

# Bio-chemistry aspects of myopathies



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paper: Bio-Chemistry Aspects of Myopathies paper? Myopathies or muscular dystrophies in children are mainly caused due to chromosomal gene mutation unlike other Myopathies occurring in adults it doesn't affect the central or peripheral nervous system. Although majority of the dystrophies looks same but, the pattern of inheritance and clinical features of the affected child are the main parameters in the diagnosis or differentiating of Myopathies. In 1868 a French neurologist named Duchene first described a myopathy called Duchene muscular dystrophy (DMD) transmitted via x-linked recessive gene affecting mostly the male infants or adolescents.

Presenting with the clinical features of progressive symmetrical weakness, hypotonia and muscle wasting or pseudo hypertrophy of extensor group of muscles in the both upper and lower extremities and most commonly evident in the 4th year of life with the history of frequent falls, difficulty in walking, running, climbing stairs, difficulty in maintaining the balance and demonstrating a typical standing pattern while assuming upright position from the floor known as Gower's sign. The exact pathology of DMD is not fully understood but recent studies have shown that an enzyme called dystrophin has been found deficient in the striated muscles of the affected children. Bio-chemical analysis will show an increase in the levels of serum CPK (creatine phosphokinase-an enzyme found in muscle), LDH, Creatinine, AST (the normal range is 10 to 34 IU/L. An increase has many indications, one of them being progressive MD), Aldolase (this test is indicator of muscle damage). As the muscle weakness and wasting progresses the child may have difficulty in maintaining the normal respiratory function due to weakness of the respiratory muscles which may become fatal in the second

or third decade of life. Symptomatic treatment and physiotherapy plays an important role in minimizing the secondary complications of DMD.

Conclusion: DMD is a progressive muscular dystrophy of the striated muscles occurring commonly in the male infants and adolescents. Bio-chemistry analysis is of great importance in understanding the progress of muscle damage caused due to muscle dystrophies. In this modern Era of science and technology where clinical trials and research has answered many unresolved questions: but the root treatment of the muscular dystrophies still remained unclear. World wide charitable societies of Muscular Dystrophies are playing a vital role in providing all forms of assistance to the patients and parents of Muscular Dystrophies but the fatality rate of patients with DMD has been of great concern. Maintaining good functional independence and well programmed psychological and physical rehabilitation can lengthen the time to develop secondary complications of DMD which are the primary cause of morbidity and mortality. (paper) ;,? (paper) paper paper (paper) -X