

Sickle cell screening Hot Topic – Dr Kim Jemmett

Q. Your patient is a 6-year-old ASA 1 boy born in England and of African descent. He is scheduled for an elective procedure under tourniquet. Do you insist on a sickle cell screen?

A. yes or no

The question of who should be tested for Sickle Cell disease and other haemoglobinopathies in the pre-operative period still appears to be an area of some confusion. A 2011 APA survey showed that there was significant variation of opinion amongst anaesthetists on this topic (1). Various factors may affect the anaesthetists decision to test a child for Sickle Cell Disease, including age, haemoglobin level, family history, ethnicity and type of surgery. Unfortunately the reliability and safety of using these factors is unknown.

There are guidelines set out by the AAGBI stating Sickle Cell Disease (SCD) testing should be carried out in 'susceptible populations' (2). The NICE guidelines state that testing should occur in African, Afro-Caribbean and people from other ethnic groups considered to be at risk (3). However, defining and identifying at risk groups will be difficult. Sickle cell disease has been seen in people from the Eastern Mediterranean, the Middle East, Asia, Cyprus and other White ethnic groups. Statistics gathered from the neonatal screening programme showed that 1.6% of babies identified as having a significant haemoglobinopathy were described as White British (4).

In 2001 the Sickle Cell and Thalassaemia screening program was set up. This includes antenatal testing of the pregnant women and their partners (if risk identified from maternal screening) and then subsequent testing of the neonate as part of the newborn screening programme ('blood spot' or 'heel prick test'). Screening is offered to all pregnant women and their neonate within the UK. The screening programme was fully established within England by 2006, Scotland by 2010, Northern Ireland by 2012 and Wales by 2013. Screening data shows that the rate of decline of testing is low at 0.1% (4); so it is very unlikely that a child will have an undiagnosed significant haemoglobinopathy if they were born within the UK after the date of screening implementation.

Results from newborn screening are made available at the child's routine 6 week follow up and can then be found in the child's health record held by the parent or guardian. Results can also be accessed by contacting the child's local health authority. However; a child diagnosed with a significant haemoglobinopathy will have regular follow up within the health system and the diagnosis will therefore be known to the parent or guardian of the child.

A recent article in Paediatric Anaesthesia outlined a Pre-operative Sickle Cell Screening guideline used in a UK District General Hospital (5). The guideline utilises the Sickle Cell and Thalassaemia screening programme to simplify and minimise testing in the pre-operative period. The guideline suggests that testing should only be carried out on children from at risk groups if they were born

before the introduction of the screening programme. Although, as discussed above, identifying those groups can prove difficult. Currently children aged 10 and under born in England will have been tested, however this age will obviously increase, and by 2022 all children aged 16 and under will have been part of the neonatal screening programme. As a child gets older the likelihood of having undiagnosed Sickle cell disease significantly reduces. A study in Nigeria (where routine testing at birth is not performed) to determine the age at diagnosis of children with Sickle cell disease showed that by the age of 5, 90% of patients had been diagnosed and by the age of 10, 98% had been diagnosed (6).

The safety of using a tourniquet to aid surgery in patients with Sickle Cell Disease is still not fully known. A review article in the Southern Medical Journal in 2010 summarizing current available evidence concluded that a tourniquet could be used with relative safety in most patients with Sickle cell disease and trait provided proper perioperative management and safety precaution were taken (7). Careful exsanguination of blood when applying the tourniquet was considered important.

So, given the above I feel that testing the child in the question for Sickle Cell Disease is unnecessary. Subjecting the child to a blood test in this case will cause unnecessary stress and discomfort as well as being a waste of resources. I have trust in the newborn screening programme and I am therefore assured that the child does not have a significant hameoglobinopathy. This well child may have sickle cell trait. Although Sickle trait will also be diagnosed as part of the newborn screening programme, this information is not always retained or volunteered by parents. Sickle Trait is not considered a disease state and a diagnosis of this would not affect my anaesthetic. I may however discuss with the surgeons the importance of careful exsanguination of blood before application of the tourniquet.

Reference:

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