NORTH EAST AND YORKSHIRE REGIONAL TRANSFUSION COMMITTEE ANNUAL EDUCATION SYMPOSIUM

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ACUTE PAIN IN SICKLE CELL DISEASE

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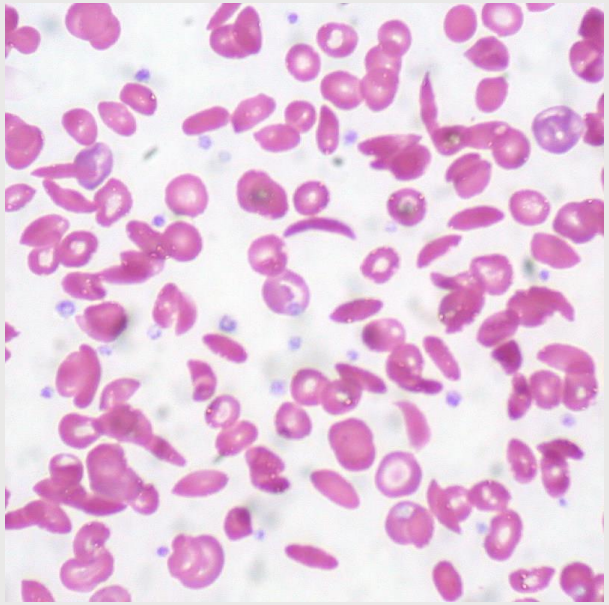
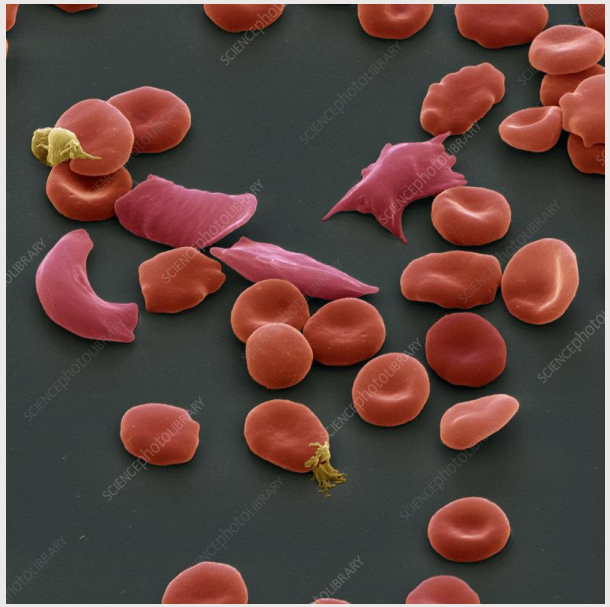
Vaso-Occlusive Crisis (VOC)



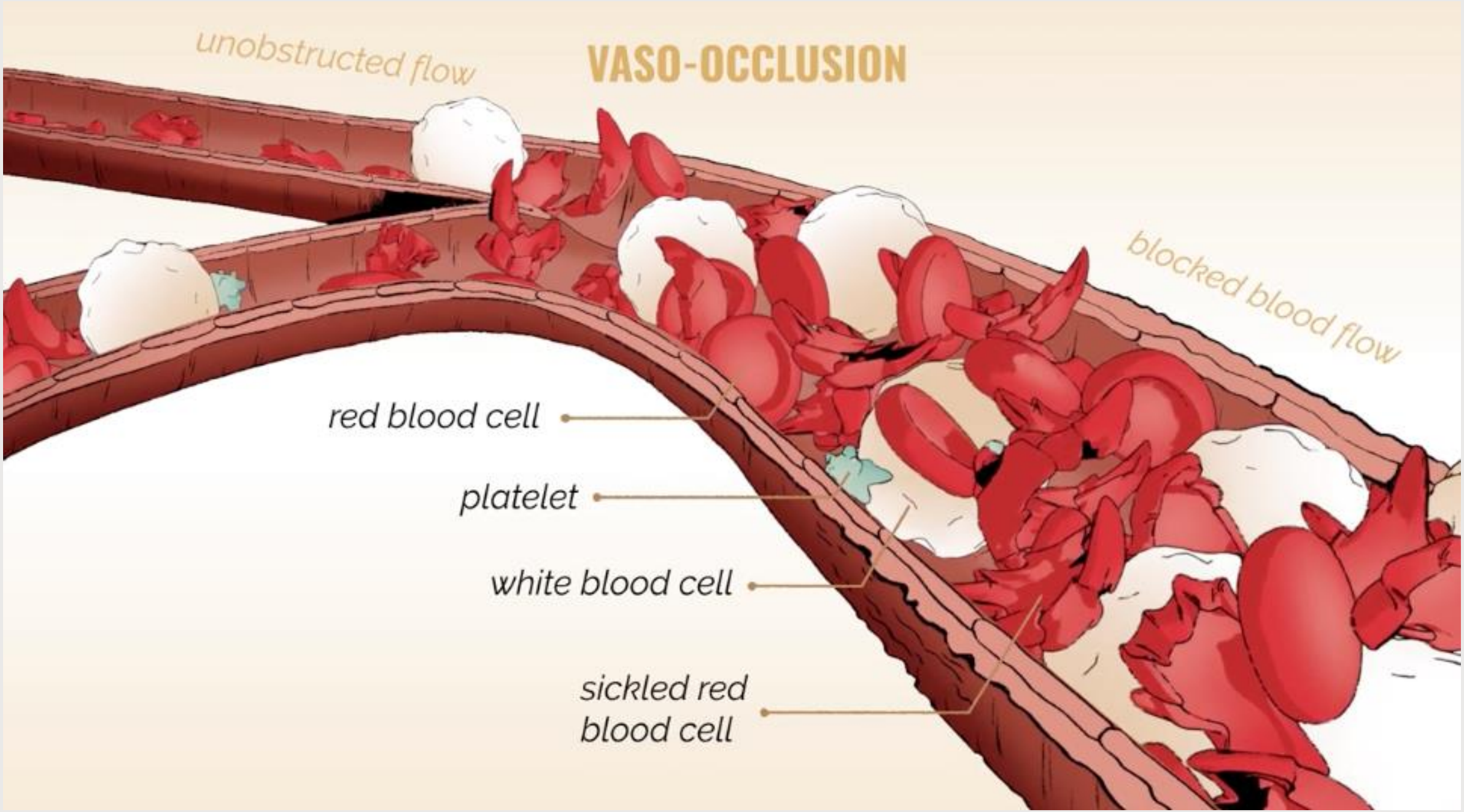


Overview

- Sickle cell disease is characterised by the presence of **distorted red blood cells** in the blood stream.
- The manifestations of the disease primarily arise from either **haemolysis**, involving the premature destruction of red blood cells, or **vaso-occlusion**, which is the most prevalent clinical feature (Al Hajeri *et al* 2016).
- **Most common cause of hospitalisation** (Ballas 2005)

Vaso-Occlusion Mechanism

- Sickled red cells adhere to the vascular endothelium, which results in the blockage of small blood vessels, leading to reduced blood flow, tissue ischaemia, and inflammation (Lutz *et* al 2015). Endothelial dysfunction, immune activation and neurogenic sensitisation further amplify the pain through complex interactions between red cells, white cells, platelets and inflammatory mediators.
- Vaso-occlusion can lead to acute, painful crisis. Known as a sickle cell crisis, VOC or vaso-occlusive episode.
- Pain is often reported in the joints, extremities, back or chest, but it can occur anywhere and can last several days or weeks.


unobstructed flow
VASO-OCCLUSION
blocked blood flow
red blood cell
platelet
white blood cell
sickled red blood cell

Triggers for VOC

Environmental Factors

Temperature changes, and high altitudes can trigger acute pain episodes. Cold weather may lead to vasoconstriction, worsening sickle cell complications.

Physical Stress

Excessive physical exertion or trauma (surgery) can precipitate pain crises. Activities that deprive the body of oxygen can lead to vaso-occlusive events.

Dehydration and Infection

Dehydration reduces blood volume and increases sickling. Infections can trigger inflammatory responses, intensifying existing pain and leading to more frequent crises.

Additionally:

Cronin *et al* 2019 increased rates of hospital admission associated with social, environmental and psychological factors such as depression, financial insecurity and spirituality.

No precipitating factors

NICE 2012b “Medical emergency”

The key points from guidance

- **Rapid pain relief**: people with an acute painful sickle cell crisis should receive prompt assessment and pain relief. NICE recommends administering analgesia within **30 minutes** of presentation.
- **Individual pain management plans**: pain management should be guided by the individualised care plan.
- **Regular pain assessments**: the effectiveness of pain relief should be reviewed regularly (every 30 minutes until pain is controlled)

Additional points

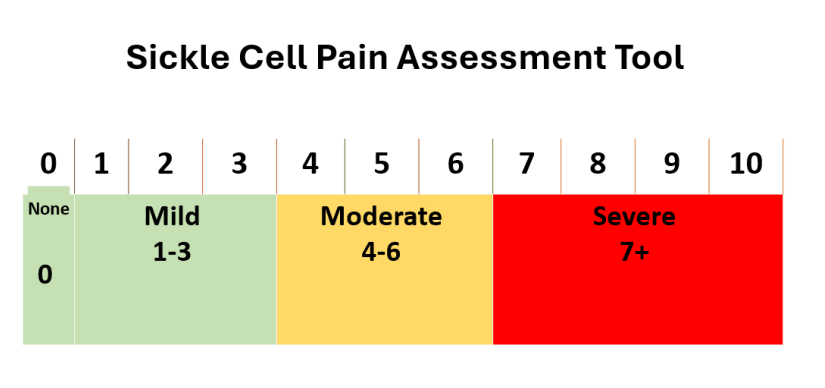
- **Mild to moderate pain**: NICE suggest the use of non-opioid analgesics – paracetamol or NSAIDs (unless contraindicated)
- **Severe pain**: Opioids should be primary treatment, often via intravenous or subcutaneous routes.
- **Hydration**: Dehydration can worsen sickling. **ORAL or Intravenous**.
- **Oxygen therapy**: Should be provided if there is evidence of hypoxia.

How to assess a patient with suspected crisis

History taking:

- Where is the pain?
- What brought it on?
- Does it feel like sickle pain?
- How severe is the pain out of ten?
- What pain relief works best?
- What have you taken prior to hospital?
- Have you had a fever or cough recently?

- The patient is the expert in their sickle cell.
- Balance empathy, speed and thoroughness.
- Don’t let history taking delay analgesia administration.
- Investigations and treatment specific interventions may be found on the individualised care plan (ICP).
- Common differentials to consider: Arthritis, osteomyelitis, myocardial infarction, pulmonary embolism, cholecystitis, pyelonephritis, appendicitis, peptic ulceration.
- Consider acute chest syndrome in any patient presenting with respiratory symptoms (fever, hypoxia, chest x-ray changes)



ICP

Sickle Cell Disease Patient Individualised Care Plan

Patient details:

Please alert haematology registrar or consultant on call immediately if this patient presents to any healthcare setting unless for planned outpatient review. This is a medical emergency.

This patient has a diagnosis of sickle cell disease and is under the care of Sheffield Teaching Hospitals Specialist Haemoglobinopathy Team. The patient must be considered an expert in their condition and must be believed when reporting severity of pain.

Sickle cell disease is a severe, inherited blood disorder which results in:

- Acute, severe pain which require strong analgesia to be given urgently
- Acute chest crisis, which can be rapidly life-threatening
- Stroke (ischaemic or haemorrhagic)
- Overwhelming bacterial sepsis
- Severe and complicated disease with COVID-19
- Acute and chronic kidney, liver and eye disease
- Risk of acute anaemia

If presenting with moderate or severe pain, give:

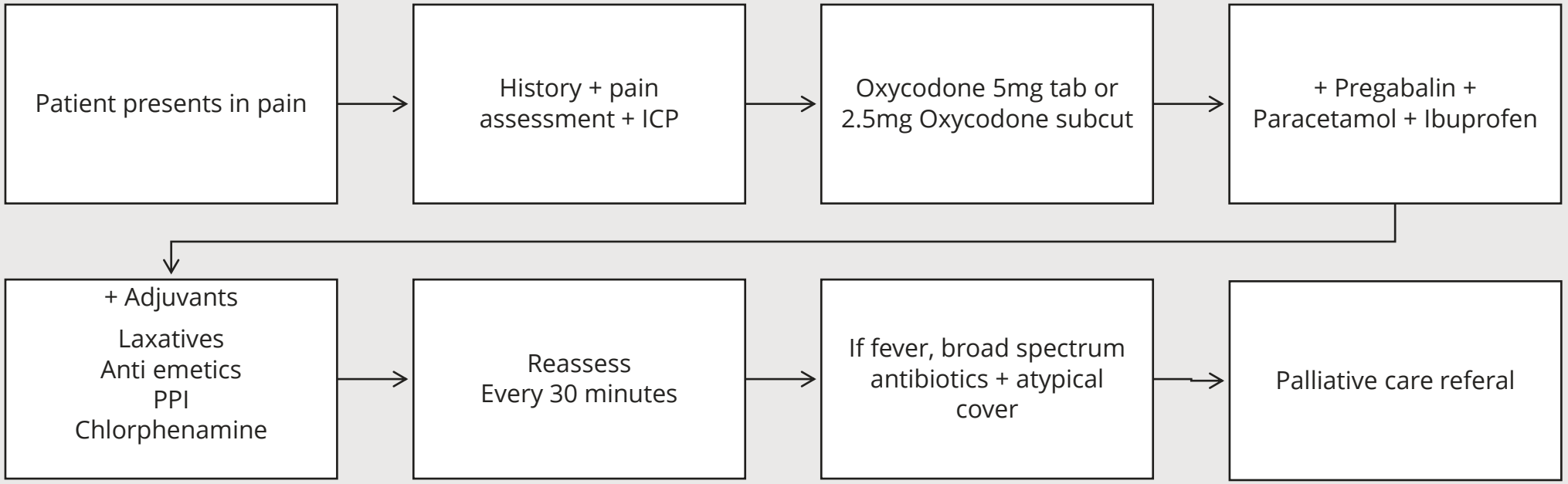
1. 1st analgesia doses **immediately**:
2. Oxycodone 5mg oral OR 2.5mg subcutaneous - *use subcutaneous dosing if pain is severe or patient vomiting*

   + Paracetamol 1g (oral or intravenous)
   + Ibuprofen 400mg (oral)
3. Pain must be reassessed at least every 30 minutes and further doses of oxycodone given until pain controlled (dose and route as above). Once pain score has reduced to VAS<4/10, maintain PRN oxycodone dose 1-2h maximum 8 doses in 24h (exceeding this should trigger a review).
4. Oxygen to maintain saturations ≥95%
5. IV fluids if severe pain or lack of certainty that oral hydration will be adequate
6. A full set of observations including pain score must be taken on admission and at least every 1 hour for a minimum of 6 hours and until pain controlled

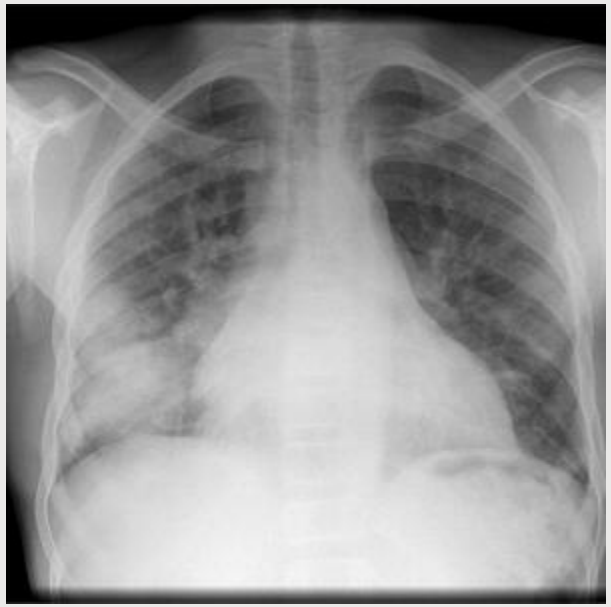
Contact haematology registrar or consultant on call via switchboard: 0114 243 4343

- Misconceptions and stigma regarding SCD related pain among HCP leads to inconsistent and untreated pain.
- Ineffective pain management increases the likelihood of readmission within days to weeks of discharge (Aljuburi *et al* 2013).
- Lack of consistency in care leads to loss of patient confidence in pain treatments and worsened pain control.
- This further compounds the financial burden of inpatient stays related to SCD and negatively affects the care given to SCD patients who already suffer stigma.
- A single centre US study (Welch-Coltrane *et al* 2021) demonstrated that ICPs can significantly reduce hospital admission rates, LOS and financial burden without compromising pain management for high-utilising patients with SCD.

Management


Patient presents in pain
History + pain assessment + ICP
Oxycodone 5mg tab or 2.5mg Oxycodone subcut
+ Pregabalin + Paracetamol + Ibuprofen
+ Adjuvants
Laxatives
Anti emetics
PPI
Chlorphenamine
Reassess Every 30 minutes
If fever, broad spectrum antibiotics + atypical cover
Palliative care referal

Acute Chest Syndrome



Definition

- Acute chest syndrome (ACS) is defined as an acute illness characterized by fever and/or respiratory symptoms, accompanied by a new pulmonary infiltrate on chest X-ray (Howard *et al* 2018)

Signs	Children	Adults
Fever	+++	++
Cough	++	++
Chest pain	+	++
Dyspnoea	+	++
Tachypnoea	+	+
Wheezing	+	+/−
Intercostal recession/nasal flaring	+	+/−
Skeletal pain	+	++
Hypoxia	++	+++
Haemoptysis	+/−	+

•+++ frequent (>80%), ++ common (50–79), +less common (10–49%), +/− infrequent (<10%).

- **Important:** ACS will often develop 24–72 h after the onset of severe pain (Gladwin & Vichinsky, 2008)
- Adequate analgesia, particularly in patients with rib, thoracic or abdominal pain is important to prevent splinting of the diaphragm and hypoventilation, with the consequent cycle of atelectasis, hypoxia and sickling that can occur.
- Broad spectrum antibiotics with atypical bacterial cover should be started such as: amoxicillin 1g IV three times per day & clarithromycin 500 mg PO twice daily.
- Exchange **or** transfusion

Thank you for listening

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