HA Convention 2017
Master Class 4: Multi-disciplinary management of Neurometabolic Disorders

Physiotherapy Management for Children with Neurometabolic Disorders

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Neurometabolic Diseases

• Inheritable inborn error of metabolism
• Neurological involvement
• Individually rare but large number as a group
Considerations for Rehabilitation Planning

- Neurological and multiple organs system involvement
- Variations in onset, severity and progression
- Specific treatment, supportive care
Rehabilitation Model in DKCH

Multidisciplinary rehabilitation

- Multiple rehabilitation programs
- Community linkage
- Multispecialty involvement

Child
Physiotherapy Role and Intervention

- Developmental training
- Neuromotor rehabilitation
- Movement disorders and spasticity management
- Musculoskeletal rehabilitation
- Pain management
- Pulmonary rehabilitation
- Ventilator Care

- Adaptive mobility aids
- Care givers support
- Community program

Assessment and Monitoring
ICF Model

Training and Intervention

Education

Community Linkage
## Motor Training: Infant Stage

<table>
<thead>
<tr>
<th>Developmental training</th>
<th>Sensory stimulation</th>
<th>Position changes</th>
<th>Care giver education and participation</th>
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</thead>
</table>

[Image of children engaged in motor training activities]
**Motor Training: Infant to Preschool**

<table>
<thead>
<tr>
<th>Ambulation</th>
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<tbody>
<tr>
<td>Functional exercise and play</td>
</tr>
<tr>
<td>Gross motor skills</td>
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<tr>
<td>Balance skills</td>
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</tbody>
</table>
Motor Training: Pre school to School Age

- Balance and coordination
- Strength and range
- Stretching program
- Aerobic capacity training
- Hydrotherapy
Mitochondrial Disease: Exercise Considerations

Fatigue and Exercise intolerance

To Exercise
## Mitochondrial Disease: Exercise Recommendation

Consensus Statement from Mitochondrial Medicine Society, *Genetics in Medicine* 2015 17(9): 689-701

<table>
<thead>
<tr>
<th>Exercise Recommendations</th>
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<tbody>
<tr>
<td>Close exercise supervision</td>
</tr>
<tr>
<td>Aerobic form of light physical exercise</td>
</tr>
<tr>
<td>Gradual and slow progression for endurance training</td>
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<tr>
<td>Prolong rest for resistance and strength training</td>
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**Fuel before and refuel after exercise**

**Pain, muscle soreness and cramps**
Exercise for Glycogen Storage Disease Type II with Enzyme Therapy


**DO’s**
- Sub-maximal aerobic exercise
- Appropriate rest
- Cardiopulmonary monitoring
- Energy conservation techniques, biomechanical advantages

**DON’T**
- Overwork with exercise fatigue
- Excessive resistance
- Eccentric exercise
- Disuse atrophy
Neurological involvement: Movement Disorders

Monitor changes for drug titration and therapy
Neuromotor rehabilitation for treatable diseases

<table>
<thead>
<tr>
<th>Fahn –Marsden Score for dystonia</th>
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<tbody>
<tr>
<td>EYES</td>
</tr>
<tr>
<td>SPEECH and SWALLOWING</td>
</tr>
<tr>
<td>RIGHT ARM</td>
</tr>
<tr>
<td>RIGHT LEG</td>
</tr>
<tr>
<td>TRUNK</td>
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</tbody>
</table>
Neurological involvement: Spasticity Management

- Stretching
- Positioning
- Splintage
- Instrumented Gait analysis
Spasticity Management

• Botox Injection Program

Pre Botox

Muscle tone
Muscle function
Functional assessment

Post Botox

Muscle training
Stretching program
Functional retraining
Intrathecal Baclofen Pump Insertion Program

**Neuromotor Joint Clinic**

- Neuromotor Joint clinic evaluation (DKCHI)
- ITB trial
  - Orthopaedic ward 56 admission
  - Pre-and-post trial PT Ax (protocol)
- Negative trial
  - Not suitable for ITB pump implantation
  - Consider other neuromotor rehabilitation intervention
  - Referral to MSW for application of funding support if necessary
- Positive trial
  - Suitable for ITB pump implantation
  - ITB pump implantation date
  - Ward 78 orthopaedic ward admission
  - Pre-op PT Ax (Protocol)
  - ITB Pump educational information to parents
- Discharge within 1 week
  - PT Ax before discharge (Protocol)
  - Discharge with letter* (ITB pump dose, withdrawal or overdose symptoms, urgent contact)
- Follow-up and ITB dose adjustment
  - Orthopaedic (Dr Wong YW clinic): every 2-4 weeks
  - Neuromotor joint clinic: every 4-5 mths for 1st year, then every 6 months
  - Pre-Neuromotor joint clinic PT update assessment (according to protocol)
Musculoskeletal Involvement: Posture and Pain Control

24 hours postural management:

**Day time:** seating and work table height

**Night time:** hip health, knee, ankle position

Multidisciplinary
Paediatric orthopaedic and
Seating clinic

Electrotherapy for pain control
Osteoporosis and Bone Health

- Complications and fracture risk
- **Weight bearing** exercise program
- **Low dose ultrasound** for fracture delay union
- Case reports using **vibration therapy**
Respiratory Involvement
Physiotherapy Pulmonary Rehabilitation Program

- Lung function test
- Aerobic exercise capacity assessment
- Airways clearance techniques
- Supervised exercise program
- Care givers education Home program
Multi-disciplinary Ventilator Care Program in DKCH

Ventilator Care Pathway

Ventilator-Assisted Children’s Voyage Allies & Companions

長隨呼吸機兒童護理計劃

為您打氣 伴您同途

Department of Physiotherapy  The Duchess of Kent Children’s Hospital at Sandy Bay
Community and School Support

- Integrate rehab program as school activities
- Mobility aids prescription
- Family support and education

Quality of Life
Thank You