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Case Report

Enhancing surgical outcomes: The effects of speech therapy on a school-aged girl with Moebius Syndrome

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ABSTRACT

Purpose: Moebius Syndrome is a rare congenital neurological condition often characterized by multiple cranial nerve involvement. This case study presents an eight-year old girl with Moebius Syndrome (MC) who received 30 sessions of speech therapy. This occurred after presenting to clinic 11 months after left facial reanimation with gracilis thigh muscle transfer surgery. On examination, only flickers of left facial movement were observed. There was no movement on the right side of the face. As a consequence of the minimal movement, MC presented with drooling and unintelligible speech. The purpose of speech therapy was three fold: minimise the pooling of saliva, improve the placement of the articulators so that articulation of speech sounds would be more accurate, and gain advances in overall intelligibility.
Methods: Therapy focussed on speech, facial movement and saliva management using a combination of speech drills, evidence-based articulation therapies, facial exercises with surface electromyography biofeedback, self-awareness training and compensatory saliva management strategies.
Results: After a course of 30 one-hour speech therapy sessions, substantial improvements were seen in speech sound accuracy, overall intelligibility, facial movement and saliva control.
Conclusions: The combination of surgery and speech therapy led to functional gains that surgery alone did not achieve. The impact of speech therapy on surgical outcomes in individuals with Moebius syndrome deserves further investigation.

1. Introduction

Moebius Syndrome is a rare congenital neurological condition that in its pure form presents as impairments of cranial nerves VI (Abducens) and VII (Facial), which control eye movements and muscles of the face. Bilateral facial palsy of varying severity is a common feature. Children can, however, present with multiple cranial nerve involvement including cranial nerves II, V, VI, VII,, IX, X and/. r XII. Coexisting tongue weakness occurs in two thirds of cases [1–4]. In some cases, orofacial problems can be complex involving cleft lip and/or palate, microglossia, micrognatia, as well as oromotor dysfunction [1]. These impairments often lead to difficulties in speech intelligibility, vision, hearing, swallowing, saliva management, self-esteem and social competence [1–4].

Speech therapy can target swallowing, saliva management, facial movement, speech intelligibility, social skills and/or overall quality of communication. Yet, little has been published about the efficacy of speech therapy in children with this rare syndrome [5–7]. With improved surgical techniques such as re-innervated gracilis thigh transplants to treat the damaged cranial nerves in Moebius Syndrome [8,9], there is a need for paediatric teams to consider the role for post-surgical speech therapy interventions to maximise the results made possible by surgery. Goldberg and colleagues report on 12 patients following bilateral gracilis muscle transplant surgery. No details of speech therapy are provided, however, all children had increased intelligibility and reduced use of compensatory strategies after receiving post-surgical therapy [7]. Robust research is complicated not only by the rare nature of the disease but also by the variability in: levels of cranial nerve damage, timing and outcomes of surgery, availability of speech therapy services and the range of intervention techniques utilised. Previously, articulation therapy focussed on strengthening and fine motor control of the cheeks, lips and velopharyngeal valve has been advocated for this population [5].

The ability to compensate is considered an important factor in

Abbreviations: sEMG, surface electromyography.

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therapeutic success and strategies reported previously include labiodental and jaw protusion use to improve bilabial approximation [4]. Despite its common use in Bells Palsy [10], to our knowledge, only one published case study has described the use of surface electromyography (sEMG) as a useful adjunct to muscle training in Moebius Syndrome [6].

2. Case report

This case study describes the speech therapy interventions and outcomes of an eight year-old child, MC who had Moebius Syndrome. MC lived at home with both parents and an older sister who has no congenital anomaly. MC attended a mainstream school and performed at national standards across reading, writing and maths on national education testing. MC had bilateral club feet, obstructive sleep apnoea, moderate sensorineural hearing loss requiring bilateral hearing aids, and short sightedness requiring glasses. At the age of six, MC's medical team proceeded with a left facial reanimation surgery, using a free gracilis muscle transfer from the thigh. This involved transfer of the gracilis muscle as microvascular transfer to the left upper lip using the masseter nerve for anastomosis. For a variety of reasons, only one side was treated. Removal and transfer of the muscle from the thigh to the upper lip requires complex and lengthy surgery; the potential success of the surgery was unknown, her coexisting sleep apnoea made her more vulnerable to risks and the available funding was also restricted. Eleven months after the surgery, MC presented to the speech therapy clinic because there had been no noticeable changes in her facial movement, speech intelligibility, articulation or saliva management.

2.1. Language abilities

Prior to speech therapy commencing, MC's language was assessed as within normal range. MC scored age-appropriate on the South Tyneside Assessment of Syntactic Structures (STASS) [11]: a standardised assessment of a child's use of complex sentences, pronouns, verb tenses and prepositions. MC used meaningful complex sentences, for example, “Look, Dad's giving a present to the girl cos she's a good girl. “ He fell off his bike and his Mum said “It's okay”.

2.2. Facial movement, saliva and feeding management

On oral examination, MC presented with multiple oromotor difficulties (Table 1). Her Sunnybrook Facial Grading System [12] score prior to therapy was 13 out of a maximum of 100 (Table 5). MC did not describe significant swallowing difficulties herself (Table 5) but her mother graded her eating and drinking more severely. Food needed to be soft or chopped. She was a slow and messy eater. Her mother often handed her a tissue and regularly reminded her to clear the frothy saliva that pooled in her anterior and lateral sulci.

2.3. Speech and articulation

As a pre-schooler, MC had received monthly group speech therapy focussed on language and interactive pragmatic skills. Previous speech therapy had not focussed on oral-motor aspects of speech production or articulation. Baseline assessment was administered by the treating clinician using the “New Zealand Articulation Test (NZAT) [13] (Table 2). The NZAT requires a child to name 80 individual pictures assessing all English phonemes. Phonemes are evaluated at the beginning, middle and end of words, for example. ‘p’ in pie, apple, cup. MC achieved an overall percentile rank of <1 resulting in a classification as a severe speech sound disorder. Her speech was extremely difficult to understand. MC often produced two or even three errors within one word (Tables 3 and 4). Error patterns included several atypical production patterns including hypernasality, “stopping” of continuent fricative sounds and exaggerated upward movement of the jaw to push the tongue up to the palate so that some type of alveolar sounds ‘d’ and ‘t’ could be attempted in a few words. During connected speech, the treating clinician determined that MC's speech intelligibility was below 50%. Intelligibility was rated during a 3-min sample of spontaneous speech. The treating clinician and mother both transcribed the speech sample using the mother's transcription as accurate and measuring intelligibility on the treating clinician's transcription. MC's rate of speech was rapid: 270 syllables per minute in comparison to norms for her age (150–180 syllables per minute) [14]. Rate of production was calculated using the same 3-min sample of spontaneous speech.

3. Intervention

MC received 30 one-hour speech therapy every two weeks for 15 months with the two authors and a speech-language therapy student. Weekly sessions were not possible due to financial and geographical constraints. Sessions were directed toward the following goals:

1. Improve articulation of speech sounds within targeted words and short phrases. In particular, improve the articulation of words which contain bilabials: p,b,m,w.

Table 1
Oral examination results prior to therapy.

Structure	Function
Oral cavity	++ pooling of saliva in lateral sulci
	Moist and clean oral cavity
Jaw	Normal jaw opening but reduced rotary movement
	Compensatory use of jaw when requested to move tongue or face
Lips	No visible movement of right face/lips
	Flickers of movement in left cheek to command
	Mouth in open position at rest
	Unable to close lips to command
Tongue	Minimal lateral, anterior or posterior movements to command
	Microglossia (as diagnosed by paediatrician)
Palate	Symmetrical palate movement
	Hypernasality in speech
Swallow	Able to trigger a swallow to command
Cough	Weak cough to command