

**GSTFT Clinical Practice Guideline**

**Management of children newly diagnosed  
with sickle cell disease, or new to the  
service**

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## Organisation and attendance

1. First paediatric haematology clinic appointment within 1 month of referral letter or inpatient admission.

## Diagnosis/investigation at first attendance

2. Haemoglobin phenotype to be confirmed and results filed in notes within 1 month of attendance.
3. Parents' and siblings' haemoglobin phenotype to be determined and recorded by 6 months.
4. Document:
  - (a) HbA<sub>2</sub>%
  - (b) HbF% (repeat and annually until 5 years if elevated, then 5-yearly thereafter)
  - (c) FBC (when well)
  - (d) Reticulocytes (when well)
  - (e) LFTs and U+E/Cr (when well)
  - (f) Parvovirus serology
  - (g) Hepatitis B serology (HBsAg and HbcAb)
  - (h) Hepatitis C for those born abroad
  - (i) ABO RhD blood group and extended RBC phenotype
  - (j) G6PD status

## Clinical History and Examination

5. Complete age-specific proforma at 2, 4, 10 and 14 years.
6. At each visit document:
  - a) Presence or absence of jaundice
  - b) Spleen size
  - c) Height and weight (centile chart annually)

## Prophylaxis

7. Penicillin V prophylaxis to start if <10 years old:

<1 year	62.5 mgs bd
1-5 years	125 mg bd
>5 years	250 mg bd

Erythromycin is the alternative.
8. If >10 years and not previously taking penicillin, discuss benefits and risks.
9. Prevenar to be given <2 years old or >2 years and high risk eg. recurrent chest infections
10. Determine immunization history (including pneumovax) and document in the notes. Pneumovax; re-immunize every 5 years or, if not previously given, immunize in clinic and re-immunize 5-yearly.
11. Hepatitis B: If not immune, immunize from 1 year of age (1<sup>st</sup> and 2<sup>nd</sup> doses 1 month apart, 3<sup>rd</sup> dose 6 months from 1<sup>st</sup> dose. Check HBsAb 1 year later - if levels <100 mIU/ml give 4<sup>th</sup> immunization. Recheck serology 5 yearly and re-immunize if levels <100 mIU/ml.
12. Recommend annual Influenza A vaccine (to be given by GP). For travel to Africa recommend Hepatitis A vaccine (if not immune) and Meningitis A + C vaccine. Advise about the need for malarial prophylaxis if travelling to a malarial area.

## Communication with Professionals

13. GP to be informed of confirmed diagnosis  
Health Visitor/School Nurse to be informed of confirmed diagnosis by six months  
Follow up letters to GP (copy to HV <5, or school nurse >5 years) at least annually.

### Minimum Follow Up

14. < 2 years of age All patients 4x/year for the first 2 years  
(once every 3 months in year 1)
- 2 – 5 years of age HbSS, HbSC, HbS $\beta^0$  and HbS $\beta^+$  thalassaemia – 2x /year
- >5 years Annually

### Non-Attendees

15. If a child does not attend on 2 occasions: parent to be contacted by letter/phone.  
If a child does not attend annually: parents to be contacted by letter/phone.  
Counsellor, HV and GP to be notified if child DNAs twice or if has not attended for annual appointment.

### Minimum Standards for Counsellors

16. Initial contact  
(a) To have contacted the family within 6 weeks of the clinic appointment.  
(b) Initial counseling session in the home setting.  
(c) Literature offered at the first visit
17. Information and education  
(a) Basic information as appropriate covering -  
Genetic inheritance  
Availability of Prenatal diagnosis  
Regime of prophylactic antibiotics and immunization
- (b) Information about the complications and management of the disease covering:  
Fever  
Splenomegaly  
Anaemia  
Painful episodes
18. Haemoglobinopathy card to be given.

### **Continuing Family Contact**

19. Ongoing contact with family (minimum every 6 months)
20. Offer haemoglobinopathy screening for other family members.