Supplemental Material

The following images represent the range of central nervous system complications that occur in children and adults with sickle cell disease. We have assembled over two decades of neuroimaging cases to provide some assistance to hematologists, neurologists, and neuroradiologists that are often faced with making challenging clinical decisions in individuals with sickle cell disease without an imaging reference.

Figure 1 Neuroimaging in acute headache and hemorrhage

A. Intracerebral hemorrhage in a previously well child with sickle cell anemia who presented with headache. Arteriogram was normal.

B. Subdural hemorrhage in a 9 month old infant with sickle cell anemia secondary to hyperviscous syndrome after splenic autotransfusion coupled with multiple simple transfusions over a course of 6 weeks.

C. Posterior circulation aneurysm (black arrow; suitable for coil obliteration) in an adult presenting with subarachnoid hemorrhage

D. Anterior circulation aneurysm (white arrow; too small to coil) in an asymptomatic child

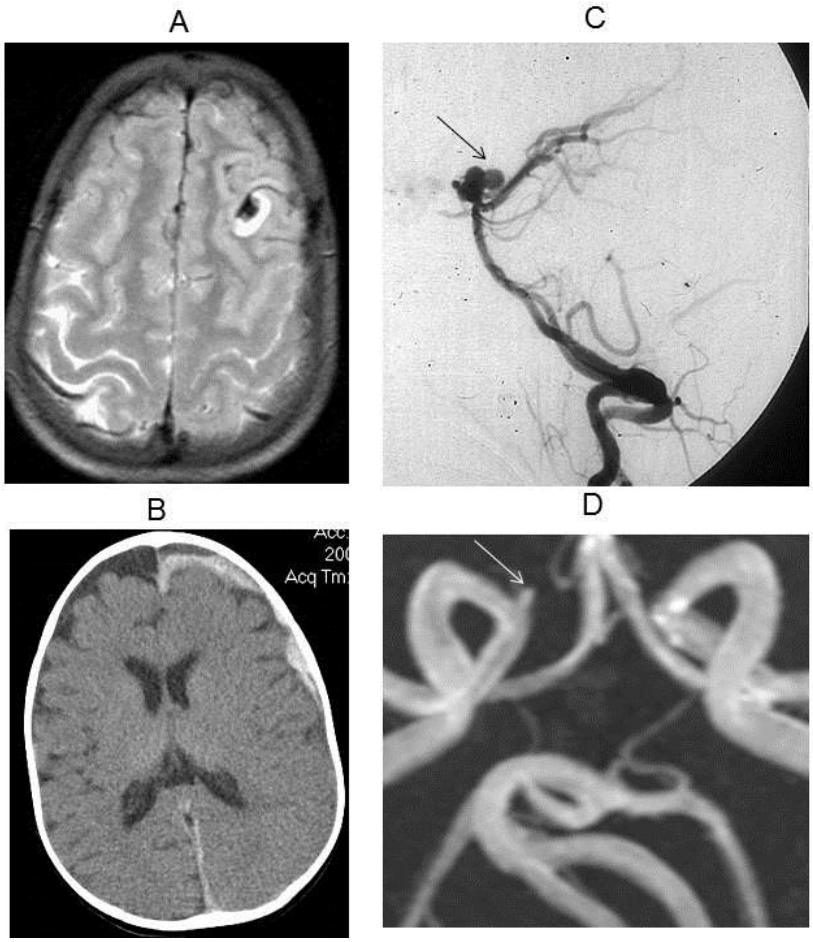
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Figure 2 Neuroimaging in patients with sickle cell disease presenting with acute seizures.

A. Signal change in the grey and white matter (arrows; posterior reversible encephalopathy syndrome) in a 9 year old boy with sickle cell anemia and nephrotic syndrome who had seizures after cyclosporin therapy

B. Bilateral borderzone ischemia in a 25 year old woman with sickle cell anemia who collapsed with seizures soon after discharge after acute chest crisis

C. Infarction in both anterior and posterior borderzones in an 8 year old boy with previously uncomplicated sickle cell anemia who developed seizures and coma after surgery to drain a painful swelling of his left cheek associated with fever

D. Left occipital infarct in a boy with sickle cell anemia who developed seizures during an acute chest crisis

E. Empty delta sign (arrow) of venous sinus thrombosis in a 18 month old child with sickle cell anemia and pneumococcal meningitis

F. Straight sinus thrombosis (arrow) in a 6 year old child with hemoglobin SC disease presenting with seizures in the context of previous neonatal post-hemorrhagic hydrocephalus

G. From the patient in panel F, widespread cerebral edema with no filling of the venous sinuses 2 days later in the same patient, who later fulfilled the clinical criteria for brain death

H. Abscess in an 18 month old boy with sickle cell anemia and seizures.

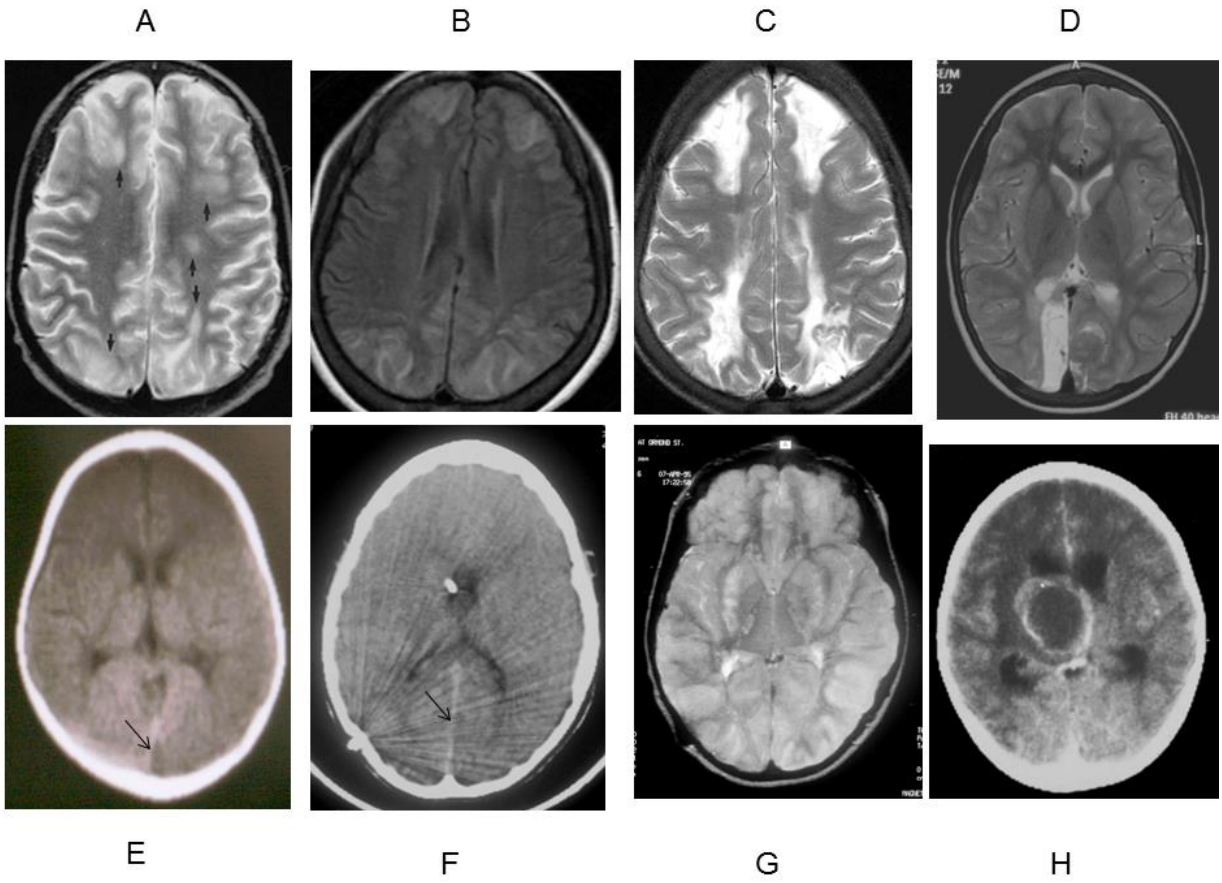
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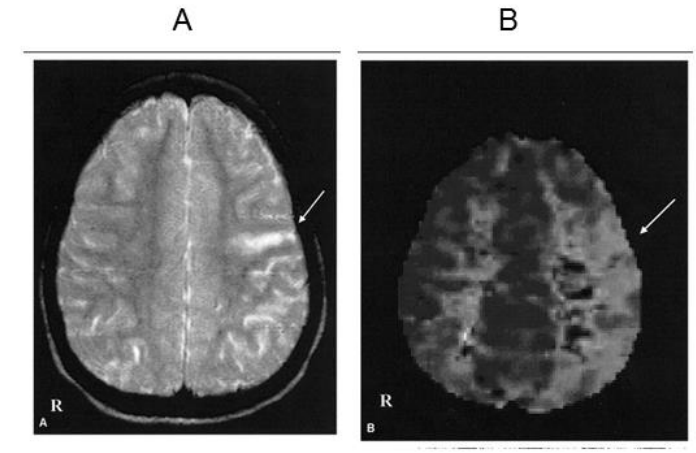
Figure 3. Neuroimaging in children with sickle cell disease and epilepsy

A. Infarct in the precentral cortex in a 7 year old girl with sickle cell anemia and a right hemiparesis and seizures (right facial twitching and blank spells). The electroencephalopgram showed epileptic activity over the left centro-temporal region

B. Mean transit time map from gadolinium perfusion imaging in the same patient shows much more widespread cortical and subcortical perfusion abnormality

C. Transcranial Doppler shows normal right and abnormal left middle cerebral artery velocities (right 137 cm/sec; left 250 cm/sec)

D. Magnetic resonance angiogram shows abnormally narrowed left middle cerebral artery.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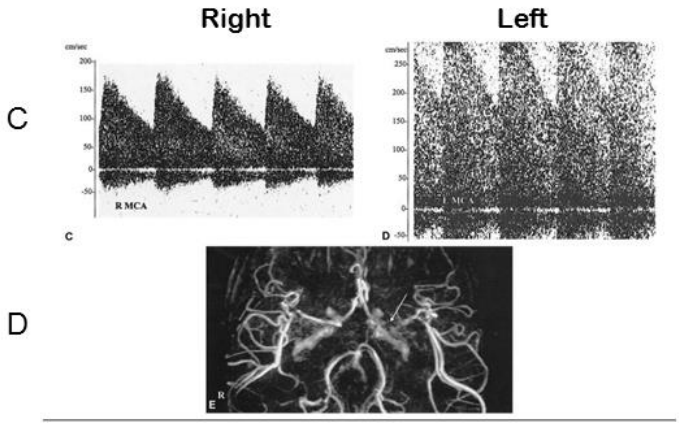
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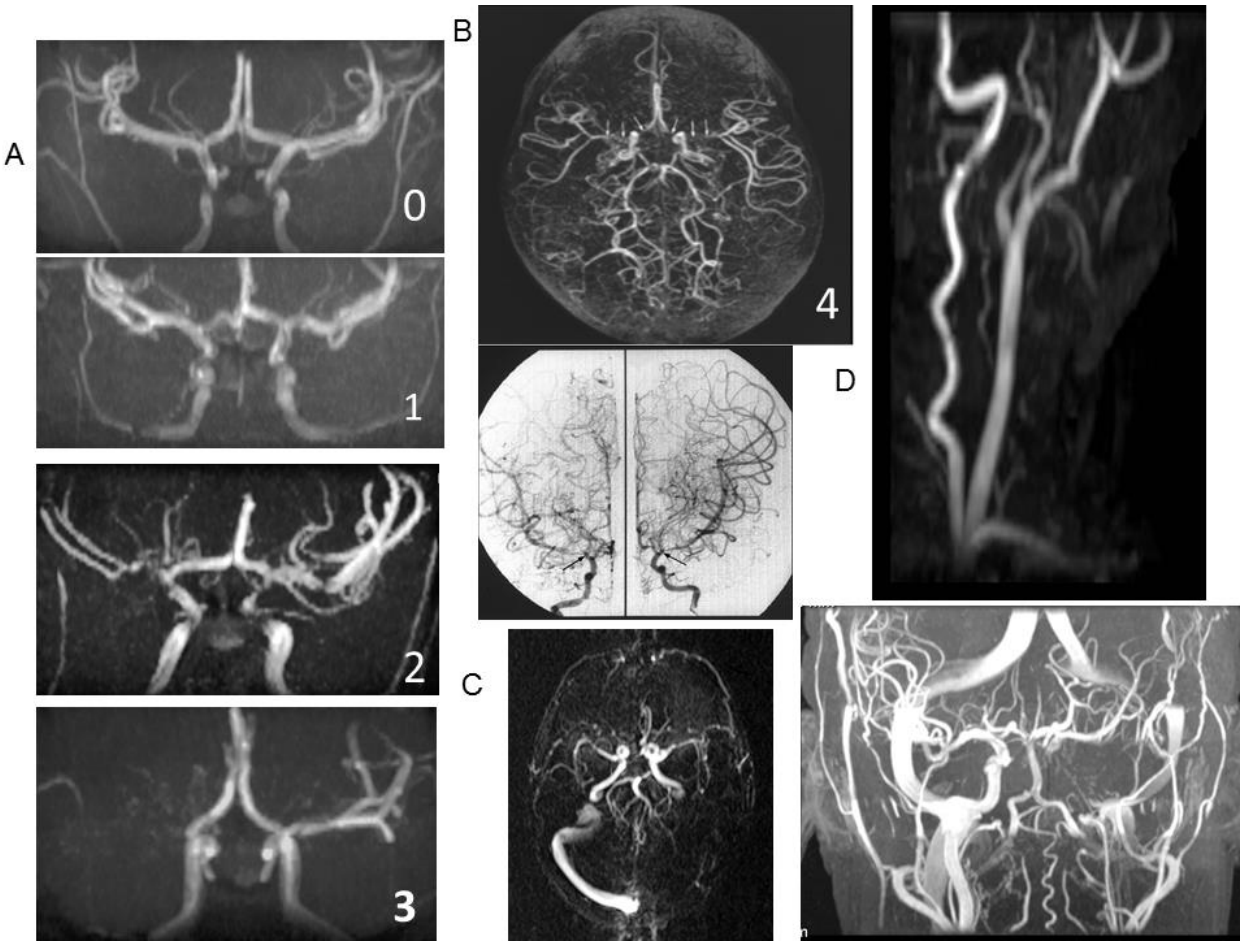
Figure 4. Vasculopathy in sickle cell disease (see also Figure 1 C and D for aneurysms)

A. Magnetic resonance angiography – 4 of the 5 grades of turbulence or signal dropout in the intracranial vessels in sickle cell anemia: (0) normal or grade 0, (1) Grade 1 turbulence in an asymptomatic child with sickle cell anemia and silent cerebral infarction, (2) Grade 2 turbulence in an asymptomatic child with sickle cell anemia and a normal MRI, (3) Grade 2 turbulence in the right terminal internal carotid artery, A1, M1 and M2 and in the left M1 in a boy with sickle beta zero thalassemia

B. Moyamoya or grade 4, the worst grade. Upper panel: Magnetic resonance angiography showing severe middle cerebral artery narrowing with moyamoya collaterals in a child with sickle cell anemia and transient ischemic attacks. Lower panel: Conventional arteriography in a child with moyamoya

C. Venogram showing transverse sinus thrombosis in a girl with sickle cell anemia presenting with seizures (Courtesy of Dr K Braun)

D. Extracranial vasculopathy. Upper panel: Tapering occlusion compatible with internal carotid dissection in a 9 year old girl with sickle cell anemia and an acute hemiparesis. Lower panel: Occlusion of the internal carotid artery in the neck in a 15 year old boy with sickle cell anemia with a past medical history of ataxia presenting with an acute hemiparesis (Courtesy of Dr P Telfer)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