

Speech-4

by

FILE	SP_04_MANUSCRIPT.DOCX (51.66K)		
TIME SUBMITTED	06-OCT-2015 06:19PM	WORD COUNT	3031
SUBMISSION ID	580711922	CHARACTER COUNT	17801

1 **Introduction**

2 Cerebral Palsy (CP) is a cluster of clinical signs which includes abnormal muscle tone
3 and movement, and associated loss of function due to a non-progressive lesion or abnormality
4 of brain. It displays a variability of disability profiles in cognitive performance,
5 communication and behaviour, epilepsy, visual-spatial or perceptual problems, or a
6 combination of these features (Himmelman, Beckung, Hagberg & Uvebrant, 2006; Shevell,
7 Dagenais & Hall, 2009). A variety of prenatal and perinatal risk factors have been identified
8 for CP including low birth weight, premature delivery, infections during pregnancy, jaundice
9 and so on. Apart from these risk factors that occur prior to or around the time of birth, post
10 natal factors can also lead to CP, which is termed as post neonatal CP. CP of post neonatal
11 origin is defined by the presence of a specific event or episode that happened after 28 days of
12 age (Cans et al., 2004; Odding, Roebroeck, & Stam, 2006). Various viral infections,
13 especially the acute exanthematous diseases of childhood, may be accompanied by, or
14 followed by, clinical manifestations of widespread involvement of the Central Nervous
15 System (CNS) which is often far more serious than the primary infection. This can even
16 result in post-infectious (or para-infectious) encephalomyelitis or immune mediated
17 encephalomyelitis, a serious condition which needs to be taken care immediately. It is
18 indicated that bacterial meningitis is one of the leading postnatal cause of developmental
19 disabilities. Other factors like head injury, paediatric stroke, epileptic attack can also lead to
20 post neonatal CP (Paneth, 1993; Odding, Roebroeck & Stam, 2006).

21
22 Of these risk factors, Status epilepticus (SE) is a medical emergency associated with
23 significant mortality and major morbidity which presents with continuous seizure lasting
24 more than 30 min, or two or more seizures without full recovery of consciousness between

25 any of them (Lowenstein & Alldredge, 1998). Longer duration of seizures is associated with
26 increased risk of morbidity and mortality. Delayed treatment may increase the frequency of
27 SE. Therefore, early seizure termination is paramount for the treatment of SE (Eriksson &
28 Koivikko, 1997). This is followed by early management options. Various disorders with
29 communication and physical disability remain unchanged due to lack of early intervention.
30 Thus, rehabilitation at the earliest is an important factor to be noted in the prognosis of
31 disordered population.

32 Patients with no prior history of epilepsy who present in status epilepticus are most
33 likely to have meningitis, encephalitis, hypoxic-ischemic encephalopathy, or a large stroke.
34 This may be accompanied by, or followed by, clinical manifestations of widespread
35 involvement of the CNS. The prognosis for these patients is worse. The longer a patient is in
36 status, the worse the outcome (Brazier & Coceani, 1976). This is partly because the duration
37 is more likely to be prolonged in patients with more severe underlying disease, but there is
38 also a great deal of experimental evidence that status epilepticus itself damages the brain and
39 that the degree of damage is a function of duration. There are some evidence that convulsive
40 status epilepticus (CSE), especially febrile CSE, might cause hippocampal injury (Raspall-
41 Chaure, 2006).

42
43 Sadarangani et al. (2008) provided a data on the incidence and outcome of convulsive
44 status epilepticus which stated that prevention and appropriate early management of seizures
45 might reduce the incidence and improve the outcome of convulsive status epilepticus in
46 children. Few studies reported that longer seizure duration, cerebral insult, and refractory
47 convulsive status epilepticus are strongly associated with poor outcomes (Legriel, Azoulay,
48 Resche-Rigon, Lemiale, Mourvillier, & Kouatchet, 2010). Specific learning difficulties have

49 been reported in children who have febrile convulsions which include problems with drawing
50 and arithmetic and a delay in speech development. Behavior disorders and attention deficits
51 have also been found after febrile convulsions. The incidence of neurologic sequelae and
52 mental retardation was reported to be higher when the first febrile convulsion occurred at an
53 early age (Verity, Greenwood, & Golding, 1998). The characteristic features pave way for a
54 holistic and multifaceted early rehabilitative process.

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

64
65 Profiling a condition like this in terms of its characteristics, mainly communication
66 and motor skills would be an effective tool to track the prognosis. Thus, the present study was
67 aimed at profiling the condition of post neonatal CP due to status epilepticus in a 14 year old
68 child and highlighting the importance of early team rehabilitation.

69 **Case Report**

70 The participant in this study, "AZ", was 10 years when they reported to our institute.
71 She had an attack of febrile seizures at the age of 4.6 years. At the time of her illness onset,

72 she was enjoying the rhymes and plays of kindergarten. There was no prior medical history of
73 note.

74

75 AZ was initially admitted to a local hospital following high fever (102⁰ F) with
76 seizures. She was in a complete unconscious state. She was kept in ICU for 2 weeks and
77 followed by a week in ventilator. Asleep EEG report reveals generalized epileptiform
78 discharges and bihemispheric dysfunction. She was medically diagnosed as status epilepticus
79 secondary to encephalitis sequelae. She had several episodes of very high temperatures with
80 seizures for the next 2 years. She was under medications which included Tegretol (5 ml) and
81 Epilex (8 ml) for the attack of seizures. However, there was no marked difference in the
82 episodes of seizures and she was discharged from the hospital. Later, medications such as
83 Parkin (2 mg), Lamictal (50 mg) and Epilexin (8ml) were included.

84

85 10 years after discharge, the child was brought to our center on with the complaint of
86 poor speech and language skills and inability to walk and stand. She presented with very
87 limited responses to external sensory stimuli and ¹⁴ was completely dependent for her activities
88 of daily living. A detailed speech-language, physical, psychological and neurological
89 evaluations were carried out. On speech and language evaluation, she was unable to vocalize
90 meaningfully and exhibited severe drooling. She also presented with difficulty in swallowing
91 and choking. Frequent attack of seizures about two times a day was present. Motor evaluation
92 revealed she was wheel chair bounded and was hypotonic except Tendo Achilles (TA). Poor
93 psychomotor coordination and impaired sitting balance were other associated features
94 present. Despite these, she exhibited overflow of movements when her body hits somewhere,
95 sleeplessness and unable to sit in one place for more than few minutes. Psychological
96 evaluation using Vineland Social Maturity Scale was carried out to evaluate the social

97 competence of the child and revealed a functional level of 1.6 years. On neurological
98 evaluation, the child was given a diagnosis of post encephalitic sequelae.

99

100 **Speech language Evaluation**

101 Comprehensive speech and language evaluations were done. Receptive Expressive
102 Emergent Language Scale (REELS) was initially used obtain a screening level of the
103 receptive and expressive language age of the child. The scores was found to be between 4-5
104 months for both reception and expression. To elaborate, child was able to localize familiar
105 voice and could respond to her own name by producing the sound /m/ with 50% consistency
106 (5/10 times). The child was also able to produce vowel like sounds similar to /o/ and /u/ in
107 response to vocal play. She exhibited poor response to name call and poor communication
108 intent. Pre-linguistic skills were relatively poor. The child was not able to attend to stimuli for
109 more than 4 seconds. She has poor attention and concentration to speech stimuli (3-4 sec).
110 Informal visual assessment reveals poor visual attention and visual scanning.

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

120 was obtained, indicating poor oromotor skills. The scores obtained in individual subsections
121 of the checklist displayed greater difficulties in the lip movements and speech.

122 **Physiotherapy Evaluation**

123 Motor Assessment Scale (MAS) to evaluate the motor functions of the child and Berg
124 Balance Scale (BBS) to test the static and dynamic balance abilities was used in
125 physiotherapy assessment. MAS Score of grade 4 and BBS score of 23 was obtained and the
126 scores reveals TA tightness, poor static and dynamic balance and poor posture.

127 Overall, the results indicated a generalized and severe communicative and motor
128 regression. On the basis of these assessment results, AZ was diagnosed as Global regression
129 secondary to post neonatal CP (encephalitis sequelae) and was enrolled for speech language
130 and physical rehabilitation at the institute.

131 **Rehabilitation profile**

132 Given evidence to severe communication and physical difficulties, it was
133 indispensable to introduce a transdisciplinary approach. As the main concern of AZ's parents
134 was communication and motor movements, the focus was enforced on restorative and
135 compensatory speech-language therapy and physiotherapy goals. The speech-language
136 therapy focused to:

- 137 a) enhance pre-linguistic skills attention and concentration span, visual stimulation,
- 138 b) communicative responses to auditory stimuli such as response to name call
- 139 c) comprehension of daily communicative vocabulary and simple commands,
140 greetings
- 141 d) vocalizations and expressions of basic needs
- 142 e) reduce drooling, improve lip closure, and oromotor strength

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

146 **Review Assessment**

147 With evidence of diminished drooling and good balance, a re-evaluation was carried
148 out after 8 months. A slight degree of improvement was evident. The drooling has decreased
149 but it generally depended on the attack of seizures and associated medical issues. The
150 severity of drooling has decreased as it doesn't drip onto clothing. The lip seal has partially
151 improved with lip strengthening exercises. Attention and concentration span has improved to
152 6-8 seconds. Visual scanning, especially to glowing objects presented in a contrastive
153 background has improved. Localising by slight head turn is present for name call. Receptive
154 Expressive Emergent Language Scale (REELS) was carried out and a language age of 5-6
155 months (scattered) was obtained for both receptive and expressive age. A total score of 14
156 was obtained on administering the Com-DEALL Oro Motor Checklist suggesting slight
157 progress in the oromotor skills, mainly in jaw and tongue movements.

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

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166 Table 1.

167 *Review assessment result of speech language therapy*

S.No	Goals focussed	Review Assessment Result
1	Drooling	Partially improved- drool doesn't drip off onto clothing
2	Lip closure	Tries to do /m/ 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

168

169 Table 2.

170 *Review assessment result of physiotherapy*

S.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

171

172 Comparing pre and post therapeutic results, it is evident that the goals taken up

173 for therapy were significant as there was improvement in all the targeted areas. Pre and post

174 comparison result are shown in Table 3 & 4. Although there was 8 months interval between

175 the assessments, the minimal progress suggests impact of transdisciplinary rehabilitation. The

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

185 Table 3.

186 *Pre and post therapeutic results in speech language therapy*

S.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 /m/ 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 also using 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

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191 Table 4.

192 *Pre and post therapeutic results in physiotherapy*

S.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

193

194 **Conclusion**

195 This study profiled the condition, post neonatal cerebral palsy due to status epilepticus
196 in speech-language and motor skills. The profiling offers an understanding that the condition
197 can remain unchanged if appropriate and effective management strategies are not being taken
198 at the right time. It provides an insight that a subtle improvement would be more appreciable
199 than a state being unchanged. The article has also highlighted the importance of team
200 management in bringing back the lost functional abilities and quality of life. As a future
201 direction, the ¹³ use of Augmentative And Alternative Communication (AAC) would be
202 suggested in maintaining the fundamental communication.

203

204

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