

TABLE 51 Responses to clinicians' questionnaire

Question reference	Questions	Subquestion	Original value, if applicable	Responses (%)				Average (as %)	
				R1	R2	R3	R4		
1	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?		NA	20	90	90	100	75	
2	<i>Table 1</i> presents data that we obtained on the method of transfusion. Do you agree with these data? If you disagree please fill out the last column (Your opinion) in <i>Table 1</i>	Proportion of patients on simple transfusion	63	NA	10	63	67	47	
		Proportion of patients on exchange transfusion	12	NA	90	12	33	45	
		Proportion of patients on combined transfusion	25	NA	0	25	0	8	
2a	Do the data set out in question 2 apply to this age group? If no, what proportions of patients receive the following methods of transfusion?	2–7 years	NA	No	No	No	No	No	
		8–18 years	NA	No	No	Yes	Yes	No/yes	
		19–30 years	NA	Yes	Yes	Yes	50/50	Yes	
		31+ years	NA	Yes	Yes	Yes	NA	Yes	
		Age 2–7 years	Simple transfusion	63	100	100	90	100	98
			Exchange transfusion	12	0	0	0	0	0
			Combined transfusion	25	0	0	10	0	3
		Age 8–18 years	Simple transfusion	63	65	80	0.63	0.67	69
			Exchange transfusion	12	15	20	0.12	0.33	20
			Combined transfusion	25	20	0	0.25	0	11
		Age 19–30 years	Simple transfusion	63	0.63	0.1	0.63	0.67	51
			Exchange transfusion	12	0.12	0.9	0.12	0.33	37
			Combined transfusion	25	0.25	0	0.25	0	13
		Age 31+ years	Simple transfusion	63	0.63	0.1	0.63	0.63	50
			Exchange transfusion	12	0.12	0.9	0.12	0.12	32
Combined transfusion	25		0.25	0	0.25	0.25	19		
3	<i>Table 2</i> presents data that we obtained on hospital admissions for sickle cell patients (<i>not</i> due to stroke/post-stroke complications). Do you agree with these data? If you disagree please fill out the last column (Your opinion) in <i>Table 2</i>	Probability per year of hospital admission on transfusion	3	10	2.63	2.63	1	4	
		Probability per year of hospital admission off transfusion	44	43.90	20	43.90	50	39	

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3a	The group on which these data were collected had a mean age of 12 years. Can we assume these probabilities for other age groups (2–7, 19–30, 31+ years)?	2–7 years	NA	No	Not sure	Yes	NA	No/yes		
		8–18 years	NA	No	No	Yes	NA	No		
		19–30	NA	No	No	Yes	NA	No		
		31+ years	NA	No	No	Yes	NA	No		
		Age 2–7 years	On transfusion	3	NA	NA	2.63	1.50	2	
			Off transfusion	44	NA	NA	43.90	50	47	
		Age 8–18 years	On transfusion	3	2.63	2.63	2.63	1.50	2	
			Off transfusion	44	43.90	43.90	43.90	43.90	44	
		Age 19–30 years	On transfusion	3	NA	2.50	2.63	1.50	2	
			Off transfusion	44	NA	20	43.90	50	38	
		Age 31+	On transfusion	3	NA	2.50	2.63	NA	3	
			Off transfusion	44	NA	20	43.90	NA	32	
		4	Is there a difference in occurrence of splenic sequestration among sickle cell patients when on or off transfusion?	Yes	NA	NA	NA	1	1	100
				No	NA	NA	NA	0	0	0
If yes, out of 100 patients, how many are likely to have splenic sequestration in any year?	On transfusion		NA	0	NA	NA	0	0		
	Off transfusion		NA	3	NA	NA	3.5	3		
Per patient, how many times per year does splenic sequestration occur when:	On transfusion		NA	0	NA	NA	0	0		
	Off transfusion		NA	NA	NA	NA	NA	NA		
Are there differences in the likelihood of splenic sequestration between age groups?	Yes		NA	1	NA	1	1	100		
	No		NA	0	NA	0	0	0		
Age 2–7 years	On transfusion		NA	0	NA	5	0	2		
	Off transfusion		NA	1.60	NA	0	5.50	2		
Age 8–18 years	On transfusion		NA	0	NA	0	0	0		
	Off transfusion		NA	0.50	NA	0	1.50	1		
Age 19–30 years	On transfusion		NA	0	NA	0	0	0		
	Off transfusion		NA	0.50	NA	0	0	0		
Age 31+ years	On transfusion		NA	0	NA	0	0	0		
	Off transfusion		NA	0.50	NA	0	0	0		

continued

TABLE 51 Responses to clinicians' questionnaire (*continued*)

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5	Per 100 transfusions, what proportion of patients become alloimmunised when on simple, exchange or combined transfusion?	Proportion of patients on:	Simple transfusion	NA	5	NA	5	2	4
			Exchange transfusion	NA	10	NA	15	5.50	10
			Combined transfusion	NA	NA	NA	10	NA	10
6	<i>Table 4</i> shows the proportions of transfused sickle cell patients who are treated with oral and injection chelation. These proportions are applicable to the age groups 2–6 years and 7–18 years. Do you agree with these figures? If you disagree please fill out the last column (Your opinion) in <i>Table 4</i>	Age 2–7 years	Proportion on oral chelation (deferiprone/ Exjade)	10	50	90	90	90	80
			Proportion on injection chelation (deferoxamine)	90	50	10	10	10	20
		Age 8–18 years	Proportion on oral chelation (deferiprone/ Exjade)	NA	50	90	90	90	80
			Proportion on injection chelation (deferoxamine)	NA	50	10	10	10	20
		Age 19–30 years	Proportion on oral chelation (deferiprone/ Exjade)	NA	50	95	90	NA	78
			Proportion on injection chelation (deferoxamine)	NA	37.50	5	10	NA	18
		Age 31+ years	Proportion on oral chelation (deferiprone/ Exjade)	NA	20	95	90	NA	68
Proportion on injection chelation (deferoxamine)	NA		50	5	10	NA	22		

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7	What is the non-stroke-related mortality rate for patients who are not on transfusion? Of 100 patients not on transfusion and whose TCD scan is > 200 cm/second, how many are likely to die in each year (excluding stroke deaths)?		NA	0.27	NA	NA	0.50	0	
			NA	NA	NA	NA	0.50	1	
	How does the mortality rate vary between age groups?	Age 2–7 years	TCD scan is < 200 cm/second	NA	NA	NA	NA	NA	–
			TCD scan is > 200 cm/second	NA	NA	NA	NA	NA	–
		Age 8–18 years	TCD scan is < 200 cm/second	NA	NA	NA	NA	NA	–
			TCD scan is > 200 cm/second	NA	NA	NA	NA	NA	–
		Age 19–30 years	TCD scan is < 200 cm/second	NA	NA	NA	NA	NA	–
			TCD scan is > 200 cm/second	NA	NA	NA	NA	NA	–
		Age 31+ years	TCD scan is < 200 cm/second	NA	NA	NA	NA	NA	–
			TCD scan is > 200 cm/second	NA	NA	NA	NA	NA	–
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 years age groups and in the 31+ years age group?	19–30 years	NA	0.50	NA	2	NA	1	
		31+ years	NA	0.50	NA	2	NA	1	

continued

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9	16.4 of SC patients aged 8–18 years per annum have a stroke when off transfusion and 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 2–7 years	TCD scan is < 200 cm/second	NA	NA	NA	NA	1	1
			TCD scan is > 200 cm/second	NA	NA	NA	NA	10	10
		Age 8–18 years	TCD scan is < 200 cm/second	NA	2.40	NA	2.40	2.40	2
			TCD scan is > 200 cm/second	NA	16.40	NA	16.40	16.40	16
		Age 19–30 years	TCD scan is < 200 cm/second	NA	NA	NA	NA	2.40	2
			TCD scan is > 200 cm/second	NA	NA	NA	NA	16.40	16
		Age 31+ years	TCD scan is < 200 cm/second	NA	NA	NA	NA	2.40	2
			TCD scan is > 200 cm/second	NA	NA	NA	NA	16.40	16

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10	<i>Table 5</i> provides data on the outcome of stroke for the 8–18 years age groups. Do you agree with these? If you disagree please fill out the last column (Your opinion) in <i>Table 5</i>	Mild	18	NA	25	0.18	20	21	
		Moderate	45	NA	45	0.45	70	53	
		Severe	36	NA	25	0.36	10	24	
		Death	0	NA	5	0	0	2	
		Age 2–7 years	Mild	NA	30	NA	25	NA	28
			Moderate	NA	50	NA	50	NA	50
			Severe	NA	20	NA	25	NA	23
			Death	NA	0	NA	0	NA	0
		Age 8–18 years	Mild	18	25	18	18	18	20
			Moderate	45	50	45	45	45	46
			Severe	36	25	36	36	36	33
			Death	0	0	0	0	0	0
		Age 19–30 years	Mild	NA	15	30	15	20	20
			Moderate	NA	40	40	30	50	40
			Severe	NA	25	25	50	28	32
			Death	NA	20	5	5	2	8
		Age 31+ years	Mild	NA	25	NA	10	NA	18
Moderate	NA		30	NA	20	NA	25		
Severe	NA		30	NA	60	NA	45		
Death	NA		15	NA	10	NA	13		

NA, not applicable.