

## Chapter 1

# A Call for Biomechanics to Understand Hypotonia and Speech Movement Disorders in Down Syndrome

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## Abstract

While a neurotypically developing child reaches 100% speech intelligibility by age 4, it is unusual for children with Down syndrome (DS) to achieve this level of intelligibility at any age [1]. This is due, in part, to altered biomechanics and physical growth delays that affect craniofacial and thoracic/limb structure, along with a pervasive movement disorder characterized by hypotonia and motor delay. The predominance of reports focused on limb-body features and movement characteristics, justifies the call for research on the biomechanics and neuromotor control features of the vocal and aerodigestive tract as it relates to speech and feeding in children with DS. This chapter considers available data on clinical measurement and new approaches to orofacial biomechanics in the context of craniofacial motor control and speech impairment in DS. Additionally, qualitative and quantitative methods of assessing orofacial biomechanics are presented to lead to more efficacious therapeutic interventions and functional outcomes for speakers with DS.

## Introduction

Although variable, speech production in Down syndrome (DS) is associated with impairments in voice, phonology, fluency, prosody, and intelligibility [2]. An assessment of the neuromuscular system in DS usually includes the term 'hypotonia' to characterize the decreased resistance of a limb to passive stretch [3-4]. Many infants and children with DS are late to achieve early motor

milestones, including grasping, rolling, sitting, standing, walking, sucking, babbling, and speech [5]. Often, speech treatment programs for children with DS include an exercise program (e.g., oral muscle strengthening, alternating rapid movement sequences) at the beginning of a speech treatment session as a “warm-up,” followed by direct work on speech production [6-7]. The rationale that is given for the use of such exercises is that they will normalize muscle tone and that normal muscle tone is a necessary condition for skilled movement control in the orofacial system. A quick Google with the search string (oral exercise, speech therapy, Down syndrome), provides a many of speech-language pathologists blogs describing how they implement oral motor exercise in children with DS with no clinical evidence base [8]. Despite the paucity of evidence-based research, some of the speech-language-pathology treatment manuals used today continue to advocate the use of oral strength and movement exercises to treat hypotonia associated with DS [9-10].

Interventions for modifying muscle tone include oral motor exercise such as lip massage, whistling, blowing, using tubes to practice sucking, tongue resistance work to increase tongue strength, and using a bite block to increase jaw stabilization [6,11]. While studies and intervention in the limb control literature have reported that DS limb motor control improved after training [12-13], it follows that similar training in the orofacial mechanism may improve motor function (i.e., improving labial muscle tone,

sucking, and feeding abilities). Although muscle tone of children with DS has been studied in the limbs, direct translation from limb dynamics to orofacial dynamics is limited given the fundamental differences in biomechanics and neuromotor control strategies for mass-dominant limbs versus the elasticity-dominant low-mass articulatory systems (e.g., upper and lower lips) of the orofacial mechanism.

## What is Muscle Tone?

The term muscle tone, as commonly used in clinical practice, is generally based upon the examiner’s manual perception of passive resistance to an imposed motion or stretch [13]. This non-instrumental method for assessing muscle tone relies on palpation and passive displacement of an extremity (e.g., limb, jaw, lip) [14]. For example, an examiner may displace a person’s limb around a joint slowly, with the subject instructed to relax and not resist the motion. The examiner judges the feeling of resistance during passive displacement based upon his/her previous experience. The examiner then compares the observed resistance to his/her internal perception of ‘normal muscle tone.’ When the perception of resistance is lower than expected, it is called ‘hypotonia,’ ‘low muscle tone,’ or ‘low rigidity.’ The quantitative counterpart to hypotonia is the biomechanical measurement of ‘stiffness’ which requires the instrumental measurement of tissue displacement divided by reactive force. Hence, ‘hypotonia’ is the clinical

perception of lower-than-normal resistance to the motion, whereas 'stiffness' is the instrumental quantification of resistance to an imposed displacement.

Insults to the neuromuscular system can disrupt normal tone and lead to various speech disorders [14-15]. Damage to lower motor neurons may impair normal function of the efferent component of the stretch reflex, resulting in hypotonia, as observed in flaccid dysarthria. Congenital or acquired forms of the Upper Motor Neuron Syndrome typically manifest exaggerated stretch reflexes and hypertonia and are associated with spastic dysarthria. Basal ganglia disease is associated with hypokinetic dysarthria, a rigid form of hypertonia. Although muscle tone has been assessed in select forms of dysarthria [15], the relation between hypotonia and speech impairment in DS remains unknown.

## From Oral Feeding to Speech in DS

The mandibular oscillations performed by infants during sucking and mastication have been hypothesized to frame a subset of coordinative synergies (e.g., jaw elevation/lowering, lip opening/closing) apparent during the production of canonical babbling and syllable production [16-17]. Through ontogeny, adaptation of the mastication central pattern generator (mCPG) and other heteroarchitectural cerebral-subcortical networks involving Broca's area and orofacial sensorimotor cortex may be modified for

communication and feeding [16,18-21]. In a study of three twin pairs (3 children with DS and 3 healthy children, ages 11-27 months), Spender et al. (1995) [22] found that children with DS demonstrated more oral motor dysfunction, including improper lip closure, excessive tongue protrusion, and poorly controlled jaw function. It was believed that hypotonia and weakness of the orofacial musculature contributed to an impaired ability to modulate tension of the articulators DS [23]. An impaired oromotor system in DS children may be associated with poor intelligibility (i.e., clarity of speech perceived by listeners) [24].

A retrospective chart review of 49 children with DS showed that 80% had feeding problems [25], and required more time to develop the motor coordination necessary for normal feeding [26]. Oromotor function in DS children not only lags behind intellectual development but also follows an irregular pathway [27]. Impairments in the timing of jaw and tongue movement initiation leads to a breakdown in maintaining a smooth sequence of feeding actions. The mean age to introduce solid foods in a group of 44 children with DS was found to be delayed relative to normal controls, which was presumed to be deleterious to oral motor development [28].

In addition to feeding difficulties, parents of DS children also reported impairments in oromotor, limb motor, and speech skills in a sample of 937 questionnaires [29]. Another questionnaire based on a new cohort of partici-

pants reported the presence of verbal apraxia in 15% of 1620 DS children [1]. Oromotor impairments in children with DS are widely reported by parents and speech-language pathologists, yet systematic research on the biobehavioral and biomechanical aspects of the orofacial mechanism and its relation to speech production and feeding in this underserved clinical population is lacking.

Delayed onset of speech production is common among children with DS, although some start to talk at the expected age [30]. For example, Lynch et al. (1995) [31] found that the average age of onset for canonical babbling was 2 months later for infants with DS compared to typically developing infants, and the rate of babbling progression was reduced and more variable. This relative instability in canonical babbling may be a consequence of motor delay and muscle hypotonicity associated with DS [31]. Because babbling has similar basic phonetic properties of speech sounds, it is reasonable to conclude that typical babbling pattern is linked to better performance on the speech and language skills [32]. In addition, the onset of meaningful speech in children with DS is significantly delayed (14 months for typically developed children compared to 21 months for infants with DS), and that majority of their utterances are non-meaningful [33]. Only 2% of utterances by 21-month old DS infants were judged meaningful, and at 30 months the proportion of meaningful utterances remained below 5%. Children with DS manifest

significant speech intelligibility difficulties at these early stages which may persist through adolescence and into adulthood [34-35]. Unfortunately, most studies of speech motor control exclude children with DS based on genetic exclusion criteria. This means clinicians are left to rely on relatively subjective estimates of neuromotor status and oromotor performance. Speech and language evaluation of a child with DS is typically descriptive in nature, and reliant on the child's ability to imitate the clinician's speech sound model, where a conversational speech sample is favored [36]. By the time many children with DS are able to produce a speech sample of 100 utterances for standard speech sample analyses; they may be in middle school or older [1]. Thus, a critical period for early intervention may have passed and clinicians would have missed the valuable treatment time.

Duffy (2005) [15] commented that specific methods for identifying abnormal tone are lacking in the clinical examination, except for the notation of facial droop as one indicator of hypotonia in assessing motor speech disorders. In the sections that follow, we provide a brief overview of the current qualitative methods and biomechanical methods for objective assessment of orofacial muscle tone, which herein is known as orofacial stiffness.

## Assessment of Muscle Tone for Orofacial Muscles

### Qualitative (Non-Instrumental) Measurement

Qualitative measurements of muscle tone usually involve an element of perceived muscle tension or stiffness at rest assessed by the tester, a readiness to move or hold a position, or a subjective estimate of passive resistance during imposed stretch of an agonist muscle system (i.e., that a person does not actively contract against the applied stretch, in which the resistance noted is attributed to resting muscle tone rather than to voluntary muscle contraction [37-38]. For example, the examiner extends the client's arm, and judges the magnitude of resistance. Therefore, stretching the bicep fibers results in some finite reactive force during forelimb extension. In contrast to the upper and lower limbs, a comparable form of muscle tone assessment is not readily accessible in the peri- and intraoral mechanism given the unique fiber orientation, origin/insertions, and complex muscle geometry in the orofacial system [39]. For example, reactive force to an imposed displacement at one corner of the mouth is the geometric sum of multiple muscle compartments, integument and connective tissues for what is essentially a floating musculo-cutaneous complex with origins and insertions predominantly in skin and integument [40]. One

exception is the mandible, with its relatively well defined muscle levators and depressors organized around the temporomandibular joint and acting on the mandible.

The Dworkin-Culatta Oral Mechanism Examination [41] is a commonly used perceptual rating scale for facial and oral functions. Orofacial status (lip, tongue, jaw) is scored by the examiner as 'abnormal', 'normal', or 'questionable.' Other clinical assessment tests have been developed by clinicians to compare nonspeech and speech function. For instance, the clinician may ask for a pucker, retraction (unilateral and bilateral), or sequential movements of the lips when assessing lip function. Or, the clinician may ask the client to protrude the tongue, move it from side to side, touch the nose or chin with the tip of the tongue, or perform a series of movements in sequences [36]. These are used to judge if there are abnormal movements associated with orofacial structure such as hypokinesia, groping, or tremor.

The main drawbacks of these tasks lie in their non-standardized and subjective nature [42], making them unlikely to be sensitive tools for the identification of abnormality. In addition, subjective measures may be insensitive to changes over time that occurs as a result of treatment or disease progression [43]. Unfortunately, few devices have been developed to objectively measure orofacial muscle tone.

## Quantitative (Instrumental) Measurement

Quantitative analyses of stiffness, or muscle tension, are usually obtained in one of two ways. The first involves imposing a known displacement ( $\Delta X$ ) and measuring the resultant force ( $\Delta F$ ). Dividing the resultant force by the displacement yields a stiffness coefficient ( $\Delta F/\Delta X$ ) [40,44-48]. The second method involves forcing the limb and extracting an impedance measure from the sampled force (torque) to displacement curve [49-50]. The ratio of the force resisting motion to joint angular displacement represents the stiffness.

Biomechanical studies of stiffness in the orofacial mechanism have focused on the lips (individually, or perioral), tongue, and/or mandible. Quantitative measures of perioral stiffness have been studied during a nonparticipatory task (i.e., one in which participants are instructed not to contract the involved musculature) by having a computer-controlled linear motor servo programmed to impose displacements over a span of approximately 24 mm at the oral angle in both female and male adults [45-46]. This linear motor servo device allows the examiner to automate data acquisition and analysis of resultant force and displacement to provide real time measures of perioral tissue stiffness during a 'do-not-contract' condition. This instrument, however, required head stabilization during sampling in patients with dyskinesia as well as in pediatric populations, such as children with DS.

A similar generation '2' device, known as OroSTIFF, allows real-time perioral stiffness measurements without head restraint and has been successfully applied in adolescence with cleft lip (age between 14 to 17 years) [48] and in adults with idiopathic Parkinson's disease [40,47]. The face-referenced OroSTIFF device allows the examiner to impose a sequence of tissue displacements (e.g., stretch to increase the distance between the corners of the mouth, aka interangle span) while simultaneously sampling force and displacement in real time with miniaturized sensors, actuators, and custom software. Individual interangle stretch trials are completed within 10 seconds, with a complete perioral stiffness protocol sampled in approximately 5 minutes per subject. Barlow et al. (2012) [48] demonstrated the efficacy of applying this objective measurement to map differences in perioral tissue stiffness among young patients with congenital cleft lip/palate who had undergone lip revision. The resultant non-linear stiffness functions were significantly higher in the patients with cleft lip/palate who did not have lip revision compared with the noncleft controls [48]. The elevated stiffness values were attributed to the mechanical properties of scar tissue along suture lines in the upper lip among children with clefts. A third instrument, Myoton-3 (Myoton AS, Estonia, EU), has been used to measure stiffness of lateral tongue and mid-cheek in lower motor neuron (LMN) and upper motor neuron patients [51]. Tongue stiffness was reported to be significantly lower for the LMN group than for the normal control group, consistent with the hy-

potonia that accompanies flaccidity. This device delivers a brief pulse perturbation with a non-invasive thin probe and senses the resulting oscillation with an accelerometer [52]. Stiffness was derived from the frequency of oscillation in response to an 8-ms pulse perturbation. Such device could be used to obtain objective measurements of tongue stiffness in children with DS.

Inferences about lips/tongue stiffness have also been made on the basis of force measures during active contraction of muscle. For instance, lip/tongue compression force (strength) and endurance are commonly assessed by speech-language pathologists using the Iowa Performance Instrument (IOPI®, IOPI Medical, Redmond, WA). Strength assessment involves brief maximal efforts (i.e., push against the pneumatic bulb as hard as you can), and endurance assesses the ability of sustained the maximal efforts (i.e., maintain the position as long as you can) [53]. This tool measures the pressure that is generated when an air-filled bulb is compressed by one of the articulators [43]. For instance, by pressing the tongue against the bulb, pressure increases within the bulb are transduced and displayed in real time for clinicians and participants. This tool has been studied in normal speakers as well as those with dysarthria and/or dysphagia [54-56].

Using a computer controlled robotic device (Phantom Premium 1.0, Sensable Technologies, Woburn, MA) programmed to deliver mechanical perturbations to the jaw

in the mid-sagittal plane, Shiller et al. (2002) [57] demonstrated that increased jaw stiffness was associated with decreased kinematic variability during vowel production (e.g., /i/, /e/, /a/, /æ/). This instrument consists of a force transducer, a metal dental appliance that attached to the subject's mandibular teeth, and head-restraint to restrict motion during testing, thereby limiting the use of this system for experimental and theoretical modeling. Hence, applying this instrument in children with DS might be challenging but theoretical modeling of the DS jaw stiffness may be derived with such device.

For the most part, clinical assessments of orofacial muscle tone involve perceptual or subjective estimates of muscle tension or resistance to an imposed stretch or manual displacement of the lips or depression of the mandible. Some emergent technologies are making it possible for the first time to perform biomechanical measurements of stiffness and the dynamics associated with movement. Future application of orofacial biomechanics to the study of movement disorders in children with DS is expected to inform the development of new diagnostic and treatment protocols to remediate speech motor impairments. Knowledge about the pathophysiology of muscle tone and its relation to speech movement disorders is expected to offer new strategies in developing evidence-based treatment plans. Although muscle tone of children with DS has been studied in the limbs, direct translation from limb dynamics to orofacial dynamics is limited given the

fundamental differences in biomechanics and neuromotor control strategies for mass-dominant limbs versus the elasticity-dominant low-mass articular systems (e.g., upper and lower lips) of the orofacial mechanism (discussed below).

## A Comparison of Limb and Speech (Orofacial) Motor Systems

Speech is a complex sensorimotor behavior involving coordinated activity among an extensive array of more than 120 muscles distributed across the respiratory (chestwall), phonatory, velopharyngeal, and orofacial systems [58]. Multiple neural control elements regulate contraction dynamics including cortical and subcortical control processes, sensory feedback primarily from auditory and somatosensory channels [59], and a complex network of integrative central pattern generators (CPG) serving as premotor inputs to lower motor neurons throughout the pons and medulla [19,21] (Figure 1). Speech motor impairments in DS may be complicated by several other factors, including anatomic differences in the vocal tract, hearing status, impaired motor programming/planning (apraxia), and language and cognitive dysfunctions [2]. From the motor development perspective, early oromotor control difficulties (i.e., feeding difficulties) in DS may engender specific constraints on early articulation coordination. The transition of a specific motor task in early motor development involves differentiation (i.e., modi-

fication of a pre-existing behavior to a more specialized one). For example, the arm segments move as a unit during early grasping, with the hand being moved by rotation of proximal joints [63]. As the child develops, this same motor task manifests an increasing degree of motor independence among limb segments, including the arm, hand, and fingers [64]. Speech motor control also may exhibit a similar trend, as the sensorimotor reflex pathways involved in perioral motor control show an increasing degree of specificity during the first decade of life [65]. Next, we consider some of the anatomic and physiologic similarities between the limb and orofacial systems.

Dexterous movements of the upper limbs (e.g., manipulation) share some features of orofacial movements during speech production. Both are highly practiced, complex motor behaviors and are relegated to automaticity in the mature system. Object manipulation and speech operate at the lower end of their force operating ranges (< 20% MVC), involving a predominance of small motor units to achieve precise movement patterns [66]. The somatosensory receptive fields subserving manipulation and speech tend to be small (<5 mm) with a combination of rapid and slow adapting mechanoreceptors that are well suited to support exploration and encode the consequences of skilled motor output [67]. Both limb and orofacial motor skills utilize load compensation mechanisms to rapidly correct for unanticipated perturbations in the local environment [68-70]. The site and extent of lesion

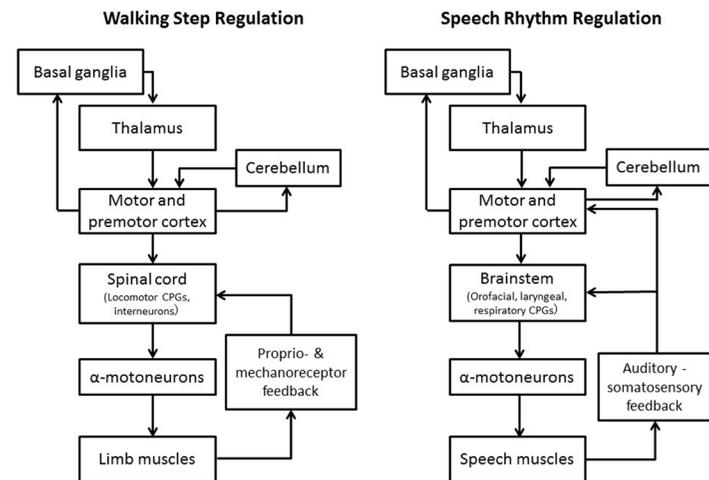
associated with progressive neuromotor disease, traumatic brain injury, or neuro developmental disorders such as DS determine the impact on fine motor skills, including speech and manipulation. There may be common features to the control impairment (e.g., dyskinesia, bradykinesia, hypotonia, dysmetria, etc.) that are manifest in both limb and orofacial movements, or with localized lesions it is possible to observe degradation in motor control in the upper limb/hand with orofacial and vocal tract control systems spared. Thus, important diagnostic information may be gained by looking at impairments across muscle subsystems involved in the control of manipulation and speech in DS.

In recent years, scientists have challenged the notion that divergent motor systems involved in the control of the hand (manipulation) and orofacial (speech, deglutition) systems are distinctly represented in the nervous system [71] (Table 1). For example, Gentilucci et al. (2001) [72] found that reaching and grasping the larger of two objects resulted in increased lip opening and vocal loudness when contrasted with smaller object manipulation, suggesting a cross-system interaction in these different motor behaviors. Despite the difference in biomechanical properties, parallels between the motor skills of speech and upper limb control have been suggested. For instance, kinematic patterns for tongue dorsum movements showed an increase in the slope of the displacement/peak-velocity relationship for unstressed versus stressed vowels [73]. At each stress level, the correlation between displacement and

peak velocity was characterized by a relatively constant interval from the initiation of the movement to the point of maximum velocity. Similar patterns have been shown in the rapid movement about the elbow, whereby the amplitude and peak velocity of the movement vary systematically with the amplitude of the electromyography (EMG) activity [74], suggesting common features between limb and speech movements. In adults with DS, degraded motor performance was observed in those adults who stutter compared to those with fluent speech [75]. Individuals with DS demonstrated dyskinesia, including slower initiation of movement, increased variability in movement trajectory, and bradykinesia for both limb and speech movements [76]. Although some unique control elements may exist for a specific behavior, these findings show interactions between limb and speech motor systems.

Recent neuroimaging techniques provide evidence that motor control for speech, nonspeech oral, and limb movement share certain underlying neural substrates. For instance, Broca's area (BA 44-45) is activated during speech production and complex hand movements [77], with increased blood flow during performance of grasping gestures [78]. Activation of the inferior frontal cortex, mesial and lateral premotor cortices, posterior and anterior regions of the superior temporal gyrus activities were found during finger tapping movements and rhythmic one-syllable speech production [79]. Despite the fact that speech and upper limb systems share some common features of motor control in health and disease, simple

extrapolation of findings from the limbs to the orofacial system is tenuous because of the significant differences in musculoskeletal anatomy, physiology, biomechanics, sensorimotor innervation, and task dynamics. Limb muscles have attachments to the skeleton via tendons and connective tissue, are arranged around joints into agonistic-antagonistic groups, and have muscle spindle-Golgi tendon afferents and spinal cord reflex mechanisms that are presumed to play a significant role in postural control, stiffness regulation, and load compensation [80]. The distribution of proprioceptors is most prominent among anti-gravity muscle systems (i.e., upper limb flexors, and lower limb extensors). Conversely, muscle fibers of the lower face (i.e., perioral muscles) do not have muscle spindles, tendon organs, or any known mechanoreceptor mediating a monosynaptic stretch reflex as typically found among limb muscles [39,81]. Unlike the limb system, perioral muscle fibers vary greatly in plane and orientation and do not have well defined insertion points [82]. For example, numerous perioral muscle fibers have their origins and insertions into the integument of the skin, or are linked to suspensory tendinous nodes (e.g., modiolus located lateral to the oral angle) [83]. As a result, estimating orofacial muscle tone from whole-body measurements of stiffness is not accurate.



**Figure 1:** A simplified schematic model for the regulation of the locomotion (left) and speech (right). This schematic model was based on the literature suggesting that locomotion is largely dependent upon the central pattern generator (CPG consisting of networks of interneurons) which is premotor, providing inputs to the lower motor neurons within the spinal cord [60]. The model on the right depicts circuits for respiration (rCPG), sucking (sCPG), chewing (mCPG), and swallowing (swCPG) which are located in the brainstem [19,21,61-62]. The role of brainstem and/or corticobulbar CPGs in the production of speech and voice remains unknown. Each box represents a set of neurons in the model, and arrows correspond to major synaptic projections. Both locomotion and speech movement involves cortical and subcortical systems that projects to  $\alpha$ -lower motor neurons for postural and phasic motor output.

The ability to modify stiffness could serve to reduce variability during speech production and play a role in achieving the motor precision necessary for different speech sounds [84]. For example, the tongue must be precisely positioned relative to the palate to produce fricative consonants. During vowel production, however, acoustic variation is more sensitive to changes in the degree of vocal tract constriction rather than to the location of the constriction along the vocal tract [85-86]. Likewise, when the jaw was forced open by external mechanical perturbation during the production of the bilabial fricative consonant / $\Phi$ /, passive properties (stiffness) of the lips and jaw contribute as a compensatory mechanism for accomplishing speech tasks [87-88]. Alterations in stiffness, including increased or decreased stiffness are regarded as a negative motor sign which may contribute to dysarthria [15].

## Muscle Tone in Limb System and Speech Impairment

Findings from the limb systems in an attempt to understand the relation between hypotonia and postural control may provide insights towards the development of physiologically-based assessment tools for the orofacial mechanism. For instance, Davis and Kelso (1982) [89] showed that individuals with DS were able to up-regulate stiffness when asked to tense their right index finger muscles against load change. However, they did not increase stiffness to the same degree as normal controls and were

found to be impaired on the precise specification of stiffness tasks, suggesting the impairment affects the scaling dynamics of muscle co-contraction with sufficient duration or intensity [89]. This study suggested that aberrant muscle stiffness in the digits of individuals with DS could be a sensitive index of hypotonia.

**Table 1:** Summary of limb and orofacial system that shows cross-system evidence.

Source	Motor system	Method	Summary of results
Gentilucci et al. (2001) [72]	<ul style="list-style-type: none"> <li>● Finger reaching and grasping of objects</li> <li>● Lip movement and vocal loudness</li> </ul>	<i>Kinematic:</i> Three-dimensional (3D)-optoelectronic ELITE system (B.T.S. Milan, Italy)	Mouth opening and sound production were affected by the grasped object size
Ostry & Kelly (1983) [73]	<ul style="list-style-type: none"> <li>● Tongue</li> </ul>	<i>Kinematic:</i> Tongue dorsum movements during consonant--vowel syllables in which speech rate and stress were varied were studied with pulsed ultrasound	Kinematic patterns of tongue dorsum movements were comparable to those observed in the rapid movement of the arms and hands

A muscle activation abnormality has been linked to increased co-activation of antagonistic muscles (i.e., simultaneous activation of muscles acting in opposite directions) in individuals with DS during anticipatory postural adjustments where participants were asked to performed bilateral shoulder flexion and extension movements while standing on a force platform [90]. Recently, Gontijo et al. (2008) [91] proposed that elevated co-contraction was a

compensatory neural mechanism for low muscle stiffness in DS. This compensatory mechanism also shows that the motor control system in DS exhibits plasticity, being adaptive and responsive to training.

Prior to training an arm flexion task, Almeida et al. (1994) [12] found that 3 of 8 DS participants showed increased muscle co-contraction, whereas none of the normal controls demonstrated this pattern. With training, individuals with DS increased the quantity of agonist activity and decreased antagonist activity; achieving motor performance similar to their normal control counterparts. For this study, simple limb training exercises resulted in a reduction in antagonist muscle co-activation among some DS participants. It is unknown whether a similar therapeutic approach would yield benefits among orofacial muscle systems.

Clinically, individuals with DS have been reported to show impaired strength and precision of articulatory movements contributing to dysarthria and poor speech intelligibility [92]. Hypotonia is presumed to underlie these speech production difficulties, manifest as reduced stiffness and an inability to modulate tension among orofacial articulators which contributes to presumed weakness of the orofacial musculature and possible hyposensitivity of the lips [93]. It should be noted here that Yarter (1980) [93] did not measure orofacial muscle force nor cutaneous sensitivity, but did offer the supposition that hypotonia may be due to reduced stiffness and lip stimulation/

exercises designed to strengthen the lip/sucking abilities should be initiated in the first year of life.

One study suggested that altered jaw stiffness did not affect orofacial muscles enough to influence speech production in children with DS [94]. Although the pattern of muscle co-activation in this study did not reflect the hypothesis that hypotonia affects jaw movement sufficiently to influence speech production, these authors suggested that children with DS may rely on muscle co-contraction to a greater extent than do age-matched controls, as a compensatory mechanism for hypotonia. These findings are consistent with the idea that increases in jaw mechanical stiffness by co-activation of agonists and antagonists facilitates rapid movements of speech production [95]. Increased muscle co-activation in the jaw appears to be consistent with a gait acquisition study where toddlers with DS showed greater lower limb (i.e., hip, knee, ankle and leg) co-contraction indices (CCIs) during the swing phase of locomotion [91]. No comparable biomechanical data exist for the tongue and perioral muscle systems in children with DS. Future studies with larger participant groups should be conducted using contemporary biomechanical methods to generate the primary data needed to accurately model the potential relation between reduced stiffness (hypotonia) and orofacial kinematics.

## Conclusion

Orofacial muscle tone per se is rarely assessed using quantitative biomechanical measurements due to lack of instrumentation and real-time data acquisition and analysis software for routine clinical use. Most studies of the oral mechanism in DS are based on gross anatomical observation, subjective estimates of muscle 'resistance', or observation of orofacial posture (facial droop, tongue protrusion, open jaw) or visual impressions of posture rather than biomechanical measurements of passive and active force, or stiffness [96-97]. In some cases, the evaluation of orofacial muscle strength was inferred from clinical ratings of facial movements [36,41]. In sum, current experimental data which indicate a causal relation between hypotonia and speech impairment are generally lacking for individuals with DS. Thus, an informed clinical intervention can only be developed following a comprehensive investigation of the movement disorder, including biomechanics, vocal tract performance anatomy, sensory capacity, muscle physiology, sensorimotor integration, and kinematics to better understand speech production in individuals with DS.

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