

AYURVEDIC APPROACH IN THE MANAGEMENT OF GLYCOGEN STORAGE DISORDER- A CASE STUDY

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ABSTRACT

Glycogen storage diseases are inherited metabolic disorders of glycogen metabolism. It is an autosomal recessive disorder. Inborn errors of metabolism result from the lack of level of specific enzymes that are needed to insufficiently convert fat. Whenever glycogen is turned from glucose a different enzyme is needed in each step and when one of the enzymes becomes defective it fails to complete the step. These defects in the enzyme can lead to Glycogen storage disease. Type I GSD is a condition where there is a lack of glucose 6 phosphates enzyme it affects the liver, spleen, kidney, and small intestine. Common symptoms are hypoglycemia, lactic acidosis, and Hepatosplenomegaly. Treatment of type I GSD is symptomatic. The patient experienced significant relief from all the symptoms. i.e., Hepatosplenomegaly, Abdominal distension, loss of appetite, Generalised weakness. Her efforts were made to manage this condition by the Ayurvedic medicine administration. A female child aged 6 years from Alandi, Pune, Maharashtra, has a known case of Glycogen storage disorder in the last 4.5 years. Through her physical, clinical, and anthropometric examinations, she was diagnosed with Case

Glycogen storage disorder. The patient has complaints, of generalized weakness, distension of the abdomen, and anorexia. The patient was examined, there was a Hepatosplenomegaly and diagnosed as Yakritodara & Pleehodara & treated with Shamanoaushadi. Distension of

abdomen reduced; generalized weakness reduced with increased appetite. The patient gave a remarkable improvement in symptoms. In this case study, we are going to throw light on GSD and its treatment through Ayurvedic medicine.

KEYWORDS: - Glycogen storage disease, Yakritodara, Pleehodara, Metabolic disorder, GSD.

INTRODUCTION

Glycogen storage disease type I (GSD I), also known as Von Gierke disease, is an inherited disorder caused by deficiencies of specific enzymes in the glycogen metabolism pathway. comprises 2 major subtypes, GSD Ia and GSD Ib. In GSD Ia, there is a deficiency of the enzyme glucose-6-phosphatase (G6Pase) which cleaves glycogen to glucose thus leading to hypoglycemia and lactic acidosis. Patients with GSD 1b have normal G6Pase enzyme activity but have a deficiency of the transporter enzyme, glucose-6-phosphate translocase (G6PT). Patients present with manifestations of hypoglycemia and metabolic acidosis typically around one year of age. Genetic testing is the investigation of choice in patients suspected of having the disease to confirm the diagnosis. Dietary treatment prevents hypoglycemia and improves the life expectancy of patients. However, animal models of GSD I are being developed to study the disease more closely and develop new treatment strategies such as gene therapy to prevent long-term complications such as hepatic adenomas and renal failure.^[1]

Glycogen storage disease type I (GSD I) is an autosomal recessive disorder resulting from insufficient activity of glucose-6 phosphide an enzyme that Catalyses the hydrolysis of glucose and inorganic phosphate, a key. step in maintaining glucose homeostasis. There are two major GSD to I sub type: GD GSD type I a which is the result of a mutation affecting the G6Pase alpha catalytic subunit which is caused by translocase and GSD type I b by a defect in G6P.^[2]

The main targets for the management of GSD I are the prevention of acute metabolic derangement, prevention of acute and long-term complications, attainment of normal psychological development, and good quality of life.^[3]

AIM AND OBJECTIVE

To demonstrate the role of Ayurveda in the management of Glycogen storage disorder.

METHODS

The current article is about a Glycogen storage disorder case that was treated by shaman Aushadhi specifically Asav-Arishta and Churn Kalpana.

CASE REPORT

A 6-years-old female child resident of Pune, brought by her parents to the Out Patient Department of Kaumarbhritya, Dr D.Y. Patil College of Ayurveda and research center Pimpri, Pune with chief complaints of Abdominal distension, Generalized weakness, loss of appetite with a known case of Type I GSD in the past 4.5 years.

Associated symptoms (Study focus symptoms)

- Loss of appetite.
- Weight gain.
- Generalized weakness.
- Giddiness.

HISTORY OF PRESENT ILLNESS

The patient is a known case of Type I GSD. At the age of 1 year, her Parents had taken her for consultation in a private clinic nearby their home and started with allopathy medicines which were taken for 3 years. But they didn't get satisfactory relief to the patient, after taking medicines. After that, they brought the child for a better evaluation and management in Dr. D.Y. Patil College of Ayurveda and Research Centre, Pimpri Pune. After a detailed interrogation with the parents regarding the diet, lifestyle, and habits of the child and the history of her present illness and after a thorough evaluation regarding the present condition of the child. Patients had a complaint of Abdominal Distention, Loss of appetite, Generalised weakness, and Intermittent giddiness. Then she started shamanaushadhi medicines.

BIRTH HISTORY

A Full-term female child delivered through LSCS having birth weight 2.8 kg. Baby cried immediately after birth. NICU stay for 3 days in view of Neonatal Hypoglycaemia.

PAST HISTORY

Known case of Type I Glycogen Storage Disorder, diagnosed at the age of 1 year.

PERSONAL HISTORY: Vegetarian in diet.

FAMILY HISTORY

No any history of major illness in family.

ASHTAVIDH PARIKSHAN

1. Nadi: Kapha Pradhan Pitta nadi
2. Mala: Intermittent Hard Stool
3. Mutra: Samyak
4. Jivha: Niram
5. Shabda: Prakrut
6. Sparsha: Anushna
7. Druk: Prakrut
8. Akrti: Madhyam

EXAMINATION ASSESSMENT OF GENERAL CONDITION OF CHILD

Weight: 18.3 kg	Height : 118 cm
Heart Rate : 96/min	Trushna: Samyak
Respiratory Rate: 26/min	Nidra: Samyak
Temperature: 98.2 F	Kshudha: Kshudha Mandya
Abdominal Girth: 71 cm	

PER ABDOMINAL EXAMINATION

Inspection: Abdominal Distension++

Palpation: Hepatosplenomegaly ++

SYSTEMIC EXAMINATION

RS: AEBE clear

CVS: S1S2 normal

CNS: Conscious and Oriented

TREATMENT GIVEN: 3 MONTHS TREATMENT**1. Primary**

- a) Kamdudha rasa 250 mg BD
- b) Sutsekhar rasa 250 ml BD with Dugdha(Milk)
- c) Gandharv haritaki 125 mg HS
- d) Praval panchamrut 125 mg TDS
- e) Arvindasav 5 ml BD with 1 cup of Koshna Jala
- f) Syrup Amlicure DS 5ml BD

2. 1st follow up

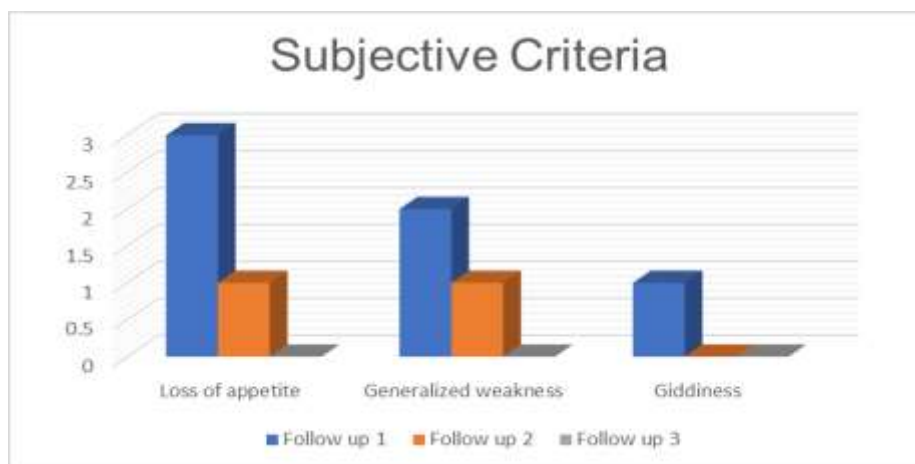
- a) Kamdudha rasa 250 mg BD
- b) Sutsekhar rasa 250 ml BD with dugdha
- c) Gandharv haritaki 125 mg HS
- d) Praval panchamrut 125 mg TDS
- e) Arvindasav 5 ml BD with 1 cup of Koshna Jala
- f) Syrup Amlicure DS 5ml BD

3. 2nd follow up

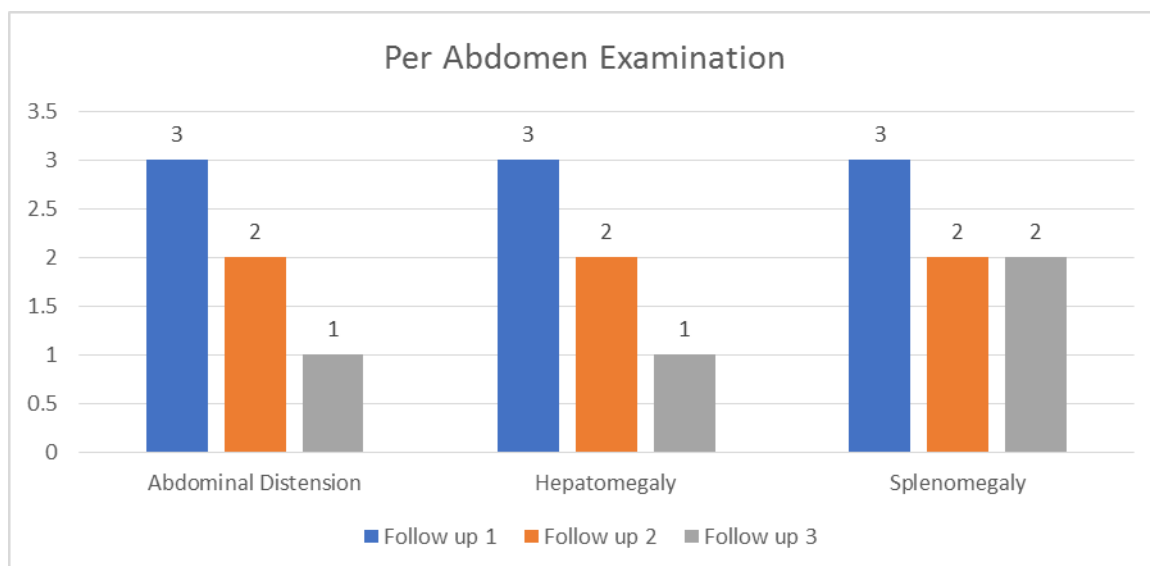
- 1) Krumimudga Rasa 250mg 1 BD
- 2) Laghusutashekhar Rasa 250mg 1BD
- 3) Rohitakarishtha 5 ml BD with 1 cup of Koshna jala
- 4) Arvindasav 5 ml BD with 1 cup of Koshna Jala
- 5) Syrup Amlicure DS 5ml BD
- 6) Kwath BD of the following drugs; Each 125 mg Churna of the following drugs
Manjishtha, Lodhra, Gokshur, Guduchi, Sariva, Vidang, Aamalaki, Kutaki.

OBSERVATION

Sr. no.	Criteria	Follow up 1	Follow up 2	Follow up 3
1.	Loss of appetite	+++	+	-
2.	Generalized weakness	++	+	-
3.	Giddiness	+	-	-
4.	Abdominal Distension	++	+	+
5.	Hepatomegaly	+++	++	+
6.	Splenomegaly	+++	+++	++



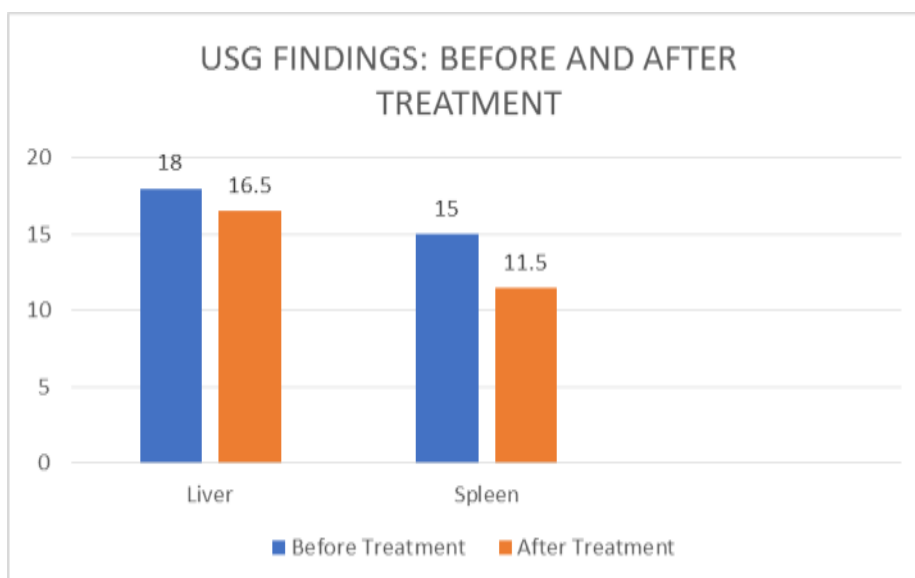
Graph 1: The graphical representation of Subjective criteria (in gradings) on 0th, 30th, & 60th day.



Graph 2: The graphical representation of per abdomen examination (in gradings) on 0th, 30th, & 60th day.

BEFORE AND AFTER TREATMENT

USG Abdomen and Pelvis		Before	After 3 months
1.	Liver	18 cm	16.5 cm
2.	Spleen	15 cm	11.5 cm
3.	Enteritis	Positive	Negative



Graph 3: The graphical representation of USG FINDINGS: BEFORE AND AFTER TREATMENT (in cm) on 0th, 30th, & 60th day.

DISCUSSION

In Ayurvedic texts, so many herbs, mineral drugs and preparations (compound drugs) are available for the successful treatment and prevention of complications in the case of GSD and other conditions. The Ayurvedic system of medicine plays a vital role in the treatment of liver and spleen disorders by providing effective treatments. The basic need while treating these patients is to evaluate the patient on the parameters of Ayurvedic Rogi pariksha to diagnose the disease. Precipitating factors for hepatomegaly such as evaluation of liver function, avoiding factors, diet regimen, and care for bladder and bowel has to be evaluated and maintained, or else patients might get.^[4]

These Ayurvedic drugs can be used in different forms as *kwath-Manjishtha*, *Lodhra*, *Gokshur*, *Guduchi*, *Sariva*, *Vidang*, *Aamalaki*, *Kutaki*, Most of the drugs are *Tikta*, *Kashayain Rasa*, *Madhura in Vipaka* and has following properties i.e., *Pittahara*, *Pittarrechak*, *Yakriduttejak*, *Deepan*, *Rechan*, *Pachaka*, *Shothhara*, *Jwarahara*, *Panduhara*, *Yakrit* and *Raktvikarhara*, *Tridoshara*, *Rasayana*, *Mutrajanana*, *Pittasaraka*, *Anulomaka*, *Dahaprashamana*, and *Raktapittahara*. *Krumimudga Rasa*, *Laghusutashekhar Rasa*, *Rohitakarishtha*, and *Arvindasav* are herbs-mineral compounds used in the treatment of splenomegaly, hepatomegaly, and inflammatory conditions.^[5,6,7]

CONCLUSION

Ayurvedic herbal preparations have many advantages an effective 3 months of Ayurveda treatment gives shortening of disease period, early regeneration of parenchymal cells, avoidance of post-GSD symptoms and complications such as hepatic encephalopathy, splenomegaly also Distension of the abdomen reduced, generalized weakness reduced with increased appetite. Many research studies have been carried out and are being conducted globally on different parameters of liver disease for the well-being of mankind.

SUMMARY

A 6 years old female child with a known case of Type 1 Glycogen Storage Disorder was treated with Shamanaushadhi for 3 months. The child got relief from symptoms.

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