

Appendix 5

Clinicians' questionnaire

- Out of 100 SC patients who are **on transfusion**:
 - How many continue transfusions past the age of 18 years for the rest of their lives?
 ___/100 patients
 - Other comments:
- Table 1 presents data that we obtained on the method of transfusion. If you disagree please fill out the last column ('Your opinion').

TABLE 1 Transfusion method

	%	Your opinion
Proportion of patients on simple transfusion	63	
Proportion of patients on exchange transfusion	12	
Proportion of patients on combined transfusion	25	

Can we assume this (or your suggested) distribution for all age groups?

Age group (years)	Do the data apply to this age group?		If no, what proportions of patients receive the following methods of transfusion?		
	Yes	No	Simple	Exchange	Combined
2–7					
8–18					
19–30					
31 +					

- Table 2 presents data that we obtained on hospital admissions for sickle cell patients (**NOT due to stroke**). If you disagree please fill out the last column ('Your opinion').

TABLE 2 Hospital admission (average age 12 years old)

	%	Your opinion
Probability/year of hospital admission <i>on</i> transfusion	2.63	
Probability/year of hospital admission <i>off</i> transfusion	43.9	

The group on which these data were collected had a mean age of 12 years old. Can we assume these probabilities for other age groups (2–7, 19–30, 31+ years)?

Age group (years)	Do the data apply to this age group?		If no, what proportions of patients are hospitalised (<i>not due to stroke/post-stroke complications</i>) when:	
	Yes	No	On transfusion	Off transfusion
2–7				
19–30				
31+				

4. Is there a difference in occurrence of splenic sequestration among sickle cell patients when on or off transfusion?

Yes/no

If yes, out of 100 patients, how many are likely to have splenic sequestration in any year?

- On transfusion: ___ /100 patients
- Off transfusion: ___ /100 patients.

Per patient, how many times/year splenic sequestration occurs when:

- On transfusion: ___ /year
- Off transfusion: ___ /year.

Are there differences in the likelihood of splenic sequestration between age groups?

Yes/no

If yes, please indicate the proportions of different age groups experiencing splenic sequestration when on/off transfusion in the table below:

Age group (years)	Proportion of patients experiencing splenic sequestration:	
	On transfusion	Off transfusion
2–7	___ /100 patients	___ /100 patients
8–18	___ /100 patients	___ /100 patients
19–30	___ /100 patients	___ /100 patients
31+	___ /100 patients	___ /100 patients

Comments:

5. Per 100 transfusions, what proportion of patients become alloimmunised when on simple, exchange or combined transfusion?

TABLE 3 Alloimmunisation: % of sickle cell patients alloimmunised per year

Alloimmunisation when on	%
Simple transfusion	
Exchange transfusion	
Combined transfusion	

6. In Table 4, are the data that we obtained for age groups 2–6 years and 7–18 years on the proportion of sickle cell patients on transfusion who are also on oral and injection chelation? If you disagree please fill out the last column ('Your opinion').

TABLE 4 Chelation types

Chelation types	%	Your opinion
Proportion on oral chelation (deferiprone/Exjade)	10	
Proportion on injection chelation (desferoxamine)	90	

Can we assume this distribution for other age groups?

Age group (years)	Do the data apply to this age group?		If no, what proportions of patients are on:	
	Yes	No	Oral chelation	Injection chelation
19–30				
31+				

7. What is the **non-stroke**-related mortality rate for patients who are **not on transfusion**? Specifically:

- Of 100 patients not on transfusion and whose **TCD scan is <200 cm/second**, how many are likely to die in each year (not due to stroke deaths only)?

___ /100 patients

- Of 100 patients not on transfusion and whose **TCD scan is >200 cm/second**, how many are likely to die in each year (not due to stroke deaths only)?

___ /100 patients.

- How does the mortality rate vary between age groups?

Age group (years)	Annual non-stroke-related deaths among 100 patients who are not on transfusion and whose:	
	TCD scan is < 200 cm/second	TCD scan is > 200 cm/second
2–7	___ /100 patients	___ /100 patients
8–18	___ /100 patients	___ /100 patients
19–30	___ /100 patients	___ /100 patients
31+	___ /100 patients	___ /100 patients

Comments:

8. Annual stroke rate: Out of 100 sickle cell patients **on transfusion**, within a one year period how many would have their first stroke in the 19–30 age group and in the 31+ age group?

Age group (years)	Annual stroke rate among 100 patients who are on transfusion
19–30	___ /100 patients
31+	___ /100 patients

Comments:

9. 16.4% of sickle cell patients aged 7–18 years per annum have a stroke when **off transfusion** if their **TCD is > 200 cm/second**. This figure drops to 2.4% for the same age group, also off transfusion, but with a **TCD of < 200 cm/second**. What are the equivalent proportions for sickle cell patients having a stroke each year for other age groups, off transfusion, and with TCDs of > 200 cm/second and < 200 cm/second?

Age group (years)	Annual stroke rate among 100 patients who are off transfusion and:	
	TCD scan is < 200 cm/second	TCD scan is > 200 cm/second
2–7	___ /100 patients	___ /100 patients
8–18	2.4/100 patients	16.4/100 patients
19–30	___ /100 patients	___ /100 patients
31+	___ /100 patients	___ /100 patients

Comments:

10. Table 5 provides data on the outcome of stroke for the 7–18 age group. If you disagree please fill out the last column ('Your opinion').

For the purpose of this study, stroke outcomes have been defined as follows:

- *mild* minor health impact (minor stroke)
- *moderate* temporary disability, some complications post stroke (minor stroke)
- *severe* permanent disability, severe complications post stroke (major stroke).

Note: Definitions of states: <http://stroke.ahajournals.org/cgi/content/short/32/6/1425>

TABLE 5 One-year stroke outcomes, 7- to 18-year-olds

Outcome post first stroke	% of patients	Your opinion
Mild	18	
Moderate	45	
Severe	36	
Death	0	

Given the above estimates for stroke outcomes for 7- to 18-year-olds, what outcomes would be expected for other age groups?

Age group (years)	Outcome, 1 year post stroke (%)			
	Mild	Moderate	Severe	Death
2-7				
8-18	18	45	36	0
19-30				
31+				