



Cardiac Involvement In Children With Inborn Errors Of Metabolism

Thesis

Submitted for partial fulfillment of medical doctorate in Pediatrics

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Abstract

Background: The heart is a metabolically active organ and it can be adversely affected by inborn errors of metabolism.

The aim of the present study: was to evaluate cardiac involvement in a group of infants and Children with Inborn Errors of Metabolism.

Subjects and methods: This study was performed in Fayoum university hospital in the period from October 2021 to May 2023. A total number of 100 children with inborn errors of metabolism were included in the study. All patients were subjected to ECG, conventional echocardiography and tissue Doppler echocardiography.

Results: our study included a total number of 100 cases with inborn errors of metabolism. Our studied cases were diagnosed with different metabolic disorders where GSD was the largest group forming 27% of cases, Gaucher disease were 19%, MPs were 14% and Wilson disease were 9%. Miscellaneous cases were 31%. ECG revealed that all cases had regular rhythm and 17% had LVH and Prolonged QT interval was found in 31% of cases. Echocardiography showed that mitral regurge was present in 12% of cases and AS was present in 3% of cases. As for congenital heart disease (CHD) was present in 14%. pulmonary HTN was present in 14% of cases and LVH was present in 18% of cases. and 8% of our cases had impaired systolic function.

Conclusion: Cardiac assessment in children with inborn errors of metabolism by ECG and echocardiography is significant and essential.

Key words: Inborn errors of metabolism, heart, valvular defects, cardiomyopathy, ECG and echocardiography.

