Stroke Prevention Trial in Sickle Cell Anemia

The Stroke Prevention Trial in Sickle Cell Anemia (STOP) was a randomized trial to evaluate whether chronic transfusion could prevent initial stroke in children with sickle-cell anemia at high risk as determined by transcranial Doppler (TCD). The trial demonstrated a large benefit of transfusion and was halted early.

Moyamoya syndrome increases the risk of stroke in sickle cell disease, but revascularization surgery can modify this risk. Collaborative management between hematology and neurosurgery offers effective strategies to reduce stroke risk in these patients. Slingerland et al. described a challenging case where a patient with sickle cell disease undergoing standard of care management as prescribed by the Stroke Prevention Trial in Sickle Cell Anemia (STOP) and revascularization with pial synangiosis subsequently developed rapidly progressive disease in other cerebral vessels and suffered ischemic hemispheric stroke. This case demonstrates the success of management in accordance with American Heart Association (AHA) and American Stroke Association (ASA) guidelines, but also demonstrates critical areas where we lack understanding of disease progression ¹⁾.

1)

Slingerland AL, Karsten MB, Smith ER, Sobota AE, See AP. Two Sides of a Coin: Case Report of Unilateral Synangiosis and Contralateral Stroke Highlighting Consequences of Disease Progression and Efficacy of Revascularization in Sickle Cell Disease Associated Moyamoya Syndrome. Acta Haematol. 2021 Dec 8. doi: 10.1159/000521361. Epub ahead of print. PMID: 34879377.

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