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ISSN: 2167-7662
Bioenergetics: Open Access

Commentary

Mitochondrial Myopathy and its Features in Primary Mitochondrial Myopathies

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DESCRIPTION

A mitochondrial disease known as a mitochondrial myopathy only affects the muscles and is distinguished from a mitochondrial encephalomyopathy, which affects both the muscles and the nervous system. The energy requirements of a typical human cell are satisfied by hundreds of mitochondria. Because a person can have a different combination of healthy and damaged mitochondria with a different distribution in the body, mitochondrial disease symptoms can differ. The majority of the time, mitochondrial disease is a multisystem condition that affects several cell, tissue, and organ types. Since muscle and nerve cells require a disproportionately high amount of energy, mitochondrial illness frequently varies as muscular and neurological issues. Impaired vision, cardiac arrhythmia (abnormal heartbeat), diabetes, and stunted growth are some more frequent consequences. A person with a mitochondrial disease typically has two or more of these diseases, some of which occur together frequently enough to be classified as syndromes.

Primary mitochondrial myopathies

The term Primary Mitochondrial Myopathies (PMM) refers to a collection of diseases that primarily affect the skeletal muscle and are linked to genetic abnormalities (such as depletions, deletions, or mutations) in the mitochondrial DNA (mtDNA) or in genes outside the mitochondria (nuclear DNA). Mitochondria, found by the hundreds within every cell of the body, regulate the production of cellular energy and carry the genetic blueprints for this process within their own unique DNA (mtDNA). The ability of the affected cells to digest food, absorb oxygen, and make energy is frequently hampered by these illnesses. Furthermore, person with signs of muscle illness but impacted organs (such as the brain, liver, kidney, etc.) are not regarded to have PMM and may fall under the umbrella of a more basic clinical diagnosis such as Kearns-Sayre syndrome, MELAS syndrome, etc. Other organs, including the brain, may also have issues as a result of some mitochondrial myopathies. Myopathies that also significantly affect the nervous system may be categorised as mitochondrial encephalomyopathies.

The start of the Mitochondrial Encephalopathy, Lactic Acidosis, and Stroke like Syndrome (MELAS) ranges from childhood to early adulthood. Lactic acidosis, encephalomyopathy with seizures and/or dementia, and recurrent stroke-like events are the distinguishing features of MELAS. These episodes are not conventional strokes, which are acute neurological symptoms brought on by disruptions in the blood flow to the brain. The myopathy variant of Mitochondrial DNA Depletion Syndromes (MDDS), which eventually affects the respiratory muscles, is characterised by weakness. Alpers syndrome, one type of MDDS, is characterised by increasing liver illness and abnormalities of the brain.

These groups could include mitochondrial DNA common mutation syndromes or mitochondrial DNA deletion syndromes, for example, and would include forms of mitochondrial myopathy that have been identified based on genetic origins. For instance, Kearns-Sayre disease is both a type of mitochondrial DNA deletion syndrome and a form of mitochondrial myopathy. Myopathy can result from mutations in either the nuclear or Mitochondrial DNA (mtDNA), which both code for the mitochondrial proteins. It's interesting to note that mtDNA alterations, most frequently mt-tRNA mutations, are linked to cardiomyopathies with worse results. Additionally Kearnes-Sayre syndrome, which is brought on by a significant deletion in the mtDNA genome.

CONCLUSION

Numerous well-known conditions linked to mitochondrial diseases feature pronounced muscle symptoms. Early onset of symptoms is common, with exertion causing a heavy sensation in the limbs and aching muscles. Patients may feel exhausted, nauseous, and out of breath after exercising. Over time, symptoms could worsen to the point that patients can only perform a certain amount of exercise. Some treatments under investigation aim to fix or avoid the damaged mitochondria rather than concentrating on particular difficulties of mitochondrial illness. These medications are dietary supplements based on three organic compounds that are important in our cells' generation of ATP.

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Received: 19-Aug-2022, Manuscript No. BEG-22-18232; **Editor assigned:** 23-Aug-2022, PreQC No. BEG-22-18232 (PQ); **Reviewed:** 12-Sep-2022, QC No BEG-22-18232; **Revised:** 20-Sep-2022, Manuscript No. BEG-22-18232 (R); **Published:** 28-Sep-2022, DOI: 10.35841/2167-7662.22.10.181

Citation: Methner S (2022) Mitochondrial Myopathy and its Features in Primary Mitochondrial Myopathies. J Bio Energetics. 10:181.