

Recent advancement in treatment of Duchenne muscular dystrophy (DMD) in Ayurveda.

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ABSTRACT

Duchenne Muscular Dystrophy (DMD) defines the most common childhood muscular dystrophies; it is an inherited disorder (X-linked recessive) with progressive degeneration of muscle. The incidence is 1:3500 live male births. Duchenne muscular dystrophy is present at birth, but the disorder usually becomes apparent between ages 3 to 6 years. The child fall frequently and have difficulty keeping up with their friends when playing, running, jumping and on getting up from the floor, the child uses his hands to lift up himself (Gower's maneuver). By the age 16 to 18, patients are predisposed to serious, and sometimes fatal, pulmonary infections. In Ayurveda it has been classified under Adibala Parvritta Vyadhi. Here pathogenesis occurs due to the Bheejabagahaavyava Dusti which lead to Medomamsa dusti further vitiates the Vata. DMD can be compared to Paurasadini jataharini (in which delivered child dies before 16 years of age) one of the AsadhyaJataharini mentioned in Kashyapsamhita. Muscular dystrophy is diagnose by blood CPK level, Muscle biopsy, DNA testing, EMG and NCV. There are no 100% specific treatments in any system of medicine that can reverse the progression of deterioration. So it is more important to consider complementary and approaches of alternative treatment. Main aim of treatment is to prevent complications such as contracture, weakness, reduced mobility and slow down muscle atrophy. By the use of Ayurvedic Rasayana, Herbomineral drugs, Snehana, Swedana, Yoga and Pranayam (breathing exercises) have shown the influence of protection and longer survival in the muscular dystrophy. This article focuses on recent advancement in treatment of Duchenne muscular dystrophy (DMD) in Ayurveda.

Keywords: Duchenne muscular dystrophy, Snehana, Swedana, Rasayana.

Introduction:-

The term Dystrophy literally means 'faulty nourishment' (dys = faulty/abnormal, trope = nourishment). Muscular dystrophy (MD) is a group of genetic diseases involving progressive weakness and degeneration of the skeletal or voluntary muscles which control movement. In some form of muscular dystrophy, the heart muscles and other involuntary muscles as well as other organs are affected. There are 9 distinct types of muscular dystrophy. Myotonic muscular dystrophy is most common among adult while Duchenne Muscular Dystrophy is most common among children. The disease was first described by Neapolitan physician Giovanni Semmola in 1834 and Gaetano Conte in 1836. However DMD was named by French neurologist Guillaume-Benjamin-Amend Duchene in 1806-1875¹. Duchenne muscular dystrophy (DMD) is an X-linked disease that affects 1 in 3000-3500 live male births^{2,3,4}. It is commonly seen in males and in this disease females are the carriers⁵. Affected individuals can have mildly delayed motor milestones and most are unable to run and jump properly due to proximal muscle weakness, which also results in the use of the classic Gowers' sign when arising from the floor. Most patients are diagnosed at approximately 5 years of age⁶. Untreated, muscle strength deteriorates, and boys require the use of a wheelchair without intervention, the mean age at death is around 19 years. Non-progressive cognitive dysfunction might also be present⁷. Respiratory failure is the most common cause of premature death in patients with DMD^{8,9}. Muscular dystrophy is diagnose by blood CPK level, Muscle biopsy, DNA testing, EMG and NCV. In Ayurveda the references of this disease is not available, but as for clinical features it can be compared with *Mamsagatavata* and *Mamsakshaya*. DMD can be compared to *PaurasadiniJataharini* (in which delivered child dies before 16 years of age) one of the *AsadhyaJataharini* mentioned in *Kashyapsamhita*. Muscular dystrophy is an incurable and fatal disease. No treatment is present in any system of medicine which has any definite influences upon muscular dystrophy. Therapeutic approach of muscular dystrophy in modern medicine is represents on corticosteroids, physical therapy, respiration assistance and gene therapy. In Ayurveda treatment of muscular dystrophy is aimed to treating the basic pathology of diseases, preventing disability and improves quality of life.

Pathogenesis of DMD:-

Dystrophy is a genetic defect and main cause of Duchenne and Becker type of muscular dystrophy is lack of a single muscle protein Dystrophin. Dystrophin protein is found in muscle fiber membrane. As a result of dystrophin deficiency, patients with DMD (including those who receive optimal treatment) experience a progressive loss of muscle fiber that eventually affects the respiratory muscles¹⁰.

In Ayurveda, it is described that *Dhatvagnis* are responsible for the process of metabolism. According to *Chraka Mamsa-kshaya* may be present when there is prolonged *Majjakupit Vata*. This is always followed by depletion of *Vata*.

Classification of muscle dystrophy:-

The classification of muscular dystrophy based upon the inheritances is as follows: i. X-1 inked recessive muscular dystrophies. It is mainly two types-

1. Duchenne muscular atrophy (DMD)
2. Becker muscular dystrophy (BMD)

Duchenne Muscular Dystrophy:-

It is also known as Pseudo-hypertrophic muscular dystrophy which occurs in male baby. It is the most common one and characterized by rapid progression of muscle degeneration. Duchenne dystrophy is caused by a mutation of the gene responsible for producing Dystrophin. Duchenne dystrophy is present at birth, but the disorder usually becomes apparent in less than 6 years age. In younger children, the calf muscles are usually enlarged called Pseudo-hypertrophy and muscle is replaced by fat, there will be inability to walk after the age of 12 years. Contractures become fixed, and a progressive scoliosis often develops which may be associated with pain. By the age 16 to 18, patients are predisposed to serious, and sometimes fatal, pulmonary infections. Other causes of death include aspiration of food and acute gastric dilation.

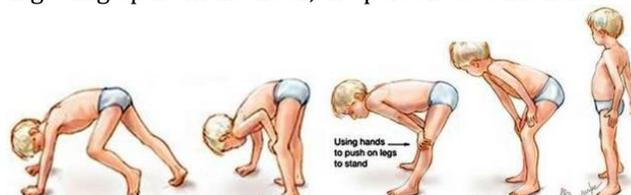
Becker muscular dystrophy: - This is another kind of muscle disorder, which also affects boys. In this case, the symptoms are less severe. People with this disease have breathing, bone, muscle, joints and heart problems. However, in this case the person will not be confined to wheelchair immediately.

Differences from Duchenne muscular atrophy and Becker muscular Dystrophy ¹¹:-

Features	Duchenne Dystrophy	Becker Dystrophy
Onset of weakness	Earlier	Later
Age till ambulatory	10-12 years	Late adolescence or early adult
Death	Commonly <18 years	25-40 years

Sign and Symptoms:-

1. Waddling gait, Gower sign, and calf muscle pseudo hypertrophy are classical findings at this stage.
2. Pseudo hypertrophy of calf muscles and tongue. The enlarged muscle tissue is eventually replaced by fat and connective tissue hence uses the term pseudo hypertrophy.
3. Awkward manner of walking, stepping or running. (Patients can walk on their forefeet)
4. Frequent fall and fatigue.
5. Eventual loss of ability to walk (usually by the age of 12 years)
6. Increased lumbar lordosis leading to shorting of the hip-flexor muscle.
7. Muscle contractures of Achillestendon and hamstrings impair functionally.
8. Weakness of intercostals and diaphragmatic muscles with spinal deformity affects the respiratory function.
9. Dropping of vital capacity <20% of normal leads to nocturnal hypoventilation.
10. Cardiomyopathy and arrhythmias are the major cardiac manifestations in Duchenne muscular dystrophy. The cause of death in Duchenne muscular dystrophy patients is usually a combination of respiratory insufficiency and cardiomyopathy.
11. **Gower's sign:** - On getting up from the floor, the person uses his hands to lift up himself.

**(Figure -Gower's maneuver)**

Diagnosis of DMD: - Five tests are helpful for the diagnosis of muscular dystrophy-**1. Blood test:-**

- Creatinine kinase (CK) or Creatinephospho kinase (CPK) levels.
- Damaged muscles release enzymes such as Creatine Kinase (CK) into the blood. High blood levels of CK suggest a muscle dystrophy.
- Normal value: 10-135 IU/L in female; 10-170 IU/L in males. It is invariably elevated to between 20 and 100 times than normal.

2. Electromyography:

- It is very useful when CPK levels are not elevated to the degree expected in Dystrophin deficiency.
- EMG demonstrates features typical of myopathy.

3. Muscle biopsy:-

- It shows muscle fibers of varying size, as well as small groups of necrotic and regenerating fibers, connective tissue and fat replace lost muscle fibers.

4. Nerve conduction velocity: - It is decreases in DMD.**5. Other tests:-**

- Immunohistochemistry for Dystrophin I, II, III reveals absent Dystrophin in DMD and reduced patchy Dystrophin staining in BMD.
- The new test called Single Condition Amplification Primer Sequencing (SCAID) allows clinicians and Geneticists to sequence the entire dystrophin gene to find mutations that confirm DMD.

Treatment of DMD: - There is currently no cure for any form of muscular dystrophy in any system of medicine. Treatment is only helps in prevention or reduction in deformities and improves quality of life.

Treatment in Modern medical science:-

- Management of a child with Duchenne muscular dystrophy requires a multidisciplinary team.
- **Steroids Therapy:-** Steroids such as prednisone can slow the rate of muscle deterioration in DMD and help children retain strength and prolong independent walking by as much as several years. However, these medicines have side effects such as weight gain and bone fragility that can be especially troubling in children^{12,13}.
- **Stem cell-based therapy:-** The muscle cells of dystrophy patients lack a critical protein, such as dystrophin in DMD so possibility that the missing protein can be replaced by introducing muscle stem cells that restore muscle function in affected persons.
- **Gene replacement therapy:** Cure for DMD might be obtained if the defective dystrophin gene could be replaced by a functional gene.

Treatment in Ayurveda:-

Treatment is aimed at keeping the patient independent for as long as possible and preventing complications that result from weakness, reduced mobility, and cardiac and respiratory difficulties. In Ayurveda the concept of *Dosha-dhatu-Mala* theory is unique in the treatment of various diseases. In this disease the main vitiated *dosha* is *Vata*, due to improper *dhatvagni* and which may lead to *Mamsadhatukshaya*. Hence in this disease the main treatment principle is correction of the *Dhatvagni* and pacifying the *Vata*. Treatment may involve a combination of approaches, including drug therapy, *Purva karma* and *Panch karma* therapy, *Pranayam* and *Yoga* therapy.

1. Drug therapy:-

- In this therapy mainly *Rasayana* and herbomineral drugs are uses. Administration of *Rasayana* is useful in the long term management of muscular dystrophy which helps in reversing the progressive muscular degeneration.
- **Rasayana Drugs:** - *Nagabala rasayana* and *Mandookaparni rasayana* etc.
- **Herbomineral drugs:** - *Swarnabhasma*, *Vatagajankusha rasa*, *Arogyavardhini vati*, *Pravalapishiti rasa* etc.

2. Purvakarma and Panchkarma: -*Purvakarma* procedures are very much essential for DMD.

- ❖ It is very well understood in the treatment principle of *Vataroga* by *Charaka* that *upakarmas* like *Snehana* and *Svedana* are having prime treatment modalities¹⁴.
- ❖ *Abhyanga* is a variety of *Bhayaasnehana* with oil like *Mahanaryana Taila* and *Mahamamsaditaila* helps in subside the *VataDosha*, improves the tonicity of the muscle and compactness the body.
- ❖ Whereas *Swedana* like *Shastikashaali pindaswedana* also improves the tone of the body. *Swedanakarma* increases the metabolic activity which in turn increases the oxygen demand and blood flow.

- ❖ Rise in temperature by *Swedana* induces muscle relaxation and increases the efficacy of muscle action as the increased blood supply ensures the optimum condition for the muscle contraction.
 - ❖ *BrihmanaBasti* is useful in Duchenne Muscular Dystrophy (DMD). Mainly *Mamsa rasa Basti* and *YapanaBasti* are beneficial.
3. **Yoga therapy:** -Practicing yoga has also been used with good results in muscular dystrophy¹⁵.Yoga however doesn't cost energy of patients, but provides with a fresh flow of energy. Yoga increased flexibility, increased muscle strength and tone Improved respiration, energy and vitality etc.
 4. **Pranayama therapy:** - In DMD respiratory capacity decreases by approximately 5% per year¹⁵.Pranayama strengthens the heart and lungs, improves the digestion and calms the mind.This technical definition refers to a particular system of breath control with three processes as *Puraka* (to take the breath inside), *Kumbhak* (to retain it), and *Rechak* (to discharge it)¹⁶.

Discussion & Conclusion:-

Muscular dystrophy is genetic disorder with no satisfactory treatment in any system of medicine.DMD is a relatively rare and progressive disease with an expected survival of 16-19 years of age. In Ayurveda main line of treatment of this disease is correction of the *dhatvagni* and pacifying the *Vata*.The administration of *Rasayana*and Herbomineral drugs, are useful in the long term management of muscular dystrophy. *Snehana* improves the tonicity of the muscle and compactness the body.*Swedana karma* increases the metabolic activity which in turn increases the oxygen demand and blood flow.*Brihmana basti* is especially useful for *Vata shaman*.*Yoga* increased flexibility, increased muscle strength, muscle tone Improved respiratory muscle capacity.*Pranayama* strengthens the heart and lungs, improves the digestion and calms the mind.

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