Exercise strategies for strength maintenance in pediatric NMD

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Neuromuscular Dystrophy (NMD)

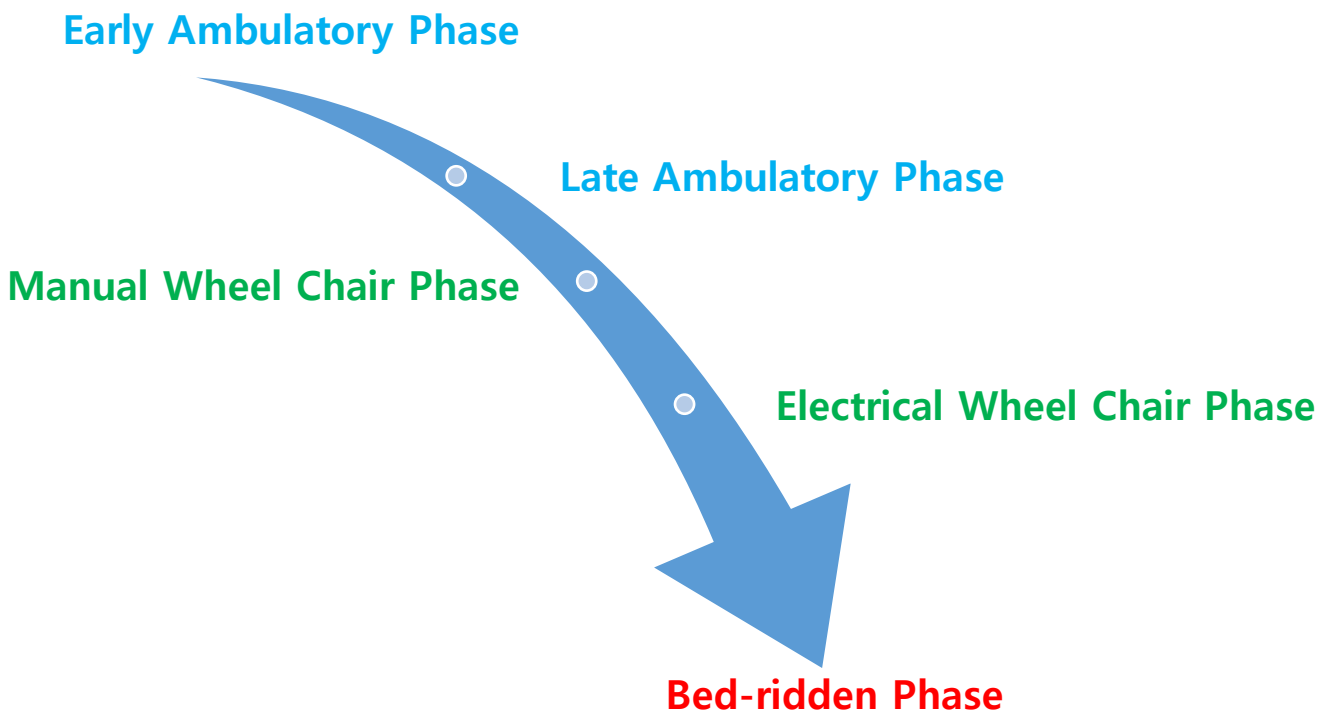
- Muscular dystrophies : Duchenne and Becker muscular dystrophy, Other types of muscular dystrophy (Myotonic, Facioscapulohumeral, Congenital, Limb-girdle)
- Neuropathies : Guillain-Barré syndrome, etc.
- Motor neuron diseases : spinal muscular atrophy, etc.
- Neuromuscular junction disorders : myasthenia gravis, etc.

3 major time points in NMD

- Physiatrist meets 3 major time points for NMD, when begin to easy fall-down, difficult to gait, and difficult to cough.

- Donders J, Taneja C. Neurobehavioral Characteristics of Children with Duchenne Muscular Dystrophy. Child Neuropsychol. 2009 Jan 22. 1-10.

Recommending Exercise in each major time points



Early Ambulatory Phase, 4-7 years

- Resisted exercises should not be prescribed as there is **no evidence** that they are useful, but they may accelerate **muscle damage**.
- Active exercise particularly in the **hydrotherapy pool** is recommended.
- Children taking **steroids** may acquire additional strength and endurance

Early Ambulatory Phase

- Use of **steroids** are the gold standard treatment for ambulant children with DMD.
- Timing: well before the loss of ambulation
- Efficacy: muscle strength, Forced Vital Capacity (FVC)
- **Side effects**: weight gain, reduction of bone mineral density
- No evidence for any beneficial effect of starting steroids after the loss of ambulation
- European Neuro Muscular centre (ENMC) (http://www.enmc.org/workshops/reports.cfm?p=157)

Late Ambulatory Phase, 7~10 years

- Contracted joint can limit walking, even if overall limb musculature has sufficient strength.
- **Passive stretching** and the use of ankle-foot-orthoses (AFOs) worn at night are the mainstay of treatment in the ambulant stage to delay development of contractures.
- Daytime AFOs are not recommended as they may impede walking ability.
- Active exercise particularly in the **hydrotherapy pool** is recommended.
- Prolonging ambulation by use of **steroids and surgical intervention** for release of fibrotic contractures

Late Ambulatory Phase

- The maintenance of **good ranges of movement** and bilateral **symmetry** are important to allow optimum movement and functional positioning, to maintain ambulation, prevent development of fixed deformities, and maintain skin integrity.

- During both the ambulatory and non-ambulatory phases, **regular stretching** at the ankle, knee, and hip is necessary.

- ROM exercise, Postural Training, Passive Stretching, Active Ex.

Manual wheel chair stage, 10~13 years

- In Emery's work, **99th** percentile for loss of ambulation in DMD is age at **13.2** years

- Emery AE. Duchenne's muscular dystrophy. In: Oxford Monographs on Medical Genetics Series #24. 2nd ed. Oxford, United Kingdom: Oxford University Press;. 1993.

Manual wheel chair stage

- Manual wheelchairs are useful for the conservation of energy where walking long distances is tiring in DMD.
- **Monitoring** for the development of **scoliosis** should begin before the loss of ambulation.
- Special seating and a head rest should be introduced early before there are detrimental postural adaptations.
- Non-ambulant children should be provided with **sitting AFOs** and **passive or active assisted exercise** should be continued for comfort, aesthetics and contracture prevention

Manual wheel chair stage

- **Stretching** and **positioning**
- Active-assisted, and/or passive stretching to prevent or minimise contractures should be done a minimum of **4–6 days per week**
- During the non-ambulatory phase, regular stretching of the upper extremities, including the long finger flexors and wrist, elbow, and shoulder joints, also becomes necessary.

Manual wheel chair stage

- **Orthoses** : Resting ankle–foot orthoses (AFOs) used at night. Knee–ankle–foot orthoses (KAFOs) allow standing and limited ambulation for therapeutic purposes. Use of AFOs during the daytime can be appropriate for full-time wheelchair users.

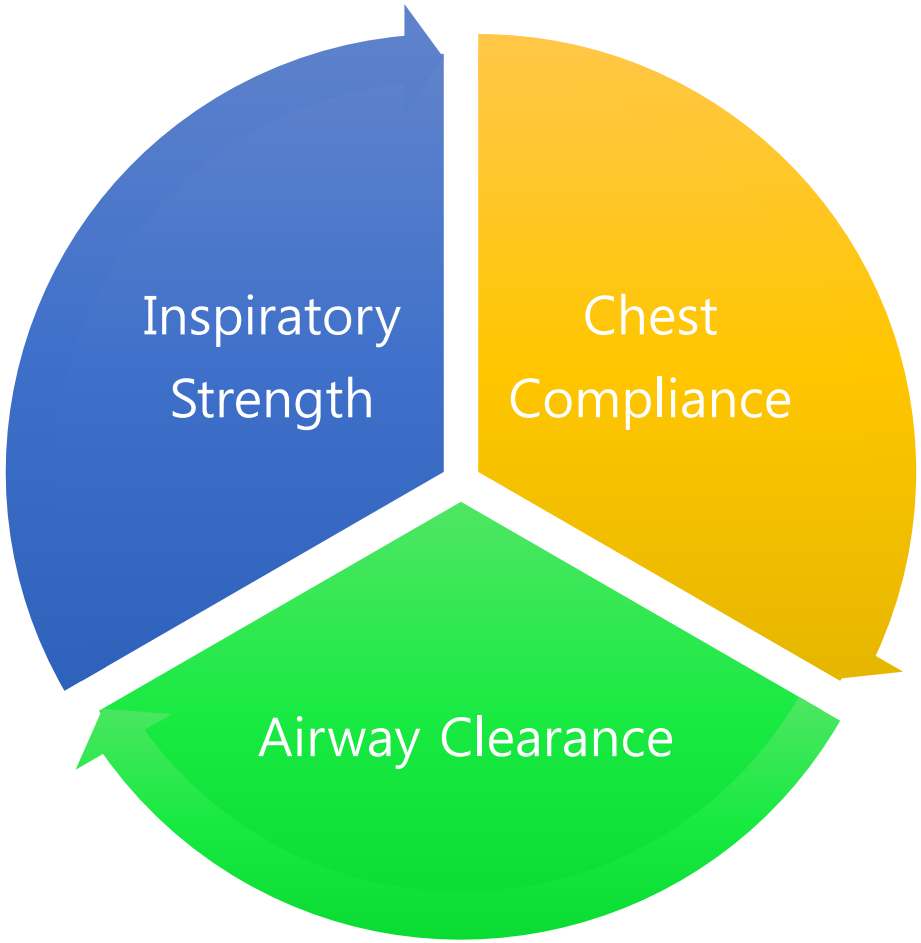
- **Standing devices** : A passive standing device for patients with either no or mild hip, knee, or ankle contractures is necessary for late ambulatory and early non-ambulatory stages.

Manual wheel chair stage

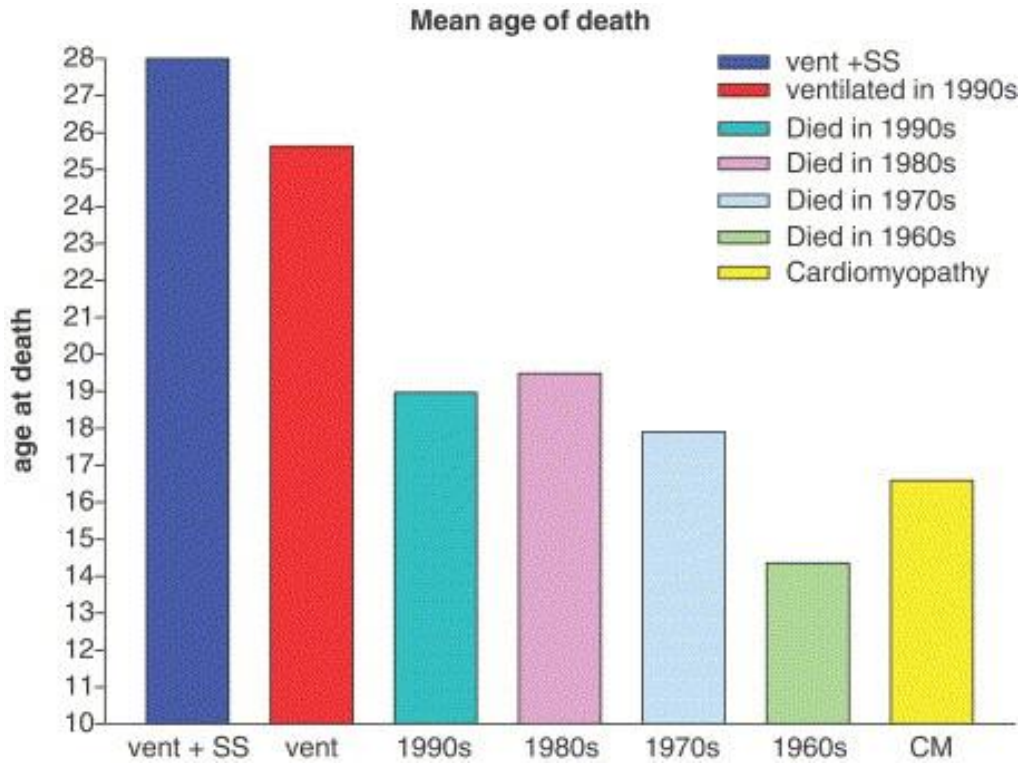
- The respiratory problems in DMD tend to be very predictable and correlate with overall muscle strength so that children who lose ambulation early are likely to **require ventilation sooner** than those who walk longer.

- Essentially respiratory function in ambulant boys is normal and problems relating to respiratory impairment are not usually seen until after the loss of independent ambulation.

Respiratory Management



Respiratory management in DMD



The change in life expectancy in DMD for boys dying in the decades since the 1960s. The yellow column represents the boys dying from an early and prominent cardiomyopathy, the red column the boys who were ventilated in the 1990s and the dark blue column the boys who had both spinal surgery and subsequently were ventilated.
(Current Paediatrics (2005) 15, 292–300. in Newcastle center)

Electrical wheel chair stage

- Over 14~18 years in DMD
- **Tilt-in-space electric wheelchairs** with supportive seating should be supplied early to avoid postural contractures and poor sitting posture.

Electrical wheel chair stage

- The mean age at institution of elective ventilation is around **17** years.
- **Chest physiotherapy** such as postural drainage and assisted coughing should be taught when coughing is ineffective and may need to be supplemented with cough aids such as the **in/exsufflator**.
- Once FVC drops to **1.25 ℓ** or **<40%** predicted value, then serial measurement of overnight oxymetry allows the recognition of the development of nocturnal respiratory failure. **symptomatic nocturnal hypoventilation** is an indication for elective **non-invasive** nocturnal **ventilation**.

Bed-Ridden stage

- During cardiopulmonary compromise
- Duchenne MD is a terminal disease in which death usually occurs by the **third decade** of life.
- Becker MD is similar to Duchenne MD, but features occur later and are more mild. Patients tend to live past the fourth or fifth decades.

- Dubowitz V. Muscle Disorders in Childhood. 2nd ed. Philadelphia, Pa: WB Saunders. 1995: 34-132.

Cardiovascular management

- 20–30% of DMD boys have left ventricular impairment on echocardiography by age 10 years.
- Cardiac investigation (**echocardiogram and ECG**) is indicated at diagnosis, **every 2 years** thereafter to age 10 and then annually, or more often.
- For the prevent of **left ventricular dysfunction**
- ACE-inhibitor and beta-blocker seem routine prescription of medication

Recommended Exercises in Muscle Disease

Exercise purpose	Examples	Notes
improve endurance and conditioning	walking, running, swimming, cycling	• recommended for most people with neuromuscular diseases who choose to adopt an exercise regimen, with intensity and duration depending on ability and cardiac function • walking or running downhill not recommended • not to be done to exhaustion
build strength and muscle mass	weight lifting, working against resistance with equipment such as stretchable bands	• Lifting light weights and working against light resistance are usually OK for most people with neuromuscular disease, unless in acute phase of an inflammatory muscle disease; stop at pain and before exhaustion
increase agility, coordination and balance	wheelchair sports, some video games, balance ball, horseback riding	• OK for most people with neuromuscular disease
maintain or increase flexibility	active or passive range-of-motion exercise; gentle stretching	• OK for most people with neuromuscular disease

Effect of Exercise on Different Muscle Diseases

Disease category	Nerve or muscle problem	Implications
Muscular Dystrophies Duchenne, Becker, congenital, distal, Emery-Dreifuss, facioscapulohumeral, limb-gridle, myotonic, oculopharyngeal	These are degenerative muscle diseases, meaning the normal ability of muscle to regenerate and repair damage is limited, resulting in a net loss of muscle tissue over time. Some (BMD, DMD and some LGMD forms) involve fragile membranes around skeletal and/or cardiac muscle fibers. Many muscular dystrophies (DMD, BMD, some LGMD, EDMD, myotonic MD) involve cardiac muscle deterioration or abnormal heart rhythms.	• Muscle fibers can be damaged by strenuous exercise, and the damaged sustained may be permanent. • In DMD, BMD and the sarcoglycan-deficient LGMDs, exercise involving eccentric (lengthening) contractions is particularly damaging. • If the heart's pumping ability or rhythm is affected by the disease, sudden, strenuous exercise could trigger an acute heart problem, respiratory problem or even death. A cardiologist should be consulted before undertaking an exercise program. Warning signs of an acute cardiac problem are chest pain, shortness of breath, nausea, sweating without strenuous exercise, a feeling of fullness or pressure in the chest, or a gurgling sound during breathing (a sign that fluid may be backing up into the lungs).
Motor Neuron Diseases Amyotrophic lateral sclerosis, spinal muscular atrophy (all types), spinal-bulbar muscular atrophy	Nerve cells in the central nervous system (spinal cord and brain) that control muscle movement are lost, leaving muscles "orphaned " and undernourished.	• Theoretically, remaining nerve cells can become overburdened if too much is required of them with exercise.

Effect of Exercise on Different Muscle Diseases

Inflammatory Myopathies Dermatomyositis, polymyositis	Muscle fibers are attacked by the immune system, causing inflammation and tissue destruction.	• During severe disease activity, when strength is very poor, assisted or passive range-of motion exercise can be undertaken. • During periods of mild to moderate disease activity, when muscles can work against gravity, light aerobic exercise, isometric strengthening, and active range-of-motion exercise are recommended. • When the disease is inactive (in remission), recreational aerobic and progressive resistance exercise are OK.
Diseases of the Neuromuscular Junction Myasthenia gravis	Communication between nerve fibers and muscle fibers is compromised, in most cases by a mistaken attack on the neuromuscular junction by the immune system.	• Exercise can be undertaken when the disease is under good control. In periods of acute disease exacerbation, or when the disease is not well controlled, exercise may increase weakness. • Exhaustion, lasting joint or muscle pain, or shortness of breath means the exercise is too strenuous.
Peripheral Nerve Diseases Charcot-Marie-Tooth disease, Dejerine-Sottas disease, Friedreich's ataxia	In CMT, and DSS, nerve fibers and/or the insulation around them are abnormal, compromising communication to muscle fibers. In FA, an abnormal cellular distribution of iron leads to diminished cellular energy production, with damage to the heart and nerves	• Exercise is good for the heart and lungs but will not overcome muscle wasting, which is the result of damage to nerve fibers and loss of input from the nervous system. • Low-impact exercise, such as swimming or biking, or yoga, is better than high-impact exercise. • Moderate exercise may help slow the loss of motor skills in FA and help offset weight gain. • Cardiac precautions apply. No exercise should be undertaken without consulting a cardiologist.

Exercise strategies for strength maintenance

- Limited research
- Submaximum, aerobic exercise/activity is recommended
- Avoid overexertion and pain : High-resistance strength training and eccentric exercise are inappropriate → induce fiber injury

Exercise strategies for strength maintenance

- To avoid disuse atrophy and other secondary complications of inactivity,
- To participate in regular submaximum (gentle) functional strengthening/activity (swimming-pool exercises and recreation-based exercises in the community)
- Swimming, which might have benefits for aerobic conditioning and respiratory exercise, is highly recommended from the early ambulatory to early non-ambulatory phases and could be continued in the non-ambulatory phase as long as it is medically safe.

Exercise strategies for strength maintenance

- Additional benefits might be provided by low-resistance strength training and optimization of upper body function.

- Significant muscle pain or myoglobinuria in the 24-h period after a specific activity is a sign of overexertion and contraction-induced injury, and if this occurs the activity should be modified.

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- Bushby, Katherine, et al. "The multidisciplinary management of Duchenne muscular dystrophy." Current Paediatrics 15.4 (2005): 292-300.
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- http://www.mayoclinic.org/diseases-conditions/muscular-dystrophy/basics/symptoms/con-20021240