# Probing production-perception relationships in children with myotonic dystrophy

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## ABSTRACT

Myotonic dystrophy, a neurodegenerative disease that causes muscle weakness and difficulties with muscle relaxation after contraction, frequently affects orofacial articulatory dynamics, leading to decreased speech intelligibility, particularly in children. We investigated the effects of myotonic dystrophy on sensorimotor relationships in children's speech by studying compensations for a lip-tube perturbation. Nine French-speaking children diagnosed with myotonic dystrophy and nine age-matched typically developing children were recruited. They were asked to produce repetitions of the vowel /u/ with and without a 17-mm-diameter tube inserted between the lips. A synchronized ultrasound system, Optotrak motion tracking system, and audio recording system were used to track lip and jaw displacement, as well as tongue shape and position. Combined with articulatory and acoustic results, perceptual ratings of the produced vowels suggest that children with myotonic dystrophy rely more on auditory feedback than their typically developing peers.

**Keywords**: clinical speech, production-perception relationships, lip-tube perturbation, speech development.

## 1. INTRODUCTION

It is well established that speech production and perception processes interact with each other. From a theoretical point of view (for a review see [1]), during speech production, the central nervous system generates an efferent signal, corresponding to the estimated auditory (or somatosensory) consequences of the articulatory movements. This efferent signal is then compared to the actual auditory (or somatosensory) feedback signal produced by the speaker (reafferent sensory input). When a discrepancy is observed, an error signal occurs and the motor system adapts to minimize the discrepancy between the expected and actual auditory or somatosensory feedback. In the course of speech development, children first learn to control phonemic goals through feedback-based mechanisms (auditory and somatosensory). Although several studies have used the artificial perturbation paradigm to probe the contribution of each of those mechanisms (feedback-based and feedforward) to speech production in typically developing speakers, much less is known about speakers with a motor control impairment. In this paper, we report on a study of speech production and perception using a lip-tube perturbation with children diagnosed with myotonic dystrophy.

### 1.1. Myotonic dystrophy

Myotonic dystrophy (MD) is a genetic neuromuscular disorder characterized by muscle weakness (myopathy) and difficulty relaxing muscles (myotonia). Myotonic dystrophy type 1, also known as Steinert's disease, is the most common form of muscular dystrophy. Its global prevalence is estimated at 1 in 20,000 inhabitants, but its proportions are greater in certain regions of Quebec, particularly the Estrie region, where the approximate prevalence is 1/1,500 ([2]), and in the Charlevoix and Saguenay-Lac-St-Jean regions, where the prevalence is approximately 1/600 ([2, 3]). One of the most common features in people with MD is bilateral weakness of the facial and jaw muscles. Children with MD often display reduced labial and lingual displacement in non-speech tasks, and produce speech sounds perceived as being less intelligible than those produced by typically developing peers ([4, 5]). To our knowledge, no data on French-speaking children are available. To gain a deeper understanding of the speech motor abilities of children diagnosed with this medical condition, a group of speech language pathologists from CHU Ste-Justine in Montreal reached out to us to gather quantitative data on their patient cohort. Consequently, the study was collaboratively developed by both clinicians and researchers.

### 1.2. The lip-tube perturbation paradigm

Perturbation experiments have been widely used to study reliance on sensory feedback ([6]). A lip tube is an appropriate perturbation to affect the geometry of the vocal tract, since it increases or decreases the lip area compared with the typical configuration. In the French vowel /u/, for example, the lips are rounded so that the lip area is small (around 0.3 $cm^2$ for adults; [7]); the associated tongue position is elevated towards the velopalatal region. It has been found that, when a 2.5-cm-diameter lip tube keeps them from rounding their lips, French adult speakers who are asked to produce /u/ cannot immediately compensate for the perturbation. After a learning session, during which a gradual improvement in the quality of /u/ was observed, some speakers proved better able to compensate than others; their compensation strategies involved a high and backward movement of the tongue towards the velo-pharyngeal region as a mean to correct for the auditory feedback error ([7, 8]).

A more recent study using a lip tube with children ([9]) showed that 4-year-olds could successfully compensate for a lip-tube perturbation (as assessed by the intelligibility of their produced vowels), but the compensation was based on trial and error and did not seem to be guided by a systematic error correction mechanism. We would expect a systematic correction mechanism to be associated, first, with a continuous evolution towards a better production of the vowel and, second, with the end of the correction process once the production is considered to be appropriate. It is important to note that, in both children and adults, compensatory responses were never complete: acoustic and articulatory values in the lip-tube condition remained significantly different from values in the normal pre-perturbed condition. This difference could be ascribed to the competing demands of somatosensory feedback, which limits articulatory displacement (which is required to compensate for auditory feedback error).

The objective of this study is to investigate the compensatory response of children with MD and typically developing children to a lip-tube perturbation during production of the French vowel /u/. The effects of the compensation will be assessed with acoustic, articulatory, and perceptual analyses.

## 2. METHODS

A speech production task and a perceptual task were administered to different groups of participants.

### 2.1. Speech production

#### *2.1.1. Participants*

Nine children ranging in age from 6 years old to 9 years old (mean age: 7.6; 5 males) were recruited at Ste-Justine paediatric hospital in Montreal (the “MD group”). These children had received a diagnosis of MD several years ago. Nine age-matched and gender-matched typically developing children were also recruited. They reported no speech or language disorder. All children were native speakers of Canadian French and were screened for auditory thresholds. The study received approval from both UQAM and the hospital's Institutional Review Boards. Additionally, all parents of the participating children provided written consent for the experiment.

#### *2.1.2. Experimental procedure*

Children had to produce 30 tokens of isolated /u/, in three conditions: first, without the lip tube, in a normal condition (referred to as the normal pre-perturbed condition). In the second condition, children had to produce the vowel /u/ 10 times with a 1.7-cm-diameter lip tube between their lips (referred to as the lip-tube condition). This diameter was chosen based on the results of a previous study with typically developing children ([9]). In the third condition, children were asked to produce another set of 10 /u/ vowels without the lip tube (referred to as the normal post-perturbation condition).

#### *2.1.3. Data analysis*

Articulatory and acoustic data were recorded using a combined ultrasound system (Sonosite 180Plus) and Optotrak system (NDI Certus). The synchronized acoustic signal was also recorded using a SHURE unidirectional microphone, with a sampling frequency of 22050 Hz. Ultrasound images corresponding to each vowel’s midpoint were selected. Tongue contours were semi-automatically tracked using the GetContour program (described in [10]) and corrected for head movements using the Optotrak measures (HOCUS; [11]). Measures of tongue maximal curvature index (MCI) and tongue height, as described in [10], were also extracted. Acoustic data were analysed using Praat ([12]). The first (F1) and second (F2) formant frequencies were also extracted at the vowel midpoint, using the Burg algorithm embedded in Praat. In order to account for between speaker variability in vocal tract size, all data were normalized by dividing the value of a given trial by the mean value measured in the normal pre-pre-perturbation phase.

Linear mixed-effects models ([13]) were built using R. The dependent variables were articulatory and acoustic measures (one model for each variable), and the independent variables corresponded to the experimental phase and the speaker group. The participant was included in the models as a random effect (slope).

### 2.2. Perceptual assessment

For each participant, the first two and last two trials of each experimental phase (normal pre-perturbation, lip-tube perturbation, normal post-perturbation) were selected and used as stimuli in a speech perception test administered to 20 French-speaking adults. Listeners did not report any speech or language disorder or any hearing deficits. They ranged in age from 20 to 29 years old. Stimuli were presented in randomized orders. Listeners' first task was to indicate, by selecting the corresponding button on a computer screen, whether the vowel they had heard was /u/ or not (open choice). Then they had to rate the quality of the vowel on a scale ranging from 1 (poor) to 7 (excellent). The percentage of /u/ responses and average quality ratings were calculated for each trial. Linear mixed effects models were built for each of the dependent variables (percent correct and quality rating), with experimental phase and speaker group as the independent variables and participant as the random effect.

## 3. RESULTS

### 3.1. Speech production results

The mean F2 values for each trial, averaged across speakers within each group, are presented in Figure 1. As the graph shows, control and MD groups produced higher values of F2 in the lip-tube condition than in the normal pre-perturbation condition ($\chi^2(2)=38.62$; $p<0.001$). This pattern can be interpreted as reflecting incomplete compensation. However, unlike the control group, the MD group had significantly reduced F2 values during the 10 trials with the lip tube. This effect was significant ($\chi^2(5)=15.58$; $p<0.01$). In the normal post-perturbation condition, no significant difference was observed between groups, nor between this condition and the normal pre-perturbed condition.

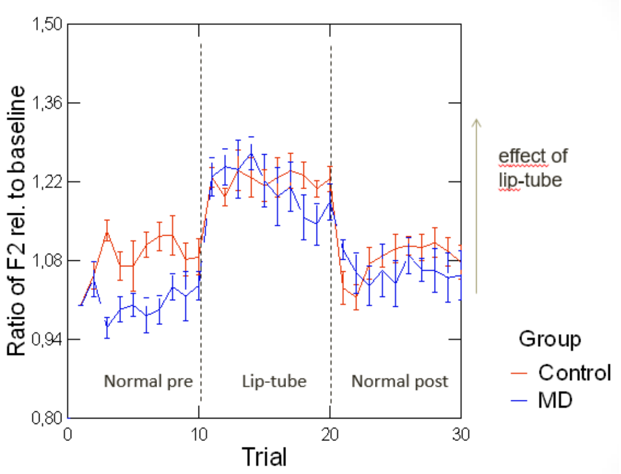

**Figure 1**: Mean normalized values of F2, averaged across speakers within each group.

The mean F1 values, depicted in Figure 2, show a similar pattern. Children with MD produced significantly lower F1 values than typically developing children in the lip-tube condition ($\chi^2(2)=37.91$; $p<0.001$). Furthermore, as was the case for the F2 values (Figure 1), F1 values decreased significantly over the course of the perturbed condition for speakers of the MD group only ($p<0.01$).

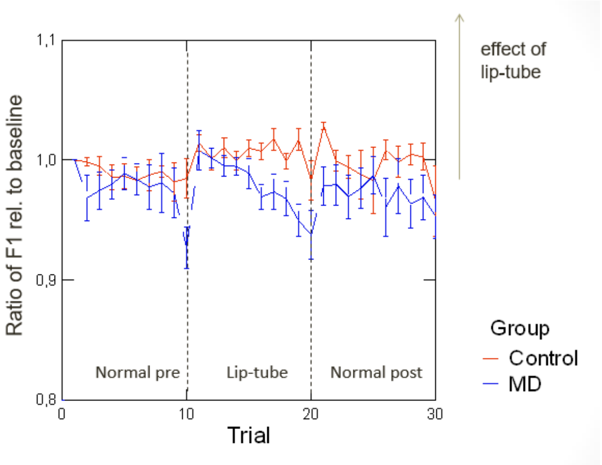

**Figure 2**: Mean normalized values of F1, averaged across speakers within each group.

In the articulatory domain, tongue displacement showed a similar trend (Figure 3). The expected compensatory manoeuvre –elevating the tongue – was produced in the lip-tube condition by children with MD, but not control children ($\chi^2(5)=21.42$; $p<0.001$). Again, a significant relationship between trial number and tongue height for the 10 trials of the perturbed condition ($p<0.01$) was observed only for the MD group.

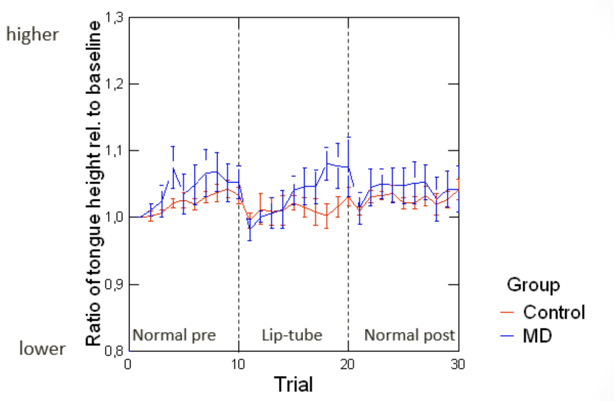

**Figure 3**: Mean normalized values of tongue height, averaged across speakers within each group.

### 3.2. Perceptual results

The perceptual test did not reveal any significant differences between the percentages of correct responses for /u/ produced by the children with MD and the typically developing children. However, mean quality-rating values exhibit different patterns, as shown in Figure 4. A significant effect of condition is noted, with /u/ produced in the lip-tube condition being perceived significantly better than /u/ produced in either of the two normal conditions ($\chi^2(2)=24.15$; $p<0.001$). Furthermore, children in the MD group produced /u/ vowels that corresponded overall to lower quality than their typically developing peers ($\chi^2(3)=25.42$; $p<0.001$). Importantly, a significant effect of the interaction between trial and group was noted in the lip-tube condition ($\chi^2(5)=28.42$; $p<0.001$): for the MD group only, the last two trials received higher scores than the first two trials, confirming the tendencies described above in the articulatory and acoustic domains.

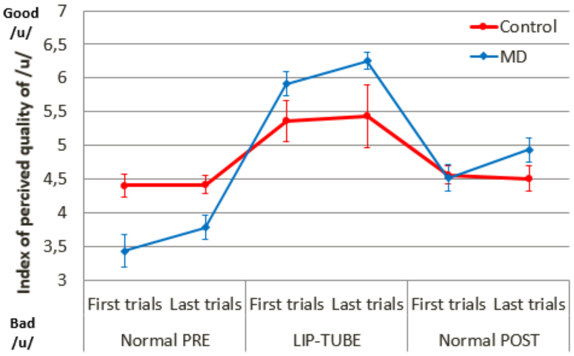

**Figure 4**: Mean quality rating values of /u/, averaged across speakers within each group.

## 4. DISCUSSION

In this study, we used a lip-tube perturbation to examine production-perception relationships in children with motor deficits (myotonic dystrophy) compared to typically developing children. Our results showed that children with MD exhibited better abilities than age-matched typically developing children in using their auditory feedback to compensate for the lip-tube perturbation. Indeed, despite the reduced motility of the tongue reported in the literature, children in this group were able to modify their tongue position and reduce formant values in the expected direction in the perturbed condition. This pattern emerged gradually from the first to the last trials, suggesting a sensory-based error correction mechanism. Importantly, typically developing children did not display this compensatory response, in line with previous studies ([9]).

Our results also provide information about the reliance on auditory and somatosensory feedback in children with MD. In line with Lametti et al., 2012 ([14]), we can consider the magnitude of the compensatory response as an index of sensory reliance in both speaker groups. Since children with MD produced a greater compensation in the acoustic dimension, they seem to rely more heavily on auditory feedback than typically developing children when producing French /u/. However, since the MD group produced larger tongue displacements compared to the normal pre-perturbed condition than the control group, they induced a larger discrepancy between the actual somatosensory feedback and the somatosensory feedback predicted by the central nervous system (see section 1). Children with MD thus attached less weight to somatosensory feedback than typically developing children.

The results reported here could have important consequences for rehabilitation strategies clinicians use for children with myotonic dystrophy. Indeed, since an artificial perturbation such as a lip tube elicits larger displacement of the tongue, more exercises of this type, taking advantage of the strong reliance of children with MD on auditory feedback, could be administered to ensure these children explore the entire lingual articulatory space and map it to their sensory experiences. Further experiments to assess the clinical value of such perturbed conditions are currently under way.

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