

Radiologic MR Findings of Inner Ear and
Its Relation to Hearing Loss in Patients
with Sickle Cell Anemia

Thesis

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List of Abbreviations

Abb.	Full term
5-FU	5-fluorouracil
ACA.....	Anterior cerebral artery
ACS.....	Acute chest syndrome
BCL11A,	B-cell lymphoma/leukemia 11A
BCL11A,	B-cell lymphoma/leukemia 11A
BNP	A brain natriuretic peptide
BNP	A brain natriuretic peptide
CAPD.....	Central auditory processing disorder
chRR	Chronic relative reticulocytopenia
CMV.....	Cytomegalovirus
CNS.....	Central nervous system
CT	Computaried tomography
CVA.....	Cerebrovascular accident
dB.....	Decibel
EPO.....	Erythropiotin
FLAIR.....	The fluid attenuation inversion recovery technique
G-CSF	Granulocyte colony-stimulating factor
Hb	Hemoglobin
HbS	Hemoglobin S
HHI.....	Hereditary hearing impairment
ICA.....	Internal carotid artery
ICH	Intracerebral hemorrhage

List of Abbreviations (Cont...)

Abb.	Full term
IgG	Immunoglobulin G
IgM.....	Immunoglobulin M
MCA.....	Middle cerebral artery
MR	Magnetic resonance
MRA.....	Magnetic resonance angiography
MRI.....	Magnetic resonance imaging
NO.....	Nitric oxide
PCA.....	Posterior cerebral artery
PCV13.....	The 13-valent pneumococcal conjugate vaccine
PCV13.....	The 13-valent pneumococcal conjugate vaccine
SCA.....	Sickle cell anemia
SCD.....	Sickle Cell Disease
SNHL.....	Sensorineural hearing loss
SSNHL.....	Sudden sensorineural hearing loss
STOP.....	Stroke prevention trial in sickle cell anemia
TCD.....	Transcranial Doppler
TIA.....	Transient ischemic attack



Introduction



INTRODUCTION

Homozygous sickle cell anemia (SS) is the most severe form of sickle cell disease (SCD), but it is phenotypically variable, and the course of disease is difficult to predict. Some individuals with sickle cell anemia have frequent vaso-occlusive complications and die prematurely (*Quinine, 2007*).

SCD vaso-occlusive phenomenon has been described as a complex event with the participation of stressed reticulocytes, sickled erythrocytes, leukocytes, platelets and endothelium activation (*Stuart and Nagel, 2004*).

In a study of sensorineural hearing loss in sickle cell patients in France despite high medical standards. A hearing loss was found of greater than 20 dB at two or more frequencies was found in 45.57 % of sickle cell patients, 47.22 % HbSC patients and 43.59 % of HbSS patients. The hearing loss in HbSS patients was neural in nature and of earlier onset (*Jovanovic-Bateman and Hedreville, 2006*). The most accepted pathogenesis of SNHL is recurrent vaso-occlusion of the labyrinthine blood vessels—either in the distribution of the anteroinferior cerebellar artery or a branch of the basilar artery—which can result in labyrinthine hemorrhage (LH) and labyrinthitis ossificans (LO). Labyrinthine hemorrhage is thought to result from altered capillary hemodynamics or

reperfusion injury (*Whitehead et al., 1998*). In another study of inner ear radiologic findings in patients with SCD, LH and LO were found in approximately one-third of patients with inner ear symptoms and preferentially affected males (*Saito et al., 2011*).