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UNDERSTANDING THE MANAGEMENT OF DUCHENNE MUSCULAR DYSTROPHY AS PER AYURVEDA

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ABSTRACT

The muscular dystrophies are genitacally determined,progressive,primary disorders of muscle.Common presenting symptoms include delayed mile stones,abnormal mode of walking,difficulty in running,climbing stairs and getting up from floor,frequent falls,weakness of arms or face,ocular symptoms,enlargement or wasting of muscles and recurrent chest infections.The symptoms are progressive and a family history of similar illness may be obtained.Duchenne muscular dystrophy(D.M.D) is a severe muscle wasting disorder,resulting in early confinement to wheel chair and often death by the age of 20 years. There is no cure available for this disorder at present,prevention & treatment of complications is of prime importance as per modern medicine, but Ayurvedic treatments show significant results in all signs and symptoms of disease.

Keywords: D.M.D,*Mamsadhathu vikara*,Ayurvedic management.

INTRODUCTION

Duchenne Muscular Dystrophy is an inherited neuromuscular disorder characterized by muscular weakness,pseudohypertrophy,fibrous tissue prolifiration in the muscles & mental retardation. It is X-linked recessive disease. The abnormal gene causing DMD is located on the short arm of the X-Chromosome at theXp21 site. Since it is an X-linked disease,it ,it is seen in males with history of similar

illness in the meternal uncle[1] the pooled global D.M.D birth prevelance was 2.8 cases per 100,000 live male births[2]. It is a severe muscle wasting disorder, resulting early confinement to wheel chair and often death by the age of 20 years.Hypertrophy of calf muscles may be observed by the age of 4-5 years,onset of cardiac involvement being usually after the age of 10 yrs,Wheel chair dependency typically occurs by about12 years of age,Death in DMD most commonly results from pulmonary insufficiency and respiratory infections at about 20 yrs of age.The average intelligence quotient is approximately one standered deviation below the mean.[3]

PATHOGENISIS:

Dystrophin is a protein which is present in the skeletal muscles,smooth muscles&cardiac muscles.It is produced by the normal functioning of the gene at Xp21 site of the chromosome. The protein Dystrophin is required for structural support of the muscle cells.Its loss causes excess permeability of muscle cell membrane eventually leading to cell death.Beacause of delition or mutation of this gene, dystrophin production is either reduced substantially(leading to DMD) or abnormal dystrophin which is partially functional may be produced(leading to Backer'myopathy).[4]

Reduced/Absent dystrophin

↓

Reduced glycoproteins in the muscle cells

↓

Become more prone to necrosis

↓

Muscular wasting(necrosis)

CLINICAL FEATURES:[5]

- Though present from birth,clinical features of DMD become apparent only between 3-5 years of age.
- Thus, the early motor milestones such as Roll over,sitting,walking are achieved at normal age. However, a keen mother or clinician may notice poor head control, frequent falls &difficulty in keeping up with children of same age group while running.
- Gower's sign(demonstrated by asking the child to get up from supine position, resulting in the child climbing up onto himself using his hands)
- Pseudo hypertrophy of calves is a distinctive feature of DMD.
- Extramuscular involvement in DMD
  - Mental retardation
  - Cardiomyopathy
  - Macroglossia
  - Dilatation of the stomach
  - Intestinal obstruction
  - Convulsions
  - Death usually occurs by 20 years of age.

Causes of death in DMD are:

- ❖ Respiratory failure due to:
- ➢ Repeated respiratory infections& pneumonia
- ➢ Weak chest muscles
- ➢ Aspiration
- ➢ Scoliosis leading to severely reduced respiratory reserve

❖ Cardiac causes:
- ➢ Cardiomyopathy causing intractable Congestive Cardiac Failure(CCF)
- ➢ Arrhythmias

❖ GI causes:
- ➢ Acute Dilatation of the stomach
- ➢ Intestinal obstruction

INVESTIGATIONS:[6]
- o Creatine phospokinase levels are elevated in children with DMD .

CPK values are >2000U/L in DMD(Normal values are <160-200U/L)
- o Muscle biopsy is diagnostic of DMD
- o PCR: It is done on blood sample to detect the mutation of gene encoding dystrophin
- o Defect in dystrophin protein demonstrated in muscle biopsy sample(Western blot method)

To rule out complications:
- ▪ X ray chest- To rule out respiratory tract infections & cardiomegaly
- ▪ ECG –For assessment of the cardiac function
- ▪ Pulmonary function test – To determine the respiratory reserve
- ▪ IQ/DQ test – For Mental retardation

Prenatal diagnosis:
- ➢ **Chorian villus biopsy-**Earliest diagnosis of DMD can be done at about 10 weeks of gestation by Chorian Villus Biopsy.
- ➢ **Amniocentesis –** If chorian villus biopsy is not available, then amniocentesis can be performed at16-18 weeks of gestation.
- ➢ **Fetal blood sampling –** It can be done at18 weeks of gestation. It shows increased CPK levels in affected fetuses.

MODERN TREATMENT OF D.M.D[7]:

There is no cure available for this disorder at present. Prevention & treatment of complications is of prime importance.
- Physiotherapy
- Orthopedic surgery
- Use of steroids
- Genetic counselling
- Treatment of respiratory infections etc

AYURVEDIC MANAGEMENT:

DMD cannot be correlated directly with any of the diseases mentioned in classics,it may be compared with *mamsa dhatu vikara* due to *vata vaisamya*.According to *Charaka mamsa dhatu gata vata lakshanas* like *Gouravam*(heaviness of the body parts),*Atyartha toda*(severe pain),*Danda musti hatam peeda*(severe pain as if beaten with fist/sticks),*Saruk*(distress),*Atyartha srama*(excessive exhaustion).*Niruha vasti, Virechana,Samana chikistha* is treatment of *mamsa* & *medho dhatu gata vata*.[8] So practically *panchakarma* procedures like *Abhyangam, Shastika sali pinda swedam, Niruha vasti* plays significant role for management of D.M,D.*Samana chikistha* like *Deepana,pachana* drugs,*vata hara* drugs,*balya,brumhaniya* drugs,*jeevaneeya* drugs, medhya,hrudya drugs are so beneficial for in this condition. *Deepana,pachana* drugs acts as *amapachana,*helps in proper *dhatu parinama,Balya,brumhaniya,jeevaneeya* drugs acts as muscle bulk promoting and *Medhya* drugs acts as I.Q improvement.*Hrudhya* drugs helps in prevention of cardiac complications which is commonly seen in D.M.D.

DISCUSSION

DMD cannot be correlated directly with any of the diseases mentioned in the classics.In Ayurveda DMD pathogenesis can be clearly understood by the concept of *Adibala pravritha vyadhi* which is in *Susrutha's vyadhi vargikarana* comes under *Adhyathmika vyadhi*.*Adibala pravritha* diseases are those produced by the abnormalities of *sukra&sonitha*.Hence the disease occurs due to *matrija beeja bhaga avayavadusti*. Here the disease is purely due to *mamsa dhatu vikara* which means the part of *matrija beeja* which is responsible for formation of *mamsa dhatu.*The complex variety of pathogenesis its responsible for indeed is responsible for the progressive wasting and necrosis of muscle fibres.Therefore the disease manifests due to *vata vaishamya* and considered under *vata vikara*.

In DMD the main symptoms are degeneration of muscle&cardiac involvement and in some cases mental retardation or poor IQ also.So along with *sodhana chikista* a group of *Balya,*

Jeevaniya,Brumhaneeya,Rasayana,Hridya,Medhya& vatahara drugs etc are advised.

CONCLUSION

D.M.D is a neuromuscular disorder,there is no safe& significant treatment for D.M.D at present.Ayurvedic treatments like *shodhana,samana* theraphies are somewhat effective on slowdown the bad prognosis of disease condition and also preventing th complications.

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