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A SINGLE CASE STUDY ON HALLERVORDEN-SPATZ DISEASE W.S.R. *KAMPAVATA* – AN INTERVENTIONAL CLINICAL STUDY

Patil. Deepa C[1], Rao Veena G [2] , S.Rashmi [3]

[1]Reader ,Department of KayaChikitsa, JSS Ayurveda medical college, Mysuru, India570028.

[2]Professor,Department of Panchakarma, JSS Ayurveda medical college, Mysuru, India570028

[3]PG scholar, Department of Panchakarma, JSS Ayurveda medical college, Mysuru, India570028.

ABSTRACT

Pantothenate kinase - associated neurodegeneration (PKAN) , formerly called Hallervorden – Spatz Disease (HSD) , is a rare disorder , usually begins in childhood and is characterized by progressive dystonia , rigidity , tremor , and choreoathetosis . Extensor plantar reponses , dysarthria , and intellectual deterioration become evident during adolescense .Based on symptoms this disease is correlated to *Kampavata* in *ayurveda* . *Acharya Basavarajiyam* has given a clear description by explaining the clinical pictures as *karapadatala kampa* (tremor, dystonia , choreoathetosis), *deha bhramana* (postural instability) , *matiksheena* (dementia) and *nidrabhanga* (disturbed sleep). A case report of 28 year old male presented with *kampa* (tremor, cervical dystonia , choreoathetosis), intellectual deterioration, *stambha*(spasticity in both upper and lower limb),*gamanekashtatha*(difficulty in walking and sitting without support) , *vakkruchrata*(dysarthria), drooling of saliva and was treated in lines of *ayurveda* in 2 sittings for a period of 20 days each with a gap of one month and the results were encouraging .

Key words: Hallervorden – Spatz Disease(HSD), *Kampavata* , *Vyatyasat Chikitsa* , *Kaphavruta Vata* , PKAN (Pantothenate kinase - associated neurodegeneration).

INTRODUCTION: Hallervorden-Spatz Disease (HSD), is a rare disorder characterized by progressive extrapyramidal dysfunction and dementia. The disease was first described in 1922 by two German physicians, Hallervorden and Spatz, as a form of familial brain degeneration characterized by cerebral iron deposition. The term neurodegeneration with brain iron accumulation type 1 (NBIA-1), eventually came to be used for this condition[1], Although the most recent term for the disorder is pantothenate kinase-associated neurodegeneration[2].

The exact etiology of PKAN is not well understood. One of the proposed hypothesis is that aberrant oxidation of lipofuscin to neuromelanin and insufficient cystine dioxygenase leading to abnormal iron accumulation in the brain. While portions of the globus pallidus and pars reticularis of substantia nigra have relative higher iron content in healthy individuals, individuals with PKAN have excessive amounts of iron accumulated in these nuclei[3].

HSD is relentlessly progressive. It is characterized by most common symptoms like corticospinal signs (e.g., spasticity, hyper-reflexia , Extensor plantar responses), and extrapyramidal signs, including spasticity, dystonia, and choreoathetosis, distorting muscle

contractions occurring in the face, trunk, and limbs; involuntary muscle contractions that cause abnormal posture and uncoordinated movements (Ataxia) and also includes other clinical features like Confusion , disorientation , seizures ,progressive dementia, weakness , drooling of saliva , dysphagia and unclear speech. The course of the disease usually proceeds over 10-12 years and affected individuals typically die in the second or third decade, but case reports describe patients surviving up to 30 years[4,5].

Prevalence data regarding this disorder remains incomplete, however it is estimated that anywhere between 1 in 1,000,000 to 3 in 1,000,000 individuals will be afflicted with this disorder (based upon observed cases in a population), but once again this is only an estimate as the disease is so rare and is difficult to statistically and accurately ascertain[6]. The classic presentation is in the late part of the first decade or the early part of the second decade, between ages 7 and 15 years. However, the disease onset has been reported in all age groups including infancy and adulthood[3].

Magnetic resonance imaging (MRI) has increased the likelihood of early diagnosis of Pantothenate kinase-associated neurodegeneration (PKAN)[7,8,9]. The typical MRI findings include bilaterally symmetrical, hyperintense signal changes in the anterior medial globus pallidus, with surrounding hypointensity in the globus pallidus, on T2-weighted images. These imaging features, which are fairly diagnostic of PKAN, have been termed the **"eye-of-the-tiger sign."** The hyperintensity represents pathologic changes, including gliosis, demyelination, neuronal loss, and axonal swelling. The surrounding hypointensity is due to loss of signal secondary to iron deposition[10].

According to *ayurveda* the disease HSD is compared with Kampavata based on the similarities in some of the symptoms like *kampa* , *stambha* , *dehabramana* , *matiksheena*[11] with Tremor , choreoathetosis , dystonia rigidity , postural instability , dementia . Increased *chala guna* of *vata* produces *kampa*[12] , dystonia , choreoathetosis , whereas increased *manda guna* of *kapha* in turn decreases *vata* and there producing *stambha*[13] (Rigidity / Spasticity) . Increased *vata* at one site and decreased at other site explains the process of *avarana* . Here *Kaphavrutavata* needs *Vyatyasat chikitsa*[14] in the form of *snigdha* and *ruksha* line of treatments alternatively . As it is a *vatananatmaja vyadhi anulomana* , *basti*[15] and *shamanoushadhi* are also been described in classics.

Herein details of a HSD patient, effectively intervened with *ayurvedic* treatment modalities, have been described. A substantial improvement was observed in some of the symptoms like cervical dystonia , tremor , choreoathetosis , spasticity , mental status and muscle power after a period of 20 days of treatment in first sitting and the condition was maintained without aggravation of symptoms after 2nd sitting of treatment .

CASE REPORT :

A 28-year-old male patient , pre diagnosed as a HSD on MRI report , was admitted in J.S.S.A.M.C & H , Mysuru , Karnataka .As per the history given by the patient mother, he was apparently healthy till his 12 yrs of age, later had a fall from height

16yrs back , had a injury to right middle finger and to right elbow and after few days he developed difficulty to write and to do his daily activities alone . Later there was sankocha (flexion) of wrist joint and elbow joint of right side followed by that of left side . Gradually he developed loss of muscle power in both upper and lower limbs and involuntary movements like *kampa* i.e. tremor , choreoathetosis , then he developed *gamanekashtata* (diificulty in walking and sitting without support) . Patient also had complaints of drooling of saliva since 5 months, *vakkruchrata (dysarthria)* since 6 months and flexion of the neck towards left side with cervical dystonia since 3 yrs .Patient was not a known case of Hypertension / Diabetis Mellitus / Bronchial Asthma / Epilepsy . Patient was on medication like muscle relaxant , anticonvulsant and anticholinergic drugs .

TABLE 1 : SHOWING THE CLINICAL FINDINGS

CLINICAL FINDINGS	
Subjective Findings	- Dysphagia. - Drolling of saliva . - Painful muscle spasm.
Objective Findings	- Physical disposition - Decorticated both UL and foot inversion - Mental disposition - Mentally retarded. - Cervical dystonia. - Choreoathetosis. - Involuntary muscle contractions - Present - Spasticity in both upper & lower limbs. - Tremor in both upper & lower limbs. - Deep tendon reflexes – Exaggerated in both upper limbs and lower limbs. - Babinski sign – Positive . - Muscle power - 4/5 in both upper & lower limbs. - **MRI Brain** - Bilateral Globus pallidus Hypointense signals with central hyperintense signals s/o Hallerverodan - spatz disease .

FIGURE 1 : MRI REPORT SHOWING THE " EYE OF A TIGER SIGN "

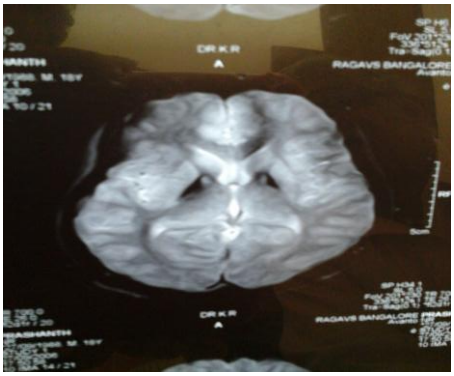

TABLE 2 : SHOWING THE TREATMENT INTERVENTION IN 1ST SITTING

INTERVENTION IN 1ST SITTING (06/06/2016 – 26/06/2016)	**DURATION OF TREATMENT**	**DESCRIPTION**
• *Sarvanga shashtika shali pinda sweda* with *maha masha taila*[16] *abhyanga.* • *Sarvanga udwarthana* with *kolakulathadi churna*[17].	On alternative days for 20 days	*Snigdha* & *Ruksha chikitsa* alternatively is *avarana vatahara* .
• *Nasya* with *yashtimadhu taila*[18] .	8 drops to each nostril - for 8 days	*Brihmana* & *srotoshodhana*
• *Shiro pichu* with K*sheera bala taila*[19] & *bramhi ghritha*[20].	For 20 days.	*Vatashamana* & *medhya*
• Physiotherapy[21] - Gait training - Bridging - Strengthening exercises for lower limb - Stretching for upper limb.	For 10 days	Improves muscle power

TABLE 3 : SHOWING THE TREATMENT INTERVENTION IN 2ND SITTING

INTERVENTION IN 2ND SITTING (28/07/2016 –11/08/2016)	**DURATION OF TREATMENT**	**DESCRIPTION**
• *Sarvanga shashtika shali pinda sweda* with *maha masha taila*[16] *abhyanga.* • *Sarvanga udwarthana* with *kolakulathadi churna*[17] .	On alternative days for 15 days	*Snigdha* & *ruksha chikitsa* alternatively is *avarana vatahara*
• *Yoga basti* *a) Anuvasana basti* *Brihat saindhavadi taila*[22,23] – 60 ml. *b) Niruha basti* - *Madhu* – 70ml - *Saindhava lavana* – 6 gms - *Brihat saindhavadi taila*[22,23] – 70 ml + *Yashtimadhu taila*[18] – 70ml. - *Shatapushpa kalka* – 20gms. - *Dashamula + Balamula + Erandamula + Amrita – Kashaya* – 350ml. - *Gomutra* – 50ml	For 8 days	*Vatahara* & *srotoshodhana* .
• *Ksheera dhuma*	For 10 days	*Kaphahara* and

		avaranahara.
• Physiotherapy[21] - Gait training - Bridging - Strengthening exercises for lower limb - Stretching for upper limb	For 10 days	Improves muscle power

TABLE 4 : SHOWING THE ORAL MEDICATION IN 1ST AND 2ND SITTING

SL NO	ORAL MEDICATION IN 1ST AND 2ND SITTING	DURATION	DESCRIPTION
1.	*Gandharva hastadi eranda taila* - 10ml + *Shunti kashaya*[24] -10ml.	Before food in morning time for20 days	*Anulomana* & *Vatakaphahara*
2.	*Swalpa rasona pinda kalka*[25] - 10 gms + *Erandamula kwatha* - 30ml	Before food in morning time for 10 days	*Amapachana* , *srotoshodhana* & *rasayana* .
3.	Tab .*Vishatinduka vati*[26] - 2-0-2	After food	*Amapachana* & *srotoshodhana*
4.	Tab.*Kampavatari rasa*[27] - 1-0-0	After food	*Vyadhihara* .
5.	Tab.*Shiva gutika*[28,29] : 1-0-1	After food	*Avarana vata rasayana.*

TABLE 5 : SHOWING THE OBSERVATION AND RESULTS OF THE STUDY BY COMPARING BT AND AT

PARAMETERS	BT	AFTER 1ST SITTING	AFTER 2ND SITTING
Cervical Dystonia	++	+	+
Choreoathetosis	++	+	+
Spasticity	++	+	+
Muscle power	4/5 in both upper limb and lower limb	4+/5 in both upper limb and lower limb	4+/5 in both upper limb and lower limb
Tremor	+	+	+
Dysarthria	++	++	+
Drooling of saliva	++	++	+
Babinski sign	Positive	Positive	Positive
Deep tendon reflexes	Exaggerated	Exaggerated	Exaggerated
Involuntary muscle contractions – Decorticate limbs	+++	++	++
Gait	Pt was not able	Pt was able to sit	Pt was able to sit

	sit without support . Pt was able to walk with maximum assistance .	without support . Pt was able to walk with minimal assistance	without support . Pt was able to walk with minimal assistance .
Mental status	Mentally retarded – Was not responding to Verbal stimulus – not oriented	Was responding to Verbal stimulus – Oriented	Was responding to Verbal stimulus – Oriented .

DISCUSSION :HSD is a rare, inherited , neurodegenerative disorder characterized by iron deposition in brain , can be compared with the *Kampavata* in *Ayurveda* where most of the *lakshanas* of *Kampavata* like *karapadatala kampa* , *stambha* , *dehabramana* , *matiksheena* etc are similar with symptom's of HSD i.e.tremor , rigidity , dystonia , choreoathetosis ,postural instability due to decorticate limbs , dementia etc and also the main pathology in both, lies in *Masthiska* (Brain) .

Vata is one among the *pradhana dosha* among *tridosha*[30], as it is considered to be a *'Prana'* and is responsible for the vital functions in the *Shareera* . *Kampavata* is one among the most disabling *vata vyadhi*[31] . This disease is being an challenge to the medical science since time immemorial because of its crippling nature and non availability of curative treatment . The specific *nidana* and *samprapti* for *kampavata* is not been mentioned in any of the classics, but based on lakshanas like *kampa* , *stambha* , *gatisanga* , *cheshtasanga* etc, *kaphavruta vata samprapti* can be derived and *avarana chikitsa*[32] line of treatment can be followed.

Patient was treated initially with *Gandharva hastadi eranda taila* and *shunti kashaya* orally for 20 days which is one of the best *vatanulomana* & *vatashamana dravya* , helpful in *stambhanigrahana* (spasticity / Rigidity) and controling *dehabramana* i.e Postural instability due to decorticate limbs . *Kampavata* is a *kaphavruta vata vyadhi* which requires *vyatyasat chikitsa* alternatively in the form of *snigdha* and *ruksha* . Hence *Shashtika Shali pinda sweda* and *udwartana* was done on alternative days which helped in reducing *kampa* & *stambha* ; later was treated with *nasya* for 8 days , which gave an improvement in cervical dystonia . *Ksheera dhuma* which is having *kaphahara* & *srotoshodhana karma* in *mukha* helped in reducing drooling of saliva and mild improvement was seen in the speech of the patient .

Later *yoga basti* was given which was *srotoshodhana* & *vatahara*, which helped in improving the overall well being of the patient. Along with *Ayurvedic* treatment , Physiotherapy was also done which helped in improving the muscle power and gait of the patient .

As *lashuna* and *shilajitu* are choice of drug for *kaphavrita vata* , it was given in the form of *swalpa rasona pinda* and *shivagutika* . *vishatinduka vati* is

stambhahara and *kampavatari rasa* here is , *vyadhihara* .

Overall *shodhana* and *shamana* treatment were helpful in reducing the clinical features evidently in this particular patient

CONCLUSION :HSD can be correlated to *Kampavata* based on the symptoms which has been explained clearly by the *acharya basavarajiyam* . Deriving *kaphavrita vata samprapti* and hence following the *vyatyasat chikitsa* in the form of *snigdha* and *ruksha chikitsa* will be helpful . Treatment has given moderately evident improvement in some of the symptoms like cervical dystonia , tremor , choreoathetosis , spasticity , mental status & muscle power. This set of *shodhana* , *shamana* and Physiotherapy helped in clinically appreciable reduction in the symptoms .

REFERENCES:

1. Neumann M, Adler S, Schlüter O, Kremmer E, Benecke R, Kretzschmar HA. Alpha-synuclein accumulation in a case of neurodegeneration with brain iron accumulation type 1 (NBIA-1, formerly Hallervorden-Spatz syndrome) with widespread cortical and brainstem-type Lewy bodies. Acta Neuropathol. 2000;100:568–74. [PubMed]

2. Schneider SA, Hardy J, Bhatia KP. Iron accumulation in syndromes of neurodegeneration with brain iron accumulation 1 and 2: Causative or consequential? J Neurol Neurosurg Psychiatry. 2009;80:589–90. [PubMed]

3. *https://www.ncbi.nlm.nih.gov/books/NBK430689*

4. Saito Y, Kawai M, Inoue K, Sasaki R, Arai H, Nanba E, et al. Widespread expression of alpha-synuclein and tau immunoreactivity in Hallervorden-Spatz syndrome with protracted clinical course. J Neurol Sci. 2000;177:48–59. [PubMed]

5. Hickman SJ, Ward NS, Surtees RA, Stevens JM, Farmer SF. How broad is the phenotype of Hallervorden-Spatz disease? Acta Neurol Scand. 2001;103:201–3. [PubMed]

6. *https://en.wikipedia.org/wiki/Pantothenate_kinase-associated_neurodegeneration*

7. Feliciani M, Curatolo P. Early clinical and imaging (high-field MRI) diagnosis of Hallervorden-Spatz disease. Neuroradiology. 1994;36:247–8. [PubMed]

8. Shah J, Patkar D, Patankar T, Krishnan A, Prasad S, Limdi J. Hallervorden Spatz disease: MR imaging. J Postgrad Med. 1999;45:114–7. [PubMed]

9. 25. McNeill A, Birchall D, Hayflick SJ, Gregory A, Schenk JF, Zimmerman EA, et al. T2* and FSE MRI distinguishes four subtypes of neurodegeneration with brain iron accumulation. Neurology. 2008;70:1614–9. [PMC free article] [PubMed]

10. https://emedicine.medscape.com/article/1150519-overview *Dec 07 2016*

11. Basavaraj, Basavarajeeyam, Choukhamba sankrit pratishtana,Delhi,edition 2005, chapter 6; pp101.

12. T.Sreekumar; Translation of Astanga Hridaya, Sutrasthana; pub: Harisree hospital ,Trissur,Kerala; edition 2007; chapter 11/5-6; pp-290.

13. T.Sreekumar; Translation of Astanga Hridaya, Sutrasthana; pub: Harisree hospital ,Trissur,Kerala; edition 2007; chapter 12/52-53; pp-327.

14. Dash Vaidya Bhagwan, Sharma Ram Karan. Charaka Samhita Chikitsa sthana , Chakrapanidatta's Ayurveda Dipika Commentary with English

translation. Choukhamba Sanskrit Series Office , Varanasi; reprint 2008; chapter 28/184; pp – 69.

15. Dash Vaidya Bhagwan, Sharma Ram Karan. Charaka Samhita Chikitsa Sthana , Chakrapanidatta's Ayurveda Dipika Commentary with English translation. Choukhamba Sanskrit Series Office , Varanasi; Reprint edition -2008; chapter 28/187; pp – 69.

16. Sharma P.V; edited Chakradatta; Chaukambha publishers Varanasi, edition 1998; chapter 22/165-171; pp-201.

17. Dash Vaidya Bhagwan, Sharma Ram Karan. Charaka Samhita Sutra Sthana , Chakrapanidatta's Ayurveda Dipika Commentary with English translation. Choukhamba Sanskrit Series Office , Varanasi; reprint 2008; chapter 3/18; pp – 78.

18. Mishra Bramhashankar; Translation of Bhasijya Ratnavali; Chaukambha publications, New Delhi; edition 2005; Vol 3 , chapter 60/146; pp-194-195.

19. Sharma Ramanivas and Sharma Surendra , SahasraYoga ; Choukhamba sanskrit pratishtana, Delhi, ;edition 2009; pp- 75.

20. Sharma Ramanivas and Sharma Surendra , SahasraYoga ; Choukhamba sanskrit pratishtana, Delhi, ;edition 2009; pp- 304.

21. Porter stuart ; Tidy's physiotherapy ;Published by Elsevier , a division of Reed Elsevier India private limited, New Delhi, 14th edition , reprinted 2011 ; chapter 17 ; pp -419-420 , 423-424.

22. Translation of Bhasijya Ratnavali; Chaukambha publications, New Delhi; edition 2005; chapter 29/222-227; pp-321.

23. Sharma P.V; edited Chakradatta; Chaukambha publishers Varanasi, edition 1998; chapter 25/48-51; pp-.232.

24. Sharma P.V; edited Chakradatta; Chaukambha publishers Varanasi, edition 1998; chapter 22/58; pp-190.

25. Sharma P.V; edited Chakradatta; Chaukambha publishers Varanasi, edition 1998; chapter 22/35; pp-187.

26. Kaleda Krishnagopala , Sidda prayoga samgraha , Prathama khanda ;Krishna gopala ayurveda bhavana , edition 2003; pp-647.

27. Pathak Shrikrishna Lalagaj Dattaram; Ras Raj Sundar, Uttarakhand (uttarabhag), Kamala prakash, Mathura, edition 1888, Vatavyadhi chikitsa adhyaya, pp-549-550.

28. Sharma P.V; edited Chakradatta; Chaukambha publishers Varanasi, edition 1998; chapter 65/172-193; pp-592-593.

29. Murthy K.R.S; Translation of Ashtanga Samgraha Uttara sthana; Chaukhambha orientalia Varanasi; reprint 2012; chapter 49/309-325; pp-506-507.

30. T.Sreekumar; Translation of Astanga Hridaya, Sutrasthana; pub: Harisree hospital ,Trissur,Kerala; edition 2007,chapter 1/7; pp-29.

31. Dash Vaidya Bhagwan, Sharma Ram Karan. Charaka Samhita Sutra Sthana , Chakrapanidatta's Ayurveda Dipika Commentary with English translation. Choukhamba Sanskrit Series Office , Varanasi; reprint 2008; chapter 20/11; pp – 364.

32. Dash Vaidya Bhagwan, Sharma Ram Karan. Charaka Samhita Chikitsa Sthana , Chakrapanidatta's Ayurveda Dipika Commentary with English translation. Choukhamba Sanskrit Series Office , Varanasi; reprint 2008; chapter 28/239,245; pp – 84.

Corresponding Author: Dr.Deepa. C Patil. Reader ,Department of KayaChikitsa, JSS Ayurveda medical college, Mysuru, India570028.
Email: rashmik190@gmail.com

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