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AYURVEDIC MANAGEMENT OF HEREDITARY SPHEROCYTOSIS: A CASE REPORT

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ABSTRACT

Hereditary Spherocytosis is an inherited abnormality of the red blood cell caused by defects in structural membrane protein clinically characterized by anemia, Jaundice & Spleenomegaly. The treatment of Hereditary Spherocytosis is splenectomy. Although the red cell defect persists the breakup of the red cells (hemolysis) ceases. Persons with Hereditary Spherocytosis should take supplemental folic acid. In *Susruta Samhita* following clinical condition can be correlated with Hereditary Spherocytosis i.e. *Paittija Pandu*, *Kostha-Shakhaashrita Kamla* and Plihodar. In the light of management of these disease explained in Ayurvedic texts we can provide satisfactory Ayurvedic management of Hereditary Spherocytosis. In the present case study a confirmed diagnosed patient with Hereditary Spherocytosis was treated with *Samshaman Chikitsa* for two months.

Keywords: Hereditary Spherocytosis, Chausath Prahari Pippli, Rohitakarist, Tapyadi Lauha.

INTRODUCTION

"Hereditary Spherocytosis is a genetic disorder of the red blood cell membrane clinically characterized by anemia, jaundice and spleenomegaly." In Hereditary Spherocytosis the red cells are smaller, rounder and more fragile than normal. The red cells have a spherical rather than the biconcave disk shape of the normal red cell. These rotund red cells (spherocytes) are osmotically fragile and less flexible than normal red cells and tend to get trapped in narrow blood passages, particularly in the spleen and they break up (hemolyze) leading to hemolytic anemia. The clogging of the spleen with red cells almost invariably causes spleenomegaly. The breakup of the red cells releases hemoglobin and the heme part gives rise to bilirubin leads to the formation of gallstones, even in childhood. There is also often iron overload due to the excess destruction of iron-rich red cells. (https://www.google.co.in/search?q=hereditary+spherocytosis&oq=heridito&aqs=chrome.5.69i57j0l5.7962j0j8&sourceid=chrome&ie=UTF-8#).

Hereditary Spherocytosis is the most common disorder of the red cell membrane and affects 1 in 2000 people of Northern European ancestry. It often shows up in infancy or early childhood causing

anemia and jaundice. The bone marrow has to work extra hard to make more red cells. So if in the course of an ordinary viral illness, the bone marrow stops making red cells, the anemia can quickly become profound. This is termed an aplastic crisis.

Hereditary Spherocytosis is due to a deficiency of a protein called spectrin and anakyrin. Anakyrins are cell membrane proteins. Hereditary Spherocytosis is inherited as a dominant trait so if a person with Hereditary Spherocytosis reproduces; their child has a 50% chance to have Hereditary Spherocytosis. Laboratrical studies show evidence not only of many spherocytes but also increased numbers of reticulocytes, hyperbilirubiniemia and increased osmotic fragility of the red cells.

The treatment of Hereditary Spherocytosis is splenectomy. Although the red cell defect persist the breakup of the red cells (hemolysis) ceases. Persons with Hereditary Spherocytosis should take supplemental folic acid.

According to Ayurveda following clinical condition can be correlated with Hereditary Spherocytosis *Paittika Pandu*, *Kosthashakhaashrita Kamla*, *Plihodar*. Hereditary Spherocytosis seems to be a state of *Vayadhi sankarya*.

CASE REPORT:

In this present study a male patient of age 21 years old, Hindu, unmarried, vegetarian, nondiabetic & normotensive, known case of Hereditary Spherocytosis diagnosed in 2003 in PGI Chandigarh at the age of 6 years, with no history of injury/surgery/allergy/joint disorder, registered in OPD No. K-II-5719/123723 dated 14.09.2017 in Dept. of Kayachikitsa in S.K.D. Govt., Ay. College & Hospital, Rampur, Muzaffarnagar. Patient attended OPD with complaints of generalized weakness, fatigue, fullness of abdomen and occasional pain in abdomen.

MATARIAL AND METHODS:

PHYSICAL EXAMINATION:

Height	-	167 cm.	Cynosis	-	Absent
Weight	-	62 kg.	Clubbing	-	Absent
B.P.	-	130/78 mm of Hg	Oedema	-	Absent
Temp.	-	98.7^0 F	JVP	-	Normal insignificant
P.R.	-	76/mt.	Lymphedenopathy	-	Absent
R.R.	-	17/mt.	Hair	-	Normal
Pallor	-	Present (++)	Nail & Skin	-	Mild pale in color
Icterus	-	Present (+)			

SYSTEMIC EXAMINATION:

GIT: Shape of abdomen- Mild distended, Umblical-normal inverted no prominent vein/scar mark / visible pulsation. **Nontender palpable spleen upto 4 fingers,** no hepatomegaly, no tenderness, no ascitis, **dullness on percusion over left hypochondric region**, no shifting dullness/fluid thrill, bowel sounds are normal.

RS: B/L Lung clears no wheezing / crepitation, normal vasicular bredth sound with B/L symmetrical chest movement.

CVS: S_1 & S_2 wnl, no added sound.

CNS, LS & Genitourinary: Showed no abnormality.

AYURVEDIC EXAMINATION: Ayurvedic examination done on the parameter of Dashavidha and Astavidha Pariksha as shown in table -1.

MANAGEMENT:

(A) Aims of treatment:

1. To increase & maintenance of normal Hb level.
2. To decrease heamolysis and belirubin level.
3. To revert the spleenomegaly.

4. To improve physical well being & reliving associated symptoms.
5. To enhance normal reticulocyte formation.

(B) Medicinal treatment:

1. *Chausath Prahari Pippli* 500 mg., *Mandur Bhasma* 500 mg., *Shudha Gairika* 500 mg., *Aamalki Churn* 3 gm. all mixed well and taken with honey twice in a day.
2. *Arogyavardhani vati* 500 BD with lukewarm water.
3. *Tapyadi Loh* 250 mg. BD with lukewarm water.
4. *Rohitakarisht* 20 ml BD after meal.
5. *Goksharan Arka* 20ml BD with lukewarm water after meal.
6. *Panchtikta ghrita* 10 gm. OD as *Samshaman ghrita.*

(C) **Educate patient about the nature and course of disease**

(D) **Diet**: Use healthy nutritious and rich in iron as green leafy vegetables, fruits & pulses and avoid Cold drink, fast food, refined & packed food and heavy exercise to avoid trauma to spleen.

OBSERVATION AND RESULTS:

With the help of above regimen in the form of *sanshaman chikitsa* and dietics, important positive results were noticed in laboratorial parameters and symptoms after 2 month of treatment.

Table - 2: Showing symptomatological analysis.

Table - 3: Showing laboratorial analysis.

Table - 4: USG whole abdomen.

DISCUSSION

Hereditary Spherocytosis is an inherited disease. The nature of disease is incurable despite the huge advancement in modern medical science. As Hereditary Spherocytosis is a *Kulaja Vikara* (in hereted disease) so *Raktagni* and *Ranjakagni* Mandhyakar Hetu was in born due to *Rakta Dhatu Beej Bhaga Avyava Dusti.* (Molecular defect in genes that code for the RBC membrane proteins spectrin, anakyrin and other proteins). The disease is *Pitta* predominant & *Rakta* being the main *Dushya*. Due to inadequate action of *Ranjakagni* and *Raktagni* the *Rakta Sadharmiansh* in *Poshak Rasa* is not converting properly in *Poshya Rakta Dhatu* leading to *Rakta Kshya.* Improper *Rakta Dhatu* formation and *Alparaktata* also in part responsible for next *Dhatu Poshan Alpta and Kshya* i.e. *Meda Alpta* or *Kshya, thats* why *Sarakta Medalpta* leads to *Pliha Vridhi* (Su.Su. 15/13) in Hereditary Spherocytosis.

Above *Samprapti Vightana* achieved with *Pittashamaka, Raktavardhaka* and *Raktavaha Srotomula Shodhaka Dravya*. *Sarakta Meda* and *Majja Vridhi* with help of *Panch Tikta Ghrita* and *Vyadhi Vipreet Chikitsa* by *Rohitakarisht* and *Tapyadi lauha* help in reducing *Plihavridhi*. Because the disease is a *Kulaja Vikar* by its origin so it is not curable but can be managed effectively with classical Ayurvedic regimen.

CONCLUSION

So in Hereditary Spherocytosis Ayurveda may provide an effective alternate to impending compulsion of spleenectomy. Finally it can be concluded that such types of small study in a rare disorder can provide more option for the further work in this field for the new scholars at different higher institutions, where number of such type of cases is more.

Limitation of this study since it's a single case study hence we cannot reach on the final result on the statistically criteria. Advantage of these types of studies having both texts & personally experience based provides more options for comparative study on this chronic disease. Due to *Pitta dosha* & *Rakta dyshya* predominance disease *Virechan & Jalaukavacharana* in the form of *Samshodha*n therapy prior to start *Samshamana* therapy may give better result before starting *samshaman* regimen. This study may be applicable for further studies on larger no. of patients for the successful management of this chronic disease.

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Table - 1

Dashvidha Pariksha		***Ashtavidha Priksha***	
Prakriti	*Kapha Pittaja*	*Naadi*	*Kaphaja, 76/mt*
Vikriti	*Vikriti Visham Samveta*	*Mala*	*Shwet-peeta, 1-2 / day*
Saar	*Madhyam*	*Mutra*	*6-7/day, Peetabh*
Sanghanana	*Madhyam*	*Jivha*	*Aardra, Shweta peeta*
Praman	*Madhyam*	*Shabda*	*Prakrita Avaikarik*
Satmya	*Madhyam*	*Sparsha*	*Ishat Ushna, Mridu*
Satwa	*Madhyam*	*Drika*	*Shweta peeta*
Vyayam	*Avar*	*Aakriti*	*Madhyama Kaya*
Yaya	*Madhyam*		

Table - 2

S. No.	Symptoms	Before Treatment	After 20 days	After 40 days	After 60 days with treatment
1	Gen. weakness	++	++	+	–
2	Fatigue	++	++	+	–
3	Fullness of abdomen	++	++	++	+
4	Occasional pain abdomen	+	+	+	+

Table -3

S. No.	**Symptoms**	**Before Treatment**	**After 60 days with treatment**
1	Hb %	6.5 gm	11.4 gm.
2	ESR	86 mm/hr	30
3	Platelet count	1,04,000	2,10,000
4	S. Bilirubin (T)	5.84	3.3
	Indirect Bilirubin	5.35	2.9
	Direct Bilirubin	0.49	0.4

Table -4

Before Treatment	**After 60 days with treatment**
Spleen is increased in size and echotexture size 18.1 cm in vertical section	Spleen is increased in size with normal echotexture size 16 mm in vertical section.

Source of Support: Nil
Conflict Of Interest: None Declared

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