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AYURVEDIC MANAGEMENT OF LIMB GIRDLE MUSCULAR DYSTROPHY: A CASE REPORT

Athreya Sharma M[1], Niranjan Rao[2], Swathi N[3]

[1]MD Scholar 3rd Year, [2]Dean PG and PHD Studies, HOD, [3]Assistant Professor
PG department of panchakarma, SDM college of Ayurveda, Udupi, Karnataka, India

Corresponding Author: athreya95@gmail.com

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ABSTRACT

Limb-girdle muscular dystrophy is a group of rare genetic disorders characterised by progressive wasting and weakness of the voluntary muscles in the limb girdle area whose many subtypes are identified based on mutations in the genes. There is no definitive cure for the disease which on progression in successive years would lead to disability. A case of autosomal recessive limb girdle muscular dystrophy in a child aged about 10 years which can be diagnosed with *Mamsagatha vaata* leading to progressive *Maamsa shosha* managed through the *Ayurvedic* line of management by *Rajayapana Basti* with follow-up period of 3 years of *Maathra Basti* through continuous monitoring of serum creatine phosphokinase.

Keywords: Limb girdle muscular dystrophy, autosomal disorder, *Maamsagatha Vata, Bheeja Dushti*, creatinine phosphokinase, *Rajayapana basti, Maathrabasti, Agni Chikithsa Lepa.*

INTRODUCTION

Limb girdle muscular dystrophy type 2 is an autosomal recessive inherited neuromuscular disorder affecting children at young age whose prevalence is estimated at the most 1 in 14,500. It is characterised by proximal muscle weakness primarily affecting lower limbs presenting with difficulty to climb stairs, running and increased fatigue. This gradual progress leads to disability of patient causing dependability on a wheelchair in 20-30years of a lifetime.

Diagnosed based on high creatine phosphokinase levels and EMG to detect myopathic patterns of the disease.

Clinically based on predominant proximal muscle involvement, independent walking achieved at one point in a lifetime, muscle fibre loss, dystrophic changes in muscle histology, inheritance pattern. Laboratory findings constantly monitored are creatinine phosphokinase which shows massive elevations.

The disease shows slow progression in patients of young age but has no definitive cure or plan of management in any medical science as of now leading to neglect and reduced quality of life in these children affected by the disorder.

In *Ayurveda*, the understanding of this particular case in person can be understood as *Maamsagatha Vaata* due to the presentation of symptoms at the age of maturation of *Maamsa Dhathu* and vitiation in the functions of *Vaata* like *Cheshta*. Though the *Nidhaana* for the same can fall back to *Bheejaavayava Dhushti*. The *Upashoshana of Maamsa Dhathu* due to subsequent *Vaata Prakopa* leads to progressive *Mamsa Dhathu Kshaya*. This *Dhathu Kshaya* caused by *Bheeja Dhushti* leading to *Vaata Prakopa* further progresses the *Vyadhi* making the patient *Durbala*.

Thus, the treatment for the same is in line with *Vaata Shamana* and *Dhathu Poshana* by choice of *Maathra Basti* and *Rajayapana Basti.*

CASE HISTORY

The child aged about 8 years, complaints of inability to get up from squatting position without support noticed by parents since 3 months. The child was born through caesarean section, new-born baby cried soon after birth and weighed 3 kgs. History of repeated fever at an interval of 15 days to 30 days at the age of 6 years and child's parents observed weight loss, usage of hands to stand up from squatting position and climb stairs with hands on the knees from that time the child is not able to achieve good grades. Repeated falling while running or walking. The patient consulted KMC Manipal in 2015. On investigation cpk 13748 U/L. Mutation analysis showed no deletion or duplication in heterozygous or homozygous state in dmd gene. On clinical examination, she had bilateral calf muscle pseudohypertrophy with an absent knee jerk and proximal muscle weakness in lower limbs and Gowers's sign was positive. EMG suggested a myopathy pattern.

General examination		
	Nadi - 80/min *Mala* - Once a day *Moothra - Samyak 3-4* times per day *Jihva -Aliptha* *Shabdha – Prakrutha* *Sparsha – Prakrutha* *Druk – Prakrutha* *Akruthi -Krusha* *Saara – Madhyama* *Sathva - Avara* *Samhanana - Avara* *Ahaara shakthi - Avara* *Vyayama shakthi – Heena*	Pallor – Present Icterus – Absent Lymph nodes – Not palpable Clubbing – Absent Oedema – Absent Weight -17kg
Systemic examination		
CNS	Conscious well oriented, Higher mental functions intact	
	Cranial nerves and sensory-No Deficits	
	Involuntary movements: Absent	
RS	NVBS heard	
CVS	S1S2 heard	
P/A	Soft Non tender No organomegaly	
Muculoskeletal System	Inspection-Musculature: Calf muscle Pseudohypertrophy Waddling gait seen No Muscular twitching	
	Palpation-No pain or tenderness in calf muscles Muscle power 4/5 in bilateral lower limbs Tone –Overall hypertonia	

	Reflexes-Deep tendon reflexes diminished Upper extremities and patella Intact-Achilles' reflex, Superficial reflexes
	Signs-Gowers's sign: Positive

Investigations did before the treatment:

24.1.2019-Mutation analysis: Duchene muscular dystrophy could not be confirmed in her

17.4.2019-DNA test report impression: Limb girdle muscular dystrophy-2

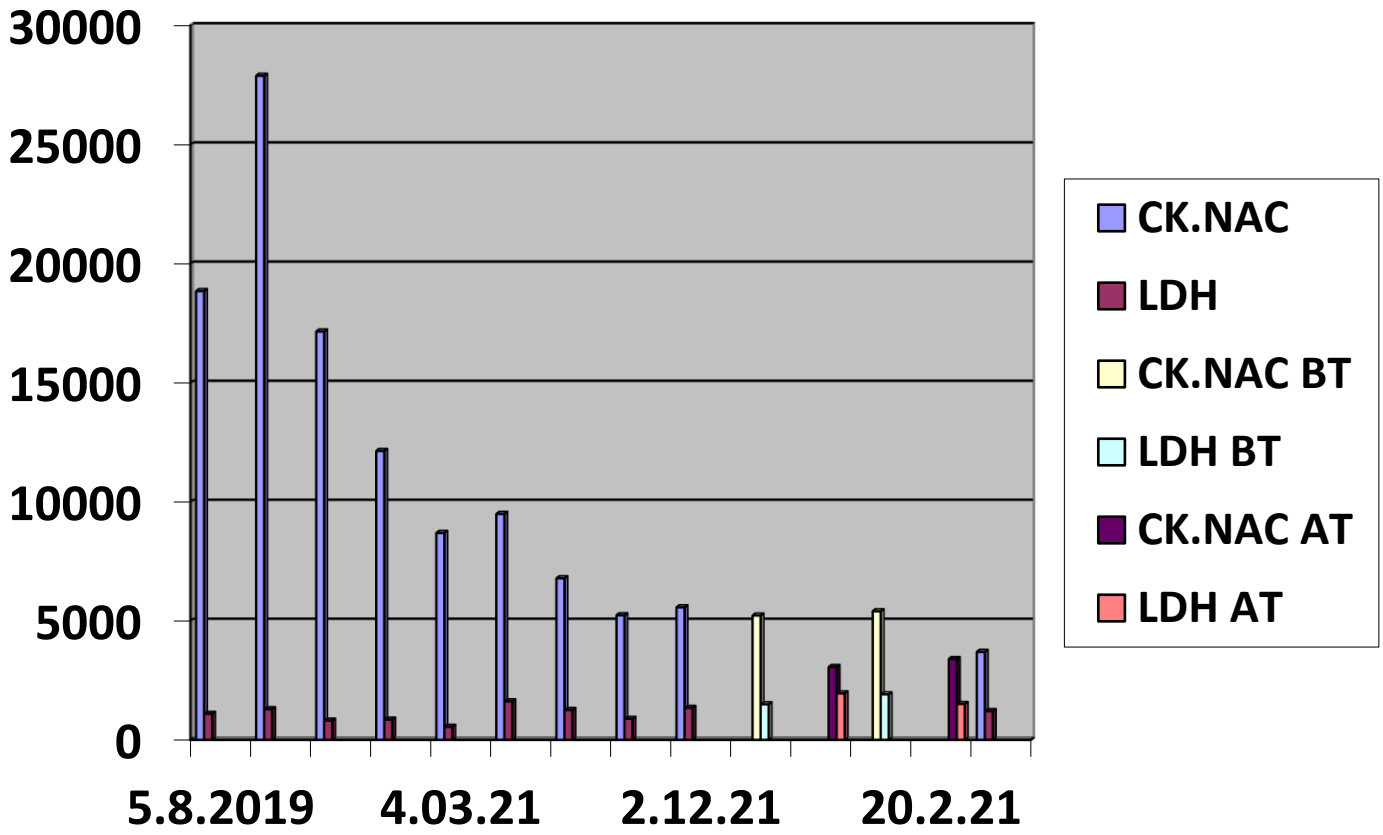
05.08.2019- Creatine phosphokinase 18,850 u/l

10.10.2019-Creatine phosphokinase 27,885u/l

Intervention planned:

Treatment plan				
Panchakarma *05.08.2019 to 02.12.2020*	*Alepa*	Ingredients: *Agnimantha (Kshudra and Bruhat), Nirgundi, Papata, Bandha, Ttulasi* as wet drugs. *Haridra, Maricha, Lashuna, Shunti* and *Lavanga* in dry form	External application for 7 days for 4hours daily	*Laghu ahara* advised *(Manda and Yusha)*
	Mathra basti	*10ml Dhanvanthara taila*	*10ml Mathra Basti* given after lunch for 7 days	*Basti* retention observed between 6-12 hours
2.12.2020 to 28.07.2021	*Rajayapana basti*	*Rajayapana Kwath Choorna- Mustha, Ushira, Bala, Aragvada, Rasna, Manjishta, Trayamana, Punarnava, Paata, Vibhithaki, Guduchi, Shalaparni, Bruhathi, Thiktha, Kantakari, Prishniparni, Gokshura* *Rajayapana Basti Kalka Choorna- Shathapushpa, Madhuka, Kutajaphala, Daruharidra, Priyangu* *Madhu, Moorchitha Tila Taila, Mamsa Rasa, Saindhava*	Quantity-120ml/day Given after light food	*Basti* retention period between 5-10minutes
	Mathra basti	*Dhanvanthara Taila 10ml /day*	*10ml Mathra Basti* given after lunch	*Basti* retention period between 6-12 hours
Internal Medication				
05.8.2019 to 28.07.2021	*Kooshmanda Rasayana* 1teaspoon twice a day *Yogaraja Guggulu* ½ twice a day			

Date	CK.NAC	LDH
5.8.2019	18850	1093
10.10.2019	27885	1287
11.11.2019	17155	809
6.1.2020	12140	850
4.3.2020	8695	1546
13.7.2020	9495	1619
26.8.2020	6790	1255
1.9.2020	5237	882
2.12.20	5570	1344
16.12.20	5225	1495
22.12.20	3059	1951
17.2.21	5406	1910
20.2.21	3396	1506
28.07.2021	3695	1205



DISCUSSION

Limb girdle muscular dystrophy was first published in 1954 describing a group of heterogeneous conditions distinct from Duchene muscular dystrophy. The autosomal recessive variant is caused by a mutation in the alpha-sarcoglycan gene.

Alpha sarcoglycan gene encodes a component of dystrophin-glycoprotein complex which is critical to the stability of muscle fibre membranes and the linking of the actin cytoskeleton to the extracellular matrix. Though its expression is thought to be restricted to striated muscles only. The creatine phosphokinase presented with 18,850 u/l whose normal values for females' range at 26-192 u/l. In *Ayurvedic* perspective will indicate *Maamsa Kshaya* with functional disruption in *gamana* and other similar functions of *Vaata* related to the *Paada* lead to the conclusion of M*aamsagatatva* of *Vaata*.

The symptoms presenting at the age of development of muscles and maturation of muscles indicate to *Dushti* of *Bheeja Bhaaga Avayava* leading to the diagnosis of *Maamsagatatva* of *Dushta Vaata* during *Garbha* formation leading to the loss of muscles.

The prognosis is *Asaadhya* since *Nidhaana* traverse back to *Bheejabhaagavayava Dushti*. Laying importance to the management by *Parivarjana* of *Nidhaana* contributing to the disease, *Shamana* of *Prakopita Dosha* and *Dhathu Poshana*.

Nidhaana Parivarjana involves curtailing the physical activity of children which can further lead to *Vaata P*rakopa or damage to muscle fibres which have naturally lower stability.

Shamana of *Prakupitha Dosha – Vaata Dosha. Maathra Basti* was first chosen as a line of treatment for being *Sneha Basti* and indicated in *Durbala* and *Vaata Vyadhi*, also the practically easiest way of administration of *Sneha* in children affected by *Vaata Vyadhi*.

Alepa consists of *Vaata Shamana* drugs such as *Nirgundi, Agnimantha, Sarshapa, Maricha, Lashuna* among ingredients.

Once the patient presents stability in symptoms from falling during walking to walking with limping,

climbing stairs withholding of external support to climbing stairs supporting hand on knees independently, physician capable of shifting focus on *Dhathu Poshana* along with *Vaata Prashamana*.

Shamana of *Vaata Dosha* along with *Dhathu Poshana* – *Rajayapana Basti* course in *Kaala* pattern which includes 6 *Rajayapana Basthi* alternating with *Maathra basthi*. "*Maamsam Maamsena Vardhathe*"- *Maamsa Rasa* is one of the ingredients of *Basthi Dravya* thus *Rajayapana Basthi* is administered at a reduced dosage of 120ml / day does *Poshana* of *Maamsa Dhathu*.

The internal medications with these treatments included *Kooshmanda Rasayana*, A *Rasayana yoga* which has targeted action on *Maamsa Dhathu* and *Yogaraja Guggulu* for its targeted action on *Vaata Prashamana*.

CONCLUSION

This management of limb girdle muscular dystrophy for a period of 3years by careful observation and monitoring of signs and symptoms along with blood parameters is described. In the time when the sole treatment thought of for the disease exercise therapy to have backfired and gene therapy was not conclusive and heavy on the pocket the treatment management through *Ayurvedic* principles give clarity and confidence in managing and providing a better quality of life to the child and parents inclusively.

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