SESLHD PROCEDURE COVER SHEET

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KEY TERMS	Sickle cell, crisis, management
SUMMARY	This procedure has been developed to assist clinicians in the management of patients in Sickle Cell Crisis.

Sickle Cell Crisis – Management of Patients Presenting in Sickle Cell Crisis

SESLHDPR/609

1. POLICY STATEMENT

The purpose of this procedure is to provide clinical guidance and a framework to ensure the safe management of patients who present with sickle cell crisis.

2. BACKGROUND

Sickle cell disease is defined as genetic conditions in which at least one β globin gene allele carries a mutation (β 6 V mutation- Hb S). Sickle cell disease can arise from either homozygosity for the mutation (sickle cell anaemia) or coexistence of the Hb S mutation with other β gene variants or β thalassaemia (e.g. Hb SC, HbS/b0 thalassaemia).

Sickle cell trait (HbAS) and HbS/hereditary persistence of fetal haemoglobin only very rarely cause symptoms and thus should not be considered the cause of pain.

The abnormal haemoglobin results in abnormal rheology of sickle red blood cells (RBCs) with cellular dehydration, abnormal RBC deformability and mechanical fragility of cells. Acute painful sickle cell episodes (also known as painful crises) are caused by blockage of small blood vessels due to the abnormal rheology of sickle RBCs.

Circumstances that may provoke sickle cell crises include:

- Acute infection, febrile illness
- Hypoxia, such as asthma
- Dehydration
- Surgery
- Exposure to cold or high altitude (mountains or unpressurised planes)
- Glucocorticoid administration, emotional or stressful events
- Other: prolonged involuntary arterial compression, systemic hypertension, exposure to adrenergic agents.

3. RESPONSIBILITIES

3.1 Employees will:

All SESLHD staff providing treatment to patients in sickle cell crisis will act in accordance with this procedure:

- Medical Staff
- Nursing staff
- Allied Health staff.

3.2 Line Managers will:

Ensure this procedure is followed by all relevant staff.

3.3 District Managers / Service Managers will:

Provide support to staff in the implementation of this procedure as required.

3.4 Medical staff will:

- Assess patient, institute management and document findings, management plan, fluid and medications
- Liaise with nursing staff in the management of the patient.

4. PROCEDURE

MANAGEMENT OF SICKLE CELL CRISIS

Patients presenting to Sutherland and Sydney/Sydney Eye Hospital – contact Haematologist on-call via St George or Prince of Wales Hospital Switchboard. The patient will require transfer to either St George Hospital or Prince of Wales Hospital for further management.

Most mild pain can be treated with simple analgesics at home – e.g. paracetamol alone or in combination with oxycodone with orally administered NSAIDs.

4.1 SEVERE CRISIS MANAGEMENT

Rapid Clinical Assessment:

- Full blood count, EUC, CMP, LFT, Coags and G&H. HbEPG. Blood gases
- Temperature, pulse, blood pressure, pulse oximetry
- Acute chest symptoms, cardiovascular parameters
- Neurological assessment
- Abdominal evaluation
- Urinary assessment (e.g. Priapism, UTI).
- Consider Chest Xray

5.2 ANALGESIA

The following guidelines relate to opioid-naïve adults to be admitted as an inpatient for acute management of their sickle cell disease.
Reduced opioid doses are required for elderly, low body mass and patients with renal impairment.
Consider patient specific factors e.g. co-morbidities prior to prescribing.
Seek specialist advice for patients who are opioid tolerant.

Pain management for a patient presenting with a vaso-occlusive pain crisis not responding to simple analgesia:

- Assess pain – Patient self-report of pain is the most appropriate method.
- Administer loading dose of Morphine 2.5-5mg IV or Oxycodone 2.5-5mg IV
- Titrate subsequent doses every 20 minutes to a maximum of 15mg Morphine / Oxycodone in the first hour following presentation.
- Commence Patient Controlled Analgesia (PCA), under guidance of Acute Pain Service
    - Morphine or
    - Oxycodone

- Commence simple analgesia (Paracetamol)
- Judicious use of PO NSAIDs – titrated to benefit, renal function and adverse risk profile. Avoid NSAIDS in patients with renal impairment
- Maintain continuous pulse oximetry
- If pain remains severe and is refractory to opioid analgesia, continue consultation with Acute Pain Service, and consider commencement of adjunctive Ketamine infusion 4mg/h. Titrate dose to maximum of 16mg/h.
- Red cell exchange ASAP if recommended by Haematologist
- As crisis improves PCA may be ceased and replaced with oral analgesia – eg. Endone PRN.
- Avoid Slow release opioids in acute vaso-occlusive crises
- Prescribe laxatives routinely e.g. Macrogol oral powder one sachet twice daily +/- coloxyl with senna two to four tablets daily
- Prescribe antiemetics if required
- Consider anxiolytic: haloperidol 1 to 3mg oral/IM every 2 hours as required, to a maximum of 15mg in 24 hours (reduce dose in elderly)

- Undertake clinical observations as per:
  SGSHHS Clinical Business Rule: Pain Management - Patient Controlled Analgesia (PCA) in Adults
  POWH CLIN064 Patient controlled analgesia PCA

- **Activate a Clinical Review, Rapid Response or Code Blue Code call as per the facility's Clinical Emergency Response System Clinical Business Rule.**

5.3 FLUID REPLACEMENT THERAPY

- Strict fluid balance in all patients
- Give approximately three (3) Litres over 24 hours, adjust according to size and cardiac function
- Give first 1000mL at a faster rate, then reduce rate as clinically appropriate.

5.4 OXYGEN

- Administer oxygen to maintain oxygen saturation >95% since desaturation will potentially lead to life threatening pulmonary sickling.

5.5 ANTIBIOTICS

- If febrile, and generally unwell, has chest symptoms or signs of infection suspected, commence broad spectrum antibiotics with pneumococcal cover within one hour of patient presentation to Emergency Department
- Antibiotic choice: ceftriaxone 1g IV daily or BD depending on index of suspicion of severe pneumococcal infection + macrolide (if chest signs) - in adults
- For patients with immediate hypersensitivity to penicillins or cephalosporins use vancomycin plus ciprofloxacin (400mg IV BD or 750mg oral BD). A macrolide is not required with this combination
- If on desferrioxamine iron chelation therapy and the patient has abdominal pain or

diarrhoea, stop chelation and send blood and stool cultures. Commence ciprofloxacin 500mg BD oral or 400mg IV BD to cover possible Yersinia infections
- If on deferiprone, check FBC to exclude neutropenia (deferiprone can cause agranulocytosis).
- Note that antibiotic doses above may need to be reduced in renal or hepatic impairment

5.6 VENOUS ACCESS

- Patient may have an AV fistula insitu. Accredited registered nurses will be required to access for Red Blood Cell exchange
- Peripheral cannulation may be difficult. Cannulation may need to be under ultrasound guidance
- IJ or femoral vascath may need to be inserted for Red Blood Cell exchange. Please consult ICU or interventional radiology early for vascath insertion.

5.7 RED CELL EXCHANGE

- Red Blood Cell exchange is considered if the patient experiences:
  1. Any early signs of organ failure
  2. Any neurological defect - confusion, motor defects, epilepsy
  3. Worsening respiratory failure or new pulmonary infiltrates or desaturation
  4. Intractable pain or opioid intolerance
  5. Haemodynamic instability
  6. Acute worsening of anaemia or cardiovascular insufficiency
  7. Acute enlargement of the spleen or liver
  8. Priapism.

- Liaise with on-call Haematologist or Haematology Registrar to organise procedure
- Crossmatch eight (8) units of packed red blood cells
- Red Blood Cell exchange is performed via Apheresis Service at both Prince of Wales Hospital and St George Hospital
- Current consent for Red Blood Cell Exchange is required
- Current consent for Blood Transfusion is required.
- Patient will require appropriate vascular access (as above)

ST GEORGE HOSPITAL

- During business hours Monday to Friday (8:00am – 4:30pm) please contact the Apheresis CNC (pager 1162)
- Out of hours please contact the Haematologist on-call who will contact after hours nurse manager who will contact the Apheresis Operator on-call
- Refer to SGSHHS WPI: Red Blood Cell Exchange (RBCX)

PRINCE OF WALES HOSPITAL

- During business hours Monday to Friday (8:00am – 4:30pm) please contact the Haematology CNC (pager 45465), Transfusion CNC (pager 45155) and HODC NUM (pager 45102)

- Out of hours service, arrangements will be made through the on-call Haematology Registrar
- Refer to POWH Clinical Business Rule CLIN036: Guidelines for the care of patients undergoing a Red Blood Cell Exchange or Therapeutic Plasma Exchange

5.8 BLOOD TRANSFUSIONS

- Red Blood Cell exchange is preferred to simple transfusion
- Hb may fall 10-20g/L in an uncomplicated painful crisis
- Routine blood transfusions are not required unless patient develops signs or symptoms which may be due to anaemia (e.g. tachycardia, dyspnoea, fatigue)
- Typically, blood transfusion will not be necessary unless fall >20g/L and is below 70g/L
- Avoid raising the haematocrit to >30% (Hb >100g/L) which will potentially increase whole blood viscosity
- Discuss with Blood Bank if the patient has any special transfusion requirements (e.g. previously required triple washed blood, Rh phenotype matching etc.).

6. DEFINITIONS

EUC: Electrolytes, Urea, Creatinine. This test is a measure of kidney function.

LFT: This test measures several liver enzymes. Alanine aminotransferase (ALT), Alkaline phosphatase (ALP) and Aspartate aminotransferase (AST)

COAGULATION studies: Assess the coagulation profile.

G&H: A group and hold screen is a group of tests that determines blood group and assesses for red cell antibodies.

HbEPG: This refers to a group of blood tests that can detect different types of hemoglobin. This test is performed on the pre-transfusion sample but the result will not be available for several working days.

BLOOD GASES: A blood gas test measures the amount of oxygen and carbon dioxide in the blood. It may also be used to determine the pH of the blood, or how acidic it is. The test is commonly known as a blood gas analysis or arterial blood gas (ABG) test.

PCA: Patient controlled analgesia

DOCUMENTATION

Nil

7. AUDIT

As per clinical requirements.

8. REFERENCES

- SESLHDPR/371 - Ketamine Infusions for Adult Patients with Acute and Chronic Non-Malignant Pain
- SGSHHS WPI: Red Blood Cell Exchange (RBCX)
- SGSHHS Clinical Business Rule: Pain Management - Intravenous Opioid Infusion - St George Hospital
- SGSHHS Clinical Business Rule: Pain Management - Patient Controlled Analgesia (PCA) in Adults
- POWH Clinical Business Rule CLIN036: Guidelines for the care of patients undergoing a Red Blood Cell Exchange or Therapeutic Plasma Exchange
- Clinical Excellence Commission: Adult Antibiotic Guideline - Severe Sepsis and Septic Shock
- Brandow, AM,et al. (2020), American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain, Blood Advances, 4(12), 2656-2701.
- DeBaun, MR & Vichinsky, EP (2020), Acute vaso-occlusive pain management in sickle cell disease. In Mahoney Jr, DH & Tirnauer, JS (Eds.), UpToDate, Retrieved August 12, 2020, from https://www.uptodate.com.acs.hcn.com.au/contents/acute-vaso-occlusive-pain-management-in-sickle-cell-disease?search=sicklcrisis&source=search_result&selectedTitle=3~150&usage_type=default&display_rank=3
- Schug, SA, Palmer, GM, Scott, DA, Halliwell, R, Trinca, J;APM:SE Working group of the Australian and New Zealand College of Anaesthetists and Faculty of Pain Medicine (2015), Acute Pain Management: Scientific Evidence (4th edition), ANZCA & FPM, Melbourne

9. REVISION AND APPROVAL HISTORY

Date	Revision No.	Author and Approval
February 2018	DRAFT	Dr Shir-Jing Ho Department Head Haematology SGH Cassandra Hobbs CNC Apheresis SGH Dr Giselle Kidson-Gerber Haematologist POWH
April 2018	DRAFT	Draft for Comment until 10 May 2018
May 2018	DRAFT	Processed by Executive Services prior to submission to SESLHD Drugs and Quality Use of Medicine Committee
August 2018	1	Endorsed by SESLHD Quality Use of Medicines Committee and SESLHD Clinical and Quality Council.
February 2020	1	Revised Section 5.2 to remove reference to PACE and updated to reflect CERS. Minor review approved by Executive Sponsor.
September	2	Updating of section 5.2 ANALGESIA.

2020		Removal of flow chart as does not reflect what the document recommends. Analgesia section updated. Approved by Executive Sponsor. Formatted by Executive Services prior to tabling at September Quality Use of Medicines Committee.
October 2020	2	Quality Use of Medicines Committee advised the procedure was *Deferred – and requested that consideration be given to removing Hydromorphone from section 5.2.*
October 2020	3	Hydromorphone removed from procedure. Approved by Executive Sponsor. Processed by Executive Services prior to tabling at November Quality Use of Medicines Committee.
November 2020	3	Approved by Quality Use of Medicines Committee. Published by Executive Services.