NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Adult Guideline - Sickle Cell Outpatient Management

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

*Usually only required at New Patient Attendance

This guideline is designed to support the clinician when meeting a patient for the first time, or when performing an Annual Review.

Annual review/ *New Patient Attendance

The following should be considered at each annual review visit (* or new patient attendance):

- General wellbeing

- Sickle related complications including their current and proposed management
    - Acute chest syndrome
    - Bone Problems including avascular necrosis (AVN), osteopenia, osteoporosis
    - Osteomyelitis
    - Gallstones
    - Chronic Hepatitis B, C or HIV
    - Hepatic or splenic sequestration
    - Hyperhaemolysis
    - Leg ulcers
    - Multi-organ failure
    - Neurological including stroke/TIA
    - Bacteraemia
    - Pneumococcal infection
    - Parvovirus
    - Priapism
    - Pulmonary hypertension
    - Renal failure
    - Sickle Retinopathy
    - Sickle hepatopathy

- Outcomes of specialist clinic reviews in past 12 months

- Number of emergency attendances in past 12 months
    - Number of days off work/education
    - Home pain plan including opiate use
    - Acute care pain plan, if necessary

- Upcoming procedures
  In particular to consider the support required peri-procedure (refer to Perioperative management in sickle cell disease guideline that is available on the SPAH website)

- Current medications, side effects and compliance issues
    - Penicillin V (erythromycin)

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  - Folic acid
  - ACE inhibitors
  - Hydroxycarbamide
  - Vitamin D
  - Iron Chelation
  - Analgesia
  - Others including potential new treatment options available

- Immunisation status
  All adults with sickle cell disease should receive

Vaccine	Frequency
Pneumococcal PPV23	Every 5 years
Influenza immunisation	Yearly
Covid immunisation	Currently yearly, though check green book as may change with viral prevalence
Primary Immunisation Hepatitis B (if not received in childhood – routine after 2017)	3 doses in total at 0, 1 and 2 months Booster 12 months

  Note: Patients may have moved as adults from out with the UK. It is therefore important that their previous immunisation history is obtained, and if unclear, that they are referred for 'catch up' vaccinations.

- Social issues (Work, housing, benefit related issues)

- Pregnancy, family planning and contraception
  - For pregnancies, include details including mode of delivery, fetal gestation and outcome
  - If considering pregnancy - Discussion re: partner haemoglobinopathy testing and extended family screening, if not previously done. Consider referral to haem/obs clinic in advance of pregnancy to discuss associated risks.

- Transfusion history
  - Would patient accept transfusion if clinically indicated?
  - Countries where transfused
  - Indications for transfusion
  - Need for regular transfusion programme?
  - If on exchange programme – any issues e.g. access/attendance etc.
  - Any significant red cell antibodies – is patient aware?

- *Details of diagnosis including previous clinicians and hospitals where treated

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- *Country of origin
- Travel plans - ensure appropriate vaccination and malarial prophylaxis advice sought
- Any other issues of concern for patient

Examination

- Weight
- Height
- Blood pressure
- Oxygen saturation
- Cardiovascular examination (right heart strain)
- Respiratory examination
- Abdominal examination (organomegaly)
- Lower limb examination (leg ulcers)

Investigations

- New patients to service/UK
  - *Haemoglobinopathy diagnostic confirmation including genetic testing of alpha and beta genes via Edinburgh genetics lab if desired
  - *G&S and extended RC phenotype (or genotype if recently transfused) – **ensure local blood bank aware of SCD status re. special requirements**
  - *G6PD screening

- Routine Investigations (Checked at least annually)
  - FBC and reticulocytes
  - U&E
  - LFTs including bilirubin
  - LDH
  - Ferritin
  - Microalbuminuria and PCR
  - Vitamin D
  - Virology

Transfusion dependent patients - Manage as per iron chelation guidelines that are available on the SPAH website.

Specialist Investigations

- ECG – if cardiorespiratory symptoms
- ECHO – at baseline and
  - a) every 3 - 5years, if normal and asymptomatic

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    - b) annually if raised tricuspid regurgitant jet velocity but no right heart catheterisation
    - c) if symptomatic or new hypoxia
  - MRI
    - Bone for AVN/osteomyelitis, if indicated
    - Liver +/- cardiac T2*MRI if heavily transfused and ferritin persistently > 1000
  - Other radiology – e.g. DEXA, if pathological fracture
  - Pulmonary function tests – patients with respiratory symptoms or chronic hypoxia
  - Consider Sleep Study – patients with disturbed sleep, excessive daytime somnolence, SpO2 < 95% RA, snoring, priapism, early morning headaches (often done in specialist Haemoglobinopathy centres but evidence limited)

Specialist Referrals

- Consider annual ophthalmology/optometry review for retinopathy screening for a) HbSC patients or b) patients on desferrioxamine/deferasirox
- Recommend local optometrist review for other sickle cell disease patients with referral to ophthalmology if required
- Consider neurology review for patients with chronic headaches and migraines
- Refer to pulmonary hypertension centre if a) raised tricuspid regurgitant jet velocity 250-290 cm/sec and symptoms of pulmonary hypertension or b) raised tricuspid regurgitant jet velocity > 290 cm/sec
- Refer to renal physician if a) acute kidney injury; b) declining renal function (GFR falling > 5ml/min/year or < 60 ml/min); c)ACR/ PCR > 50 mg/mmol
- Refer to urology if a) new onset haematuria or b) problematic priapism
- Refer to hepatology if sickle related liver disease
- Refer to orthopaedics if AVN
- Refer to psychology and/or social work if indicated
- Refer to BSH guideline Management of sickle cell anaemia disease in pregnancy or Local Guidelines

References: Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK (2018) https://www.sicklecellsociety.org/resource/sicklecellstandards/

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