Transcranial Doppler Scanning for Children with Sickle Cell Disease

Standards and Guidance (March 2009)
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Foreword

Diane Abbott MP

Effective and comprehensive care, particularly preventative care, for patients with sickle cell and thalassaemia has historically been an overlooked area. Health services have been slow to adapt to effectively serve this group - patients who often face significant health inequalities.

As Chair of the recently formed All Party Parliamentary Group on Sickle Cell and Thalassaemia I am therefore delighted to be able to welcome the first edition of these Standards for Transcranial Doppler Scanning for Children with Sickle Cell Disease.

Good quality TCD scanning has the potential to have a major impact on effectively identifying those children most at risk of stroke. With early intervention and preventative action it will be possible to significantly reduce the incidence of strokes among this at-risk group and, ultimately save lives.

Of course TCD scanning is only one of the elements of care necessary to ensure comprehensive improvement of clinical services across England but it is a great step and, alongside the developing clinical networks, I am confident that it is an example of the wider improvements that will develop in care for these diseases.

I would like to thank the NHS Sickle Cell and Thalassaemia Screening Programme for leading on this process; I know they appreciated the very supportive level of response they received from both clinicians and voluntary organisations, particularly the Sickle Cell Society, who contributed to the drafting of these standards. David Worthington took the challenging role of lead editor and deserves particular mention for drawing together all comments into this final whole.

Diane Abbott MP
Chair
All Party Parliamentary Group on Sickle Cell & Thalassaemia
Introduction

Stroke is a major cause of mortality and morbidity in children with sickle cell disease. Strong evidence from studies in the United States has demonstrated that early detection of narrowing of arteries in the brain, and subsequent transfusions to bring down the concentration of sickle haemoglobin, can decrease the risk of stroke dramatically. Doppler ultrasound can be used to measure the arterial blood flow and is a relatively straightforward technique that is well tolerated by even very young children.

These standards and guidelines have been written to help those clinical staff looking after children with sickle cell disease. National standards have already been published for the clinical care of children with sickle cell disease and these recommend regular transcranial Doppler (TCD) ultrasound for children aged two and upwards. What has been lacking in the past has been national guidance on how the TCD scan should be performed, the cut-off values that should be used to decide further action and the frequency of the scans. It is also important that all ultrasound operators perform the scan in a similar way and for this reason guidance on training and quality assurance has needed to be considered as well. The ultimate aim is to ensure that all children are offered a scan of high quality wherever they happen to reside.

Alongside the publication of these standards, the Sickle Cell and Thalassaemia Screening Programme is commissioning a training programme for those ultrasonographers who will be undertaking the TCD scanning. It is anticipated that a network of training centres will be established where the theoretical background will be taught and ‘hands on’ practical training will be given.

We would welcome any comments that you have about these standards and guidelines. This document is very much a ‘first edition’ and may need to be modified or extended in the future. We will be reviewing the contents in the Spring of 2010.

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, Professor Fenella Kirkham and Dr David Rees.

Dr David Worthington
Editor
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 scanning, with the method of choice depending on local circumstances.

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 clinician taking responsibility for directing the TCD scanning services within any particular locality.

All parents/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. This should be backed up with appropriate written information. Sufficient verbal and written information should be given to enable an informed decision to be made about the desirability of the TCD scan and accepting the consequences of chronic transfusion if an abnormality is detected. The Programme Centre intends to provide a suitable patient information leaflet that could be used by those centres performing the scans.
Scanning protocols and follow-up

The protocols for TCD non-imaging scanning and categorisation of results are based on the criteria developed from the first Stroke Prevention Trial in Sickle Cell Anaemia (STOP 1 trial) in the United States. TCD imaging techniques were not investigated in the STOP trials and the recommendations for the imaging protocols have been based on more recent comparison data between the two methods. Because of these limitations, the method of scanning (TCD or TCDi) must be quoted on the report.

Arterial blood velocities must be examined in the distal intracranial internal carotid artery (ICA) and middle cerebral artery (MCA) on both sides of the head. Velocities in the anterior cerebral arteries (ACA) and posterior cerebral arteries (PCA) should also be examined for additional information and to help refine the stroke risk assessment but are not used in the STOP classification of risk.

The scan results should be divided into five categories depending on the time averaged maximal mean (TAMM) velocity recorded, whether in the ICA or MCA or the bifurcation of the two arteries:-

- Inadequate image
- Unusual low velocity
- Normal velocity - ‘low risk’
- Borderline velocity - ‘conditional’
- High velocity - ‘high risk’

A TCD scan would be defined as inadequate if for whatever reason unsatisfactory results were obtained. This might be due to such causes 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), previous stroke, etc. The time interval for a repeat scan would depend on clinical judgement and considered on a ‘case by case’ basis.

Low velocities of <70 cm/s in the MCA, or a velocity <50% of the contralateral MCA, are indicative of possible occlusion and should prompt further investigations with MRI or CT. Assessment of the blood velocities in the ACAs and extracranial ICAs at the time of the TCD scan might provide useful additional information in these cases.

The TAMM blood velocities used as cut-offs to define the risk limits would be:

<table>
<thead>
<tr>
<th>TCD non-imaging</th>
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</thead>
<tbody>
<tr>
<td>Normal velocity - ‘standard risk’</td>
<td>&lt;170 cm/s</td>
</tr>
<tr>
<td>Borderline velocity - ‘conditional’</td>
<td>170 to 199 cm/s</td>
</tr>
<tr>
<td>High velocity - ‘high risk’</td>
<td>≥200 cm/s</td>
</tr>
</tbody>
</table>
TCDi using duplex scanners can be used to examine children, although some studies have shown that TCDi velocities can be up to 15% lower than those measured by non-imaging TCD. Improved technique and a change in imaging parameters can reduce this difference to 10% or less. If TCDi is used as the scanning technique, appropriate allowances should be made for velocities that are within 10% of the conditional or abnormal risk thresholds. (Further work will be undertaken to examine the reasons for these discrepancies and decide whether separate cut-offs are justified.) The (rounded) values for the cut-offs for TCD imaging based on this 10% allowance would be:

<table>
<thead>
<tr>
<th>TCD imaging</th>
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</thead>
<tbody>
<tr>
<td>Normal velocity - ‘standard risk’</td>
<td>&lt;155 cm/s</td>
</tr>
<tr>
<td>Borderline velocity - ‘conditional’</td>
<td>155 to 179 cm/s</td>
</tr>
<tr>
<td>High velocity - ‘high risk’</td>
<td>≥180 cm/s</td>
</tr>
</tbody>
</table>

This classification should be based on the maximal velocity recorded during the examination, with appropriate regard to the settings of the scanning equipment. Adequate training should ensure that the optimum reading is taken.

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. This will 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.

Assessment of extracranial arteries are not used in the formal protocols but may be used locally to refine a diagnosis and prognosis.

Appropriate magnetic resonance or CT imaging studies to assess the extent of the cerebrovascular disease should be considered if the child is placed in the high risk category, requiring blood transfusions, although treatment should not be altered or delayed for this reason.

There will be a need for some TCD scans once a child has started on chronic transfusions to ensure that blood velocities have decreased to acceptable levels. The time intervals for performing these scans will depend on individual clinical circumstances and should be considered on a case-by-case basis.
Velocities are TCD non-imaging, time-averaged, maximal mean velocities (TAMMV). Decisions apply to TAMMVs in the distal ICA, bifurcation and/or MCA only. For bilateral or multifocal TAMMVs $\geq 170 \text{ cm/s}$, choose the highest single value for the decision tree. Recurrent inadequate scans or low velocities may indicate severe stenosis. Consider using other imaging techniques. For any particular child, detailed clinical knowledge and judgement might override this guidance.
Training

All operators performing TCD or TCDi scanning on children must have had appropriate training in the technique.

Some aspects of training will be organised nationally, incorporating three elements:

- a training day on the theory of TCD scanning, protocols, equipment, etc. with some TCD scanning practice on adults/other trainees.
- a secondment of at least one day for each trainee to a ‘centre of excellence’ to receive ‘hands on’ TCD training with children in a clinic environment. Further practice can be carried out locally but only under adequate supervision in a clinic environment.
- a visit to the trainee’s place of work by a tutor to observe their TCD scanning in practice and review traces/images, etc.

Trainees will be expected to carry out specific scanning exercises designed to develop their technique. These can be carried out on adult volunteers at their own centre.

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 scan at least 20 children to gain competence in the technique.

The Programme Centre will evaluate the need for a training manual/workbook and the resources required to produce copies for all students.

When a trainee has demonstrated a thorough understanding of the theoretical and practical aspects of TCD scanning in children, they would be ‘signed off’ by their line manager (in conjunction with their tutor) as competent to scan unsupervised. The Programme Centre will work towards establishing a national register of practitioners competent in paediatric TCD scanning and a national certificate of competence to practice.
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 those operators seeing a sufficient number of children to enable them to remain proficient in the process. The guideline number of TCD scans to achieve this proficiency is considered to be a minimum of 40 per year. It would be appropriate to consider refresher training if operators performed fewer than this number. Operators performing TCD or similar techniques in other groups of patients might justify fewer than 40 TCD scans on sickle cell children. Network centres must determine the best way to provide sufficient numbers of staff to provide a quality TCD service for the sickle cell population in their area.

The Programme Centre will work towards the establishment of a national quality assurance scheme, whereby a number of scans for each operator will be assessed for their quality by a peer review system. This is likely to be by a process of local peer review within the clinical networks or possibly by submission to an independent central body. Unsatisfactory images will result in a requirement for a further period of training.

The Programme Centre 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 hence audit the effectiveness of the scanning programme. This will also form part of a quality assurance scheme whereby centres might be identified whose TCD results or clinical outcomes differ from their peers.
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