NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Transcranial Doppler (TCD) screening for Sickle Cell Disease (SCD)

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

Background

HbSS and HbSβ^0 patients

All children and young adults with Hb SS and HbSβ^0 thalassaemia subtypes of sickle cell disease, should be offered annual TCD scans from age 2 years until at least age 16 years. This is based on the findings of a randomised controlled trial on the benefits of transfusion in children with raised cerebral blood-flow velocities in the reduction of stroke. The NHS Sickle Cell and Thalassaemia Screening Programme has published Standards and Guidelines for Transcranial Doppler scanning in Children with SCD.[1,2]

HbSC patients

There is no national recommendation for the screening of children with HbSC sickle cell disease. There is still a risk of cerebrovascular disease, but this is lower than for children with HbSS or HbSβ^0 and the yearly screening and normal values for the other sickle cell groups are thought to be inappropriate as this group of children are not anaemic to the same extent. In Scotland we have developed an approach for this group based on guidance obtained from Kings College Hospital who provided us with their protocol and normal values, based on their assessment of many children with this genotype[3] and which is in keeping with other publications.[4]

Patient/parent information

Prior to the TCD, parents and carers should be given appropriate verbal and written information by the referring team to enable an informed decision to be made about the necessity of the TCD scan and for families to understand and accept the need for of chronic transfusion and its consequences if an abnormality is detected. The association between high blood velocity in the cerebral arteries and the risk of a stroke should be made clear and hence the purpose of the test. SPAH patient information leaflets are available, for those living within NHS GG&C and those who live outwith NHS GG&C click here.

The information leaflet also explains the TCD process and the limitations of the procedure will be sent to parents and carers with the first appointment. There will be a brief discussion about the results with the RHC Haematologist on the day. If an abnormality is found, this will be explained along with the follow up process for further TCD and a letter with results will be sent back to the referring clinician.

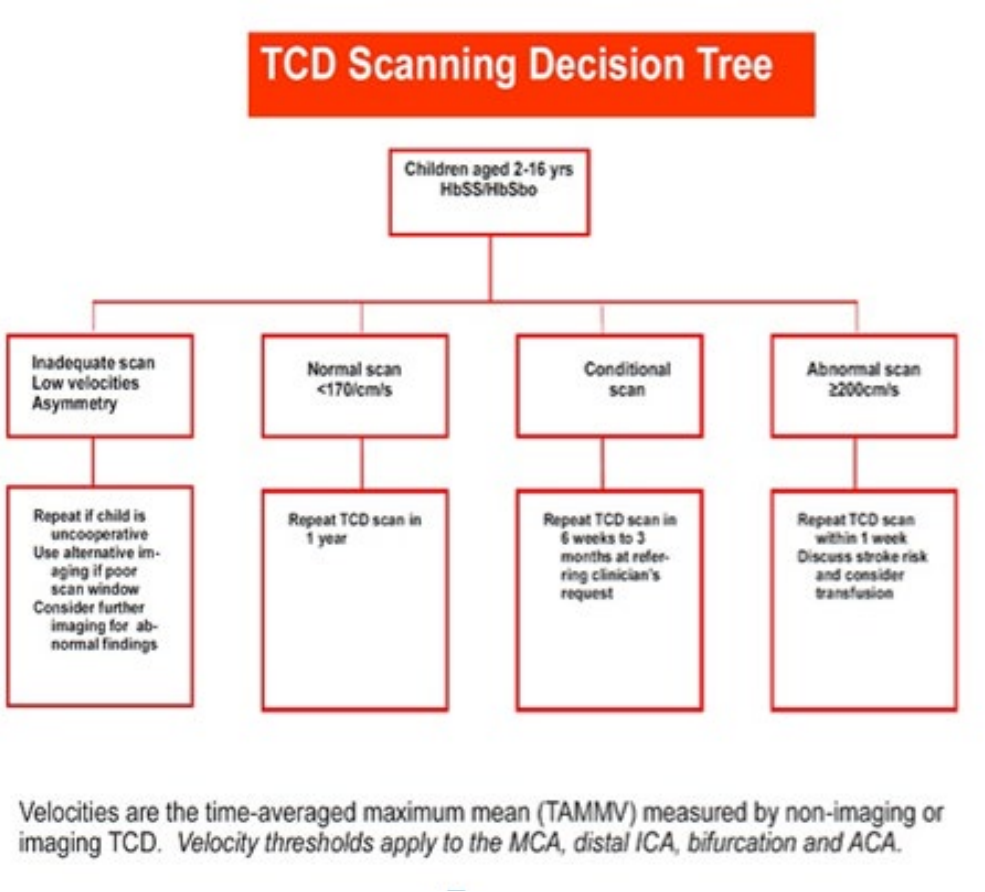
TCD referral procedure

In Scotland TCD studies are carried out centrally at RCH Glasgow. The referral form is available on the SPAH website here and should be sent to the **RHC Non-malignant team on ggc.rhctcdreferrals@ggc.scot.nhs.uk**

Results will be returned to the referring clinician with a covering letter. In the event that the patient DNAs they will be automatically reappointed and the referring clinician will be informed.
Families may be eligible to reclaim travel costs from the local health board and should be given the appropriate claim form by their local team and advised to keep their receipts.

HbSS and HbSβ^0 TCD protocol

As per national protocol TCD screening should be offered at least annually from the age of 2 years to all children with HbSS and HbSβ^0 sickle cell disease. Timings of repeat scanning depend on the TCD results as below.



Velocities are the time-averaged maximum mean (TAMMV) measured by non-imaging or imaging TCD. *Velocity thresholds apply to the MCA, distal ICA, bifurcation and ACA.*

HbSC TCD procedure

Children with HbSC should receive a transcranial Doppler scan at

2yrs of age,
5 yrs of age and
10 yrs of age

If TAPV readings are <132 cm/s then no additional scans are required
If TAPV readings of > 132 cm/s are obtained then closer review and further imaging should be considered in discussion with the clinical MDT.

Actions to be taken based on results of TCD

Interpretation of TCD velocities and recommendations for further investigation and clinical management can be found in the Transcranial Doppler Scanning for Children with Sickle Cell Disease Standards and Guidance, Second Edition September 2016.

References

1. Standards and Guidance for Clinical Care, 2nd Edition 2019
2. Transcranial Doppler Scanning for Children with Sickle Cell Disease Standards and Guidance Second Edition September 2016 - UK Forum (haemoglobin.org.uk)
3. Communication form Dr Sue Height and Dr Colin Deane, Kings college Hospital NHS trust
4. Transcranial Doppler in haemoglobin SC disease. Viera C et al; Paediatric Blood and Cancer E26342, Volume 65 issue 4