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A Review on Muscular Dystrophy in Children & It's Management

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Abstract:

Muscular dystrophies are a group of genetic disorders which causes weakness & muscle wasting of skeletal muscle. The term dystrophy is derived from Greek word, Dys means faulty & trophy means nourishment. Muscular dystrophy is a genetic disorder with X-linked recessive inheritance.

Most of the muscular dystrophies are progressive in nature and they worsen with time. Modern drugs give temporary relief from symptoms but they can not delay the process of weakness of muscles. Ayurveda slow down progression of disease by reducing kapha kshaya & consequently Mansa kshaya.

Ayurveda gives nutrion to muscles through Bruhan & Rasayan chikitsa which gives energy to affected child & keeps him ambulatory. Ayurvedic management includes Yoga which keeps child physically as well as mentally fit & happy.

Introduction:

Ayurveda is the most ancient and effective medical science. The pledged purpose of Ayurveda as a medical system is to ensure a healthier and longer life to the humanity. In Ashtanga Ayurveda Kaumarbhritya tantra is as precious as 'Agni' among all gods.

Muscular dystrophy means abnormal growth and poor nourishment of muscle fibres. The term dystrophy is taken from Greek word Dys means faulty, trophy means nourishment. Any disorder caused due to faulty nutrition is called as dystrophy.

Muscular dystrophy is a genetic disorder which leads to weakness and muscle wasting. Most of the muscular dystrophies are progressive in nature and conditions worsen with the time. It is a genetic disorder with X-linked recessive inheritance with affected males and carrier females. Modern science has got few limitations in management of muscular dystrophies. Modern drugs have shown temporary improvement in muscle weakness but it can not affect course of the disease. Physiotherapy helps in delaying the process of weakness of muscles. Ayurvedic treatment is effective in muscular dystrophy so patients are approaching ayurveda.

Muscular dystrophies are

a) Hereditary
b) Progressive in nature
c) Primary myopathies of normally formed muscles

All muscular dystrophies have different genetic trait and each differing in its clinical course and expression. Some diseases are severe at birth and rapid progressing and other follow very slow progression.

Types of Muscular dystrophy:-

1) Duchenne muscular dystrophy-

It is a X-linked recessive disease with affected males and carrier female due to deletion of one or more exons of the DMD gene, located on Xp21 locus. However 30% cases may be new mutations. Girls are very rarely affected, if having Turner syndrome (single X chromosome) or random inactivation of normal chromosome(Lyon hypothesis). It is a commonest hereditary myopathy in Indian children.

Primary product of DMD gene is an important cytoskeletal protein- **Dystrophin,** present in skeletal & smooth muscles, brain, peripheral nerves and many other tissues. This dystrophin is absent or severely deficient in DMD.

Boys are asymptomatic at birth with normal early development with normal early development including walking. In a typical

case of DMD, weakness usually begins at 2-3 years of age with

- Mild waddling (Trendelenburg) gait with a lordotic posture to adopt for gluteal weakness.
- Positive Gower sign, I.e. difficulty in standing from recumbent position due to pelvic girdle weakness. When asked to do so, he typically turns to his side, lifts his trunk up by supporting his weight on his arms and then stands up as if climbing upon his body with hand support.
- Pseudohypertrophy of calf muscles with wasting of thigh muscles. Tongue and forearms may also develop Pseudohypertrophy. Distal deep jerks, e.g. ankle and brachioradialis are stronger than proximal knee or biceps/triceps jerks.
- Progressive weakness of proximal muscles including respiratory involvement with weak cough and recurrent respiratory infection, and/or pharyngeal muscle weakness with recurrent aspirations, nasal twang and regurgitation. Distal muscle functions e.g. writing, etc. Are spared till terminal stage.

Associated features, e.g. cardiomyopathy and mild mental retardation or learning disabilities are present in nearly all cases with variable severity, not necessarily correlating with severity of muscular disease. Cardiomyopathy usually appears at ~10 years, though CCF and arrhythmia are uncommon till terminal stage and usually precipitated by intercurrent infections. Intermittent gastric dilatation with abdominal pain and vomiting is common.

Course:- Patient gradually develops muscle weakness and becomes bedridden by 7-10 years of age with development of contracture and scoliosis and die by 15-20 years due to respiratory failure, aspiration or congestive cardiac failure.

2) Becker muscular dystrophy-
It is milder variant, which usually present in late childhood with inability to raise hand above shoulder. Child remain ambulatory till 15-20 years and survive till 4^{th}-5^{th} decade. Milder deficiency of Dystrophin is present in BMD.

3) Limb Girdle muscular dystrophy-
It is group of slowly progressive inherited disorders, mainly affecting muscles of the hip and shoulder girdle. Some of them are autosomal dominant and others are recessive. Most cases present in adolescence.

4) Facio-Scapulo-Humaral muscular dystrophy-
It is autosomal dominant muscular dystrophy. It manifests at about puberty with facial weakness (inability to close eyes, whistle, smile) shoulder girdle weakness and upper limb weakness.

5) Myotonic Muscular dystrophy-
It presents with typical facial features, progressive weakness of distal muscles eg. hands. Presence of myotonia I.e. slow relaxation of muscles after contraction.

6) Congenital muscular dystrophy-
It is autosomal recessive muscular dystrophy consists of many distinct disorders all with severe diffuse hypotonia and proximal muscle weakness at birth. Facial, ocular and bulbar muscles are relatively spared. However further course is slow progressive and many cases are able to stand and walk with support in late childhood, unless contractures have developed.

CNS involvement is common with mental retardation and white matter changes on MRI. Cpk is moderately elevated and biopsy shows signs of muscular dystrophy with merosin deficiency on immuno histo chemist.

Diagnosis :-

1) Blood test
   a) Creatinine phospho kinase level-
   It is elevated in most of the carrier females and in patients in early stages even before clinical manifestations become obivious.
   b) SGOT
   c) SGPT

2) Electro Myography
EMG shows characteristic myopathic features. It can give the dystrophy diagnosis but can not distinguish the specific type of muscular dystrophy.

3) Histo pathology studies

Muscle biopsy shows diffuse changes of degeneration, variation in size and central nuclei of muscle fibres.

4) Prenatal diagnosis

It is possible as early as by 12 weeks by PCR DNA analysis on chronic villi samples.

Management according to Modern science-

- It is entirely supportive at present though experimental studies after discovery of dystrophin molecule are promising.
- Proper nutritional support should be given.
- Physiotherapy delays but does not prevent contractures.
- Treatment of respiratory infection. Diagnosis and management of congestive cardiac failure.
- Calcium supplements to prevent osteoporosis.
- Psychological support
- Steroids (Orally prednisolone 0.75 mg/kg/d for 10days)

Ayurvedic view

According to prachin ayurvedic samhitas muscular dystrophy presents as 'Mans-shosh'. In this vyadhi because of vitiated doshas there is progressive impairement in movements of child. It affects day today lifestyle of child.

It is considered under Adibalpravrutt vyadhis.

● **Nidan –**

Causes of muscular dystrophy according to ayurveda-

1) Defect in matruj bhava as Mansa is derived from maternal factors.
2) Partly vitiated shukra or shonita.
3) Specific Beejabhag or Beejabhagavaya defect.

● **Roop –**

Progressive impairment in movements of child shows vitiation of Vata.

Metabolism is impaired because of Pitta dushti.

Because of Kapha dushti quality like 'Sthiratva' is impaired.

● **Samprapti –**

Due to defect in specific beej bhag(specific gene) or Matruj bhav- Mansa, there is defect in muscle.

According to ‘ Dhatu-parinaman Nyay' mansa is prepared from Sar bhag of Rakt dhatu. If Rakt dhatu is deficient in enzymes, proper conversion of rakt sar bhag into mansa does not occur which leads to accumulation of rakt sar bhag producing ‘Aam'. The part of Rakt sar bhag is necessary for proper development of other succeeding dhatus.

Because of improper development of Rakt sar bhag, there is improper development of mansa which results in muscle wasting and muscle dystrophy.

The clinical signs and symptoms with respect to doshas shows that there is dominance of Pitta in initial stage which is marked by defective dhatu-parinaman. When there is non production of concern enzymes due to genetic involvement resulting in Rakt dushti and vitiation of Pitta which results in Kapha-kshay. Progressive impairment in movements of child shows involvement of Vata. Mansa dhatu kshay is the chief reason for vitiation of Vata.

Samprapti Ghatak-

- Dosh- Tridosh
- Dushya- Ras, Rakt, Mansa
- Agni- Jatharagni, Rakt and Mansa dhatu agni
- Adhishthan- Mansa (muscle)

Sadhyasadhyatva

It is Asadhya but can be made Yapya by early detection and proper management.

Aim of the treatment-

¡) To slow down the progression of disease to maintain independent walking of child

¡¡) To help child for living his normal life.

Chikitsa according to Ayurveda-

- As muscular dystrophy has genetic origin, Sannipatik nature and Yapya prognosis of disease, the management is done for bringing back the equilibrium of vitiated doshas by proper, timely and continuous langhan and bruhan procedures.
- The etiopathogenesis of the disease is mainly concerned with Sannipatik with Pitta predominance and dhatvagni mandya in initial stage, and hence management should be deepan, pachan followed by Pittahara ghratpan and virechan with madhur, tikta and shit dravyas.
- In the second stage, the vitiation of Pitta results in Kapha-kshay and consequently mansa kshay. So treatment should have bruhan and rasayan .
- In third stage Vatik complications are caused due to Kapha kshay and consequent dhatu-kshay. Hence these complications should be managed by snehan and mridu swedan.

Hence Treatment protocol for management of muscular dystrophy includes-

- Deepan, Pachan
- Mrudu-Shodhan with madhur, tikta and shit dravyas
- Bruhan and Rasayan

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