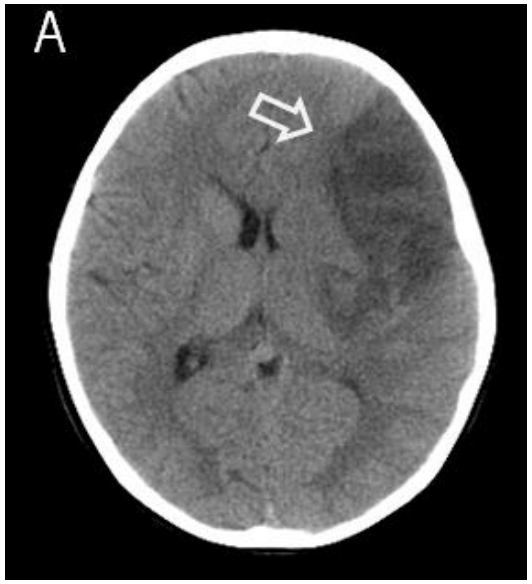
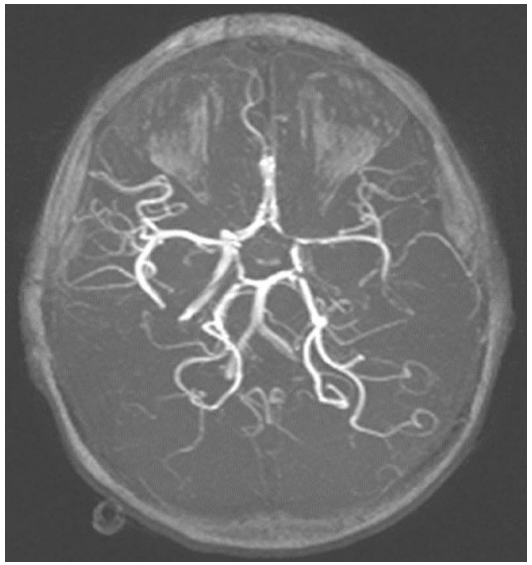
Stroke in Pediatric Sickle Cell Disease

- Sudden stoppage of blood flow to part of brain
- It is one of the major complications and is a <u>MEDICAL EMERGENCY</u>
- Used to occur in about 10% of patients (now we can prevent most strokes)
- More common in HbSS & HbSβ^0 thalassemia
- Often patients have no pain
- Symptoms: asymmetry of face, paralysis of one side or extremity, seizure, altered level of consciousness, sudden change in behavior, speech impairment
- Risk factors:
  - Silent stroke
  - High blood pressure
  - Low oxygentation and low hemoglobin
  - Genetic risk factors





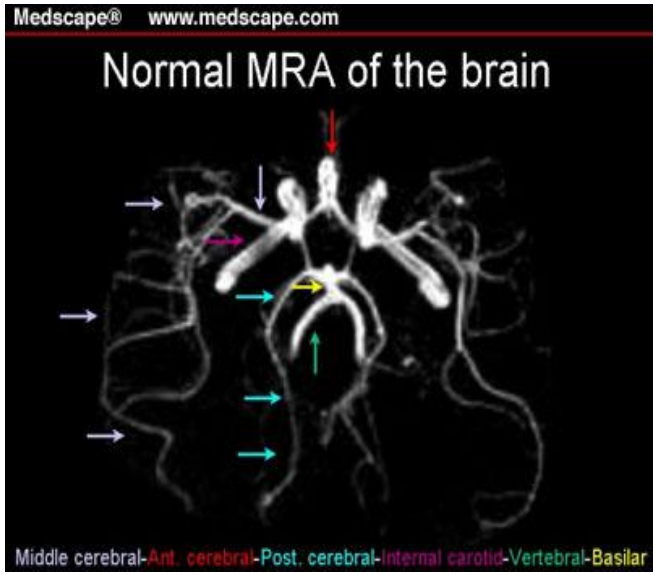
Stroke in Pediatric Sickle Cell Disease

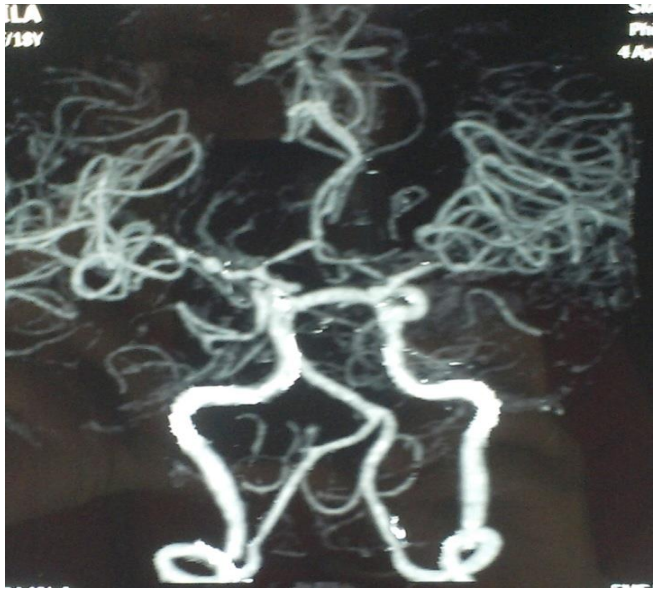
- Complications:
  - Neurological deficit
  - Cognitive impairment
  - Recurrent stroke
  - Moya moya syndrome
  - Death

- Treatment includes emergent exchange blood transfusion and supportive care afterwards

- Often patients need life-long chronic blood transfusion therapy and/or bone marrow transplantation

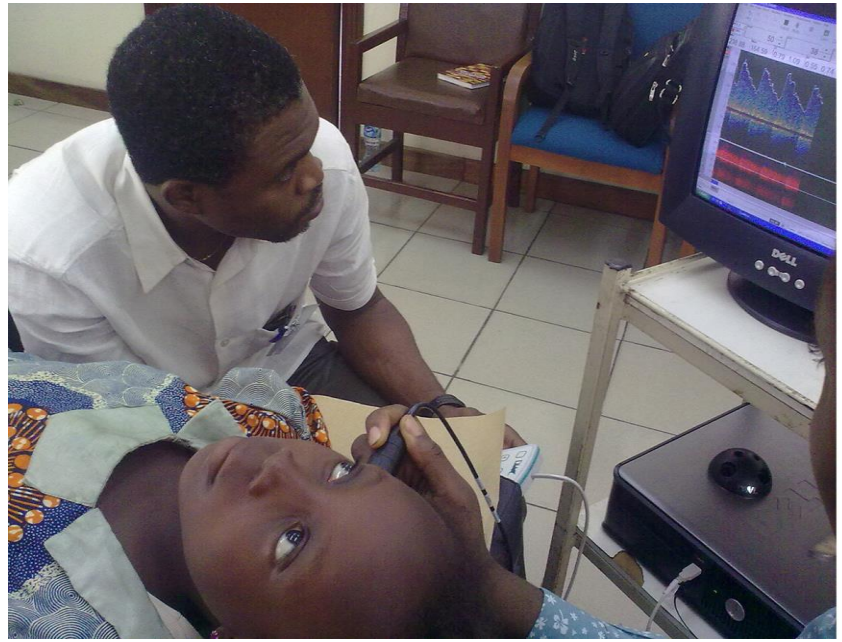
- Complications of blood transfusion:
  - Iron overload
  - Alloimmunizations
  - Recurrent stroke
  - Port placement and complications
  - Monthly appointments

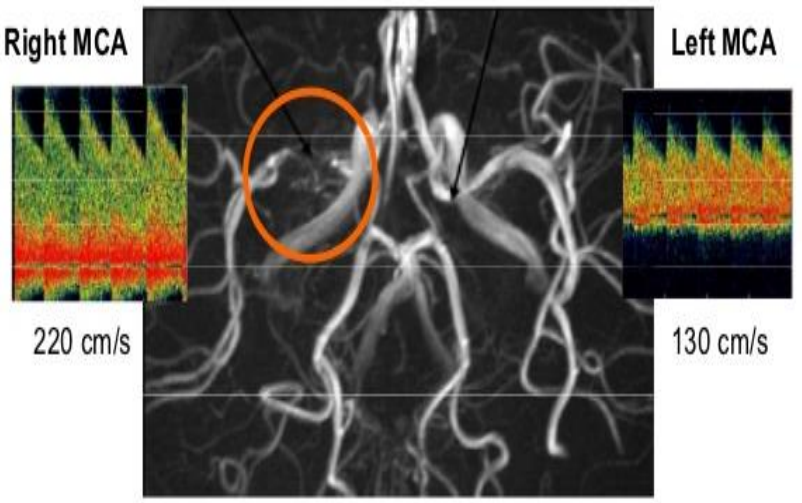




Stroke Prevention in Pediatric Sickle Cell Disease

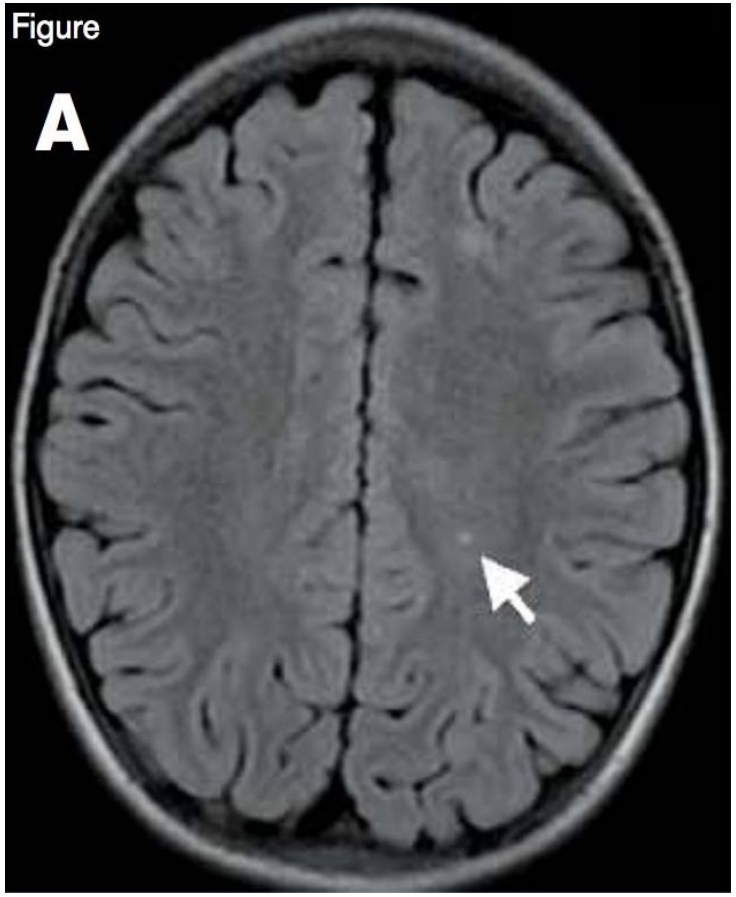
- Transcranial Doppler (TCD) is a great non-invasive screening tool
- It measures the blood flow velocity of major blood vessels in the brain
- Abnormal TCD velocity is highly predictive of developing stroke
- STOP trial showed routine implementation starting at age 2 and initiating chronic blood transfusion greatly decreased primary stroke incidence
- Recent study (TWiTCH) showed hydroxyurea seems to be a safe alternative to blood transfusion
- Studies showed that starting hydroxyurea for babies (1 year age) decrease the development of abnormal TCD





Silent Stroke in Pediatric Sickle Cell Disease

- Smaller lesions in the brain without clinical symptoms of stroke
- Occurs in 1/3rd of children with SCD and again more common in HbSS & HbSβ^0 thalassemia
- Most of these lesions occur by age 6 and most of the patients have normal TCD
- Risk factors: high blood pressure, low hemoglobin high WBC, low pain rate, history of seizure
- Complications:
  - Overt stroke,
  - Neurocognitive impairment: poor academic achievement and low employment rate
  - Increase in number of lesions with increasing age
- Chronic blood transfusion (SIT trial) seems to decrease the progression of new lesions and development of new lesions



Summary

- Clinical stroke is uncommon now due to routine implementation of TCD

- However, **stroke awareness and compliance is very important**
  - Annual TCD screening to detect patients at risk of developing stroke
  - Chronic blood transfusions/Hydroxyurea for patients with abnormal TCD
  - Chronic blood transfusions to prevent recurrent stroke
  - Hydroxyurea to prevent the development of abnormal TCD

- More research is needed to
  - Optimize blood transfusion strategies
  - Increase hydroxyurea compliance
  - Develop prevention strategies for silent stroke