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SCIENTIFIC LETTERS

Sickle cell disease in pediatric intensive care☆

Enfermedad de células falciformes en cuidados intensivos pediátricos

To the editor:

Sickle cell disease (SCD) is a structural haemoglobinopathy with an autosomal codominant pattern of inheritance[1] characterised by the presence of sickle haemoglobin (HbS) in red blood cells. In 2017, 826 affected patients were reported to the Spanish Register of Haemoglobinopathies and Rare Anaemias. In Spain, there has been a recent increase in the incidence of SCD on account of immigration. This disease can result in severe complications requiring admission to a paediatric intensive care unit (PICU), and few studies have analysed these complications.[2–5]

We conducted a retrospective descriptive study with the aim of describing the characteristics and outcomes of children with SCD who required admission to the PICU.

We collected data on demographic characteristics (age, sex, racial/ethnic background), phenotype, reason for admission, laboratory values (HbS, C-reactive protein [CRP], procalcitonin [PCT]), treatment, complications and mortality in the patients with SCD admitted to the PICU between January 2017 and January 2020. Patients with SCD amounted to 0.8% of the total admissions of a 14-bed tertiary care PICU in a referral hospital that manages SCD in the region.

The study included 11 patients with SCD (8 of African descent, 2 of Latin American descent and 1 of Asian descent), 8 were male, and the median age was 9.8 years (interquartile range [RIC], 8.2 years). Ten had the HbSS phenotype, 1 the HbSC phenotype, and 2 had previously undergone haematopoietic stem cell transplantation.

Table 1 summarises the reason for admission to the PICU. Four patients (36.4%) had developed acute chest syndrome (ACS), 2 posterior reversible encephalopathy syndrome (PRES) and 1 stroke involving the right middle cerebral artery. The median score in the Paediatric Risk of Mortality (PRISM) III scale at admission was 4 (IQR, 5). During the stay in the PICU, 2 patients with ACS developed other complications (1 a vaso-occlusive crisis and 1 multiple organ failure and PRES). Six patients required respiratory support (in 2 cases, invasive mechanical ventilation due to decreased level of consciousness) All 4 patients with ACS required respiratory support: 1 invasive mechanical ventilation and 3 non-invasive ventilation. Nine patients required transfusion therapy (3 exchange transfusion and 6 erythrocytapheresis) due to a percentage of HbS greater than 30%. Table 2 presents the haematological outcomes of treatment. None of the patients died. The median length of stay in the PICU was 2 days (IQR, 6 days). Few studies have analysed the characteristics of children with SCD who require admission to the PICU.[2] The predominant phenotype in our patients was HbSS, which is the most frequent and severe.[1] The most frequent reason for admission was AST, in agreement with other studies.[2,3] We ought to highlight that there are patients who develop more than one complication, as was the case in 2 of our patients.

Table 1 Reason for admission to the paediatric intensive care unit.

Reason for admission	n (%)
Acute chest syndrome	4 (36.4%)
Vaso-occlusive crisis	2 (18.2%)
Altered level of consciousness	2 (18.2%)
Establishment of central venous access for exchange transfusion or erythrocytapheresis	2 (18.2%)
Seizures	1 (9.1%)

Only one patient with ACS required invasive ventilation[4] and the other 3 had favourable outcomes with non-invasive ventilation, which may be initiated early to prevent progression to respiratory failure.

Some studies have reported a frequency of recurrence of ACS of up to 80%.[2] In our study, 3 of the 4 patients with ACS had had previous episodes of it. This disease is the leading cause of death in children or young adults with SCD.[4]

Ischaemic attacks in children with SCD most frequently result from occlusion of the middle cerebral artery, as was the case in our patient. This is an important reason for admission that is also associated with mortality and sequelae.[5] Three patients had PRES, a complication previously described in the context of SCD. It manifests with high blood pressure, altered level of consciousness, seizures and visual disturbances, and it is reversible in most patients once blood pressure is controlled.[6]

Infectious complications are one of the causes of death in the PICU described in other case series.[2] However, none of our patients was admitted due to infection.

In recent years, the use of automated erythrocytapheresis has increased, as it is quicker and achieves lower percentages of HbS compared to exchange transfusion.[1]

Despite its severity, the mortality in children with SCD that require admission to the PICU is very low[2]; none of the patients in our series died.

Our study has the limitations characteristic of a retrospective and descriptive study conducted in a single centre and with a small sample size, but it reviews the most frequent complications that develop in children with SCD.

In conclusion, ACS and neurologic manifestations were the most frequent reasons for admission to the PICU in children with SCD. The mortality is low, but the morbidity is significant, so we recommend centralising the care of this disease in specialised referral hospitals.

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Table 2 Changes in haemoglobin values.

Patient	Transfusion therapy	Hb at admission (mg/dL)	Pre-transfusion HbS (%)	Post-transfusion HbS (%)
1	Not needed	14.1		
2	Not needed	13.8		
3	Manual exchange transfusion	8.9	53.3%	19%
4	Manual exchange transfusion	8.8	60%	22.5%
5	Erythrocytapheresis	10.6	30.6%	13.5%
6	Erythrocytapheresis	7.4	57.8%	12.9%
7	Erythrocytapheresis	8.8	43.2%	11.6%
8	Erythrocytapheresis	9.9	72.8%	9.5%
9	Erythrocytapheresis	10	Unknown	Unknown
10	Erythrocytapheresis	9.6	66%	26%
11	Manual exchange transfusion	9	64.9%	29.7%

Hb, haemoglobin; HbS, sickle haemoglobin.

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