Maternally inherited diabetes and deafness (MIDD)

Maternally inherited diabetes and deafness, also known as MIDD, is a rare mitochondrial condition that slows down the mitochondria's ability to make proteins. This causes the mitochondria to not work as well as they should, and is thought to impact the body's ability to trigger the release of an important hormone called insulin.

Mitochondria are the parts of a cell that help turn the energy we get from food into energy that the body can use. They are also important in the communication between body parts and creating other materials the body needs. Mitochondrial conditions can cause a variety of signs and symptoms in many parts of the body, particularly those that use a lot of energy like muscles and the brain.

Alternate names

- MIDD;
- Diabetes and deafness, maternally inherited;
- Ballinger Wallace syndrome;
- Diabetes mellitus type II with deafness;
- Diabetes-deafness syndrome, maternally transmitted;
- non insulin-dependent diabetes mellitus with deafness;
- NIDDM with deafness

Cause and Genetics

MIDD is an inherited genetic condition, meaning it is passed down in a family. MIDD is caused by changes in the mitochondrial genome (mtDNA), or set of DNA contained in the mitochondria of a cell. Mitochondria are inherited from the mother through her eggs. People typically do not inherit any mitochondria from sperm, making it very unlikely for mitochondrial conditions to be passed down from a father. Both males and females can have MIDD.

Changes, sometimes called mutations or variants, in several genes can cause MIDD. These include MT-TL1, MT-TE, and MT-TK. The most common cause of MIDD is a change in the gene MT-TL1 called 3243A-G.

Frequency

Around 1% of people with diabetes have MIDD. MIDD occurs around the world, but is most common in Japan.

Signs and Symptoms

MIDD causes a range of signs and symptoms. These may differ greatly, even among members of the same family, so always check with your provider if new symptoms appear or you are concerned.

The hallmark symptoms of MIDD are diabetes and hearing loss. Hearing loss usually occurs in mid-adulthood and especially affects high tones. Diabetes usually occurs after hearing loss begins, but also usually in mid-adulthood.

Some people with MIDD may have additional signs and symptoms similar to those that can be seen in other mitochondrial conditions. These may include:

- An eye condition called macular retinal dystrophy, which are colored patches on the back of the eye. This does not usually cause vision concerns.
- People with MIDD sometimes have other eye features including vision loss, night blindness, blind spots, and spots of bright light.
- Dropping of the eyelid (ptosis)
- An enlarged heart (cardiomyopathy) or other heart features
- Muscle weakness (myopathy) or cramps, especially after exercise
- Kidney conditions
- Constipation or other gastrointestinal disease
- Mental health symptoms
- Stroke
- Shorter height than family members and peers

Diagnosis

- MIDD can be diagnosed by:
- Measuring biochemical markers in blood and urine
- Hearing evaluation
- Performing a genetic test to look for changes in genes known to cause MIDD

MIDD is not included on newborn screening panels. If there is a known family history of MIDD, or if a mother is a known carrier, prenatal testing can be performed on amniotic fluid (the fluid surrounding a baby) or chorionic villi (a specific part of the placenta). Results interpretation of this testing for mitochondrial conditions is complicated and genetic counseling is recommended.

Treatment and Management

Before beginning any treatment or therapy, please consult with your physician.

There is currently no FDA-approved therapy for MIDD. Treatment and management of MIDD is symptomatic and supportive. This may include:

- Individualized treatment of diabetes
- Hearing aids or cochlear implants for hearing loss
- Avoidance of mitochondrial toxins like certain drugs, tobacco and alcohol
- Mitochondrial supplements
- Special schooling arrangements depending on age of onset

Individuals living with MIDD typically work with several healthcare providers regularly based on their symptoms, which may include:

- Audiology for hearing
- Endocrinology for diabetes
- Optometry and Ophthalmology for vision and eye concerns
- Neurology for muscles and the brain
- Nephrology for the kidneys
- Cardiology for the heart
- Gastroenterology for constipation or other GI concerns

If you or someone you care for has MIDD, ask a clinician if an emergency protocol letter would be helpful

in case of an emergency. These letters, which are written and signed by a doctor, share details about prescribed treatment during a crisis and in emergency room settings. Also discuss if your clinician is willing to write a "travel letter" to help you safely travel through TSA with your medical supplies and food for your diabetes.

Clinical Trials

For specific details on other clinical trials visit the MitoAction Clinical Trials page or www.clinicaltrials.gov.

Resources

- GARD Rare Disease Information — Maternally inherited diabetes and deafness
- MedlinePlus Genetics — Maternally inherited diabetes and deafness
- DiabetesGenes — Maternally Inherited Diabetes and Deafness (MIDD)

Connecting with others who are impacted by a rare disease allows for important information to be shared about day-to day life, prevents isolation, and gives hope. Please contact MitoAction for peer support opportunities at 888-MITO-411 or email mito411@mitoaction.org. Other resources we recommend are:

- New Patient Kit for Mitochondrial Conditions
- Planning and Preparation
- Monthly Expert Series
- Energy in Action Podcast

Sources

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- DiabetesGenes — Maternally Inherited Diabetes and Deafness (MIDD)
- Ali, Aleena Shujaat, Elif Ilhan Ekinci, and Felicity Pyrlis. "Maternally inherited diabetes and deafness (MIDD): an uncommon but important cause of diabetes." Endocrine and Metabolic Science 2 (2021): 100074. Maternally inherited diabetes and deafness (MIDD): An uncommon but important cause of diabetes - ScienceDirect

MitoAction does not provide medical advice, diagnosis, treatment, or legal advice. It is essential that all those living with or caring for someone with a Mitochondrial or FAOD disease have an emergency protocol letter. These letters, which are written and signed by a doctor, share details about prescribed treatment during crises and in emergency room settings. Always check with your doctor if you or your child has concerns as everyone may present with symptoms differently. Before beginning any treatment or therapy, please consult with your physician.