Study of Retinal and Choroidal Changes in Children with Sickle Cell Disease using Optical Coherence Tomography and Angiography

Thesis

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List of Abbreviations

Abb.	Full term
CBC	Complete blood count
CC	Choriocapillaris
DCP	Deep capillary plexus
DVC	Deep vascular complex
FAZ	Foveal avascular zone
FFA	Fundus fluorescein angiography
GCC	Ganglion cell complex
HbS	Hemoglobin sickle
ICGA	Indocyanine green angiography
ICP	Intermediate capillary plexus
ILM	Internal limiting membrane
IOP	Intraocular pressure
IS/OS	Inner segment/ outer segment
NPSR	Non-proliferative Sickle retinopathy
OCTA	Optical coherence tomography angiography
ONH	Optic nerve head
PAMM	Paracentral acute middle maculopathy
pRNFL	Peripapillary retinal nerve fiber layer
PSR	Proliferative sickle cell retinopathy
RPCP	Radial peri-papillary plexus
SCD	Sickle cell disease
SCP	Superficial capillary plexus
SCR	Sickle cell retinopathy
SD-OCT	Spectral domain optical coherence tomography
SNR	Signal to noise ratio

List of Abbreviations Cont...

Abb.	Full term
SSADA	Split-spectrum amplitude-decorrelation angiography
SS-OCT	Swept-Source OCT
SVC	Superficial vascular complex
TD-OCT	Time domain-OCT
VD	Vessel density
VEGF	Vascular endothelial growth factor

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INTRODUCTION

Sickle cell disease is an inherited autosomal recessive hematological disease caused by a single point mutation in the beta globin gene that results in the formation of abnormal hemoglobin (HbS). Patients may inherit two sickle genes resulting in HbSS genotype (sickle cell disease), or one sickle gene in combination with a beta globin gene variant resulting in HbSC (sickle cell trait) or HbSb thalassemia genotype ***(Fox et al., 1990).*** Under states of deoxygenation, this abnormal hemoglobin aggregates resulting in sickling of the erythrocytes in which they acquire a rigid elongated form that can disrupt the blood flow in small vessels and cause vascular occlusions ***(Ware et al., 2017).*** These vascular occlusions can occur anywhere in the body resulting in various systemic manifestations.

Microvascular insults that occur in the eye result in sickle cell retinopathy. Vaso-occlusion of the retinal vessels leads to ischemia and may trigger the production of vascular endothelial growth factor (VEGF) resulting in pathologic neovascularization. This can cause vision loss due to vitreous hemorrhage, traction retinal detachment, retinal vascular occlusions or neovascular glaucoma.