Hypoglycemia in Mitochondrial Disorders

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The electron transport chain (ETC) in mitochondria functions to produce energy in the form of adenosine triphosphate (ATP). Defects in the mitochondrial or nuclear DNA that codes for components of the ETC lead to mitochondrial disorders (MTDs). MTDs are multi-system conditions affecting the heart, muscles, and especially brain. The endocrine system is commonly affected in MTDs, and diabetes and hyperglycemia are established secondary diagnoses. Rates of non-iatrogenic hypoglycemia have not been studied in individuals with MTDs. This study aims to investigate the frequency of hypoglycemia in patients with MTDs. Individuals diagnosed with a ‘definite’ or ‘probable’ MTD according to the modified Walker criteria at The University of Texas, Mitochondrial Center of Excellence were included in this study. Exclusion criteria included diagnosis of diabetes or adrenal insufficiency or past or present use of hydrocortisone or prednisone. Patient charts were reviewed retrospectively for blood glucose values. Individuals with at least two values were recorded. Patients were classified as neonatal (≤28 days of life) or non-neonatal (>28 days of life) at the time of measurement. Data analysis included descriptive statistics, mixed-model regression, and two-sample tests of proportion. All data analysis was done using Stata® (v.13, College Station, TX). Statistical significance was assumed at $p<0.05$. Of the 116 patients included in this study, 22 (18.97%) experienced at least one episode of hypoglycemia. This is significantly higher ($p<0.05$) than the 6% general population rate of hypoglycemia. Neonatal readings were also found to be 30mg/dL lower than non-neonatal readings, on average, a significant difference ($p<0.05$). Patients with MTD are more likely to experience hypoglycemia compared to the general population with especially low blood glucose readings during the neonatal period. This demonstrates hypoglycemia may be contributing to the high rate of neurological symptoms reported in MTDs and supports that MTDs should be in the differential diagnosis in cases of hypoglycemia, especially during the neonatal period. Additional and earlier monitoring could reduce negative outcomes such as decreased cognitive outcome, developmental delays, seizures, or brain damage in patients with MTDs.

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