



Research Article

AYURVEDIC MANAGEMENT OF DUCHENNE MUSCULAR DYSTROPHY

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ABSTRACT

DMD is one among the most common muscular disorders. The incidence is 1:3600 live born infant boys worldwide. It is X-linked recessive disease caused by a deficiency of a normal muscle protein called dystrophin, which maintains the integrity of the muscle cell wall. Eventually irreversible progressive muscle weakness and degeneration of the skeletal or voluntary muscles, which control movement leading to lose the ability to stand, walk and loss of ambulation before 10years, with progression of the disease, most patients succumb to death in their early 20s. As there is no specific treatment in any system of medicine and the disease prognosis is unpreventable but if we start treatment in early stages of ambulant DMD boys, it may slow or stop the progressive degeneration of muscles. DMD cannot be directly co-related with any single disease in Ayurveda. All most all major neuromuscular disorders are identified with *Vata dosha*. In Ayurveda this pathogenesis can 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 study was under taken to increase functional and physical capabilities, minimizing disability to delay further progression of disease and to maintain the ambulation for longer time and to improve quality in the activities of daily living. 30 children with DMD were taken up for the study of age between 05-16years and were given treatment for 2 weeks consisting of *Abhyanga* with *Bala taila*, *shastika shali Pinda swedana* followed by *Vasti*. The study has been subjected to statistical analysis, in which the results were found to be encouraging.

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INTRODUCTION

Among the more than 20 different genetic muscular disorders, the most common fatal one is DMD. It is ubiquitous and it occurs all around the world, causing considerable hardships to affected individuals and their families. DMD is x-linked recessive disease caused by a deficiency of normal muscle protein called dystrophin which maintains the integrity of the muscle cell wall. This leads to progressive muscle weakness and exhibits the symptoms like difficulty in getting up from floor, difficulty in walking, climbing stairs, frequent falls while walk and finally loss of ambulation¹. The term dystrophy means "abnormal growth" or "faulty nourishment" or "poor nourishment" was popularised at the close of the 19th century, derived from Greek word "trophe" meaning nourishment. The incidence is 1:3600 live born infant

boys. The abnormal dystrophin gene on Xp21.1 locus and is one of the largest mammalian gene. In affected families which 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, 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. Patho physiology in brief, Dystrophin is a large, rod shaped cytoskeletal protein (molecular wt: 427000 Daltons) transcribed from a 14000 base mRNA, that is encoded by 79exons within the enormous 2.4million base DMD gene located at Xp21.2. Although dystrophin accounts for only 0.002% of total muscle protein, its absence reduction or abnormality and

changes in its associated membrane proteins appear to be the primary lesion in a no. of related myopathic or dystrophic disorders, resulting in variable phenotype of muscle wasting². Clinical presentation, Early gross motor skills such as rolling over, sitting, standing are usually achieved at the appropriate ages or may mildly delayed, Around 2-3 years of age, boys with DMD may appear to be “somewhat clumsy” due to hip girdle weakness. By the age of 3-5years they often have difficulty keeping with their peers on the play ground. In some children they can never run in their life. Toddlers might assume a lordotic posture when standing to compensate for gluteal weakness. An early “Gower’s sign” is often evident by age 3years and it is fully expressed by age of 5 or 6 years, delayed walking, frequent falls while walk, trouble in running, difficulty in climbing upstairs, toe walking, pseudo hypertrophy of calves, relentlessly progressive weakness of muscles, leading to wheel chair confinement at 12years. Death occurs usually at about 18-20years of age³. The cause of death are respiratory failure in sleep, intractable heart failure, pneumonia occasionally aspiration and airway obstruction.

This disease cannot be directly co-related with any single disease in Ayurveda. All most all major neuro muscular disorders are identified with *Vata dosha*. This disorder can be considered under *Vata vikara* due to *Bheaja dusti* because of *Adibalapravritta*⁴. The cardinal symptoms is ‘*Chestahani*’ which indicates a decrease in the *Chala guna* and *Pravarthaka chestana* of *Vayu*. Finally causes paraplegia and *Sarvanga vata*.

The important causative factors can be brought under *Aatma karmaja* and *beeja dosha*⁵. These

factors bring *Khavaigunya* at *Mamsa dhatu* levels leading to vitiation of *Vata* which causes *Bhutagni* impairment⁶. Thus these two causes attribute to DMD may be analysed as *Bheaja dusti* and *Atma karma* are responsible for the X-linked recessive disease as well as gene mutations.

SAMPRAPTHI can be sort out as it is result of *Mamsa-vata-kshaya* due to *Bheaja dosha* which leads to *Vata Vaishamya* of *Mamsa dhatu*. This vitiated *Vayu* causes improper formation of *Mamsa dhatu* by its influence on the *Dhatvagni* of *Mamsa*⁷. So depletion of *Mamsagni* causes formation of *Ama* which leads to faulty nutrition and causes progressive relentless degeneration of muscle tissue⁸. Considering DMD as *Vata vikara*, *Vasti* treatment is adopted, as this best treatment for diseases of *Vata*. The treatment modality is applied mainly to combat

Vata dosha along with drugs having *Balya*, *Rasayana*, *Agni vardhaka* & *Vatahara* properties. Any *Shodana* procedure should always be preceded by *Snehana* and *Swedana*.

- a) **ABHYANGAM**⁹–*Bala taila*.
- b) **PINDA SWEDAM**¹⁰ – *Shali dhanya (Rakta)*, *Balamoola kwath*, Milk, cloth and 2 vessels.
- c) **VASTI**¹¹– *Madhu*, *saindavam*, *Shatapuspa*, *Tailam (Bala tail)*, *Gomutram*, *Kashaya* – prepared with *Balamoola*, *Dasamoola*, *Yasti*, *Raasna quath churna*
- d) **Instruments for Vasti** – 50ml disposable syringe, Nel’s catheter 16 size.

Table 1: Method of Preparation and Administration of Vasti

Ingredients		05 - 10yrs (150ml)	11- 16 yrs (200ml)
<i>Madhu</i>		4 ml	6ml
<i>Saindavam</i>		½ g	2g
<i>Satapushpa</i>		1 g	2g
<i>Kashayam</i>	4 g }	66ml	80ml
	2 g }		
	1 g }		
<i>Tailam – Bala Taila</i>		75 ml	100ml
<i>Gomutram</i>		5 ml	10ml

The above mentioned quantity of drugs is measured based on total dosage required for the particular age group.

Method of preparation of Vasti

Step 1: *Madhu* is first taken in the *Khalwa* and then *Saindavam* is added. They are mixed uniformly till the sticky sound disappears.

Step 2: *Tailam* is added till the mixture becomes homogenous.

Step 3: *Shatapuspa* is added, mixed carefully.

Step 4: *Kwatha* prepared using above mentioned drugs in the ratio 1:8 and reduce to ¼ quantity, appropriate amount is added to *Vasti* preparation.

Step 5: *Gomutram* was added slowly & mixed uniformly.

Step 6: *Vasti dravya* was made lukewarm by keeping it in hot water.

Method of Administration

- The child was given very light food neither too *Snigdha* nor too *Ruksha*. It was checked that the child has passed the stool routinely in the morning. As *Abhyangam* with *Bala taila* and *Pinda swedana* has been done, after these *Purvakarma* the patient was made to lie in left lateral position with left lower extremity straight and right lower extremity flexed on knee and hip joint on the *Vasti*

table. Then the anal orifice was smeared with oil for lubrication.

- Required quantity of *Vasti dravya* was taken as per age and then was taken into syringe connected to Catheter (Nel's catheter - 16 size). Then the Catheter was smeared with oil and after removing air bubbles the Catheter was introduced into rectum for about 4 inches and then *Vasti dravya* was gently pushed inside.
- During administration of *Vasti*, the child was made to take deep breath through his mouth.
- After the procedure the child was made to lie down on supine posture after gently tapping his buttocks. Then the child was made to lie down and mother/ attendant were advised to notice the time of retention.
- The same procedure is done for 11 days. The patients were advised to take usual food.

The review of follow up is done & asked to come for next sitting with gap of 2 months

Clinical Plan

Patients were selected from OPD /IPD of S.V. Ayurvedic hospital in *department of Kaumarabhritya*, based on the inclusion criteria and clinical symptoms.

Inclusion Criteria

- Patients between the age group 5 - 16 yrs.
- Children having mild to moderate physical disability
- Children with DMD.

Exclusion Criteria

- children with advanced stage of DMD with cardio myopathies, respiratory complications
- Congenital muscular dystrophies
- Myelopathies
- Congenital myopathies
- Metabolic muscle diseases
- Becker'muscular dystrophy

Diagnosis: already diagnosed cases were taken and also repeated serum creatine kinase muscle enzyme, which is sensitive indication of active muscular dystrophy.

Study design

All the patients were given *Vasti* for 11 days. All were subjected to *Abhyangam* with *Bala taila* and *Pinda swedana* with *Rakta shali* as per classical method prior to *Vasti* therapy for 14 days. During this period oral medication is also given which is continued between the sittings also

Table 2: Treatment plan

Group	Treatment	Dose	Duration
05- 10 Yrs	<i>Abhyangam</i> with <i>Bala taila</i> (20 - 25minutes)	30 - 40 ml	14 days
	<i>Pinda swedam</i> (10 - 25 minutes)		14 days
	<i>Vasti</i>	150ml	11 days
11- 16 Yrs	<i>Abhyangam</i> with <i>Bala taila</i> (20 - 30 minutes)	40 - 50 ml	14 days
	<i>Pinda swedam</i> (20 - 35 minutes)		14 days
	<i>Vasti</i>	200ml	11 days

Table 3: Assessment criteria

Criteria	3- severe	2- Moderate	1- Mild	0- No complaint
Symptoms				
Falling down while walk	>5 times /day	1-5 times/day	1-5 times/week	ceased
Difficulty in walking	Difficulty in walking even small distance	Difficulty in walking long distance with stops	Difficulty in walking long distances without stops	Able to walk without difficulty
Difficulty in climbing stairs	With assistance	With support	Without support	Without support & difficulty
Muscle power in the upper limbs	G ₁	G ₂	G ₃	G ₄
Toe walking	continuous	often	occasional	absent

2) Barthel Index:¹²

Grooming

- Needs help with personal care
- Independent face/hair/teeth/shaving/brushing

Feeding

- Unable
- Needs help cutting/spreading butter, etc
- Independent

Toilet use

- Dependent
- Needs some help, but can do something alone
- Independent (on and off, dressing, wiping)

Bathing

- 0- Dependent
- 1- Independent

Dressing

- 0- Dependent
- 1- Needs help (but can do about half unaided)
- 2- Independent (including buttons, zips, laces)

Stairs

- 0- Unable
- 1- Needs help (verbal/physical/carrying aid)
- 2- Independent

Transfer

- 0- Unable- no sitting balance

- 1- Major help (one or two people, can sit)
- 2- Minor help (verbal or physical)
- 3- Independent

Mobility

- 0- Immobile
- 1- Wheel chair independent
- 2- Walks with help of one person
- 3- Independent (but may use any aid)

The efficacy of therapy was studied on various parameters and the results were derived after subjecting to statistical methodology by using Student paired t-test.

Results

Table 4: The effects of the therapy on various parameters were as follows

S. No	Characteristics	No. of Observations	Mean Score		SD	P	Remarks
			BT	AT			
1	Falling down while walk	27	2.5	0.4	0.9595	<0.001	Highly Significant
2	Difficulty in walking	28	2.3	1.4333	0.7761	<0.001	Highly Significant
3	Difficulty in climbing stairs	26	2.1	1.3	0.6644	<0.01	Highly Significant
4	Muscle power in the upper limbs	30	2.4667	0.8667	0.7701	<0.001	Highly Significant
5	Toe walking	27	2.4667	1.1	0.7112	<0.001	Highly Significant

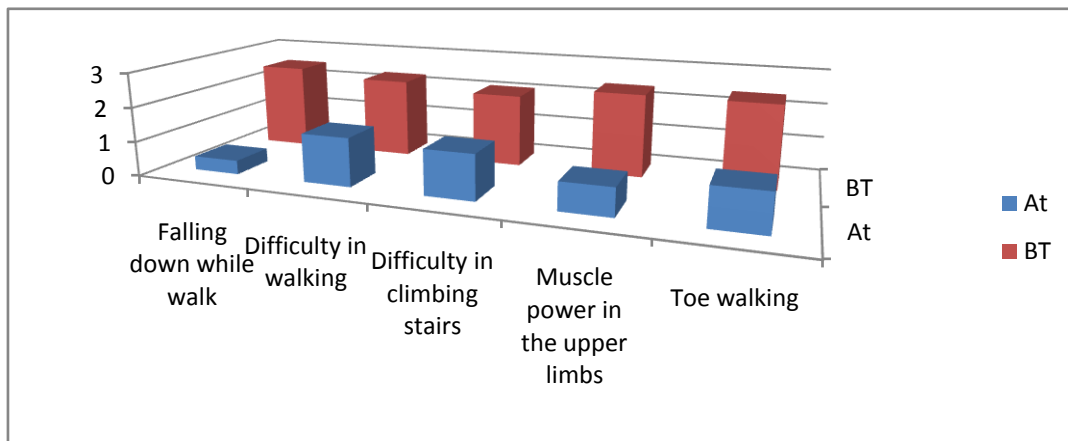
Improvement in the ADL

Improvement in the ADL was assessed by Barthel index which is useful for measuring the functional status of the individual. In this score total possible score ranges from 0 to 20. If used to measure improvement after treatment, changes of more than two points in the total score reflects a probable genuine change and is also likely to be reliable. After treatment 04 points improved in two cases, 03 points improvement in 14 cases, 02 points improved in 10 cases, one point improved in two cases while remaining 02 cases were unchanged.

The overall change in the total score is 2.6 which denote genuine change after the treatment.

Table 5: Percentage of overall improvement in each ailment after treatment in 30 patients with respect to assessment criteria

Ailment	Percentage improvement
Falling down while walk	84%
Difficulty in walking	37.68%
Difficulty in climbing stairs	38.09%
Muscle power in the upper limbs	64.86%
Toe walking	54.05%



DISCUSSION

By this treatment encouraging progress observed in the following major complaints Frequent falls while walk completely ceased in 18 patients, good progress in 6 cases, while remaining 3 children got moderate results. Overall 84% of progress is observed may be due to increased muscle power in the both lower limbs and softening of the pseudo hypertrophy of calves. After treatment there was good result in 15 patients with improvement in quality of walking, in 7 patients there is moderate improvement and in 6 patients no improvement.

Overall percentage of progress is 37.68%

Difficulty in climbing stairs in 09 there was moderate progress and no improvement in 07 patients.

Overall percentage of progress is 38.09%. It was statistically highly significant.

Muscle power in the upper limbs: There was an overall progress of 64.86%. Among 27 cases there is excellent improvement in 03 cases, 24 cases got good improvement, while remaining 03 cases got moderate improvement. After treatment most of the patients were able to raise both hands by overcoming the gravity due to improved muscle power.

In Toe walking there was excellent result in one case with complete correction of toe walking. In 14 cases there was good improvement, moderate results in 08 cases and remaining 04 patients had no improvement. This result is due to reduction of spasticity in the Achilles tendons and by lengthening of already contracted connective tissue. And there may be reduced hyper tonus in calves, increased muscle power.

Overall progress was 54.05%.

The overall change in the total score of Barthel index is 2.6

Effect of therapies on other ailments

Contractures

Most often involve the Ankles, Knees, Hips and elbows, because the muscle fibers shorten and fibrosis occurs in connective tissue. Among 30 one child had contractures at knees. He got good result with improvement in the extension of knees (increased range of motion).

Pseudo hypertrophy of calves

As per the observations all young ambulant boys had the complaint of pseudo hypertrophy of calves with hard consistency. After treatment there was marked change in consistency i.e. calves become soft and size is also reduced in all the cases of DMD. Frequent falls and toe walking has come down due to decreased tonicity in the calves.

Gower's sign

An early Gower's sign is often evident by age 3 years and is fully expressed by age 5 or 6 years. This is

a clinical assessment of pelvic muscle weakness in which considerable result was not observed and Gower's sign is remained persisted.

All these results were obtained by *Abhyanga*, *Pindasweda* and *Vasti*. Their mode of action presumed to be as follows.

Abhyanga stimulates circulatory system, enhancing cell activity, increases blood flow, vasodilatation result in nourishment of the muscles, strengthening the muscles, releasing facial constrictions, assisting in reducing connective tissue thickening and provide flexibility by decreasing fibrous adhesions from muscle tissue injury. Due to *Abhyanga* progress is seen in reduction of toe walking, relieving contractures, nourishment of atrophied muscles, increasing muscle power and assisting muscle tone.¹³

Heating the skin has been demonstrated to produce a decrease in gamma activity. With a decrease in gamma activity, the stretch on the muscle spindle would be less, thus reducing afferent firing from the spindle. This indirect method ultimately results in decreased alpha motor neuron firing and thus less muscle spasm. Elevating muscle temperature can also alter strength and endurance. Heating can result in decreased joint stiffness and increased tissue extensibility, thus facilitating ease of motion and gains in range of movements¹⁴.

Vasti is having two actions, expelling the *Dosa* & nourishing the body as it is indicated in *Gambhiragata vata* also¹⁵. First, potency of the *Vasti* drugs gets absorbed to have its systemic action. Its second major action is related with the facilitation of excretion of morbid substances responsible for the disease process into the colon, from where they are evacuated.

CONCLUSION

The present study was under taken to increase functional and physical capabilities, minimizing disability to delay further progression of disease and to maintain the ambulation for longer time and to improve quality in the activities of daily living.

30 children with DMD were taken up for the study of age between 05-16 years and were given treatment for 2 weeks consisting of *Abhyanga* with *Bala taila*, *Shastika shali Pinda swedana* followed by *Balamooladi Vasti* which was continued even during the interval period of 2 months.

Based on the observations and results it is concluded that

- The given treatment is efficacious.
- Frequent fall down while walk was stopped in many patients, while it was reduced in some other patients. Overall 84% of progress is observed.

- Considerable improvement in difficulty (37.68%) in walking and climbing stairs (38.09%) was observed.
- The muscle power in the upper limbs improved (64.86%) significantly which was expressed by raising hands above the shoulder without difficulty.
- Toe (54.05%) walking also shown considerable reduction.
- Contractures also released to some extent.

There is overall progress observed functionally, physically, psychologically, and nutritionally in these children with minimum or no side effects, which is much economical.

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