

# Physiotherapy for children with Neuromuscular Disorders

Special considerations for therapists,  
schools and parents,  
recommendations and latest research.

Chiara Tewierik, Neurosciences Physiotherapist,  
The Royal Childrens Hospital Melbourne, 2008

# Objectives

1. To explain the role of the Neuromuscular clinic at the RCH, and the role of Physiotherapy in this clinic.
2. To outline the Physiotherapy role in the management of Neuromuscular Disorders, and the supporting research.

# Objectives

3. To provide information on how to access Physiotherapy at school, funding, and planning for a child with a Neuromuscular disorder at school.
4. To provide contact details for any enquiries regarding Physiotherapy for a child with NMD

# The History

- Physiotherapists have been involved in the care of the neuromuscular community at RCH for many years.
- The service begins when a child is referred by the consultant to the physiotherapist for monitoring, and plays a role across the lifespan of the child during their involvement with the hospital.
- Previously these children were seen in the Physiotherapy Department, and now are being seen in the Neuromuscular clinic in the multidisciplinary setting.

# Where RCH Physiotherapists fit into the big picture

The ideal Physiotherapy management of children with Neuromuscular Disorders is a combined effort between children, their families and community physiotherapists at home or school.

The Physiotherapist at the RCH will offer support to this process, and communicate any recommendations or management changes that result from the Neuromuscular clinic visit.

# What happens at Clinic?

- Children are seen by all the medical specialists at one place, in one day
- When they visit Physiotherapy, the child will be assessed on repeated objective measures, any issues will be identified with families, exercises and stretches may be prescribed
- If the child is enrolled in any of the studies, the study testing will replace the Physiotherapy assessment for that clinic visit.

# What happens at NMD Clinic?

- NMD clinic will also be where many decisions about the ongoing management of a child's disease will occur, such as implementation of steroids, waitlisting for orthopaedic surgery, or trialling BiPAP for respiratory support.
- If you have any concerns or queries regarding any of these decisions, please contact the professionals involved in the clinic, either by phone, letter or email.

# The Role of the Physiotherapist in the NMD clinic at RCH

- Monitor and maintain range of movement of joints, muscles, and postural changes
- Monitor and maintain functional activities
- Education about the Physiotherapy Respiratory management and training around use of equipment

# The Role of the Physiotherapist in the NMD clinic at RCH

- Liaison with families, schools and community therapists re best management of any physical issues with exercise, modifications, or equipment
- Liaison with medical staff in the clinic re child's condition as required
- Liaison with acute therapy team re child's management if hospitalised at RCH

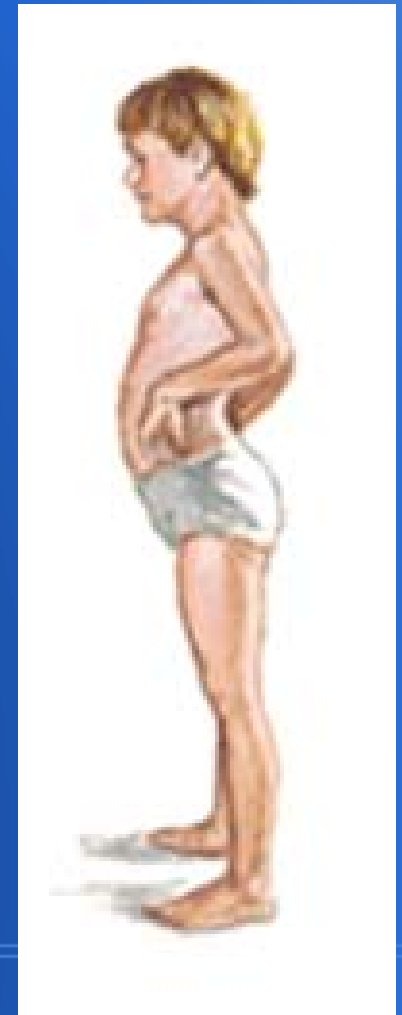
# Objective Measures monitored in NMD clinic

- Range of Movement of joints and muscle length
- Timed Functional activities
- Functional activity scales
- Balance
- Respiratory function, along with Pulmonary function testing

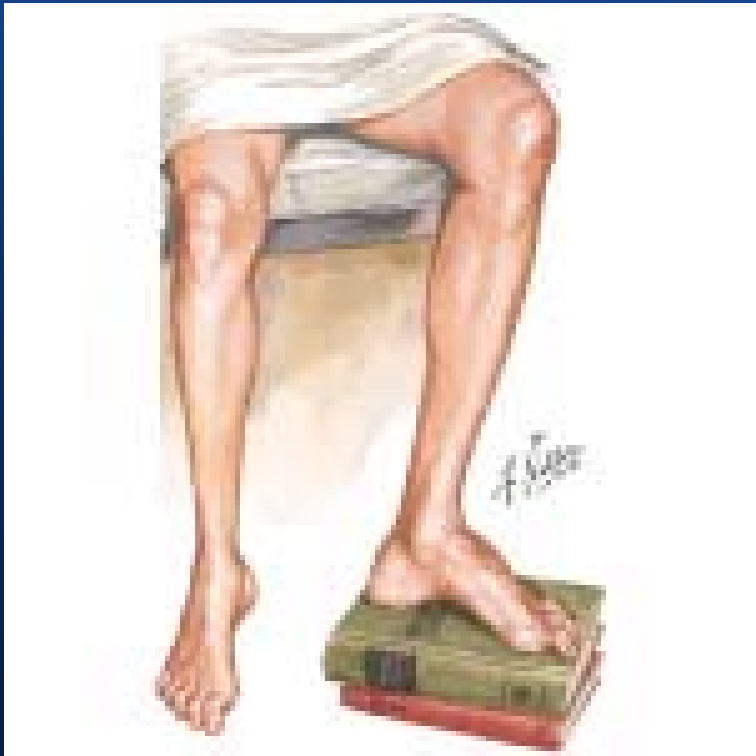
# Range of movement of joints and muscle length

For Muscular Dystrophies, muscles that are most often measured include:

- Hip flexors
- Hip abductors
- Hamstrings
- Calves
- Elbow flexors
- Wrist flexors
- These muscles are commonly tight for this population.

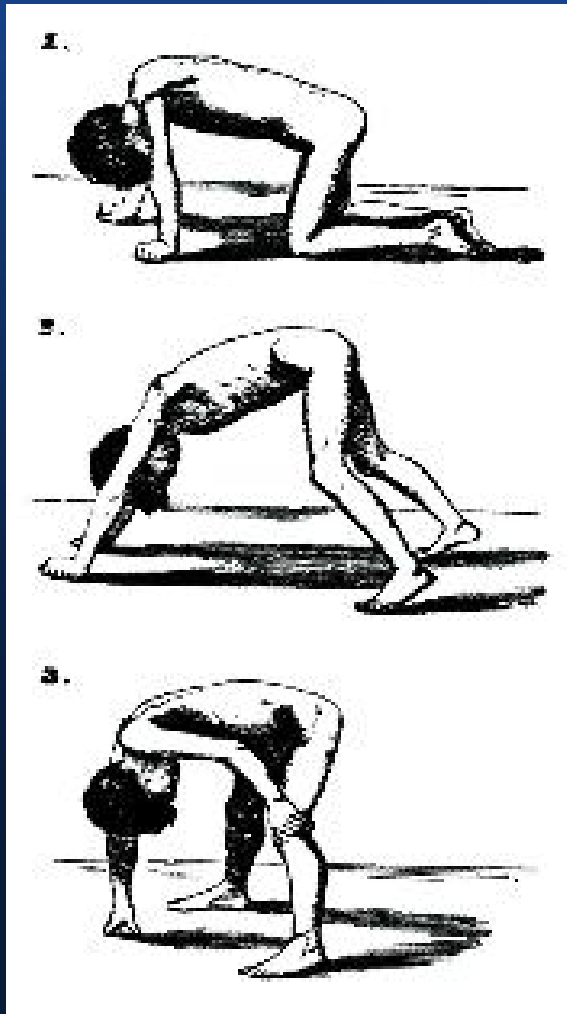


# Range of movement of joints and muscle length 2



- For CMT:
  - -as for muscular dystrophies, also ankle everters and inverters, and grip strength.
  - -Photographs taken for the Foot Posture Index.
- For SMA:
  - -Range of movement and muscle length less problematic, so are not routinely assessed.

# • Timed Functional activities



- Assessment of tasks that are important for everyday life completed for those who are able:
- Timed 10 metre walk/run
- Timed supine to stand – standing up from lying on the floor
- Timed climbing 4 stairs

# • **Functional activity scales**

- These scales group activities together that are most likely to be effected in specific Neuromuscular disorders.
- The tasks may include activities such as sitting, rolling, walking, jumping, hopping, and standing on one leg.

# • **Functional activity scales**

Some examples used in the clinic are:

- Hammersmith Motor Ability Scale
- Motor Function Measure
- HiMAT
- North Star ambulatory Assessment
- Egan Klassification scale

# Functional scales in Neuromuscular Disorders

- In 2007 a workshop was had with the Treat NMD clinicians to develop consensus of the types and procedures for the outcome measures to be used in the monitoring and associated research for children with DMD and SMA. (Mercuri et al 2008)
- The measures used in the Neuromuscular clinic at the RCH comply with these recommendations.

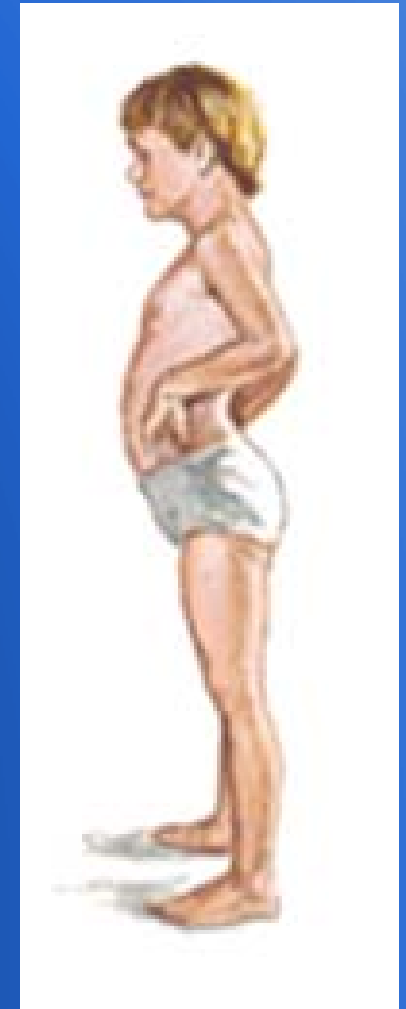
# Physiotherapy Exercises

Your Physiotherapist will identify with you any issues that need to be addressed. This may include the prescription of exercises:

- specific stretches for tight muscles
- strengthening exercises when appropriate
- respiratory exercises
- activities to promote health, function and participation

# Stretches

- Tight muscles often occur with children who have Neuromuscular disorders. When muscles become very tight, joints also get stiff and can become contracted.
- Your physiotherapist will prescribe specific stretching to help keep muscles long, so that children are able to move easily and to try to limit contractures.



# Stretches

- Stretches are most often for hip flexors, hip abductors, hamstrings and calves.



- Night splinting can be used to help with calf stretches
- Use of standing frames may also help with calf stretching in children who are unable to walk.

# Research on range of movement and muscle length

- Maintenance of calf length in DMD through use of night splinting and passive stretching is better than passive stretching alone. (Hyde et al 2000, Bushby et al 2005, TREAT\_NMD Recommendations accessed 2008)
- Newer studies are trialing other techniques such as serial casting, but evidence is not yet substantial enough to trial in our population. (Main, et al 2007; Glanzman et al 2008)

# Strengthening

Strengthening is important in all neuromuscular disorders, it is the type of strengthening that differs.

# Strengthening

- For muscular dystrophies:
  - - Strengthening should be low to medium intensity, with a focus on function, but most of the evidence is not very strong as it is limited to small numbers of children. (Ansved 2003).
  - - The evidence still recommends that high resistance exercise may be harmful in this population. (Sayers 2000).

# Strengthening

- For CMT:

- Strengthening in children should be functional, although progressive resistance exercise can also be effective when the child is old enough to participate, (Chetlin et al 2004)

- For SMA:

- Strengthening is more focussed on developing functional skills, such as rolling, sitting and transitions if able.

# Respiratory Function in NMD

Reduced respiratory muscle strength in muscular dystrophies and SMA can result in:

- Susceptibility to infection
- Reduced ability to cough and clear secretions
- Difficulty in taking deep breaths to keep lungs inflated
- Breathing fatigue and hypoventilation
- (Finder et al 2004, Wang et al 2007)

# Respiratory Physiotherapy at home

Your Physiotherapist may prescribe the following to help with maintenance of a healthy respiratory system:

- Bubble PEP – to assist with maintenance of lung volumes and secretion removal
- Manual Assisted Coughing – physical assistance with co-ordinated pressure applied to the external chest wall to increase force of cough.

# Respiratory Physiotherapy when in hospital

- If you become an inpatient at the RCH for impaired respiratory function, the following supplementary equipment may be used:
  - BiPAP machine – to help take bigger breaths to help shift secretions
  - In/exsufflation (Cough Assist machine) – to help increase efficiency of cough.
  - Suction – to aid removal of secretions from mouth

# Scoliosis management

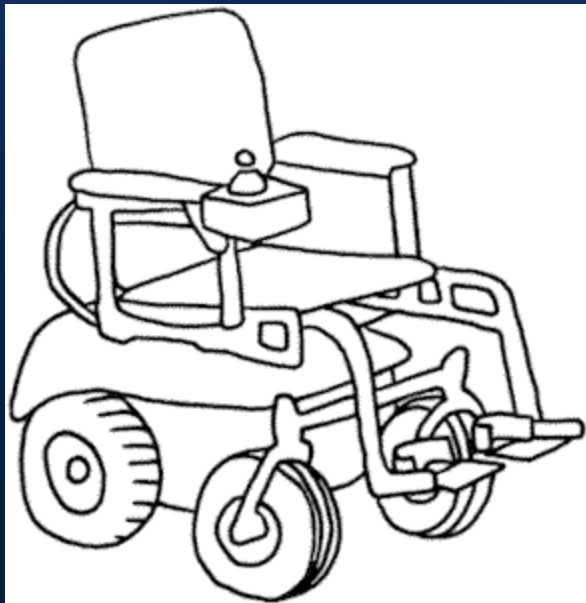
Scoliosis development can occur due to muscle weakness causing muscle imbalance.

“Scoliosis can lead to compromised seating and trunk balance, discomfort, pain and difficult attendant care, (and) exacerbation of any underlying cardio-respiratory dysfunction” (Kinali et al 2007)



# Scoliosis management

- Physiotherapists assist scoliosis management through stretching and postural controls
- Standing frames are used to aid upright posture
  - Specialised postural support in wheelchairs is important to maintain symmetry.
  - If a child requires surgery for management of their scoliosis, Respiratory Physiotherapy may be required to aid recovery.



# Activities to promote health, function and participation

All children are encouraged to participate in activities that they will enjoy and feel able to complete.

Physiotherapy can aid in playground activities and skills such as balance, jumping and climbing, as well as other gross motor skill development.

Swimming and hydrotherapy and tai chi are helpful, low impact activities for strength and range of movement in Neuromuscular disorders.



# Fatigue

- Fatigue is often a very limiting factor with children who have neuromuscular disorders.
- Children often need to pace their daily activities, and this may need to be guided by teachers and parents to ensure that children can participate as much as possible.
- Wheelchairs are often prescribed early, before children have trouble with their walking, so that they can use wheelchairs to limit fatigue.

# Wheelchairs

- Manual wheelchairs are used when children are experiencing fatigue, or for long distances.
- Electric Wheelchairs are important when children are unable to walk safely, so they have the independence to move around.
- At the RCH, Physiotherapy prescribe wheelchairs for children. In the community wheelchairs may be prescribed by either Physiotherapists or Occupational Therapists.

# Wheelchairs

- Wheelchairs for children with NMD are prescribed and maintained by community therapists, who have regular contact with the children.
- Physiotherapists at the RCH may assist in this process if required, but in close consultation with the community team.
- Wheelchairs need to be considered in advance, as there is a long waiting time between organising a wheelchair and actually receiving it.

# Families and Exercise

- In a study of families of children with a chronic medical condition, 66% of families report some non-compliance in physiotherapy home exercise program. (Rone-Adams, Stern and Walker, 2004)
- Also, the more stressed the family, the more likely it was that they would not complete their home exercise program.

# Families and Exercise

- Any exercise that is prescribed will be modified to ensure families and children are able to complete regularly.
- These are regularly reviewed and changed to ensure the burden to families is kept minimal.

# Community Physiotherapists

- Many children will have a community Physiotherapist, either through Early Intervention (0-school age), at school, or in a local Community Health Service or privately.
- The Physiotherapist at RCH will initially refer children to a community therapist, and will liaise with this therapist about the management plan as developed in the Neuromuscular clinic, and offer any support required with the management of this complex set of disorders

# Physiotherapy in the School Environment

School aged children with Neuromuscular Disorders will often receive their primary Physiotherapy service through their school.

# The role of Physiotherapy at School

- To assist children to access their school environment and curriculum, Physiotherapists consider the following:
  - Safety – in classroom and playground
  - Fatigue – in walking, climbing stairs, on excursions
  - Education of teachers and aides to the physical needs of the child at school
  - The potential for progression of the disorder, and the need to put plans in in advance

# Physiotherapy at School

- Often Physiotherapy review is needed once per term to monitor school program and environment to achieve these goals. If there is a change in a child's condition, increased frequency may be indicated.
- Integration aids are often taught stretching programs to assist maintenance of mobility and therefore safety at school.
- Children who require regular active therapy sessions may see a Physiotherapist at school more frequently.

# How to access therapy at school

- Schools in Victoria are required to make 'reasonable adjustments' to assist a student to participate in school and to use its facilities and services.
- This mandate is regardless of whether or not the child has received funding from the Department of Education and Childhood Development (DEECD) for integration.
- Access to therapy services at school fits under these 'reasonable adjustments'.

# Funding for Physiotherapy at school

- Most schools will apply for integration funding to support a child with a Neuromuscular disorder at school, and will most often apply for this funding based on Physical Disability.
- Funding is also available for all types of schools, including pre-school.
- This is the source of funding to provide Physiotherapy for the child at school, to be able to access all educational opportunities.

# Integration Funding

- To access Integration funding, an educational needs questionnaire needs to be completed between May and July for the following year.
- Most often the Physiotherapist is required to provide supportive documentation to fill out this form.
- For new school starters applications are accepted later in the year, but it pays to be organised about the school your child will attend.

# How to find Physiotherapy for school

- Most schools access Physiotherapy through SCOPE or Yooralla services. Other private Physiotherapists may also be available. If you are having difficulty sourcing a Physiotherapist please contact the RCH.
- If you need assistance on how to access therapy at school, information available from Ed institute at RCH website – [www.rch.org.au/edinst.OTphysio](http://www.rch.org.au/edinst.OTphysio)

# Preplanning for NMD at school

- Information on the condition accessible through the MDA
- Information on supporting the child to access education and educational supports through the Education Institute at the RCH
- Liaise with treating team to ascertain need for integration funding

# Preplanning for NMD at school

- Apply for integration funding for aide time, as well as for Physiotherapy and Occupational Therapy to ensure an environment that will allow access to the curriculum
- Consider environment for teaching, does the child need to climb stairs to get to classroom?

# Contact Details

Chiara Tewierik

Physiotherapy Department,  
The Royal Childrens Hospital, Melbourne  
Ph: 9345 5411, pager 5406  
[chiara.tewierik@rch.org.au](mailto:chiara.tewierik@rch.org.au)

# References 1

- (2008) Standards of care for Duchenne muscular dystrophy, Brief TREAT-NMD recommendations. Accessed from [http://www.parentprojectmd.org/site/DocServer/TREAT-NMD\\_DMD\\_interim\\_recommendations.pdf?docID=3781-NMD\\_DMD\\_interim\\_recommendations.pdf?docID=3781](http://www.parentprojectmd.org/site/DocServer/TREAT-NMD_DMD_interim_recommendations.pdf?docID=3781-NMD_DMD_interim_recommendations.pdf?docID=3781), October 2008.
- Ansved T. (2003) 'Muscular Dystrophies: influence of physical conditioning on the disease evolution'. Current opinion in Clinical Nutrition and Metabolic Care. 6(4):435-9
- Bushby, K., Bourke, J., Bullock, R., Eagle, M., Gibson, M., Quinby, J. 'The multidisciplinary management of DMD' Current Paediatrics 15, 292–300
- Chetlin, R., Gutmann, L., Tarnopolsky, M., Ullrich, I., yeater, R. (2004) 'Resistance Training Effectiveness in Patients With Charcot-Marie-Tooth Disease: Recommendations for Exercise Prescription' Archives of Physical Medicine Rehabilitation Vol 85, August p1217 - 1223.
- Glanzman, AM., Flickinger, JM., Dholakia, KD., Bonneman CG, Finkel RS. (2008) 'Serial casting in children with DMD'. Pediatric Physical Therapy. 20(1); 106-7.

# References 2

- Hyde S.A., Filytrup, I., Glent, S., Krosmark, A.K., Salling, B., Steffensen, BF., Werlauf, U., Erlandsen, M. (2000) 'A randomised comparative study of two methods for controlling tendo achilles contracture in DMD'. Neuromuscular Disorders. 10:257-63
- Finder, JD., Birnkrant, D., Carl, J., Farber, HJ., Gozal, D., Iannaccone T., Koveski, T., Kravitz, RM., Panitch, H., Schramm, C., Schroth, M., Sharma, G., Sievers, L., Silvestri, JM., Sterni, L., and the American Thoracic Society. (2004) 'Respiratory care of the patient with DMD: ATS consensus statement.' American Journal of Respiratory and Critical Care Medicine. 170 (4) 456-465.
- Kinali, M., Main, M., Mercuri, M., Muntoni, F. (2007) 'Evolution of abnormal postures in Duchenne muscular dystrophy' Annals of Indian Academy of Neurology 10 (Supplement 1):S44-54
- Main M., Mercuri, E., Haliloglu, G., Baker, R., Kinali, M., Muntoni, F. (2007) 'Serial casting of ankles in DMD: can it be an alternative to surgery?' Neuromuscular Disorders. 17(3):227-30

# References 3

- Mercuri, E., Mayhew, A., Muntoni, F. (2008) 'Towards Harmonisation of Outcome measures for DMD and SMA within TREAT-NMD; Report of three expert workshops...' Neuromuscular Disorders. 18, 894-903.
- Rone-Adams,S, Stern, D., Walker, V. (2004) 'Stress and Compliance with a Home Exercise Program Among Caregivers of Children with Disabilities' Pediatric Physical Therapy 16:140–148
- Sayers, SP., (2000) 'The role of exercise as a therapy for children with DMD.' Pediatric Exercise Science. 12:23-33
- Wang, CH., Finkel, RS., Bertinin, ES., Schroth, M., Simonds, A., Wong, B., Aloysius, A., Morrison, L., Main, M., Crawford, TO., Trela, A., and participants of the International Conference on SMA Standard of Care (2007) 'Consensus statement for standard of care in Spinal Muscular Atrophy' Journal of Child Neurology 22 (8): 1027 - 1049