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Neuromusculoskeletal Rehabilitation of Severe Cerebral Palsy

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Abstract

Persons with Gross Motor Function Classification System (GMFCS) levels IV and V are considered as severe cerebral palsy (CP) and are non-ambulatory. These persons are at a higher risk of complications such as hip displacement (sub-luxation or dislocation), spinopelvic deformities, musculoskeletal pain, low bone mineral density and low energy fracture. The recommended management strategy at present for this group is wheelchair-aided mobility, with which none of these complications can be prevented. There is a strong need to evaluate alternative methods of treatment that can allow assisted ambulation in persons with severe CP. The role of Single Event Multilevel Lever Arm Restoration and AntiSpasticity Surgery (SEMLARASS) and protocol-based active rehabilitation on gross motor function and ambulation of non-ambulatory persons with CP at GMFCS levels IV and V is examined. Active rehabilitation involves making the person with severe CP active through most of the waking hours and participating actively in the rehabilitation. A well-planned and executed SEMLARASS, followed by intensive, protocol-based, sequenced multidisciplinary active rehabilitation, provides the persons with GMFCS levels IV and V a significant functional improvement in gross motor function and mobility.

Keywords: cerebral palsy, neuromusculoskeletal rehabilitation, SEMLARASS, active rehabilitation, GMFCS

1. Introduction

Cerebral palsy (CP) is a non-progressive disorder affecting the individual's posture, movement, and causing limitation in the activities that are permanent, caused due to damage in developing brain of neonates or infants. CP causes not only motor disturbances, but also sensory,

cognitive, social, behaviour, speech and communication, seizure disorder, respiratory illness and other musculoskeletal disorders [1]. A total of 17 million persons are estimated to have CP worldwide and CP is one of the most common causes of physical disability among children. The prevalence of CP is currently estimated to be 2.11/1000 live births [2] and varies between 1 and 5/1000 live births in different countries. 28% of persons with CP have epilepsy, 58% have difficulties with communication, 42% have visual problems and 23–56% have learning disabilities [3].

Clinicians classify patients with CP to describe the specific problem, to predict prognosis and to guide treatment. Classification is based on the change in muscle tone, anatomical region of involvement and severity of the problem. Types of CP according to muscle tone are spastic (hemiplegia, diplegia and quadriplegia based on anatomical region of involvement), ataxic and dyskinetic (dystonia and choreoathetosis). Even though these terms do not have specific reliability among the observers, these terms are helpful for understanding the CP condition [4]. Gross Motor Function Classification System (GMFCS) is another system to classify gross motor function of children and youths with CP on the basis of their self-initiated movement with particular emphasis on sitting, walking and wheeled mobility. Children and youths with GMFCS levels IV and V are non-ambulatory and are considered as severe CP. Persons in GMFCS level IV use wheelchair, either manual or automatic, for transportation and persons in GMFCS level V are highly dependent because of the lack of balance in head, neck and trunk, and will require major assistive devices for physical assistance [5].

According to data from North India, 69% of persons with CP had spastic quadriplegia or dyskinetic CP and were non-ambulatory [6]. These persons are at a higher risk of developing complications such as hip displacement (sub-luxation or dislocation), spinopelvic deformities, musculoskeletal pain, low bone mineral density and low energy fracture. The most common and serious structural change in persons who have severe CP is hip displacement. This deformity is seldom present at birth but develops as the child grows older and experiences abnormal muscle pull from spastic muscles, increased femoral anteversion and the lack of weight bearing on the lower extremities. The reported rates of hip displacement in persons with CP vary from 1 to 75% [7]. The incidence of hip displacement in CP is related to the severity of involvement, varying from 1% in children with spastic hemiplegia up to 75% in those with spastic quadriplegia [8]. In two population studies, the rate of hip displacement was found to be one-third and was not related to the movement disorder but was directly related to gross motor function as determined by the GMFCS [9, 10].

The prevalence of hip pain in severe CP is reported to be 47.2% [11]. The common type of pain identified was provoked pain (e.g. during mobilisation, palpation and weight bearing on the lower extremities). A study of 2777 children (57% boys) at a median age of 7 years reported 32.4% children in pain, with significantly more girls than boys experiencing pain, and significantly more children at GMFCS levels III and V than GMFCS level I. The frequency of pain increased with age. Pain in the abdomen and hips was most frequent at GMFCS V, whereas knee pain was most frequent at level III and foot pain at level I [12].

Markedly low bone mass in children and adults with severe CP has been reported to place these persons at the risk of osteopenia, osteoporosis and low-energy fracture [13].

Increasing problems with reduced mobility lead to problems with daily activities especially in instrumental activities of daily living (IADL). Social participation, sexual relationships, employment and leisure activities are restricted among many youths and young adults with severe CP [14–18]. Severities of physical and/or cognitive impairment are predictors for limited participation, but limited participation is not necessarily synonymous with a poorer quality of life [14, 19, 20]. Some studies also show that pain, falling stamina and functional deterioration have a negative impact on the quality of life.

Several potential benefits exist for making a person with severe CP ambulate in a therapeutic setting:

1. The ability to retain standing transfers in adolescents and young adults with CP means that they do not require lifting or hoisting by their caregivers, thereby reducing the risk of musculoskeletal disorders in caregivers [21].
2. Supported walking and standing in a therapeutic setting for non-ambulatory children with CP seem to improve participation in activities of daily living and social roles, as well as pulmonary and gastrointestinal functions [22].
3. Improved bone mineral density [23].
4. Less musculoskeletal pain.
5. Reduced risk of hip displacement [9, 10].
6. Lesser risk of pressure sores, aspiration and early death.

2. Goals of treatment

The goals of treatment for persons with severe CP are different from those for ambulatory persons with CP. Persons with severe CP are at increased risk of developing displacement of the hip, spinal deformities and joint contracture, which may altogether hinder and interfere with caregiving, positioning, sitting and transferring the person. Other comorbidities include cognitive disorders, visual and hearing impairment, epilepsy, difficulties in chewing and swallowing, drooling, speech, digestive disorder, respiratory illness and bowel and bladder problems [24].

The management aims [24, 25] of severe CP are to

1. Relieve or prevent pain and discomfort.
2. Facilitate ease of care: dressing, toileting, bathing/hygiene; positioning: seating and lying down; transfers and mobility.
3. Preserve or improve health.
4. Improve the quality of life.

More specific therapeutic goals [26] include the following:

1. Adequate tone control.
2. Straight spine and level pelvis (to allow comfortable sitting and positioning).
3. Stable, enlocated, mobile and painless hips.
4. Mobile knees that can flex to sit and extend to brace for transfer.
5. Plantigrade feet.

3. Treatment options

Management aims of severe CP are very challenging and the ultimate goal of the rehabilitation process is to make the child independent at community and household level. The primary aim is to prevent any secondary complications. The rehabilitation of severe CP is an intensive process in which the patient's goals are prioritised with the help of a team of physiotherapists, occupational therapists, speech therapists, psychologists, special educators, etc., headed by a rehabilitation physician to improve the person's function physically, mentally and socially. This process also requires active participation by the patient and caregivers. No two persons with severe CP are the same. However, the impact of rehabilitation techniques on one person must be taken for reference while rehabilitating the other.

The usual management of severe CP at present consists of physical therapy, sometimes followed by multiple, and often concurrent, medical and surgical interventions, most intensively in early childhood through pre-adolescence. While a growing list of treatments, e.g. oral antispasticity medication, alcohol, phenol or botulinum toxin injections, have been shown to individually improve some motor outcomes, few definitive practice guidelines have been proposed for the management of CP due to limited and fragmented scientific evidence to support multidisciplinary intervention approaches [27]. Persons with severe CP usually do not fulfil the selection criteria for selective dorsal rhizotomy. Intrathecal baclofen is a therapeutic option in this population, but the disadvantages include high cost and serious complications like infection, neurological injury and hip dislocation.

In ambulatory patients (GMFCS levels I, II and III), single-event multilevel surgery (SEMLS) has become widely accepted to be effective in improving gait parameters and the quality of life [28]. However, the effectiveness of orthopaedic surgery (OS) to improve and maintain mobility in children with lower functional levels (GMFCS levels IV and V) has not been ascertained. A study conducted in 2012 revealed that orthopaedic surgery in children with CP at GMFCS IV was unlikely to restore or maintain mobility. The study reported that the following results:

- Only 36.4% of the patients achieved their goals.
- The Functional Mobility Scale (FMS) remained the same in 95.4% of the patients.
- Most children lost their ability to perform assisted walking and standing transfers at 2 years' post-surgery.

The authors concluded that the role of orthopaedic surgery for children with CP at GMFCS level IV is limited to the treatment and prevention of spastic hip disease and scoliosis [29].

The aims of OS in GMFCS level IV are to optimise

1. Foot positioning for standing and walking/transferring.
2. Knee extension for standing and walking/transferring.
3. Hips to prevent progressive sub-luxation and dislocation.

The aims of OS in GMFCS level V are to optimise

1. Foot positioning for feet on footplate of wheelchair.
2. Hips to prevent progressive sub-luxation and dislocation.
3. Other surgery may be indicated if impacting on the persons' quality of life (e.g. pain) or ability to be positioned comfortably in their wheelchair.

None of the conventional therapeutic approaches reported so far have shown any significant improvement in gross motor function or the ability to ambulate in persons with severe CP. Consequently, the recommended rehabilitation strategy across the world at present for severe CP is wheelchair-aided mobility. Hence, there is a strong need to evaluate alternative methods of treatment that can allow assisted ambulation in persons with severe CP.

4. Orthopaedic selective spasticity-control surgery (OSSCS)

OSSCS, a Japanese OS approach, has been proposed with the aims of selective reduction of a specific muscle's spasticity, dystonia and athetosis, and improvement of anti-gravity posture control and movement [30]. The principles of OSSCS are as follows:

1. Longer muscles are selected for surgical release on the assumption that spasticity of shorter muscles limits anti-gravity function in persons with CP.
2. Longer muscles that are considered are always multi-articular and inserted at the more distal portion in the same muscle group.
3. The longer and hyperactive muscle fibres can be selectively sectioned with intramuscular tendon lengthening and controlled sliding tendon lengthening.
4. Simultaneous release of flexor and extensor muscle groups is performed in each joint (except at wrists, hands and feet).

The main surgical techniques in OSSCS are intramuscular release and controlled sliding lengthening [30].

The advantages of OSSCS over conventional OS [30, 31] are as follows:

1. There is no loss of anti-gravity activity and weakness of the muscles because monoarticular muscles are preserved.

2. Over lengthening of tendons is avoided because of the surgical technique of controlled sliding tendon lengthening.
3. It controls spasticity, produces reciprocal movements to facilitate anti-gravity muscles and improves functional skills and voluntary movement of the hand.
4. It leads to significant functional improvement in the severely involved spastic quadriplegia, athetoid or dystonia.
5. There is no loss of sensation or sense of stereognosis.
6. There is no increase in the occurrence of dislocations.

5. Single Event Multilevel Lever Arm Restoration and Anti-Spasticity Surgery (SEMLARASS)

SEMLARASS is an advancement of the concept of OSSCS [32]. The additional principles of SEMLARASS include the following:

1. Operating between the ages of 4 and 6 years (preferably) to avoid joint decompensation and over lengthening of tendons that happen due to continued usage of deformed joints.
2. Simultaneous restoration of lever arm dysfunction (LAD) is essential for spasticity and contracture correction as well as to reduce chances of recurrence of deformities and repeat surgery at a later stage, and to improve the direction of pull of muscles and facilitating strengthening.
3. Minimally invasive procedures using image intensification that do not require large skin incisions and consequent risk of blood loss and infection.
4. Use of only external fixators that do not require a second operation for removal, and are technically superior to internal fixation in enabling reduction of dislocated hips and preventing stress shielding of the bone and consequent fractures after implant removal.
5. All surgeries to restore LAD are extra-articular to allow for the maximum growth potential of children's bones.
6. Power generators are preserved: tendon transfers of spastic muscles may lead to further weakness and worsen, lead to an opposite deformity, e.g. genu recurvatum following Eggers transfer.
7. For non-reducible hip dislocation, the preferred salvage operation is redirection of femoral head and tectoplasty while preserving the femoral head (**Figure 1** and **2**).
8. The surgery is followed by a structured, intensive, institutional, physician-directed, multidisciplinary rehabilitation protocol.



Figure 1. Percutaneous varus derotation osteotomies with external fixators.

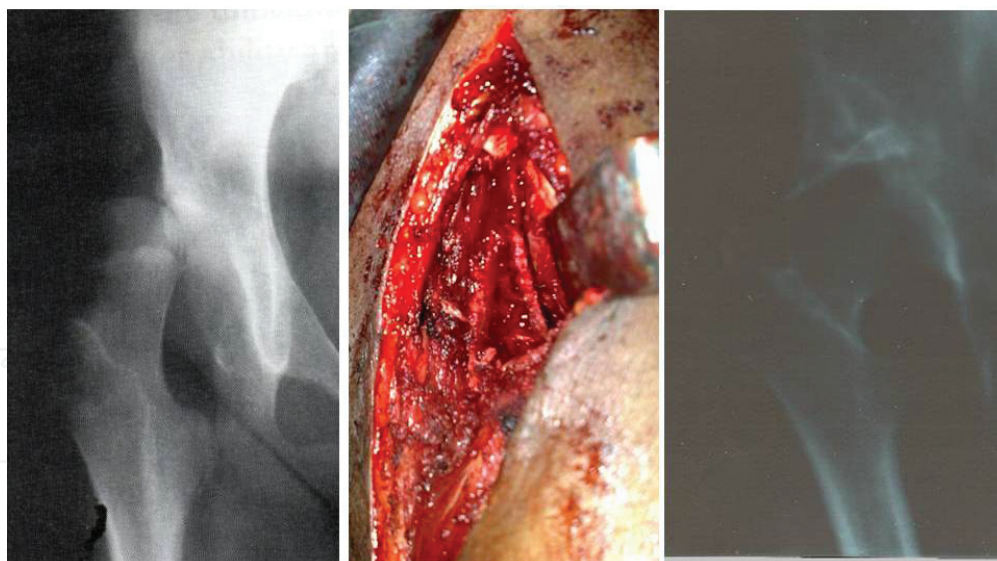


Figure 2. Tectoplasty.

Following are the components of SEMLARASS:

1. Single event: under a single anaesthesia, requiring only one hospital admission and one period of rehabilitation, all surgeries are completed.

2. Multilevel: simultaneous correction of all the affected regions and all orthopaedic deformities (soft tissue and bony) as joints are interdependent.
3. Lever arm restoration: to improve the direction of pull of muscles and to facilitate muscle strengthening post-operatively, LAD corrections are done simultaneously.
4. Anti-spasticity surgery: using the principles of OSSCS.

6. Rehabilitation approaches used with SEMLARASS

SEMLARASS is followed by a protocol-based rehabilitation that has already been published [32]. This rehabilitation approach is comprehensive, and includes physiotherapy, occupational therapy, speech therapy, orthosis and other adaptive equipment, recreational activities, school and education adaptation and psychosocial support, etc. [33]. Rehabilitation in severe CP can differ due to clinical type and severity of conditions, physiological age and socioeconomic factors. In addition, visual, auditory, cognitive disorders, seizures, learning disabilities and emotional problems may influence intervention outcomes [34]. Physiotherapy plays a central role in managing the condition; it focuses on function, movement and optimal use of the person's potential. Physiotherapy uses physical approaches to promote, maintain and restore physical, psychological and social well-being. The rehabilitation occurs not only at the rehabilitation centre but also involves the functional strategies at home, community, school or work and other recreational environments, where the therapists works on making the person with CP to become independent by concentrating on gross motor activities, functional mobility and ambulation either with or without assistive devices [33].

Active rehabilitation has been the choice of functional treatment for CP according to the present evidence available in the literature [35]. The rehabilitation is focussed on a combination of aspects involving physical, mental and social functions. It involves a time-bound interaction between the patient, therapist and other persons involved in the rehabilitation process to show a reduction in the disability on the person's day-to-day activities by his or her condition [36]. Active rehabilitation involves making the person with severe CP active through most of the waking hours and participating actively in the rehabilitation. A variety of therapeutic programmes such as aquatic therapy, virtual reality-based therapy, physiotherapy, occupational therapy, hippotherapy (HT), whole body vibration therapy (WBVT), body-weight-supported treadmill training, EMG biofeedback and functional activity training are used. In addition, supportive therapies such as psychological counselling, special education, neurotherapy, yoga therapy and relaxation exercises also form a part of the treatment regimen. In the post-operative phase, a person with severe CP undergoes 5–6 hours of the above therapies in a programmed manner through a phased multidisciplinary treatment protocol for 6–9 months and less intensively thereafter [32].

6.1. Aquatic therapy

Aquatic therapy is one of the most popular and important rehabilitation strategies in persons with severe CP [37]. Water is an equalising medium; its gravity-minimising nature reduces

compressive joint forces, providing a better exercise environment for patients with medical conditions that may restrict physical training on land [38]. Adapted aquatic exercises have been particularly recommended as a part of physical activity programmes for persons with severe CP. The buoyant nature of water provides persons with severe CP the opportunity to feel their bodies free from the constraints they experience on land [39]. Aquatic exercises have benefits on joint range of motion, strength of muscles, pain, muscle spasms, circulation and respiratory function, speech, balance, coordination and posture [40]. Ease of movement and weight relief allows safe movement exploration, strengthening and functional activity training with a reduced level of joint loading and impact, providing a gentler environment for persons who experience persistent abnormal loading [39]. In addition, aquatic physical activities are important for the teaching-learning process and might promote greater independence, better manual ability and, as a consequence, increase social participation in persons with severe CP [41]. Despite the fact that swimming is one of the most frequently reported physical activities in children and adolescents with CP, there is no consensus on optimal concepts of aquatic physical activity regarding duration of intervention period, duration of a single treatment, frequency per week of treatment, individual/group work, water temperature and swimming pool size and depth [38]. In our practice, the aquatic therapy consisted of 5 minutes of light warm-up in the temperature-controlled swimming pool (forward and backward walking, jumping and other such exercises), 20 minutes of exercise on swimming techniques (prone and back gliding from the wall, prone and back floating and blowing bubbles) and 5 minutes of play (ball games, chasing games, etc.). The therapy was focused and performed individually (**Figure 3**). To ensure active participation, the intervention was customised to maximise enjoyment by each individual. Depending on the improved performance demonstrated by each person and related functional ability, the complexity of the exercises was increased. In addition, some interventions focused more on arm movements than on leg movements and vice versa. Thus, the goals and progression of each person could be followed individually, and every instructor was able to easily continue onto the next lesson with each child.



Figure 3. Aquatic therapy.

6.2. Body-weight-supported treadmill training (BWSTT)

Persons with severe CP are non-ambulatory. Treadmill training has shown positive outcomes in improving ambulation in CP [42]. Animal studies of supported treadmill training have demonstrated restoration of coordinated stepping movements in spinalised cats [43]. BWSTT is an active, repetitive, task-specific approach used to facilitate attainment of stepping and locomotion and to achieve a more normalised gait pattern. It is a method of task-oriented ambulatory training using the overhead suspension system and harness to support a percentage of the person's body weight while walking on a treadmill. Other effects of BWSTT include increase in walking speed, improved balance and increased endurance [44]. We use an indigenously constructed BWSTT device to aid persons with severe CP during the gait training phase of rehabilitation (**Figure 4**). The body weight support device can facilitate walking, both on treadmill and on level ground. In addition to its effectiveness in improving the person's walking ability, it also reduces the physical workload of physiotherapists and caregivers handling the persons with severe CP.



Figure 4. Body-weight-supported treadmill training.

6.3. Functional electrical stimulation (FES)

FES is used to stimulate targeted muscles during ambulation, especially in enhancing quadriceps function, allowing for better range in knee extension. It is used to improve gait control and trunk control [45–47]. Persons with severe CP have difficulty in generating sufficient muscle force. So, combining exercise with FES programme is a good option for increasing the intensity and effectiveness of the strengthening programme. Studies show participants in an FES programme can make measurable gains in body structure and function, activity and participation. The evidence supporting the efficacy of FES in improving gait quality, gait symmetry and muscle strength and motor control in persons with CP is growing steadily [48–51]. Given the complex nature of the gait deviations seen in children with CP, many paediatric FES studies investigate the effect of multi-channel FES systems on abnormal gait. Most multi-channel systems include stimulation of the anterior tibialis muscle as a treatment for drop foot [45, 51, 52]. Two of the FES review studies investigated single-channel FES systems that operate as neuroprostheses by stimulating the peroneal nerve to alleviate drop foot [45, 48]. In our setting also, we use a single-channel FES to facilitate the activation of tibialis anterior and knee extension for gait training, especially during treadmill training (**Figure 5**).



Figure 5. Functional electrical stimulation.

6.4. Whole body vibration therapy (WBVT)

Studies on vibration therapy at a specific frequency and amplitude have many negative effects on humans. However, recent studies have shown that vibration therapy for humans at very low amplitude and frequency is safe and beneficial to the human musculoskeletal structures. WBVT can be used as a form of exercise targeted for improving muscle strength, power, flexibility and coordination. Persons using this tool stand on the vibrating platform so that the whole body was stimulated with a sinusoidal vibration [52]. WBVT has been utilised to deliver mechanical accelerations to the appendicular and axial skeletons to elicit increased bone mass. WBVT has been shown to be specifically effective in improving bone mineral density especially in hip and spine for persons with severe CP after plaster immobilisation [53, 54]. The advantage of WBVT lies in its ability to be applied in a low-impact manner, which is critical for persons with impaired mobility and muscle strength (**Figure 6**). In our practice, WBVT is used, initially with suspension similar to BWSTT, in persons with severe CP who are in the weight bearing phase of rehabilitation.



Figure 6. Whole body vibration therapy.

6.5. Virtual reality-based therapy (VRBT)

Virtual reality is the use of interactive simulations using computer hardware and software to present users with opportunities to perform rehabilitation in virtual environments that appear,

sound, and less frequently, feel similar to real-world objects and events. The advantage of VRBT is that it allows a more elaborate and complex interaction between the virtual environment and the user. We have reported on the successful use of Nintendo Wii for VRBT to improve the balance, motivation and participation of children with CP following SEMLARASS [55] (**Figure 7**). Virtual reality-based bilateral arm training shows improvement in upper limb motor skills on the affected sites and in bilateral coordination ability. It can also improve trunk control and concentration [56]. Active participation, receiving feedback and repetition of movements during the VRBT intervention, assists in motor learning that induces cortical reorganisation and neural plasticity changes in the brain [57–59]. VRBT also increases the exercise compliance level in achieving selective motor control and enhances the effectiveness of conventional physiotherapy [60]. Participants were highly motivated by the feedback, challenge, variability and other competitive factors involved in the VRBT and overall give a sense of achievement mimicking the real world [61–63]. We also use Microsoft X-Box with Kinect and virtual reality headsets in neurorehabilitation. Initially, suspension is used for non-ambulatory persons.



Figure 7. Virtual reality-based therapy.

6.6. EMG biofeedback

Biofeedback uses the principle of measuring and processing normal and/or abnormal neuromuscular activity in the form of auditory or visual feedback by means of an electromechanical

instrument. EMG biofeedback specifically uses surface electrodes and records the muscular activity and gives the feedback that can be therapeutically used to facilitate or inhibit the muscular activity. This helps to create a better awareness about the physiological process of the abnormal body movements in specific activities of the individuals. Pressure, temperature, angular and positional systems can also be used as other modes of biofeedback similar to EMG biofeedback [64–67]. EMG biofeedback is now widely used in the rehabilitation of upper motor neuron lesions and also found that it helps in improving the spastic muscles by relaxing them [68–70]. In CP rehabilitation, EMG biofeedback has not been evaluated with any major controlled studies. Studies with small sample sizes have reported reduction in spasticity in the gastrocnemius [71] and increases in active range of motion and ankle dorsiflexion strength [72]. Although EMG is used to reduce muscle spasticity, there was no degree of changes in muscle contracture using EMG biofeedback. However, reversing muscle contracture can be achieved by combining surgical procedures that lengthen the muscles along with biofeedback training in order to maintain the lengthened muscle. [73]. In our setup, we use a single-channel EMG biofeedback device that provides an individual with supplemental information about the response of muscular activity, allowing the individual to attempt to control a given output associated with this process (**Figure 8**). Through trial and error, the person receives feedback on his or her success in adapting their behaviour to achieve the desired output (reduced spasticity or improved muscle strength). A variety of instruments are being used in research and in the clinical setting including surface EMG, computer-assisted feedback and a variety of simple auditory and visual feedback such as providing an auditory cue on the heel of a child to encourage heel-toe gait [74].



Figure 8. EMG biofeedback.

6.7. Activity monitor (AM)

AMs can be used for persons with severe CP as a biofeedback device, especially in rehabilitation for improving specific gait parameters. It is economical, easy to use and provides a real-

time feedback [75]. Physical activity seems to be one of the most important factors in effective CP rehabilitation or prevention of functional decline in older persons [76]. Small, lightweight, body-worn accelerometers that are able to record activity over longer periods of time now are available commercially. Body positions, movement and number of steps taken by the subject are detected in the sensory systems by inbuilt pre-installed software. This pre-installed software helps to extract data of the body's movement such as no of steps taken, time, speed, kilometers travelled and calories burnt. In our setup, we use a small, lightweight AM worn on persons' body for a particular period of time and subsequently monitoring the output and fixing targets to achieve, thus acting as a biofeedback device for persons with severe CP (**Figure 9**). This accurate measurement of free-living physical activity using advanced dynamic acceleration and inclination logging technology allows medical and rehabilitation professionals to assess patient compliance with exercise and treatment protocols, and also patient response to novel treatment interventions.



Figure 9. Tracking the training with activity monitor.

6.8. Hippotherapy

HT is a form of physical, occupational or speech therapy utilising a trained horse. The movement of the horse affects a rider's posture, balance, coordination, strength and sensorimotor systems. Recently, mechanical HT devices have been used in the rehabilitation of

persons with severe CP. The mechanical HT is the modern form of providing therapeutic horse riding benefits by a pre-programmed mechanically operated device, mimicking a horse riding experience (**Figure 10**). We use mechanical HT to improve head and trunk control, sitting balance, posture and promote functional activities [77–80]. HT should be avoided in the presence of hip displacement or hip osteotomies (till the stage of bony consolidation).



Figure 10. Mechanical Hippotherapy.

6.9. Velcro platform (VP)

Persons with CP often have difficulty in balancing post-SEMLARASS. VP is a novel method developed and designed by us for improving standing balance. The principle behind the use of Velcro for increasing the stability of base of support was that the sensory input from the varied rehabilitation strategies using Velcro for support helps in increased muscular activation, thereby improving functional performance [81]. The majority of persons with severe CP have severe muscular weakness and fear of falling, post-SEMLARASS, and have difficulties in achieving standing balance. VP consists of a wide wooden board attached with foot-contoured pads enforced with straps that accommodate the person's feet. The positive and negative pieces of a conventional Velcro form the contact surfaces of the platform base and the foot pads. The person is made to stand on the board fastening the Velcro straps. The device provides better foot contact to the ground, which aids in better weight bearing, proper

biomechanical alignment and acts as an indirect psychological assistance, which can make a person stand independently without or with minimal manual assistance (**Figure 11**). It helps to reduce the gravitational insecurities, fear of falling and gives confidence to a patient. Although the pilot study reported significant improvement in balance parameters and reduction in fear of falls, a larger clinical trial investigating the effectiveness of the VP is under progress.



Figure 11. Balance training with Velcro platform.

6.10. Head-held laser illumination device (HHLID)

Similar to VP, HHLID is also an indigenously developed device that can be a useful tool in the post-SEMLARASS rehabilitation programme for the improvement of sitting or standing balance and head or trunk control in persons with severe CP. A Laser-pointer-based focussed trajectory exercises were found to improve the hip extensor activation in comparison to erector spinae activation during pelvic bridging exercise [82]. A laser pointer device attached to the pelvis has been used to assess impaired balance [83]. HHLID consists of a laser-emitting device that can be fixed to the person's head or pelvis and the target is to focus on a variable screen

with mazes that challenges the person's balance and control abilities (**Figure 12**). HHLID works by giving visual feedback and it requires active correction of the patient, initially guided and assisted by the physiotherapist. The pilot study reported significant improvements in sitting balance, and a larger clinical trial investigating the effectiveness of the HHLID is under progress.



Figure 12. Child fixed with a head held laser illumination device.

6.11. Gaze-assistive rehabilitation technology (GART)

Gaze stabilisation during head motion is an important aspect of rehabilitation of individuals with severe physical impairments with problems in communication and speech disorders. For such persons, gaze stabilisation and control-based technological devices help in communication and interaction without the use of upper or lower extremities [84]. GART works on the principle of video-based corneal reflection eye trackers [85]. A few studies have reported the use of GART in severe CP helping them perform certain activities and increase participation [86, 87]. We are currently studying the Samsung EyeCan+ eye mouse, a futuristic device that enables people with severe CP to use computers only through eye movements. The EyeCan+ is a simple portable box positioned near the computer monitor and users are not required to wear glasses or other special equipment. Users can work with the device either sitting or lying down and just need to be a couple of feet from the monitor. This device allows people to compose and edit documents as well as browse the web (click, double click, scroll, drag, etc.) through simple eye movements. Eighteen different mouse menus allow the person with severe CP to communicate, play games and participate in rehabilitation (**Figure 13**).



Figure 13. EyeCan+ device fixed to a personal computer.

Other supportive therapies in the rehabilitation protocol include occupational therapy involving specific goal-oriented therapies such as constraint-induced movement therapy (CIMT), mirror therapy, play therapy, music and art therapy, psychological counselling, behavioural training, special education, yoga therapy, speech therapy and dance movement therapy.

6.12. Results of SEMLARASS for severe CP

A study was conducted on 170 children with severe CP (GMFCS levels IV and V) to find out the functional outcome of SEMLARASS and rehabilitation. The mean age of the participants was 9.68 ± 4.77 years. The follow-up ranged from 2 to 10 years (mean = 4 years). The outcome measures such as component of Gross Motor Function Measure (GMFM-88), Functional Mobility Scale, Physicians Rating Scale (PRS), Manual Ability Classification System (MACS) were used to compare the functional status of the child before and after SEMLARASS.

The results showed a significant improvement in all GMFM-88 components and the values were lying and rolling (A): GMFM 5: $t=9.77$ ($P < 0.001$); GMFM 4: $t=8.56$ ($P < 0.001$); sitting (B): GMFM 5: $t=20.01$ ($P < 0.001$); GMFM 4: $t=12.61$ ($P < 0.001$); crawling and kneeling (C): GMFM 5: $t=22.26$ ($P < 0.001$); GMFM 4: $t=21.01$ ($P < 0.001$); standing (D): GMFM 5: $t=20.01$ ($P < 0.001$); GMFM 4: $t=22.64$ ($P < 0.001$); standing (D): GMFM 5: $t=20.01$ ($P < 0.001$); GMFM 4: $t=22.64$ ($P < 0.001$); walking, running and jumping (E): GMFM 5: $t=12.71$ ($P < 0.001$); GMFM 4: $t=15.65$ ($P < 0.001$) and total GMFM-88: GMFM 5: $t=31.55$ ($P < 0.001$); GMFM 4: $t=32.86$ ($P < 0.001$), respectively. The result of pre-post PRS evaluation showed a significant improvement for both sides ($P < 0.01$). Correlation studies showed median value of Functional Mobility Scale of 1 before surgery and 3 after surgery. Before surgery the median value of Gross Motor Functional Classification System was level IV and after surgery it was level II. The GMFCS improved two

levels on average. Before surgery, the mean value of Pediatric QOL (PQOL) was 23.11 ± 14.02 ; after surgery, the mean value was 39.64 ± 17.49 . Before surgery median value of Manual Ability Classification System was 3 and after surgery it was 1. No child was wheelchair bound at the end of the rehabilitation and all the children were able to walk at least with help of a walking aid. A significant improvement was noted in their participation levels, motivation and a significant improvement in the overall quality of life [88]. Over 50 patients have been followed up for 10 years and there have been no significant recurrence of deformities or significant deterioration of gross motor function.

7. Discussion

The currently practised treatment options have little impact on gross motor function and mobility in non-ambulatory persons with spastic quadriplegia and dyskinetic CP, which constitute nearly 70% of all cases of CP. In particular, OS is considered to have minimal role in this patient population. A retrospective cohort study of 107 children with bilateral spastic CP, classified as GMFCS level II or III, who underwent surgery at a single tertiary institution in Australia between 1997 and 2008, reported that the GMFCS levels remained stable and unchanged in 95% of children and improved by one level in 5% of children [89]. Khan reported a series of previously untreated 85 non-walker children with diplegic CP who underwent multilevel surgery. All patients improved and became walkers. However, since the GMFCS was not used, their cohort cannot be compared to this study [90]. Blumetti et al. found a low rate of success after surgery in patients with GMFCS level IV with only 36.4% of the patients achieving their goals. The FMS scores remained the same in 95.4% of the patients. Only one patient maintained an FMS score of 2, 1, 1 at 2 years' follow-up. Most children lost their ability to do supported walking and standing transfers at 2 years' post-surgery [29]. Some external factors are known to influence the outcomes after OS, including post-operative rehabilitation, use of orthotic devices, pain-controlling strategies, adequate tone control, and presence of co-morbidities [91]. However, all patients in this series received a standardised rehabilitation programme as described previously and were closely monitored by the team of medical and rehabilitation professionals. Unlike previous studies the current study showed that GMFCS levels improved at least by two levels and significant improvement in gross motor function and mobility was recorded. The main limitation of the study was the lack of a control group.

8. Conclusion

A well-planned and executed SEMLARASS, followed by intensive, protocol-based, sequenced multidisciplinary active rehabilitation, provides the person with severe CP, a significant functional improvement in gross motor function and mobility. SEMLARASS is the only documented treatment for CP till date that has been able to address all the three key problems in CP—selective reduction of spasticity, dyskinetic movements and LAD. The best age for SEMLARASS is 4–6 years before the LAD become severe or joints become decompensated.

Intensive, protocol-based and medically supervised rehabilitation for several years and close follow-up are needed at least till skeletal maturity. SEMLARASS provides a patient with severe CP with the best hope for a dramatic, predictable and lasting functional improvement.

9. Case study 1

9.1. Pre-op status

(A) A 13-year-old girl with spastic quadriplegia was confined to bed and completely dependent on caregivers for all her daily activities. She had no neck control or sitting balance. When held upright by an adult, there was severe crouching at hips and knees. Her GMFCS level was V.

9.2. Treatment

She underwent SEMLARASS in two stages: the first stage with OSCSS of bilateral hamstring, psoas, rectus femoris and gracilis along with femoral derotation and tibial derotation osteotomies and the second stage (after 8 weeks) with OSCSS of bilateral forearm flexors, pronators and hand intrinsics.

9.3. Current functional status

At a follow-up of 13 months, she was able to walk with the assistance of walker with forearm gutter. She was able to sit independently either long or cross sitting and able to perform some of her daily activities such as feeding, brushing and upper body dressing on her own. Her present GMFCS score was III (**Figure 14**).



Figure 14. Pre- and post-rehabilitation status of child A.

‘Our daughter, till the age of 12, could only sit with maximal support. But after SEMLARASS and intensive rehabilitation in RECOUP, our daughter is now able to sit without support in both cross sitting and long sitting and now she is able to stand and walk with walking frame with minimum support. She now feels more confident’ – parents of A.

10. Case study 2

10.1. Pre-op status

(S) A 12-year-old boy with spastic diplegia was not able to walk and perform his daily activities. His GMFCS level was IV.

10.2. Treatment

He underwent SEMLARASS with OSCSS of bilateral hamstring, psoas and rectus femoris along with bilateral femoral varus derotation osteotomy.

10.3. Current functional status

At a follow-up of 12 months, he was able to walk independently for over 100 metres and able to climb up stairs by holding the hand rails and started going to school independently. Current GMFCS score was 2 (**Figure 15**).



Figure 15. Pre- and post-rehabilitation status of child S.

‘Our son, born with Spastic Diplegia, could not walk and needed constant support even for few steps and frequent falls and imbalance were a constant issue. Now, after the treatment at

RECOUP, he started walking independently with lot of confidence and enthusiasm’ — parents of S.

11. Case study 3

11.1. Pre-op status

(D) A 6-year-old girl with spastic quadriplegia and bilateral hip dislocations was confined to bed and could only bunny hop or creep from one point to other and was totally dependent on caregivers. Her GMFCS level was IV.

11.2. Treatment

She underwent SEMLARASS in two stages: the first stage with OSCSS of bilateral hamstring, psoas, rectus femoris, gracilis and tibialis anterior along with femoral varus derotation and tibial derotation osteotomies and the second stage (after 8 weeks) with OSCSS of bilateral forearm flexors and pronators.

11.3. Current functional status

After 12 months of follow-up, she was able to walk independently with walker and able to sit up in bed by herself and stand to sit with support. She was able to carry out some basic activities of daily living by her own. Her current GMFCS score was III (**Figure 16**).



Figure 16. Pre- and post-rehabilitation status of child D.

‘The SEMLARASS surgery and rehabilitation at RECOUP had made a lot of difference in her and now she can walk with a walker’ — parents of D.

12. Case study 4

12.1. Pre-op status

(MA) A 13-year-old boy with spastic dystonic quadriplegia could not walk, even with support. His GMFCS level was IV.

12.2. Treatment

He underwent SEMLARASS in two stages: the first stage with OSCSS of bilateral hamstring, psoas, rectus femoris and gracilis along with bilateral femoral derotation and tibial derotation osteotomies and the second stage (after 8 weeks) with OSCSS of bilateral forearm flexors and pronators.

12.3. Current functional status

At a follow-up of 10 months, he was able to walk independently using walking frame. He is now able to climb up stairs with 50% assistance. His current GMFCS status was III (**Figure 17**).



Figure 17. Pre- and post-rehabilitation status of child MA.

‘Our son could only walk on toes with his knee bent and used to get a lot of pain and confined to bed. But now, after the surgery and intensive rehabilitation at RECOUP, he started walking independently with a walker and looking for a brighter future ahead’ — parents of MA.

13. Case study 5

13.1. Pre-op status

(R) A 5-year-old boy with spastic athetoid quadriplegia with bilateral dislocated hips was not able to sit or stand or use his hands. His GMFCS level was V.

13.2. Treatment

He underwent SEMLARASS with OSCSS of bilateral hamstring, psoas and rectus femoris along with femoral varus derotation osteotomy.

13.3. Current functional status

After 7 years of follow-up, he was able to stand with minimal support and walk with rollator independently. He was now able to climb up stairs with assistance and able to walk on ramp with rollator. His current GMFCS was III (**Figure 18**).



Figure 18. Pre- and post-rehabilitation status of child R.

‘Our son born with spastic athetoid quadriplegia till the age of 5 and was not able to sit, stand or walk; after surgery and intensive rehabilitation in RECOUP, our son is now able to sit and stand independently and walk with rollator more confidently’ – parents of R.

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