Transcranial Ultrasound in Pediatric Patients with Sickle Cell Disease

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PURPOSE

Transcranial Doppler ultrasonography (TCD) can identify children with sickle cell disease at risk for stroke and in need of blood transfusion. Data from the Stroke Prevention Trial in Sickle Cell Anemia (STOP) shows that persistently increased TCD velocities in the distal internal carotid and/or middle cerebral arteries indicates a high risk for first time stroke. TCD detection of abnormal time averaged maximum mean flow velocities in two consecutive exams, followed by blood transfusion resulted in 90% risk reduction for first time stroke. Transcranial color duplex imaging (TCDI) can also be used to perform these exams, but variations in velocities have been reported when compared to TCD studies.

APPROPRIATE INDICATIONS

- Patients aged 2-16 years old with a diagnosis of sickle cell anemia or thalassemia
- Abnormal time averaged maximum mean velocities from previous TCD exam(s)

CONTRAINDICATIONS AND LIMITATIONS

- Children should be evaluated when healthy: hypoxia, hypercarbia, fever, sickle chest syndrome, pneumonia, hypoglycemia and other processes can result in increased cerebral blood flow velocities
- Hypocarbia and recent blood transfusions can decrease cerebral blood flow velocities. It is crucial to document if/when a transfusion has occurred
- Children must remain awake and alert during the TCD examination: sleeping results in increased CO2 and can cause increased velocities and misdiagnosis; crying may also result in hyperventilation, which can impact velocities
- Restless or agitated children may be unable to maintain appropriate positioning for a complete exam. Note any variations in exam performance in technical report.

PATIENT COMMUNICATION

Prior to beginning the exam, the sonographer or examiner should:

- Introduce self and explain why the examination is being performed and indicate how much time the examination will take.
- Verify the patient’s name and date of birth or utilize facility specific patient identifiers.
- Explain the procedure, taking into consideration the age and mental status of the child or parent.
- Respond to questions and concerns about any aspect of the evaluation.
- Refer specific diagnostic, treatment or prognosis questions to the patient's physician.
- Remove patient’s eyeglasses, contacts and/or head-coverings.
- Explain the importance of remaining awake and breathing normally throughout the exam.
PATIENT ASSESSMENT

Patient assessment should be completed before the evaluation is performed. This includes assessment of the patient's ability to tolerate the procedure and evaluation of any contraindications to the procedure. The sonographer or examiner should obtain a complete, pertinent history by interview of the patient or parent and review the patient's medical record, if available. A pertinent history includes:

- Previous cardiovascular surgeries
- Current medications or therapies
- Results of other relevant diagnostic procedures
- Review of prior examinations to ensure the evaluation duplicates prior Doppler parameters.
- Assess current medical status; patient should be without fever, cough or any symptoms of illness that could impact physiology.
- If child has had recent transfusion, document date.

PATIENT POSITIONING

Review patient positioning requirements for the examination and determine patient’s ability to maintain proper positioning for all portions of the exam:

- Sitting or supine position with head supported or stabilized for transtemporal, orbital and sub-mandibular approach.
- Turned to side with neck flexed/chin toward chest to optimize access to foramen magnum for sub-occipital (transforamenal) approach.
- Alternatively, the sub-occipital exam can be performed with patient in sitting position, arms crossed and supported by stretcher or bedside table, head resting on arms so that neck is supported and relaxed.
- Patient should be kept awake throughout the exam due to changes in CO₂ during sleep which may affect velocities and result in misdiagnosis.
- The TCD examiner should be positioned at the patient’s head with arms supported.
- Exam positions may be adapted to keep patient from moving, crying or becoming restless.

INSTRUMENTATION

Velocity reference standards for predicting stroke in children with sickle cell disease were obtained with non-imaging TCD equipment. Distinct equipment is used for TCD and TCDI, but both analyze spectral Doppler waveforms to assess flow.
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- **Transcranial Doppler Ultrasound Equipment (TCD)**
  - Utilizes a single crystal 1.5-2.5MHz pulsed Doppler probe
  - Equipment and software specifically designed for TCD applications
    - Displays all data in real time including:
      - Sample volume, sample depth
      - Time averaged maximum mean velocity
      - Peak systolic velocity/end diastolic velocity
      - Acceleration time
      - Pulsatility (PI) or resistive indices (RI)
      - Doppler power output and frequency
    - Direction sensitive Doppler blood flow meter
    - Bi-directional Doppler waveform display
    - Audible output and permanent recording of the spectral waveform

- **Transcranial Color Duplex Imaging Equipment (TCDI)**
  - Duplex imaging scanner with B-Mode, color and spectral Doppler capabilities
  - Utilizes a 1-5MHz transducer probe
    - Displays all data in real time including:
      - Sample volume, sample depth, Doppler angle
      - Time averaged maximum mean velocity
      - Peak systolic velocity/end diastolic velocity
      - Acceleration time
      - Pulsatility (PI) or resistive indices (RI)
      - Doppler power output and frequency
    - Direction sensitive Doppler blood flow meter
    - Bi-directional Doppler waveform display
    - Visual display, audible output and permanent recording of the spectral waveforms with corresponding B-Mode/Color image

**EXAM PROTOCOL**

Follow a standard imaging protocol for this examination. A complete evaluation includes spectral Doppler analysis of all accessible portions of the major intracranial arteries. Velocities can be measured by automatic tracing or manual cursor placement. Bilateral evaluations are essential for a complete evaluation.
Spectral Doppler waveforms include the following vessels and approaches (without angle correction). Waveforms are obtained at 2-5mm increments, or walked through each vessel of interest:

**Submandibular Window:**
- Distal cervical internal carotid arteries (ICA)

**Transtemporal Window:**
- Terminal internal carotid arteries (TICA)
- M1 segment of the middle cerebral arteries (MCA)
  - Should obtain proximal, mid and distal measurements
- A1 segment of the anterior cerebral arteries (ACA)
- Anterior communicating artery, if detectable (ACoA)
- P1 and P2 segments of the posterior cerebral arteries
- Posterior communicating arteries, if detectable (PCoA)

**Transforamenal/Suboccipital Window:**
- Terminal vertebral arteries (VA)
- Proximal and distal segments of the basilar artery (BA)

**Orbital Window:** (not typically utilized for this exam)
- Ophthalmic Artery (OA)
- Carotid siphon

**Technical Considerations:**
- Time averaged maximum mean (TAMM) flow velocities are used for interpretation
- Mean flow velocities are classified as normal, conditional or abnormal
- Diagnosis applies specifically to the internal carotid and middle cerebral arteries
- TCD detection of abnormal mean flow velocities on two separate exams determines the need for blood transfusions to reduce stroke risk
- Abnormal studies should be verified with a second exam, preferably within a one to two week period
- Conditional exams should be repeated within 3-6 months to document progression
- In young children, the temporal bone is easy to penetrate. It may be possible to complete most of the exam from one side. This must be documented to prevent confusion and misdiagnosis of flow direction or velocity.
• The STOP Trial did not include the orbital exam; however, if a temporal window cannot be identified, the orbital exam may be useful, as it provides access to the carotid siphon (power must be decreased for this approach).

REVIEW OF THE DIAGNOSTIC EXAM FINDINGS

The sonographer or examiner should:
• Review data acquired during the exam to ensure a complete and comprehensive evaluation has been performed and documented.
• Explain and document any exceptions or limitations to the protocol.
• Document any changes compared with previous exams.
• Record technical findings on a worksheet or other appropriate method (e.g., computer software), and classify results according to the laboratory diagnostic criteria.
• Document the exam date, clinical indications, performing sonographer and exam summary in patient’s medical record.

PRESENTATION OF EXAM FINDINGS

The sonographer or examiner should:
• Provide preliminary results when necessary as provided for by laboratory specific guidelines.
• Present record of diagnostic images, data, explanations, and technical worksheet to the interpreting physician. Interpretation must be available within two business days.
• Sonographer and interpreting physician name must appear on the final report. Finalized/signed report should be available within four business days.
• Alert vascular laboratory Medical Director or appropriate health care provider when immediate medical attention is indicated based on departmental guidelines and procedures.

EXAM TIME RECOMMENDATIONS

High quality, accurate results are fundamental elements of TCD and TCDI examinations. A combination of indirect and direct exam components is the foundation for maximizing exam quality and accuracy.

• Indirect exam components include:
  o Pre-exam activities: obtaining previous exam data, initiating exam worksheet and paperwork, equipment and exam room preparation, patient assessment and positioning, patient communication
Post-exam activities: exam room cleanup, compiling and processing exam data for preliminary and/or formal interpretation, and exam billing activities.

- Direct exam components include:
  - Equipment optimization and the actual hands-on, examination process.

- While study times may vary depending on testing protocols, patient condition, and clinical complexity of the evaluation being performed, these are the times necessary to provide a quality diagnostic evaluation. Listed are the recommended examination times for performing each CPT related to this guideline, which were derived from the direct time inputs from the Resource Based Relative Value Scale (RBRVS).
  - 93886  73 minutes
  - 93888  49 minutes
REFERENCES


