[HOSPITAL / HEALTH AUTHORITY NAME]

SICKLE CELL EMERGENCIES PATHWAY

Protocol 45: Vaso-Occlusive Pain, Acute Chest Syndrome, Stroke, Sepsis, Severe Anaemia, Sequestration, Priapism, Transfusion Complications, and Safe Disposition

DRAFT FOR EMERGENCY MEDICINE, INTERNAL MEDICINE, PAEDIATRICS, HAEMATOLOGY, NURSING, PHARMACY, TRANSFUSION MEDICINE, CRITICAL CARE, RESPIRATORY MEDICINE, NEUROLOGY / STROKE, UROLOGY, OBSTETRICS, RADIOLOGY, LABORATORY SERVICES, PAIN SERVICES, AND CLINICAL GOVERNANCE

STATUS: This is a draft clinical-governance document. It must be adapted to local sickle cell epidemiology and genotype patterns, paediatric and adult services, individual pain plans, controlled-drug policy, medication formularies, blood-bank capability, red-cell antigen matching, exchange transfusion access, critical-care and imaging capacity, specialist availability, transport, and referral pathways before implementation.

SICKLE CELL SAFETY RULE: Pain is a medical emergency and the patient is the expert in their usual episode and effective treatment. Give first analgesia rapidly - target within 30 minutes of triage and no later than 60 minutes after arrival - while simultaneously looking for acute chest syndrome, sepsis, stroke, severe anaemia, sequestration, priapism, pulmonary embolism, pregnancy complications, and other dangerous diagnoses.

Document control	Details
Document owner	Emergency Department / Medical Services Directorate / Paediatrics / Haematology / Nursing Services / Clinical Governance
Clinical leads	Emergency Medicine; Internal Medicine; Paediatrics; Haematology; Nursing; Pharmacy; Transfusion Medicine; Critical Care; Respiratory Medicine; Neurology / Stroke; Urology; Obstetrics; Radiology; Laboratory Services; Pain Service
Applies to	Children, adolescents, pregnant people and adults with known or suspected sickle cell disease presenting with acute pain or any possible sickle-related complication. It does not apply to sickle cell trait alone unless another acute diagnosis is present.
Interfaces	Protocol 1 Patient Journey; Protocol 2 Triage; Protocol 3 Resuscitation / Sepsis / Shock; Protocol 4 Assessment and Documentation; Protocol 6 Pain Management; Protocol 14 Fever / Sepsis; Protocol 17 Altered Mental Status; Protocol 18 Stroke / TIA; Protocol 19 Seizures; Protocol 20 Acute Headache; Protocol 24 Abdominal Pain; Protocol 25 GI Bleeding; Protocol 26 Dehydration / Electrolytes; Protocol 28 AKI; Protocol 29 Poisoning; Protocol 30 Anaphylaxis; Protocol 38 Obstetric Emergencies; Protocol 40 Paediatric Assessment; Protocol 41 Neonatal Emergencies; Protocol 46 Immunocompromised / Oncology; Protocol 48 Airway / Ventilation; Protocol 49 Major Haemorrhage / Transfusion; Protocol 53 Observation Care.
Version / status	Draft 1.0 for local multidisciplinary validation
Review cycle	After any death or serious harm involving delayed analgesia, missed acute chest syndrome / sepsis / stroke, transfusion reaction, hyperhaemolysis, deterioration during observation or transfer, or at least every 2 years.
Required approval	Emergency Department; Internal Medicine; Paediatrics; Haematology; Nursing; Pharmacy; Transfusion Medicine; Critical Care; Neurology / Stroke; Respiratory Medicine; Urology; Obstetrics; Radiology; Laboratory Services; Pain Service; Clinical Governance.

1. Purpose

To provide a standardized emergency-department pathway for rapid, equitable and safe assessment of people with sickle cell disease; immediate treatment of vaso-occlusive pain; early recognition and stabilization of life-threatening complications; safe transfusion decisions; specialist escalation; and reliable discharge, admission or transfer.

2. Scope

- Includes acute pain / vaso-occlusive episode, acute chest syndrome, fever and sepsis, stroke / TIA, seizures, severe anaemia, aplastic crisis, splenic or hepatic sequestration, delayed haemolytic transfusion reaction and hyperhaemolysis, priapism, renal / abdominal / musculoskeletal emergencies, pregnancy-related risk and acute complications in children.
- Use alongside standard emergency pathways. A person with sickle cell disease can have myocardial infarction, pulmonary embolism, appendicitis, ectopic pregnancy, meningitis, trauma, toxic exposure or any other emergency.
- Use the patient-specific emergency and analgesia plan whenever available, after confirming identity and clinical appropriateness. The absence of a plan must not delay treatment.
- This protocol does not replace specialist advice for exchange transfusion, complex alloimmunization, hyperhaemolysis, pregnancy, neonatal disease or critical illness.

3. Core policy statements

- Triage pain as an acute medical emergency. Do not delay analgesia for registration completion, blood tests, IV access, imaging, urine testing or proof of diagnosis.
- Use respectful, non-stigmatizing communication. The patient-reported pain score, usual pattern, effective medicines and previous adverse effects are valid clinical data.
- Assess for complications at every presentation and at every reassessment. A patient may develop acute chest syndrome or sepsis after initially presenting with uncomplicated pain.
- Use oxygen for hypoxaemia or a fall below the patient's baseline, not routinely for normal saturations. Avoid both dehydration and fluid overload.
- Do not transfuse for uncomplicated pain or stable chronic anaemia. Compare haemoglobin with the patient's baseline and involve haematology / transfusion medicine for acute indications.

- Any new focal neurological deficit, seizure, severe altered mental state or sudden visual loss requires immediate stroke / neurological activation and urgent transfusion planning.
- Any painful erection lasting or approaching 4 hours requires urgent urological intervention; do not delay aspiration / irrigation while arranging transfusion.
- Discharge only after clinical improvement, complication screening, a feasible home plan, medicines and follow-up are confirmed, with explicit return precautions.

4. Definitions and clinical framework

Term	Operational definition
Sickle cell disease (SCD)	A group of inherited haemoglobin disorders in which HbS causes haemolysis, vaso-occlusion and progressive organ injury. Genotype and baseline haemoglobin vary; obtain the patient's documented genotype where possible.
Vaso-occlusive episode (VOE)	Acute pain attributable to vaso-occlusion after assessment for other diagnoses and sickle complications. Pain severity cannot be inferred from appearance, vital signs or laboratory results.
Acute chest syndrome (ACS)	A new pulmonary infiltrate with fever and/or respiratory symptoms in a person with SCD. Early imaging can be normal; evolving hypoxia, chest pain, cough, tachypnoea or falling haemoglobin requires repeat evaluation.
Baseline haemoglobin	The person's usual steady-state haemoglobin, reticulocyte count and oxygen saturation. These are often more useful than population reference ranges.
Severe acute anaemia	A clinically important fall from baseline, especially with symptoms, haemodynamic compromise, low reticulocytes, sequestration, haemolysis or organ dysfunction.
Splenic / hepatic sequestration	Rapid trapping of blood in the spleen or liver causing enlargement, acute anaemia and potentially shock. Splenic sequestration is especially important in infants and children.
Delayed haemolytic transfusion reaction (DHTR)	New haemolysis usually occurring days to weeks after transfusion, sometimes with pain, fever, jaundice, dark urine and haemoglobin below the pre-transfusion level. Hyperhaemolysis may occur with a negative antibody screen or DAT.
Stuttering / ischaemic priapism	Recurrent painful erections under 4 hours may precede a prolonged episode. Ischaemic priapism lasting 4 hours or more is a time-critical urological emergency.

5. Roles and accountability

Role	Minimum responsibilities
Triage nurse	Identify SCD, pain severity, fever, chest / neurological / priapism symptoms, pregnancy and high-risk history; obtain complete observations; activate rapid analgesia and immediate escalation criteria.
ED nurse	Administer analgesia promptly, reassess pain and adverse effects, monitor oxygenation and sedation, support hydration and incentive spirometry, and escalate deterioration.
ED clinician	Perform parallel pain treatment and complication assessment, use the individual plan, order targeted investigations, recognize transfusion indications, obtain specialist input and determine disposition.
Senior ED clinician	Review severe / refractory pain, repeated dosing, diagnostic uncertainty, high-risk complications, opioid or sedation concerns, discharge after complex presentations, and all transfer decisions.
Haematology / paediatrics / internal medicine	Advise on severe complications, baseline values, transfusion mode and targets, alloantibodies, DHTR / hyperhaemolysis, disease-modifying therapy and admission / transfer.
Transfusion laboratory / blood bank	Retrieve historical antibodies, communicate compatibility limitations, provide appropriately selected HbS-negative and antigen-matched units according to policy, and support urgent exchange planning.
Critical care / respiratory team	Support deteriorating ACS, severe hypoxaemia, shock, multiorgan failure, non-invasive or invasive ventilation and high-dependency transfer.
Neurology / stroke team	Coordinate imaging, reperfusion assessment and neurological management while urgent SCD transfusion treatment proceeds.
Urology	Provide urgent corporal aspiration / irrigation and intracavernosal sympathomimetic treatment for ischaemic priapism.
Pharmacy / pain service	Support individualized opioid regimens, multimodal analgesia, adverse-effect prevention, controlled-drug safety and difficult pain management.

6. Required readiness

Capability	Minimum standard
Identification	Electronic / paper SCD flag, genotype and baseline data when known, transfusion and antibody history, individualized pain plan and specialist contacts.
Rapid analgesia	Triage-initiated or nurse-initiated protocol approved for oral, intranasal, subcutaneous or IV routes; age-appropriate pain scoring; monitoring and reversal capability.
Monitoring	Pulse oximetry, cardiac monitoring when indicated, capnography where required by sedation / opioid policy, serial pain and sedation scoring, paediatric observation capability.
Respiratory support	Oxygen, incentive spirometers, nebulization, high-flow / non-invasive support where available, and rapid critical-care escalation.
Laboratory	24-hour FBC with reticulocytes, chemistry, bilirubin / LDH, blood cultures, group and screen / crossmatch, antibody history retrieval and urgent blood-gas testing.

Capability	Minimum standard
Transfusion	Approved simple and exchange transfusion pathways, compatible HbS-negative red-cell access, antigen matching policy, emergency release process, and regional transfer agreement when exchange is unavailable.
Specialists	Haematology / paediatrics / internal medicine, critical care, stroke / neurology, urology, obstetrics, radiology and transfusion medicine contacts available at all times.
Patient support	Warm environment, hydration, interpreter and disability access, privacy, family / caregiver involvement, and written discharge information.

7. Triage and immediate danger recognition

Finding	Immediate response
Airway compromise, severe respiratory distress, SpO2 below baseline or <95% on room air, shock, collapse, severe drowsiness or rapidly worsening pain	Move to resuscitation; ABCDE; oxygen for hypoxaemia; senior clinician; critical-care / haematology escalation.
Chest pain, cough, fever, dyspnoea, tachypnoea, wheeze, new oxygen requirement or falling saturation	Treat as possible ACS / pneumonia / pulmonary embolism. Continuous oximetry, chest imaging, FBC / reticulocytes, cultures as indicated and early antibiotics / specialist review.
Fever, rigors, toxic appearance, hypotension, altered mental state, meningism or indwelling line	High-risk sepsis pathway. Cultures if this does not delay therapy; empiric IV antibiotics within 60 minutes or sooner if shock.
New focal deficit, speech / visual disturbance, severe headache, seizure, altered consciousness or acute ataxia	Activate stroke / neurological pathway; document last-known-well; urgent imaging and haematology / transfusion planning.
Pallor, tachycardia, syncope, jaundice, dark urine, rapidly enlarging spleen / liver or recent transfusion	Urgent FBC / reticulocytes, haemolysis tests, group and screen, senior review; consider sequestration, aplasia, haemolysis or DHTR.
Painful erection persisting or approaching 4 hours	Immediate analgesia and urgent urology; prepare aspiration / irrigation. Do not wait for spontaneous resolution or transfusion.
Pregnancy with pain, bleeding, contractions, reduced fetal movement, hypertension, dyspnoea or fever	Simultaneous obstetric and sickle emergency assessment; early maternal-fetal and haematology review.
Child under 5 with fever, lethargy, poor feeding, pallor, abdominal enlargement or parental concern	High-priority paediatric assessment; palpate spleen gently, check glucose and obtain baseline history.

DO NOT USE THE LABEL "SICKLE CRISIS" AS THE DIAGNOSIS. Document the actual syndrome and risks: acute pain / VOE, possible ACS, fever / sepsis, stroke, anaemia / sequestration, priapism, abdominal emergency, pregnancy complication or another diagnosis.

8. The first 30 minutes

1. Complete ABCDE, full vital signs, oxygen saturation, temperature, pain score, sedation score, weight and bedside glucose. Compare saturation with the patient's usual baseline if known.
2. Ask immediately about chest symptoms, fever, focal neurology, headache, seizure, pallor, jaundice, dark urine, abdominal enlargement, priapism, pregnancy, recent transfusion and previous critical complications.
3. Retrieve the individualized emergency / analgesia plan, baseline haemoglobin and saturation, genotype, regular medicines, opioid tolerance, allergies, renal / hepatic disease and transfusion antibody history.
4. Administer first analgesia by the fastest safe route. Do not wait for IV access. Use the individualized dose when available; otherwise use a locally approved weight-based or opioid-exposure-based protocol.
5. Apply warmth, position comfortably and offer oral fluids if safe. Obtain IV or intraosseous access when clinically indicated, but avoid repeated traumatic cannulation attempts solely to give the first dose.
6. Reassess pain, respiratory rate, oxygenation, sedation, nausea and pruritus after each parenteral opioid dose, usually every 15-30 minutes during titration; repeat or escalate according to the approved plan.
7. Order targeted investigations based on presentation. Uncomplicated pain in a well patient does not require indiscriminate testing, but a lower threshold applies to fever, chest symptoms, severe pain, anaemia, pregnancy, renal disease or atypical features.
8. Contact senior ED and haematology / paediatrics / internal medicine early for severe complications, recent transfusion, difficult pain control, high-risk pregnancy, repeated presentation or likely admission / transfer.
9. Document times of arrival, triage, pain assessment, first analgesia, each reassessment, clinician review, antibiotics and escalation. Delay requires contemporaneous explanation and mitigation.

9. Focused history and examination

Domain	Minimum assessment
Pain pattern	Onset, sites, severity, similarity to usual VOE, provoking factors, home treatment and response, effective prior regimen, opioid tolerance and adverse effects.
High-risk symptoms	Fever / rigors, cough, dyspnoea, chest pain, haemoptysis, syncope, focal neurology, severe headache, seizure, visual loss, abdominal distension, vomiting, priapism and reduced urine.
Recent events	Infection exposure, travel, dehydration / heat, altitude, exertion, surgery, pregnancy, immobilization, missed medicines, new drugs and recent hospital attendance.
SCD history	Genotype, baseline Hb / reticulocytes / SpO2, prior ACS, stroke, sequestration, splenectomy, renal disease, pulmonary hypertension, avascular necrosis, chronic pain and frequent admissions.
Transfusion history	Date and indication of recent transfusions, exchange programme, previous antibodies, DHTR / hyperhaemolysis and transfusion card.
Medicines	Hydroxyurea / hydroxycarbamide and other disease-modifying therapy, penicillin prophylaxis, anticoagulant, opioids, NSAIDs, pregnancy-related changes and adherence.

Domain	Minimum assessment
Examination	General appearance, hydration, pallor, jaundice, perfusion, respiratory and cardiac examination, abdomen / spleen / liver, full neurology, painful joints / bones and skin / line sites.
Psychosocial	Patient goals, prior negative care experiences, mental health, caregiver support, safe medicine storage and barriers to follow-up.

10. Acute vaso-occlusive pain

- Treat reported pain promptly and proportionately. Normal observations, a calm appearance, sleep, phone use or frequent attendance do not invalidate severe pain.
- Prefer the patient-specific plan. If absent, select analgesia from pain severity, age / weight, current home opioid exposure, renal / hepatic function, allergies, pregnancy and previous response.
- For severe pain use a parenteral opioid by IV or subcutaneous route; intranasal options may be used under local paediatric or adult protocols when access is delayed. Oral opioids may be suitable for moderate pain when absorption is reliable.
- Use regular paracetamol and a short course of NSAID when appropriate, after assessing renal function, dehydration, GI bleeding, anticoagulation, asthma, pregnancy and cardiovascular risk. Avoid duplicate products.
- After initial titration, use scheduled dosing or patient-controlled analgesia rather than long gaps between PRN doses for admitted patients with ongoing severe pain. Prescribe antiemetic, bowel regimen and monitoring as appropriate.
- Offer non-pharmacological measures such as local heat, distraction, relaxation and quiet surroundings. Avoid cold packs on painful areas.
- Do not use pethidine / meperidine routinely because of neurotoxic metabolite accumulation and seizure risk. Do not use corticosteroids solely for uncomplicated VOE.
- Refractory pain requires senior review for missed complication, opioid-induced hyperalgesia, chronic pain phenotype, substance withdrawal, renal / hepatic dysfunction and specialist analgesic adjuncts. Monitored low-dose ketamine may be considered only under an approved protocol.
- Do not transfuse for uncomplicated VOE. Repeated severe pain should trigger specialist review of disease-modifying and longitudinal care after the acute episode.

PAIN REASSESSMENT IS A TREATMENT DECISION: Record the patient's pain relief, function, respiratory rate, oxygen saturation and sedation after every parenteral dose. Persistent pain means reassess, repeat or escalate - not simply wait.

11. Fluids, oxygen and supportive care

Intervention	Standard
Oxygen	Give for hypoxaemia, a fall below baseline, respiratory distress, shock or another clinical indication. Investigate any new oxygen requirement. Do not use routine oxygen as a substitute for diagnosing ACS.
Hydration	Encourage oral fluids when safe. If unable to drink or clinically dehydrated, give cautious isotonic IV fluid with frequent reassessment. Avoid routine boluses or overhydration in euvolaemic patients because pulmonary oedema and ACS may worsen.
Temperature	Maintain warmth and treat fever / sepsis. Avoid exposure to cold.
Incentive spirometry	Use for hospitalized VOE, especially chest / back / upper abdominal pain, reduced mobility or opioid treatment, according to age and local protocol. Encourage regular deep breathing and early mobilization when safe.
Thrombosis prevention	Assess VTE risk in admitted adults, pregnancy and reduced mobility. Use pharmacological prophylaxis when indicated and not contraindicated; investigate PE when clinically suspected.
Regular medicines	Reconcile and continue time-critical medicines. Do not automatically stop disease-modifying therapy; seek specialist advice for cytopenia, organ failure, pregnancy or suspected adverse effect.

12. Acute chest syndrome

- Suspect ACS with any new chest pain, cough, fever, dyspnoea, tachypnoea, wheeze, hypoxaemia, reduced exercise tolerance or unexplained fall in haemoglobin. It can evolve after admission for pain.
- Obtain chest X-ray, FBC with reticulocytes, chemistry, group and screen and infection investigations. A normal early chest X-ray does not exclude evolving ACS; repeat imaging if symptoms or oxygenation worsen.
- Provide oxygen to restore the person's usual saturation or local target, continuous oximetry, prompt analgesia that permits ventilation, incentive spirometry and careful fluid balance.
- Give empiric antibiotics covering typical and atypical respiratory pathogens according to local age-specific guidance; investigate viral infection and obtain cultures when appropriate.
- Use bronchodilator for wheeze / bronchospasm. Corticosteroids are not routine for ACS but may be required for a clear concurrent indication such as significant asthma, with specialist awareness of rebound VOE risk.
- Discuss transfusion early. Simple transfusion may be appropriate for moderate ACS with anaemia / significant fall from baseline. Urgent red-cell exchange is preferred for severe or rapidly progressive ACS, severe hypoxaemia, multiorgan involvement, high pre-transfusion haemoglobin or failure to improve after simple transfusion.
- Escalate immediately for increasing oxygen need, multilobar infiltrates, rising respiratory rate, acidosis, falling platelets / haemoglobin, altered mental state, haemodynamic compromise or need for non-invasive / invasive ventilation.
- Consider pulmonary embolism, fat embolism, pneumonia, fluid overload, asthma and cardiac dysfunction. Arrange post-episode haematology review and disease-modifying care.

13. Fever, infection and sepsis

- Functional asplenia and prior splenectomy increase the risk of rapid invasive bacterial infection. Fever of 38.0 degrees C or higher, reported fever with unwellness, rigors or hypothermia warrants urgent assessment; local paediatric thresholds may be more specific.
- Complete sepsis assessment, FBC / reticulocytes, cultures, renal / liver tests, lactate when indicated and focused imaging. Consider malaria after relevant travel, dengue and other local / travel infections, line infection, osteomyelitis, septic arthritis, meningitis and ACS.
- Give empiric IV antibiotics within 60 minutes for suspected serious infection and immediately in shock, using local age-specific regimens that cover encapsulated organisms and the suspected source. Do not delay for imaging or difficult access.
- Use cautious fluid resuscitation with repeated perfusion and respiratory assessment. SCD patients can deteriorate from both under-resuscitation and fluid overload.
- Admit or observe children and adults according to clinical appearance, age, genotype / splenic function, source, cultures, immunization / prophylaxis history, comorbidity and reliable follow-up. A single normal test does not make an unwell patient low risk.
- If discharge after specialist review is appropriate, ensure a clear antimicrobial plan, culture follow-up owner, direct return route and review within the required timeframe.

14. Acute neurological emergency and stroke

1. Activate Protocol 18 immediately for any focal deficit, speech or visual change, severe acute headache, seizure, acute ataxia, altered consciousness or suspected TIA. Record exact last-known-well and glucose.
2. Obtain urgent brain and vascular imaging according to the stroke pathway, but do not allow imaging or subspecialty delay to postpone urgent SCD transfusion treatment when stroke is strongly suspected.
3. Contact haematology and transfusion medicine immediately. Aim to begin transfusion as soon as possible and no later than 2 hours after presentation when feasible.
4. Exchange transfusion is preferred for acute ischaemic stroke. If exchange is not immediately available and haemoglobin is sufficiently low, give a carefully planned simple transfusion while arranging exchange / transfer. Avoid hyperviscosity from excessive haemoglobin elevation.
5. Continue standard assessment for thrombolysis or thrombectomy eligibility; transfusion complements rather than replaces stroke reperfusion and supportive care.
6. Treat fever, hypoxaemia, hypotension and seizures promptly. Avoid unnecessary delays in transfer to a centre capable of neurocritical and exchange-transfusion care.
7. Include retinal artery occlusion or sudden monocular visual loss as a stroke-equivalent emergency requiring urgent ophthalmology, stroke and haematology input.

15. Acute anaemia, aplastic crisis and sequestration

Syndrome	Clues and immediate management
Aplastic crisis	Acute pallor / fatigue with haemoglobin below baseline and inappropriately low reticulocyte count, often after viral symptoms. Test for parvovirus B19 as locally available; use infection precautions; transfuse if clinically indicated after specialist discussion.
Splenic sequestration	Sudden splenic enlargement, abdominal pain / distension, pallor, tachycardia, shock and acute Hb fall with reticulocytosis. Resuscitate, urgent haematology / paediatrics and cautious red-cell transfusion. Avoid overcorrection because sequestered blood may remobilize.
Hepatic sequestration / severe hepatic crisis	Enlarging tender liver, jaundice, falling Hb, reticulocytosis and possible coagulopathy / organ failure. Urgent specialist and critical-care assessment; transfusion or exchange may be required.
Accelerated haemolysis	Jaundice, dark urine, rising bilirubin / LDH and falling Hb. Investigate infection, drugs, G6PD-related triggers, autoimmune haemolysis and transfusion reaction.
Bleeding or other cause	Do not attribute every Hb fall to SCD. Assess menstrual / obstetric, GI, surgical and traumatic bleeding, renal loss and nutritional deficiency.

COMPARE WITH BASELINE: A haemoglobin that looks "low" by population standards may be normal for the patient, while a seemingly acceptable value may represent a dangerous acute fall. Always review baseline Hb and reticulocyte response.

16. Delayed haemolytic transfusion reaction and hyperhaemolysis

- Suspect DHTR in any patient presenting within days to weeks of transfusion with pain, fever, jaundice, dark urine, fatigue or an unexplained haemoglobin fall. It can mimic VOE.
- Immediately contact haematology and transfusion medicine / blood bank. Retrieve all historical antibodies and transfusion records.
- Obtain FBC, reticulocytes, bilirubin, LDH, haptoglobin where interpretable, urinalysis, DAT, antibody screen / identification and haemoglobin fractionation as available. Repeat testing may be needed because antibodies and DAT can initially be negative.
- Hyperhaemolysis is suggested when post-transfusion haemoglobin falls below the pre-transfusion level with destruction of donor and patient red cells.
- Avoid reflex additional transfusion in a stable patient because it may worsen haemolysis. If anaemia is life-threatening, transfuse only after urgent joint planning using the safest matched blood and the minimum necessary exposure.
- Immunosuppressive and supportive treatment is specialist-directed. Monitor haemoglobin, reticulocytes, bilirubin, LDH, renal function and cardiopulmonary status closely; admit or transfer to an experienced centre.

17. Transfusion principles in the emergency department

Principle	Requirement
Indications	Use for acute stroke, severe / progressive ACS, clinically significant acute anaemia, sequestration, aplasia with symptoms, life-threatening haemolysis or other specialist-defined indications - not uncomplicated pain or stable chronic anaemia.
Pre-transfusion checks	Confirm baseline Hb, current Hb / reticulocytes, indication, target, recent transfusions, pregnancy, cardiac / renal status, antibody history and prior DHTR / hyperhaemolysis.
Blood selection	Notify the blood bank that the patient has SCD. Use HbS-negative, leucocyte-reduced and antigen-matched red cells according to local / regional policy and all current or historical antibodies.
Simple transfusion	Useful when oxygen-carrying capacity must increase and pre-transfusion Hb is low. Avoid raising total Hb excessively because hyperviscosity can worsen vaso-occlusion.
Exchange transfusion	Preferred for acute stroke and severe / rapidly progressive ACS, and considered for selected multiorgan or severe hepatic complications. Start transfer early if unavailable locally.
Monitoring	Use standard transfusion observation plus close respiratory and volume-status monitoring. Stop and investigate any suspected reaction.
Communication	Record unit identifiers, phenotype / genotype matching, antibody information, target and response. Give the patient updated transfusion information before discharge.

18. Priapism

1. Ask directly and privately about erection duration in boys, adolescents and men with lower abdominal, genital or unexplained pain. Record the start time and previous episodes.
2. Provide immediate analgesia, IV access if needed, hydration only if dehydrated and oxygen only for hypoxaemia. Do not use ice packs or cold baths.
3. Contact urology early for an episode persisting beyond 2 hours or not resolving with the patient's approved home plan. Ischaemic priapism lasting 4 hours or more requires emergency corporal aspiration / irrigation and intracavernosal sympathomimetic treatment under monitoring.
4. Do not delay definitive urological intervention while arranging transfusion. Routine simple or exchange transfusion is not first-line acute treatment.
5. Admit / observe according to response and complications. Arrange haematology and urology follow-up for stuttering priapism and prevention planning.

19. Abdominal, musculoskeletal, renal and ocular emergencies

Presentation	Do not miss
Abdominal pain / vomiting	Splenic or hepatic sequestration, cholecystitis / cholangitis, pancreatitis, appendicitis, bowel ischaemia / obstruction, renal colic, pyelonephritis, pregnancy complications and opioid-related constipation.
Bone or joint pain	Osteomyelitis, septic arthritis, fracture, avascular necrosis, compartment syndrome and gout. Fever, focal swelling, inability to bear weight or persistent single-site pain requires targeted investigation.
Haematuria / flank pain	Papillary necrosis, infection, stone, renal infarction, AKI and renal medullary carcinoma, especially with persistent or gross haematuria.
Sudden visual symptoms	Retinal vascular occlusion, vitreous haemorrhage, retinal detachment and acute glaucoma. Urgent ophthalmology; treat sudden visual loss as a neurological emergency.
Leg swelling / dyspnoea	DVT / pulmonary embolism. SCD increases thrombotic risk; use standard diagnostic pathways and do not attribute symptoms to pain alone.
Severe diffuse pain with organ dysfunction	Consider acute multiorgan failure, fat embolism, sepsis, rhabdomyolysis and severe ACS; obtain critical-care and haematology support.

20. Pregnancy and postpartum presentations

- Pregnancy with SCD is high risk. Involve obstetrics and haematology early for pain, fever, chest symptoms, anaemia, hypertension, bleeding, contractions or reduced fetal movement.
- Assess and treat maternal emergencies first while arranging gestation-appropriate fetal assessment. Use standard obstetric pathways for ectopic pregnancy, pre-eclampsia / eclampsia, haemorrhage, labour and thromboembolism.
- Use the individualized pain plan and pregnancy-safe multimodal analgesia. NSAID use depends on gestation and obstetric guidance. Avoid withholding adequate opioids when clinically needed.
- Maintain oxygenation, normothermia and careful hydration. Have a low threshold for admission with ACS, infection, hypoxaemia, dehydration, uncontrolled pain or obstetric concern.
- Transfusion decisions require joint obstetric-haematology review and attention to alloantibodies and haemolytic disease risk. Routine transfusion is not automatic, but acute complications may require urgent simple or exchange transfusion.
- Postpartum patients remain at high risk of pain, ACS, infection and VTE. Ensure thromboprophylaxis assessment and coordinated follow-up.

21. Paediatric considerations

- Use Protocol 40 and age-specific observations, pain tools and medicine dosing. Weigh the child; do not estimate when avoidable.
- Fever, poor feeding, lethargy, respiratory symptoms, pallor or parental concern can signal severe infection or acute anaemia. Infants may not localize pain.

- Ask caregivers about the child's usual spleen size and whether they have been taught palpation. Gently assess for new splenomegaly; mark / document the spleen edge when sequestration is suspected.
- Use rapid-access analgesia, including intranasal routes where locally approved. Reassess behaviour, function, respiratory status and sedation, not only a numeric score.
- Consider dactylitis, osteomyelitis, septic arthritis, splenic sequestration, aplastic crisis, ACS and stroke. Any new weakness, speech change, seizure or severe headache requires immediate stroke pathway.
- Engage the child and caregiver, maintain warmth, minimize repeated procedures and provide clear home / return instructions. Confirm prophylactic antibiotics and immunization follow-up without delaying acute care.

22. Investigations

Clinical context	Suggested initial tests - tailor to presentation
Uncomplicated typical VOE, clinically well	No mandatory broad panel. Consider FBC / reticulocytes and chemistry if severe, prolonged, atypical, pregnancy, renal disease, admission or no recent baseline.
Fever / sepsis	FBC / reticulocytes, cultures, renal / liver tests, lactate when indicated, urine and source-specific tests, chest imaging for respiratory symptoms, viral / travel testing as relevant.
Chest symptoms / hypoxia	FBC / reticulocytes, chemistry, group and screen, chest X-ray, cultures / viral testing, blood gas when severe, ECG / troponin / PE work-up as clinically indicated.
Neurological symptoms	Glucose, FBC / reticulocytes, chemistry, group and screen / crossmatch, urgent CT / CTA or MRI / MRA per stroke pathway; do not delay transfusion planning.
Acute anaemia / jaundice	FBC / reticulocytes, bilirubin, LDH, chemistry, DAT / antibody screen, urinalysis and haemoglobin fractionation as available; assess spleen / liver and bleeding.
Recent transfusion	Full haemolysis and transfusion-reaction work-up with immediate blood-bank involvement.
Priapism	Primarily clinical; targeted FBC / reticulocytes, chemistry and group and screen based on severity and procedure. Do not delay urological treatment.

23. Observation and senior reassessment

- Observation is active treatment. Set explicit goals for pain, function, oral intake, oxygenation, respiratory findings, temperature, haemoglobin trend and diagnosis.
- Repeat vital signs, pain and sedation after each opioid dose and at intervals appropriate to risk. Continuous oximetry is required for ACS, hypoxaemia, significant sedation, respiratory disease or escalating parenteral opioids.
- Re-examine the chest and reassess for fever, neurological change, abdominal enlargement, urine output and priapism. ACS frequently emerges after initial presentation.
- Failure to improve after repeated analgesia requires senior reassessment, not automatic discharge or indefinite dosing without a revised plan.
- Use Protocol 53 only when observation capability, monitoring, review intervals and conversion-to-admission criteria are met. Do not place a high-risk SCD patient in an unmonitored waiting area.

24. Admission, critical care and transfer

Disposition	Indications / requirements
Critical care / urgent tertiary transfer	Severe or progressive ACS, escalating oxygen / ventilation, shock, acute stroke, multiorgan failure, severe haemolysis / hyperhaemolysis, exchange transfusion need, refractory seizures or other critical illness.
Hospital admission	Uncontrolled pain, repeated parenteral dosing / PCA need, fever / infection, new oxygen requirement, ACS, significant Hb fall, AKI, vomiting / dehydration, priapism after procedure, pregnancy complication, unsafe home plan or diagnostic uncertainty.
Observation / short stay	Stable physiology with typical VOE, improving pain and a defined reassessment plan; no high-risk complication; reliable monitoring, specialist access and clear maximum duration.
Discharge	Pain and function improved to a manageable level, vital signs stable, no evolving ACS / sepsis / neurological / anaemia concern, oral medicines and fluids tolerated, and follow-up / return plan verified.
Transfer preparation	Stabilize ABCDE, begin analgesia / antibiotics / oxygen / transfusion as indicated, send imaging and laboratory data, communicate antibodies and baseline values, and confirm receiving clinician, transport capability and contingency plan.

25. Safe discharge bundle

☐ Pain is improved and manageable with the agreed home regimen; patient can mobilize / function sufficiently for destination.
☐ No new fever, hypoxaemia, chest symptoms, neurological findings, significant anaemia, abdominal enlargement, priapism or other dangerous alternative diagnosis.
☐ Final observations and oxygen saturation documented and compared with baseline where known.
☐ Medicines supplied / available, with clear dose, interval, maximum daily dose, side-effect prevention and safe opioid storage / disposal advice.
☐ Hydration, warmth, incentive spirometry / breathing advice and activity guidance provided as appropriate.
☐ Specific red flags explained: fever, chest pain / breathlessness, severe headache / weakness / visual change, worsening pallor / jaundice, enlarging abdomen, reduced urine, uncontrolled pain or erection approaching 4 hours.
☐ Direct contact route for sickle cell / paediatric / haematology service and primary care follow-up confirmed; early review arranged after significant episodes.
☐ Pending cultures, imaging or laboratory results have a named owner and reliable contact method.
☐ Transfusion details and new antibody information provided when relevant.

☐ Patient / caregiver understands the plan using teach-back and agrees that discharge is safe.

26. Documentation and handover

☐ Genotype, baseline Hb / reticulocytes / SpO2, prior ACS / stroke / sequestration / DHTR and relevant comorbidities.
☐ Individual pain plan used or reason unavailable; home opioid use and effective previous regimen.
☐ Arrival, triage, pain score, first analgesia and every reassessment / repeat-dose time.
☐ Full observations, oxygen need, respiratory / neurological / abdominal findings and serial changes.
☐ Complications actively considered and reasons for diagnosis / exclusion.
☐ Investigations and comparison with baseline, including reticulocyte response.
☐ Fluid, oxygen, antibiotics, incentive spirometry, transfusion and specialist advice.
☐ Transfusion indication, target, blood-bank discussion, antibody history and response.
☐ Capacity, pregnancy status, safeguarding / psychosocial issues and patient preferences.
☐ Destination, receiving clinician / service, pending-results owner, discharge medicines, follow-up and return advice.

27. Quality indicators and audit

Indicator	Suggested measure
Time to first analgesia	Median and percentage receiving analgesia within 30 minutes of triage and within 60 minutes of arrival; stratify by age, sex and presentation frequency.
Pain reassessment	Percentage with documented pain, sedation and respiratory reassessment within the approved interval after parenteral opioid.
Individualized plans	Percentage of frequent attenders with accessible, current patient-specific pain / emergency plans.
Complication recognition	Missed or delayed ACS, sepsis, stroke, sequestration, priapism, PE, pregnancy complication and DHTR / hyperhaemolysis.
ACS care	Time to oxygen when hypoxaemic, antibiotics, imaging, incentive spirometry, specialist / critical-care review and transfusion decision.
Stroke care	Time from arrival to stroke activation, imaging, haematology contact and initiation of transfusion.
Transfusion safety	Appropriate indication, antibody-history retrieval, matching compliance, transfusion reactions and avoidable hyperviscosity / fluid overload.
Disposition	Unplanned return within 72 hours / 7 days, admission after observation, transfer delay and discharge with unresolved red flags.
Patient experience / equity	Respect, involvement, confidence in pain care, stigma reports, delay disparities and access to follow-up.

28. Training and implementation

- Provide mandatory orientation and recurrent simulation for triage, rapid analgesia, opioid monitoring, ACS, stroke, sequestration, priapism, transfusion reaction and paediatric deterioration.
- Co-design individualized plans, patient information and quality review with people living with SCD and caregivers.
- Maintain electronic order sets, accessible baseline and antibody information, and a clear route to haematology / transfusion medicine at all hours.
- Use case review to identify bias, stigma, delay, repeated cannulation, undertreatment, over-sedation, missed deterioration and unsafe discharge.
- Ensure regional agreements for exchange transfusion, neuroimaging, critical care and paediatric transfer, including time targets and transport requirements.

29. Local configuration before approval

☐ Named adult and paediatric clinical leads and 24-hour specialist contacts.
☐ Approved rapid-analgesia order set with adult / paediatric doses, routes, reassessment intervals, monitoring, naloxone and escalation.
☐ Local fever / sepsis antibiotic regimens and thresholds.
☐ ACS oxygen target, antibiotic bundle, incentive-spirometry process, critical-care triggers and transfusion pathway.
☐ Acute stroke transfusion / exchange and transfer pathway with 2-hour operational target.
☐ Blood selection, antigen matching, emergency release, antibody retrieval, DHTR / hyperhaemolysis and exchange-transfusion policy.
☐ Urology pathway and monitored intracavernosal sympathomimetic protocol for priapism.
☐ Pregnancy / obstetric and paediatric pathways.
☐ Observation eligibility, staffing, maximum duration and discharge criteria.
☐ Patient-facing red-flag and discharge information; direct follow-up contacts.
☐ Audit dashboard and serious-incident review process.

30. Source guidance for local adaptation

Source	Key use in this protocol
American Society of Hematology. Clinical Practice Guidelines on Sickle Cell Disease: Management of Acute and Chronic Pain. 2020; expert review completed 2023.	Rapid patient-centred assessment, analgesia within 1 hour, individualized dosing, repeated reassessment, multimodal care and avoidance of bias.
ASH. Sickle Cell Disease Quality Measures and Methodology Reports. 2024-2025.	Emergency-department measures for median time to pain medication and equitable guideline-adherent treatment.
National Institute for Health and Care Excellence. CG143: Sickle Cell Disease - Managing Acute Painful Episodes in Hospital. 2012; surveillance review 2023.	Treating acute pain as a medical emergency, analgesia within 30 minutes, assessment, reassessment and discharge information.

Source	Key use in this protocol
American Society of Hematology. Clinical Practice Guidelines: Cerebrovascular Disease and Transfusion Support. 2020; expert review 2023.	Urgent transfusion for acute neurological deficits, exchange transfusion for stroke / severe ACS, antigen profiling and DHTR / hyperhaemolysis.
British Society for Haematology. Management of Acute Chest Syndrome in Sickle Cell Disease. 2015.	ACS definition, monitoring, imaging, infection treatment, incentive spirometry, early transfusion and critical-care escalation.
British Society for Haematology. Red Cell Transfusion in Sickle Cell Disease, Parts I and II. Part II reviewed January 2024.	Acute transfusion indications, sequestration / aplasia / ACS, blood selection, hyperviscosity avoidance and specialist planning.
WHO Regional Office for Africa. WHO SICKLE Package of Interventions and Harmonized Guide for Sickle Cell Disease Management in Africa. 2024.	Integrated, age-attuned, resource-aware SCD care, emergency recognition, comprehensive pathways and patient education.
Local protocols and formularies	Pain medicines, sepsis, stroke, respiratory support, transfusion, pregnancy, paediatrics, controlled drugs, transfer and clinical governance.

Annex A. One-page sickle cell emergency workflow

Stage	Action
1. Recognize	Identify SCD, pain and danger symptoms at triage. Obtain full observations, SpO2, temperature, pain score, weight and glucose.
2. Treat pain now	Retrieve individual plan and give first analgesia by fastest safe route - target within 30 minutes of triage / 60 minutes of arrival.
3. Screen complications	Chest / hypoxia, fever / sepsis, stroke / seizure, anaemia / spleen / liver, priapism, recent transfusion, pregnancy and other diagnoses.
4. Investigate selectively	Use presentation and baseline values. FBC + reticulocytes and group / screen for high-risk cases; imaging and cultures without delaying treatment.
5. Support safely	Warmth, oral fluids or cautious IV hydration, oxygen for hypoxaemia, incentive spirometry and monitored opioid care.
6. Escalate	Early senior, haematology / paediatrics / internal medicine; critical care for ACS / shock; stroke team; urology; obstetrics; blood bank.
7. Reassess	Pain and sedation after each dose; serial chest, neurology, oxygenation, temperature, haemoglobin / reticulocytes and response.
8. Decide disposition	Admit / transfer for complication, uncontrolled pain or uncertainty; discharge only after improvement, safe medicines, follow-up and teach-back.

Annex B. Rapid analgesia and monitoring checklist

☐ Pain score / functional impact recorded using age-appropriate tool.
☐ Individual pain plan retrieved; home opioid use and last dose confirmed.
☐ First analgesia administered at _____; route _____; arrival _____; triage _____.
☐ Paracetamol / NSAID suitability assessed; duplicate medicines avoided.
☐ Parenteral opioid dose and monitoring comply with local policy; naloxone available.
☐ Reassessment after each dose: pain, respiratory rate, SpO2, sedation, nausea / pruritus.
☐ Repeat / escalation decision recorded every 15-30 minutes during titration.
☐ Warmth, oral fluids if safe, local heat and patient-preferred non-drug measures offered.
☐ Complications re-screened at every reassessment.
☐ If ongoing severe pain: senior review, PCA / scheduled regimen, admission and pain / haematology input considered.

Annex C. Acute chest syndrome bundle

☐ Continuous oximetry; baseline SpO2 obtained if known; oxygen for hypoxaemia / fall below baseline.
☐ Chest X-ray obtained; repeat planned if early film normal but symptoms progress.
☐ FBC / reticulocytes, chemistry, group and screen, cultures / viral tests as indicated.
☐ Prompt analgesia with respiratory and sedation monitoring.
☐ Empiric age-appropriate typical + atypical respiratory antimicrobial coverage.
☐ Incentive spirometry / deep-breathing plan and early mobilization when safe.
☐ Careful hydration and fluid balance; avoid overhydration.
☐ Bronchodilator if wheeze / bronchospasm.
☐ Haematology and senior ED / paediatric / medical review; simple versus exchange transfusion decision documented.
☐ Critical-care review for deterioration, increasing oxygen, multilobar disease, acidosis, shock or ventilatory support need.
☐ Reassessment times and transfer contingency documented.

Annex D. Acute anaemia and transfusion decision aid

Question	Document
What is the patient's baseline Hb / reticulocyte count?	Baseline: _____ / _____ Source: _____
What is the current Hb / reticulocyte count and clinical effect?	Current: _____ / _____ Symptoms / perfusion: _____
Is there sequestration, aplasia, bleeding, haemolysis, ACS or stroke?	_____
Any transfusion in past 3 months? Date / units / indication?	_____
Historical antibodies or DHTR / hyperhaemolysis?	_____
Indication and target for transfusion?	_____
Simple or exchange, and why?	_____
Blood bank / haematology contacted; compatible units / transfer plan?	Time: _____ Clinician: _____

Question	Document
Response and repeat Hb / clinical endpoint?	_____

Annex E. Neurological emergency checklist

☐ Last-known-well / symptom onset recorded: _____.
☐ Glucose, ABCDE and full neurological examination / stroke score completed.
☐ Stroke / neurology pathway activated; urgent CT / CTA or MRI / MRA requested.
☐ Haematology and transfusion medicine contacted immediately.
☐ Transfusion planned to start as soon as possible and within 2 hours when feasible.
☐ Exchange preferred; if delayed and Hb low, interim simple transfusion considered with hyperviscosity precautions.
☐ Reperfusion / thrombectomy eligibility assessed under standard stroke protocol.
☐ Fever, hypoxaemia, hypotension and seizures treated.
☐ Receiving centre and transport capability confirmed if exchange / neurocritical care unavailable.

Annex F. Sickle cell discharge and return-safety checklist

☐ Pain and function manageable with available home treatment.
☐ No fever, chest symptoms, hypoxaemia, neurological change, significant anaemia / jaundice, enlarging spleen / liver, priapism or concerning alternative diagnosis.
☐ Final vital signs, SpO2 and examination documented.
☐ Medicine names, doses, maximums, adverse effects, constipation / nausea plan and safe opioid storage reviewed.
☐ Patient has fluids, warmth, transport, caregiver support and ability to obtain follow-up.
☐ Return immediately for fever; chest pain / breathlessness; weakness / speech / vision change; severe headache / seizure; worsening pallor / jaundice; abdominal enlargement; reduced urine; uncontrolled pain; or erection approaching 4 hours.
☐ Sickle cell / haematology / paediatric contact and review date provided.
☐ Pending results and named follow-up owner documented.
☐ Transfusion / antibody information updated when applicable.
☐ Patient / caregiver used teach-back and agrees with discharge plan.

END OF PROTOCOL 45 - DRAFT 1.0 FOR LOCAL MULTIDISCIPLINARY VALIDATION