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## Highlighting the speech profile in hyperkinetic Dysarthria: oromandibular (Jaw opening type) and lingual dystonia (A single case study)

**Garima Dixit, Priyanka Dubey and Ayushi Shukla**

### Abstract

Neurological impairment affecting the motor programming or neuromuscular execution of speech that result in speech disturbances are termed as neuromotor speech disorders. Out of all the communication disorders 36.5% are motor speech disorders and out of which 46.3% are dysarthria. Hyperkinetic dysarthria are a perceptually distinguishable group of disorders that are most associated with the diseases of the basal ganglia control circuits. The deviant speech characteristics are a result of rapid or slow, involuntary, rhythmic and unpredictable movements. Hyperkinetic dysarthria can be due to multiple aetiologies ranging from Ballismus, Chorea, and Myoclonus to Dystonia and Dyskinesia. Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both. Dystonic movements are typically patterned, twisting, and may be tremulous. Dystonia is classified along two axis: clinical characteristics and aetiology. Oromandibular dystonia involves the masticatory, lingual, perioral, and platysma muscles. The estimated annual incidence of Oro mandibular dystonia is 3.3 to 6.9 cases per 1 million people. Oromandibular dystonia can either be a primary disease or secondary to other disorders. Patients are categorized as having focal, segmental, multifocal, or generalized dystonia. OMD can be focal, but more often is part of a segmental or general dystonia. The different clinical forms encountered are jaw-opening oromandibular dystonia (JOOD), jaw-closing oromandibular dystonia (JCOD), and mixed OMD (patients with two or more combinations of either JCOD, or jaw deviation (JD), and with no clinical predominance of any subtype. (Linda Slaim *et al.*). A case 32years/Male was confronted bedside in the neurology ward of the SAIMS Medical College and hospital, who reported complaint of abnormal involuntary movements of the tongue for one year. The patient's assessment was completed by Neurologist and Speech Language Pathologist. Neurologist clinically evaluated and diagnosed the case as oromandibular Dystonia (Jaw Opening Type) and Lingual Dystonia. A speech language pathologist completed the assessment by administering the different standardized test in which detail evaluation of Oral Peripheral Examination, reflex examination, perceptual evaluation of speech characteristics and cranial nerve examination was done. The results of assessment revealed severely deviant speech characteristics with significant effects on respiratory, phonatory, articulatory, resonatory and prosodic speech subsystems. Hyperkinetic dysarthria has significant impact on all speech sub systems. In an integrated manner it deteriorates the quality of life of an individual by degrading professional as well as personal life. Speech therapy and swallowing rehabilitation will help in improving the quality of life of the patient by reducing the effects of hyperkinetic dysarthria and maximizing functional communication skills. However, for intervention to be effective, understanding the severity and characteristics of the disorder is of paramount importance and hence up to the mark assessment protocol is to be followed.

**Keywords:** Oromandibular dystonia, lingual dystonia, speech

### Introduction

Speech is amongst the most powerful neurological tools possessed by humans. Under most circumstances speech is produced with ease that belies the complexities of operations underlying it. Unfortunately, presence of neurological disease can inhibit this tool. Neurological impairment affecting the motor programming or neuromuscular execution of speech that results in speech disturbances are termed as neuromotor speech disorders. A group of disorders resulting from impairments in neuromuscular control over the speech mechanism due to damage in the central or peripheral nervous system are known as dysarthria. Out of all the communication disorders 36.5% are motor speech disorders and out of which 46.3% are dysarthria. Hyperkinetic dysarthria are a perceptually distinguishable group of disorders that are most associated with the diseases of the basal ganglia control circuits.

The deviant speech characteristics are a result of rapid or slow, involuntary, rhythmic and unpredictable movements. Hyperkinetic Dysarthria can be due to multiple aetiologies ranging from Ballismus, Chorea, and Myoclonus to Dystonia and Dyskinesia. (Joseph. R. Duffy, 2005) <sup>[1]</sup>. Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both. Dystonia is classified along two axis: clinical characteristics and aetiology. Oromandibular dystonia involves the masticatory, lingual, perioral, and platysma muscles. The estimated annual incidence of Oro mandibular dystonia is 3.3 to 6.9 cases per 1 million people. OMD is a chronic condition affecting speech and swallowing, leading to an impaired quality of life. Oromandibular dystonia can either be a primary disease or secondary to other disorders. Patients are categorized as having focal, segmental, multifocal, or generalized dystonia. OMD can be focal, but more often is part of a segmental or general dystonia. The different clinical forms encountered are jaw-opening oromandibular dystonia (JOD), jaw-closing oromandibular dystonia (JCOD), and mixed OMD (patients with two or more combinations of either JOD, JCOD, or jaw deviation (JD), and with no clinical predominance of any subtype. (Slaim *et al.* 2018) <sup>[4]</sup>. The following are the prevalence of occurrence of Oromandibular Dystonia in various domains which include Male versus Female ratio, Etiology, Prevalence of Dystonia, Symptoms coexisting with Oromandibular Dystonia and associated movement disorders. (Slaim *et al.* 2018) <sup>[4]</sup> The need of this study was to evaluate the effects of Oromandibular Dystonia on speech, language, swallowing, cognition and quality of life of the patient. The incidence of the disorder persisting according to a study conducted

within the Speech Pathology program in the Department of Neurology at Mayo Clinic revealed that, 57% cases were diagnosed with motor speech disorders out of which 20% of whom were diagnosed with dysarthria. (Julie. M. Barkemeier *et al.* 2017). It is significantly important to understand the impact of the disorder on the quality of life of the patient resulting from speech and swallowing disturbances in order to plan effective intervention programme. One important need of this study is to contribute to existing knowledge with respect to the speech, language, cognition and swallowing along with facilitating evidence-based practice.

The need of this study was to evaluate the effects of Oromandibular Dystonia on speech, language, swallowing, cognition and quality of life of the patient. The incidence of the disorder persisting according to a study conducted within the Speech Pathology program in the Department of Neurology at Mayo Clinic revealed that, 57% cases were diagnosed with motor speech disorders out of which 20% of whom were diagnosed with dysarthria. (Julie. M. Barkemeier *et al.* 2017). It is significantly important to understand the impact of the disorder on the quality of life of the patient resulting from speech and swallowing disturbances in order to plan effective intervention programme. One important need of this study is to contribute to existing knowledge with respect to the speech, language, cognition and swallowing along with facilitating evidence-based practice.

The investigation was aimed at highlighting the clinical features of speech, language, cognition and swallowing in a case of hyperkinetic dysarthria caused by oromandibular dystonia (jaw opening type) & Lingual Dystonia; along with understanding the impact on the quality of life of the patient.

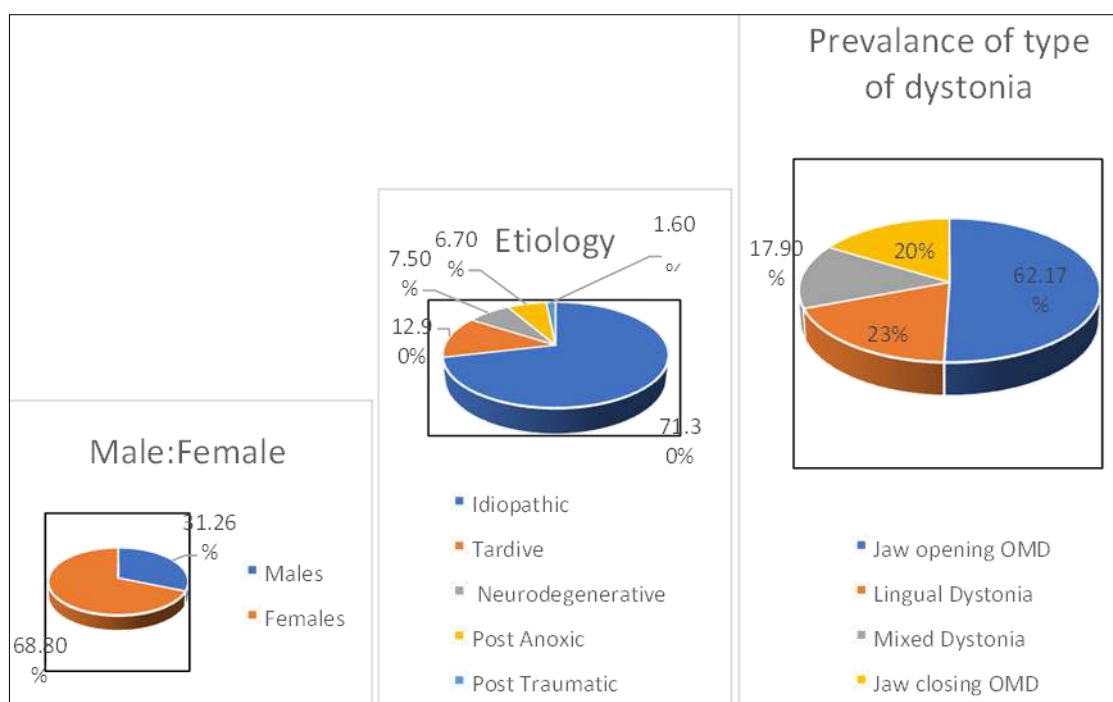


Fig 1: Showing incidence, etiology & prevalence of type of dystonia

## Method

A case 32years/Male was confronted bedside in the neurology ward of the SAIMS Medical College and hospital, who reported complaint of abnormal involuntary movements of the tongue since one year. The patient's

assessment was completed by Neurologist, Psychiatrist and Speech Language Pathologist.

Neurologist clinically evaluated and diagnosed the case as Oromandibular Dystonia (Jaw Opening Type) and Lingual Dystonia.

presence of obsessive compulsive disorders). Also, the patient had a prospective of indulging in Botox Injection Therapy. Patient self-report of disorder impact on quality of life included disturbed professional and personal life, embarrassment and reduction in socializing and self-isolation.

### Detail Speech, language, swallowing and cognitive evaluation profile

Speech evaluation was done which included Frenchay Dysarthria Assessment, Oral Peripheral Examination, Cranial Nerve Examination, and Perceptual evaluation of speech characteristics, The Montreal Cognitive Assessment, Gugging Swallow Screening and WAB Bedside Screener. The results of assessment revealed severely deviant speech characteristics with significant effects on respiratory, phonatory, articulatory, resonatory and prosodic speech subsystems. Frenchay dysarthria assessment interpretation leaned towards the presence of hyperkinetic dysarthria.

		Patient/ Proband																										
		Re fle x	Res pir atio n	Lip Movements			Jaw	Palate		Voice		Tongue			Intelligi bility													
Normal function ↓	9																											
	a																											
	8																											
	b																											
	7																											
	6																											
	c																											
	5																											
	4																											
	d																											
No function ↓	3																											
	2																											
	e																											
	1																											
	FDA total=																											
	co u g h	Swallow	Dribble/drool	At rest	In speech	At rest	snread	seal	alternate	In speech	At rest	In speech	While eating	function	In speech	time	pitch	volume	In speech	At rest	protrusion	elevation	Lateral	Alternate	In speech	words	sentences	conversation

Oral Structures	At Rest	During Movement (With Resistance)	During Movement (Without Resistance)	Sustained Posture
Face	Abnormal, asymmetrical, presence of involuntary movements.	Increased tone, excessive strength, endurance couldn't be measured due to the presence of involuntary movements.	Involuntary movements increased in attempt of voluntary movements.	Couldn't sustain any facial posture for > 2-3 seconds.
Lips	Absence of lip seal, drooling.	Inability to sustain resistance due to difficult in maintaining voluntary control.	Absence of lip seal, difficulty puffing cheeks, difficulty sucking, difficulty puckering, difficulty maintaining retraction, difficulty performing alternate lip pucker-retraction.	Mouth opened due to inability of voluntary lip closure.
Jaw	Opened, inability of jaw closure.	Increased spasticity. Cannot close jaw against resistance.	Can move jaw from right to left.	Inability to make maxilla and mandible meet, can keep the jaw apart.
Tongue	Consistent involuntary movements throughout the day. Presence of tremors even while sleeping.	Inability to sustain finger pressure against either of the cheek, inability to push tongue against a tongue blade.	Difficulty sticking tongue out, difficulty elevating, lateralization, retro flexion.	Absence of maintaining any kind of lingual sustained posture.

Speech musculature assessment during non-speech oral movements revealed decreased range, decreased rate, increased tone, significantly reduced steadiness, and significantly reduced accuracy, presence of fasciculation, evident spasticity, and evident synkinesis. No visually

evident atrophy, weakness or facial myokymia were noted. Speech characteristics, when evaluated perceptually, on a scale of 0-4, the patient scored 4 indicating severely deviant speech characteristics.

**Table 2:** Results of Speech Subsystem Analysis

Respiratory	Phonatory	Articulatory	Prosody	Resonatory
Forced inspiration, Forced expiration.	Strained voice quality, pitch breaks, excessive loudness, loudness decay and myoclonus. <b>GRBAS Rating Scale</b> G:2 R:1 B:2 A:0 S:2	Imprecise consonants, prolonged, irregular articulatory breakdowns, inappropriate vocal noises.	Short phrases, reduced stress, variable rate, prolonged intervals, inappropriate silences, short rushes of speech.	Weak pressure consonants, altered resonance (hyper nasality).

**Table 3:** Depict speech and non-speech characteristics observed in client

Speech Characteristics	Non-Speech Characteristics
Reduced loudness level	Atrophy
Irregular articulatory breakdown	Fasciculation
Imprecise consonants	Hypoactive gag
Short phrases	Hypotonia
Rapid deterioration and recovery after rest	Reduced range of motion on AMR tasks
Pitch breaks	
Slow and regular alternate motion rates	
Mono pitch	
Mono loudness	

### Cranial Nerve Examination

The cranial nerve examination involves a number of steps.

The only nerves that could be evaluated in this patient were the trigeminal, Facial, Glossopharyngeal and Hypoglossal.

**Table 4:** Results of Cranial Nerves

<b>Trigeminal Nerve</b>	Inadequate Jaw Pressure, Inability to Clench Jaw and Teeth, Presence of Pain sensation, presence of corneal reflex, inability to whistle.
<b>Facial Nerve</b>	Facial asymmetry at rest, facial asymmetry during the task like eyebrows raising, closing eyes to resistance, smiling, frowning, showing teeth and puffing cheeks.
<b>Glossopharyngeal nerve</b>	Absence of Gag Reflex, normal elevation of soft palate, presence of taste sensation.
<b>Hypoglossal Nerve</b>	Normal force, No deviations, Presence of tremors at rest as well as during movement. Increased tone.

**Table 5:** Results of speech motor tasks

Speech Motor Behaviours	Interpretation
Maximum repetition rate (MRR) for single syllable and trisyllabic sequence ((e.g., /pAtAkA/ etc.)	Severely affected, Unable to combine and produce
Alternate motion rate (AMR)	Severely affected, /p/: 7 in 20 sec /t/: 9 in 20 sec /k/: 9 in 20 sec
Maximum sound prolongations of vowels	Severely affected, 8 sec
Non-word repetitions (NWR)	Severely affected, 30%

Speech intelligibility rating: speech intelligibility was assessed using speech intelligibility rating scale it suggests rating 3.

### Swallowing Evaluation

Gugging Swallow Screening was administered. It is a two phase examination involving a preliminary examination involving saliva swallow test and a direct swallow test

involving swallowing test with different consistencies of semisolid, liquid and solid texture. Preliminary test score of the patient was 5 suggesting passed in saliva swallow. Direct swallow test implied passed in semisolid and liquid consistencies whereas failed in solid consistency. A total score of 19/20 was obtained that revealed the presence of a mild swallowing disorder.

**Table 6:** Depict the score of domains and sub domains of Montreal cognitive assessment (MoCA) and Gugging Swallowing Screening (GUSS) tests

Montreal cognitive assessment (MoCA) Hindi								
Domains	Visuospatial	Naming	Memory	Attention	Language	Abstraction	Delayed Recall	Orientation
Raw score	5/5	5/5	--	6/6	3/3	1/2	4/5	6/6
Total score	30/30							
impression	Above mention score suggestive of “ cognitive impairment”							
Gugging swallow screening (GUSS)								
Domains	Indirect Swallow Test		Direct Swallow Test					
Sub-domains			Semisolid		Liquid		Solid	
Raw score	5		5		5		4	
Total score	19/20							
Impression : “Mild signs of dysphagia”								

Language evaluation was done using Web-Screener which revealed absence of any language issues.

Common SLP treatment approaches include techniques that optimize respiratory–phonatory coordination techniques for improved pitch and loudness control, phrasing, and consistency of sound production. Facilitative speaking strategies may be used to improve comprehensibility by

modifying speaking tempo, phrase length, and incorporate purposeful pauses during speaking. This was the tailor made plan for our patient with this specific symptoms.

The patient had the following symptoms and the management program will take place accordingly. (Linda Slaim *et al.*)<sup>[4]</sup>.

**Table 7:** Showing Symptoms and Management program

Symptom	Intervention
Inhalation appears inadequate, prolonged, or speaking initiation occurs at unusual locations within the respiratory cycle, or utterance	Maximum inhalation/exhalation tasks, or sustained phonation tasks to improve respiratory/phonatory coordination and steadiness
Few words or syllables produced per breath group.	Body positioning to optimize breathing and respiratory efficiency during speaking
Abnormal posture or movements associated with volitional respiratory-phonatory coordination during speaking	Rehearse taking deeper inhalations prior to speaking and implementing increased respiratory effort during speaking
Poor integrity, Variable pitch and loudness.	Laryngeal relaxation techniques such as easy voice onset, Speech Tasks
Abnormal speech pattern or rate.	Modify speaking rate (typically encourage slower), Speech rhythm techniques

## Conclusion

Hyperkinetic dysarthria has significant impact on all speech sub systems. In an integrated manner it deteriorates the quality of life of an individual by degrading professional as well as personal life. Oromandibular dystonia has significant impact on speech & swallowing skills. Research can be a stepping stone in terms of exploring information for assessment and intervention protocol in case of hyperkinetic dysarthria and adequate awareness and counselling of care givers and professionals are helpful to improve the overall quality of life of the patient. Speech therapy and swallowing rehabilitation may help in improving the quality of life of the patient by reducing the effects of hyperkinetic dysarthria and maximizing functional communication skills. However, understanding the severity of Speech & swallowing characteristics is of paramount importance as it severely impairs an individual's quality of life.

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