

HA Convention 2017

Master Class 4: Multi-disciplinary management  
of Neurometabolic Disorders

# Physiotherapy Management for Children with Neurometabolic Disorders

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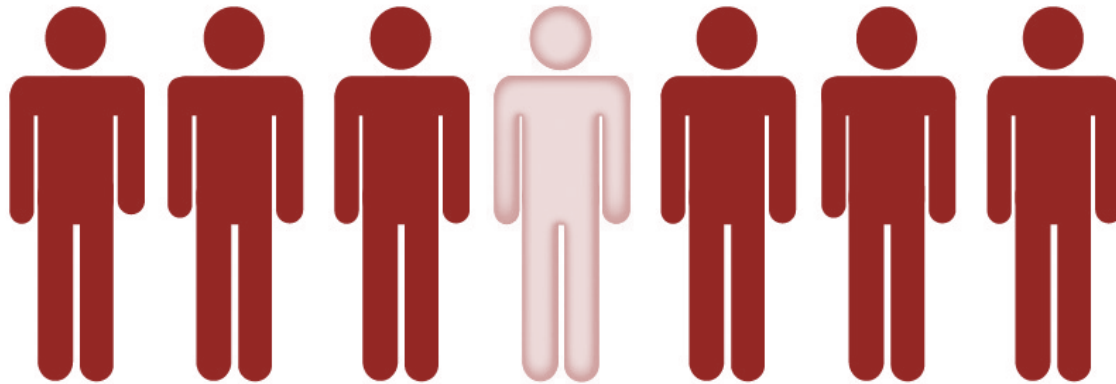
The Duchess of Kent Children's Hospital at Sandy Bay



# Neurometabolic Diseases

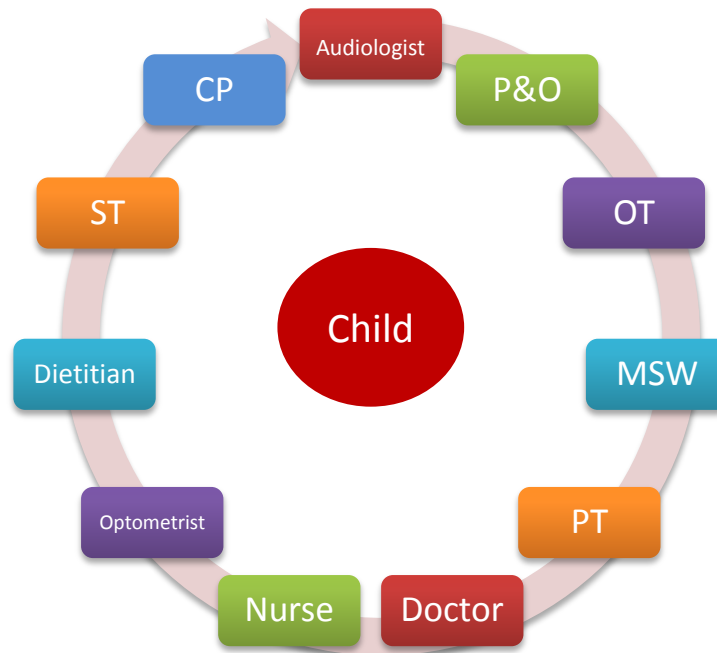
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- Inheritable inborn error of metabolism
- Neurological involvement
- Individually rare but large number as a group



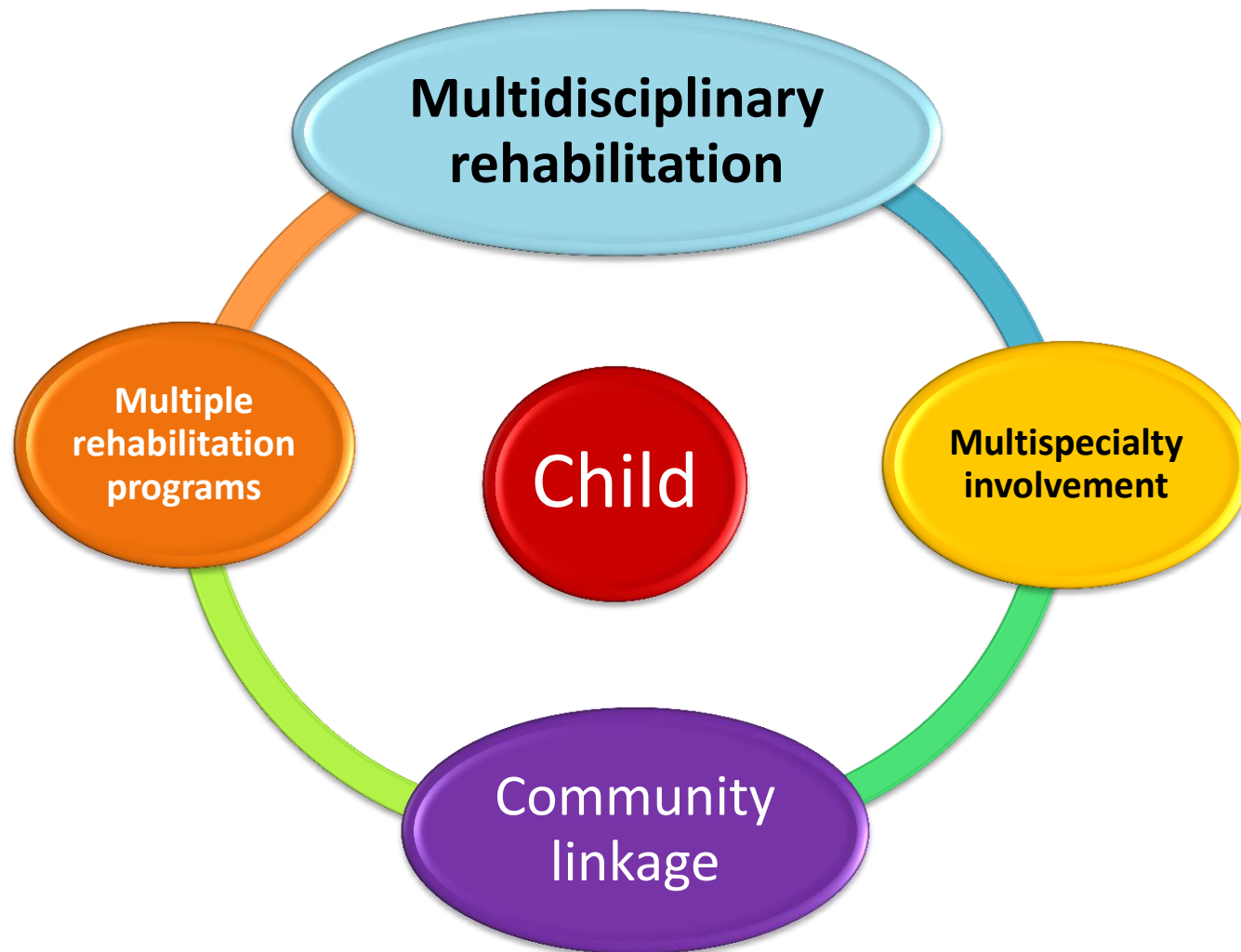
# Considerations for Rehabilitation Planning

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

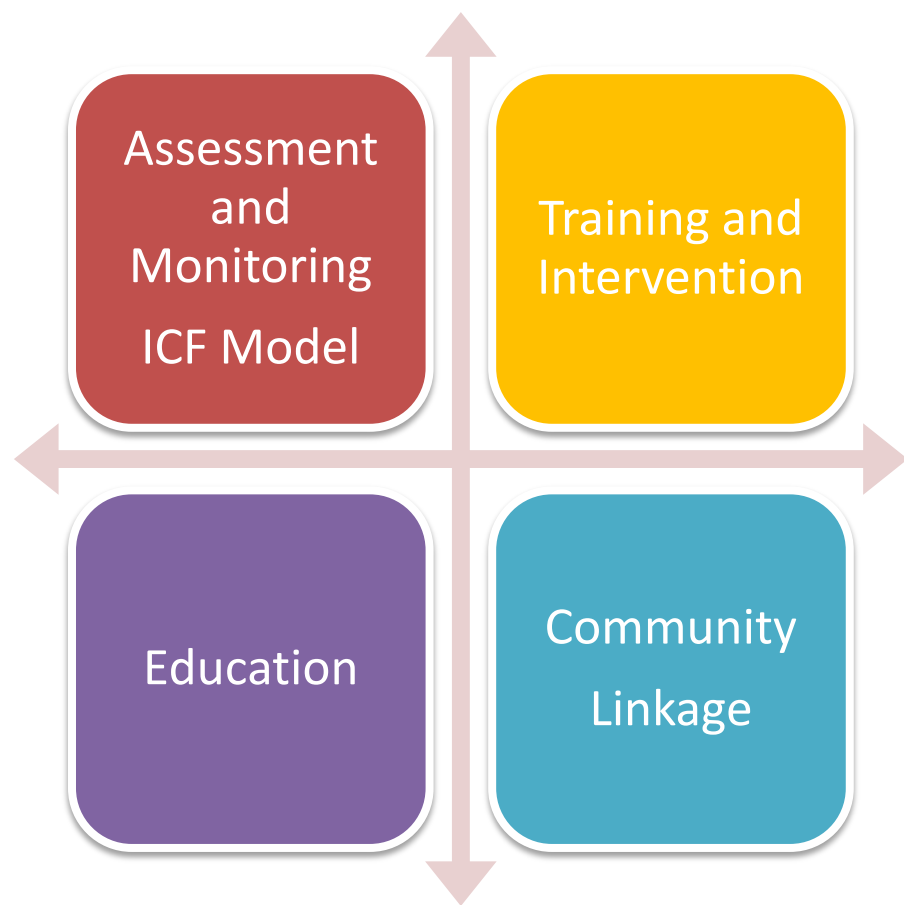


# Rehabilitation Model in DKCH

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# 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**



# Motor Training: Infant Stage

Developmental training

Sensory stimulation

Position changes

Care giver education and participation



# Motor Training: Infant to Preschool

Ambulation

Functional exercise and play

Gross motor skills

Balance skills



# Motor Training : Pre school to School Age

Balance and coordination

Strength and range

Stretching program

Aerobic capacity training

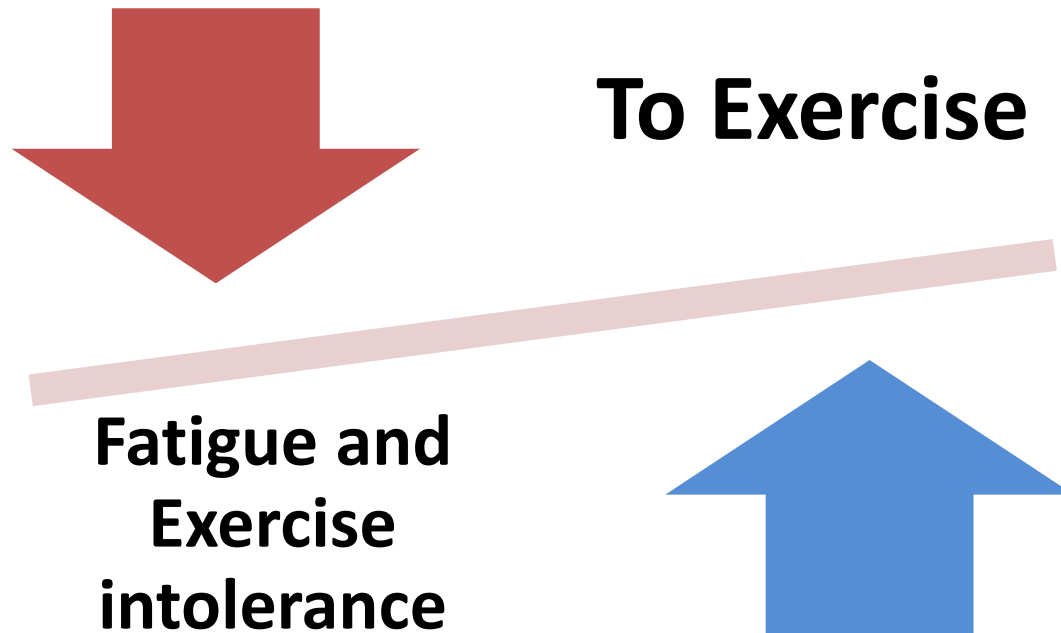
Hydrotherapy





# Mitochondrial Disease: Exercise Considerations

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# Mitochondrial Disease: Exercise Recommendation

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

- Close exercise supervision
- Aerobic form of light physical exercise
- Gradual and slow progression for endurance training
- Prolong rest for resistance and strength training

***Fuel before and refuel after exercise***



***Pain , muscle soreness and cramps***



# Exercise for Glycogen Storage Disease Type II with Enzyme Therapy

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Update on Exercise considerations *Am J Med Genet Part C 160C: 69-79 2012*

## 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

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

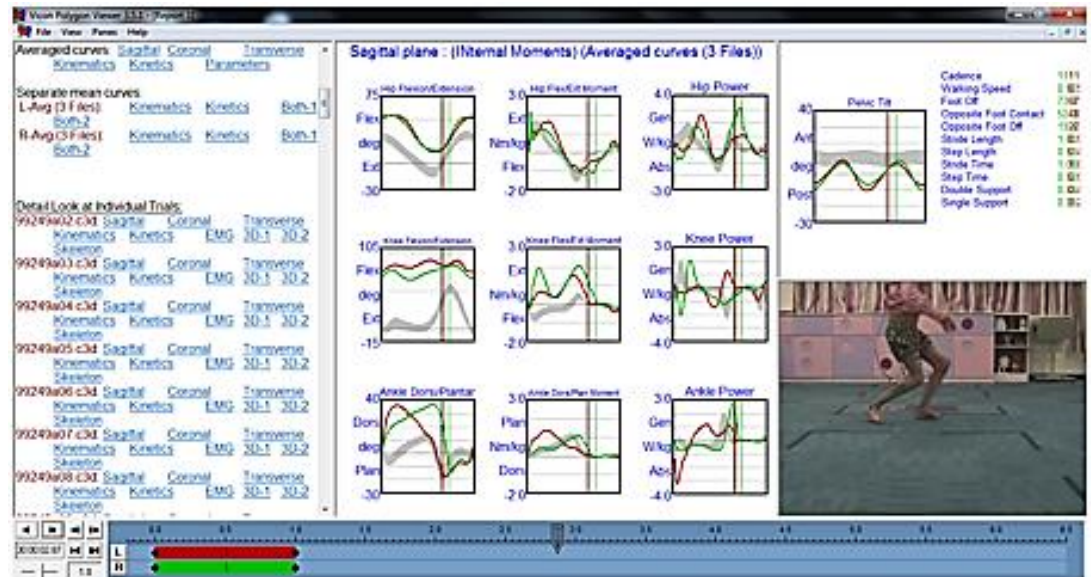
## Fahn –Marsden Score for dystonia

EYES	MOUTH
SPEECH and SWALLOWING	NECK
RIGHT ARM	LEFT ARM
RIGHT LEG	LEFT LEG
TRUNK	



# Neurological involvement: Spasticity Management

- Stretching
- Positioning
- Splintage
- Instrumented Gait analysis



# Spasticity Management

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- Botox Injection Program

Pre Botox

Muscle tone  
Muscle function  
Functional  
assessment

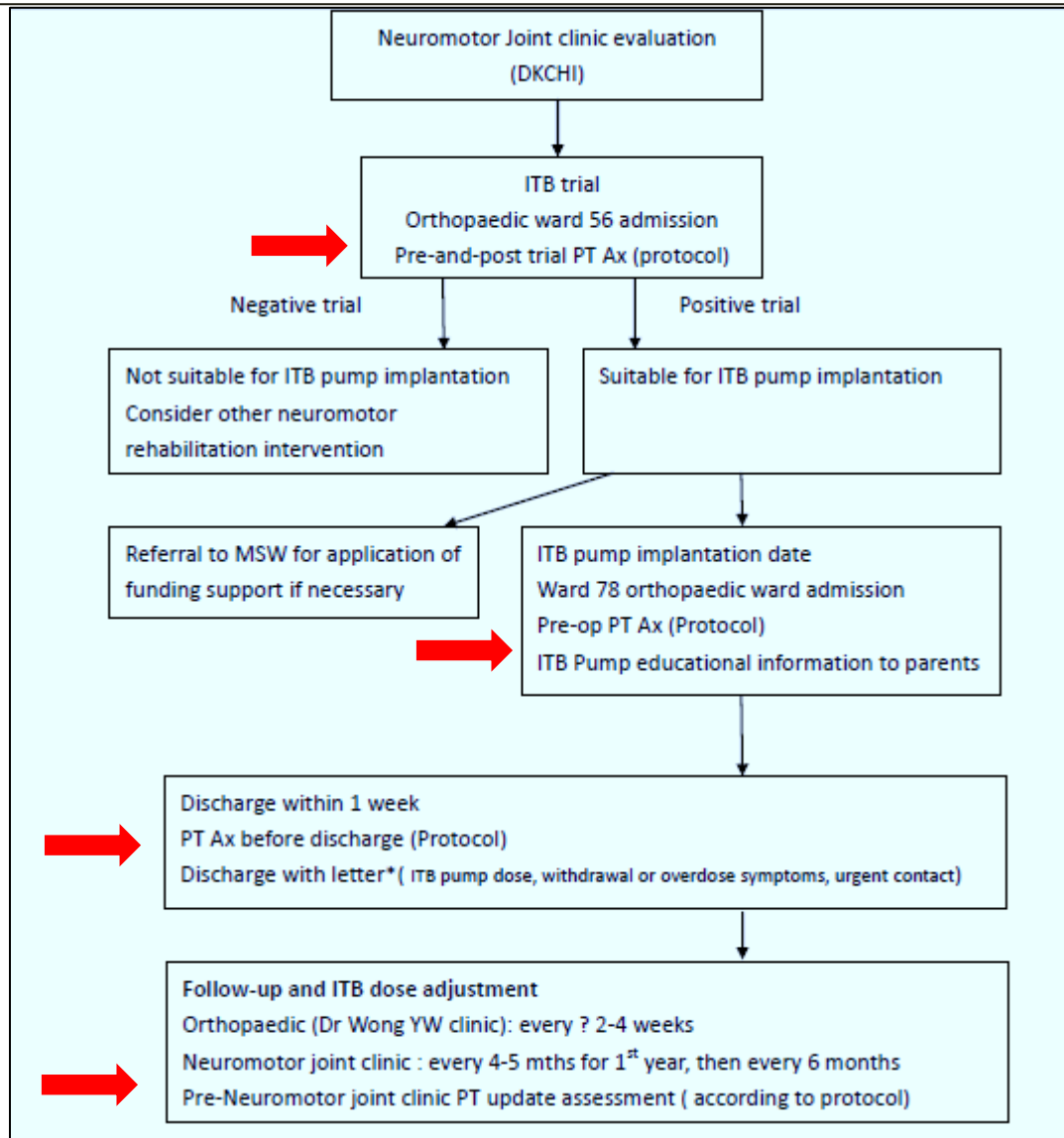
Post Botox

Muscle training  
Stretching program  
Functional retraining



# Neuromotor Joint Clinic

## Intrathecal Baclofen Pump Insertion Program



# Musculoskeletal Involvement: Posture and Pain Control

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## 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**



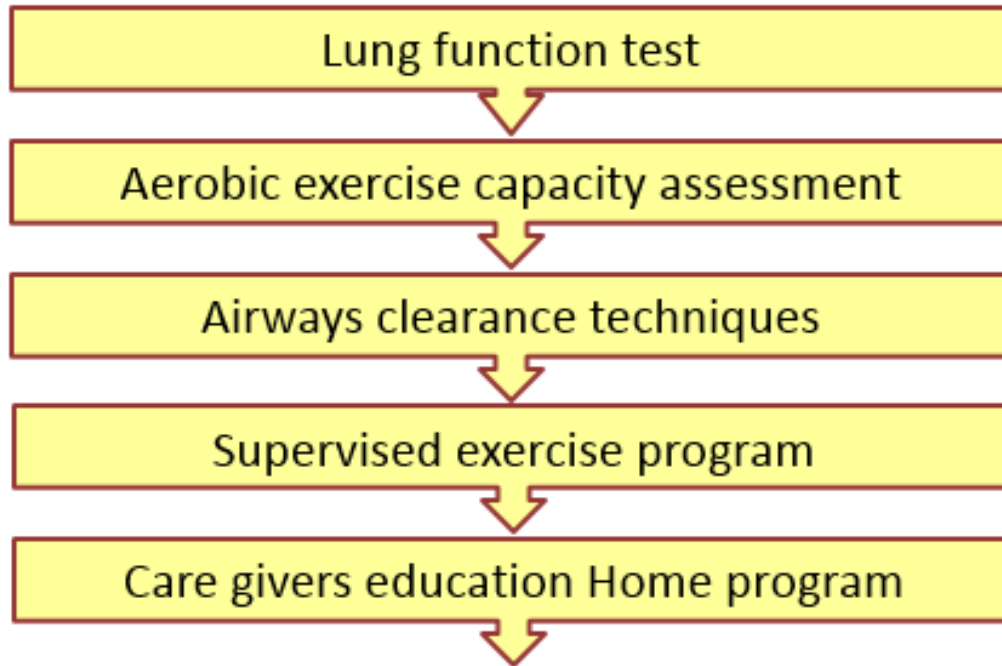


# Osteoporosis and Bone Health

- 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



# Multi-disciplinary Ventilator Care Program in DKCH



## Ventilator Care Pathway

The Duchess of Kent Children's Hospital at Sandy Bay  
Children's Habilitation Institute – Ward3/4

Gum Label

### Pathway of VAC patients

Week 0 – 2 <i>Orientation, multidisciplinary assessment and rehabilitation planning</i>	
Doctor (_____)	<input type="checkbox"/> Establish respiratory care plan <input type="checkbox"/> Referral to multidisciplinary therapists for assessment <input type="checkbox"/> Review all other medical conditions and management <input type="checkbox"/> Developmental assessment
Case Coordinator (_____)	<input type="checkbox"/> An initial family orientation in which the education and discharge processes are outlined <input type="checkbox"/> Provide DK education manuals <input type="checkbox"/> Schedule training sessions by nursing team / allied health teams <input type="checkbox"/> Refer caregivers to CPR course <input type="checkbox"/> Clarification of expectations of family regarding the change in care and management goals
Nursing (_____)	<input type="checkbox"/> Daily care / bath <input type="checkbox"/> Tracheostomy care training <input type="checkbox"/> Basic home ventilator training <input type="checkbox"/> Pulse oximeter training <input type="checkbox"/> Bedside emergency set up <input type="checkbox"/> Emergency Go – bag set up <input type="checkbox"/> Feeding assessment and growth chart
Allied Health – PT (_____)	<input type="checkbox"/> Pulmonary assessment ± lung function testing <input type="checkbox"/> Musculoskeletal assessment <input type="checkbox"/> Pain assessment <input type="checkbox"/> Developmental assessment <input type="checkbox"/> Transfer and mobility assessment <input type="checkbox"/> Chest physiotherapy <input type="checkbox"/> Caregiver training on suctioning <input type="checkbox"/> Developmental assessment
Allied Health – OT (_____)	<input type="checkbox"/> ADL assessment including oromotor & feeding, bed-mobility, grooming, dressing & toileting <input type="checkbox"/> Explore home environment (with MSW) and anticipate need of home modification and assistive device
Allied Health – ST (_____)	<input type="checkbox"/> Initial assessment on oromotor function and communicative ability <input type="checkbox"/> Developmental assessment
Allied Health –MSW (_____)	<input type="checkbox"/> Psychosocial assessment <input type="checkbox"/> Emotional support to enhance caregiver confidence <input type="checkbox"/> Financial and home environment assessment <input type="checkbox"/> Referral to DA /other funding agency when needed <input type="checkbox"/> Assess backup support /assist helper recruitment <input type="checkbox"/> Introduction to community resources / schooling support
Allied Health – CP (_____)	<input type="checkbox"/> Assess patient /family attitudes towards ventilator care; psychosocial support if necessary <input type="checkbox"/> Cognitive assessment
Allied Health - Audi & Opto	<input type="checkbox"/> Baseline assessment

Ward3/4, DKCH  
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# Community and School Support

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

**Quality of Life**



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# Thank You

