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Review Article

AN AYURVEDIC PERSPECTIVE ABOUT DUCHENNE MUSCULAR DYSTROPHY

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ABSTRACT

Duchenne muscular dystrophy is a neuromuscular disease. This is an X-linked recessive disease caused by mutations in the dystrophin protein gene located on the short arm of the X chromosome, in the Xp21 region. It causes progressive muscle weakness due to wasting of muscle fibers. It causes loss of ability to stand, walk, and move before age 10; most patients die in their 20s. Since there is no specific treatment in any medical system and the prognosis of the disease is uncertain, if we start treatment at an early stage, children with DMD can walk, it can slow or stop progressive muscle degeneration. DMD cannot be directly related to a single disease in Ayurveda. Most major neuromuscular disorders are determined by *Vata dosha*. In Ayurveda, this pathogenesis can be clearly understood by the concept of *Adibala Pravrutta Vyadhi* (genetic disease). Here the pathogenesis is due to *Beeja Bhagavayava Dusti* (partial chromosomal defect) leading to *Mamsa Vata Dushti*.

INTRODUCTION

The term dystrophy, which means "abnormal growth" or "faulty nourishment" or "poor nourishment," was popularized towards the end of the 19th century. It was derived from the Greek word "trophe," meaning nourishment.[1] The disease was initially described by Neapolitan physician Giovanni Semmola in 1834 and Gaetano Conte in 1836. However, DMD, as we know it today, was named by the French neurologist Guillaume-Benjamin-Amend Duchene between 1806 and 1875.[2]

Among the myriad of more than 20 different genetic muscular disorders, the most common and ultimately fatal is DMD. It occurs all around the world, causing significant hardships for affected individuals and their families. Dystrophinopathies, a group of disorders resulting from mutations in the dystrophin gene, are found on the short arm of the X chromosome in the Xp21 region. Duchenne muscular dystrophy is known as an x-linked recessive disorder.

The incidence is 1:3600 live born infant boys [3]. In affected families, the mother is known to be a carrier of the gene, 50% of boys will be affected, and 50% of the girls will be carriers. Despite the X-linked recessive inheritance in DMD (Duchenne Muscular Dystrophy), about 30% of cases are new mutations and the mother is not a carrier.

The new mutation rate in the dystrophin gene is high. Most cases are born now in families with no prior history of the disease. Dystrophin is cytoskeletal protein localized in the inner surface of the muscle membrane. It forms dystroglycal-glycoprotein complex; this complex helps to maintain the integrity of muscle cells [4].

So absence of dystrophin results in the destabilization of the entire dystroglycan-glycoprotein complex. So, muscle mass is not growing well and cause to weakness of muscles. This condition is most apparent or symptomatic in skeletal muscle only. Heart and diaphragm muscle often involved too.

Most patients die because of heart failure or respiratory problems. Diagnosis of this disorder is based on clinical presentation, genetic testing, muscle biopsy, and muscle imaging.

No any treatment is available at present in DMD definitely in any medical field. Therapeutic approach of muscular dystrophy represents on corticosteroids. Physical therapy, respiration assistance, and gene therapy are the best ways to manage this condition.

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Pathogenesis

Dystrophy is a genetic defect caused by a deficiency in a single muscle protein, dystrophin. DMD and BMD are due to different changes in the dystrophin gene, which contains protein information important for the proper functioning of muscle cells.

This gene is located on chromosome X Dystrophin is localized to the sarcolemma of normal skeletal muscle but is completely absent in the muscle of DMD patients. A hypothesis states a defect in the sarcolemma membrane that allows an unknown substance, possibly calcium, to freely penetrate into the muscle fibres, where it activates neutral proteases, thus leading to excessive muscle catabolism and muscle necrosis.[5-8]

Clinical Features

Children with Duchenne muscular dystrophy often have symptoms before age 5 and may even have a history of delayed walking. Gait disorder usually becomes apparent when a child is 3 or 4 years old. Waddling gait, Gower sign, and pseudohypertrophy of the calf muscles are classical signs at this stage.[9] Weakness of the neck flexors was observed early. The progression of muscle weakness may reach a plateau between the ages of 3 and 6 years.

Subsequently, there is increasing difficulty in walking, development of spasticity, and lumbar lordosis. Studies showed that the age of loss of independent ambulation in cases of untreated Duchenne muscular dystrophy ranged from 8.8 to 10.5 years [10]. After losing the ability to walk, kyphosis and scoliosis become more severe, upper limb weakness increases, and bulbar dysfunction occurs.

Weakness of the intercostal muscles and diaphragm accompanied by spinal deformity affecting respiratory function and thereby reduces vital capacity

Examination of the Child

Evaluate following test results –

Blood Tests - CK Levels - Weak or damaged muscles release an enzyme called CK (creatinine phosphokinase) into the blood. High CK levels in children may be a sign of DMD or another muscle disease.

Genetic testing- Tests a blood sample for abnormal genes, for DMD

Muscle biopsy- A biopsy is only needed if first genetic test is negative.

This happens in about one-third (30%) of cases.

Modern DMD Treatment

The Neuromuscular and Neurometabolic Canter's care team will recommend a treatment plan to meet your child's needs. The goal of treatment is to control symptoms, move the muscles for as long as possible, and give your child the best possible quality of life. There is no cure for DMD.

- **Physical therapy:** It is important for your child to stay active. Using their muscles helps them stay as healthy as possible. Inactivity, such as bed rest, can cause muscle weakness more quickly
- **Corticosteroids:** Steroid medications can slow the progression of the disease.

Pathogenesis of DMD according to Ayurveda

The direct correlation of DMD with any other disease in Ayurveda is not possible. But considering the symptoms in the children with DMD, it may be due to *Beejabhagavayava Dushti* (defect in fraction of part of chromosome) due to *Adibala Pravrutta* (Genetic disease) cause[11]. This *Beejabhagavayava Dushti* further leads to *Tridoshdusti* (vitiation of *Vata, Pitta, Kapha*). Among the *Tridoshas Vata* is affected more. This *Tridosha dusti* further leads to *Dhatu Vaishamya* (imbalance of the tissue component of the body) mainly *Mansa Dhatu* (muscle tissue).

So this *Mansagata Vata Kshaya* (decrease of *Vata* in muscle tissue) due to *Beejabhagavayava Dushti* (defect in fraction of part of chromosome) causes *Mansadhatwagni Mandya* (decrease in the fire of muscle tissue) [12], which leads to improper formation of *Mansadhatu* (muscle tissue) and *Mansadhatu Kshaya* (decrease in muscle tissue) which can be correlated with muscle atrophy. As per Ayurveda *Siddhant* (principles of Ayurveda), each *Dhatu* (tissue) has its self *Dhatwagni* (the fraction of *Agni* that functions at various body tissues for nourishment and metabolism) and according to *Uttarottar Dhatu Poshan Nyaya* (each tissue possesses nutrient portion of consecutive tissue) formation of *Dhatu* takes place serially. The *Sukshmansha* (small portion) of each *Dhatu* is nutrient portion of next *Dhatu* and this *Sukshmansha* gets transformed into Next *Dhatu* when it goes to specific *Srotas* (micro channels of circulation in the body) using *Dhatwagni* of that specific *Dhatu*.

So due to vitiation of *Mansadhatu* there is improper formation of further *Dhatus* like *Meda* (fat), *Asthi* (bone), *Majja* (bone marrow), which causes more vitiation of *Vata*. As per *Acharya Charak*- the symptoms of *Mansamedogata Vata* like *Guruanga Tudyate Atyartha Danda Mushtihatam Tatha*[13]. These symptoms can be correlated with DMD.

Similarly, according to *Acharya Charaka* the symptoms of *Asthimajjagata Vata* like *Mansakshaya* (decrease in muscle tissue) and *Balakshaya* (loss of physical strength or weakness) and *Sandhishool* (joint pain)[14] can be correlated with symptoms of DMD.

Among the 5 types of *Vata,* the *Vyana Vayu* is mainly vitiated. As we know the *Karmas* (function) of *Vyana Vayu* are- *Gati* (movements), *Apakshepana* (downward movement of limbs), *Utkshepa* (elevation of limbs), *Nimesha* (closing of eyelids), *Unmesha* (opening of eyelids). So all the movements are controlled by *Vyana Vayu* and due to its vitiation these

movements are hampered [15]. The *Vyana vayu* acts on autonomic nervous system. The location of *Vyana Vayu* is whole body, so whole body movements including pumping of heart, movements of diaphragm, respiration are under control of *Vyana Vayu.* So its vitiation causes weakness in skeletal muscles, muscles of heart, diaphragm etc which further leads to respiratory failure and cardiac failure in the end stage of disease.

As per *Acharya Charaka* the location of *Prana Vayu is Murdha* (head)*, Ura* (chest)*, Kantha* (throat)*, Jivha* (tongue)*, Asya* (mouth)*, Nasika* (nose)[16]. As the *Prana Vayu* is located in *Murdha* (cranium) and do the function of respiration, deglutition and other body activities and spiritual understandings. Hence *Prana Vayu* is controlling each and every cell of the body. In DMD along with *Vyana Vayu, Prana Vayu* is also vitiated.

Ayurvedic management of DMD

Acharya Charaka has explained *Chikitsasutra* (treatment guidelines) of *Mansavritta Vata* in *Charak Chikitsasthan* [17]. As there is *Mansadhatwagni Mandya, Deepan* (metabolism boosting) and *Pachan* (detoxification) *Chikitsa* is started to improve *Agni* (digestive fire).

Snehana- It can be internal as well as external. External *Snehana* i.e., *Abhyanga* (oil massage/oleation) stimulates receptors of the skin. It also stimulates circulatory system. It causes vasodilatation and thereby enhances the blood flow. It provides more nourishment to the tissue resulting into strengthening of the muscle tissue. It also provides flexibility to the muscle cells by decreasing the fibrous adhesions in the muscles and increases muscle tone and power[18] e.g. *Mahamasha Taila, Kshirbala Taila.*

For internal *Snehana Tikta Rasa* (bitter) is used. According to *Acharya Charaka Tikta Rasa* is *Twak Mansa Sthirikaranam,* so *Tiktarasayukta Ghrita* is used for *Snehapana.* (E.g. *Panchtikta Ghrita)*

***Swedana* (body heating):** After *Snehana*, heat application decreases gamma activity, so that the stretch on muscle spindle is decreased. It further decreases afferent firing from spindle which causes decrease in alpha motor neuron firing. It decreases muscle spasm. *Swedana* decreases joint stiffness and increases tissue extensibility thus improves the range of movements[19]. E.g. *Shashtikshali pindasweda, Mashapindasweda.*

Basti (enema) is the main treatment for *Vatavyadhi* (disorders due to *Vata* component of body). *Basti* drugs get absorbed through colon to show their systemic actions. It treats *Gambhir Dhatugata Vata* and excretes the morbid substances responsible for disease progression [20]. E.g. *Mansarasa Basti*, as per *Samanya Vishesha Siddhanta* (similarity of all substances is always the cause of increase and dissimilarity is the cause of decrease) *Mansa* can be used to strengthen the muscles.

For *Anuvasan Basti* (enema of medicated oil) *Tikta Ghrita, Chhaglyadi Ghrita* and *Ashwagandha Ghrita* can be used [21,22]

Nasya (intranasal drug application) - *Nasa hi shirso dwaram* (The entry point of brain is nose) So drugs administered through nose acts on brain, nerves etc. So that the cognitive impairment associated with DMD can be managed[23] e.g. *Goghrita Nasya, Brahmighrita Nasya, Panchendriyavardhen Taila Nasya.*

Yoga improves strength of muscle and helps to maintain equilibrium between mind and body. It improves positivity and works as *Satvavajaya Chikitsa* (Ayurvedic psychotherapy).

In *Niramavastha* state (not associated with toxins), *Brihana* (building, nourishing or strengthening) *Yogas* are used. E.g. *Ashwagandha Churna, Shatawari Churna, Amalaki Churna,Balarishta, Ashwagandharishta, Trayodashanga Guggula*. Daily *Sevan* of *Mansarasa* (medicated flesh soup) is advised.

DISCUSSION

DMD is neuromuscular disorder. As we know modern medicine uses corticosteroids only for the treatment of DMD which has adverse effects on the body. There is no cure for this disease in other pathies except Ayurveda. All *Panchkarma* procedures with internal medications can be used without any side effects. It improves functional capacity of activity, also reduces rate of disability. It slows down disease progression and retains the ambulation for longer period of time, hence supports the activities of daily routine.

CONCLUSION

Using Ayurveda principles, we can't totally cure the disease but can improve the quality of life of the patient. There is vast scope of research in this area as other pathies has no satisfactory treatment for this disease, this article promotes the need of research in DMD.

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