USANA Technical Bulletin

Muscular Dystrophy

Description

- Muscular dystrophies are rare, inherited disorders of the muscles associated with progressive muscle weakness, loss of coordination, difficulty in walking, destruction and degeneration of the muscle fibers, and eventual replacement of the muscle fibers by fibrous and fatty connective tissue.[1]

Causes

- Muscular dystrophies are genetic disorders.

Types

- There are many forms of muscular dystrophy, the most common of which are Duchenne and Becker dystrophies.
- Duchenne dystrophy produces symptoms during early childhood, and leads to the inability to walk near the end of the first 10-12 years of life. Flexion contractures and scoliosis ultimately occur, and most patients die by 20 years of age.
- Early symptoms of Duchenne dystrophy include developmental delays, difficulty in running or climbing stairs, frequent falls, and enlargement of the calves. The muscles of respiration and the heart are commonly affected. Brain involvement is indicated by a lower than normal intelligence, and mild atrophy of the brain.
- Becker dystrophy is less common, and is a milder syndrome than Duchenne dystrophy. The symptoms of this disorder begin later and progress more slowly than those of Duchenne dystrophy. The average age of onset of symptoms is 12 years, patients become chair-bound at around 30 years and die at approximately 42 years.[1,2]

At Risk

- Those with the genetic predisposition to muscular dystrophies are the only ones at risk. Duchenne and other x-linked muscular dystrophies occur almost exclusively in males, with rare exceptions.

Prevention and Management

- These diseases cannot be prevented. Prenatal diagnosis in families with known histories of muscular dystrophy, carrier detection , and genetic counseling are the only controlling measures. There is no specific treatment of any of the muscular

dystrophies. Physical therapy, and corrective orthopedic surgery can be utilized at various stages of these disorders in order to improve quality of life. Recent advances in molecular genetics have raised the possibility of gene therapy of Duchenne dystrophy, but this approach has not yet been proven.[1]

- There is no particular diet or nutritional supplement that has been shown to be helpful in treatment of muscular dystrophy. Caloric requirements are, in most cases, less than normal. In any case, these patients should be encouraged to eat wisely in order to avoid becoming overweight, as obesity creates further burdens on the patient's quality of life.[2]

Sources of Additional Information

- http://www.upmc.edu/news/mdresbg.htm
- http://www.mgen.pitt.edu/res2res/res2resf.htm
- http://www.mdausa.org/index.html
- http://www.mda.org.au/

References

[1] Cecil Textbook of Medicine, 20th Ed. Philadelphia:W.B. Saunders Company, 1996; p 2161.
[2] The Merck Manual of Diagnosis and Therapy, 16th Ed. Rahway, (NJ):Merck Research Laboratories, 1992; pp 1526-1527.