Psychosocial support should be provided to the child and family who are struggling with the emotional challenges of lymphoedema and its associated deformities.

Surgical treatment:

Surgery may be recommended for children with a longer life expectancy and unacceptable symptoms or recurrent infection.

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IMMUNE RECONSTITUTION SYNDROME IN CHILDREN

DEFINITION:

A condition seen in some cases of immunosuppression (including AIDS), in which the immune system begins to recover, but then responds to a previously acquired opportunistic infection with a profound inflammatory response that paradoxically makes the symptoms of infection worse.

Up to 21% of children after starting Anti-retroviral therapy will develop IRIS.

IRIS in children has not been associated with increased mortality rates, but under-recognition and under-reporting are very likely.

ASSESSMENT:

In patients on ARVs, verify duration of time since initiation of ARVs, current drug regimen, and whether or not regimen was ever changed/adjusted.

Consider obtaining microbiological confirmation of TB and determining whether or not the child has been adherent to TB therapy if previously initiated.

DETERMINING AND TREATING THE UNDERLYING CAUSE:

IRIS can be associated with:

- mycobacterial organisms
- varicella zoster virus
- herpes simplex virus
- cutaneous mycoses
- progressive multifocal leucoencephalopathy
- *Cryptococcus neoformans*
- *Bacillus Calmette Guerin* (BCG)-associated IRIS (common in South Africa)

General Management Approach:

For more severe IRIS cases, anti-inflammatory therapy may be considered. Although there are no randomised controlled trials of IRIS treatment in children, corticosteroids have proven to be of benefit in adults with moderate-to-severe TB-IRIS manifestations. Risks include reactivation of herpes virus infections, Kaposi sarcoma and metabolic side effects if prolonged use is necessary. Corticosteroids should be used in the setting of life-threatening space-occupying lesions such as inflammatory lymph nodes obstructing airways or intracranial swellings such as tuberculomata.

Other aspects of Management are similar in Children and Adults:

- Explanation to patient and family

As in adults this is the most important intervention. Explanations to the child need to be age appropriate and child friendly.

- Institute non-pharmacological interventions

Always consider using non-pharmacological approaches such as distraction, play, relaxation techniques and positioning in symptomatic children.

- Prescribe appropriate first line treatment

For pain in children we are now using the WHO 2 step approach. See the section on pain in children.

Refer to and discuss with local paediatric experts

- Review assessment and management

Prevention

- PMTCT
- Improved TB screening
- INH prophylaxis
- Early initiation of ART
- Prevention of malnutrition

The high estimated risk of BCG disease in HIV-infected children and the lack of evidence for protective efficacy of BCG vaccine in HIV infected children, led the World Health Organisation (WHO) in 2007 to publish revised recommendations to defer BCG vaccination in HIV exposed infants and to avoid BCG vaccination in infants who are confirmed to be HIV-infected.

Bacillus Calmette Guerin (BCG)-associated IRIS:

Management:

There are no prospective studies evaluating whether antimycobacterial therapy directed against *M. bovis* BCG or anti-inflammatory therapy alters the outcome of local or regional BCG-IRIS; There seems to be no difference in time to resolution between those who receive antimycobacterial therapy and/or corticosteroid treatment and those who received no medication.

Distant/disseminated disease should be treated with Antimycobacterial therapy as case fatality rates >70% have been reported in infants. *M. bovis* BCG is inherently resistant to pyrazinamide and therapy usually comprises rifampicin, isoniazid, ethionamide or ethambutol and a fluoroquinolone such as ofloxacin. Pharmacokinetic interactions involving rifampicin and protease inhibitors or nevirapine may result in subtherapeutic antiretroviral plasma concentrations and require modification of the ART regimen adding complexity and challenges to adherence.

Although surgical excision or debulking of lymph nodes or inflammatory tissue has not shown to improve outcome significantly, the therapeutic aspiration of pus appears to result in less scarring and better outcome.

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PAEDIATRIC PALLIATIVE CARE EMERGENCIES:

Uncontrolled and distressing symptoms in children should be considered a medical emergency and need to be actively treated. Emergencies one might encounter in pediatric palliative care include:

- Severe pain
- Seizures
- Hemorrhage
- Agitation
- Difficulty breathing and airway obstruction
- SVC Syndrome
- Spinal cord compression
- Neuro-irritability
- Urinary retention

Most emergencies can be anticipated by knowing the natural history of a disease and from knowledge of the individual child's history. These emergencies should be thought about in advance and a proposed management plan should be written in an ADVANCED CARE PLAN as early in the child's course of illness as possible.

Families should be forewarned about possible emergencies and walked through the approach to management. In the advanced care planning stage, it is important to determine family's preference for where they want to be in an emergency (e.g., stay at home, vs. admit to hospital or hospice). If the family's preference is to stay at home, one must ensure that they have adequate access to supplies and medications needed to manage potential emergencies. Providing them with an emergency contact number for a health professional who can be of assistance if need be is also of immense value.

ASSESSMENT:

Investigation, management and treatment of palliative care emergencies With all emergencies it is important to consider^{XI}:

- Do I need to know the underlying cause or can I manage the symptom effectively without confirming the cause?
- Is the underlying cause likely to be treatable?
- Are investigations of the underlying cause appropriate (for example, are they invasive, do they require being in hospital etc.)?
- Will treating the underlying cause improve prognosis?
- How effective could any potential treatment be?
- How harmful could any potential treatment be?
- Will the child have to move to another location for the investigation and/or treatment? Will this be possible?
- What are the wishes of the child and family in the event of this emergency?

DETERMINING AND TREATING THE UNDERLYING CAUSE^{XI}:

The emergencies of severe pain, hemorrhage, seizure, and anxiety were handled in previous sections of this guideline. Please refer to those sections for a management approach to those specific problems.

Breathlessness/Profound dyspnoea:

Breathlessness should be anticipated in the following situations:

- Reduced lung volume (e.g., pulmonary hypoplasia, tumour growth, chronic lung disease)
- Upper airway obstruction (e.g., tumour)
- Pneumothorax (may occur in pulmonary metastasis/lung tumour)
- SVC syndrome
- Pulmonary oedema (eg cardiac failure)

Treatment of the underlying cause should always be considered, but may not be appropriate or possible. Treatment options may include:

- Steroids and radiotherapy or chemotherapy for malignant disease
- Chest drain for pneumothorax
- Diuretics in pulmonary oedema
- Antibiotics for chest infection

Frequently, when severe dyspnoea or breathlessness suddenly occurs in the palliative care patient, it is a terminal event. Comfort can be maximized with the following approach:

Give buccal midazolam 0.5mg/kg and buccal morphine 0.1 mg/kg X 1. Repeat every 10 -30 minutes as needed until the child is settled. Consider setting up a continuous subcutaneous or intravenous infusion of Morphine and Midazolam or Lorazepam, if this is an option.

Consider adding IV Furosemide in children with suspected pulmonary edema if this is an option.

Superior Vena Cava (SVC) Obstruction:

SVC obstruction is most likely to occur in children with mediastinal tumours. Typical signs of SVC obstruction are:

- Breathlessness
- Headache
- Visual changes
- Dizziness
- Swelling of face, neck, arms

Emergency treatment is usually with steroids, usually dexamethasone (1-2 mg/kg/day up to 16mg maximum). Radiotherapy and/or chemotherapy may then be considered. It is important to remember to manage associated dyspnoea with opioids and diuretics as appropriate.

Spinal Cord Compression:

This is a true medical emergency and prompt appropriate treatment is essential. It is most commonly seen in children with intramedullary metastases, intradural metastases or extradural compression (vertebral body metastases, vertebral collapse, interruption of vascular supply).

Early signs of spinal cord compression include: • Back pain

- Leg weakness
- Sensory impairment in the legs

Late signs of spinal cord compression include:

- Profound weakness
- Noting a sensory level
- Disturbance in sphincter function

Emergency treatment is with steroids, usually dexamethasone (1-2 mg/kg/day up to 16mg maximum). Radiotherapy and/or chemotherapy may then be considered. Spinal surgery may also be an option.

Neuro-irritability:

This diagnosis is often one of exclusion. It is most frequently a problem in children with severe birth asphyxia and associated cerebral palsy, but may also be seen in children with neurodegenerative disorders, metabolic disorders/inborn errors of metabolism, and epilepsy syndromes. In regards to symptoms, these children can cry for hours, without any response to comfort or analgesia.

Medication that can be helpful includes:

Phenobarbital (1-4mg/kg once to twice daily)

Buccal midazolam (0.5mg/kg as needed)

Haloperidol (0.5-1 mg PO once daily-TD PRN. Safe for use in children >3 yrs of age. May be increased to max of 5 mg in 24 hours. Obtain EKG prior to starting to r/o prolonged QT syndrome).

Gabapentin: 5 – 10mg/kg/dose 8 -12 hourly.

Midazolam and Haloperidol can be used in a crisis situation when the child needs something to break the cycle of crying. It should not be considered as ‘treatment’ for the irritability, but as an essential drug for crisis management.

Urine retention:

The most usual causes of urine retention are:

- Side effect of morphine
- Constipation
- Spinal cord compression
- Solid tumours

Address by treating the underlying cause. Opioid associated urinary retention can be addressed with the following approaches:

- Consider rotating opioids every few weeks
- Warm baths to encourage the child to pass urine
- Creating a relaxed environment
- Suprapubic massage
- Short-term catheterization

In the case of constipation, manage as best possible from “above” (with enteral agents and dietary adjustments) and “below” (with enemas and/or suppositories).

Urinary retention secondary to cord compression or solid tumour may be managed with steroids and radiation, if an appropriate and available option.

Be very cautious if considering catheterisation in a child with a solid tumour obstructing urinary outflow. Is likely that they may need suprapubic catheterization.

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