Current Practice

Cerebral palsy – understanding the disabilities and planning intervention

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Introduction

Cerebral palsy (CP) is an umbrella term covering a group of persistent, non-progressive, but often changing, disorders of movement and posture secondary to lesions or anomalies of the brain which occur in the early stages of development1. Although the brain lesion is static, the condition itself is progressive as a result of changes associated with nervous system maturation, growth and experience.

The global incidence of CP has remained constant around 2.5 per thousand live births. However, there have been changes in the epidemiological pattern and clinical signs[1]. Increasing survival of the extremely preterm infant (<28 weeks gestational age) has resulted in a changed clinical picture of CP.

The common pathology of the infant born around 28 weeks gestational age is periventricular leucomalacia (PVL) which usually results in spastic diplegia, the clinical picture often being complicated by the presence of specific visual and perceptual problems. If the lesion is more extensive and extends into sub-cortical areas, the result is usually spastic quadriplegia often associated with seizures in addition to significant visual, cognitive and perceptual problems.

Extreme prematurity or severe asphyxia of full term neonates have produced children with low tone proximally, especially in the trunk, usually accompanied by marked hypertonia of the limbs.

The child showing the typical picture of spastic diplegia has bilateral equinovarus deformities, whereas the child who demonsrates truncal instability has valgus deformity of his feet due to co-existence of hypertonia and hypotonia, the latter allowing for hypermobility of the ankles and forefeet[1].

Although the major problem of CP is related to motor function, there are often associated problems which make rehabilitation of these children a real challenge. These are problems related to:

- Intelligence
- Cognitive functions
- Eating and drinking
- Communication
- Epilepsy
- Co-ordination
- Emotions and behaviour
- Hearing and vision
- Sensation and perception
- Activities of daily living

The Upper Motor Neurone Syndrome

A lesion of the upper motor neurone (UMN) is very complex. For an easier understanding, the characteristics are divided into positive, negative and adaptive (rheologic) features[2].

Positive features

1. *Spasticity*

    - Increased muscle tone
    - Exaggerated tendon reflexes
    - Stretch reflexes spread to extensors
    - Repetitive stretch reflex discharges clonus

2. *Released flexor reflexes*

    - Babinski response
    - Mass synergy patterns

Negative features

- Loss of finger dexterity
- Weakness
  Inadequate force generation
  Slow movements
- Loss of co-ordination

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Adaptive (rheologic) features

- Stiffness
- Contracture
- Fibrosis
- Atrophy

In a patient with an UMN syndrome the negative features are more disabling than the positive.

Spasticity

Spasticity may be defined as a motor disorder characterized by a velocity dependent increase in tonic stretch reflexes with exaggerated tendon reflexes resulting from hyperexcitability of the stretch reflex as one component of the UMN syndrome[2]. Spasticity is associated with a tendency to limit the range of movements and may lead to contractures.

Spasticity is best assessed in the supine position when the patient is fully relaxed. It is tested by quick passive movements from full flexion to extension done at several joints, and graded according to Ashworth scale taking into account the increase of tone and limitation of joint movement.

Ashworth Scale

0 - No increase of tone; full range of movement.

1 - Little increase of tone; nearly full range of movement; repeated clonus may occur.

2 - Noticeable increase of tone; the range of movement decreases to about 80%.

3 - Extensive increase in tone; range of movements decreases to about 15%.

4 - Hardly any movements possible with quick passive movements.

In the limbs spasticity causes a chain of events progressing through reduced muscle excursion, dynamic contracture (fully reversible), mixed muscle, contracture (partially reversible) fixed musculo-tendinous contracture (not reversible), short muscles bony torsional abnormalities and joint instability. These changes are time dependent, so that younger children mostly exhibit spasticity with few contractures, whereas older children have a more complex problem with spasticity, contracture and secondary bony deformities.

Tone

Tone may be described as a state of readiness (firmness) in the muscles and is assessed by checking the resistance to passive movements. It is contributed to by

1) *Neural mechanisms – [central]* – spasticity/rigidity.

2) *Non neural*

(a) Adaptive (Rheologic) – viscoelastic properties of muscles.

(b) Weakness of antagonistic muscles which fail to stretch the affected muscles fully.

Tone may be further subdivided into intrinsic and postural tone[3]. Intrinsic or true tone is specified by the reactivity of the stretch reflex, and in the normal child can only be elicited by the tendon tap. Postural tone is the state of continuous and unfatiguable contraction of postural muscles needed to overcome gravity and maintain posture and as such is decreased if the postural mechanism is impaired as is often the case in cerebral palsy. Therefore, it is not uncommon for a 'spastic' child – one whose deep tendon reflexes are increased – to be floppy due to impaired postural mechanisms.

Pathophysiology of movement disorder in cerebral palsy[1]

1. Lack of cortical control of the spinal motor neurone pool results in
   - Hyperreflexia
   - Reduction in the number of motor units recruited in voluntary activity resulting in slow initiation, slow, laboured ineffective, weak less forceful muscle contractions.
   - Ineffective manipulative skills due to limited ability to perform relatively independent finger movements.
2. Lack of synchronization of synergistic muscles such as biceps and brachialis, which in normal circumstances would produce a sustained forceful contraction to effect quick, smooth elbow flexion

3. Impaired interaction between agonist and antagonist muscle pairs. In children with spastic CP agonist and antagonistic muscles in the limbs co-contract which result in inefficient limb movements. Alternatively the trunk muscles contract reciprocally and hence does not provide sufficient trunk stability for postural control and independent limb movements.

4. Impairment of postural control – The ability to maintain stability and balance in a certain posture.

Intervention

The overall aim is to improve the quality of life for the individual person and his/her family and to prepare for improved quality of life during adult years. Depending on the age of the child and the severity of the disability three general aims can be identified.

- Improve existing skills – motor, social, play, cognitive, communication and activities of daily living.
- Maintain existing functional level in a child who has already reached his/her full rehabilitation potential.
- General management and minimization of contractures and deformities.

Priorities for intervention are as follows:

- Communication
- Activities of daily living
- Hand function
- Mobility / Walking

Rehabilitation of the child with CP is done by the team comprising medical and non-medical people using a multidisciplinary approach. The three most important people in this team are the physiotherapist, occupational therapist and speech therapist. A rehabilitation consultant would supervise the therapists' work and co-ordinate between the other members. Various skills of the child are helped by therapists and figure 1 is a guide to referral to those therapy departments.

		Physiotherapy	*Occupational therapy*	*Speech and Language*
1	Posture	X		
2	Movement	X		
3	Handskills [hand eye coordination, reaching, grasping]		X	
4	Play		X	
5	Learning [concentration, awareness of environment, cognition]		X	
6	Self care activities [washing, dressing and toileting]		X	
7	Eating, drinking and swallowing		X	X
8	Communication [Interaction, speech and language]			X

Figure 1. *Guide to referral*.

Principles of therapy approaches

a. **Reduction of the effects of abnormal muscle tone / postural tone**

- Muscle stretching
- Orthosis / Casting
- Handling / Positioning
- Botulinum toxin A
- Selective dorsal rhizotomy
- Intrathecal baclofen
- Single event multilevel surgery

The above techniques are used primarily to improve / facilitate functions or skills. In a more disabled or an older child these may be used to help position or comfort a child. Sometimes these may be helpful to the carer in attending to the child's daily needs.

Muscle stretching is one of the most important aspects of physiotherapy. One of the conditions required for the growth of skeletal muscle is that the muscle should be stretched in its relaxed condition[4]. In stretching, the origin of the muscle has to be moved away from the insertion. This loosens cross bridges between actin and myosin and helps to reduce the sensitivity of muscle spindles. Stretching can be done passively by the therapist in the small child. Positioning and orthosis may be used for the same purpose. In the more able child stretching can be performed actively. Stretching improves the range of movement and stimulates linear growth of muscles. However, it is only by activating the person in optimal and useful ways that any long lasting reduction of the effect of spasticity (or tone) can be achieved.

Botulinum toxin A[4]

Botulinum toxin (BTx) A has been used in the management of CP since 1990. Growth of skeletal muscle depends on the muscles being stretched in its relaxed state. ***The main use of BTx is in the management of spasticity or dynamic contracture interfering with function in the absence of a fixed myostatic contracture.*** Target muscles are those muscles affected by spasticity interfering with function and traditionally developing significant contractures needing surgery e.g. biceps, brachialis, pronator teres, flexors of wrist, fingers, adductor pollicis, flexor pollicis brevis, hip flexors, hip adductors, hamstrings, gastrocnemius, soleus.

The dose of BTX is 4-6 u/kg body weight per muscle. Children between two to six years respond well and the effect lasts for 3-6 months. They may need definitive surgery later. Earlier period of use of toxin is recognized as a short term treatment during the key early years when functional skills in walking are being developed. Children with mild spasticity may never need surgery or may need less extensive surgery.

Handling

Handling is a therapy concept meaning using the hands and the body of the carer in all situations of child's daily life. The aims of handling are:

1. To influence tone – to reduce tone when it is high and to increase tone when it is low.
2. To increase the range of movement.
3. To introduce and encourage different ways of movement.
    - To increase the child's repertoire of skills.
    - To enable new experiences.
    - To participate as much as possible in daily activities.

Positioning

A normal child will go through a variety of positions such as supine, side-lying, prone, sitting and standing during the first year of life. A child with CP will spend most of his first year in the supine position. While supine position is extremely stable and comfortable it results in limited opportunities to develop other skills. Therefore it is important to use other positions in therapy after carefully weighing the advantages and disadvantages.

Aims of positioning are:

- To influence tone for more activity.
- Facilitate body symmetry and alignment.
- To provide more stability as a background for more functional mobility and to increase skills.

- To increase sensory input.
- To prevent / influence contractures.
- To influence growth of muscles and bones.
- To prevent pressure sores

b. Musculoskeletal problems

Muscle length

Hypertonic muscles do not elongate as well as healthy muscles during growth. Therapy involves stretching, positioning, and using orthosis and equipment to elongate muscles.

Muscles weakness

The pre-occupation with management of abnormal tone has resulted in less attention being paid to muscle strength the thought being that it is secondary to the primary problem of tone. Muscle weakness is a direct effect of reduced cortical output as it leads to reduced voluntary drive on to spinal motor neurones resulting in reduction in the number of motor units recruited. This leads to slow initiation and activation of muscles with less forceful contractions. This is complicated by impaired reciprocal inhibition and co-contraction of antagonistic muscles which aggravate the inefficiency of muscle contractions. Therefore any therapy programme should have a pleasurable activity programme for the child specifically aimed at strengthening various muscle groups.

c. Working for activity in a functional context

Neural reorganization can occur as a result of specific or altered sensory input or can be activity driven. Therapists use various sensory inputs such as tactile, proprioceptive and vestibular to try to improve the sensory control of a child's function. From a motor perspective therapists attempt through handling techniques, stretching, specific activation of muscle patterns and helping a child to modify the way functional activities are carried out, to promote optimal development of activities of daily life. Patterns of muscle activation should be learnt in appropriate functional context. Unfortunately many therapists do not place enough importance on the practice of functional activities in their intervention programmes. Therefore practice and training of functional activities and goal directed therapy programmes are extremely valuable for the child with CP. Constraint induced therapy where sound side is restrained in patients with hemiplegia forcing the use of the affected side promotes improved function and neural reorganization at the level of the cortex.

d. Providing opportunities for practice

Therapy should not be confined to the therapy session, nor should a situation exist in which patients are dependent on the therapist. Rather, therapist should be a facilitator enabling ideas and modifications to be integrated into all aspects of the child's life at home, school, and all environments encountered during the child's daily life. This necessitates the training not only of the child but also of parents, carers and family members in the most useful ways to help the child. This requires both common sense and a setting of realistic, achievable and relevant goals for each individual person. Eating and drinking session and other self care activities such as dressing, toileting and washing provide enough opportunities for therapy intervention.

Play provides another excellent opportunity for intervention. Play may be defined as pleasurable interaction with people and exploration of the environment. Play forms a natural part of a child's development and is an enjoyable form of learning. Children with disabilities need extra help to play. Play can be used effectively to develop:

- Sensory skills – seeing, hearing, feeling, touching, smelling, tasting.
- Physical skills – balance, movement, co-ordination, hand function.
- Intellectual skills – concentration, thinking, talking, planning.
- Social skills – relationships, behaviour, turn taking.
- Emotions – feelings.

Careful planning and adapting play is often needed when play is used as a therapy idea in children with disabilities.

- Position – depending on child's physical skills one needs to find a play position which is stable and can be maintained for a sufficient time. Additional support may be provided externally.
- Toys may have to be adapted to make it easier for the child to handle.
- Toys will have to be carefully selected taking into account child's age and skills.

- The activity may have to be adapted according to the skill level.

- Activity has to be varied as children's concentration is limited.

Rehabilitation – current Sri Lankan set up

All teaching hospitals, general hospitals and some base hospitals are currently served by physiotherapists. Speech and language therapists are becoming available to most hospitals. However the passing out of occupational therapists occur at an extremely slow rate (1-2 per two years). At present occupational therapy services are available for neuro rehabilitation only at Ragama Rehabilitation Hospital, Lady Ridgeway Hospital and at Teaching Hospital Kandy (THK). THK does not run a separate service for children. Like all other hospitals in Sri Lanka THK too treats children and adults together in therapy departments. This is extremely unsatisfactory. Therefore at present the best centres available for children with disabilities in Sri Lanka are Lady Ridgeway Hospital and Ragama Rehabilitation Hospital.

Multidisciplinary approach is limited to a mere word in the Sri Lankan set-up. Rather than discussing relevant clinical issues among the team members at meetings and taking collective decisions, each member has a strong tendency to take his or her own decision. In addition, it is extremely rare to see the physiotherapist, occupational therapist, speech and language therapist working together with one child at the same time.

Another main drawback in our set-up is the guilt and over protectiveness of the parents in Sri Lanka. They tend to do most things for children and do not give enough opportunities and encouragement for children to practise and develop their skills.

In conclusion the following are important concepts of the therapy intervention:

- Maintenance of muscle length.

- Attention to muscle strength.

- Acceptance of the limitations of the damaged nervous system while not neglecting the potential for positive adaptation and change.

- A consideration of the need to prepare for adult life.

- A programme which aims to achieve meaningful and relevant functional goals.

- A programme which offers a manageable routine compatible with quality of family life.

Drastic changes are not required in what therapists do at present. Rather, a more rational approach, explanation and individualized management would be more beneficial for the families of children with cerebral palsy

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