Three cases of focal embouchure dystonia: classifications and successful therapy using a dental splint

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Abstract

Background – Focal embouchure dystonia (FED) is a type of task specific dystonia affecting wind instruments players, and frequently ends the professional careers. The results of the treatments of FED in reported literatures are disappointed. Objective – We report three patients with FED who were brass instrument players. In one patient, we evaluated the usefulness of dental splint. Methods and Results – Patient 1 was a 28-year-old trumpeter who was suffering from an intermittent tremor of the lower jaw. Following the usage of a dental splint, her symptoms completely disappeared. Patient 2 was a 29-year-old horn player with atonia of the lower facial muscles. In this case, medication with various drugs brought no benefit, and he ended his career. Patient 3 was a 43-year-old trombone player who suffered involuntary contractions and relaxations of perioral muscles along with clumsiness of tongue movement. Extraction of double teeth and long-term cessation of playing for twenty years did not bring any positive effects. Conclusion - We propose that FED can be categorized into three subgroups according to masticatory, facial, and lingual types, and that the dental splint might be a useful therapeutic strategy for the masticatory type of FED.

Key words: focal embouchure dystonia (FED), sensory trick, dental splint
Introduction

Playing a brass instrument puts extraordinary demands on the muscles of the face, jaw, and tongue [1]. The term embouchure describes the oral mechanisms for playing a wind instrument, and employs the lips, tongue, and lower facial muscles, as well as jaw and teeth structures [2]. The embouchure not only directs the considerable force needed to produce the sound, but also exquisitely modulates airflow, changing timbre, pitch, and articulation [1]. Task Specific Dystonias (TSDs) are a subset of action dystonias that occur exclusively or primarily when the patient is performing a specific task [3]. Focal dystonia in musicians, also known as musician’s dystonia (MD), is a type of TSDs, which presents itself as muscular incoordination or loss of voluntary motor control of extensively trained movements while a musician is only playing the instruments [4]. Focal embouchure dystonia (FED) is a type of MD affecting musicians who play wind instruments [5]. It is often occupationally induced by extensive, repetitive, and effortful movements [6], and is characterized by playing-related inappropriate control of the orofacial muscles that adjust the airflow into the mouthpiece [7]. The pathophysiological mechanism of MD, including why it is task specific, remains to be clarified, but recent studies have indicated that the functioning of the motor and somatosensory system during playing instruments may be abnormal in such patients as follows: 1) reduced inhibition at different levels of the central nervous system, 2) maladaptive plasticity and altered sensory perception, and 3) alteration in sensorimotor integration [4]. For example, using functional MRI, Haslinger and his colleagues studied sensorimotor activation patterns in brass players with FED [7]. Compared with healthy brass players, patients with FED showed significantly increased activation of somatotopic face representations within the bilateral primary sensorimotor cortex and of the bilateral premotor cortex. Overactive premotor cortices has been interpreted as an abnormal modulation of the cortico-striato-pallido-thalamo-cortical circuit in dystonia. So, the authors suggested that sensorimotor overactivity could reflect deficient subcortical and intracortical inhibition as well as abnormal sensorimotor integration and reorganization in musicians with FED. In addition to abnormal motor and sensory function mentioned above, a genetic contribution to MD was also reported [8]. In psychological studies, musicians with dystonia have more anxiety and perfectionist tendencies than healthy musicians [9]. This finding strengthens the assumption that behavioral factors may be involved in the etiology of MD.

The incidence of FED is unknown, but according to a review by Brandfonbrener [10], FED was observed in 16 patients among 113 musicians with focal dystonia (14.2%). It takes 8.3 years on average (range: 0-28 years) for patients with FED to consult a medical doctor for the first time [11], so many individuals are suffering for a long time
without necessarily being aware of any underlying pathology. There is general agreement that, once present, symptoms of FED do not improve [11], and this disorder frequently ends the professional careers of brass or wind players who become affected [6]. Frucht [11] described 26 patients with FED and, according to their symptoms, categorized them into three subgroups: tremor of lips (T-type), lateral pulling of the lips (LP-type), and isolated involuntary movements of the jaw (J-type). Therapies for FED have been disappointing [1]. Medications such as anticholinergics (e.g., trihexyphenidyl) [12], levodopa, baclofen [5], and gabapentin [3], as well as direct injection of Botulinum toxin A [1,5,11], had only a limited effect on the observed dystonia. In contrast to traditional pharmacotherapies, sensory tricks are often useful in ameliorating symptoms. Sensory tricks are defined as simple touches or stimulations near the area of abnormal movement that can decrease the dystonia [12]. Frucht [11] reported that among their four FED patients belonging to J-type, placing a straw or piece of plastic between their teeth improved the abnormal movements in two patients. Frucht [11] also tried using a larger-diameter mouthpiece (for example, a typical trombone mouthpiece for a horn player). Exposure to cold temperatures also produced a transient relief of symptoms [5]. Thus, a great deal of effort has been expended to uncover possible treatments for FED, but as far as we know no cases have shown complete remission.

We encountered three patients of FED among brass instrument players, and were able to see complete remission in one patient by the use of a dental splint. This is the first reported case of a FED patient whose symptoms have completely disappeared. Below we present the clinical courses of each patient and describe the effectiveness that was brought by a dental splint in one patient.

**Patient 1**

A 28-year-old, right-handed female, who was a professional trumpeter, presented herself to our hospital with a 2-month history of progressive difficulty in performing. At first, she felt that her embouchure was simply becoming uncomfortable. But a week later, involuntary tremor of her lower jaw appeared, and then gradually worsened. Upon examination, there was no dystonia of the lips, face, or tongue with routine movement. Speech was normal, there were no perioral or facial sensory deficits, and no facial weaknesses were present. However, when she played the trumpet, there were involuntary 1-3 Hz dysrhythmic tremors of the lower jaw, which appeared symmetrically and intermittently. A typical attack consisted of 1-6 tremors and lasted 1-5 seconds. She could produce the intended sound itself, but the jaw tremor caused the production of unnatural and nonmusical sound vibrations, moderate deterioration of the
control of high and low pitch tones, and mild deterioration of the timbre and quality of sounds. The patient started to play the trumpet at age 10, and had typically practiced for 3-8 hours per day. Immediately before the onset of symptoms, she did not significantly alter her embouchure or prolong her playing time. The patient was neither exposed to neuroleptics nor did she have a family history of dystonia or any perioral trauma. Brain magnetic resonance imaging (MRI) showed no abnormalities. During the performance, brass instrument players have to blow the air into their instruments, so they are exposed to the hyperventilation state which decreases the threshold of epileptic discharge. So, we performed the electroencephalography (EEG), including hyperventilation stimulation, and it revealed normal findings.

The therapy was carried out by the use of a dental splint. We noticed that, upon mildly biting a piece of cotton, her jaw tremor was reduced while playing. In order to make the dental splint, we referred her to the department of dental surgery in our hospital. When playing brass instruments, the degree of mouth opening is different depending on the desired pitch of the tone. When the player sounds a low versus a high tone, the mouth is opened widely or narrowly, respectively. In other words, the distance between the front teeth of the upper and lower jaw is relatively long in low tones and short in high tones. With this in mind, we used the elastic material ethylvinyl acetate in making the dental splint. First, a gypsum model of the upper and lower teeth was made. Then, four dental splints that covered the two molars of the bilateral upper and lower jaw were made out of ethylvinyl acetate (Fig. 1). She applied these splints with the assistance of a dental surgeon who made sure that her upper and lower jaw was set at the best position. As a result, her upper and lower front teeth were 6.5 mm apart when playing low tones, and 2.5 mm apart for high tones. The dental splints were completed 1 month after her first consultation, and her lower jaw tremor did not appear while she had them on. Since then, she has always used them when playing her trumpet. Within 2 months (approximately 3 months after her first consultation to our hospital) her abnormal movements began to disappear. Moreover, 6 months after her first consultation (5 months after beginning the usage of dental splints) her symptoms had entirely disappeared. She gradually decreased the use of her dental splints over a few months, and then completely stopped the intervention. Since then, reoccurrence of symptoms has not been observed for 2 years.

**Patient 2**

The patient was a 29-year-old, right-handed professional horn player. Two years before the first consultation at our hospital, he developed difficulty in playing due to an atonia of lower facial muscles with significant air leak from the corners of his mouth. At
first, he felt that his embouchure was simply unstable, but the atonia gradually worsened. Two weeks later, he could not play even a single tone at all. He had resorted to several therapeutic strategies such as massage, acupuncture, and moxibustion, with no effect. One year after the onset, he significantly altered his embouchure, and became able to play some tones within an octave, but his performance could only resemble that of a beginner. Upon our examination, there was no dystonia of the lips, face, or tongue with routine movement. His speech was normal, there were no perioral or facial sensory deficits, and no facial weakness was present. When he played the horn, the lower facial muscles were completely relaxed, and his cheeks were expanded bilaterally. Air leaked from the bilateral corners of his mouth. In comparison, the upper facial muscles were tightly contracted, which was the primary gain in voluntary movement resulting from his changing his embouchure. When he adjusted his embouchure in the mirror without the mouthpiece or instrument, he could correctly control his embouchure. But, as soon as the mouthpiece touched his lips, his embouchure was distorted out of shape. He could only play some tones within the midrange, but could not play short easy phrases. He did not show a lower jaw tremor. He started to play the horn at age 12, and had practiced for 5-8 hours a day. Before the onset of symptoms, he did not significantly alter his embouchure or lengthen his playing time. He was neither exposed to neuroleptics nor did he have a family history of dystonia or any perioral trauma. Brain MRI and EEG showed no abnormal findings.

Medications such as trihexyphenidyl, levodopa, clonazepam, valproate, diazepam, and Selective Serotonin Reuptake Inhibitors (SSRIs) were prescribed, but these drugs had little effect on the abnormal movements. Various sensory tricks, such as biting on a piece of cotton, playing with larger mouthpieces, and cooling his facial muscles, also did not show any effect. Three years later (five years after symptom onset), he decided to end his career as a professional horn player with great disappointment.

**Patient 3**

The patient was a 43-year-old, right-handed man. At age 18, he entered a musical college and played trombone. One year later, he realized a difficulty in performing. He first felt that his embouchure was uncomfortable, and then involuntary and abnormal contractions and perioral muscle atonias appeared. When he played the trombone, the open space between his lips became narrow and round, and the bilateral corners of his mouth were relaxed. Lower jaw tremors were not observed. At first he only had trouble with playing some tones in midrange, but symptoms gradually worsened accompanied by aggravating air leaks. Tone onset was unclear because of the clumsiness of tongue movement. He changed and retrained his embouchure a few times. Each change brought
some improvement, but only for a few months, followed by a worsening of symptoms. At age 21 (two years after the onset of symptoms) he could not produce tones at all. He had his double teeth removed in a dental clinic with no effect. He graduated from the musical college with minimum scores, and, since then, has not played trombone for twenty years.

At age 42, he recognized his symptoms as FED and consulted our hospital. He wanted to try pharmacological approaches for the disorder. We prescribed trihexyphenidyl, levodopa, and diazepam, in that order. However, no improvement was observed. Like 20 years ago, he could not play a single tone at all. Various sensory tricks tried in the patient 2 also did not show any effect. He completely gave up playing the trombone.

Discussion

Table 1 summarizes the characteristics of our patients. It is noteworthy that the symptoms of patient 1 completely disappeared by the use of a dental splint. As far as we know, this is the first case of FED where therapy was successful. Below we discuss the subtype classifications and the recommended therapy for a certain subtype of FED.

Many muscles participate in the formation of embouchure. According to the symptoms, Frucht [11] categorized FED into the following types: tremor of lips, lateral pulling of lips, and isolated involuntary movements of the jaw. However, considering the lack of the lingual symptoms by Frucht’s classification, and based on the participating muscles and their innervations, we propose that FED could be classified into the following three subgroups: masticatory, facial, and lingual type. First, the masticatory muscles (namely the masseter, temporal, and pterygoideus muscles) are innervated by the first branch of the trigeminal nerve. By stabilizing the position of the lower jaw, these muscles build the “frame” of the embouchure. The involuntary lower jaw tremor observed in patient 1 might be caused by dysfunction of these masticatory muscles. Second, the facial muscles innervated by the facial nerve could play a central role in FED. Facial muscles include the orbicularis oris, levator labii, zygomaticus major and minor, buccinators, depressor anguli oris, and many others [11]. When playing brass instruments, sound is produced by the vibration of the lips. In order to produce discrete sounds, a fine balance of tension and relaxation of lower facial muscles is necessary. As observed in patients 2 and 3, the dysfunction of lower facial muscles might seriously impair sound production itself. Finally, the tongue is used to create a clear start to a note, which is called “tonguing”. The lingual muscles are innervated by the hypoglossal nerve. Clumsiness of tongue movement, also known as dysfunction of tonguing, was observed in patient 3. Though other muscles might participate in
embouchure (such as the orbicularis oris and platysma muscles), we might say that embouchure is primarily formed by three components: the frame by masticatory muscles, the sound production via the lips controlled by facial muscles, and the tonguing by lingual muscles. The dysfunction of any or all of these components might contribute to FED.

It must be noted that, for the masticatory type of FED, therapy with the use of a dental splint was beneficial. We believe that the different responses to intervention of dental splint between our patient 1 and patient 2 and 3 was caused by the differences in clinical characteristic. First, the symptom of FED was restricted to jaw tremor in patient 1, but the patient 2 and 3 showed other symptoms, such as atonia and/or involuntary contraction of perioral muscles, and clumsiness of tongue movement, without jaw tremor. Second, as shown in the clinical course of each patient, the improvement of symptom by mildly biting a piece of cotton while playing instruments was only observed in patient 1. In the patient 2 and 3, various sensory tricks did not show any effect. This effectiveness might be brought on by two mechanisms. First, by biting the dental splint, the lower jaw was mechanically stabilized, so that the jaw tremors could be prevented. While playing brass instruments, the relative positions of the upper and lower jaw need to be changed finely and frequently according to the desired pitch of tone. In this regard, the use of elastic material for the dental splint made fine control possible. Second, we can say that sensory tricks may also participate in the remission of the masticatory type of FED. There are several reports evaluating the efficacy of sensory tricks using either a piece of plastic [11], large diameter mouthpieces [11], or a 5 minute-cooling of perioral muscles [5] for the treatment of FED. However, an inflexible piece of plastic does not permit adequate mobility of the lower jaw, the larger mouthpiece could not be used in an actual performance, and the effect of cooling is only transient. The involuntary muscle contractions of FED could be due, at least in part, to abnormal sensory processing of spindle afferent information [13]. The use of dental splint might influence such information. As shown in the Introduction, the MD might be caused by abnormal function of cortical and subcortical system. So, the alteration of sensory perception and/or sensorimotor integration by the use of dental splint might bring about the good response in patient 1. In short, by using the dental splint, the frame of the embouchure made by masticatory muscles might be stable both structurally and functionally.

There are some limitations in this study. The first is the sample size. As demonstrated by the typically long duration between the onset of symptoms and the initial consultation in Frucht’s report [11] and also in patient 3 in the present study, FED is not well known to wind players, and may go underreported. The sample size of the present
study is almost the same as the previous ones with a single or double case reports [1,3,5,14]. Second, needle electromyographies (EMGs) were not carried out for our patients. In involuntary movement disorders, the needle EMG can provide much useful information. Abnormal contractions and relaxations of several muscles, and/or a dysregulated balance between them, might underlie FED. In order to clarify the relationships between symptoms and responsible muscles, it is necessary to assess the needle EMG of each muscle during performance. As described above, so many muscles participate in the formation of embouchure that there are only a few data of muscle activity in normal subjects [15]. All of our patients refused to undergo needle EMG recordings. Third, the duration after the onset of FED was relatively short in the patient 1 compared with the patient 2 and 3. It is possible that the difference of the duration from the onset of FED might participate in the difference of treatment effect between the patient 1 and patient 2 and 3. Finally, repetitive transcranial magnetic stimulation (rTMS) was not performed. Some investigators have reported that rTMS was effective in treating writer’s cramp, which is one of the most prominent and famous symptoms of TSD [16,17]. To date, there are no studies that have investigated the effects of rTMS on FED. We have no established strategy for the treatment of FED which has high impact on musical career of the patients. So, we should apply any therapeutic strategy which might be useful to reduce the symptom of FED.

In conclusion, we report three cases of FED. We propose that FED could be categorized into three subgroups according to masticatory, facial, and lingual types. For one patient with the masticatory type of FED, a dental splint was effective, and sensory tricks might have played a role in this improvement. As far as we know, this is the first case of FED that completely subsided. The other two patients with symptoms affecting facial and lingual muscles and longer etiology showed no improvement from either the sensory tricks or various medications. The ultimate goal of FED treatment is to prevent this disabling disorder from affecting musicians. The reduction of psychological stress will be needed to prevent the occurrence and worsening of FED. Compensatory techniques for dystonia by retraining their embouchure might be somewhat effective. It is important to expand knowledge regarding FED among wind or brass instrument players, for the early diagnosis and treatment.
References
Legend for Fig.1

Fig.1 Left: dental splints for patient 1. Upper and lower splints are for upper and lower molars, respectively. Left and right splints in the photo are for the right and left molars of the patient, respectively. Right: two molars of the bilateral upper and lower jaw are covered by splints.