

A REVIEW ARTICLE ON MUSCULAR DYSTROPHY AND ITS MANAGEMENT THROUGH AYURVEDA

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ABSTRACT

Duchenne muscular dystrophy (DMD) is the most common muscular dystrophy caused by the absence of dystrophin gene. It is an X-linked inherited disorder without involvement of nervous system, progressive degeneration of group of muscle characterized by mildly delayed motor milestones, slow and progressive muscle weakness start presenting soon after the age of 3 years. Severe weakness and wasting occurs in proximal muscles of lower limbs. The Incidence of Duchenne muscular dystrophy is 1 in 3500 live male. Most common cause of death in DMD is cardiomyopathy and respiratory

insufficiency. In modern medicine no successful therapy for treating DMD cases except corticosteroid's i.e. Deflazacort. Muscular dystrophy cannot be directly co-related with any single disease in *Ayurvedic* texts. It can be correlated to *Vata vyadhi in Ayurvedic text*. In *Ayurveda* muscular dystrophy may be clearly understood by the concept of *Adhi bala pravritta vyadhi* here the pathogenesis occurs due to the *Beeja bhagavayava dusti* which leads to *Mamsa vata dushti*. The present review article reveals highlights on various *Ayurvedic* medicines along with *Panchkarma* (*Snehan, SSPS, and Basti karma*) therapy for treatment and prevention of muscular dystrophy.

KEYWORDS: Muscular dystrophy, dystrophin, *Adhi bala pravritta vyadhi, Panchkarma*.

INTRODUCTION

The incidence of muscular dystrophy is commonly seen in male children's and females are the carriers. The term Dystrophy literally formed by (dys = faulty/abnormal, trophe = nourishment) means 'faulty nourishment' of the child and they are related with a group of hereditary progressive diseases and each type of muscular dystrophy has unique phenotypic and genetic presenting features. Muscular dystrophies is the result of mutation in gene i.e.

dystrophin. The dystrophin is a part of a large complex of sarcolemmal proteins and glycoproteins. It is one of the largest identified human genes, estimated at 2000 kb in size, localized to the short arm of the X chromosomes at Xp21.

MUSCULAR DYSTROPHY

The classification of muscular dystrophy based upon the inheritances is as follows:

I. X-linked recessive muscular dystrophies

- Duchenne muscular atrophy
- Becker muscular dystrophy

II. Autosomal recessive muscular dystrophies

- Limb – girdle dystrophy
- Congenital muscular dystrophy

III. Autosomal dominant muscular dystrophies

- Myotonic dystrophy
- Facioscapulohumeral muscular dystrophy
- Oculopharyngeal dystrophy
- Distal myopathies

DUCHENNE MUSCULAR DYSTROPHY

It is the commonest and severe,^[1] form hereditary muscular dystrophy in children, affecting all races and ethnic groups which is recognized by progressive weakness, intellectual impairment, hypertrophy of the calves and proliferation of connective tissue in the muscle.^[2]

It is the result of mutations (mainly deletions) in the dystrophin gene located on the locus Xp21.2 chromosomes and mutations lead to an absence of or defect in the protein, i.e. dystrophin, which results in progressive muscle degeneration leading to loss of independent ambulation by the age of 13 years.^[3]

Its prevalence estimated to affect 1 in 3,800-6,000 live male births.^[4] The DMD is diagnosed between 4 to 5 years of age group^[5], when children's start to show signs of physical disability including walking and climbing along with mild delayed developmental milestones of affected child, which also results in the use of the classic Gowers' manoeuvre when arising from the floor.^[6] It is suspected during infancy to early childhood on the basis of mildly

delayed developmental milestones and non-progressive cognitive dysfunction with increased serum creatine kinase activities.^[7]

The mean age of death is ~ 19 years in DMD due to cardiomyopathy, respiratory insufficiency and orthopaedic complications and patients not treated on appropriated time.

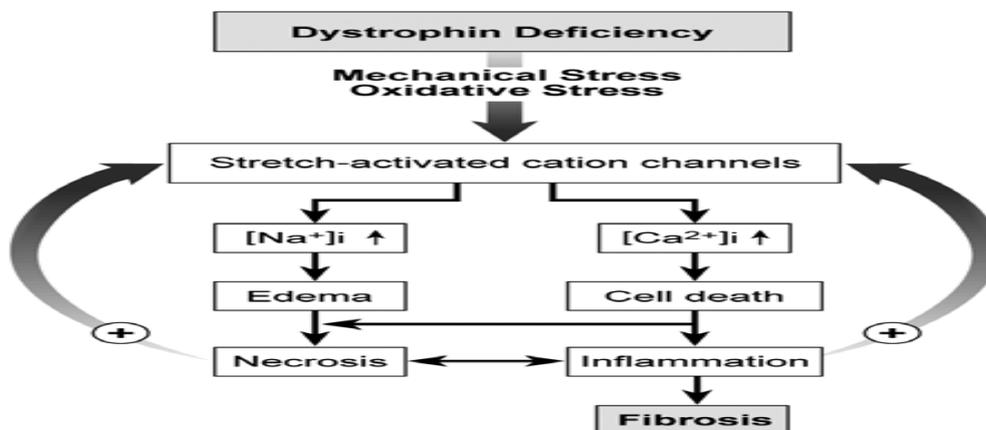


Fig 1: Flowchart: from dystrophin deficiency to fibrosis.

When to suspect DMD

Suspicion of the diagnosis of DMD should be considered irrespective of family history and is usually triggered in one of three ways:

- (1) Most commonly, the abnormal muscle function in a male child.
- (2) Increase serum creatine kinase tested for unrelated indications.
- (3) Increased transaminases (aspartate aminotransferase and alanine aminotransferase), which are produced by muscle as well as liver cells.

CONFIRM DIAGNOSIS

The blood sample is always necessary even if DMD is first confirmed by the absence of dystrophin protein expression on muscle biopsy.

The following investigation should be done for confirm diagnosis of DMD:

- i. The genetic tests commonly used to identify dystrophin mutations are multiplex PCR.^[8]
- ii. Multiplex ligation-dependent probe amplification.^[9]
- iii. Single-condition amplification/internal primer.^[10] Multiplex amplifiable probe hybridization.^[11]
- iv. The creatine kinase is a good screening test for muscle disease in clinical practice because levels rise in conditions with active muscle fiber necrosis and injury.^[12] Usually very elevated and are between 5000 and 150 000 IU/L (normal is less than 200 IU/L).

CLINICAL MANIFESTATION

The clinical features of muscular dystrophy are depends on the age group of child such as:

In infancy: In this age group boys are rarely symptomatic at birth or in early infancy, although some are mildly hypotonic. Early gross motor skills, such as rolling over, sitting, and standing, are usually achieved at the appropriate ages or may be mildly delayed except poor head control may be the first sign of weakness in the infancy.

In later childhood: a “transverse” or horizontal smile may be seen. Walking is often accomplished at the normal age of approximately 12 month, but hip girdle weakness may be seen in subtle form as early as the 2nd yr.

Toddlers: In this age group the affected child developed lordotic posture when standing to compensate for gluteal weakness and Gowers sign is often evident by age 3 year which is fully expressed by 5 or 6 years of age group as well as a Trendelenburg gait, or hip waddle, appears at this time.

The most common clinical presentations in toddlers are delayed walking, frequent falling, toe walking and trouble running or walking upstairs, developmental delay and less often, malignant hyperthermia after anesthesia.^[13] The classic features of DMD are enlargement of the calves (pseudohypertrophy) and wasting of thigh muscles.

Contractures: The most common site of contractures in DMD is often involve the ankles, knees, hips and elbows joints and Scoliosis is common defects. Unless ankle contractures are severe, the ankle deep tendon reflexes remain well preserved until terminal stages while the knee deep tendon reflexes may be present until about 6 years of age but are they less brisk in comparative the ankle jerks and are eventually lost. The brachioradialis reflex is usually stronger than the biceps or triceps brachii reflexes in case of DMD.

Cardiomyopathy: Persistent tachycardia and myocardial failure, is affected in 50-80% of patients with DMD.

Intellectual impairment: 20-30% has an IQ <70 in case of DMD affected children's. The learning disabilities are mostly found in affected patients but still allow them to function in a regular classroom, particularly with remedial help.

AYURVEDA AND MUSCULAR DYSTROPHY

Muscular dystrophy is cannot be directly correlated to any of a particular disease in *Ayurvedic* science. This type of phenomena may be attributed under the concept of *adibalapravritta vyadhi* as which occurs due to the *beejadustiand aatma karma*^[14] (selfdeeds), leads to *khavaigunya* of *mamsavahastrotas* causing *dhatwagni* impairment,^[15] its means *dhatwagni* play a major role in the pathogenesis of Muscular dystrophy. It is the result of an imbalance of *vata dosha*, *saptadhatu* (basic elements for formation of *garbha* both functional as well as structural - to the level of *dhatwagni*) and *ojas* considering its progressive degeneration to systemic involvement.

Pathogenesis of Muscular disorders

Vitiated *Vata dosha* play a major role in muscular dystrophy that is developed after mutation in the genome because the *Vata Dosha* is more provoked & obstructed (*Avrita*) by *Rasa*, *Raktadi Dhatus*, then mainly *Mansa* and *Meda* are affected as they are the main constituents of our body (As the body of human being is mainly supported by skeletons & muscles, which are the chief sites of *Vata Dosha*. As a result of this, *Meda & Mansa Dhatu Kshaya as well as Virldhi* (Degeneration and regeneration of muscle fibers, particularly calf muscles that is responsible for psseudohypertropy of calf muscles and hypertrophy of tongue) occurs, by which nervous tissues supplying to that affected parts lacks proper nutrition & gets deactivated. This pathophysiology leads to Muscular Disorders.

The cardinal feature is *chestahani* (frequent falling, difficulty in climbing and standing), which indicates decrease in *chalaguna* of *Vata*.

GENERAL PRINCIPLES OF TREATMENT AND PREVENTION OF MEDA-MAMSADHATU DUSTHI (MUSCULAR DYSTROPHY)

In *Ayurvedic* texts muscular dystrophy is considered under *Meda-Mamsadhatu dusthi* due to vitiated *Vata dosha* play a major role in this disease, so line of treatment manly at three levels such as:

- First line –*Srotosodhana* including *lekhana aushadhi*, *dhatwagni deepanapacana (rukshana)*
- Second line – *Dhatukshaya janyavatavyadhi Chikitsa* (to promote tissue metabolism)
- Third line – followed by *Brumhanachikitsa*.

Shodhanachikitsa

- i. *Deepana* and *pachana* (like *udvartana*, *pariseka* with *dhanyamla*^[16] at tissue level
- ii. *Sadupakarmas* (*Charka*) therapy (Six major treatment categories) consisting of *Langhana* – *Brimhana*, *Rukshana* – *Snehana*, *Swedan* - *Stambhana*.
- iii. *Snehana* (Oleation Therapy): It is indicated in *Krisha* (Emaciated) and *Vatarogas* (Neurological disorders,^[17] depending upon the application of *Snehana*, it can be divided into 2 types, viz: *Abhyantara Snehana* (Internal Oleation) and *Bahya Snehana* (Massage).

Snehapana^[18] *Snehapana* should be done with *Tikta Gruta*, *Dashamularasnadi Ghruta*:

- If there is a simple provocation of *Vata* without any *Upastambha* or *Avarana*, it should be treated at first with oral administration of unctuous preparations such as *Ghrta*, *Tailaa*, *Vasa* and *Majja*.
- The person, when overstrained by the *Snehana* should be comforted and again *Snehana* should be done with milk.
- Further he should be treated with oleated *Yusas*, meat juices of domestic, wet land and aquatic animals mixed with unctuous articles.
- Preparations of milk and *Krisara* may be given for eating.

Bahya Snehana: The muscular dystrophy is especially muscular disorders so *bahya Snehana* or massage plays a major role as following properties of the massage therapy:

- i. Massage with *Bala* oil mix with *Balaashwagadhalakshadi Taila*, *Mahanarayana Taila*, *Mahamashadi Taila*,^[19] oil should be applied which is used for relaxation of contractures and giving nourishment to the muscles, for promoting the strength of muscles and nerves system.
- ii. The strokes used in *Abhyanga* also have effects like a local increase in circulation in the treated area,^[20] which helping in the remove of despite west products to the muscles tissue.
- iii. *Snehana* pacifies *Vata*; lubricates and softens the *Dosha* and improve the digestion, regularizes bowels, improve the strength and complexion.^[21]

Shashtika Shali Pinda Sweda

1. It should be applied followed by *Snehan Karma*; muscular dystrophy is the result of *Medo-Mams* imbalance disorders that's why *SSPS* was found very effective for

nourishment as well as providing strength of the muscles and nervous tissue to the affected child.

2. *Brimhaniya Snehika* sudation performed by bolus of boiled *Shashtika Shali* (*Oryza Sativa* Linn) with *Vatahara Kwatha* and milk. *Dashamula Kwatha* as *Vatahara Kwatha* for SSPS was used due to its *Kapha-Pitta-Vatahara* properties,^[22] and *Rasanasaptaka kwatha* was its ability to alleviate *Saptadhatugata-vata* (normalize the functions of *Vayu* in all the tissues of body,^[23] due to its main ingredient is *Rasana* (*Pluchea lanceolata* Linn), which is one of the best drug in alleviating *Vata dosha*.^[24] *Nirgundi* leaves decoction is also very effective in DMD due to *Vata* and *Kapha* are the main causative factors for Muscular dystrophy and these *Doshas* can be pacified by *Tikta* and *Katu Rasa*, *Katu Vipaka* and *Ushna Virya* of *Nirgundi*.^[25]

Anuvasana Basti

Basti Krama is the best treatment for *Vata Vyadhis* (diseases occurring due to vitiation of *Vata*), when the *Vata* is controlled in the *Pakwashaya*, which is the center of administration of *Vata*, the other subtypes of *Vata* located in all the parts of the body will be automatically controlled and provide nourish to the muscles tissue and improve strength of child due to its *brmuhana* property some examples of basti which broadly used in muscular dystrophy e.i. *Mamsa rasa basti*, *Mustadiyapana basti*.

Udvardana

Udvardana by *Yava*, *Masa* coarse powder also very effective to treat hypertrophy of calf muscles and strength of muscles due to its *medo-kaphar*^[26] properties and stimulates nerve ending, relax muscles and relieves pain.^[27]

Samshan chikitshya

- i. *Samsamana* of imbalances *dosha* and *dhatu*s by appropriately planned diet, drug, & life style interventions.
- ii. *Rasayana* therapy: *Rasayana* group of herbo-mineral or gold based medicine are special *Ayurvedic* resources that increase enzymatic essence of each *dhatu* starting from *Rasa dhatu* and previous studies has shown the pure gold *bhasma* in low dose has been used successfully in the management of degenerative diseases of *mamsa* and *Majja dhatu*.^[28]
- iii. *Yougic* supports have shown definite protective influence and longer survival upon muscular dystrophy in children's.

Certain *Ayurvedic* herbs used for their *Rasayana* effects are being scientifically verified for their possible effect in the management of muscular dystrophy such as:

- i. ***Curcuma longa***: Previous studies shown the clinical implications for the pharmacological treatment of patients with Muscular dystrophy.^[29]
- ii. ***Withania somnifera***: It contains Withanolide which induces significant regeneration of axons, dendrites, pre-synapses and post synapses in the neurons, it also suppresses free radical generation and ameliorates neuronal dysfunction.^[30] It is commonly used in emaciation of children (when given with milk, it is the best tonic for children), vitiated conditions of *vata* and nervous breakdown,^[31] and induced immunomodulatory actions^[32] and longevity in the users.^[33]
- iii. ***Terminalia Arjuna***: Cardiomyopathy and arrhythmias are the major cause of cardiac manifestation in DMD and death occurs due to respiratory insufficiency and cardiomyopathy that's why the.^[34] *Terminalia Arjuna* is used in treatment of DMD due to its cardio-protective properties.^[35]
- iv. ***Tinospora cordifolia***: It is one of the most important immunomodulatory plant in *Ayurveda*,^[36] it acts as *Rasayana* (that which promotes health, provides defense against disease and promotes longevity).^[37] It also acts as an immunomodulator.^[38,39] Regular physical exercise and physiotherapy should be done along.
- v. *Yogic* exercise particularly *Pawanmuktasana* and *Bhastrica Prayanayama* is effective for prevention of respiratory problems in muscular dystrophy because respiratory disease in DMD is a major cause of mortality.^[40]

CONCLUSION

Muscular dystrophy is a genetic disorder with no specific treatment in any system of medicine and disease prognosis is unpreventable. In *Ayurvedic* classics, muscular dystrophy is characterized by *Adhi bala pravritta vyadhi* occurs due to the *Beeja bhagavayava dusti* which leads to *Mamsa vata dushti* with manifestation of vitiated *Vata Dosha*. The *Dhatupaka avastha* clearly signifies the importance of *Agni* which is responsible for the formation of next *Dhatu*. *Purva-Panchakarma therapies (Snehana, SSPS)*, with *Anuvasana Basti* is useful in the long term management of DMD. *Basti Karma* offers the *shamana* of provoked *vata* which is evident in Muscular Dystrophy as the reason brings a chain of *Dhatu Kshaya* leading to *Vata prakopa* and further *Dhatu Kshaya* due to *Vata prakopa*. Thus, administration of *Basti* should be done for the correction of *Agni*, balancing *Doshas* eliminating metabolic toxins from *Dhatu* and nourish to the various *dhatu*s. Various research works have been done

on *Vatavyadhi* with special reference to DMD in various institute of India, where it can be concluded that herbo-mineral medicine along with *Panchakarma* therapy has a major role to prevent further complication of DMD. The *Yogic* exercise with *Panchkarma* therapy is very effective for prevention as well as treatment of various complications in muscular dystrophy in children. Single *Ayurvedic* drugs possessing properties like *Medhya* (memory boosting), *Balya* (strengthening), *Rasayana* (rejuvenative), *Agnivardhana* (digestive & carminative) & *Vatadoshahara* are administered both internally and externally as a principle guideline for nourishment, followed by strengthening and rejuvenation of *Mamsadhatu*.

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