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## **Outcome of children with sickle cell disease admitted to intensive care - a single institution experience**

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## Outcome of children with sickle cell disease admitted to intensive care - a single institution experience



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3 Abstract  
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6 We retrospectively audited children with sickle cell disease admitted to paediatric  
7 intensive care (PICU) at King's College Hospital between January 2000 and  
8 December 2008. 46 children with SCD were admitted, on 49 separate occasions. Ages  
9 ranged from 4 months to 15 years (median 7.6 years). Three children died in PICU,  
10 however two presented to hospital in cardiorespiratory arrest; overall mortality was  
11 6%. The most common reason for admission was acute chest syndrome (43%). 88%  
12 of admissions required blood transfusion, of which 74% had exchange blood  
13 transfusions. The mortality among children with SCD admitted to PICU is low.  
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27 Keywords  
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29 Sickle cell disease. Children. Paediatric intensive care unit.  
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8 Outcome of children with sickle cell disease admitted to intensive care - a single  
9 institution experience  
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12 Introduction

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15 Sickle cell disease (SCD) is one of the commonest severe genetic disorders in  
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17 England, affecting over 1 in 2000 live births (Streetly *et al*, 2009). Children with SCD  
18  
19 can become critically ill requiring referral to intensive care units. The most common  
20  
21 causes of death in children with SCD are acute, and include infection, acute chest  
22  
23 syndrome (ACS) and stroke (Manci *et al*, 2003; Leikin *et al*, 1989). To our  
24  
25 knowledge, there are no published data on the outcome of children with SCD  
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27 admitted to PICU.  
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34 Population based data on the epidemiology of all children admitted to PICU in the UK  
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36 are reported to the Paediatric Intensive Care Audit Network (PICANet) (Paediatric  
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38 Intensive Care Audit, 2008), established in 2002. However, there is little published  
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40 experience about children with SCD admitted to PICU. There have been several  
41  
42 studies reporting outcomes for children with other conditions admitted to PICU with  
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44 mortality rates of 44% and 38% for children with bone marrow transplants (Jacobe *et*  
45  
46 *al*, 2003) and HIV (Cooper *et al*, 2004) respectively. We audited the admission of  
47  
48 children with SCD admitted to PICU at King's College Hospital (KCH) over 9 years,  
49  
50 to define the reasons for admission and outcome for this group, and to make  
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52 comparisons with all children admitted to PICU.  
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## Methods

We retrospectively audited all children with SCD admitted to our PICU between 1<sup>st</sup> January 2000 and 31<sup>st</sup> December 2008. KCH looks after about 400 children with SCD. We included all patients admitted to PICU with SCD from birth to 16 years. Information collected included presentation, treatment and outcome; outcome data included reason for and length of admission, the need for respiratory and inotropic support, use of blood transfusion and antibiotics, time to hospital discharge and mortality. Data were collected from PICU admission books, patient case notes and electronic records.

## Results

### *Patient Demographics*

Forty-six children with SCD were admitted to PICU 49 times (3 HbSC, 46 HbSS), accounting for 2% PICU admissions over this 9 year period (Table 1). Approximately 1000 children with SCD live in the area served by KCH PICU, giving an admission rate of approximately 0.5/100 patient years.

### *Reasons for Admission*

The most frequent reasons for admission to PICU were ACS (43%, n=21) and stroke (24%, n=12), including three subarachnoid haemorrhages (SAH) (Table 1). Four (8%) elective postoperative admission were planned due to a history of complications. Acute surgical admissions comprised two patients with acute hydrocephalus (1 immediately post stent and coil embolisation for cerebral aneurysm; 1 with a brain tumour) and 1 patient with slipped upper femoral epiphysis for central venous access and urgent exchange transfusion pre operatively. One patient was admitted with upper

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3 airway obstruction due to massively enlarged tonsils. The patients with other  
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5 neurological problems include two with isolated seizures, one with pneumococcal  
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7 meningitis, and one with cerebrovascular disease for urgent exchange transfusion.  
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10 Two patients with acute sepsis presented to hospital in cardiac arrest and rapidly  
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12 developed multi-organ failure.  
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#### 15 16 17 18 *Length of admission pre admission to PICU*

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20 Twenty-eight percent (n=13) patient were admitted directly from the Emergency  
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22 department to PICU. The majority of patients were admitted to normal hospital wards  
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24 for at least 24 hours (with 22% being admitted for more than 4 days) before requiring  
25  
26 admission to PICU. The majority of patients with CVA presented with acute  
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28 neurological symptoms and were transferred directly to PICU on day 0, whereas  
29  
30 typically patients with ACS were in hospital for at least 24 hours before being  
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32 transferred to PICU.  
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#### 36 37 38 39 *Respiratory support*

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41 Thirty-five percent (n=17) required respiratory support, 1 following CVA, 2 with  
42  
43 multiorgan failure and 14 with ACS. The median length of respiratory support  
44  
45 (including non invasive ventilation) was two days (range 1-18 days). High frequency  
46  
47 oscillatory ventilation (HFOV) was used in seven admissions (14%). Five of these  
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49 admissions were for ACS and two had multi-organ failure secondary to sepsis. Five  
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51 (10%) were treated with inhaled nitric oxide (iNO); all had ACS and survived to at  
52  
53 least 6 months follow-up (Table 2).  
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#### 57 58 59 60 *Inotropic support*

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3 Sixteen percent (n=8) patients required inotropic support: 3 ACS, 2 multiorgan  
4 failure, 1 cerebral aneurysm, 1 pneumococcal meningitis, 1 transient aplasia. Three of  
5 these died (both multiorgan failure patients and the child with the cerebral aneurysm).  
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9 The five remaining patients survived to at least six months follow-up.  
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### 12 13 14 15 *Blood Transfusion*

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17 Eighty-eight percent (n=43) required blood transfusion on PICU, of which 74%  
18 (n=32) had exchange transfusions. Exchange blood transfusion was most commonly  
19 performed for ACS (35%, n=17), which accounted for 80% of transfusions in those  
20 patients with ACS; the remaining patients with ACS all had simple transfusion. 10 out  
21 of the 12 patients with CVA had exchange transfusion, and the other two were already  
22 on a transfusion programme, with satisfactory parameters at the time of admission.  
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### 34 *Antibiotics and lines*

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36 Eighty-two percent (n=40) were prescribed antibiotics; all had an intravenous  
37 cephalosporins. 61%, (n=30) also had a macrolide. All admissions had peripheral  
38 venous access. 30 % (n=15) had central venous catheters and 71% (n=35) had arterial  
39 lines inserted.  
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### 50 *Length of stay, treatment related morbidity and discharge outcome*

51 The median length of stay was 1 day (range 1-18 days). There was one central line  
52 associated thromboembolism, one delayed transfusion reaction and one death was due  
53 to complications post neurosurgical procedure. Three patients (6%) died on PICU,  
54 although two of these were transferred straight from the Accident and Emergency  
55 department, having presented in cardiorespiratory arrest. Median time to discharge  
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3 from hospital after discharge from PICU was 4 days (range 0 - 95 days). There were  
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5 no further deaths before discharge from hospital; one death occurred 4 months post  
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7 discharge due to complication of a brain tumour, unrelated to SCD.  
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#### 10 11 12 *Comparison with PICANet Data*

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15 Comparison of our data with that from all PICU admissions in the UK (2005-2007)  
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17 showed that children with SCD were more significantly less likely to require  
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19 ventilatory or inotropic support, and had a similar mortality rate (Table 3).  
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#### 24 Discussion

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27 This is the first study to characterise the use of PICU by children with SCD. ACS and  
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29 CVA were the commonest reasons for admission. Three patients died during their  
30  
31 admission to PICU. One death was post-stent and coil embolisation for cerebral  
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33 aneurysm causing a SAH, a procedure which has a recognised mortality rate (Kurre &  
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35 Berkefeld, 2008). The other two deaths were as a result of multi-organ failure  
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37 secondary to presumed sepsis. Both of these children were less than 2.5 years old,  
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39 presented directly to the emergency unit in cardiorespiratory arrest, and died within  
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41 24 hours of admission. A study by Schindler looking at out-of-hospital paediatric  
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43 cardiac arrest found that survival to hospital discharge in children requiring CPR in  
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45 the emergency department was low at 15% (Schindler *et al*, 1996).  
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53 ACS was the most common reason for admission. There were no deaths in this  
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55 group. The outcome for ventilated patients with ACS was very good, even those  
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57 requiring maximal support with HFOV and iNO. Our study confirms previous reports  
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59 that ACS is the leading cause of admission to PICU in SCD (Platt *et al*, 1994). CVA  
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3 was the second commonest reason for admission to PICU. All CVA patients received  
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5 blood transfusion, with 83% having exchange transfusions (Vichinsky *et al*, 2000);  
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7 those not exchanged were already on a transfusion programme with low HbS levels.  
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9 In young children, exchange transfusion is particularly difficult and is often facilitated  
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11 by the use of an arterial line for blood removal, which necessitates admission to  
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13 PICU. PICUs are also useful in the care of children with acute neurological symptoms  
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15 in that close observation and neuroprotection are easier than on a general ward.  
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22 One child presented with severe hypovolaemic shock secondary to acute anaemia,  
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24 with a haemoglobin of 1.7 g/dl; the reticulocyte count was low initially but repeated  
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26 Parvovirus B19 serology and nucleic acid testing were negative. The patient required  
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28 full PICU support but made a good recovery. Only three children were admitted with  
29  
30 septicaemia, and in only one case was pneumococcus isolated. Other studies in the  
31  
32 United Kingdom have also observed lower than expected rates of pneumococcal  
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34 sepsis in patients with SCD (Telfer *et al*, 2007), suggesting the benefit of prophylactic  
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36 antibiotics and vaccination.  
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43 Fifteen (33%) of the patients admitted to PICU had a history of either ACS or CVA.  
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45 Thirty-eight percent of patients admitted to PICU with ACS had a previous history of  
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47 ACS and 63% of those previously required admission to PICU. Three patients with  
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49 CVA had previously had CVA; 2 of these patients were not on regular transfusion  
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51 programme as secondary stroke prevention; following prolonged discussions and  
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53 counselling, one family had decided to stop regular transfusions and the other had  
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55 started hydroxyurea. This gives further support to the use of transfusion to prevent  
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3 recurrent stroke and demonstrates that discontinuing transfusion increases the risk of  
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5 further strokes (Ohene-Frempong *et al*, 1998; Pegelow *et al*, 1995).  
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10 Mortality was similar in the SCD patients to that for all patients admitted to PICU  
11 (Table 3), although children with SCD were significantly less likely to need  
12 ventilatory or inotropic support. The most common reasons for admission in the  
13 PICANet data were: ventricular septal defect 8%, respiratory failure 6.5%, status  
14 epilepticus 5.5%, sepsis 4.8%, pneumonia 4.1%, asthma 2% and intracranial tumour  
15 2%. Although the reasons for admission were different, length of stay was similar.  
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17 Limited inferences can be made from our audit because it is a single-centre,  
18 retrospective study involving relatively small numbers, although outcome appears to  
19 be equivalent to unselected PICU admissions.  
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5 Cooper, S., Lyall, H., Walters, S., Tudor-Williams, G., Habibi, P., de Munter, C.,  
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7 Britto, J. & Nadel, S. (2004) Children with human immuno deficiency virus admitted  
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9 to a paediatric intensive care unit in the United Kingdom over 10-year period.  
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11 *Intensive Care Medicine*, 30, 113-118.  
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Jacobe, S.J., Hassan, A., Veys, P., & Mok, Q. (2003) Outcome of children requiring admission to an intensive care unit after bone marrow transplant. *Critical Care Medicine*, 31, 1299-1305.

Kurre, W. & Berkefeld, J. (2008) Materials and techniques for coiling of cerebral aneurysms: how much scientific evidence do we have? *Neuroradiology*, 50, 909-927.

Leikin, S.L., Gallagher, D., Kinney, T.R., Sloane, D., Klug, P., & Rida, W. (1989) Mortality in children and adolescents with sickle cell disease. *Pediatrics*, 84, 500-508.

Manci, E.A., Culberson, D.E., Yang, Y.M., Gardner, T.M., Powell, R., Hyanes, J. Jr, Shah, A.K., & Mankad, V.N. (2003) Causes of death in sickle cell disease: an autopsy study. *British Journal of Haematology*, 123, 359-365.

Ohene-Frempong, K., Weiner, S.J., Sleeper, L.A., Miller, S.T., Embury, S., Moohr, J.W., Wethers, D.L., Pegelow, C.H. & Gill, F.M. (1998) Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood*, 91, 288-294.

1  
2  
3  
4  
5  
6 Paediatric Intensive Care Audit Network (PICANET) coordinated by the Universities  
7  
8 of Leeds and Leicester. Fifth PICANET National Report (2008).

9  
10 <http://www.picanet.org.uk/documentation.html>  
11  
12

13  
14  
15  
16 Platt, O.S., Brambilla, D.J., Rosse, W.F., Milner, P.F., Castro, O., Steinberg, M.H. &  
17  
18 Klug, P.P. (1994) Mortality in sickle cell disease: life expectancy and risk factors for  
19  
20 early death. *New England Journal of Medicine*, 330, 1639-1644.  
21  
22

23  
24  
25  
26  
27  
28 Pegelow, C.H., Adams, R.J., McKie, V., Abboud, M., Berman, B., Miller, S.T.,  
29  
30 Olivieri, N., Vinchinsky, E., Wang, W. & Brambilla, D. (1995) Risk of recurrent  
31  
32 stroke in patients with sickle cell disease treated with erythrocyte transfusions.  
33  
34 *Journal of Pediatrics*, 126, 896-899.  
35  
36

37  
38  
39  
40  
41  
42 Schindler, M.B., Bohn, D., Cox, P.N., McCrindle, B.W., Jarvis, A., Edmonds, J. &  
43  
44 Barker, G. (1996) Outcome of out-of-hospital cardiac or respiratory arrest in  
45  
46 children. *New England Journal of Medicine*, 335, 1473-1479.  
47  
48  
49

50  
51  
52  
53  
54 Streetly, A., Latinovic, R., Hall, K. & Henthorn, J. (2009) Implementation of  
55  
56 universal newborn bloodspot screening for sickle cell disease and other clinically  
57  
58 significant haemoglobinopathies in England: screening results for 2005-7. *Journal of*  
59  
60 *Clinical Pathology*, 62, 26-30.

1  
2  
3  
4  
5  
6 Telfer, P., Coen, P., Chakravorty, S., Wilkey, O., Evans, J., Newell, H., Smalling, B.,  
7  
8 Amos, R., Stephens, A., Rogers, D. & Kirkham, F. (2007) Clinical outcomes in  
9  
10 children with sickle cell disease living in England: a neonatal cohort in East London.  
11  
12 *Haematologica*, 92, 905-912.

13  
14  
15  
16 Vichinsky, E.P., Neumayr, L.D., Earles, A.N., Williams, R., Lennette, E.T., Dean,  
17  
18 D., Nickerson, B., Orringer, E., McKie, V., Bellevue, R., Daeschner, C. & Mancini,  
19  
20 E.A. (2000) Causes and outcomes of the acute chest syndrome in sickle cell disease.  
21  
22 *New England Journal of Medicine*, 342, 1855-1865.  
23  
24  
25  
26  
27  
28  
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Genotype	HbSS 43, HbSC3
Median age (range)	7.6 years (0.4-15.7)
Hydroxyurea	2 (4%)
History of previous ACS	10 (22%)
Prescribed penicillin prophylaxis	46 (100%)
History of previous CVA	5 (11%)
Receiving regular blood transfusions	1 (2%)
Previous admission to PICU	5 (11%)
Transferred from outside hospital	13 (28%)

ACS acute chest syndrome; CVA cerebrovascular accident

Table 1: Demographics of patients admitted to ITU (n=46)

Reason for Admission	No. (%) of Total Admissions (n=49)	Ventilatory support (% of Admission Group)	Survived to PICU discharge (% of Admission Group)
ACS	21 (43)	6 (29)	21 (100)
CVA	12 (24)	2 (17)	11 (92)
Overwhelming sepsis	2 (4)	2 (100)	0 (0)
Other neurological	4 (8)	2 (50)	3 (75)
Elective post operative	4 (8)	1 (25)	4 (100)
Acute surgical	3 (6)	2 (67)	2 (100)
Upper airways obstruction	1 (2)	1 (100)	1 (100)
Pain control	1 (2)	0 (0)	1 (100)
Aplasia unknown cause	1 (2)	1 (100)	1 (100)
Total	49 (100)	17 (35)	46 (94)

PICU paediatric intensive care unit; ACS acute chest syndrome; CVA cerebrovascular accident

Table 2: Reason for admission to paediatric intensive care, need for ventilatory support, and outcome

	PICANet 2005-2007 n=43841	SCD cohort KCH 2000-2008 n=49	P
% needing non-invasive ventilation	13%	2%	0.039
% needing invasive ventilation	66%	33%	<0.0001
% needing inotropes	31%	16%	0.039
Mortality	5%	6%	0.97
Length of stay (days)	32% stayed 1-3 days	median 1; mean 2	

PICANet paediatric intensive care audit network; SCD sickle cell disease; KCH King's College Hospital NHS Foundation trust

Table 3 Comparison of our experience and with PICANet data. P-values calculated using Chi-squared test with Yates' correction.