

MUSCULAR DYSTROPHY: AN INTRODUCTION

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Summary

Muscular dystrophy is a rigorous disease recognized by the weakness of muscles of skeleton. About four types of this disease are known which are described on the basis of the pattern of inheritance. They are Dechunne, Becker, Facio-scapula-humeral, limb girdle and myotonic muscular dystrophies. Abnormalities of skeletal muscles on the microscopic value are found in almost each type. Dechunne is recessive abnormality which is X linked. Mostly boys are the patients of Dechunne. Becker is the mild type of Dechunne type of muscular dystrophy. The Facio-scapula type affects both the sex, also the limb girdle form. Disease shows its affects sooner and in upcoming generations of the diseased family. Dystrophy has its origin in genetic mutation. As specified cure is not found, general measures like physically and professionally treatment is in use.

Key words: Muscular dystrophy, Loss muscular function, Dystrophin

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Introduction

Muscular dystrophy is a rigorous disease ⁽¹⁾ recognized by the weakness of muscles of skeleton. With the passage of time, disease progress. Its affects are extreme weakning and lowering of function and mass of muscles ⁽²⁾.

With passage of time a person with muscular dystrophy needs some support to move around. More than four clinical types of the disease are known ⁽³⁾. These types were established on pattern of inheritance, the age when symptoms are recognized first time and on the pattern of muscles` distribution. Abnormalities of skeletal muscles on microscopic value are available in each form of this disease ⁽⁴⁾.

ETIOLOGY

All forms of muscular dystrophy are inherited. The loss of gene which cause Duchenne and Becker disease produce a protein named dystrophin. Dystrophin helps the muscle to keep its shape and its function normal.

TYPES AND SYMPTOMS

Following are the different forms and their respective symptoms:

1- Duchenne Form of Muscular Dystrophy:

In the Duchenne form of illness, symptoms are commonly noticed in the age before 5⁽⁵⁾. Muscles which affect firstly are of pelvic and the trunk region. Which results in the deshape of spinal cord and also disturb the gait of the person. Later in the teen age weakning of all muscle groups can also be increased. Also death can occur by respiratory weakness or may be by the involvement of heart muscles. Inheritance of this type is through the mechanism of X linked recessive trait. Mostly all the patients if this disease are boys.

2- Becker form of Muscular Dystrophy:

Becker muscular dystrophy is a mild type of Duchenne form of muscular dystrophy.

3-Facio-Scapulo-Humeral Form of Muscular Dystrophy:

Facio-scapula-humeral type of sickness affects both sexes in equal proportions which results in fragility of girdle of shoulder and upper arms. This is mostly recognized on the start of puberty. The specific weakness of muscles of face may some times be noticed during the early life. Many patients of this type of disease can walk in the early stage till the advanced stage start.

4-Limb-Girdle Form of Muscular Dystrophy:

Limb-girdle type of disease affects both sexes. Skeletal muscles which are involved may be of the shoulder or of hip muscles, or can be of both. The ailment can start in early or late life and progress slowly. In late stage most of the body muscles can be affected⁽⁶⁾.

5- Myotonic Form of Muscular Dystrophy:

In myotonic muscular dystrophy, along with the weakness and wasting also there is the delayed relaxation of muscles⁽⁷⁾. Flush of eyes by both eyes can happen and reproduction can also be disturbed Age at which disease start and complexity of symptoms are different in this type. The sickness try to occur sooner and in upcoming offspring of the diseased family, it may be more severe. It also affects both the sexes. But offspring of sick mothers are most probably inherited the severe type of illness than the offspring of diseased fathers.

The other types include Mitochondrial, Distal, Oropharyngeal, Emery driefuss muscular dystrophies.

DIAGNOSIS

Muscular dystrophy is originated from mutations in genes ⁽⁸⁾. Biochemical procedure through which defect in the genetic material is removed by the degenerative process in the muscle is not surely in knowledge to us.

Tests for the genetic material to find mutations in the different genes; which are responsible to cause muscular dystrophy; give fast and up to the mark diagnosis of disease ⁽⁹⁾. When becker and dechunne muscular dystrophies are under the study in the genetic tests, detection of female carriers is acceptable ⁽¹⁰⁾.

MANAGEMENT

As specified treatment is not found so commonly physically and professionally pathways are in use ⁽¹¹⁾.

Some times physicians perscribe the drugs like corticosteroids which slow down the progress procedure Dechunne muscular dystrophy. The action and accuracy of action of this medicine in the case of Becker type of disease is not in known ⁽¹²⁾.

Different ranges of exercises on movement applied on the patients may help to keep the position normal and their bodies flexible and able to move ⁽¹³⁻¹⁵⁾.

Park and Oh (2006) have tried to treat the disease by stem cells therapy. When they treated the dogs, who had a muscular disease which was some how similar to the Dechunne type of muscular dystrophy, by stem cells therapy, they found improvement in the symptoms ⁽¹⁵⁾.

References

- 1- Kianzowa M, Saraceni O, Wilhelm JM, Kieffer P. Lyme disease disclosed by disabling amyotrophy. *French. Rev Med Interne*. 1993; 14(7):723-6.
- 2- You P, Ma Q, Tao T. Gene diagnosis of oculopharyngeal muscular dystrophy in a Chinese family by a GeneScan method. *J Clin Lab Anal*. 2010; 24(6.): 422-5.
- 3- Lusakowska A, Sulek-Piatkowska A. Myotonic dystrophy - a new insight into a well-known disease. *Neurol Neurochir Pol*. 2010; 44(3):264-76.
- 4- Mochida K, Ito K, Harino H, Onduka T, Kakuno A, Fujii K. Muscular Dystrophy. *Environ Toxicol Chem*. 2008; 27(2):367-74.
- 5- Nilsson J, Nilsson J, Larson G, Grahn A. Characterization of site-specific O-glycan structures within the mucin-like domain of alpha-dystroglycan from human skeletal muscle. *Glycobiology*. 2010; 20(9):1160-9.
- 6- Pathak P, Sharma MC, Sarkar C, Jha P, Suri V, Mohd H, Singh S, Bhatia R, Gulati S. Limb girdle muscular dystrophy type 2A in India. *Neurol India*. 2010; 58(4):549-54
- 7- Berlit P, Stegaru-Hellring B. The heart in muscular dystrophy: an electrocardiographic and ultrasound study of 20 patients. *Eur Arch Psychiatry Clin Neurosci*. 1991; 241(3):177-80.

- 8- Awano H, Malueka RG, Yagi M, Okizuka Y, Takeshima Y, Matsuo M. Contemporary retrotransposition of a novel non-coding gene induces exon-skipping in dystrophin mRNA. *J Hum Genet.* 2010. [ahead of print]
- 9- Tarnopolsky MA, Pearce E, Matteliano A, James C, Armstrong D. Muscular dystrophy. *Muscle Nerve.* 2010; 42(6):853-5.
- 10- Nasiłowski J, Szkulmowski Z, Migdał M, Andrzejewski W, Drozd W, Czajkowska Malinowska M, Opuchlik A, Chazan R. Prevalence of home mechanical ventilation in Poland. *Pneumonol Alergol Pol.* 2010; 78(6):392-8.
- 11- Call JA, McKeehen JN, Novotny SA, Lowe DA. Progressive resistance voluntary wheel running in the mdx mouse. *Muscle Nerve.* 2010; 42(6):871-80.
- 12- Xiong F, Xu Y, Zheng H, Lu X, Feng S, Shang Y, Li Y, Zhang Y, Jin S, Zhang C. Microdystrophin delivery in dystrophin-deficient (mdx) mice by genetically-corrected syngeneic MSCs transplantation. *Transplant Proc.* 2010;42(7):2731-9
- 13- Yin H, Moulton H, Betts C, Wood M. CPP-directed oligonucleotide exon skipping in animal models of Duchenne muscular dystrophy. *Methods Mol Biol.* 2011; 683:321-38.
- 14- Rousseau J, Dumont N, Lebel C, Quenneville SP, Côté CH, Frenette J, Tremblay JP. muscular dystrophy. *Cell Transplant.* 2010;19(5):589-96.
- 15- Park KS, Oh D. Gene therapy for muscular dystrophies: progress and challenges. *J Clin Neurol.* 2010;6(3):111-6.