

## Limb Girdle Muscular Dystrophy (LGMD) and its Management through Unani system of medicine- A Case Study

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### ABSTRACT:

Muscular dystrophies refer to a group of hereditary progressive diseases, each with unique phenotypic and genotypic features. Limb-girdle muscular dystrophy (LGMD) and fascioscapulohumeral muscular dystrophy are the most common types of muscular dystrophy. LGMDs are a heterogeneous group of non-congenital genetic disorders that are caused by mutations in multiple genes and are generally characterized by weakness and wasting of the shoulder and pelvic girdle muscles. The clinical course of LGMD ranges from severe forms with disease onset and rapid progression within the first decade of life to milder forms with later onset and slower progression. Unani System of Medicine (USM) relies on the *Naẓariya-i-Akhlāt* (Concept of Humours), in which the four *Akhlāt* (dam (blood), *balgham* (phlegm), *safra'* (yellow bile), and *sawdā'*) are proportionately found in the body during the state of health, but when there is any dyscrasia, disequilibrium occurs in this proportion and diseases arise. According to USM, abnormal *balgham* or excess *burudat* and *rutubat* lead to *murkhi* (pseudohypertrophy), and abnormal *hadm udwi* leads to *Istirkeba'*. A condition where the muscles of an organ or the whole body become flaccid and are not able to perform function is called *Istirkeba'*. USM endeavors to use simple and natural means to cure diseases. The four main therapeutic modalities are: *Ilaj-Bi'l-Tadbir* (regimenal therapy); *Ilaj-Bi'l-Ghidba'* (dietotherapy); *Ilaj-Bi'l-Dava'* (pharmacotherapy), and *Ilaj-Bi'l-Yad* (surgery). The case study suggests that LGMD can be adequately managed with Unani System of Medicine by using pharmacotherapy and regimenal therapy.

**KEYWORDS:** Humours, *Istirkeba*, Limb-Girdle Muscular Dystrophy, Neuromuscular disorders, Regimental Therapy.

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### Quick Response Code



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**INTRODUCTION:**

Muscular Dystrophies (MDs) refer to a group of hereditary progressive diseases each with unique phenotypic and genotypic features. The prognosis of dystrophies is slow progressive weakness, although the severity and course is available between and even within subtypes. Some are associated with cardiac and respiratory muscle involvement, which leads to cause of mortality.<sup>[1]</sup> There are so many types of muscular dystrophy among which Fascio-scapulohumeral Muscular Dystrophy (FSMD) and Limb-Girdle Muscular Dystrophy (LGMD) are more common. Limb-girdle muscular dystrophy is a diverse, genetic muscle disorders which can occurs in any age group indiscriminately, primarily it's begin with weakness and wasting of the proximal limb (the hip/shoulder girdle) musculature.

The Unani System of Medicine (USM) is based on the *Nazariya-i-Akhlāt* (Concept of Humours), which states that when the body is in a state of health, the four *Akhlāt-dam* (blood), *balgham* (phlegm), *safra'* (yellow bile), and *sawdā'* are found in proportionate amounts; however, when there is dyscrasia or disequilibrium in this ratio, diseases develop.<sup>[2]</sup>

The terms sanguine, phlegmatic, choleric, and melancholy on the prevalence of the corresponding humour disclose an individual's *Mizāj* (temperaments). Temperaments are allocated to the humours themselves. Blood is *Harr* (hot) and *Ratb* (moist); phlegm, *Bārid* (cold) and *Ratb*(moist); yellow bile, *Harr* (hot) and *Yābis*(dry); black bile, *Bārid* (cold) and *Yābis* (dry).<sup>[3, 4]</sup>

According to Unani medicine, the human body is thought to consist of the following seven elements, each of which is closely related to an individual's state of health:

Element (*Arkan*): States of matter and the materials inflowing into and forming a part of the whole thing in the universe, Temperament (*Mizāj*): Bodily temperament, Humour (*Akhlāt*): Body fluids, Spirit (*Rūh*): The vital force or life force, (*A'da'*): Fully mature organs, Power (*Qumwa*): The bodily powers or faculties and Function (*Af 'al*): The corporeal functions.<sup>[4]</sup>

We know that the two main components of the body that support life are *Khilt* (e.g., blood) and *Rūh*(spirit/prana). The *Rūh* has been compared to genetic material that contains life information necessary for a variety of functions. The biological power that sustains life at the cellular level and facilitates the elimination of metabolic pollutants is rooted in the dam, or blood. Beyond blood, *Rūh* is the driving power that circulates to the cellular level in tandem with blood. The simultaneous flow of *Rūh* and blood is an expression of *Hayāt* (life). The contraction and relaxation of muscle fibers or muscular activity is brought on by this *rūh*.<sup>[5]</sup>

Galen considers *Balgham* as having a close resemblance with blood. Its availability is made parts of the body by its circulation along with the blood. Since, *balgham* includes: intracellular fluids, lymph, cerebrospinal fluids, digestive juices, seminal fluids, plasma protein, and endocrine secretion. But when *Balgham Taba'i* (normal phlegm) turns into *balgham Ghayr Taba'i* (abnormal), the chemical changes taking places in *Haḍm udwi* which has significant role in sustainability of life and cell functioning during this process certain nutrients and substrates are required to be part of body, wastes and synthesized products of the cells result to come out. Protein synthesis under the influence of Gene (DNA) can come under this function.<sup>[6, 7, 8]</sup> Pseudohypertrophy (*Murkhi*)

caused by the *Balgham-i-Ghayr taba'i* (Abnormal phlegm) or excessive *Burūdat* and *Rutūbat*, as well as abnormal *hadm udwi* which leads to *Istirkhā'*. *Istirkhā'* and *Huẓāl* (Emaciation or cachexia) defined as condition where muscles of an organ or the whole body become flaccid and are not able to perform their functions. It may be caused by (a) nutritional deficiencies e.g. intake of low-fat diet (b) weakness of the faculties of stomach due to its cold temperament (c) after discontinuation of regular and heavy exercise (d) the bilious temperament of a person (e) liver dysfunction (f) invasion of worms (g) overindulgence in catabolism e.g. in case of chronic debilitating illnesses, excessive grief, sorrow, etc.<sup>[9]</sup>

Zakaria Razi (865-925 AD) critically assessed, in his book *Al-Havi Fi'l Tibb* Volume-I, Chapter-I (An Encyclopaedia of Medicine).<sup>[10, 11]</sup> The term *Istirkhā'* as mentioned in Unani classical literature, there clinical presentation resemble to MD (muscular dystrophy, a term first coined by Erb in 1891).<sup>[12]</sup>

### **CASE REPORT:**

A 45 years old male previously diagnosed with Limb-girdle muscular dystrophy was registered through the OPD of Central Research Institute of Unani Medicine (CRIUM), Basaha, Kursi Road, Lucknow. He confined on wheelchair, came in the OPD on 03 March 2022 with the complaint of pain in lumber region, inability to stand and walk without support, and painful abduction of both arms. He was diagnosed with LGMD along with Chronic Renal Failure, Chronic Liver Disease, and Portal Hypertension at Department of Neurology, SGPGI, Lucknow. The patient was also HBsAg positive.

After taking the detailed history it was revealed that he started treatment from local

doctors and different orthopedician for 2 years. Thereafter he was referred to SGPGI, Lucknow where he was diagnosed with LMGD along with other co-morbidities. As per the medical record of the patient, his treatment started in SGPGI from 01.07.2021, and was on the following medications: NSAIDs (Local & Systemic), Corticosteroids, Calcium and Vit-D3.

### **History of present illness:**

According to the patient's statement, he complained of pain in the lumber region, inability to stand and walk without support and painful abduction of both arms. He started treatment from local doctors and different orthopaedics for 2 years. Thereafter he was referred to SGPGI, Lucknow where he was diagnosed with LMGD along with other co-morbidities. He went under treatment different doctors and hospitals but could not relief completely.

### **H/o past illness:**

The patient has a positive history of Chronic Renal Failure, Chronic Liver Disease, and Portal Hypertension, D.M, HBsAg positive.

### **Family History:**

No any history of parents and siblings related to such type of complaint was found.

### **History of Treatment:**

Patient was on NSAIDs (Local & Systemic), Corticosteroids, Calcium and Vit-D3 medication since he was diagnosed.

### **Personal history:**

Vegetarian, Non alcoholic, non-smoker and non tobacco chewer, appetite was decreased, Bowel-constipated, Bladder clear.

### **General Physical Examination:**

Patient was a young average heighted male apparently unhealthy, confined on a wheelchair; his blood pressure was 144/82mmHg, Pulse Rate 95bpm, Respiratory Rate 18/min. Weight 50.1kg SPO2 98%, Temperature 98.4F. Pallor, icterus, cyanosis and clubbing of fingers not

present and no any lymphadenopathy was found.

**Systemic Examination:**

1. **Respiratory System:** No abnormality was detected. Air entry was bilaterally symmetrical.
2. **Cardiovascular System:** S1 S2 Audible, No abnormal sound.
3. **Per Abdomen:** Soft. No distension.
4. **Central Nervous System:** Conscious of time, person, and location. Unable to walk, confined on a wheelchair.

**THERAPEUTIC INTERVENTION:**

The management carried out on the principal of treatment (*Usul-i-Ilaj*) considering the patient’s co morbidity and deep anxiety due to prolong illness. The goal of the therapy was to first give relief in pain an anxiety, besides the counselling, he was given *Musakkināt* (sedatives) and *Muḥallilat Advia* (resolvent drugs) locally and orally, treated symptomatically as well his co-morbid conditions accordingly.

Since the patient was already diagnosed LGMD, and came along with the features of Muscular Dystrophy, the patient was registered and managed according to the basic *Usul-i-ilaj* (Principal of treatment) of *Istirkhā’*, mentioned in our classical Unani literature which are as follows: *Ilāj-Bi’l-Tadbīr* (Regimenal Therapy), *Ilāj-Bi’l-*

*Ghidba*(Dietotherapy), *Ilāj-Bi’l-Dawā’*(Pharmacotherapy) and *Ilāj-Bi’l-Yad* (Surgery).<sup>[13]</sup> Principle medications given over the time period from March 2022 to November 2022 are mentioned in Table No 01.

**OBSERVATIONS AND RESULT:**

After two months of therapy, there was a noticeable improvement in discomfort, limb motions, power and reflexes. The VAS scale was used to measure the patient's pain, and the Gardner-Medwin and Point Walton ten point scales were used to measure the patient's walking capacity. Table number 02 illustrates the progress of the patient experienced over time. Following 341 days of therapy through OPD with fortnightly follow-up, the patient was admitted to the IPD section of CRIUM, where he received regimental therapies and medicine in accordance with the previously mentioned *Usul-i-Ilaj* (Principle of Management).

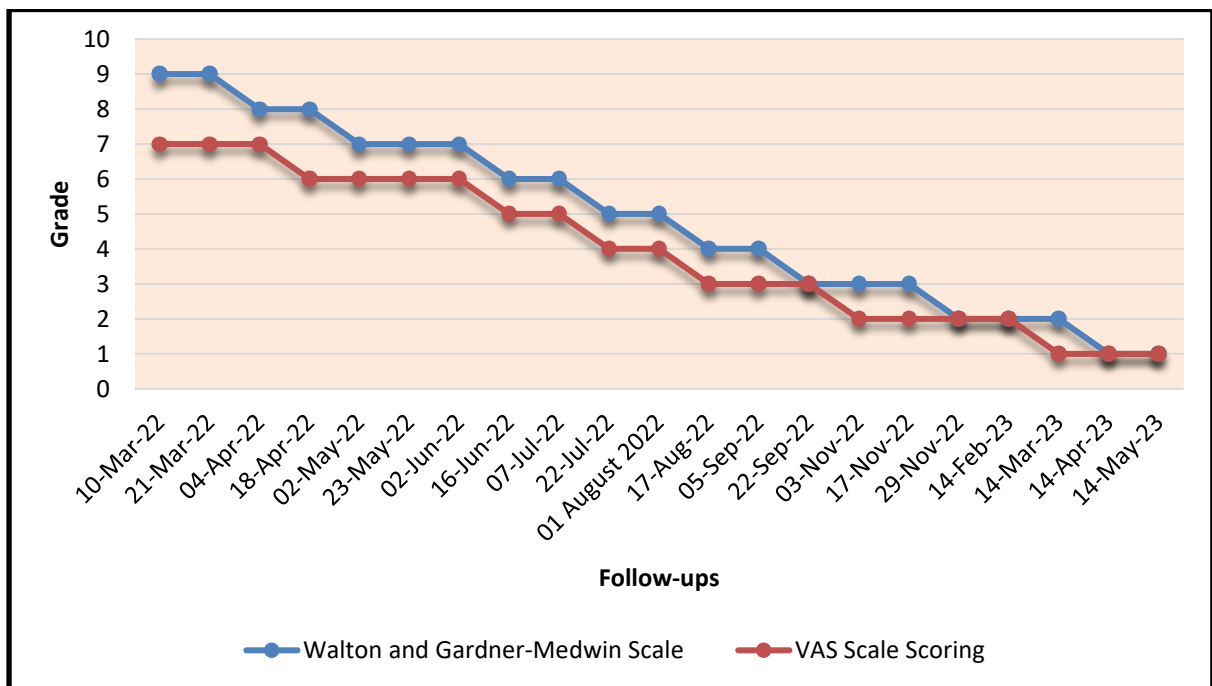
He was given instructions for a special *Riyādat* (exercise) that involved starting at 10 minutes and working your way up to 20 minutes. He was also told to perform daily medicated steam fomentation for 20 minutes all over the body. Weekly *Dalk-i-Muḥliqa* (gliding massage) was administered with *Roghan-i-Babūna* from the cervical to the lumbar regions during the IPD hospitalization.

**Table-1: Medication Summary:**

Formulations	Dosage and Timing
<i>Ḥabb-i-Sūranjān</i>	3 Tab x TDS
<i>Ḥabb-i-Asgand</i>	2 Tab x BD
<i>Ḥabb-i-Ambar Momiya’i</i>	1 Tab x OD
<i>QursDhayabitusKhās</i>	2 Tab x BD
<i>Caṭ Dabūdulward</i>	2 Tab x BD
<i>Chandraprabhavati</i>	1 Tab x BD
<i>Roghan-i-Mom</i>	For local application
<i>Roghan-i-Malkangini</i>	For local application

**Table-2: Pain and walking capacity scoring:**

	10-Mar-22	21-Mar-22	04-Apr-22	18-Apr-22	02-May-22	23-May-22	02-Jun-22	16-Jun-22	07-Jul-22	22-Jul-22	01-Aug-22	17-Aug-22	05-Sep-22	22-Sep-22	03-Nov-22	17-Nov-22	29-Nov-22	14-Feb-23	14-Mar-23	14-Apr-23	14-May-23	
<b>Walton and Gardner-Medwin Scale</b>	9	9	8	8	7	7	7	6	6	5	5	4	4	3	3	3	2	2	2	2	1	1
<b>VAS Scale</b>	7	7	7	6	6	6	6	5	5	4	4	3	3	3	2	2	2	2	1	1	1	1



**Figure-1: Progress of the patient experienced over time**

**DISCUSSION:**

Since many musculoskeletal disorders are autoimmune but the LGMD’s main pathology is gene mutation causing autosomal, hereditary disorder. At present, there is no cure for LGMD. There are supportive treatments that can help to

manage the symptoms for the treatment of LGMD. Western medicine mainly has the following treatment measures, such as the application of NSAIDs- Local & Systemic use of Corticosteroids, Calcium and Vit-D3. Thus, the MD patients in our case study were treated in accordance with the *Usūl-i-*

*Ilāj of Istirkhā'*, which is referenced in our traditional Unani literature. *Mundij-Mushil* therapy in addition to the regimen that includes medicated steam treatment throughout the body, frequent *Riyādat* and gliding message with the oils.

The polyherbal Unani formulation and their bioactive constituents have the immunomodulator properties and various pharmacological actions. These include: Nervine tonic (*Muqanwi-i-A'sāb*),<sup>[14,15]</sup> Sedative (*Musakkin*),<sup>[16]</sup> Deobstruent (*Mufattih*),<sup>[17,18]</sup> Anti-inflammatory (*Muḥallil*),<sup>[16,18]</sup> Analgesic (*Musakkin*),<sup>[16]</sup> Astringent (*Qabid*),<sup>[17]</sup> Rheumatism (*Hudār*),<sup>[19]</sup> Staphisagrine acts like curare and paralysis of the motor nerves, Stimulates heart, liver, brain (*Mubarrik-i-Dimāgh, Qalbwa Jigar*),<sup>[19]</sup> Atony (*Istirkhā'*),<sup>[17,18]</sup> therefore, it may be restricted the progression and delay the other distorting in the muscle and bone. The combination of medication given on the principle of treatment as mentioned along with local application with the consideration of their pharmacological in different neuromuscular disorders, and has nervine stimulant (*Mubarrik-i-A'sāb*), *Muḥallil-e-Waram* (Anti-inflammatory), and one of the important formulation *Habb-i-Ambar Momiya'* which is compound of such drugs have the action in strengthening the immune system and reduces stress and fatigue and is useful in the anxiety, depression, emotional trauma, insomnia and its give strength to nerve brain and heart muscle. In LGMD with other associated factors cardiomyopathy is very common.

The Unani polyherbal formulations *Habb-i-Sūranjān*, *Habb-i-Asgandh*, *Habb-i-Ambar Momiya'*, *Cap Dabidulward*, *Roghan-i-Mom*, *Roghan-i-Malkangini*, *Roghan-i-Bābūna*, *Qurs-i-Dhyābitus Khās*, *Chandraprabhavati* has analgesic, *Muḥallil-e-Waram* (anti-

inflammatory), *Muqanwi-i-Jigar* (hepatotonic), *Muqanwi-i-Mi'da* (Stomachic) properties and therapeutically used in *Fālij* and *Laqwa* (Paralysis).

***Habb-i-Ambar Momiya'***: The contents of this compound drug, basically *Muhammir*, rubefacient and *qābid*, there is increase the blood supply locally, thus causing increase nutrition in that area or a particular organ. It is also having *Muqanwi* and *Muḥarrik* effects on nerves.<sup>[20]</sup> The main constituent of this compound is *Ambar Ashhab* which has Anti-nociceptive effect.<sup>[21]</sup> It's other constituent is *Dārchinī* which helps to suppress the muscular flaccidity, thus it is useful in facial palsy & epilepsy.<sup>[20]</sup> Besides this another constituent *Jawzbuwa* has an important function which helps in maintaining/preserving natural heat (*Harārat-i-Gharizjyya*) and useful in the rheumatism, paralysis and facial palsy.<sup>[20]</sup> The drugs *Behman safed* and *Behman Surkeb* which having Adipogenous/Anabolic (*Musammin-i-badan/farba*) and *Mubarrik w aMuqanwi A'sāb* effect.<sup>[20]</sup>

*Ood Hindi*, *OodSalib*, *Khusya al-Thālab* and *Jadwār* has anti-inflammatory effects and is a *Muqanwi-i-A'sāb* (nervine tonic) /strengthening and helpful in *Ra'sha*, *Fālij* and *Laqwa*.<sup>[20]</sup>

***Habb-i-Sūranjān*** is a well-known polyherbal medicine which is used since ancient times to cure gout, rheumatism and other different Musculo-skeletal disorder. The main constituent of *habb-i-Sūranjān* is cochicine which has an effect on musculo-skeletal disorder. *Guggul* is useful in rheumatic disorders. *Zanjabil* is useful in hyperacidity which may be caused in some patients due to the use of *Sūranjān*. It is helpful in *Waja' al-Mafāsīl* (Arthralgia) and *Niqris*(gout).<sup>[22]</sup>

***Habb-i-Asgand*** is a *Muḥallil-i-Waram* (Anti-inflammatory) properties and useful in *Waja'*

*al-Mafāsīl* (Arthralgia) and *Waja' al-Warik* (Coxalgia).<sup>[22]</sup>

**Roghan-i-Bābūna** having *Musakkin-i-Alam* (Analgesic) and *Muḥallil-i-Waram* (Anti-inflammatory) properties, that helps in subsiding *Awaram* (Inflammation) and *Auja* (Pain).<sup>[23]</sup>

**Roghan-i- Malkanginī** is for External use having *Muqanwi-i-A'sāb* property. And used in *Fālij*, *Laqwa*, *Waja' al-Mafāsīl*, *Niqrīs*, *Khadar*, *Du'f-i-A'sāb*.<sup>[24]</sup>

**Roghan-i-Mom** is used externally having *Muḥarrik* and *Musakkin-i-Alam* (Analgesic) properties. It is also useful in the case of *Fālij*, *Laqwa* and *Waja' al-A'sāb*.<sup>[24]</sup>

### CONCLUSION:

Medicinal plants are being used since times immemorial, as a first line of treatment for various diseases. This case study demonstrates successful treatment of LGMD with the topical ointment without any adverse reactions. The results revealed that Unani medications are both safe and effective in the treatment of LGMD because of their potent anti-inflammatory, Nervine tonic, Analgesic and hepato-protective properties. Thus, it is postulated that regimental therapy, when combined with polyherbal formulations, have favourable effects in USM, as the case study illustrates. In the future, the management of all neuromuscular disorders, including LGMD, will conduct randomized experiments in a strictly scientific manner, taking into account the many regimental therapy modalities, based on the concepts of *Istirkhā'* and *Fālij*.

### Limitation of study:

Unani Drugs have promising, anti-inflammatory, antioxidant and hepatoprotective activities which are already proven in various scientific studies too.

However, the authors feel that a largescale study on these drugs involving larger population may be undertaken to substantiate the findings of this study.

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### Patient consent:

After properly explaining the procedure of the study to the patient, consent of the patient was taken on a written consent form.

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