



Profiling of Communication and Motor Skills in Post-Neonatal Cerebral Palsy- A Case Report

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Abstract

Status epilepticus is a medical emergency associated with significant mortality and major morbidity. This article aimed to profile the assessment and management conditions of a child with post neonatal CP due to status epilepticus in communication and motor skills. A 14 year old female child served as the subject. She was diagnosed as post neonatal cerebral palsy due to status epilepticus at the age of 4.6 years. However, the management of speech-language and physiotherapy had begun at the age of 14 years. The study also highlights on the fact about delay to early rehabilitation and progress in the overall functional abilities of the child. Focus has also been on the team management as an effective rehabilitation strategy in restoring the functional abilities in the child.

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Introduction

Cerebral Palsy (CP) is a cluster of clinical signs which includes abnormal muscle tone and movement, and associated loss of function due to a non-progressive lesion or abnormality of brain. It displays a variability of disability profiles in cognitive performance, communication and behaviour, epilepsy, visual-spatial or perceptual problems, or a combination of these features (Himmelmann, Beckung, Hagberg & Uvebrant, 2006; Shevell, Dagenais & Hall, 2009). A variety of prenatal and perinatal risk factors have been identified for CP including low birth weight, premature delivery, infections during pregnancy, jaundice and so on. Apart from these risk factors that occur prior to or around the time of birth, post natal factors can also lead to CP, which is termed as post neonatal CP. CP of post neonatal origin is defined by the presence of a specific event or episode that happened after 28 days of age (Cans et al., 2004; Odding, Roebroek & Stam, 2006). Various viral infections, especially the acute exanthematous diseases of childhood, may be accompanied by, or followed by, clinical manifestations of widespread involvement of the Central Nervous System (CNS) which is often far more serious than the primary infection. This can even result in post-infectious (or para-infectious) encephalomyelitis or immune mediated encephalomyelitis, a serious condition which needs to be taken care immedi-

ately. It is indicated that bacterial meningitis is one of the leading postnatal cause of developmental disabilities. Other factors like head injury, paediatric stroke, epileptic attack can also lead to post neonatal CP (Paneth, 1993; Odding, Roebroek & Stam, 2006).

Of these risk factors, Status epilepticus (SE) is a medical emergency associated with significant mortality and major morbidity which presents with continuous seizure lasting more than 30 min, or two or more seizures without full recovery of consciousness between any of them (Lowenstein & Alldredge, 1998). Longer duration of seizures is associated with increased risk of morbidity and mortality. Delayed treatment may increase the frequency of SE. Therefore, early seizure termination is paramount for the treatment of SE (Eriksson & Koivikko, 1997). This is followed by early management options. Various disorders with communication and physical disability remain unchanged due to lack of early intervention. Thus, rehabilitation at the earliest is an important factor to be noted in the prognosis of disordered population. Patients with no prior history of epilepsy who present in status epilepticus are most likely to have meningitis, encephalitis, hypoxic-ischemic encephalopathy, or a large stroke. This may be accompanied by, or followed by, clinical manifestations of widespread involvement of the CNS. The prognosis for these

patients is worse. The longer a patient is in status, the worse the outcome (Brazier & Coceani, 1976). This is partly because the duration is more likely to be prolonged in patients with more severe underlying disease, but there is also a great deal of experimental evidence that status epilepticus itself damages the brain and that the degree of damage is a function of duration. There are some evidences that convulsive status epilepticus (CSE), especially febrile CSE, might cause hippocampal injury (Raspall-Chaure, Chin, Neville & Scott, 2006).

Sadarangani et al. (2008) provided a data on the incidence and outcome of convulsive status epilepticus which stated that prevention and appropriate early management of seizures might reduce the incidence and improve the outcome of convulsive status epilepticus in children. Few studies reported that longer seizure duration, cerebral insult, and refractory convulsive status epilepticus are strongly associated with poor outcomes (Legriell et al., 2010). Specific learning difficulties have been reported in children who have febrile convulsions which include problems with drawing and arithmetic and a delay in speech development. Behavior disorders and attention deficits have also been found after febrile convulsions. The incidence of neurologic sequelae and mental retardation was reported to be higher when the first febrile convulsion occurred at an early age (Verity, Greenwood, & Golding, 1998). The characteristic features pave way for a holistic and multifaceted early rehabilitative process.

Transdisciplinary approach includes the sharing of roles across disciplinary boundaries so that communication, interaction, and cooperation are maximized among team members (Davies, 2007; Johnson et al., 1994). This approach adopts a framework for allowing members of an educational team to contribute knowledge and skills, collaborate with other members, and collectively determine the services that most would benefit a child. This would more beneficial when rehabilitation services are provided at an earlier age. The goals of early rehabilitation are to maintain functioning or to minimize the loss of functioning and to optimize recovery and early autonomy. The maintenance or early restoration of functioning is of particular importance in patients at risk (Johnston, Wood & Fiedler, 2003).

Profiling a condition like this in terms of its characteristics, mainly communication and motor skills would be an effective tool to track the prognosis. Thus, the present study was aimed at profiling the condition of post neonatal CP due to status epilepticus in a 14 year old child and highlighting the importance of early team rehabilitation.

Method

Participants

The study was conducted after receiving informed ethical consent from the participant's parent. The participant in this study, 'AZ', was 10 years old when she was brought to our institute. She had an attack of febrile seizures at the age of 4.6 years. At the time of her illness onset, she was enjoying the rhymes and plays of kindergarten and as reported by the parents the child's receptive and expressive language age was as similar to a 4 year old child. There was no prior medical history of note. AZ was initially admitted to a local hospital following high fever (102⁰F) with seizures. She was in a complete unconscious state. She was kept in ICU for 2 weeks and followed by a week in ventilator. Asleep EEG report reveals generalized epileptiform discharges and bihemispheric dysfunction. She was medically diagnosed as status epilepticus secondary to encephalitis sequelae. She had several episodes of very high temperatures with seizures for the next 2 years. She was under medications which included Tegretol (5 ml) and Epilex (8 ml) for the attack of seizures. However, there was no marked difference in the episodes of seizures and she was discharged from the hospital. Later, medications such as Parkin (2 mg), Lamictal (50 mg) and Epilexin (8 ml) were included.

10 years after discharge, the child was brought to our center with the complaint of poor speech and language skills and inability to walk and stand. She presented with very limited responses to external sensory stimuli and was completely dependent for her activities of daily living. A detailed speech-language, physical, psychological and neurological evaluations were carried out. On speech and language evaluation, she was unable to vocalize meaningfully and could only express her needs through crying and growling. Oromotor and vegetative skills were affected. The child exhibited severe drooling. Prelinguistic behaviors such as eye contact and attention and concentration were poor. She also presented with difficulty in swallowing and choking.

Frequent attack of seizures about two times a day was present. Motor evaluation revealed she was wheel chair bounded and was hypotonic except Tendo Achilles (TA). Poor psychomotor coordination and impaired sitting balance were other associated features present. Despite these, she exhibited overflow of movements when her body hits somewhere, sleeplessness and unable to sit in one place for more than few minutes. Psychological evaluation using Vineland Social Maturity Scale was carried out to evaluate the social competence of the child and revealed a functional level of 1.6 years. On neurological evaluation, the child was given a diagnosis of post encephalitic sequelae.

Procedure and Analyses

Comprehensive speech and language evaluations were done. Receptive Expressive Emergent Language Scale (REELS) was initially used to obtain a screening level of the receptive and expressive language age of the child. The scores were found to be between 4-5 months for both reception and expression. To elaborate, child was able to localize familiar voice and could respond to her own name by producing the sound /m/ with 50% consistency (5/10 times). The child was also able to produce vowel like sounds similar to /o/ and /u/ in response to vocal play. She exhibited poor response to name call and poor communication intent. Pre-linguistic skills were relatively poor. The child was not able to attend either to visual or auditory stimuli for more than 4 seconds. The child's attention and concentration to speech stimuli (3-4 sec) were also limited. Informal visual assessment reveals poor visual attention and visual scanning.

The child also presented with complex array of oro-motor and vegetative issues such as like drooling because of poor lip closure and feeding issues. On oro motor evaluation, lip seal was affected and excessive drooling was present. Usually, the drool drips off chin and onto clothing which suggests severe drooling. She had an open mouth posture all the time, even when a food was placed inside the mouth. The protrusion, elevation, lateralization functions of the tongue was severely affected. Tongue movements were limited for bolus formation. She had difficulty in biting and chewing solid food. She was generally on semisolid diet, fed using a syringe. She had difficulty in blowing and sucking also. On administration of the Com-DEALL Oro Motor Checklist (Archana, 2008), a total score of 10 was obtained, indicating poor oro-motor skills. The scores obtained in individual subsections of the checklist displayed greater difficulties in the lip movements and speech.

Motor Assessment Scale (MAS) to evaluate the motor functions of the child and Berg Balance Scale (BBS) to test the static and dynamic balance abilities was used in physiotherapy assessment. MAS Score of grade 4 and BBS score of 23 was obtained and the scores reveals TA tightness, poor static and dynamic balance and poor posture. Overall, the results indicated a generalized and severe communicative and motor regression. On the basis of these assessment results, AZ was diagnosed as Global regression secondary to post neonatal CP (encephalitis sequelae) and was enrolled for speech language and physical rehabilitation at the institute. Given evidence to severe communication and physical difficulties, it was indispensable to introduce a trans-disciplinary approach. As the main concern of AZ's parents was communication and motor movements, the focus was enforced on restorative and compensatory speech-language therapy and physiotherapy

goals. The speech-language therapy focused to:

- enhance pre-linguistic skills attention and concentration span, visual stimulation
- enhance communicative responses to auditory stimuli such as response to name call
- enhance comprehension of daily communicative vocabulary and simple commands, greetings
- enhance vocalizations and expressions of basic needs
- enhance reduce drooling, improve lip closure, and oromotor strength

And supportively, the physiotherapy goals targeted on balance training, positioning (better seating and neck control) to reduce the TA tightness and to facilitate active or dynamic movements.

Therapy was provided regularly for 3 days per week for 45 minutes. The child attended speech-language therapy and physiotherapy for 8 months continuously. With evidence of diminished drooling and good balance, a re-evaluation was carried out after 8 months. A slight degree of improvement was evident. The drooling has decreased but it generally depended on the attack of seizures and associated medical issues. The severity of drooling has decreased as it doesn't drip onto clothing. The lip seal has partially improved with lip strengthening exercises. Attention and concentration span has improved to 6-8 seconds. Visual scanning, especially to glowing objects presented in a contrastive background has improved. Localising by slight head turn is present for name call. Vocalisations such as /a/ and /u/ in response to 'hello' has emerged with 50 % consistency. Receptive Expressive Emergent Language Scale (REELS) was carried out and a language age of 5-6 months (scattered) was obtained for both receptive and expressive age. A total score of 14 was obtained on administering the Com-DEALL Oro Motor Checklist suggesting slight progress in the oromotor skills, mainly in jaw and tongue movements.

The re-assessment done in physiotherapy using Motor Assessment Scale and Berg Balance Scale reveals a significant improvement in motor functions and balance abilities. TA tightness has reduced and other improvements in trunk balance and control and range of motion are appreciable. Results of review assessment are shown in Table 1 and 2.

Comparing pre and post therapeutic results, it is evident that the goals taken up for therapy were significant as there was improvement in all the targeted areas. Pre and post comparison result are shown in Table 3 and 4. Although there was 8

Table 1: Review assessment result of speech language therapy

Sl.No	Goals focussed	Review Assessment Result
1	Drooling	Partially improved- drool doesn't drips off onto clothing
2	Lip closure	Tries to do /m̂/ for closing the lips when verbal prompts are given
3	Attention and concentration	Improved to 6-8 seconds Visual scanning has improved
4	Response to name call	50% consistency with unfamiliar voices also localises by head turning, Smiling present
5	Communication intent	Started looking for the object which express a need to have an object 40% consistency
6	Vocalisation	Started producing /a/ and /u/ in response to 'hello' with 50% consistency

Table 2: Review assessment result of physiotherapy

Sl.No	Goals focussed	Review Assessment Result
1	TA tightness	Tightness reduced Post MAS score of grade 1+
2	Balance training	Trunk balance and control improved Sitting: Dynamic balance improved Standing: Standing with minimal support improved. Post BBS score of 48
3	Posture Correction	Full range of motion is present. Quadripodal position improved. Kneeling with minimal support present. Initiation of active or dynamic movements Improvement in heel strike and stance phase in gait

Table 3: Pre and post therapeutic results in speech language therapy

Sl.No	Goals focussed	Pre therapy	Post therapy
1	Drooling	Frequently drool drips off chin & onto clothing	Partially improved with regular massaging of cheeks
2	Lip closure	Open mouth posture	Tries to do /m̂/ for closing the lips with lip strengthening exercises when verbal prompts are given
3	Attention and concentration	Attention span was around 3-4 seconds Poor visual scanning	Improved to 6-8 seconds Visual scanning has improved with visual stimulation activities
4	Response to name call	50% consistency with familiar voices No head turn for response to name call	50% consistency with unfamiliar voices with the use of modelling technique Localises by head turning Smiling present
5	Communication intent	Cries to express her needs	Started looking for the object which expresses a need to have an object with 40% consistency using modelling technique
6	Vocalisation	Produce vowel like sounds similar to /o/ and /u/ in response to vocal play	Started producing /a/ and /u/ in response to 'hello' with 50% consistency with the use of modelling technique

Table 4: Pre and post therapeutic results in physiotherapy

Sl.No	Goals focussed	Pre therapy	Post therapy
1	TA tightness	MAS score of grade 4	Tightness reduced MAS score of grade 1+
2	Balance training	Poor static and dynamic balance Poor posture BBS score of 23	Trunk balance and control improved Sitting: Dynamic balance improved Standing: Standing with minimal support improved. BBS score of 48
3	Posture correction	Limited range of motion	Full range of motion is present. Quadripodal position improved. Kneeling with minimal support present Initiation of active or dynamic movements Improvement in heel strike and stance phase in gait

months interval between the assessments, the minimal progress suggests impact of transdisciplinary rehabilitation. The significant improvements made in balance and position provided a facilitatory benefit to the drooling control and enhanced vocalizations. Similarly, the activities targeted to improve the neck control eventually helped in the speech-language therapy during the massaging and strengthening of the lips and cheeks muscles. The speed in progress is suggestively propositional to the age at which rehabilitation is initiated. So the delay in rehabilitation in our case could be a reasonable factor for the slow progression in overall development. These results correspond to the outcome of the systematic intervention strategies of speech-language and physical rehabilitation. Further, extensive control in the limbs and seating exercises is a prognostic indicator for the advances overall communication skills in the child.

Conclusions

This study profiled the condition, post neonatal cerebral palsy due to status epilepticus in speech-language and motor skills. The profiling offers an understanding that the condition can remain unchanged if appropriate and effective management strategies are not being taken at the right time. It provides an insight that a subtle improvement would be more appreciable than a state being unchanged. The article has also highlighted the importance of team management in bringing back the lost functional abilities and quality of life. As a future direction, the use of Augmentative and Alternative Communication (AAC) would be suggested in maintaining the fundamental communication.

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