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Paediatric Sickle Cell Pain Management Clinical Guideline

Link to Sickle Cell Pain Management Algorithms

Aim

The aim of this guideline is to optimise pain control in patients with Sickle Cell Disease, presenting with a sickle cell crisis. Children presenting to the Emergency Department (ED) should be assessed quickly to define the nature of the presenting complaint.

Definition of terms

Sickle Cell Disease (SCD), Advanced Paediatric Life Support (APLS), Full Blood Count (FBC), Non Steroidal Anti Inflammatory Drugs (NSAIDS), Basic Life Support (BLS), Glasgow Coma Scale (GCS), AVPU scale ("alert, voice, pain, unresponsive").

Target Patient Population

This guideline is to be used in the treatment of patients who present with a sickle cell crisis.

Target Users

This guide is directed at health-care professionals engaged in the care of patients with a Sickle Cell Crisis.

****** Please note the method for preparing Morphine / Oxycodone Nurse Controlled Analgesia (NCA) outlined* in this guideline reflects local practice in OLCHC. Practices in other hospitals vary. Please prescribe *Morphine / Oxycodone NCA as per local policy.******

Assessment

Initial Acute

- Basic resuscitation principals as per APLS (if required)

- Assessment must include:
  - Level of Consciousness
  - Perfusion
  - Oxygenation
  - Pulse
  - Blood pressure
  - Respiratory rate
  - Oxygen Saturations
  - Pain Assessment with Pain score

Note: All children have post triage monitoring of vital signs as per their triage category.

Ongoing assessment

Routine investigations
All children should have vascular access and the following investigations:

- FBC */ Reticulocyte count
- Blood Group & screen *
- Urea, Electrolytes *

- Creatinine*
- LFT's, LDH

(* Urgent requests)

If indicated:

- Blood cultures
- Urinalysis and urine culture
- Throat swab
- Viral serology (including Parvovirus titres)

Note: Patients on iron chelation with Desferrioxamine (DFO) or desferasirox, admitted with diarrhoea/abdominal pain, should have blood and stool screened for *Yersinia* and the iron chelation stopped whilst awaiting results of culture.

State ongoing monitoring and follow up required beyond initial presentation

Additional Investigations (if indicated)

- Chest X-ray (If there are chest or abdominal signs)
- Abdominal X-ray and amylase (If there are abdominal signs)
- Ultrasound of Abdomen
- Plain radiographs of symptomatic limbs (if differential diagnosis is other than bony sickle cell crisis)
- CT scan of brain (if neurological signs are present)

Management

This is supportive care (i.e. conservative) management unless there are indications for exchange transfusion (see below) which must be discussed first with the Consultant Haematologist on call. The aim of treatment is to break the cycle of sickling →hypoxia and acidosis → more sickling — all exacerbated by dehydration.

All patients are managed using protocol unless otherwise directed by the senior Haematologist on call.
Assess Pain regularly using developmentally appropriate pain scoring tools. Remember that Pain intensity is just one aspect of overall pain assessment.

Pain Scores
0=No pain,
Mild Pain: 1-3/10;
Moderate Pain: 4-6/10
Severe Pain: 7-10

Non-Pharmacological interventions:

- Reassurance-giving reassurance that the patient's pain will be relieved.
- Hot pack, warm bath.
- Establishing a position of comfort.
- Distraction technique (e.g. television, storytelling, computer games, school, music, play, artwork etc.) may be helpful for some children.
- Massage.
- Involve play specialist.

Analgesia

Pain in SCD may be severe and often underestimated by medical and nursing staff. The liberal use of an oral or parenteral analgesic is essential in painful crises.

Management of severe pain should include paracetamol, NSAIDS (unless contraindicated), opiate analgesics and adjuvant analgesic medication e.g. clonidine, ketamine, magnesium citrate. The following are suggestions but it is IMPERATIVE that your management strategy is decided at a local level and should include the administration of opiates by Nurse Controlled Analgesia infusion or Patient Controlled Analgesic infusion.
Please refer to Appendix 1 Algorithm - Moderate Sickle Cell Pain Episode and Appendix 2 Algorithm - Severe Sickle Cell Pain Episode by following this link

Monitoring

Observations including pain assessment, sedation, respiratory system, effectiveness of pain management are hourly in children receiving intravenous opioids as per local observation charts.

Respiratory Depression

Opioids depress all phases of respiratory activity including rate, minute volume, tidal exchange and rhythm. Respiratory depression is clinically significant when it is severe enough to require an intervention (i.e. stopping opioid infusion, providing physical stimulation or administering naloxone to reverse it and prevent respiratory arrest). Because more opioid is required to produce respiratory depression than is required to produce sedation, patients with clinically significant respiratory depression are usually sedated. **Monitoring sedation levels is as important as monitoring respiratory status**.

If excessive sedation or respiratory depression is suspected, stop any IV or Subcutaneous opioid Infusion, stimulate the patient (e.g. shout, shake &/or sternal rub), shout for help and commence Basic Life Support (BLS).

AVPU: Patient receiving opioids		Action
A	Awake, arousable, alert.	
V	Responds to voice only (drowsy & sleepy). Child may be sedated from opioid.	Obtain GCS, Consider reducing IV opioid.
P	Responds to pain stimulus only (deeply asleep, arousable only with deep or significant physical or painful stimulus).	Stop opioid, Obtain GCS. If opioid related over-sedation, contact haematology registrar on call, anaesthetist on bleep 8528, Opioid can be recommenced once child becomes more responsive.
U	Unresponsive.	Stop Opioid. Stimulate the child, administer oxygen, Call 2222, and obtain GCS. Give naloxone to reverse opioid. Inform Pain service.

Clear and useful definitions of respiratory depression in children are lacking.

General Guide for Respiratory Depression requiring an intervention

0-2 years	<18 breaths/minute
2 to 6 years	<14 breaths/minute
7 to 10 years	<10 breaths /minute
11 + years	< 8 breaths/minute

Management of opioid induced respiratory depression

SUSPECTED RESPIRATORY DEPRESSION OR OVERSEDATION:

- STOP the opioid infusion

- STOP all other infusions that could be contributing to sedation
- Attempt to rouse the patient
- **Call 2222** if appropriate
- If apnoeic: administer bag & mask ventilation with 100% oxygen
- If breathing: maintain airway, monitor oxygen saturation and administer oxygen via face mask at 8 L/min
- Check circulation. If pulseless: commence chest compressions
- Administer naloxone per instructions on the prescription if opioid toxicity is suspected
- Call PAIN SERVICE/MEDICAL TEAM for urgent review

The treatment of opioid overdose is the opioid antagonist **naloxone**[1]

Drug	Indication	Route	Dose	Caution
Naloxone	**Excess sedation when respiratory compromise is present**	Intravenous injection	2 – 10 microgram/kg, repeated as necessary to maintain reversal Max: 200 micrograms/dose.	Watch for rebound respiratory depression Naloxone has a short duration of action; repeated doses or infusion may be necessary to reverse effects of opioids with longer duration of action.

Morphine Sulphate Infusion for Sickle Cell Pain

***** Please note the method for preparing Morphine / Oxycodone Nurse Controlled Analgesia (NCA) outlined in this guideline reflects local practice in OLCHC. Practices in other hospitals vary. Please prescribe Morphine / Oxycodone NCA as per local policy.*****

When prescribing morphine infusion for children with severe pain due to sickle cell crisis:

- Give a loading dose via the infusion pump on starting the IV Infusion if no opioid has been administered PO or I.V.
- Start infusion at a minimum of 2mL/hour to ensure pain control early on.
- Increase the infusion rate if the child requires more than 4 NCA boluses each hour for two consecutive hours.
- As the pain improves the rate of infusion can be reduced.
- Prescribe NURSE CONTROLLED ANALGESIA rather than Patient Controlled Analgesia until pain is well controlled.

MORPHINE NURSE CONTROLLED ANALGESIA Criteria to provide NCA bolus:-Pain Score > 4. AVPU score V		
Drug	**Children< 50kg**	**Children > 50kg**
Morphine	**1mg/kg** to a total volume of 50 mL in Glucose 5%w/v or Sodium Chloride 0.9%w/v **1mL = 20microgram/kg**	**50 mg** to a total volume of 50mL in Glucose 5%w/v or Sodium Chloride 0.9%w/v **1mL = 1mg**
Loading Dose (if no oral or IV opioid received).	50 or 100microgram/kg (2.5mL or 5mL)	50 or 100microgram/kg
Bolus Dose	**1 – 2 mL** (20 - 40microgram/kg)	**1 – 3mL (1mg - 3mg)**
Lockout	**10 minutes**	**10 to 12 minutes**
Background Infusion **(mL / hour)**	**0.5 – 3mL/hour.** (10 to 60microgram/kg/hour.) **Start infusion at 2mL/hour.** Increase rate if pain is not controlled with frequent boluses (4 boluses/ hour in 2 consecutive hours) and a continuous infusion of 2mL/hour.	**0.5 – 4mL/hour** (0.5mg to 4mg/hour) **Start infusion at 2mL/hour.** Increase rate if pain is not controlled with frequent boluses (4 boluses/ hour in 2 consecutive hours) and a continuous infusion of 2mL/hour.
Note	Ensure Paracetamol, NSAID, Clonidine has been administered and consider Oral ketamine before increasing rate to 3mL/hour	Ensure Paracetamol, NSAID, Clonidine has been administered and consider Oral ketamine before increasing rate to 4mL/hour
Maximum 4 hourly Dose	25mL	30mL

In patients who are opioid tolerant (Those currently receiving opioids) the recommended starting doses may be ineffective. These patients may need to have their doses increased rapidly to provide adequate analgesia, and frequent reassessment is the key to providing adequate analgesia. If necessary seek specialist pain advice from the pain service or department of anaesthesia.

- Aim to reduce pain from severe to moderate and then moderate to mild.
- If pain is not controlled when the infusion is at its maximum rate for 4 hours, please liaise with Haematology team OLCHC.
- When prescribing an opioid infusion please ensure that other analgesic medication e.g., paracetamol, + NSAID,
  +/- clonidine are prescribed if there are no contraindications.
- Prescribe regular laxative. An anti-emetic is prescribed.
- Ketamine (oral) may also be prescribed
- Diazepam may also be required (muscle spasm).
- Magnesium citrate may be prescribed for pain, muscle relaxation.

Oxycodone Infusion Guideline Sickle Cell Pain

***** Please note the method for preparing Morphine / Oxycodone Nurse Controlled Analgesia (NCA) outlined in this guideline reflects local practice in OLCHC. Practices in other hospitals vary. Please prescribe Morphine / Oxycodone NCA as per local policy.*****

OXYCODONE NURSE CONTROLLED ANALGESIA **Criteria to provide NCA bolus:-Pain Score > 4. AVPU SCORE A OR V**		
NCA INFUSION	**Children< 50kg**	**Children >50kg**
OXYCODONE	**1mg/kg** to a total volume of 50 mL in Glucose 5%w/v or Sodium Chloride 0.9% w/v **1mL=20microgram/kg**	50 mg to a total volume of 50mL in 5% Glucose 5%w/v or Sodium Chloride 0.9% w/v **1mL= 1mg**
When converting from IV morphine to IV oxycodone		
Background Infusion **(mL / hour)**	**Run infusion rate at 33% less than the rate of the morphine infusion** *Example:* Morphine rate = 3mL/hour. Start oxycodone at 2mL/hour **0.5 – 3mL/hour.** (10 to 60microgram/kg/hour.) **Start infusion at 2mL/hour.** Increase rate if pain is not controlled with frequent boluses (4 boluses/hour in 2 consecutive hours) and a continuous infusion of 2mL/hour.	**Run infusion rate at 33% less than the rate of the morphine infusion** *Example:* Morphine rate = 3mL/hour. Start oxycodone at 2mL/hour **0.5 – 4mL/hour** (0.5mg to 4mg/hour) **Start infusion at 2mL/hour.** Increase rate if pain is still not controlled with frequent boluses (4 boluses/hour in 2 consecutive hours) and a continuous infusion of 2mL/hour.
Note	Ensure Paracetamol, NSAID, Clonidine has been administered and consider PO ketamine before increasing rate to 3mL/hour	Ensure Paracetamol, NSAID, Clonidine has been administered and consider PO ketamine before increasing rate to 4mL/hour

When patient starting on IV Oxycodone from start of this admission		
OXYCODONE	**1mg/kg** to a total volume of 50 mL Glucose 5%w/v or Sodium Chloride 0.9% w/v **1mL=20 microgram/kg**	**50 mg** to a total volume of 50mL in 5% Glucose 5%w/v or Sodium Chloride 0.9% w/v **1mL= 1mg**
Loading Dose **(if no PO or IV received)**	50 or 100 microgram/kg (2.5mL or 5mL)	50 or 100microgram/kg
Background Infusion **(ml / hour)**	**0 to 3mL/ hour.** (0 to 60 microgram/kg/hour.) **Start infusion at 2mL/hour.**	**0 to 4mL/hour** (0 to 4mg/hour) **Start infusion at 2mL/hour.**
Bolus Dose	**1 to 2mL** (20-40 microgram/kg)	**1mL to 3mL (1mg to 3mg)**
Lockout	10 to 15 minutes	10 to 12 minutes
Maximum 4 hourly dose	25mls	30mls

Opioid Rotation[2,3,4,5]

Opioid rotation is a strategy that refers to a switch from one opioid to another in an effort to improve clinical outcomes (benefits or harms). It begins with the selection of a new drug at a starting dose that minimizes potential risks while ideally maintaining analgesic efficacy.

"Best practice" recommendations are that, after calculating the equianalgesic dose of the new opioid it should be reduced by 25% to 50% to account for incomplete analgesic cross-tolerance (that is, physiologic tolerance established to the first opioid does not carry over completely to the new one, so the patient is more sensitive to the new opioid).[2] Doing this allows for a safety margin to help prevent overmedication; although, *initial under medication with the new agent is highly likely*.

Tolerance and Hyperalgesia

In the absence of disease progression, a decrease in the effectiveness of opioid analgesia has traditionally been attributed to opioid tolerance. It is now known that the administration of opioids can lead to both **opioid tolerance** (a desensitisation of antinociceptive pathway to opioids) and, paradoxically, to **opioid-induced hyperalgesia*** (OIH).

OIH is defined as a state of nociceptive sensitization caused by exposure to opioids. The condition is characterised by a paradoxical response whereby a patient receiving opioids for pain can become more sensitive to painful stimuli[1.]

OIH should be suspected when opioid treatment effects seems to wane in the absence of disease progression, particularly if found in the context of unexplained pain reports or diffuse allodynia** associated with the original pain and increased levels of pain with increasing dosages.

These mechanisms underlying the development of tolerance and opioid induced hyperalgesia are still not fully understood. It is thought to result from neuroplastic changes in the peripheral and central nervous system that lead to sensitization of the pronociceptive pathways.[2,5]

Treatment involves reducing the opioid dosage, weaning opioid off, or supplementation with NMDA receptor modulators (e.g. Ketamine)[2,4,5]

***Hyperalgesia** is an extreme exaggerated reaction to a stimulus which is normally painful.
****Allodynia** is a pain due to a stimulus which does not normally provoke pain.

Switching from IV to Subcutaneous Opioid

Occasionally IV access may be difficult and a child may need to be converted from **IV opioid to sub-cutaneous opioid,**

1. Calculate the total dose of opioid the child received in the past 24 hours, including any bolus doses.
2. Using this dose make up to final volume of 24mL with Sodium Chloride 0.9%w/v and run at 1mL/hour subcutaneously.
3. **Note: Do not give bolus doses** subcutaneously
4. Oral opioid can be given for breakthrough pain.(one sixth of the total daily dose)

Example.
Child 35kg receiving morphine infusions at 2mls/hr = 40microgram/kg/hour.=1.4mg/hour.
1.4 x 24hours = 33.6mg
Boli x 8 of 1mL (20microgram/kg) 20 x 35=700microgram x 8 = 5.6mg
5.6 + 33.6 = 39.2mg (round down to 39mg)
Subcutaneous infusion: morphine 39mg in final volume 24mls Sodium Chloride 0.9%w/v.
Rate of Infusion: 1mL/hour.
Oral breakthrough dose: (1/6th total daily dose PO) = morphine 6.5mg PO 1 to 4 hourly PRN
Switching from Oral to Subcutaneous Opioid

When switching from PO to Subcutaneous Opioid:

1. Calculate the total daily dose of oral opioid including any breakthrough doses.
2. Divide this dose x 2.
3. Using this dose make up to a final volume of 24mls with Sodium Chloride 0.9%w/v and run at 1mL/hour subcutaneously.
4. **Note: Do not give bolus doses** subcutaneously
5. Oral opioid can be given for breakthrough pain

Example:

Child 35kg receiving morphine sulphate 7mg PO 4 hourly = 42mg
Breakthrough dose x 3 of 7mgs = 21mg
42mg + 21mg = 63mg ÷ 2 = 30mg rounded down
Subcutaneous infusion: morphine 30mg in final volume 24mls Sodium Chloride 0.9%.
Rate of Infusion: 1mL/hour.
Oral breakthrough dose (1/6th total daily dose PO) = morphine 10mg PO 1 to 4 hourly PRN

Adjuvants

Clonidine[1]
Indications: Pain, opioid sparing
Oral/NG/PEG or IV
Child >1 month: initial dose 1 microgram/kg (max. 100micrograms) 3-4 times daily. Increase gradually as needed and tolerated to maximum of 5 microgram/kg (max. 100 micrograms) four times a day (See OLCHC formulary)

Clonidine is a mixed alpha-1 and alpha-2 agonist (mainly alpha-2). Appears to have synergistic analgesic effects with opioids and prevent opioid withdrawal symptoms.

Side effects: Licensed indication of clonidine is for the treatment of hypertension so reduction in BP is a likely side effect of use. Titrate the dose of clonidine against the symptoms and monitor BP and pulse on starting treatment and after each dose increase.
Other possible side effects include: constipation, nausea, dry mouth, vomiting, postural hypotension, dizziness, sleep disturbances, headache.

Clonidine when used as an adjunct to pain management should be weaned off before discharge from hospital.

Clonidine oral suspension is not routinely available from community pharmacies. If a child must be discharged on clonidine oral suspension please contact OLCHC pharmacy before discharging the patient.

Suggested Regime for weaning Clonidine

- Clonidine < 1 week: Stop, no need to wean.
- Clonidine >1 and < 2 weeks: Wean by 50% of dose for 24 hours and then stop.
- Clonidine > 2 weeks and < 1 month: Wean by 20% of original dose over 3-5 days.
- Clonidine > one month: Wean by 20% of original dose every 48 hours when off opioids. Wean frequency once
  dose is 0.5microgram/kg/dose.

Magnesium Citrate PO[1,6,7,8]
Magnesium has a role in pain management[1,6,7,8]. Magnesium citrate tablets contain 150mg magnesium (6.2mmol) per tablet.

Dose: 10mg/kg (max. 450mg) 8 – 12 hourly. (See OLCHC formulary[1])
Side Effects: includes loose stools, if this occurs reduce the dose.

Magnesium does not need to be weaned.
Children remain on magnesium until opioids have been stopped.
Magenesium Verla (Magnesium L-aspartate) is an alternative option for children unable to swallow tablets.
Magnesium verla sachets contain 121.5mg magnesium (5mmol).
Reduce dose if patient experiences diarrhoea.

Ketamine[1, 9-12]
Use:

- Adjuvant to opioid for Neuropathic pain.
- To reduce Methyl-D-Aspartate (NMDA) receptor wind-up pain, opioid tolerance and opioid induced hyperalgesia

Dose and route: Oral, NG, PEG
Child one month to 12 years: starting 0.5mg/kg to max 25mg 6 to 8 hourly.
Over 12 years and adult: 25mg 6 to 8 hourly prn.

Side Effects:

- Agitation, hallucinations, anxiety, dysphoria and sleep disturbance are recognised side effects. These are less
  common in children and when low doses for pain management are used.
- Ketamine can cause urinary tract symptoms- frequency, urgency, dysuria and haematuria. Consider discontinuing Ketamine if these symptoms occur.
- Caution in severe hepatic impairment, consider dose reduction.

Fluid Replacement

Use IV fluids in children with severe pain requiring parenteral analgesia, children with abdominal symptoms (e.g., pain, diarrhoea), and any children failing to maintain adequate oral intake.
Fluid of choice – Sodium Chloride 0.45%w/v and Glucose 5%w/v.
Requirement (Maintenance) – 80 - 100 mL/kg/24 hours.
Route – oral or intravenous.
Duration – stop IV fluids as soon as patient is pain free and taking adequate oral fluids.
Oxygen

Patients with simple painful crisis rarely require supplemental oxygen. Falling oxygen saturations should prompt urgent investigation with repeat FBC and CXR. Falling oxygen saturations may represent falling Hb levels or the development of a chest syndrome.
Oxygen saturations measured during a crisis should be compared with baseline values as some children have chronically low oxygen saturations.

Antibiotics

If apyrexial, continue penicillin prophylaxis (E.g. phenoxymethypencillin, see Formulary for dosing)
If pyrexial >38^{0}C, after blood and urine cultures and other appropriate microbiological investigations have been taken, start IV **Ceftriaxone** (see Formulary for dosing[1]).
If allergic to cephalosporin, start **Erythromycin** (see Formulary for dosing[1])
If there are signs or symptoms suggestive of a chest infection, start IV Ceftriaxone as above and add PO **Clarithromycin**. (see Formulary for dosing[1])
In the event of a severe deterioration, consider adding **Vancomycin (**see Formulary for dosing[1]). If osteomyelitis is suspected, refer to Microbiology for appropriate antibiotic cover for *staphylococcus aureus* and *salmonella.*
Note:

- In patients on Desferrioxamine (DFO) or Desferasirox, who have diarrhoea, the DFO should be discontinued and Ciprofloxacin started immediately (see Formulary for dosing1). Ciprofloxacin is discontinued once the diagnosis of Yersinia is excluded.
- **NEVER** aspirate an affected joint without prior discussion with the Haematologist.
- Stop oral intake if the abdomen is silent.

Special Considerations

Pain in the Shoulders or Hips

If the pain is repeated, or prolonged, X-ray the joints 4-8 weeks after the episode, to exclude aseptic necrosis.

Differential diagnosis

- Osteomyelitis
- Septic arthritis

The above conditions are distinguished by swinging pyrexia, severe systemic disorder, and positive blood cultures.
Note: swellings over long bones, muscles, and joint effusions are common in vaso-occlusion.
Special Note: Never aspirate joints as a first line investigation.

Abdominal pain

A common symptom in children may be due to mesenteric vaso-occlusion, constipation, splenic or hepatic sequestration; also consider urinary tract infection, appendicitis and other causes of an acute abdomen.

Management

Additional to analgesia and fluids:

- If there is vomiting, or the abdomen is distended, or bowel sounds are absent, give nothing by mouth and consider nasogastric suction.
- Measure abdominal girth (in centimetres at umbilicus) at regular intervals to monitor distension (1 - 4 hourly).
- Monitor liver and spleen size.
- If bowel sounds are absent, examine chest, do abdominal X-ray and arterial blood gases.
- If jaundice is worsening, perform abdominal ultrasound to evaluate the biliary system.
- Give Antibiotics: cefuroxime and metronidazole.
- If constipation is present: add Macrogol 3350 (prescribed as either Laxido® Paediatric or Movicol®) Senna or Bisacodyl.

Complex Pain

Complex Pain is a relatively new speciality in children's medicine. Complex pain may refer to patients who are significantly disabled by chronic or recurrent pain, or to pain in patients with medically complex conditions.
Chronic pain is defined as pain persisting for more than 3 months. It includes a variety of different types of pain whose aetiologies are still incompletely understood. It needs to be differentiated from a prolonged or series of sickle cell crises, which can be expected to resolve eventually without persistence of pain.

It can be due to:

- Central Sensitivity Syndromes, where there is an increased sensitivity to painful and non-painful stimuli and to other sensory inputs, such as loud noise. It appears to be precipitated and reinforced by repeated painful and stressful stimuli.
- Hyperalgesia. Exaggerated pain response to non-noxious stimuli. It can be associated with excessive opiate use.
- Neuropathic pain caused by peripheral or nerve root damage.
- Arthritis e.g. associated with AVN.

Symptoms

- Disabling severe pain, which is not localised and often described as "all over the body".
- It may vary in severity from day to day.
- During bad spells pain interferes with movement, social activities, school attendance, family life and sleeping.
- It is inadequately relieved by analgesics including opiates.
- Disease modifications are also not effective.

Diagnosis

A pattern of unremitting pain, without the characteristic episodic nature usually seen in SCD should raise the possibility.

Management

- Correct vitamin D deficiency.
- Out-rule other causes of pain.
- Complex and chronic pain require a multidisciplinary approach that includes medication, physical activates including physiotherapy and occupational therapy, and psychological therapies to help minimise the impact of pain
  on the individual.
- Neuropathic pain may require treatment with drugs such as gabapentin and amitriptyline.
- Management of sleeping problems may require education around sleep hygiene habits, medication such as melatonin or amitriptyline.

If Chest or Girdle syndromes develop, see below.

Chest pain, pain in the spine, sternum, ribs, and scapula may proceed to full blown sickle chest syndrome.

Opioid side effects and treatment

Constipation

If patients are on opioids, laxatives must be prescribed. Titrate dose to effect. Encourage increased fluid intake.

- Lactulose (osmotic laxative)
- Senna (Senokot® tabs, liquid). Consider prescribing Senna if there is no response to Lactulose.
- Movicol® and Laxido® Paediatric (iso-osmotic laxative)

Pruritis

- Chlorphenamine
- Naloxone: 1 – 4 microgram/kg (max. 200 micrograms) IV every two hours prn (unlicensed use)

Nausea/Vomiting

Ondansetron PO/IV
Cyclizine PO/ IV
For dosing information for medicines, please refer to Hospital Formulary and Prescribing Guide on Hospital Intranet

Weaning

- Wean opioids once pain is resolved.
- Wean opioid infusion by 0.5mL to 1mL per day as tolerated. This may be increased or decreased depending on patient's symptoms.
- Generally aim to wean off IV opioid over 24 to 48 hour period.
- Stop Ketamine when opioid requirements have been reduced to 50% of the maximum rate of infusion. Ketamine does not need to be weaned.
- Clonidine – wean last

Companion Documents:

Link to References