SESLHD PROCEDURE COVER SHEET

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EXECUTIVE SPONSOR or EXECUTIVE CLINICAL SPONSOR	SESLHD Clinical Stream Director, Cancer Services
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FUNCTIONAL GROUP(S)	Cancer and Palliative Care Services Critical Care and Emergency Medicine
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
- Recent COVID-19 infection (1 to 3 weeks post infection)
- 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 orally administered opioids (e.g., oxycodone) and/or non- steroidal anti-inflammatory drugs (NSAIDs). NSAIDs can be a useful adjunct to opioids in Sickle Cell pain crises. Patients who present to ED have generally failed home management.

4.1 SEVERE CRISIS MANAGEMENT

Rapid Clinical Assessment:

- Patient should be triaged as a Category 3 to ensure assessment and analgesia given within 30 minutes of triage
- 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, blood cultures (2 sets collected from different sites), MSU

Ongoing Clinical Observations to be undertaken in accordance with the relevant facility Clinical Business Rule. If observations are NOT between the flags **activate a Clinical Review, Rapid Response or Code Blue Code call as per the facility's Clinical Emergency Response System Clinical Business Rule.**

4.2 ANALGESIA

For pain management for a patient presenting to the Emergency Department with a vaso-occlusive pain crisis **not** responding to simple analgesia: commence IV opioids in accordance with usual management of moderate to severe pain.
Obtain an **Acute Pain Service** review.
Patients should receive analgesia within 60 minutes of presentation and 30 minutes triage.

4.3 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 1000 mL at a faster rate, then reduce rate as clinically appropriate.

4.4 OXYGEN

- Administer oxygen to maintain oxygen saturation >95% since desaturation will potentially lead to life threatening pulmonary sickling.
- Administration of oxygen can be nurse initiated provided a medical review occurs post initiation
- Oxygen therapy requires a documented plan from medical officers on target saturations, weaning instruction and eMR documentation in altered calling criteria reflecting this
- Oxygen is a medication and needs to be prescribed for patients receiving PCA.

4.5 ANTIBIOTICS

- If febrile, and generally unwell, has chest symptoms or signs of infection, collect 2 sets of blood cultures from different sites and MSU then commence antibiotics in accordance with eTG Antibiotic Guidelines
- Commence IV antibiotics within 1 hour of presentation
- If on desferrioxamine iron chelation therapy and the patient has abdominal pain or diarrhoea, stop chelation and send blood and stool cultures and request Yersinia cultures
- If on deferiprone, check FBC to exclude neutropenia (deferiprone can cause agranulocytosis).

4.6 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.

4.7 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
- Valid Request/Consent For Medical Procedure Treatment
- Valid Blood and Blood Products Administration and Consent Form
- 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 125 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) or HODC NUM (pager 46689)
- Out of hours service, arrangements will be made through the on-call Haematology Registrar

4.8 BLOOD TRANSFUSIONS

- Red Blood Cell exchange is preferred to simple transfusion
- Hb may fall 10-20 g/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 >20 g/L and is below 70 g/L
- Avoid raising the haematocrit to >30% (Hb >100 g/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.).

5. DEFINITIONS

CMP: Calcium, Magnesium, Potassium

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

NSAID: Non-Steroidal Anti Inflammatory

DOCUMENTATION

Request/Consent for Medical Procedure Treatment
Blood and Blood Products Administration and Consent Form
SEALS Blood Bank Issue Report
NSW Health Patient Controlled Analgesia (PCA) Adult form (SMR130.025)
Between the Flags (BTF) Observation Chart in eMR/eRIC/MOSAIQ/ARIA
Patient's Health Care Record
Daily Fluid Balance Form

6. AUDIT

As per clinical requirements.

7. REFERENCES

- SESLHDPR/371 – Pain Management - Ketamine Infusions for Adult Patients with Acute and Chronic Non Malignant Pain
- SGSHHS WPI 125 Red Blood Cell Exchange (RBCX)
- SGH CLIN126 Intravenous Opioid Infusions - Prescribing and Administration of Intravenous Opioid Infusions in General Wards and Critical Care Units for Adult Patients only at SGH
- SGH Interim CLIN725 Pain Management - Patient Controlled Analgesia (PCA) in Adults at St. George Hospital
- TSH CLIN127 Pain Management - Patient Controlled Analgesia (PCA) in Adult Patients at The Sutherland Hospital
- POWH Clinical Business Rule CLIN064 Patient Controlled Analgesia PCA
- 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, Scott DA, Mott JF, Halliwell R, Palmer GM, Alcock M; APM:SE Working Group of the Australian and New Zealand College of Anaesthetists and Faculty of Pain Medicine. 2020, Acute Pain Management: Scientific Evidence (Fifth Edition 2020) , ANZCA & FPM.

8. 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 2020	2	Updating of section 5.2 ANALGESIA. 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.
May 2023	4	Update of section 4 PROCEDURE: expansion of use of NSAIDs Update of section 4.1 SEVERE CRISIS MANAGEMENT: to include Triage Category; escalation process as per local CERS CBR Update of Section 4.2 ANALGESIA: removal of specific analgesic agents and doses and add statement on referral to Pain Management Services

		Expansion section 4.4 OXYGEN: to include nurse initiation, target saturations; weaning and altered calling criteria Update section 4.5 ANTIBIOTICS: removal of specific antibiotic agents and doses. Replace with link to eTG Antibiotic Guidelines. Approved by Executive Sponsor.
June 2023	4	Approved by SESLHD Drug and Therapeutics Committee.