

Routine hemoglobin electrophoresis for pediatric surgery day case in at-risk populations: Suggested algorithm for screening using clinical risk factors

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Abstract

Background: Hemoglobin electrophoresis (HBE) is a part of the preoperative routine requested by anesthetists. However, the prevalence of hemoglobinopathy in the population is low. This study aims to determine the clinical risk factors for hemoglobinopathies and propose clinical guidelines for preoperative screening of patients for pediatric day care surgery.

Patients and Methods: A prospective study carried out over 12 months. Consecutive patients aged 6 months and older who had day case surgery were recruited to the study. Biodata and relevant clinical data were collated and documented in a proforma and analyzed using a statistical package for social sciences version 17.

Results: There were 124 patients 106 boys and 18 girls. The median age was 3 years. Scrotal lesions were the most common conditions managed (71.7%). Seventy-eight percent of patients had HbAA, 15.3% had HbAS, and 4.8% had HbAC while 0.8% each had both HbSC and HbSS. At least one parent of 78.2% knew their Hb phenotype, of which, 79% were HbAA. A history of jaundice ($P = 0.0001$), hand and foot syndrome ($P = 0.0001$), frontal bossing ($P = 0.0001$), and low packed cell volume at surgery ($P = 0.001$) were found significant in predicting hemoglobinopathies. There was no mortality.

Conclusion: Risk factors for hemoglobinopathies from this study included a positive history of jaundice, hand and foot syndrome, frontal bossing, and anemia. Proposed guidelines for HBE screening include the presence of hemoglobinopathy in one parent if one parent has sickle cell trait, and the other parent's genotype is unknown or if any of the risk factors is present.

Key words: Algorithm, day case surgery, hemoglobin electrophoresis screening

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Introduction

Day case surgery constitutes about 40% of the pediatric surgical workload in our center. Preoperative workup includes packed

cell volume (PCV), hemoglobin electrophoresis (HBE), and electrolytes/urea/creatinine.^[1,2] HBE is included in this subregion because sickle cell anemia is endemic and catastrophic complications such as chest syndrome, stroke, and death may follow inadequate preoperative evaluation of such patients.^[3,4]

The reported perioperative mortality rates are as high as 10%, and the rate of postoperative complications is up to

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50%.^[5] Such complications may be secondary to perioperative hypoxia, hypoperfusion, and acidosis, which cause erythrocytes to sickle, thus precipitating vaso-occlusion and organ dysfunction.^[6] Accordingly, the attending anesthetist needs to be aware of sickle cell patients scheduled for surgery and ensure that they have been adequately prepared.

Despite these challenges, the prevalence of sickle cell anemia in the subregion is <2%, and in a study by Hyder *et al.*,^[4] only 3.6% of children who had undergone surgical procedures had sickle cell disease.^[7] Our earlier study on patients undergoing herniotomy in our center revealed a prevalence of 1.1% for patients with sickle cell anemia.^[8] Due to the low prevalence of HbSS, relatively high cost of HBE set against the background of pervasive poverty and improved anesthetic services, routine HBE seems unnecessary before day case surgery in the pediatric population in our region. This study proposes an algorithm for the preoperative clinical screening of HBE in pediatric patients scheduled for day care surgery.

Patients and Methods

A prospective, observational study over 12 months (February 2012–January 2013) period in children aged 6 months to 15 years scheduled for day case surgery under general anesthesia. Data collected included demographics, previous medical history of jaundice, hand and foot syndrome, frontal bossing, unexplained abdominal pain with hospital admission, history of previous blood transfusion, and family history of sickle cell disease. In addition, the Hb phenotype of parents, PCV at surgery, patient's HBE status, surgical outcome and duration of stay were documented.

Data are expressed as median and range, mean ± standard deviation, absolute numbers, or percentage, whereas the Pearson's Chi-square was used to determine the association between significant previous history suggestive of sickle cell anemia and HBE. *A P* < 0.05 was considered significant for all tests. All analyses were performed using the Statistical Package for Social Sciences for Windows version 17 (SPSS, Chicago, IL, USA).

Results

One hundred and twenty-four children aged 6 months to 15 years (median age = 3 years) were studied. There were 106 (85.48%) boys and 18 (14.52%) girls with male:female ratio of 5.9:1. Seventy-six (61.29%) of the patients were in the age group 1–5 years range while 26 (20.97%) were in within 6–10 years range. Fifteen (12.09%) patients were <6 months; 15 and 7 (5.65%) patients were between 11 and 15 years. The most common indication for surgery was inguinal hernia in 56 (45.16%), followed by hydrocele in 24 (19.45%), whereas the least were rectal

biopsy 2 (1.61%), genital cyst 2 (1.61%), and circumcision 2 (1.61%) [Figure 1].

The distribution of Hb phenotype is as shown in Figure 2. HbAA was present in 97 (78.2%) children while the least was HbSS in 1 (0.8%) patient and HbSC 1 (0.8%). In 3 out of 4 patients, the parents are aware of their Hb phenotype, none of the parents were HbSS. The most common phenotype in the parents was AA 98 (79.82%), followed by AS 16 (13.13%), whereas the least was AC 5 (2.29%) [Figure 3].

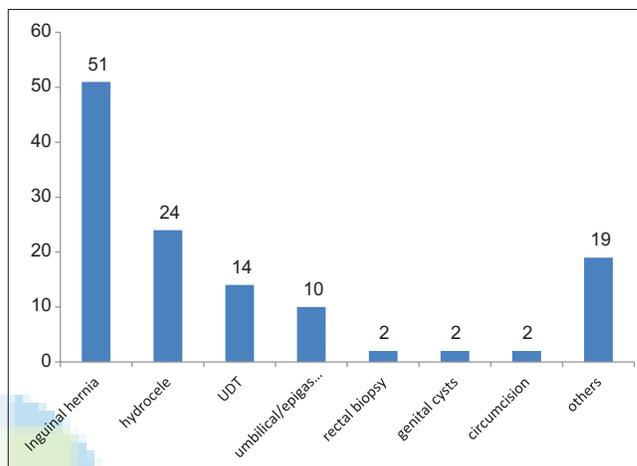


Figure 1: The indications for day case surgery in children

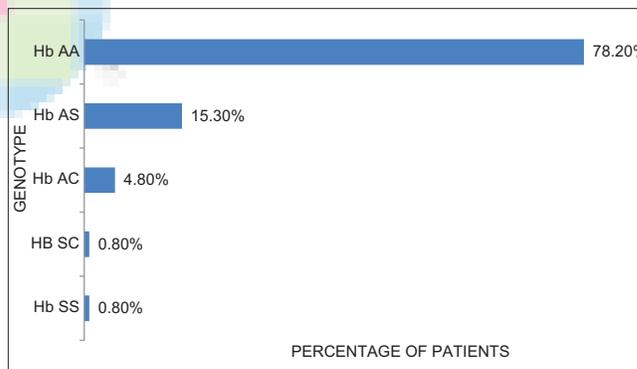


Figure 2: The hemoglobin electrophoresis pattern in children undergoing day case surgery

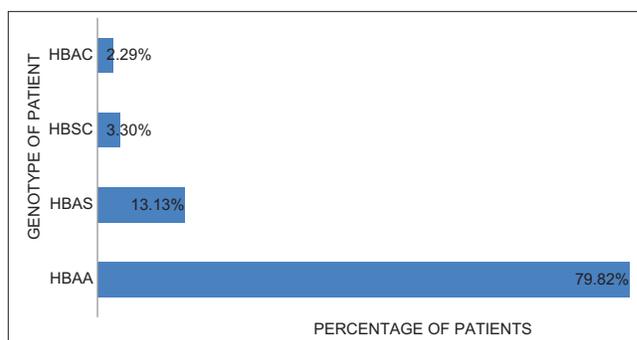


Figure 3: The distribution of parents' genotype

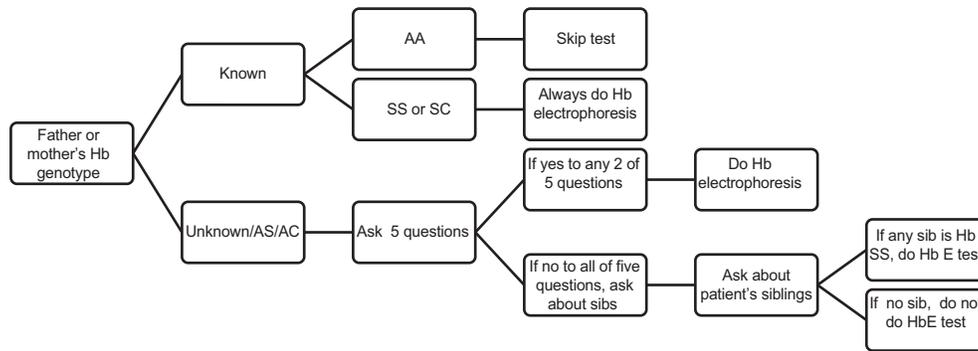


Figure 4: Lagos University Teaching Hospital algorithm for hemoglobin electrophoresis screening

Table 1: The association between history suggestive of sickle cell disease and the diagnosis of sickle cell disease

| Variable | Chi-square (χ^2) | P |
|------------------------|-------------------------|--------|
| Jaundice | 26.086 | 0.0001 |
| Hand and foot syndrome | 61.135 | 0.0001 |
| Frontal bossing | 40.701 | 0.001 |
| PCV at surgery (<28%) | 34.616 | 0.001 |

PCV=Packed cell volume

Nine children (7.3%) had a positive history of jaundice while 3 (2.4%) had frontal bossing. Two children (1.6%) had a history suggestive of hand and foot syndrome. There was no history suggestive of abdominal pain crisis or blood transfusion in any of the children. With respect to the preoperative PCV levels, five children (4.0%) had a preoperative hematocrit < 28%, 18 children (14.52%) had hematocrit of between 28% and 30%, and 101 (81.45%) had hematocrit > 30%. When these factors were subjected to a Chi-square test, the results obtained are as shown in Table 1.

There was no mortality and no readmission postsurgery.

Discussion

HBE is a routine preoperative investigation before a surgical procedure in this particular subregion.^[1,8] This has been the practice for some time because sickle cell disorder is not an uncommon coexisting medical condition in pediatric patients scheduled for surgery.^[1,8,9] A previous study from our center has, however, demonstrated that the need for HBE appears unnecessary considering the low prevalence of sickle cell anemia in our studied population (1.0%), and the relatively high cost of investigation in a resource-poor economy.^[8] Our observed prevalence in the current study is not significantly different from our earlier study and other prevalence studies in our center.^[9,10] In contrast, the prevalence, we reported was lower than 14% documented Vichinsky *et al.*^[11] in a group of children scheduled for inpatient surgical procedures. This may be because they included patients up to 18 years of age, other more complex surgical procedures such as cholecystectomy, tonsillectomy, and splenectomy. It is,

therefore, not surprising that they reported a mortality of 0.2% while we reported none.

We also observed that the age, gender distribution, and Hb phenotypes mirror the national pattern in Nigeria and West African subregion.^[9,12] This may suggest that the actual prevalence of sickle cell anemia in this region is low, and as such the use of other diagnostic criteria may be adequate to determine the presence of sickle cell disease. The previous history of jaundice, hand and foot syndrome, and frontal bossing were shown to be highly suggestive of the presence of sickle cell disease in our study. This is in addition to a hematocrit <28%. These clinical and investigative criteria are positive indicators that a child may have sickle cell anemia.

Our study also showed that about 3 of 4 parents are aware of their Hb phenotype, and 79.2% had a phenotype of HbAA; hence, inference to the likelihood of sickle cell anemia can be ruled out if one of the patient's parents has HbAA. Thus, in about 60% of patients presenting for day case surgery in our center, the likelihood of sickle cell anemia can be ruled out simply by finding out the HBE phenotype of the parents. Based on the clinical and investigative criteria of the patient and the HBE status of parents (if known), we are proposing an algorithm to streamline the screening of HBE to a specific group of patients presenting for pediatric surgical procedures [Figure 4]. It is to be noted that in patients with HbSS, adequate perioperative preparation should be ensured. In patients with sickle cell anemia, precipitating conditions such as infection, hypoxia, hypothermia, dehydration, and pain, which can precipitate a sickle cell crisis, should be avoided.^[11] Adequate oxygenation, perioperative monitoring, hydration, pain control, and the use of prophylactic antibiotics are advised.

Conclusion

Routine HBE for all patients undergoing day case surgery is unnecessary in our practice as about two-third of HBE test can be avoided by enquiring about the parents' Hb phenotype. The clinical risk factors for hemoglobinopathies

include the history of jaundice (outside the neonatal period), hand and foot syndrome, frontal bossing, and hematocrit <28% at the presentation. A simple algorithm as suggested will triage patients subjected to test.

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Conflicts of interest

There are no conflicts of interest.

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