# Maternally Inherited Diabetes and Deafness: Insights Into Mitochondrial Diabetes

*S. RAFI, H. OUAKRIM, G. El MGHARI, N. El ANSARI Department of Endocrinology, Diabetology, Metabolic Diseases and Nutrition Mohammed VI university hospital of Marrakech, Faculty of medicine and pharmacy of Marrakech, Cadi Ayyad University, Marrakech, Morocco*

**Abstract:**

**Background:**

Mitochondrial diabetes represents about 1% of diabetes but still very often misunderstood. The most frequent mutation is 3243A >G of the mtDNA, which is responsible for the maternally inherited diabetes and deafness syndrome (MIDD).

**Objective**:

We report an observation of a patient with strong suspicion of mitochondrial diabetes with the aim of highlighting the characteristics of this type of diabetes.

**Case Report:**

43-year-old female patient, with diabetes for 5 years on an Insulin Therapy.
Anamnesis: Diabetes discovered at the Age of 38 years by an acid ketosis decompensation. Strong heredity of diabetes: grand-mother, two uncles, two aunts, tree cousins, all on the mother's side. Hypoacusis in an uncle and two sisters on the mother's side. Blindness in grandmother and diabetic retinopathy in mother Diabetic nephropathy in aunt on the mother's side. Clinical examination is without particularity with BMI at 37.86 kg/m2.

Diabetes typing immunological test is negative

**Discussion And Conclusion:**

Various mutations or deletions in mitochondrial DNA (mtDNA) can lead to multi-organ syndromes, including diabetes. Maternally Inherited Diabetes and Deafness (MIDD) is the most prevalent form of mitochondrial diabetes. The 3243A >G mutation affects tRNA's tertiary structure, causing cellular energy deficits, primarily affecting highly metabolically active organs like the pancreas. Mitochondrial diabetes should be suspected when diabetes occurs at a young age, is maternally inherited, and coexists with extra-pancreatic conditions like neurosensory deafness, reticular macular dystrophy, neurological and muscular issues, and cardiomyopathy. Mutation testing confirms the diagnosis, assesses maternal transmission risk, and offers genetic counseling to relatives.
Understanding mitochondrial diabetes is crucial for early detection, comprehensive organ damage screening, specialized multidisciplinary care, and genetic counseling.

**Keywords**:

Mitochondrial diabetes, maternally inherited, Mutation testing