MUSCULAR DYSTROPHY (M.D.)

Demis Cachia SROT

WHAT IS MUSCULAR DYSTOPHY?

The word dystrophy comes from the Latin and Greek roots meaning "faulty nutrition". During the 19th century, tools were very limited in diagnosing Muscular Dystrophy. Muscles in many diseases appeared to be wasting away, and the doctors theorised that they somehow weren't being properly nourished.

Today, we know that many muscle-wasting diseases are caused by defects in genes for muscle proteins. Most of these proteins appear to play a role in supporting the structure of muscle fibres, although some may play a role in the biochemical process that go on in muscle fibres. The term M.D. refers to a group of genetic diseases marked by progressive weakness and degeneration of the skeletal or voluntary muscles, which control movement. The muscle of the heart, in some forms of M.D., is also affected.

M.D. can affect people of all age. Some forms of M.D. can appear in infancy or childhood; others may not appear until middle age or later. With the progress in medical care, particularly of problems affecting the heart and lungs, children with M.D. are living longer.

The major forms MD include:

1. Myotonic
2. Duchenne
3. Becker
4. Limb-Girdle
5. Facioscapulohumeral
6. Congenital
7. Oculopharyngeal
8. Distal and Emery-Dreifuss

Some of these names are based on the locations of affected muscles i.e. "Facioscapulohumeral" refers to muscles of face, scapula and humerus. Others are based on the muscle protein involved, the onset age or the doctors who first describe the disease.

The symptoms of this condition differ in severity, according to the age of onset, primary muscles involved, the rate at which they progress, and the way the disease is inherited.

WHAT CAUSES M.D.?

A flaw in muscle protein genes causes MD. Each cell in our bodies contains tens of thousands of genes. Each gene is a string of chemical DNA and is the "code" for a protein. If the composition of a protein is inadequate, it affects its make up and the amount produced.
All forms of MD are generally inherited but in some cases no family history of the disease may exist. MD is not contagious.

DIAGNOSIS OF M.D.

- Evaluation of patient medical history,
- Physical examination,
- Diagnostic tests used to distinguish between different forms MD including:-
  - Studying muscle tissue – muscle biopsy
  - DNA testing
  - Electromyogram (EMG)
  - Nerve Conduction Velocity (NCV)
  - Blood Enzyme Tests

THE TEAM APPROACH TO HOLISTIC CARE

In order to provide quality treatment and care of the individual with Muscular Dystrophy, a comprehensive and individualised approach is required. The health care team may include physician, neurologist, orthopaedic, nurse, social worker, rehabilitation counsellor, clinic-school liaison, occupational therapist and physiotherapist. Roles may overlap according to the needs of each client and situation. Patient care should be a co-ordinated effort and communication among team members is vital.

The roles of health care team members in treating this condition are described below:-

PHYSICIAN

The physician is responsible for diagnosis and medical management. The physician works with the nurse to direct patient care, the physical and occupational therapists to develop an individualised muscle care and adaptive living skills program, and the psychosocial staff to ensure the individual's and family's emotional well-being at home and in the community.

Responsibilities of a physician within the team are multiple: providing direct patient care, educating medical colleagues, allied health professionals, the lay public and, most importantly, individuals affected by muscular dystrophy and their families. One of the most significant roles of the physician is as co-ordinator; to obtain and utilise information and services from various allied health professionals.

NURSE

The nurse has a central role in co-ordinating daily operation of the health care team. Because the nurse provides a critical element of continuity between outpatient and inpatient services when hospitalisation is required, a close relationship often develops between the nurse, the individual affected by MD and the family. As a result of such close relationships, the nurse can often recognise and inform other team members about psychological, social and financial conditions that could affect overall management.

The nurse can provide support to an individual affected by M.D. and his family during significant events such as diagnosis, alterations in treatment and death. Experience with health care

problems that frequently occur in the disease, together with personal knowledge of each individual treated in clinic; can allow the nurse to facilitate the organisation of care provided.

PSYCHOSOCIAL STAFF

(Social Worker, Rehabilitation Counsellor, Psychologist)

M.D. has significant psychosocial implications, particularly during potential developmental crisis such as diagnosis, alterations in therapy, adolescence and death. The psychosocial members of a health care team can evaluate each family's and individual's ability to cope with issues related to M.D. and determine appropriate therapeutic intervention when needed.

Referral to a professional therapist or to support services available in the community may be helpful. In addition to reinforcing the importance of medical care, psychosocial team members can help the family and individual with M.D., function at an optimum emotional level. Because of the extensive amount of attention required by the disease and its effects upon a growing child and adolescent, relationships among siblings and parents can be affected. By providing education, support and assistance in locating community resources, problems can be reduced for the entire family.

The psychosocial staff members also assist the family and those with M.D. in obtaining financial assistance through appropriate state benefits. By providing guidance in education, community involvement and planning for the future, the psychosocial team members help those with M.D. prepare for the greatest possible degree of independent living.

CLINIC - SCHOOL LIASON

(Facilitators)

The majority of individuals with M.D. are adolescents or younger and should be involved in some type of educational program. A trained professional, familiar with general and special education programs, school law and teaching methods can be an invaluable asset to a family and to the school. By helping to educate school staff and student peers, teachers can encourage understanding and healthy adaptations that keep the student with M.D. involved in school activities.

As a liaison person between the clinic and school, the facilitator can encourage appropriate annual educational evaluations and adequate school placement with consideration of the student's continually changing abilities as the disease progresses.

PHYSICAL THERAPIST

Currently there is no treatment that can stop or reverse the progression of muscle degeneration in muscular dystrophy. However, maintenance of muscle strength through physical therapy can allow

individuals to function at an optimum physical level and avoid premature contractures and joint stiffening. Regular evaluations are necessary to provide information for development of an individualised physical therapy program.

In addition to direct care, physical therapists instruct family members, young adults with M.D., health care providers and school personnel in proper techniques for daily physical therapy exercises.

OCCUPATIONAL THERAPIST

As M.D. progresses, adaptive equipment becomes increasingly valuable in maintaining an individual's independent functioning. Assistance with activities and recreation can be provided by the occupational therapist.

Occupational and physical therapists work closely in evaluating muscle strength, abilities and limitations.

The need for adaptive equipment may help the patient to become more aware of The physical therapist can recommend exercises that will continue to maintain flexibility and strength while taking into consideration adaptive equipment recommended by the occupational therapist. The need for adaptive equipment may help the patient become more aware of his decreasing physical abilities. The emotional support of the occupational therapist, while encouraging the use of equipment, can greatly enhance the overall success of medical management.

BIBLIOGRAPHY

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