| Problem | What to do | When to refer | Who to refer to |
|---|---|---|---|
| **Abdominal/Hepatobiliary** | | | |
| **Symptoms** | Request ultrasound liver and biliary system | If gallstones identified and any evidence of obstruction OR continuing symptoms, so that lap cholecystectomy would be considered | Refer to Surgical team for consideration of cholecystectomy |
| **Liver related problems** | Conjugated and unconjugated bilirubin, full liver function tests, abdominal ultrasound and viral hepatitis screen. | Evidence of sickle cell hepatopathy (deranged LFTs, US showing hepatomegaly, increased echogenicity and histopathology shows widespread sinusoidal sickling), cirrhosis or unexplained hepatomegaly | Referral to Sickle Liver Clinic |
| **Acute Hepatitis/Liver Failure** | | Urgent discussion and transfer to specialist liver unit. | Referral to Hepatologist |
| **Circulation/Pulmonary/Cardiac** | | | |
| **Acute Anaemia/Hypovolaemia** | | | Discuss possible Haemolytic transfusion reaction with local transfusion laboratory, RCI NHSBT Laboratory and NHSBT Consultant |
| **Cardiac Problem** | Symptomatic of cardiac pathology Perform ECG, 24hour tape, ECHO.<br>If possible transfusion related iron overload arrange cardiac MRI T2* | Any abnormal cardiac investigations which require specialist input.<br>T2* <10ms | Refer to Cardiology |
| **Pulmonary Hypertension** | Routine ECHO every 1-3 years specifically reviewing Tricuspid regurgitant jet velocity | Raised TRV >2.5m/s | Refer to Sickle Pulmonary Hypertension Clinic |
| **Respiratory** | Chronic lung problems (without known cause e.g asthma/COPD) note history and request CXR and CT Chest<br><br>If chronic low baseline oxygen. Request Lung Function tests, Oximetry / Sleep Study and HR CT Chest | According to investigation results.<br><br>If sleep disordered breathing | Refer to Sickle Respiratory Clinic. |
| **Leg Ulceration** | Check appearance, take swab for MC&S, advise daily cleaning with saline solution, application of non-adherent dressing, firm strapping and elevate.<br><br>Antibiotics to treat clinical infection/cellulitis. | If not healing, deep, slough covered or heavy exudate. | Refer to Tissue Viability Service.<br><br>Discuss with Consultant Microbiologist. |
| **Endocrine** | | | |
| **Endocrinology** | LH,FSH, testosterone/oestradiol<br><br>R2 MRI scan of liver<br><br>Bone mineral density scan (DEXA) | Abnormal findings | Referral to Endocrinology |
| **Infection** | | | |
| **Infection** | Fever, rigors, MC&S | Discussion with Microbiology | Referral to Consultant Microbiologist/Infectious Diseases |
| **Mental Health** | | | |
| **Mental Health** | Discuss in outpatient clinic and ensure moos score at least annually | Referral if indicated and patient consents. | Referral to red cell psychology service. |
| **Neurology** | | | |
| **Neurological problems** | TIA/stroke usually presents acutely BUT may give history of TIA or other neurology as outpatient.<br><br>Full neurological examination, request CT scan brain with contrast, MRI and MRA brain. | | Referral to Sickle Neurology Clinic. |
| **Ophthalmology** | | | |

| | | | |
|---|---|---|---|
| **Retinopathy** | Screen according to genotype and degree of retinopathy.<br><br>Patient reports acute change in vision. | Routine screen.<br><br>Urgent referral. | Refer to Ophthalmology clinic. |
| **Orthopaedic** | | | |
| **Orthopaedic problems** | Examine range of movement of affected site.<br><br>If pain in one joint for more than 1 month request plain X ray and if no abnormalities noted then MRI scan. | If evidence of Avascular Necrosis or impingement syndrome. | Referral to Sickle Hip/Orthopaedic Clinic. |
| **Osteomyelitis** | Fever, painful swollen limb with limited range of movement.<br><br>Request inflammatory markers, Ultrasound, MRI.<br>Start Antibiotics. | Always | Sickle- Orthopaedic MDT<br>Liaise with Infectious Diseases/Consultant Microbiologist |
| **Pain** | | | |
| **Chronic Pain** | Identify patients with ongoing and multifactorial pain requiring increasing analgesic medication. | Psychologist for help with non-pharmacological interventions. | Referral to Sickle Pain Clinic/MDT. |
| **Renal / Urology** | | | |
| **Renal Disease** | U&E, Creatinine, eGFR, Urine ACR, calcium, phosphate, uric acid, renal tract ultrasound.<br><br>If urine ACR>50 check renal screen (C3, C4, ACE, autoimmune screen – anti CCP, ANA autoantibodies, protein electrophoresis, serum free light chains) | Persistently raised Urine ACR<br>Hypertension BP >140/90mmKH<br>Rising creatinine | Referral to Sickle Renal Clinic. |
| **Haematuria** | Urinalysis, MSU, US Renal tract and CT KUB.<br><br>If UTI then for antibiotic treatment | Haematuria not resulting form simple resolved UTI. | Referral to Sickle Urology Clinic. |
| **Priapism** | Check with male patients routinely.<br><br>Stuttering priapism – suggest emptying bladder, hydration, oral analgesia, warm bath and exercise.<br><br>Trial of Etilefrine nocte. | If symptoms continue despite trial of Etilefrine.<br><br>Urgent inpatient urology referral for patient who attends A&E with prolonged priapism. | Referral to Sickle Urology clinic. |
| **Obstetrics & Gynaecology** | | | |
| **Fertility** | Encourage partner testing (Wooden Spoon clinic) | Referral to specialist councillors | Referral to Fertility Clinic. |
| **Obstetric** | Patient reports pregnancy. | Always | Refer to Sickle Obstetric Clinic. |
| **Transplantation and Novel Treatment** | | | |
| **Transplantation** | Meets transplantation criteria and / or patient request.<br>HLA typing of patient and consider family members | Local and HCC MDT discussion and then referral to NHP. | Refer to HCC MDT for transplant. |
| **Blood Transfusion** | Ensure full red cell phenotyping and genotyping performed | Multiple red cell antibodies.<br><br>Rare phenotypes/genotypes.<br><br>Transfusion reactions | Liaison with Local transfusion team and NHSBT |
| **Hydroxycarbamide** | Discuss with all patients<br>Ensure adequate monitoring | | Hydroxycarbamide monitoring in CNS/OP clinic. |
| **Novel treatments and clinical trials** | Patient meets criteria | Local and HCC MDT discussion | HCC/NHP |

This has been adapted from “The Red Cell Network – Adult Sickle Cell Disorder Unified Network Guideline” with permission.