

## HUNTINGTON'S DISEASE - A CASE STUDY

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### INTRODUCTION:

Hyperkinetic movement disorders are characterized by the presence of variety of different involuntary movements<sup>1</sup>. Out of these disorders, Chorea is the most common and major form of hyperkinetic disorders. The term Chorea is used to describe jerky, semi purposive movements which may affect the limbs, face or trunk<sup>2</sup>. In other words, Chorea can be defined as explosive, fidgety movements, flitting around the body<sup>3</sup>. They are variable in manifestation even in the same patient at different times. The patient is not able to control them, although they may get worse with anxiety and always disappear during sleep<sup>4</sup>. There are so many different causes for the Chorea<sup>5</sup>. In adult life, the most common cause of Chorea is the inherited disorder Huntington's disease<sup>6</sup>.

Huntington's disease can be defined as relentlessly progressive Chorea<sup>7</sup> and psychiatric illness usually behavioral and cognitive dysfunction<sup>8</sup>. Onset is typically between the ages of 25 – 45 years (range 3 – 70 years) with a prevalence of 2 – 8 cases per 100000<sup>9</sup>. It is also known as Huntington's chorea. This is an inherited disorder with autosomal dominant transmission affecting both males and females in adult life<sup>10</sup>. Children of an affected parent have a 50% chance of developing Huntington's disease<sup>11</sup>. Huntington's disease (HD) is characterized by rapid, non-patterned, semi purposeful, involuntary choreiform movements. In the early stages, the chorea tends to be focal or segmental, but it progresses over time to involve multiple body regions. Dysarthria, gait disturbances and oculomotor abnormalities are common features<sup>12</sup>.

According to Ayurvedic literature, it can be understood with special reference to Tandava Roga<sup>13</sup>. Tandava means 'Nrutyam', dancing especially violent gesticulations and frantic dance of Shiva and his votaries. Tandava Roga Nidana (causes), Lakshanas (signs & symptoms) and Chikitsa (treatment) are explained by acharya's<sup>14, 15</sup>. This disease is mostly prevalent in adults especially in females and who are devoid of Bala<sup>16</sup>. Symptomatology of Tandava Roga are well explained in Ayurvedic literature as Vama Bahum Samarabhya Prayo Tatoparam Tatah Pado Tatoangani Chalayet Tandavamya (flitting type of movements predominantly starts from left hand after that spreads to legs and other parts of the body with time), Mustina Kimapi Dravyam Samyagdarayitum Kshmah Samarpayitumasye Vaapyadaniyam Na Tandavi (can't able to hold objects in hands, feed to itself and give to anyone), Nrityanniva Chalatyesh (dancing like flitting movements), Bibhatsee mukha cheshtitee (different expression and movements of face), Adhirah (disturbed intellect) and Nidrayam Kampavarjitah (loss of movements during sleep)<sup>17</sup>.

### OBJECTIVE:

This paper presents a case study of Huntington's disease managed through Ayurveda.

### MATERIALS AND METHODS:

A single case study, qualitative data analysis

#### Diagnostic Criteria:

1. Signs and Symptoms of Huntington's disease<sup>18, 19, 20</sup>.
2. Signs and Symptoms of Tandava Roga<sup>21</sup>.
3. Genomic DNA test<sup>22</sup>: CAG expansion (polyglutamine), Adults: 40 – 55 repeats, Childhood: > 70 repeats.
4. CT scan/MRI scan<sup>23</sup>: Caudate nucleus atrophy

#### Assessment Criteria:

1. Abnormal Involuntary Movement Scale (AIMS)<sup>24</sup>
2. Barnes Akathisia Rating Scale (BARS)<sup>25</sup>

**Approach/About the case:** A female aged about 47 years, Housewife from Hassan was admitted on 30/6/14 in SDMCAH, Hassan having OPD No. 023649 and IPD No. 04074 with the complaints of involuntary and uncontrollable movements of body (hands, legs and head), difficulty in speech, holding objects, swallowing, chewing, hearing, can't able to stand and walk its own with mild behavioral changes.

Patient was said to be apparently health 6 years back. Suddenly one day when she was talking to her family she started with the complaint of involuntary twitching type of movements of neck. After that she consulted to Neurologist and treatment started for the same. After taking treatment for few days, she got some relief in the movement of the neck but after 6 months patient got complaints of sudden onset of involuntary movements of both upper and lower limbs. In starting days, these movements used to be occurred in episodes especially present during talking to her. After the onset of 1 year, symptoms got aggravated and she was unable to do her normal daily activities because she was unable to hold any object due to jerky movements of hands. During this period she was on medications which prescribed

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by Neurologist. Within a period of 3 years, disease progressed and she started with the complaints of difficulty in swallowing, chewing and speaking. After occurring of these all symptoms they consulted to other higher Centre for further management of the condition. There she diagnosed **General examination:**

<b>General Appearance</b>	:	Sick Looking		<b>Cyanosis</b>	:	Absent		<b>Pulse Rate</b>	:	78/min
<b>Built</b>	:	Poor		<b>Lymph nodes</b>	:	Not Palpable		<b>Respiratory Rate</b>	:	16/min
<b>Nourishment</b>	:	Poor		<b>Tongue</b>	:	Coated		<b>Blood Pressure</b>	:	130/90 mm of hg
<b>Pallor</b>	:	Present		<b>Tonsils</b>	:	Normal				
<b>Icterus</b>	:	Absent		<b>Edema</b>	:	Absent		<b>Temperature</b>	:	98.2 F
<b>Clubbing</b>	:	Absent								

**Systematic examination:**

**Respiratory System:** NVBS heard  
Air entry normal  
No added sounds heard

**GIT System:**

Soft, No organomegaly  
No fluid collection  
**CVS:** S<sub>1</sub>, S<sub>2</sub> heard, No added sounds  
**CNS:** Conscious, Well oriented

**Cranial Nerves Examination:**

Result	Examination	Cranial Nerve		Result	Examination	Cranial Nerve
Absent	Eye tightening, Blowing, Whistling	Facial Nerve (VII)		Normal	Smell	Olfactory (I)
Difficult	Swallowing	Glossopharyngeal (IX), Vagus (X)		Normal	Visual Field	Optic Nerve (II)
				Normal	Visual Acuity	
Absent	Shrug shoulders	Accesory Nerve (XI)		Normal	Extra Occular Movements, Nystagmus, Pupils, Accomodation	Occulomotor (III), Trochlear (IV), Abducens (VI)
Abnormal	Tongue Movement	Hypoglossal Nerve (XII)		Difficult	Movement of Jaw	Trigeminal (V)

**Deep Tendon Reflexes:**

Grading	Power of Limbs		Result	Reflex
3/5	Right upper limb		Sluggish	Biceps
3/5	Right lower limb		Sluggish	Triceps
3/5	Left upper limb		Sluggish	Supinator
3/5	Left lower limb		Sluggish	Knee
3/5	Right Hand grip		Sluggish	Ankle
3/5	Left Hand grip			

**Investigations:**

Genomics DNA test	Blood Routine		Serology Report	Serum Electrolytes
Number of CAG repeats on of the alleles at the HD locus falls beyond the normal range of 6 – 33 i.e. 41 repeats	Hb: 10.4 gm%	PCV: 34.8 %	HIV: Negative	S. Sodium: 145.5 mmol/L
	TLC: 5800 cells/cmm	MCV: 92.3 fL	HBSAG: Negative	S. Potassium: 4.4 mmol/L

	ESR: 6 mm/hr	MCH: 30.1 Pg		S. Chloride: 112.9 mmol/L
	Neutrophils: 42%	MCHC: 48.6 fL		
	Lymphocytes: 50%	FBS: 66.3 mg/dl		
	Monocytes: 02 %	Blood Urea: 17.1 mg/dl		
	Eosinophils: 06 %	S. Creatinine: 0.9 mg/dl		
	Platelet count: 2.13 laks/cmm	RBC count: 3.75 millions/cmm		

**Diagnosis:** Huntington's disease (TandavaRoga)

**Intervention:**

1. Tab. Dhanvantari Vati 2-2-2 B/F : 10 days (30/6/14 – 9/7/14)
2. Hingwastaka Churna 1tsf BD with Ghrita between the food : 10 days (30/6/14 – 9/7/14)
3. Nasya with Ksheerabala taila<sub>101</sub> 4 drops morning : 3 days (30/6/14 – 2/7/14)
4. Shirostalam with Brahmi taila OD : 5 days (30/6/14 – 4/7/14)
5. Sarvanga Abhyanga with Ksheerabala taila and Nadi sweda : 7 days (3/7/14 – 9/7/14)
6. Matrabasti with Ksheerabala taila (30 ml) + Samwardhana Ghrita (40ml) : 7 days (3/7/14 – 9/7/14)

**RESULTS:**

**Table 1: Abnormal Involuntary Movement Scale (AIMS)**

Segment	Signs & Symptoms	BT	AT – 3 <sup>rd</sup> day	AT – 6 <sup>th</sup> day	AT – 10 <sup>th</sup> day	% improvement
<b>Facial &amp; Oral Movements</b>	Muscles of Facial Expression	4	3	3	2	50
	Lips & Perioral area	4	3	3	1	75
	Jaw	3	3	2	1	66.67
	Tongue	4	3	2	1	75
<b>Extremity Movements</b>	Upper limb	4	4	3	2	50
	Lower limb	4	4	3	2	50
<b>Trunk Movements</b>	Neck, Shoulder & Hips	4	3	3	1	75
<b>Global Judgment</b>	Severity of abnormal movements overall	4	3	3	2	50
	Incapacitation due to abnormal movements	4	3	3	2	50
	Patient's awareness of abnormal movements	3	3	2	1	66.67
<b>Dental Status</b>	Current problem with teeth/dentures	No	-	-	-	-
	Are dentures usually worn	No	-	-	-	-
	Endentia	No	-	-	-	-
	Do movements disappear with sleep	Yes	-	-	-	-

BT: before treatment, AT: after treatment, 0: none/normal, 1: minimal, 2: mild, 3: moderate, 4: severe

**Table 2: Overall assessment of segments of AIMS:**

Segment	BT	AT – 3 <sup>rd</sup> day	AT – 6 <sup>th</sup> day	AT – 10 <sup>th</sup> day	% improvement
<b>Facial &amp; Oral Movements</b>	15	12	10	5	66.67
<b>Extremity Movements</b>	8	8	6	4	50
<b>Trunk Movements</b>	4	3	3	1	75
<b>Global Judgment</b>	11	9	8	5	54.55

**Table 3: Barnes Akathisia Rating Scale (BARS)**

Category	Segment	BT	AT – 3 <sup>rd</sup> day	AT – 6 <sup>th</sup> day	AT – 10 <sup>th</sup> day	% improvement
<b>Objective</b>	Movement	3	3	2	1	66.67
<b>Subjective</b>	Awareness of restlessness	3	3	2	1	66.67
	Distress related to restlessness	3	3	2	1	66.67
	General clinical assessment of Akathisia	5	5	4	2	60

## DISCUSSION:

Tandavaroga mentioned in Sharangadhara Samhita<sup>26</sup> (Parishista 1) comprises neurodegenerative disorders including Huntington's disease (Huntington's chorea). The aetiology, pathogenesis and symptomatology of Huntington's disease is easily understood and correlated with that of Tandava Roga.

Specific aetiological factors which are Rajoavrodha, degenerative pathology (Vata vrudhi) substantiate aetiology of Huntington's disease. The pathogenesis of Tandava Roga starts with Majja Dhatu (Mastulunga Majja Dhatu) kshaya and consequently pathology will be seen in turns of Balakshaya and Pratat Vata Rogi<sup>27</sup>. Neuro degenerative disorders causing cerebellar atrophy with marked loss of neurons in caudate nucleus and putamen. In further stages it causes changes in neurotransmitters leads to progressive symptomatology of rapid, non-patterned, semipurposeful involuntary choreiform movement with myopathy and dementia (Pratat Vata Rogi & Balamamsakshaya). Hence, Nidanapanchaka's of Tandavaroga get correlated with that of Huntington's disease.

The symptomatology of Huntington's disease involves the Vata prakopa and Majja Dhatukshaya. The presenting complaints like difficulty in swallowing of food, speech, loss of strength (Balamamsakshaya) and change in behavior implies for Prana and Udana Vatavikruti<sup>28</sup>. Symptomatology of Progressive, rapid, non – patterned, semi purposeful movements of whole body gives the clue for Vyana Vatavikruti<sup>29</sup>. On the other hand, loss of appetite indicates for the pathology of Samana Vata vikruti<sup>30</sup>.

Hence, treatment is planned 1) in accordance with Agnideepana, Pranarakshna and Urdhwajatrugata Vikara chikitsa. As it is Dhatukshayajanya and Agnimandya pathology so Snehika Nasya (Brihmana) and Agnideepana chikitsa was planned. Medicines selected for above plan were Ksheerabala<sup>101</sup> drops Nasya, Hingwastaka Churna with Ghrita and Dhanwantari vati. 2) After giving treatment for Prana, Udana and SamanaVata treatment of Vyana Vata and Brihmana Chikitsa was planned as second line of treatment. For achieving this aim, Sarvanga Abhyanga with Ksheerabala taila and Nadi sweda was planned with accordance with Desha and Kala. 3) Matra Basti was planned with the combination of (Yamaka Sneh) Samwardhana Ghrita and Ksheerabala taila for the purpose of giving Chikitsa to all Pancha Vata and especially to give nourishment to the Majja and Mamsa Dhatu. The ingredients of Samwardhana Ghrita like Khadira, Prashniparni, Saindhava, Bala, Atibala, Kebuk<sup>31</sup> (kashaya form) helps for increasing the Mamsa and Majja Dhatwagni. The benefits of Samwardhana Ghrita like Shigra Nirvyadhi Vardhate (soon get free from disease and lead to growth, Samsarpatyashu Gachhati<sup>32</sup> (soon starts walking) and Ksheerabala taila like Rasayana, Brihmana, Indriya prasadana<sup>33</sup>. Samwardhana Ghrita is indicated in diseases like Pangu, Muk, Ashruti, Jadta (Dementia, Cerebellar dysfunction). Hence, Hetu Sthana Vishesha Chikitsa<sup>34</sup> is planned accordingly in the present case.

## CONCLUSION:

Tandava Roga comprises neurodegenerative disorders including Huntington's disease. Symptomatology of Huntington's disease involves the Vata Prakopa and Majja Dhatu kshaya. After 10 days of treatment, in female patient of Huntington's disease/chorea there was remarkable improvement seen in signs & symptoms.

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