Detection of Prolonged QT Interval Among Sickle Cell Diseased Children

Thesis

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Abstract

Prolonged corrected QT interval (QTc) is not uncommon among patients with sickle cell disease (SCD). **Aim of the work:** we aimed at detection of cases of SCD with prolonged QTc, correlate QTc with estimated systolic pulmonary artery pressure (ESPAP), serum ferritin and other echocardiographic dimensions. **Patients& methods:** This is a cross sectional descriptive study that included 46 SCD patients attending the hematology clinic at the new Children's hospital, Faculty of Medicine, Cairo University during the period of March 2014 to October 2015. Cases were subjected to full history, clinical examination and laboratory investigations in addition to 12 leads ECG and transthoracic color Doppler echocardiography. **Results:** The mean age of the patients was 8.93 ± 5.26 years. Prolonged QTc interval was detected in19.57% with normal QT dispersion and no significant arrhythmias. Pulmonary hypertension was detected in 13.04%. There was no significant correlation between ESPAP and QTc (P=0.990, r=0.002). There was a significant positive correlation between QT dispersion and diameter of interventricular septum (IVS) (P=0.017, r=0.350). Also there was a significant weak positive correlation between QTc and the diameter of left atrium (LA) (P=0.047, r=0.295). The left ventricular end diastolic diameter (LVIDd) correlated significantly with serum ferritin (P=0.44, r=0.301).**Conclusion:** Prolonged QTc interval (> 440 msec.) was detected in 19.57 % of SCD cases. This increases the risk of arrhythmias that could be fatal and might necessitate beta blocker management. Pulmonary hypertension (≥ 36 mmHg) was not a relatively common complication among patients with SCD in this study. It was detected in 13% of patients.

Key words: prolonged QTc, sickle cell disease, pulmonary hypertension.

Contents

List of Abbreviations

µm	Micrometer
6MWD	6-Minute Walk Distance
ACS	Acute Chest Syndrome
ADMA	Asymmetric Dimethylarginine
AHRQ	Agency for Healthcare research and Quality
ANA	Anti-Nuclear Antibody
AO	Aorta
ATG	Anti Thrombocyte Globulin
AUC	Area Under the Cover
B19V	Parvovirus B-19
BMI	Body Mass Index
CBC	Complete blood count
CBT	Cord Blood Transplantation
CERHR	Center for the Evaluation of Risks to Human Reproduction
CRP	C-reactive protein
CSF	Cerebrospinal Fluid
CSSCD	Cooperative Study of Sickle Cell Disease
CXR	Chest X-Ray
dl	deciliter
DNA	Deoxyribonucleic acid
DS	Doppler Sonography
ECG	Electrocardiogram
eNOS	endothelial Nitric Oxide Synthase
ER	Emergency Room
ESPAP	Estimated Systolic Pulmonary Artery Pressure
ET-1	Endothelin-1
FS	Fractional Shortening

G-6-PD	Glucose 6-Phosphate Dehydrogenase
GM-CSF	Granulocyte Macrophage Colony Stimulating Factor
GVHD	Graft Versus Host Disease
Hb	Hemoglobin
Hb AS	Sickle Cell Trait
Hb F	Fetal Hemoglobin
HbS	Sickle Hemoglobin
Hct	Hematocrit
HDAC	Histone Deacetylase
HIV	Human Immunodeficiency Virus
HLA	Human Leukocyte Antigen
HPFH	Hereditary Persistence of Fetal Hemoglobin
HPLC	High-Performance Liquid Chromatography
HU	Hydroxyurea
ICA	Internal Carotid Artery
IEF	Iso Electric Focusing
IgM	Immunoglobulin M
ISCs	Irreversibly Sickled Cells
IV	Intravenous
IVS	Inter Ventricular Septum
kg	Kilogram
LA	Left Atrium
LDH	Lactate Dehydrogenase
LFTs	Liver Function Tests
LMWH	Low molecular weight heparin
LV	Left Ventricle
LVIDd	Left Ventricular Internal Diameter end diastole
LVIDs	Left Ventricular Internal Diameter end Systole
LVPW	Left Ventricular Posterior Wall

MCA	Middle Cerebral Artery
MCV	Mean Cell Volume
metHb	Met Hemoglobin
mg	Milligram
mm Hg	Millimeter of mercury
mPAP	mean Pulmonary Artery Pressure
MPI	Myocardial Performance Index
msec	Millisecond
MSH	Multi-Centers Study for Hydroxyurea
NADPH	Nicotinamide Adenine Dinucleotide Phosphate
ng	Nanogram
NHLBI	National Heart, Lung and Blood Institute
NIH	National Institute of Health
NO	Nitric Oxide
$NO3^-$	Nitrate
NOS	Nitric Oxide Synthase
NSAID	NonSteroidal Anti-Inflammatory Drug
NT-proBNP	N-terminal pro–B-type natriuretic peptide
O2	Oxygen
ONOO	Peroxynitrite
PAWP	Pulmonary Artery Wedge Pressure
PCV7; Prevnar	7-valent pneumococcal conjugate vaccine
pg	Picogram
PH	Pulmonary Hypertension
PO2	Partial Pressure of Oxygen
PUSH	Pediatric Hypoxic Response
PVR	Pulmonary Vascular Resistance
QTc	Corrected QT interval

RBCs	Red Blood Corpuscles
RNA	Ribonucleic Acid
ROC	Receiver Operating Characteristic
ROS	Reactive Oxygen Species
RV S'	Right Ventricle Systolic function
RVSP	Right Ventricular Systolic Pressure
SCA	Sickle Cell Anemia
SCD	Sickle Cell disease
SD	Standard Deviation
SS	Sickle Hemoglobin
TAMMvel	Time-averaged Mean Velocity
TCD	Trans Cranial Doppler
TDI	Tissue Doppler Imaging
TGF	Transforming Growth Factor
TIAS	Transient Ischemic Attacks
TR	Tricuspid Regurge
TRM	Transplant Related Mortality
TRV	Tricuspid Regurgitation Velocity
VCAM	Vascular Cell Adhesive Molecule
VOC	Vaso-Occulusive Crisis
Walk-PHASST	Walk–Treatment of Pulmonary Hypertension and Sickle Cell Disease with Sildenafil Therapy
WBC	White Blood Cells
WHO	World Health Organization
XO	Xanthine Oxidase

List of Tables

List of Figures