Official Journal of the Irish Medical Organisation

Ir Med J; July/August 2022; Vol 115; No. 7; P631

August 18th, 2022

National Audit of Criteria For Pre-Operative Sickle Cell Screening in Children

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Abstract

Aim

We set out to identify the current practice in the anaesthesiology departments of Ireland's public hospitals that deliver paediatric anaesthesia with regard to pre-operative screening for sickle cell disease (SCD) and Sickle cell trait (SCT).

Methods

The Departments of Anaesthesiology at 14 public HSE-funded hospitals that deliver paediatric anaesthesia were contacted over a three month period in 2020. Any existing policies regarding pre-operative screening of paediatric patients for Sickle cell disease or trait were sought. Comparisons were made between any screening policies in place.

Results

A response was received from 11 of the 14 hospitals. Three out of 11 of the Anaesthesiology Departments have formal policies in place. The ethnicities identified for pre-operative screening varied across these three hospitals.

Conclusion

Despite a significant increase in the number of people of African, middle Eastern & Indian descent living in Ireland in recent years, no neonatal screening programme for Sickle cell exists here, and no national policy exists with criteria to guide the practice of pre-operative screening of patients for SCD/SCT (trait). Our survey highlights a lack of standardisation in the approach to pre-operative sickle cell screening of children across Ireland's public hospital system. In view of the increasing multiculturalism in Ireland we recommend a national review of the merits of the introduction of developing a targeted national guideline for pre-operative screening for sickle cell in at-risk children.

Introduction

Sickle cell disease (SCD) is an inherited autosomal recessive disease with multisystem complications as a result of vaso-occlusion and chronic haemolysis. [1] Complications of SCD include anaemia, ischaemic pain, acute chest syndrome, stroke, and increased risk of bacterial infections. Intra- and post-operative complications associated with SCD, including vaso-occlusive crises and acute chest syndrome, are well recognised and are a significant cause of morbidity & mortality. [1] Some of the known triggers associated with vaso-occlusive events include hypoxia, acidosis, dehydration, hypothermia, pain, and infections. [2] Perioperative complications of SCT are much less likely but can occur in extreme physiological circumstances.

The key to safe perioperative management of children with Sickle cell is planning and pre-operative optimisation.[1] Such preparation requires identification of children with SCT or SCD. The latest AAGBI guidelines published in June 2021 contain 12 key recommendations, including the requirement for a designated lead clinician to oversee the peri-operative management of patients with SCD, and a recommendation that all patients at risk of haemoglobinopathy should be screened for haemoglobinopathy before surgery.[3]

Despite a significant increase in the number of people of African, Caribbean, Mediterranean and middle eastern descent living in Ireland in recent years, no paediatric screening programme for Sickle cell exists here.

The Centers for Disease Control and Prevention (CDC) identifies SCD as being particularly common among those whose ancestors came from sub-Saharan Africa, Spanish-speaking regions in the western Hemisphere such as South America, the Caribbean and Central America; Saudi Arabia; India; and Mediterranean countries such as Turkey, Greece and Italy. [4] The NHS in the UK highlight SCD being particularly common in those with African or Caribbean family backgrounds, however screening for sickle cell disease is offered to all pregnant women in England to check if there is a risk of a child being born with the condition, and all babies are offered screening as part of the new-born heel prick test. [5]

To our knowledge, no national policy exists with criteria to guide the practice of new-born or pre-operative screening of patients for SCD/SCT. In this study we set out to identify the current practice in the Anaesthesiology Departments of Ireland's public hospitals that deliver paediatric anaesthesia regarding pre-operative screening for SCT and SCD.

Methods

The Departments of Anaesthesiology at 14 public HSE-funded hospitals that deliver paediatric anaesthesia were contacted, including Ireland's two tertiary referral paediatric centres, over a three month period in 2020. Initially, contact was made to Anaesthesiology departments through known colleagues by telephone or email.

Attempted contact was made to the departments with no known colleagues through their respective departmental emails attained through HSE website information. Following contact with the departments, email addresses were provided to us for Consultant Anaesthesiologists in departments that had not already been contacted.

Each contact was a consultant Anaesthesiologist who was currently working in their department at the time of screening and who carried out regular paediatric care in that hospital. A review of the current literature was reviewed and based on the data available plus data from NHS and CDC, a set of questions around screening were compiled. (Table 1). The contacts were asked questions either over the phone or through email. Once the information was gathered from each respondent department, comparisons were made between each department regarding any screening policies that were in place.

Table 1: Questions asked to each department.

1	Were there any existing policies regarding pre-operative screening of paediatric patients for Sickle cell disease or trait currently in place?
2	If so, who devised the screening protocol?
3	What countries/ continents were included in their screening protocol?
4	Were they testing children of whom one parent may be from an area of high Sickle cell disease?

Results

A response was received from 11 of the 14 hospitals.

Three Anaesthesiology Departments have formal policies in place with criteria for which their paediatric patients undergo pre-operative screening for sickle cell disease or trait. Two of the three departments with formal policies in place were answered and discussed during phone consultation. The other was answered by email.

Notably the ethnicities identified for pre-operative screening varied significantly across these three hospitals. In the first policy, all children of west African, central African and Afro-Caribbean ethnicity were screened for SCD. In the second policy only children of African ethnicity were screened for SCD. While in the third policy, all children from African, Middle Eastern, and Indian ethnicities were screened. This policy also stipulated that if one of the parent's ethnic origins were outside the identified regions then the child was not required to have pre-operative screening. This was not the case in the other two policies. Of the three hospitals that did have policies, each policy was compiled at a local level in conjunction with the Anaesthesiology and Haematology departments respectively.

Eight of the Anaesthesiology Departments contacted currently have no formal departmental policy in place determining which children require pre-operative sickle cell screening; in these institutions sickle cell screening occurs on a more ad hoc basis.

Discussion

This paper has highlighted that there is significant heterogeneity in the screening of children for SCD across Ireland. As it stands in Ireland some children deemed at risk may be screened for SCD prior to elective surgery. However other children may not get screened depending on their hospital policy, or lack thereof in many instances. A standardised policy approach would eliminate this potential discrepancy.

Of the three hospitals that do have formal policies in place, it is difficult to determine why certain countries, regions or continents were included in some but not in others. The individual countries/ continents included are understandable, however the significant differences between the policies cannot be easily explained. While having policies in place is seen as welcomed step towards improved identification of children with SCD in Ireland, there is significant scope to improve this process.

Using the 12 recommendations from Walker *et al* 2021 could significantly improve the screening and management of children with sickle cell disease undergoing surgery in Ireland. The paper brings together the best evidence currently available and in doing so provides an opportunistic blueprint on how to optimise national screening and management of this patient cohort. It has been specifically written to clarify organisational aspects of care and to support anaesthesiologists who manage patients with sickle cell disease.[3] We can see that some of the recommendations are already taking place in the three hospitals that have policies in place. With this framework of recommendations, it is possible to implement these strategies at a national level.

Figure 1: Recommendations[3]

Recommendations

1 Clinical teams should work in partnership with patients and their families, and endeavour to make sure they are kept informed of clinical decisions relating to their care at all times.

2 The lead clinician in the Department of Anaesthesia is responsible for making sure that there is local departmental guidance for the peri-operative management of patients with sickle cell disease, developed in partnership with the haematology team. The Department may wish to identify a specific lead for this role.

3 All patients at risk of haemoglobinopathy should be screened for haemoglobinopathy before surgery but unnecessary repeat screening should be avoided.

4 There should be a nominated lead haematologist (or for children, a lead paediatrician or paediatric haematologist) when a patient with sickle cell disease undergoes surgery. The nominated leads are responsible for deciding the peri-operative transfusion plan, with support of the specialist centre where relevant.

5 Local governance arrangements should be in place so that the surgical team booking the patient for surgery communicates the sickle cell disease diagnosis at all stages of the patient pathway, and documents this clearly in the patient record so that the relevant teams are aware: haematology; anaesthesia; transfusion laboratory; waiting list co-ordinators; pre-assessment; and ward nursing staff.

6 Patients with sickle cell disease presenting for elective surgery should be reviewed in a pre-assessment clinic, with input from the nominated lead haematologist (or for children a paediatrician/paediatric haematologist). The haematology team must be informed if a patient with sickle cell disease is admitted for emergency surgery.

7 The acute pain team should be notified in advance if a patient with sickle cell disease is undergoing major surgery, particularly if the patient has a history of chronic pain.

8 Patients with sickle cell disease should be scheduled early on the operating list to avoid prolonged starvation. Last minute cancellations for administrative reasons should be avoided, particularly if the patient has received a blood transfusion in preparation for surgery.

9 Patients are at increased risk of sickle complications (acute chest syndrome, pain, acute renal insufficiency or stroke), sepsis and venous thromboembolism in the peri-operative period. The majority of complications occur postoperatively, and there should be a low threshold to admit patients to high dependency or intensive care.

10 Patients require meticulous peri-operative care to avoid factors that may precipitate sickling: dehydration; hypoxia; acidosis; hypothermia; and pain. Routine surgery should be avoided if the patient is febrile or having a painful crisis.

11 Pregnancy confers an increased risk for patients with sickle cell disease. Patients should be managed by a multidisciplinary team and be encouraged to give birth in hospitals able to manage high-risk pregnancies and complications of sickle cell disease.

12 Patients should be managed according to standard COVID-19 care pathways, striking a careful balance between limiting hospital contact to minimise the risks of viral exposure and avoiding delays to essential treatments.

What other guidelines are available on this topic?

There are existing evidence-based guidelines and quality standards relating to the care of patients with sickle cell disease published by the National Institute for Health and Care Excellence [1,2], British Committee for Standards in Haematology [3], British Society for Haematology [4], Sickle Cell Society [5], UK Forum on Haemoglobin Disorders/West Midlands Quality Review Service [6], NHS screening programme [7], Royal College of Obstetrics and Gynaecology [8] and US National Institute of Health [9].

Why were these guidelines developed?

Sickle cell disease is one of the most common serious inherited single gene disorders worldwide and has a major impact on the health and life expectancy of the affected individual. Peri-operative complications are higher for patients with sickle cell disease than for the general surgical population, but outcomes can be improved with careful

Sickle cell is becoming increasingly prevalent in Ireland as cultural diversity increases across the country. There are approximately 500 people with SCD in the Republic of Ireland & the estimated amount of people with SCT is unknown.[3]

Many European countries including the UK & Northern Ireland have taken further strides to identify those with SCD&SCT, by introducing new-born screening into standard care. An Oireachtas committee report from 2014 identified a growing need for a national standardised approach to new-born Sickle cell screening in Ireland.[6] As globalisation, cultural diversity, and integration increases, SCD and other haemoglobinopathies will become more prevalent in Ireland. Identification of children with sickle cell pre-operatively is key to minimising peri-operative morbidity and mortality. A broader conversation on the need and plausibility of screening for these other haemoglobinopathies such as Thalassaemia is something beyond this paper but should be subject to further discussion.

Our survey highlights a lack of standardisation in the approach to pre-operative sickle cell screening of children across the Republic of Ireland's public hospital system. In view of the increasing multiculturalism of Irish society, we recommend a national review of the merits of the introduction of a targeted neonatal screening programme for sickle cell in at-risk children. In the absence of such a programme, a standardised approach to pre-operative screening across the various hospitals that provide paediatric anaesthesia would be extremely warranted and beneficial.

Declaration of Conflicts of Interest:
We declare no competing interests with this report.

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