

Speech and Language Profiling of a Young Adolescent Diagnosed with Moya-Moya Disease

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Abstract

Background: Moya-Moya disease is leading condition of stroke in children and can affect children as young as 2 years to middle aged adults.

Aim: Speech language profiling was carried out in the client after eliciting detailed on Neuro-imaging and intervention. The client reported difficulty on the expression domain and reduced clarity of speech. Western Aphasia Battery, a diagnostic test to diagnose Aphasia (verbal impairment associated with stroke/trauma/tumour) was administered and the diagnosis of Broca's aphasia was made. Following this treatment was provided for a period of 3 months. Though the client dropped out from therapy, he showed considerable improvement and was able to sustain the progress after the period of 3 years also.

Keywords: facilitation; inhibition; paraphasia; competition

Introduction

Moya-Moya disease is a very rare condition considering its prevalence. It is a progressive condition. The literal meaning of Moya-Moyain Japanese is a "puff of smoke". The other important feature of this condition is that there is narrowing of arteries and also tangling of the blocked arteries. This tangling of the blood vessels would resemble the puff of smoke; hence the condition is called as Moya-Moya Disease [1,2,3]. The tangling of the arteries is viewed as a compensatory mechanism to regulate the blood flow. At the inception the condition was seen in Japan, however in the due course the condition was seen globally and is viewed as a leading cause of childhood stroke [4,5].

Moya-Moya Disease is associated with an initial presentation of transient ischemic attack or mini stroke which may be recurrent in nature [6]. Sometimes, it can also be associated with symptoms like headache or seizures, however these symptoms are very rare and may not be seen in all children affected by this condition. The exact cause of the condition is obscure and unclear and the condition is reported in children as young as 3 years to middle aged adults around 35-40 years of age. The condition also has a high recurrence or relapse rate.

Need of the study: The evidence pertaining to Moya-Moya Disease is based on case studies, most of the published literature concentrates on the neurological profile or intervention and less details are known on speech-language profile and the "Aphasia" component associated with the condition, the current case study profiles the speech language profiling of 15-year-old adolescent affected by the condition over a period of 3 years.

Aim: The current case study profiles the speech and language disorders associated with Moya-Moya Disease in a 15-year-old adolescent affected by the condition over a period of 3 years.

Methods:

The client was 15-year-old when he reported to a hospital. complaining of headache. He also developed sudden onset weakness in the right upper and lower limb. He also complained slurring of speech. There was no complaint of vomiting. Soon he was taken to the hospital where he was diagnosed to have acute ischemic stroke-left insular infarct and with Moya-Moya Disease

Physical examination revealed Right Upper Motor Neuron Palsy. CT scan was carried immediately on the day of symptom manifestation. The CT revealed patchy sub-acute infarct, involving portions of left fronto-temporo-parietal lobe, frontal operculum, insular cortex with mild mass effect and the diagnosis of Moya-Moya Disease was made from the CT findings.

Treatment: After explaining the neurological findings to the client's family, Left mini temporal Craniotomy was carried out and medicine for heart safety and blood thinning was prescribed for a period of 1 year with a recommendation made for constant follow up. Post 1 month after surgery, the client reported to a premier speech and hearing institute with the complaint of no verbal output and a detailed speech and language profiling was carried out. The results section of the current study summarizes on the profiling and treatment.

Results and Discussion

Case Presentation

The complaint at the time of reporting included the loss of expressive language and reduced clarity of speech following acute ischemic stroke.

WAB Domains	Max score	Visit 1
Spontaneous speech		
Information content	10	3
Fluency	10	2
Comprehension		
Yes/No question	60	51
Auditory word recognition	60	54
Sequential commands	70	59
Repetition	100	14
Naming		
Objective naming	60	24
Word fluency	20	0
Sentence completion	10	4
Responsive speech	10	4
Aphasia quotient		35.6
Reading	100	70
writing	100	17
Praxis	60	57
Impression		Broca's aphasia

Table 1: Summary of performance on WAB Domains

Following this the client attended speech and language therapy for a duration of 3 months where the goals taken was to make the client express his needs verbally, to improve intelligibility/clarity of speech, to improve the comprehension and repetition abilities. The client dropped from therapy after 3 months as the Covid pandemic was at its peak during this time and he reported after 2 years which is highlighted under re-evaluation section.

Reevaluation

The findings at visit 2 were as follows: During visit 2 client was 18 years old and reported to the institute on June 2023 with complaints of word-finding difficulties and poor performance in reading and writing. MRI performed on January 2023 revealed cerebrospinal fluid signal intensity areas with gyral volume loss noted in the left parietal and right temporal lobes. No acute infarcts on diffuse weighted imaging and no obvious

The client reported 1 month after the stroke. The client was a known case of bilateral Moya-Moya disease and had undergone left mini temporal craniotomy and encephalo duro myoarterio synangiostosis as discussed in the method section.

Upon oral speech mechanism examination client had tongue and lips deviated to the right side of protrusion and reduced range of motion thus reducing speech clarity. The client had no complaints related to swallowing and on informal assessment, the result was found to be clinically normal swallow. A formal assessment was carried out using Western Aphasia Battery-Kannada [7] was used to assess language impairment. Client M had good comprehension abilities and was able to answer relevant questions in 1-word utterances with unclear speech and was able to follow 2-step commands. The client performed poorly in spontaneous speech, naming, repetition, and reading and writing domains. Based on all the evaluation client was provisionally diagnosed as having Broca's Aphasia. The scores obtained in various sub-sections of the WAB-K on both visits are presented in Table 1.

intracranial bleed. Brain angiogram reveals faint visualization of the supraclinoid part of the right internal carotid artery, and the cavernous and supraclinoid part of the left internal carotid artery. The left middle cerebral artery and left anterior cerebral artery were not visualized. The right middle cerebral artery and anterior cerebral artery were poorly visualized. Oral speech mechanism examination revealed structurally normal structures with affected tongue lateralization and elevation thus slurred speech was present. The client had poor reading abilities and his writing legibility was poor. Upon administration of WAB-K (Kannada version) and an AQ of 88.9 was derived. Client M performed relatively low in the naming domain (86 on 100) as compared to all other domains of WAB-K. Thus, Client M was diagnosed with Anomic aphasia based on the scores. The scores on WAB-K during the re-evaluation is summarised in Table 2.

WAB Domains	Max score	Visit 2
Spontaneous speech		
Information content	10	9
Fluency	10	8
Comprehension		
Yes/No question	60	57
Auditory word recognition	60	60
Sequential commands	70	68
Repetition	100	90
Naming		
Objective naming	60	60
Word fluency	20	12
Sentence completion	10	10
Responsive speech	10	10
Aphasia quotient		88.9
Reading	100	71
writing	100	
Praxis	60	60
Impression		Anomic Aphasia

Table 2: Details of re-evaluation

Therefore, as observed at visits 1 and 2, anomia was also the predominant feature of his language impairments. In addition, improvements were observed in AQ. The client was able to follow 2-3 step commands and could answer questions with inappropriate syntactic complexity. The client could narrate speech but with word-finding difficulty. Recommended Goals for the future are as follows: To work on naming skills, improving discourse, reading and writing skills, and overall speech intelligibility.

The onset of the condition was as young as 15 years which converged with many case reports reported on Moya-Moya Disease [4,5]. The condition remained stable over a period of 3 years. In fact, the client improved on his overall health as well as speech language profile in the span of 3 years. He was regular on his medicine. Though he discontinued therapy he showed remarkable progress and his clarity of speech which a major concern at the time of reporting improved over these 3 years and the diagnosis changed from Broca's Aphasia a considerably severe variant of aphasia to anomic aphasia, a milder variant of aphasia. Therapy provided immediately after the onset improved the client to achieve mastery over the deficient skills and was game changer in his case.

Conclusion

The current case study profiles a 15-year-old client diagnosed with Moya-Moya Disease. It highlights the neurological evaluation and intervention. The case study profiles the speech language formal evaluation details carried out at the time of reporting, the speech language treatment carried out over a brief period of 3 months and also the reevaluation carried out after the period of 3 years and how the condition improved over time due to the impact of therapy provided immediately after the manifestation of the condition.

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