Pak. J. Biochem. Mol. Biol., 2012; 45(2): 104-111

Effects of rehabilitation on mild hypotonic diplegic cerebral palsy child with Down syndrome: an observational case study

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Abstract: The purpose of this study was to evaluate the effects of rehabilitation on Cerebral Palsy (CP) patient suffering from Down syndrome (DS). Al-Umeed Rehabilitation Association (AURA) was visited for this purpose for one month after the consent granted by AURA authorities. A thorough observational study of the behavior, performance and rehabilitation techniques was conducted for the period of one month on the patient suffering from the dual problem. The patient exhibited dominant characteristics of CP which were precipitated as perceptional deficits, motor impairments, lack of communication and disability to perform activities of daily living (ADL). The observed DS characteristics of the patient include Mongolian face, protruding tongue, hypotonia, flexible ligaments, excessive bending of legs, repetitive opening of mouth, bulging eyes, chest and ear infections, low concentration and cognition, small oral cavity and delayed speech with impaired fine motor skills. The rehabilitation program structured by AURA which intended to ameliorate his functional and mental deficits by providing US occupational physiotherapies, visual and auditory perception therapies, tactile perception therapies, communication and speech therapies, and therapies for ADL improvement. The resultant positive rehabilitation effects on the patient were evident from his annual examination reports evaluated by AURA in the form of improved muscle tone, visual perception, auditory perception, tactile perception, and communication and ADL performances. Despite the fact that a continuous rehabilitation is being provided to the subject which definitely has had an apparent holistic improvement but it should be borne in mind that the rehabilitation techniques produce a very gradual effect. No doubt that, these effects had a positive impact and it could be suggested that they could be extended to make the patient more productive towards motor, perceptional, cognitive and ADL performances, utilizing more effective tools.

Keywords: Cerebral Palsy, Down syndrome, rehabilitation.
Received: December 10, 2011 **Accepted:** April 20, 2012
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INRODUCTION

Cerebral Palsy (CP) is a disorder caused by the non-progressive biochemical changes in brain due to brain injury in the prenatal or postnatal brain developmental period resulting in physical deformities leading to physical disabilities, abnormal mental growth and movement disorders, while Down syndrome (DS) is a congenital chromosomal abnormality characterized by the presence of an extra copy of genetic material on the 21st chromosome (Hsa21)[1], either in whole (Trisomy 21)[2] or part (such as due to translocations).

The world wide prevalence of CP is approximately 2-2.5 per 1000 live births[3]. The classification of CP depends on the predominant motor alteration and encompasses three main types; spastic; having an unusual tightness in muscles of limbs, athetoid; characterized by involuntary movements and ataxic; characterized by difficulties in motor coordination. The mixed type involves characteristics of the two type's i.e (spastic and athetoidic) at the same time[4]. Depending on the impairment of the arms relative to the spastic legs, CP can further be categorized into hemiplegic CP (hCP) with unilateral limb disorder, diplegic CP (dCP) with greater impairment of the legs than of the arms, and tetraplegic CP (tCp) with all four limbs equally affected. Children with tCP also have mental retardation, while the intellect often is spared in dCP and hCP[5]. A number of causes stated in literature survey may include prematurity, perinatal asphyxia, and deficiency in maternal iodine[6]. Placental abruption, cord prolapse, and uterine rupture sharply heighten the risk of CP. Among other prenatal factors associated with increased risk of CP are intrauterine exposure to infection or maternal fever in labor, ischemic stroke, congenital malformation and atypical intrauterine growth (restricted or excessive for gestational age) and complication of multiple gestations[7]. Acquired cases in the postnatal period are usually related to central nervous system infection, trauma, strokes and severe hypoxic events such as near drowning[8]. Neurologically, deep gray matter and mostly white matter injuries are the most probable causes of CP.

The DS characteristic features may include physical, mental and clinical abnormalities; such as speech delay[9], delayed fine motor skills that often lag behind gross motor skills, impaired cognitive ability and Alzheimer's-type dementia[10]. Mental retardation[11], stunted growth, atypical fingerprints, separation of abdominal muscles, flexible ligaments, hypotonia, brachycephaly, smaller genitelia, eyelid crease, shortened extremities, oval palate, low-set and rounded ear, small teeth, flattened nose, clinodactyly, umbilical hernia, short neck, shortened hands, congenital heart disease, single transverse palmar crease, large protruding tongue, epicanthal fold, strabismus, brushfield spots[12,] hypothyroidism[13], gastrointestinal anomalies[14], infertility along with defects in spermatogenesis in men[15], hearing loss by effusion[16], and reduced blood pressure[17] are certain other consequences of the diseases. The possible

causes of DS can be a gene (or several genes) on HSA21 (on chromosome 21) independently contributing to the risk of DS[18]. Hsa21-derived micro-RNAs (miR-155, and -802) and proteins Methyl CpG binding protein 2 (MeCP2), CAMP responsive element binding protein 1 (CREB1), and Myocyte-specific enhancer factor 2C (MEF2C) are also over-expressed in DS brain. Improper repression of MeCP2, secondary to trisomic over-expression of Hsa21-derived miRNAs, may lead to the neurochemical changes in the DS brain[1]. 1 in 800 children have DS, and the frequency becomes even higher with the age of the mother. Mothers past 35 years of age have increased chances of having a child with DS[2]. Its prevalence is highly dependent on maternal age at gestation[19]. The aim of present observational study, was to visualize CP and DS characteristics in the patient to evaluate the effects of rehabilitation offered to him at AURA.

MATERIALS AND METHODS

Case report

A 5 years old child (US) with mental age of 2 months was recommended by the doctor to the Al-Umeed Rehabilitation Association (AURA) diagnosed as the patient of Cerebral Palsy Mild Hypotonic Diplegia with Down syndrome, accompanied by motor deficit, mainly affecting his left body side and lower limbs, in 2006. The mother was 39 years at the time of his birth and was suffering from Rheumatoid Arthritis during pregnancy. He was born on 13th January, 2001. He was diagnosed with CP and DS at the age of one month. Born prematurely with Lower Segment Cesarean Section (LSCS) was having low birth weight of 1.4 kg. No delay in the birth was reported and an immediate cry was heard after the delivery. He was kept in incubator for 15 days due to low birth weight. The child had delayed milestone history. He was suffering from the symptoms of DS accompanied by CP, as taken from his case history.

The patient had mild spasticity in lower limbs, audio-visual and tactile perception problems, persistent severe ear infection, non-verbal communication, with low concentration and cognition. Small oral cavity, protruding tongue, small ears, repetitive opening of mouth and Mongolian face were his physical DS characters. Hitting his head continuously with his right hand was an exceptional characteristic of US.

The child's rehabilitation program had been focusing for the improvement of mixed muscle tone, understanding deficits, abnormal hand function, aided walking, verbal communication, ADL skills. An additional focus was to work on his deficits such as vision, cognition, auditory and tactile disabilities employing different techniques. Physiotherapy, Occupational Therapy, Visual Perception Activities, Tactile Activities, Audio Perception Activities, Hydrotherapy Pool, and Communication Skills Training, were the different modes utilized in AURA to counteract the difficulties encountered by US. No invasive therapy such as injection of Botulinum toxin injections was reported by AURA.

Conducive rehabilitation therapies conducted for the patient at AURA

AURA offered variety of therapies depending upon the kind of abnormality experienced by the patient. The specific techniques that were offered to US were as follows:

A. Motor activity training program (MATP) using occupational and physiotherapies

The rehabilitation occupational and physiotherapies which worked on Fine and Gross motor skills for improvement of movements and mobility of US patient included the use of following apparatus:

a) Wedge: It was a bed like slope apparatus on which US was placed by his tummy side and his arms laid down on the floor and pressure was applied on his arms. Legs were straight and relaxed (as shown in figure 1). Wedge therapy was used as a stretching muscle technique for his whole body.

Figure 1: Therapy procedure of a US on wedge.

b) Tilt table: It was a type of a stature on which US was laid in a straight position; knees were tied with steel plate's gaiters to keep the leg straightened. Then the stature is kept in standing position to make the feet weight bearing (Figure 2). This enabled US to be more weight bearing and helped to improve the gait posture and walking pattern.

c) Abductor lock chair: An abductor lock chair was a chair having a knob like heightened lock in the chair base (Figure 3) to separate the legs of US apart while sitting.

d) Hammock (Rotating net): In the rotating swing net, US was placed with tummy support and the

swing was moved 'to and fro', 'backward or forward', or in any other direction. The speed of the swing was determined depending upon the requirement of US. It helped in muscle extension and straightening, muscle tone enhancement, muscle relaxation and balancing (Figure 4).

e) Ball therapy: The US was placed on a physioball in a sitting posture (Figure 5) and the ball was moved 'to and fro' and 'forward and backward' very gently. It helped in elongation of muscles. Hip joint movement was increased. The muscle tone improvement was also accomplished by this therapy.

Figure 2: Rehabilitation procedure of US on tilt table.

Figure 3: Abductor lock chair.

Figure 4: Hammock with tummy base.

B. Multisensation rehabilitation therapies

The therapies for the improvement of multisensational abilities of US were as follows:

Figure 5: Therapy procedure on a physioball.

Figure 6: Visual board having model objects.

Visual perception activities

US was brought to a Snozeleen Room (multisensory room) for these activities. There were different kinds of apparatus as visual board; object's models board, face expression board, clip board, playing board, and shapes of different objects and flowers etc (Figures 6). With the help of expression board US learnt to recognize different emotional expressions, learnt to locate things in the right place, hence their proper orientation. Clipper board was used for the color recognition and improvement of visual perception of US. In this patient had to sequence the clippers of different colors in an alternate pattern. Amusements with TV and videos were part of visual rehabilitation for US.

Auditory perception activities

Auditory boards having different objects' models were used to hear and recognize sound of different things as shown in Figure 7 (Table 1). US was made to learn and recognize different sounds like street noise, nursery rhymes, home sounds, sounds of different animals and birds with the help of audio cassettes.

Tactile perception activity

Modality table, was a round table pool filled with sand or water in which US was made to explore and identify the hidden objects, sense their texture and improve recognition by tactile perception. Sometimes it was facilitated by an abductor chair. The modality sand pool apparatus is shown in figure

8. Moreover, soft and hard brushes are applied on the patient's skin to let him feel the soft and hard texture differentiation.

Figure 7: Auditory board having auditory model objects.

Figure 8: Modality and pool apparatus.

Ball pool

A pool compartment filled with balls of different colors and at the corner had an abductor lock chair. Concealed objects among the balls were explored by the US (Figure 9) to improve his tactile perception deficits, color discrimination, visual perception deficits. US got relaxed by playing with the balls.

Activities of daily living (ADL)

Patients learn activities of daily living by practicing on different models of house hold things such as water tap, telephone, ringing door bell, plugging sockets, kitchen cabinets etc (Figure 10) to enable them to be able to get hands on to these objects.

C. Communication and speech therapies

US patients had communication and speech disability. Different therapies were applied to improve his communication. These included blowing and whistling activity to improve the opening and closure control of US. Sucking straw activity exercised different facial muscle.

Repeated pronunciation of words, Pecs communication technique aided with communication book, having pictures and emotions was also used (Figures 11-13) through which he could convey his needs in the form of picture sentencing. A light ladder was also used which illuminated with the sound production of US. Greater the intensity of sound produced by US, the upright glow was also intense. This therapy enabled him to improve his quality of sound and his speech.

Figure 9: Ball pool with patients.

Figure 10: Model objectives for activities of daily living with patient's activity.

Figure 11: Pec communication book developing concept of Yes/No.

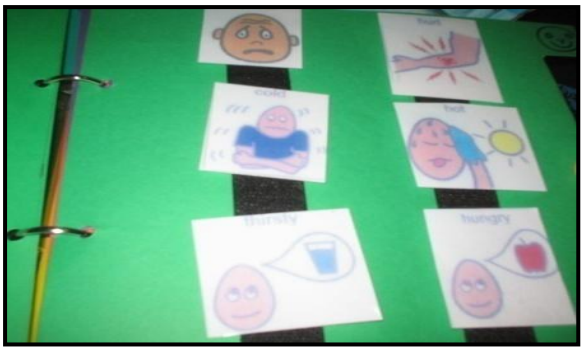

Figure 12: Pec book pictures for recognition of different perceptions.

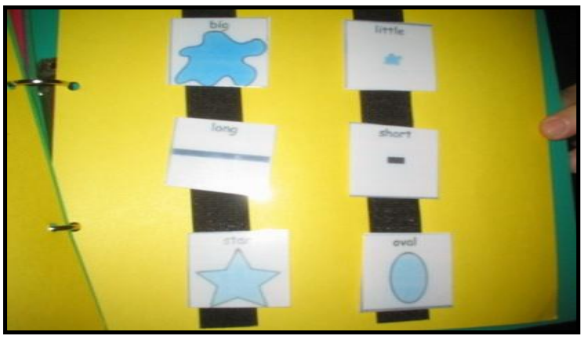

Figure 13: Identification of size of object.

RESULTS AND DISCUSSION

A thorough study of the Cerebral Palsy Mild Hypotonic Diplegic with Down syndrome patient "US" revealed that he was exhibiting inherited DS accompanied with Mild Hypotonic Diplegic CP characteristics. The risk of CP increases with decreasing gestational age, but the presence of maternal infection magnifies the risk. This may be due to the intrinsic vulnerability of the developing brain earlier in gestation[20, 21]; leading damage to the oligodendrocytes can impair myelintion resulting in the clinical manifestation of CP[22]. Hypoxia and infection both trigger synthesis and release of pro-inflammtory cytokines such as tumor necrosis factor-alpha (TNF-α), interleunkin-1 (IL-1), interleukin-6 (ILKK-6), as well as free radicals and nitric oxide which all might play role in the precipitation of CP[23].

Many other causes of CP stated in literature survey may include prematurity, perinatal asphyxia, and deficiency in maternal iodine[6]. Placental abruption, cord prolapsed, and uterine rupture sharply heightens the risk of CP. Among other prenatal factors associated with increased risk of CP are intrauterine exposure to infection or maternal fever in labor, ischemic stroke, congenital malformation and atypical intrauterine growth (restricted or excessive for gestational age) and complication of multiple gestations[7]. Acquired cases in the postnatal period are usually related to central nervous system infection, trauma, strokes, and severe hypoxic events such as near drowning[8].

Neurologically, deep gray matter and mostly white matter injuries are the most probable causes of CP. But in US no such possible causes were reported for the manifestation of CP except low birth weight evident from his case history provided by the authorities. In depth neurological evaluation might help us to evaluate the cause.

Table 1: Disease characteristics of the observed patients.

Cerebral Palsy Characters	Down syndrome Characters
Mild spasticity in lower limbs	Bulging eyes
No verbal communication	Short ears
Audiovisual perception problem	Delayed speech and fine motor skills
	Mongolian face
Induced movement therapy applied (right hand gaiters) to use left hand	Severe ear infection
Non – weight bearing	Chest congestion
Unable to walk without support	Protruding tongue
Perception problems	Cyanosis blockage
Problem in activities of daily living	Low concentration and cognition
	Repetitive opening of mouth
	Small oral cavity
	More use of functional right hand which is controlled by induced gaiter therapy (fig.18) and bending of legs and right arm despite spasticity.

The overall physical and mental growth in US was impaired. Weak immune response, slow recovery rate and persistent ear and chest infection in the subject (US) could be manifested due to protein deficiency malnutrition as US faced difficulties in eating and swallowing which is a pertinent character of the CP. Proteins are important for the growth, immune function and recovery from diseases, as well as for skeletal respiratory muscle function; which all become severely impaired when levels are low[24]. The hypoxic-ischemic encephalopathy includes specific and well-known patterns of brain injury[25,26] that interfere with the frontal/insular-basal ganglia-brainstream swallowing pathway[27-37].

Table 3: Rehabilitation therapies provided to US and their impact on his improvement.

Problems	Types of rehabilitation therapies	Impact
Motor activity	Physiotherapy and occupational therapies Physioball therapy Joint exercise Joint's weight bearing exercises Independent stool sitting Dynamic sitting Tailer sitting Platform Swinging Hammock Therapy Tilt Table Therapy	The spasticity was reduced and muscle tone is improved by different occupational and physiotherapies
Visual perception	Visual board with object models Face expression board Clipping board TV and videos	US was able to recognize basic colors and size of objects with the visual perception therapies
Auditory perception	Auditory model board Audio tapes and cassettes	US was able to respond to different sounds such as cat's sound and calling his name after the audio perception therapies
Tactile perception	Modality table Hot/cold perception Tactile brush applied to body parts	US was able to respond to hot/cold things, can also give the rough/smooth touch concept when tactile brush is applied
Communication	Blowing and whistling activity Sucking straw activity Repeated pronunciation of words Pec communication techniques	The verbal communication of patient improved learnt to recite Bismillah and Dua in Urdu and also able to pronounce certain words
Adl	Practicing on different models of house hold things. Washing hand/face. Brushing hair/teeth Dressing/undressing	Activities of daily living also improved after the rehabilitation ADL therapies

The subject had perception problems. The visual impairment as observed in the US patient may be explained in terms of the two higher visual pathways which have been described in primate brains. The dorsal stream passes between the occipital lobes and the posterior parietal lobes, and is involved in the analysis of the whole visual scene, as in attending to elements within the scene (along with the frontal lobes). It facilitates immediate visual guidance of movement through the scene, by interacting with area V5 (or the middle temporal lobes MT), the area of the brain responsible for processing perception of motion. The ventral stream passes between the occipital lobes and the temporal lobes where visual representations of the external world are stored. Functions of the ventral system lead to the recognition of faces, shapes, objects, and routes by matching visual input with their stored representations. The dorsal visual system is more vulnerable to insult occurring early in life than the ventral one, so that CP children have been reported to find it more difficult to detect motion coherence than form coherence[38], and using body-centered coordinates to locate objects rather than object-centered coordinates[39].

Cognitive impairment observed in US is his DS phenotypic character. Literature survey has shown that DS patients exhibit general decrease in brain weight (a function of cerebral and cerebellar atrophy, shortened occipital lobes, and narrowing of the superior temporal gyri), and particular deficits in pyramidal cell dendritic arborization and dendritic spine architecture[40, 41]. For example, dendritic branching and spine counts in the hippocampus[42, 43], in layers III and V of visual cortex[44, 45], and in layers of the motor cortex[46, 47], are dramatically reduced in people with DS. Moreover, the remaining spines in the visual and motor cortices adopt irregular morphologies: some spines appear long and tortuous, while others develop enlarged heads (all features of synaptic contacts with reduced plastic or learning potential[47, 44]. The striatum also represents the main input station of the basal ganglia and plays a critical integrative role within the basal ganglia circuit, receiving neural inputs from several brain areas and leading to the acquisition of motor and cognitive action sequences. However, we did not have any information with reference to morphological characteristics of this patient's brain. We could suggest that reduced cognition in US could be due to nonfunctional areas of the brain. Altered cholinergic transmission also might play a critical role in the pathophysiology of motor and cognitive deficits in DS, leading to an abnormal processing of neuronal inputs within the basal ganglia[48].

Patient's tactile and sensation problems might be due to significant alterations in white matter fibers connection to sensory cortex (corona radiate and internal capsule), suggesting that injuries might reflect disruption of sensory as well as motor connection [49, 50]. The patient encountered only some of the dominant CP characters while others such as spasticity, movement disability were partially overcome by DS characteristics. Moreover, he used

his functional right hand more than left, as is a common observation in CP patients that they use their Paretic arm less than normal[51]. An induced gaiter therapy on his right limb is used to let him use his left limb normally (as shown in fig. 18, table.2). Besides these variant diseased characters of CP and inherited DS, the main problems of the Subject at the time of admission in AURA institute were: mixed tones, poor understanding and impaired cognitive ability, walking problem, speech problem, and ADL problem. The main aims of treatment set by the AURA therapists and specialists were: to normalize the tone, to improve understanding, to improve left hand function, to improve walking, verbal communication, his perceptions, and ADL skills.

To control all his disabled characters of CP and DS both and achieve the main aims of treatment set by the therapists, different rehabilitation therapies including occupational and physiotherapy (ball therapy, joint exercises, joint's weight bearing exercises, independent stool sitting, dynamic sitting, tailor sitting, platform swinging, rotating net therapy and tilt table therapy), visual perception therapies (visual board with object models, face expression board, clipping board, TV and videos), auditory perception therapies (auditory model board, auditory tapes and cassettes), tactile perception therapies (modality table, hot/cold perception, tactile brush), communication and speech therapies (Pecs communication book, blowing and whistling activity, sucking straw activity, repeated pronunciation therapy), and therapies for ADL improvement (practicing on different models of household things, washing hand/face, brushing hair/teeth and dressing/undressing activities) (all apparatus shown in Figures 1-13) have been provided to him in AURA since he was 5 years old and are still on the go. The positive rehabilitation effects on the patient were evident from his annual examination reports not attached in the present paper.

A prolonged follow-up of the patient US at AURA would help us to reevaluate the effectiveness of the different kind of rehabilitations applied to the subject. In-depth imaging techniques of the brain would provide an insight to the morphological alterations in the examined subject.

ACKNOWLEDGMENT

The authors are thankful to Dr. Shabana (Principal-AURA) who extended their help during observations and their interpretation. Heartly thanks to the Al-Umeed Rehabilitation Association (AURA) authorities for granting the consent to conduct a thorough observational study of the patient US in their institute. The photographs of the patient are printed with their consent.

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