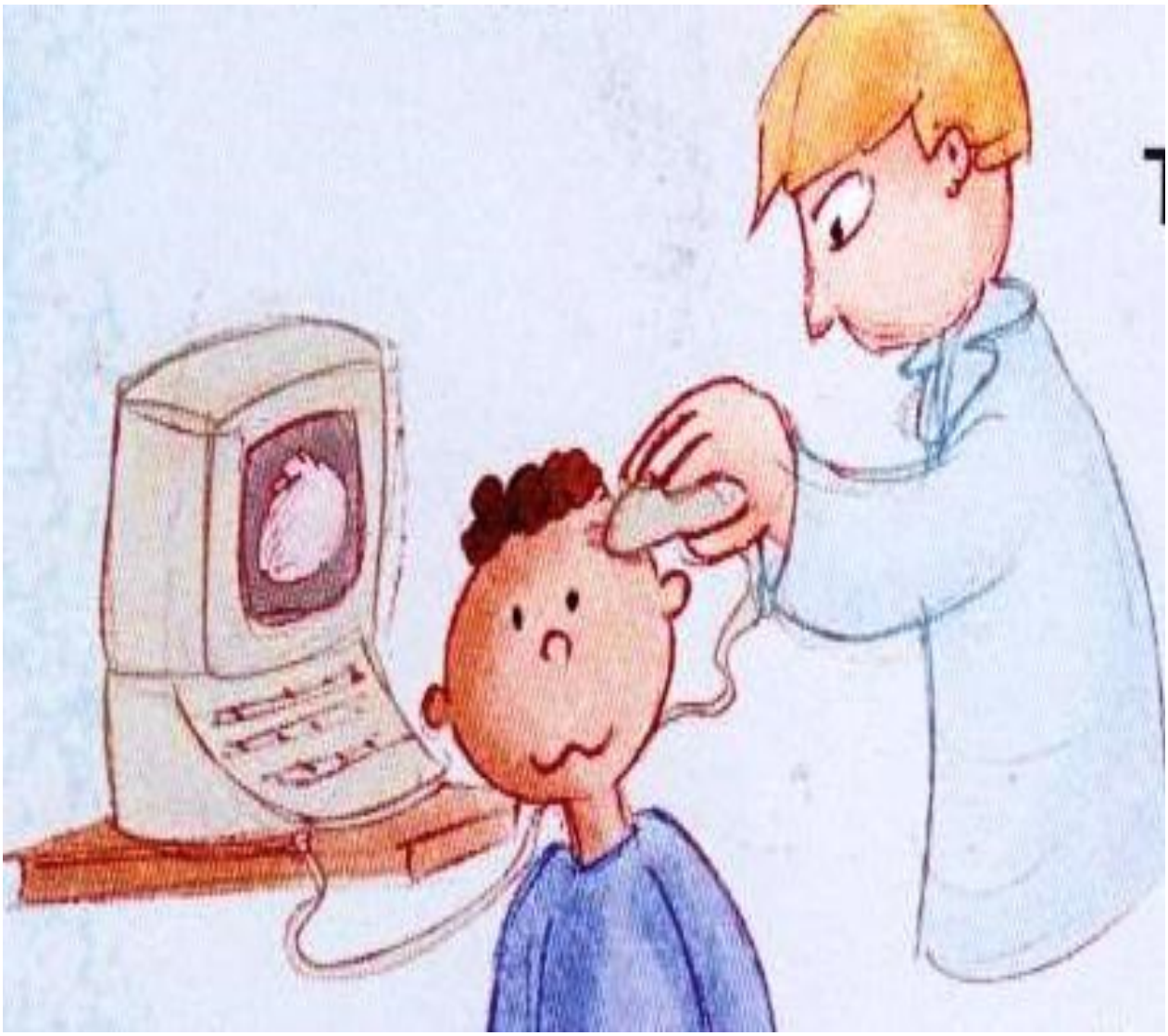
Transcranial Doppler Scanning for Children with Sickle Cell Disease Standards and Guidance

Second Edition September 2016



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Introduction

The following standards have been written to replace the original transcranial Doppler (TCD) standards published in March 2009. These updated standards include new guidance for; non-imaging and imaging TCD cut-off points and screening for extracranial internal carotid (eICA) stenosis. The standards also provide information on training, quality assurance and the TCD Practitioner Register, which will be held by the UK Forum.

List of main contributors

Many people have contributed to these standards and guidelines and we would like to thank them all for their input. Major contributions have been made by Dr Colin Deane, Dr Soundrie Padayachee, Dr David Goss, Dr Baba Inusa and Dr Paul Telfer.

Organisation of TCD scanning services

All children and young adults with sickle cell anaemia (Hb SS) and HbS ß zero thalassaemia, should be offered annual TCD scans from age 2 years until at least age 16 years. The need for children with other types of sickle cell disease to be screened should be reviewed on a case-by-case basis.

TCD or TCD imaging (TCDi) are both acceptable techniques for performing screening, with the method of choice depending on local circumstances but all scans must be performed by competent specialists.

It is expected that the mode of delivery of the service and choice of equipment will depend on the configuration of clinical services for children with sickle cell disease. This will probably be determined by the prevalence of the condition in any particular area. Children could be scanned in an outpatient clinic environment, ultrasound department or in the home. There should be a lead specialist/clinician taking responsibility for directing the TCD scanning services within any particular locality.

All parents and carers should be given a verbal explanation of the TCD scanning process and limitations of the procedure, together with an explanation of the follow up process if an abnormality is found. 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. Sufficient verbal and written information (see Appendix) should be given to enable an informed decision to be made about the necessity of the TCD scan and accepting the consequences of chronic transfusion if an abnormality is detected.

Scanning protocols and follow-up

The protocols for TCD scanning and categorisation are based on the criteria developed from the first Stroke Prevention Trial in Sickle Cell Anaemia (STOP 1 trial) in the United States; this trial used non-imaging TCD. Non-imaging TCD and TCDi (imaging TCD) using duplex scanners are both effective methods to examine children with SCD. Although early studies reported that TCDi velocities could be lower than those measured by non-imaging TCD, there is sufficient evidence that, with correct technique, optimisation and measurement, the same velocity thresholds can be applied for both methods. It is recommended that the method of scanning (TCD or TCDi) should be quoted on the report.

Operators undertaking scans must demonstrate proficiency to scan, which will be determined by attendance at a recognised training programme and successful competency evaluation. Competent operators will be eligible to apply for inclusion in the UK TCD Practitioner's Register held by the UK Forum.

Arterial blood velocities must be examined in the distal intracranial carotid artery (dICA), middle cerebral artery (MCA), anterior cerebral artery (ACA) and posterior cerebral artery (PCA) on both sides of the head.

The classification should be based on the highest time averaged maximum mean velocity (TAMMV) measured in the distal ICA or MCA during the examination. The risk category is based on the TAMMV cut-offs as identified in the STOP trial as follows:

Normal velocity - low risk	All velocities < 170 cm/s
Borderline velocity – conditional risk	Any velocity in the range 170-199 cm/s
High velocity – high risk	Velocity in the dICA/MCA ≥200 cm/s

Inadequate imaging or unusually low MCA TAMMV are also abnormal as follows:

Unusual low velocity	Velocities <70cm/s in MCA
Asymmetrical velocities	Velocity <50% of contralateral MCA
Inadequate image	Incomplete images and measurements from dICA, MCA, ACA or PCA bilaterally

Low velocities or pronounced asymmetry are indicative of possible occlusion and should prompt further investigations and alternative imaging. A TCD scan would be defined as non-diagnostic if for whatever reason unsatisfactory results were obtained. This might be due to causes such as an uncooperative child (in which case a repeat scan should be considered), poor scanning window (in which case an alternative scanning method such as MRI/MRA should be considered) or previous stroke.

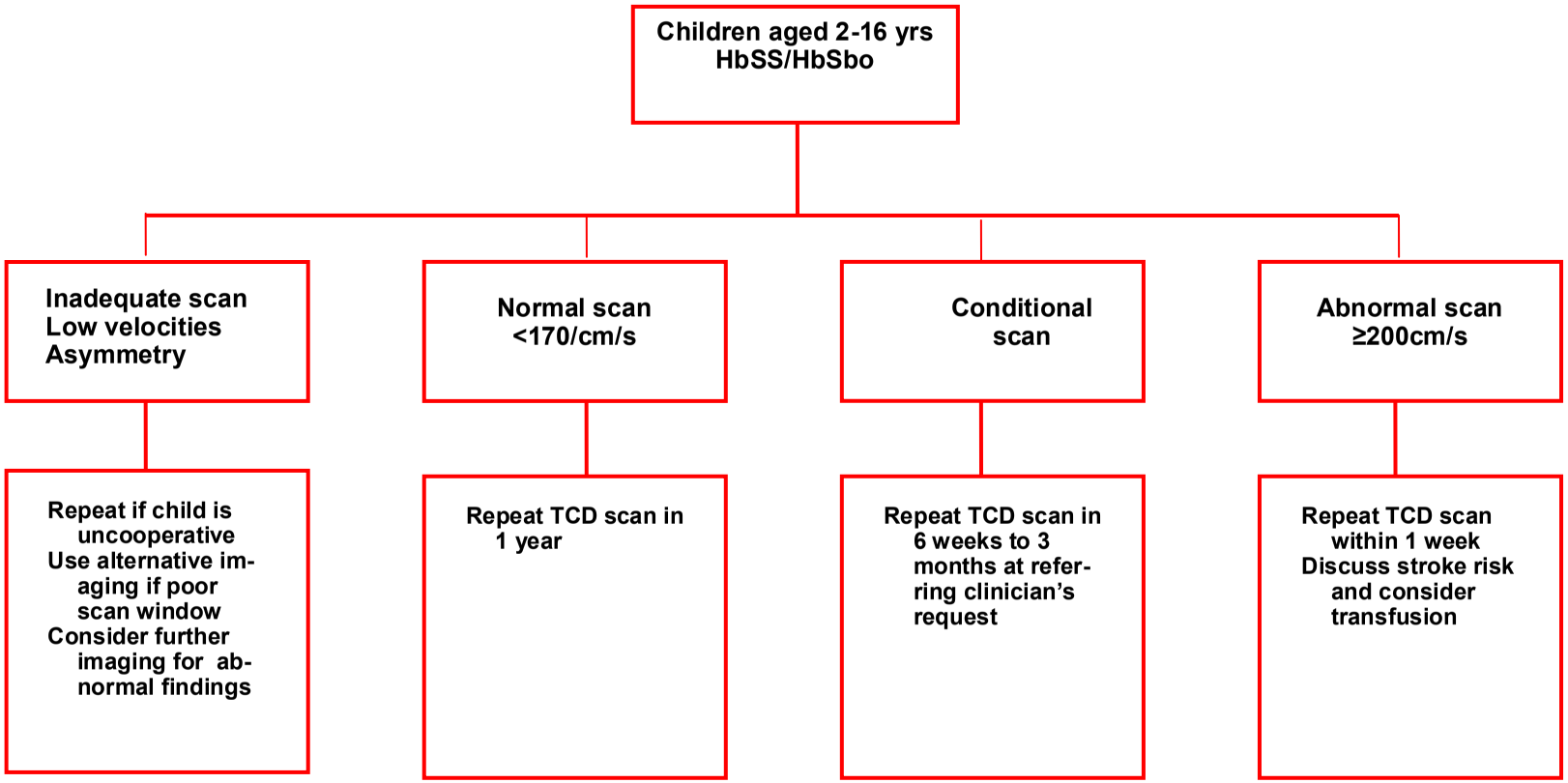
The original STOP study did not include a category for abnormal velocities in the ACA. Subsequent analyses indicate a raised risk of stroke in cases of ACA velocities ≥170 cm/s. These findings should prompt early repeat scanning with further clinical and imaging investigation for possible intervention. ACA velocities ≥ 200 cm/s should be considered as high risk.

The action taken following the categorisation of results should depend on the age of the child and follow the protocols as given in the associated algorithm. Repeat TCD scans should be undertaken at the time intervals recommended.

Because of the long-term consequences of starting chronic transfusions in children at high risk, all available data should be considered prior to beginning treatment. Repeat TCD scanning is recommended for all abnormal classifications. The assessment should also include a comprehensive neurological assessment and the results of other imaging studies such as MRI/MRA, although these are not used in the risk classification and treatment should not be altered or delayed for this reason.

Ongoing TCD scanning is recommended once a child has started on regular transfusions. Raised velocities may return to normal levels with transfusion, and if this fails to happen or if velocities increase, further investigations and interventions should be considered. The time interval for performing these scans should be yearly or shorter depending on the TCD velocity and the individual clinical circumstances.

TCD Scanning Decision Tree



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.*

Extracranial internal carotid artery examination

Examination of the cervical ICA was not part of the STOP trial and related recommendations. There is, however, increasing evidence that extracranial carotid artery (eICA) stenosis in children with sickle cell disease is an independent risk factor for silent cerebral infarction. Although there is no agreed level to classify an eICA stenosis and velocities are affected by haemoglobin level and tortuosity, two techniques have been used in published data:

Non-imaging/imaging: Use of a 2MHz phased array or TCD probe with a sub-mandibular approach, no angle correction, and a TAMMV of ≥ 160 cm/s.
Imaging: Use of a linear array with angle correction and a peak systolic velocity of ≥ 300 cm/s.

The current level of evidence indicates that scanning the eICA provides useful information on risk and that if high velocities are detected, clinicians should consider further investigations for vasculopathy and ischaemic lesions.

Training

All operators performing TCD or TCDi scanning on children must have had appropriate training in the technique. Training will be organised nationally, incorporating three elements:-

- A training day on the theory of TCD scanning, protocols and equipment with demonstration and hands-on TCD scanning practice on adults/other trainees.

  Each trainee will receive a training manual. 'Hands on' TCD training will be provided with children in a clinic environment. Further practice can be carried out locally but only under adequate supervision in a clinic environment.

- Trainees will be expected to keep a log book showing records of the subjects scanned and the procedures undertaken. It would normally be expected that trainees will conduct at least 40 TCD scans on children to gain competence in the technique unless they are performing other vascular ultrasound scans routinely, which can contribute to 50% of the log book scans.

- A final competency evaluation will be carried out either at the trainee's place of work or at the training centre. Successful trainees will be entered onto the TCD Practitioner Register held by the UK Forum.

Quality assurance

Those centres undertaking TCD scanning must be part of a network of care for sickle cell children and be part of any national approval/accreditation process of those centres.

TCD scanning should only be performed by practitioners approved by the UK Forum. All TCD practitioners will be required to complete an annual QA return, confirming the number of scans performed to enable them to remain on the register. The guideline number of TCD scans to maintain proficiency is considered to be a minimum of 40 per year. It would be appropriate to consider refresher training for those operators performing fewer than this number. Network centres must determine the best way to provide sufficient numbers of staff to maintain a quality TCD service for the sickle cell population in their area.

The UK Forum will work towards the establishment of a national database to incorporate all TCD scanning results on children with sickle cell disease. This will enable scanning results to be correlated with clinical outcome and audit the effectiveness of the screening programme. This will form the second part of the quality assurance scheme to identify centres reporting unusual TCD results or clinical outcomes that differ from their peers.

Appendix 1 - Good Clinical Practice

Publications for Good Clinical Practice are available at the following links:

1. **Nice.org.uk/guidance/qs58**
2. **http://www.hbpinfo.com/ukts-standards-2008.pdf**

Appendix 2 - TCD information leaflet

Transcranial Doppler Scanning

Contents

What is Transcranial Doppler (TCD) scanning?

TCD is a test that uses ultrasound to measure how fast the blood flows through the blood vessels within the brain. The machine detects the noise of the blood rushing through the vessels and uses this to measure the speed at which it is travelling. It is an extremely safe, easy procedure and is the only way to detect an increased risk of stroke in children with sickle cell disease. Dr Soundrie Padayachee is the specialist leading this service.

Why does my child need a TCD scan?

In sickle cell disease, blood vessels can be damaged by the sickled cells sticking to the walls. This causes the blood vessels to narrow and potentially close up. If this happens in the brain, the blood gets cut off, starving the brain of oxygen and causing a stroke.

What is a stroke?

A stroke is when the oxygen flow to part of the brain is reduced. This causes weakness in an arm or leg, difficulty talking or understanding what is said and/or memory problems. These problems may either be short-lived or permanent. Strokes can happen more than once.

What happens during the TCD scan?

The test is done by the specialist radiologist in either the x-ray department or in the sickle clinic. No preparation is required beforehand.

The scan takes about 15 minutes and your child will be asked to lie on a couch and keep still. He/she will be awake and you may stay with them throughout the scan.

The scanner will run over the side of your child's neck and forehead and is painless. Afterwards you will be able to go straight home, although if the test is done in the sickle clinic, Dr Inusa will be there to discuss the results with you.

What do the results mean?

The results can be normal, conditional or abnormal.

- A **normal** result means that nothing needs to be done now, but the test will be repeated each year.
- A **conditional** result will mean that the scan should be repeated within three months.

An **abnormal** result means that there is an increased risk of having a stroke in the future. If the result is abnormal, the scan will be repeated within a month and other tests may be arranged, such as an MRI (Magnetic Resonance Imaging) scan or a special x-ray. Neither of these will be painful for your child. All the results will be sent to Dr. Inusa for discussion with you. The treatment decision, however, will be based on the results of the TCD and not the MRI

What happens next?

Research has shown that giving a child regular blood transfusions (about every four weeks) can substantially reduce the risk of a future stroke. If your child's TDC results are abnormal and you consent to treatment, they should start regular blood transfusions as soon as possible.

How long will the transfusions continue for?

The increased risk of stroke is highest between the ages of two and 16 (peaking at seven years of age).
However, studies have shown that even when the blood flow is back to normal the increased risk of stroke remains, so it is best continue with transfusions in the long term.

What if I think my child has had a stroke?

We know that one in 10 children with sickle cell anaemia (HbSS or HbSβ0) will have developed stroke by the age of 14 if nothing is done about it. It is important to report any symptoms of a stroke such as:

- weakness in an arm or leg
- difficulty speaking and/or understanding,
- memory problems.

You should see a doctor immediately, even if you are on holiday, so that tests can be carried out to find out whether your child has had or is having a stroke. If a stroke is confirmed, you will be offered regular transfusions to help prevent further strokes in the future.

What if I have more questions?

The haemoglobinopathy team is available to answer any questions or concerns you may have.

Please add contact details:

Consultant Paediatric Haematologist
t:

Sickle Cell Nurse
t:

Specialist Community Nurse
t:

Further information

The following organisations provide further information, support and advice for patients and parents with sickle cell disease:

The sickle cell information centre
An information service covering all aspects of sickle cell disease.
w: www.scinfo.org

Sickle Cell Society
Offers advice and support on sickle cell and thalassaemia
t: 0800 001 5660 **w:** www.sicklecellsociety.org

Appendix 3 - References

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