Guidelines for adults

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Background

Sickle Cell Disease is a rare condition in many regions of Europe and general clinicians responsible for local services to patients with SCD may have relatively little experience and knowledge of the condition. Inadequate and inappropriate management may deprive patients of treatments and support shown to be beneficial, and may result in sub-optimal outcomes in the short and long-term. A recent report of fatal outcomes in patients with haemoglobin disorders in the UK (National Confidential Enquiry into Patient Outcome and Death, 2008) has highlighted a number of cases where mortality could have been avoided by adherence to standard guidelines or protocols.

General considerations

Local, national, and international guidelines need to take into account differences in health care systems, accessibility and availability of resources. Obviously guidelines for resource-poor, high prevalence countries in sub-Saharan Africa cannot be the same as those for developed countries. Pan European guidelines may be difficult to implement in individual countries because of the variability in health care systems and differing demographics, health care priorities, and funding arrangements. Local guidelines need to be specifically tailored to the local services and local patient group, and would normally be an adaptation of a national guideline.

The main purposes of guidelines include: Informing and assisting health care workers in delivering accepted standards of care, facilitating management of quality of clinical services (e.g. through use in formulating clinical audit standards and peer review of services). They may also form part of a framework for commissioning of health care services, and guide patients in understanding what management they should be receiving.

Evidence base for SCD guidelines

In the case of SCD, the optimal evidence for clinical interventions, i.e. well designed and conducted randomised controlled trials (RCT’s), is relatively sparse. Below is a list of completed RCT’s which have influenced clinical practice in SCD:

Pneumococcal prophylaxis with oral penicillin

PROPS 1 and 2: Penicillin prophylaxis recommended for children with SCD at least until age 5. (1,2)

Hydroxyurea

MSH, Baby HUG. Hydroxyurea significantly reduces frequency and severity of acute painful crisis and acute chest crisis, and is recommended for adults with a history of frequent crises. (3,4)

Transfusion

Pre-operative transfusion in sickle study group: Top-up transfusion is as effective as exchange in preventing surgical complications and resulted in fewer transfusion-related complications. TAPS: Pre-operative transfusion results in fewer adverse events than no transfusion (J

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Parts of this work were presented at the “3rd Pan-European Conference on Haemoglobinopathies and Rare Anaemias”, Limassol (Cyprus), 24-26 October 2012.
Howard et al., submitted for publication). Pre-operative top-up transfusion is recommended for patients with SCD (HbSS, HbS beta zero thalassaemia) undergoing moderate to high intensity surgical procedures. (5)

**Iron chelation**

Deferasirox is a safe alternative to desferrioxamine and has similar efficacy. (6)

**Pain**

Oral versus i.v. morphine. Oral short and long-acting morphine is as effective as i.v. morphine in managing children with acute painful crisis, once initial i.v. bolus has been given. (7).

**Primary stroke prevention**

STOP 1 and 2. Regular transfusion to maintain Hbs<30% significantly reduces the risk of stroke in children with abnormal transcranial Doppler (TCD) velocities. (8,9) Services are recommended to implement the STOP protocol of regular TCD screening and long-term transfusion of children with SCD (HbSS and HbS beta zero thalassaemia) and abnormal TCD.

**Secondary stroke prevention**

SWiTCH. Switching from transfusion (and chelation) to hydroxyurea (and venesections) is associated with a significantly higher risk of recurrent stroke compared to continuing transfusions. (10) Patients with a history of ischaemia stroke should continue on long-term transfusion.

**Pregnancy**

Regular transfusions decrease the frequency of painful crises, but do not impact on obstetric or perinatal outcomes. Transfusion is not recommended as routine practice during pregnancy. (11)

**Scope of guidelines for SCD**

It is clear that this evidence base does not cover the entirety of SCD care, and comprehensive guidelines need to take into account other published evidence from non-randomised trials, cohort studies, case series and expert opinion. Guidelines should cover the following domains of care:

**Acute and chronic pain management**

Acute complications: Acute Chest syndrome, Biliary problems, priapism, multiorgan failure, sepsis, acute anaemic episodes, ischaemic and haemorrhagic stroke etc.

Chronic complications: Chronic renal insufficiency, pulmonary hypertension, chronic sickle lung syndrome, avascular necrosis of joints and skeleton, retinopathy, lower limb ulceration, liver disease, heart disease etc.


**What is the current status of national/international guidelines for adults with SCD?**

Examples include:
- Europe: ENERCA Workpackage 4 ‘Public Health Issues and management of patients with SCD’, Paediatric guideline has been published and work is progressing on an adult guideline.

**References**

