GENOTYPE-BASED SEVERITY SCORING SYSTEM IN WOLFRAM SYNDROME

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Value Proposition: *Disease severity scoring system that can predict the progression of Wolfram syndrome.*

Technology Description

Researchers at Washington University in St. Louis have developed a genotype-based severity scoring system to predict the onset of cardinal symptoms in patients with Wolfram syndrome. Wolfram syndrome is a rare genetic disorder characterized by juvenile-onset diabetes mellitus, optic atrophy, hearing loss, and diabetes insipidus, with significant variability in disease severity. This scoring system provides valuable insights into the progression of Wolfram syndrome.

Stage of Research

Assessed the accuracy of a genotype-based severity scoring system in 324 patients with documented onset ages for diabetes mellitus.

Publications

Lee EM, Verma M, Palaniappan N, Pope EM, Lee S, Blacher L, Neerumalla P, An W, Campbell T, Brown C, Hurst S, Marshall B, Hershey T, Nunes V, de Heredia ML, Urano F. Genotype and Clinical Characteristics of Patients with Wolfram Syndrome and WFS1-related Disorders. medRxiv [Preprint]. 2023 Feb 16:2023.02.15.23284904. doi: 10.1101/2023.02.15.23284904. Update in: Front Genet. 2023 Jun 21;14:1198171. doi: 10.3389/fgene.2023.1198171. PMID: 36824811; PMCID: PMC9949199.

Applications

- Treatment of Wolfram syndrome

Key Advantages

- Accurately predicts onset and severity of symptoms

Related Web Links – Fumihiko Urano Profile; Urano Lab