TRANSCRANIAL DOPPLER AND CT ANGIOGRAPHY OF THE CEREBRAL ARTERIES IN EGYPTIAN SICKLE CELL PATIENTS

Thesis

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بِسْمِ اللَّـهِ الرَّحْمَـٰنِ الرَّحِيمِ

نَرْفَعُ دَرَجَاتٍ مَّن نَّشَاءُ وَفَوْقَ كُلِّ ذِي عِلْمٍ عَلِيمٌ

صدق الله العظيم

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Abstract

Sickle cell disease (SCD) is a life-threatening genetic disorder. Children with sickle cell disease are at risk of a neurological complication over their lifetime. To identify those children at risk, TCD can be used to measure blood flow velocity in the large arteries of the circle of Willis and CTA in detection of the large vessels stenosis and occlusions.

Key Words:

Sickle cell disease, Trans-cranial Doppler (TCD), CT Angiography (CTA).

Contents

List of Abbreviations

ACA	**Anterior Cerebral Artery**
ACOM	**Anterior communicating**
ACS	**Acute chest syndrome**
ASL	**Arterial spin labelling**
BA	**Basilar Artery**
CBF	**Cerebral blood flow**
CBFV	**Cerebral blood flow velocity**
CVSOD	**Cerebrovascular steno-occlusive disease**
DSA	**Digital Subtraction Angiography**
EDV	**End-diastolic velocity**
FLAIR	**Fluid-attenuated inversion–recovery**
HU	**Hydroxyurea**
HR	**Heart rate**
HRCT	**High-resolution computed tomography**
ICA	**Internal carotid artery**
MCA	**Middle cerebral artery**
MCV	**Middle cerebral artery Velocity**
MHz	**Mega hertz**
MIP	**Maximum intensity projection**
MMD	**Moyamoya disease**
MRA	**Magnetic resonance angiography**
MRI	**Magnetic resonance imaging**
MFV	**Mean flow velocity**
OA	**Ophthalmic artery**
PCA	**Posterior Cerebral artery**
PCOM	**Posterior communicating branch**

PI	**Pulsatility index**
PSV	**Peak systolic velocity**
RI	**Resistance index**
SCD	**Sickle cell disease**
SCI	**Silent cerebral infarct**
S/D	**Systolic diastolic ratio**
STOP trial	**Stroke Prevention Trial**
TAMM	**Time-averaged mean of the maximum velocities**
TCD	**Transcranial Doppler ultrasound**
TOF	**Time-of-flight**
VA	**Vertebral Artery**
VOCs	**Vaso-occlusive crises**

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INTRODUCTION

Sickle cell disease is a common inherited blood disorder that is characterized by the presence of sickle-shaped red blood cells. The clinical manifestations of SCD vary, but they may be attributed to three mechanisms: vaso-occlusion, chronic hemolytic anemia, and infection. **(Saito et al, 2010)**

The molecular pathologic abnormalities of SCD lead to microvascular occlusion and intravascular hemolytic anemia. Microvascular occlusion is related to painful episodes and probably causes microcirculatory problems in the brain. The most commonly recognized stroke syndrome in children with SCD is large-artery infarction. These "big strokes" are the result of a vascular process involving the large arteries of the circle of Willis leading to territorial infarctions from perfusion failure or possibly artery-to-artery embolism **(Adams, 2007)**

It is believed that damage to the intima leads to vascular stenosis and occlusion. Intimal damage may result from high velocity blood flow, the shape of the sickle cells, adherence of RBCs to the endothelium, endothelial damage, intravascular sludge, intimal hyperplasia, or thrombosis . Endothelial damage also may lead to aneurysm formation .Extracranial ICA stenosis is rare, whereas intracranial arterial stenosis that leads to moyamoya vessels is common and well recognized **.(Saito, et al, 2010)**

The Transcranial Doppler ultrasonography (US) is a noninvasive, portable technique for evaluating the intracranial vasculature. it has been

proved useful for evaluating intracranial vasculature in patients with SCD **(Kirsch et al, 2013)**

Transcranial Doppler ultrasonography can detect children who are developing cerebral vasculopathy. TCD measures blood flow velocity in the large arteries of the circle of Willis. Velocity becomes elevated in a focal manner when stenosis reduces the arterial diameter. Children with SCD who are developing high stroke risk can be detected months to years before the stroke using TCD **(Adams, 2007)**

The Stroke Prevention Trial in Sickle Cell Anemia (STOP) (1995-2000) was halted prematurely when it became evident that regular blood transfusions produced a marked (90%) reduction in first stroke. Children were selected for STOP if they had 2 TCD studies with velocities of 200 cm/s or greater. Children not undergoing transfusion had a stroke risk of 10% per year, which was reduced to less than 1% per year by regular blood transfusions **(Adams, 2007)**

An alternative method of reducing the production of Hb SS-containing RBCs is the induction of fetal hemoglobin (Hb F) by the native marrow. Hydroxyurea, a cytotoxic agent, increases production of Hb F. Hydroxyurea itself inhibits endothelial adhesion of RBCs, mildly increases overall hemoglobin concentration, and mildly suppresses neutrophil production, which may also ameliorate the effects of SCA . Hydroxyurea has been shown to decrease the incidence of ACS, painful crises, hemolytic crises. **(Lonergan et al, 2001)**

Advances in CT technology are having profound impact on imaging children and have made CT angiography possible even in neonates. (CT) has proven capable of offering a detailed analysis of the