Date of examination: \_\_\_ \_\_\_ - \_\_\_ \_\_\_ \_\_\_ - \_\_\_ \_\_\_ \_\_\_ \_\_\_ (DD-MMM-YYYY)

MRI type:

1.5T Flair

3.0T Flair

Other

MRI (as read by 3 or more neuroradiologists as an adjudication panel) – Repeat as needed

No SCI

1 SCI

2 SCI

3 or more SCIs

Please specify the location(s) of the SCI(s)

**Supplemental:**

Table for recording MRI results

| Brain MRI | Date Performed  (dd-mmm-yyyy) | Age of affected | Where Performed |
| --- | --- | --- | --- |
| 1st | Data to be entered by site | [derived field] | Data to be entered by site |
| 2nd | Data to be entered by site | [derived field] | Data to be entered by site |
| 3rd | Data to be entered by site | [derived field] | Data to be entered by site |

1. Was sedation used?  Yes  No
2. General description of field of view/ anatomical positioning:
3. Magnetic field strength of scanner used:

1.5 T  3.0 T  4.0 T  7.0 T  Other: T

1. Head circumference at time of scan: cm
2. Total time in scanner (include all studies done within each particular session): :HH:MM
3. RF receiver coil(s) and number of channels: (check all that apply)

Head coil,  Neck coil,  Spine Array,  Body coil (transmit)

1. Sequences used:  T1-weighted  T2-weighted  FLAIR  Other:
2. Specify sequence name of T1 or T2 used:
3. Contrast used:  Yes  No

If yes, name of the contracts: dosage:

1. T1-MRI sequence parameters
   1. Slice orientation:  Axial  Coronal  Sagittal  Oblique
   2. Field of view: x mm2
   3. In-plane resolution: x mm2
   4. Slice thickness: mm
   5. Gap between slices: mm or % (for 2D acquisition)
   6. Number of slices:
   7. Repetition time (TR): ms
   8. Echo time (TE): ms
   9. Acquisition time: minutes
2. T2 sequence parameters (copy the following sections if parameters are different for the 2 sequences)
   1. Slice orientation:  Axial  Coronal  Sagittal  Oblique
   2. Field of view: x mm2
   3. In-plane resolution: x mm2
   4. Slice thickness: mm
   5. Gap between slices: mm or %
   6. Number of slices:
   7. Repetition time (TR): ms
   8. Echo time (TE): ms
   9. Acquisition time: minutes
3. FLAIR sequence parameters (copy the following sections if parameters are different for the 2 sequences)
   1. Slice orientation:  Axial  Coronal  Sagittal  Oblique
   2. Field of view: x mm2
   3. In-plane resolution: x mm2
   4. Slice thickness: mm
   5. Gap between slices: mm or %
   6. Number of slices:
   7. Repetition time (TR): ms
   8. Echo time (TE): ms
   9. Acquisition time: minutes
   10. Inversion time (TI): ms

## General Instructions

The MRI studies adjudication panels are to correctly diagnose SCI in sickle cell patients. It is recommended that three (3) neuroradiologist come to consensus on the findings. The examinations are independently assess presence of absence of SCI on MRI over time. Standard training is recommended for detection of SCI on this population.

For MRA studies, vessels examined are the internal carotid artery and the first segments of the anterior, middle, and posterior cerebral arteries bilaterally; each vessel is graded as: normal, mild stenosis (25%-50%), moderate stenosis (50%-75%), severe stenosis (75%-99%), or occlusion.On the initial MRA, vasculopathy is defined as any presence of moderate or worse stenosis in any vessel segment scored. Worsening vasculopathy is defined as an increase in stenosis from one category to a higher category in any vessel segment, or any new vessel occlusion.

Reference:

Liem RI, Liu J, Gordon MO, Vendt BA, McKinstry RC 3rd, Kraut MA, Strouse JJ, Ball WS, DeBaun MR. Reproducibility of detecting silent cerebral infarcts in pediatric sickle cell anemia. J Child Neurol. 2014 Dec;29(12):1685-91. doi: 10.1177/0883073813506491. Epub 2013 Dec 5. PMID: 24309240; PMCID: PMC4096057.

Comparison of Study Definition of SCI in Sickle Cell Disease

|  |  |  |
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| French | TWiTCH | SIT |
| **Stenosis was defined by MRA as at least 20% decrease in the lumen of middle cerebral artery, anterior cerebral artery, intracranial ICA, and eICA.** | **The number of vessels involved, the length (mm) and severity were assessed** at baseline and at exit. Grading of vascular occlusion based on MRA was assessed at each cerebral hemisphere (left, right bilateral) and its corresponding vessels were reported separately because they represent functionally and structurally independent systems. **Vessels occlusions of less than or equal to5 mm in length are not counted. Stenosis is scored as mild (25-49%), moderate (50-74%), severe (75-99%), or occlusion (.99%)** | **>=50-75% stenosis or occlusion on initial or after initial any worsening as defined by an increase in stenosis from one category to a higher category in any vessel segment or any new vessel occlusion. Grading as normal, mild stenosis (25%-50%), moderate stenosis (50%-75%), severe stenosis (75%-99%), or occlusion**  **Locations assessed:** vessels examined were the internal carotid artery and the first |

|  |  |  |
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|  |  | segments of the anterior, middle, and posterior cerebral arteries bilaterally |
|  | The MRA images used a 3-dimensional time of flight technique with a short echo time and included both source and maximum intensity projection reformatted images in the anterior to posterior and right to left projections to assess the patency of the intracerebral arterial vessels. All MRI/MRA studies were masked to demographic and clinical information and centrally reviewed by a single reviewer (K.J.H.) blinded to clinical site, treatment arm, and neurological status.” | MRAs were not required for enrollment in the SIT Trial, but were commonly performed, and were collected as part of the trial, when available. Each MRA was assessed for intracranial vasculopathy by two investigators and discordant reads were reviewed by a senior neuroradiologist. For MRA studies, vessels examined were the terminal portions of the internal carotid arteries (cavernous and supraclinoid segments) and the first segments of the anterior, middle and posterior cerebral arteries bilaterally; each vessel was graded as |

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|  | “Brain MRA examinations were reviewed for vessel stenoses in both the anterior and posterior circulations. A standardized scale for vessel stenosis was developed during initial reviews to provide a means of accurately assessing the location, extent, and severity of vascular disease (Figure 1). This vasculopathy scale was then used prospectively for all baseline and exit SWiTCH brain MRA examinations. Four segments of each internal carotid artery (ICA), and 3 segments of each anterior cerebral artery (ACA) and middle cerebral artery (MCA) were examined, with results recorded for a total of 20 vessel segments per subject (10 per cerebral hemisphere). Vessel stenosis was recorded by vascular location, frequency (number of stenoses), length (millimeters), and severity (percentage vessel occlusion). Severity of vessel segment occlusion was graded as mild (25-49%), moderate (50-74%), severe (75-99%), or occlusion (.99%).” | normal, or at least moderate stenosis >50% or occlusion, similar to Hulbert et al (2011) and… described (Lell et al, 2007) using vascular boundaries based on neuroanatomical landmarks (Ropper et al, 2009).” |
|  |  | “For MRA studies, vessels examined were the internal carotid artery and the first segments of the anterior, middle, and posterior cerebral arteries bilaterally; each vessel was graded as normal, mild stenosis (25%-50%), moderate stenosis (50%-75%), severe stenosis (75%-99%), or occlusion. On the initial MRA, vasculopathy was defined as any presence of moderate or worse stenosis in any vessel segment scored. Worsening vasculopathy was defined as an increase in stenosis from one category to a higher category in any vessel segment, or any new vessel occlusion. |