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AN AYURVEDIC PERSPECTIVE OF FRIEDREICH'S ATAXIA

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ABSTRACT

Friedreich's ataxia is an autosomal recessive inherited disease that causes progressive damage to the nervous system. The recent discovery of the gene that is mutated in this condition, FRDA, has lead to rapid advance in the understanding of the pathogenesis of Friedreich's ataxia. About 98% of mutant alleles have an expansion of a GAA trinucleotide repeat in intron 1 of the gene. This leads to reduced levels of the protein Frataxin. There is mounting evidence to suggest that Freidreich's ataxia is the result of accumulation of iron in the mitochondria leading to excess production of free radicals, which then results in cellular damage and death. Currently in modern medicine there is no known treatment that alters the natural course of the pathology. The *Ayurveda* approach to Friedreich's ataxia is aimed at controlling the symptoms, treating the degeneration of nervous system and preventing long term complications by considering it as *PRANA AVRUTA VYANA UDANA VATA VYADHI.*

Keywords: Friedreich's ataxia, GAA, Frataxin, *prana avruta vyana udana vata.*

INTRODUCTION

Friedreich' ataxia, is the most common of the hereditary ataxia[1] with an incidence rate of 1 in 50,000 people with an estimated carrier prevalence of about 1:110.[2-4] It was first described by German physician Nicholaus Friedreich in the 1860's.[5-9] It is also called FA or FRDA. It is a rare inherited disease that causes progressive damage to the nervous system associated with movement problem caused due to reduced levels of Frataxin in the cells. It usually begins in childhood and leads to impaired muscle coordination (ataxia) that worsens over time. It results in symptoms ranging from gait disturbances to speech problems. It can also lead to heart disease and diabetes, but does not affect cognitive function. The disease progresses until a wheelchair is required for mobility[10]. FA is caused by a defect (mutation) in a gene labeled FXN. FXN gene codes for production of a protein called frataxin.[11] Frataxin is an iron binding protein present in the mitochondria which helps in the synthesis of Fe-Su clusters which aid in electron transportation for the production of ATP, the energy currency necessary to carry out metabolic functions in cells.[12] Without

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a normal level of frataxin, certain cells in the body (especially peripheral nerves, spinal cord, brain and heart muscle cells) cannot effectively produce energy and they have been hypothesized to build up toxic byproducts leading to what is called oxidative stress. It also may lead to increased levels of iron in the mitochondria.[12-13] When the excess iron reacts with oxygen, free radicals can be produced which then destroy cells and harm the body. According to *Ayurveda*, in FRDA, Frataxin may be correlated with *prana vata* functions, which controls *prayatna* action of *udana* and in turn *gati* induced by *vyana vata* becomes impaired. Due to decreased *prayatna* (action potential), there is less *urja karma* (ATP production) leading to decreased *bala* (cellular metabolism) and there is *ama dravya* produced at cellular level. This *ama* leads to cellular *strotorodha* (obstruction) and *vimargagamana* of iron which then reacts with oxygen to produce free radicals (*ama visha*) and destroy the cell. At the end, *vata prakopa* occurs and manifest the features of FRDA. On keen observation, *prana* as *avaraka* (which obstructs) and *udana* and *vyana* as *avarya* (which is getting obstructed) participate in pathogenesis. *Avarana* initiates pathogenesis followed by dhatu (body elements) *kshaya* (diminution) and *vata vyadhi.* Frataxin is protein, that is *mamsa poshakamsha* in *rasa dhatu,* and iron as *rakta poshaka bhava*. From this it can be said that in FRDA, *Avarka* is *Prana vata, Avarya* are *Udana and Vyana vata, Dosa* is *Vata, Dushyas* are *Rasa, Rakta* and *Mamsa dhatus.* It can be concluded that FRDA is a type of *vata vyadhi* caused by *PRANA AVRUTA VYANA, UDANA.*

METHODS

DIAGNOSIS:

A) The main clinical features for diagnosis are: [14]

Primary criteria: 1) age of onset of symptoms before the age of 25 years. 2) Progressive unremitting ataxia of limbs and of gait. 3) Absence of knee and ankle jerks. Secondary criteria: 1) dysarthria 2) extensor plantar responses.

If secondary criteria are absent, the following have to be present, 1) an affected sibling fulfilling primary and secondary criteria. 2) median motor nerve conduction of greater than 40m/s. thus excluding cases of type I hereditary motor and sensory neuropathy.

Cardiomyopathy, scoliosis, and foot deformity, are common but non-essential features.

B) Investigations for diagnosis are:

1) Nerve conduction studies show motor velocity greater than 40m/s. in arms and absent sensory action potentials.
2) MRI of brain and spinal cord shows characteristic atropic changes, particularly of the cervical spinal cord.
3) ECG - there may be ventricular hypertrophy, T- wave inversion.
4) ECHOCARDIOGRAPHY- may show ventricular hypertrophy, septal hypertrophy and hypertrophic cardiomyopathy.

TREATMENT PROTOCOL:

The cause that is the *avarana* should be treated first. Frataxin deficiency is caused by gene defect. So, *Rasayana dravyas* (rejuvenating drugs) like *Amalaki* (Phyllanthus emblica), *Haritaki* (Termina-

lia chebula) etc,. should be used, which are found to act on genes and these *dravyas* are also immunomodulators.

General line of treatment of *avarana* can also be adopted like[15]

1) *Yapana vasti* prepared with *madhura dravyas.*
2) *Anuvasana vasti.*
3) *Samsrana*
4) Administration of *rasayana* like *shilajit, rasona, eranda, gokshura, guggulu* along with milk.
5) *Chyavanaprasha along* with milk.

Specific line of treatment for *prana avrta udana and prana avrta vyana* can also be adopted.[16-17]

1) *Nasya karma*
2) *Dhuma*
3) *Gandusha*
4) *Akshitarpana*
5) *Karnapoorana* etc,.

DISCUSSION

FRDA the most common of hereditary ataxia is caused by a mutation in gene labeled FXN. As the natural course of the pathology cannot be altered by the modern medicine, the scope of Ayurveda interventions in such nervous disorders has been increasing because of its holistic approach. Understanding such disorders in Ayurveda perspective is need of the hour. Nervous system and its disorders are described under the heading of *vata vyadhi* in *Ayurveda*. In *Ayurveda*, *vata vyadhis* are divided into two broad categories namely diseases due to *VISHUDDHA VATA(*vitiation of vata alone) *and* diseases due to *AVARANA(*due to blockade in the natural flow of vata by its types or by other dosa or dhathu or mala). Proper knowledge of the types of *VATA(prana, udana, samana, vyana, apana)* , their normal functions,[18] categorizing the disorder either as *vishuddha vata janya* or *avarana janya* helps in understanding the diagnosis, prognosis and how to plan the treatment protocol. Further research work in application of these principles in understanding and treating of FRDA has to be carried out.

CONCLUSION

In FRDA which as no direct reference from the classics, the treatment protocol can be planned by understanding the underlying pathology of the disease in Ayurveda perspective. Here it can be interpreted FRDA has *prana avruta vyana, udana vata vyadhi* and *avarana chikitsa* in general and specific to particular avarana can be adopted.

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