Case Report

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A Pattern of Medication-induced Persistent Bulbar and Cervical Dystonia

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ABSTRACT

A longitudinal, analytical, and treatment oriented case study is presented describing the effects of a medication-induced impairment of neurological function. The subject is an elderly, Caucasian, female whose presenting symptoms included spasmodic eyelid movement; lack of muscle tone and control for specific muscles of the head and neck region; generalized hyperactivity; plus distortion of speech and deglutition. Clinical investigation determined that the subject's timed release antihistamine medication was the primary etiological factor in the presenting symptoms. Discontinuation of the medication resulted in a decrease, but not complete remission, of the symptoms. Clinical evaluation of the residual speech impairment included intensive therapy which was only minimally successful in reducing propositional speech distortion. The patient's neuropathy was inferred to be suprabulbar in site as a result of the speech analysis which is described as a reliable diagnostic aid in evaluation of severity of this type of neurological pathology. During the eight years of observation of this subject, documentation of the impairment was made by means of various media. Some of the results of this documentation accompany the study.

INTRODUCTION

The person of this report developed spasmodic dystonia in the lower bulbar (CN 7, 9, 10 and 12) and the cervical motor innervations during prolonged chlorpheniramine medication (Thach et al, 1975). The dystonia resembled that which may be induced by neuroleptic medications, particularly those of the phenothiazine

series (Crane, 1968 and 1973; Crane and Gardner, 1968; Crane and Naranjo, 1971; Chase, 1972). Like those, it has continued in an essentially consistent pattern, with distortions of speech and of mature pattern of feeding. The elementary performances of swallow, suckle, tidal respiration and maintenance of the pharyngeal airway were spared.

We report cinephotographic, cineradiographic and acoustic spectrographic observations of a particular pattern of dystonia of the mouth, pharynx and larynx and of abrupt darting motions of the tongue which are related in their occasions to voluntary performances. The details of their interactions with speech are particularly strategic as criteria of identification and of demonstration of this dyskinesia pattern.

CLINICAL HISTORY

The person of this report, a woman of 65 years, was referred to the Clinical Center, NIH, in November, 1971 for evaluation of involuntary facial movements of three years duration. In the Fall of 1965 she was given a prescription for a timed release capsule containing chlorpheniramine maleate 8 mg., phenylpropanolamine 50 mg. and isopropamide 2.5 mg., for treatment of allergic rhinitis. The medication was continued without

prescription. Within one year she began taking the capsules twice per day prophylactically. She was taking talbutal (5-allyl-5 secbutylbarbituric acid) 120 mg. each evening for insomnia from February, 1967. She was also taking chlordiazepoxide HC1 (10 mg. twice weekly) for nervous tension. She had taken dexamphetamine for six weeks in 1963 for weight control but had no further exposure to amphetamines or neuroleptic drugs (including phenothiazines) prior to onset of symptoms.

In the Fall of 1968 she developed blepharospasm, which continued in increasing but irregular incidence and severity; it was increased by conjunctival irritation, as from dust or soap. In 1970 she had involuntary opening of the mouth. Her speech gradually became less intelligible. Drooling, choking while swallowing and nasal regurgitation of liquids began in 1971 and increased to severely impair feeding. These impairments were increased by anxiety or fatigue and disappeared during sleep. Tranquilizing drugs (chlordiazepoxide HC1, diazepam), reserpine (40 mg. a day for eight weeks) and haldol (3 mg. $\times$ 3 for 10 days) produced no lasting change in her neurological impairment.

Her health was generally good throughout her lifetime. She was severely ill without neurological symptoms in 1918 for three days

*Illustrations by
Mrs. Diane Robertson Murayama*

at the time of an epidemic of influenza. A mild concussion was sustained in an automobile accident at the age of 33 years. Following complete dental extraction at the age of 25 years, she wore only an upper denture.

There was no family history of degenerative neurological disease.

Neurological Examinations*

At time of referral to the Clinical Center in 1971, the dyskinesia and associated impairments were conspicuous. The dystonia was externally evidenced by intermittent blepharospasm, platysmal contraction and prolonged yawn-like open-mouthed gaping. The blepharospasm was increased by tapping at the glabella. Cinephotographic studies of dystonic and darting motions of the tongue are described below. The tongue moved about extensively in the mouth. During the dystonia, a 2-4 per second undulating tremor involved the mandible, tongue and soft palate; the tremor was in the direction of the dystonia. Drooling continued from the open mouth. Oral spray was common during speech. Adjacent to overt dystonia, we occasionally noted licking and lip pursing gestures like those seen frequently in persons having oral dyskinesia following phenothiazine medication.

Akathisia was commonly present; she paced about the room, fidgeted in her chair or moved her hands. The rest of the neurological examination was negative, except for slight occasional fumbling and tremor in the hands. Manual coordination in writing, typing and fine needlework was normal. Notably absent were muscle rigidity, weakness or wasting, and evidences of impaired mentation or psychiatric disease.

No abnormalities were demonstrated by a detailed oral sensory and perceptual examination, by standard audiological and ophthalmological examinations, by hematological and chemical evaluations of blood, urine and cerebrospinal fluid, or by special studies which

included serum thyroglobulin and ceruloplasmin and a brain scan. The electroencephalogram was mildly abnormal with a diffuse dysrhythmia and an unusual frequency distribution, but no specific pathologic findings.

Course

Her continuing medications were discontinued in 1972. Within six weeks we noted diminution of oral and palatal dystonia and tremor, improved ability to maintain oral closure, and improved competence in speech and in biting and chewing. In January, 1973 she was hospitalized and given chlorpheniramine HC1 (4 mg. $\times$ 3 per day) in a double blind trial (Thach et al. 1975). One week after the first dose of medication, mandibular tremor returned. Marked worsening of blepharospasm and speech was noted by the subject and by observers. Recovery from this exacerbation promptly followed termination of the medication. After initial in-hospital trials of haldol, tetra-benzine, lithium and L-dopa, with little beneficial effect, neuropharmacologic efforts were discontinued; in the subsequent observation period medication was limited to propranolol for relief of cervical arthritic pain and chloral hydrate as a night sedative.

During the next 5 years, until the time of this report, her neurological pattern changed only slightly. The dystonia and associated disabilities varied in severity during the waking day, being minimal at morning waking and usually most severe in the afternoon and evening. The postural gestures of straightening the neck from lordosis commonly diminished severe dystonia in all of its elements. The dystonia was increased by psychic stress, such as apprehension, by physical exertion, as by carrying a load, or by exposure to cold. The dystonia was increased rapidly and specifically by feeding or speech, so that these performances were progressively impaired. Remarkably, the dystonia and disabilities were in near remission for periods of minutes or, occasionally, of several hours. The dystonia continued in its general pattern. The associated distortions of speech and of oral

feeding actions continued. The general variations in dystonia and related changes in particular performance competencies have been recorded in a daily diary for 7½ of the 8 years of observation.

She continued to have oral dysphagia and occasional choking episodes which seemed to be clearly related to attempted swallowing of insufficiently masticated food or of an overly large bolus. She devised circumstances of dining, and of physical composition of diet, which facilitated intake. During a social meal she would attempt little or no intake. When the oral dysphagia was severe, she would deliver pureed foods to the back of the tongue by use of an Asepto syringe; she was generally able to swallow the puree adequately, though occasionally the bolus would penetrate the palatopharyngeal isthmus or the larynx.

In the first year of our observation (1972-3), she reported occasional abrupt episodes of "clutching of the throat" without apnea and not related to feeding or speech. We were able to obtain no other observations suggestive of laryngospasm. In November, 1978, while dining in a restaurant with friends, she had an episode of apnea, cyanosis and generalized tonus followed by clonus. Competent attempts to extrude a possible food mass resulted in no evident expulsion. Respiration resumed spontaneously and she returned to her neurologic baseline in approximately 10-20 minutes. There were no other episodes suggestive of grand mal seizure disorder.

Pharyngeal airway maintenance was intact. She generally slept in recumbent position. However, when she was in severe dystonia at the end of the day, she initiated night sleep in semirecumbent position in a reclining chair. During sleep-EEG studies, we observed sleep in a variety of positions. It was associated with snoring but no apnea.

In 1972-1976, she reported paraesthesia in the anterior oral area, without distortion of taste. No objective changes in oral sensibilities were demonstrated. The spontaneous anteriorward or lateral-

*We acknowledge the contributions of Drs. Derek Denny-Brown, Thomas N. Chase, and Raymon B. Jenkins to the evaluations of this patient.

ward darting motions of the tongue disappeared early in 1979; the lateralward motions could no longer be elicited by touch.

She continued in good health during the years of our observation. A mild cervical osteoarthritis became symptomatic at times of

exacerbation of dystonia. Nasal allergic reactions to seasonal pollens continued. Nutrition was adequate, despite her dysphagia. Gross motor strength and stamina and fine manual skills continued essentially unimpaired. The akathisia diminished gradually in

the first 3-4 years of observation. Mental function and personality pattern remained unchanged. She has received counselling and supportive therapy and has made notably adequate adaptations to her feeding disabilities and to the problems of social interaction

Figure 1. Lateral views and radiographs of the head.

The illustrations in the upper tier are of the subject at ease, with eyes open, mouth closed and head in horizontal orientation. The illustrations in the lower tier are of the subject in position of severe dystonia, with eyes closed, mandible lowered, tongue retracted, pharyngeal palate elevated and the head in extension. In each tier, the schematic at the left is derived from the radiograph.

The central schematic, of the subject at ease, is supplemented by diagrams of the muscles which are inferred to effect the principal dystonic displacements as they are evidenced in lateral views and radiographs. These include the muscles of the mandible (particularly the pterygoideus lat.) of the hyoid (digastricus ant. and post., geniohyoideus and stylohyoideus), of the tongue (genioglossus, styloglossus, hyoglossus and lingualis longit.), of the pharyngeal palate (levator), the anterior cervical flexors (sternohyoideus, sternolaryngeus and thyrohyoideus) and the cervical extensor muscles.

Figure 2. Motions of the tongue as it attempts to approximate a tactor. Drawings of frames selected from cinema at 24 frames per second. Drawings are in sequence left to right across each line.

Frame 1. is reference showing approximation of the tactor to the tongue in midline. Subsequently, in attempting this approximation, the tongue tip is tensed into pointed contour and is protruded to the right of the tactor (Fr. 8), forcibly forward against the tactor (Fr. 14), inferior (Fr. 22) then superior (Fr. 28), pushing the tactor rightward. The tensed tongue tip is pushed against the tactor in its lateral position (Fr. 38), protruded inferior to it (Fr. 43), then far forward (Fr. 50, 60 and 66). The tongue is briefly retruded (Fr. 80), then protruded upward and forward, again pushing tactor to the right (Fr. 85).

which were occasioned by her conspicuous disorder. A 12-month period of intensive speech therapy supplemented by biofeedback was associated with improved intelligibility of speech (see below).

Special Methods of the Study

Motor functions of the face, mouth and mesopharynx were recorded by color cinephotography on 16 mm film employing an Arriflex camera, with the aid of an apparatus which provided strong illumination closely in parallel with the line of camera. Pictorial study was facilitated by tracings or by 4 × 5 prints of selected cineframes. Cinema studies of speech were performed in a sound-treated room. Sound was recorded from a shotgun condenser microphone (Sennheiser Model MKH804) suspended in air or from a skin microphone (Sennheiser Model ROV503) held on the skin beside the larynx. Acoustical analysis was supplemented by extensive narrow band and wide band sound spectrography using a Kay Sonagraph (Model 661A). Observations of sensory and reflex elicitations in the oral area were recorded on video tape (Sony, Model 3000). Night sleep was observed in two sessions by videophotography and electroencephalography.

Laryngeal function was observed using an Olympus fiberoptic endoscope, with recordings made on 16 mm film at 16 or 32 frames per second; the fiberoptic conduit penetrated the nose, epipharynx and palatopharyngeal isthmus after local anesthetization.*

Oral, pharyngeal and laryngeal positions and motions were observed by radiography. Single radiographs were obtained at posed instants with subject "at ease", during severe dystonia (Fig. 1), during phonation and during maximal passive flexion or extension at head and neck. Cineradiographic studies employed a Westinghouse radiographic apparatus and an Arriflex 35 mm camera. Cine-

radiography in lateral projection of the pharynx area was performed on three occasions at 40 frames per second, after preliminary injection of dilute barium mixture into the nose, to outline the epipharynx, and into the mouth, for evaluation of pharyngeal function during swallow, suckle feeding and selected enunciations.

OBSERVATIONS OF THE STUDY

The subject's general level of *dystonia* increased during her activity day. It was evident in sleep and it accumulated slowly following waking. When she was awakened during our night sleep studies, her speech was fluent and clearly articulated. She noted, and we confirmed, that progressive relief of dystonia was a prologue of the achievement of sleep. A brief nap during daytime was followed by a substantial diminution in the dystonia.

The dystonia accumulated at times of voluntary functions involving the mouth and pharynx area, particularly including speech and feeding. To increase the dystonia, during our studies, we required continued conversational speech or other voluntary oral actions, such as the efforts of following a tactor by the tongue (see Fig. 2). In a notable discrepancy of its manifestations, the dystonia diminished progressively to absence or minimal demonstration during times of distraction of her attention from the oral area. During cinema studies, this was accomplished by mental arithmetic problems, with the answers written or signalled manually.

The 2-4 per second *tremor* of the tongue and mandible was correlated in its demonstration with the severity of dystonia. By inspection and palpation, the motions of the tremor were in the direction of the displacements of the dystonia; it seemed to be a wavering of the dystonic coordination. The tremor of the tongue and mandible was diminished when the tongue margin was in apposition with the lower lip (Fig. 2 and 3). It was arrested when the tongue was touched by an external object or a drop of fluid. It was also diminished during speech

articulation or other voluntary tongue gestures.

The anteriorward and lateralward *darting motions* of the tongue (Fig. 3, Seq. A, B and D), as studied by Thach and Weiffenbach (1973), were grossly correlated with the severity of dystonia. The lateralward darting motions resembled in pattern those elicitable in normal infants by object or fluid touch upon the tongue. But the responses might be multiple, e.g., rightward then leftward, crossing the midline in a single continuous displacement. In this person, the displacements occurred spontaneously and were also elicitable by touch by a Semmes Weinstein (1960) nylon filament or by 5 μ l of fluid delivered by a micropipette (Eppendorf Model 22-35-000-5). The lateral darting motions, whether spontaneous or touch elicited, were not perceived by the subject. The contours of the tongue tip during the lateralward darting motions differed from those in infants, with narrowing of the tongue tip and the appearance of a discrete shallow median groove (Fig. 3, Seq. B). Each of these distortions might occur when the tongue was already in dystonia (see Fig. 3, Seq. D). These responses, like the tremor, were absent during voluntary performances, such as the enunciation of single words, but appeared soon after completion of the word, on a schedule suggestive of a rebound phenomenon.

The dystonia was generally correlated in severity with the subject's reported tensions, confirmed by overt akathisia. She was well aware of these subjective tensions, describing herself as "jumping out of my skin." She also perceptively reported her subjective sense of relation to the dystonia. When her dystonia was the most severe, prior to discontinuance of the etiologic medication, and in the brief period of trial re-exposure to it, she felt totally, or integrally involved in the dystonia. Later, when this problem was less severe, she again felt herself to be an intact, whole person, interacting with the dystonia. At times she described herself as "fighting with this thing." Notably, there was no

*We acknowledge the cooperation of Drs. J. Smiddy and D. Credle, of the Pulmonary Function Laboratory of the National Naval Medical Center.

Sequence A

Sequence B

Sequence C

Sequence D

Sequence E

Figure 3. Spontaneous motions of the tongue. Drawings of selected cineframe sequences during a filming session in which the subject was in dystonia of moderate severity. Cinema is at 24 frames per second.

Sequence A. *Forward protrusion* of the tongue in darting pattern from a common reference position (Frame 1) in which the margin of the tongue is in general apposition with the lower lip and buccal wall. The tongue rapidly protrudes over the lower lip (Fr. 5 and 8). The body of the tongue is moderately grooved in the midline during this action. 11/24 seconds later the mouth is closed at lips.

Sequence B. *Lateralward displacement* of the tongue from position of protrusion (Frame 1). The tip is displaced leftward in a continuous motion (Frames 9 and 12); a narrow median groove becomes evident in the tongue tip during this action. The tongue returns abruptly to median position.

Sequence C. *Dystonia in pattern of median grooving* of body of tongue. Frames 1, 3 and 7 demonstrate progression of dystonia in this pattern. In Frames 1 and 3, the uvula is visible through the median groove. In Frame 3, the margin of the tongue is transiently rolled upward as a rim. In Frame 10, the tongue is partially relaxed, with its margin in apposition with the lower lip, but the blade is still slightly grooved in the midline and the mandible is still lowered.

Sequence D. *Lateralward displacement* of tongue tip during dystonia. Prior to this displacement (Frame 1), the medial portion of the tongue is drawn downward and the lateral portions are drawn upward and lateralward; this contour is appropriate to coincident dystonic activity of the genioglossus and styloglossus. The uvula, tensed and protruding forward from the body of the pharyngeal palate, is seen through the groove. The tongue tip is then protruded leftward (Frames 3 and 7); its tenseness is indicated by the narrow median groove in the tongue tip. The tongue returns to median position, with its margin in general apposition with the lower lip.

Sequence E. *Dystonia in pattern of tongue retraction.* From position of moderate retrusion (Frame 1), the tongue is progressively retracted into the ovoid contour shown in Frame 18, then partially relaxed to the contour in Frame 21.

associated irritability or confusion and no apparent disturbance of affect. To the contrary, she demonstrated a remarkable tolerance of stressful procedures, such as per-nasal fiberoptic laryngoscopy, or a succession of lumbar punctures during neurochemical studies.

Dystonia: Pattern

The dystonia was stable in its motor distribution and in the directions of its abnormal displacements (Fig. 1, 4 and 3, Seq. C and D). The dystonic contractions and the tremor were demonstrated most directly in the tongue and in the facial muscles (Fig. 4). The contractions in most of the muscles of the dystonic assortment were indicated by displacements, as of the mandible and hyoid, the pharyngeal palate and the cranium and cervical vertebrae (Fig. 1).

The variations were in the degree of effort, with corresponding extent of displacement, in the associated tremor and in the incidence of imposition of the darting motions

of the tongue tip and the adventitious motions of the uvula. Severe dystonia was variably associated with a tremor at 2-4 per second which was visible in the tongue and mandible; the tremor was shown in transversely oriented cineradiography as an anteroposterior "rocking" of the mandibular condyle within the temporal fossa. Mild dystonia was externally evidenced as tensing of the zygomatic and the orbicularis oris and oculi muscles, and opening of the mouth by lowering of the mandible. During dystonia of moderate or marked severity, the mandible was drawn strongly inferiorward, so that it assumed the position shown in Fig. 1. The condyles were displaced anteriorward in the temporal fossa, but were not advanced beyond the articular eminence; the mandible was not dislocated. The hyoid bone was drawn inferiorward less than the mandible, so that the mental portion of the mandible was closer to the body of the hyoid. In dystonia of moderate or marked severity, the cervical extensor muscles were also involved, so that the cranium

was extended upon the cervical vertebrae and the vertebrae were curved in lordosis.

At lesser degrees of dystonia, the tongue was forward in the mouth, with its margin in general approximation to the inner aspect of the lower lip and cheek (Fig. 2 and 3). In dystonia of moderate severity, the body of the tongue was grooved in the midline (Fig. 3, Seq. A and C) in a contour appropriate to coincident hypertonus of the genioglossus, drawing the midline of the tongue anteriorward and inferiorward, and of the styloglossus, drawing the lateral portion of the tongue posteriorward and superiorward; the apical margin of the tongue might be raised in a ridge, or curl, as in Figure 3. In severe dystonia, the tongue was retracted into an oval contour and the mass of tongue was retruded posteriorward, intermittently approximating the posterior wall of the mesopharynx (Fig. 1 and 3, Seq. E). During dystonia of moderate or marked severity, the pharyngeal palate was drawn upward in the contour indicated in Fig. 1. The uvula might be shortened and frequently protruded anteriorward in a variety of rapid midline gestures.

The larynx was visualized only in dystonia of marked severity, appropriate to the stressful procedure of fiberoptic endoscopy. During tidal respiration, the arytenoid masses were maintained, with slight tremor, anteriorward and medialward in relation to their expected position. During phonation, the arytenoid masses were displaced still further medialward and anteriorward, but the spatial relation of the vocal folds resembled that in normal phonatory coordination. The discrepantly anterior position of the arytenoid masses during tidal respiration and phonation was demonstrated also in transversely oriented single radiographs and cineradiography.

Interaction of Dystonia with Local Touch-Cued Gestures

The interaction of the dystonia with local, or contiguous touch-

cued gestures was demonstrated by asking the subject to follow a small metal tactor with her tongue tip. As shown in Fig. 2, her effort led rapidly to dyscoordination in which the tongue tip, in tensed contour, moved rapidly and ineffectually about the tactor. In contrast, the tongue's motions in reaching toward a touched site upon the upper or lower lip were dextrous in coordination.

Interaction of Dystonia with Feeding

Dystonia was increased by the oral manipulation of solid foods. The actions of prehension, biting, chewing, bolus gathering and initiation of swallow were all progressively impaired during feeding. During even mild dystonia, labial enclosure was inadequate, allowing labial spill. When the mouth was dystonically opened, biting, chewing and gathering of bolus by the tongue were severely impaired. Cineradiography showed delivery of bolus into the pharynx and initiation of swallow to be slow and laborious. As the mouth was not emptied of its content by this maneuver, labial spill continued after swallowing efforts were completed, and subsequent speech resulted in spray. The voluntary actions of oral feeding would generally increase the dystonia, and meals commonly terminated in motor failure, with increasing spill of food at the lips, premature spill of bolus into the pharynx, or an attempt to swallow an insufficiently prepared bolus.

In contrast with the impaired oral feeding maneuvers and delivery of bolus into the pharynx, the pharyngeal swallow proceeded in a normal peristaltic pattern, except for a relative anteriorward displacement of the hyoid and larynx and corresponding anteriorward displacement of the posterior pharyngeal wall (Fig. 5). The swallow effectively emptied the pharynx, except for that remaining in the valleculae, which were expanded by dystonic displacement of the tongue anteriorward and of the epiglottis inferiorward. No

Figure 4. Frontal views of face.

The upper drawing is of the subject at ease, with eyes open, mouth closed and head in horizontal orientation.

The lower drawing is of the subject in severe dystonia, with forehead corrugated, eyes closed, mandible lowered, tongue retracted, pharyngeal palate elevated, margins of lips everted and the skin of the submental and anterior cervical areas drawn tightly.

The middle schematic is of the muscles which are inferred to effect the dystonic displacements of the face as they are shown in frontal view. These include the frontalis, orbicularis oculi and oris, and zygomaticus.

bolus penetrated the larynx or the palatopharyngeal isthmus. Suckle was performed competently; during severe dystonia milk, or liquids of equivalent density, could be effectively suckled from a firm tube. When the suckle became rhythmic, a remarkable analogy to infant suckle feeding was demonstrated in cineradiography: a portion of the suckled ingesta penetrated the apposition of the tongue and palate, to accumulate in the valleculae. This is the second, or accessory, bolus accumulation area in infantile suckle feeding (Logan and Bosma, 1967). The accumulation of bolus remained in the valleculae until the swallow, which emptied the bolus from both the primary, or principal, accumulation area, adjacent to the junction of the oral and pharyngeal palate, and this accessory area. In further analogy to infantile suckle feeding, the lips approximate a small object by motions which are only vertical, without the mature pattern of pursing by transverse contraction and corrugation which effectively close them about a small object; this less complete labial closure resulted in leakage about the sucking tube. During tasting, the dystonically-opened mouth would be closed and the suckle-like actions of tasting were performed in normal fashion.

Interactions of Dystonia with Speech

This person was consistently competent in all categories of language performance including vocabulary and syntax. Her perception of her own speech production and her self-awareness of oral articulatory problems was valid.

At times of relief of dystonia, as on waking from sleep, the cortical organization, or programming, and the motor coordination of speech were entirely competent. But when the dystonia was mild or moderate, propositional speech readily accentuated it. In a speaking sequence during moderate dystonia, the impairment of speech was variable, so that within a brief period of speech each of the

common articulations might be produced correctly. During severe dystonia, speech was not possible.

The dystonia interacted with details of the speech motions, delaying and distorting phonation, distorting the oral articulations and also secondarily impairing oral articulation by the forced opening of the mouth and separations of the oral articulators.

The interaction of speech and respiration varied with the severity of dystonia and also with the person's effort of speech. During relief of the dystonia, she was able to count rapidly from 1-20 in a single breath. During severe dystonia, speech would impede expiration, so that a phrase would be followed by conspicuous completion of expiration. During lesser dystonia the delay in word achievement and the prolongation of words resulted in prolongation of the breath unit. She might employ supplementary breath for speaking, with corresponding treble distortion and diminution in the volume of phonation. Phonation was at times extended into the succeeding inspiration. The relation of speech and respiration were particularly disturbed if respiration was coincidentally adapting to general exertion, such as rapid walking.

In light of a position distortion of the arytenoid masses which was demonstrated by cinendoscopy, phonation was of remarkably good acoustical quality. Phonation was usually in this person's central range of pitch. Tone was usually stable and firm, without wavering. Speech abnormalities were of schedule, in relation to the CVC words which we employed in the observation routine. The vowels were commonly prolonged. More remarkably, a phonation frequently preceded the initial consonant (Fig. 6). This phonation might follow a silent but visible struggle in dystonia lasting as long as 20 seconds. The preceding phonation typically was of abrupt onset and initially in vowel pattern. At its intersection with the CVC utterance the vowel pattern of the preceding phonation was modulated toward the contour appropriate to

the initiation of the pending consonant (see Fig. 6A and B).

The sound of the preceding phonation might be interrupted as in Fig. 6A, or the primary (F₀) frequency might continue (Fig. 6B). In either of these instances, the formants at the conclusion of the preceding phonation were reciprocal in contour to those of the pending consonant. Figure 6C illustrates a lesser modulation of the preceding phonation and little or no diminution in its sound volume at the intersection with the consonant. In Fig. 6B and D, the preceding phonation is not visibly modulated in formant pattern or diminished in sound volume; it continues into the pending utterance and distorts its initial consonant. The preceding phonation was not repetitive, as in stuttering.

An accessory phonation might also follow the CVC utterance. It might resemble in acoustical pattern the latter portion of the utterance and might be demarcated from the adjacent consonant by an instant of silence. Alternatively, the utterance might be followed by an expiratory sigh or, rarely, by single or series of clicks.

We found no acoustical indications of abnormal contour or articulation in the upper pharynx. This correlated well with the cineradiographic demonstration of adequate palate elevation and palatopharyngeal isthmus closure. The speech articulatory gestures of the tongue were generally reciprocal to either of the two common dystonic contours of the tongue; whether the tongue was grooved in a trough-like contour or was retracted in oval contour. Accordingly, each speech articulation gesture, in whatever its degree of accomplishment, was achieved in a context of anatomical arrangement which was alternative to its target contours and approximations. The lingual speech gestures demonstrated considerable adaptation; compensatory approximations most commonly included protrusion of the tongue, as during the articulations of "b" and "d".

The articulatory gestures of the lips were severely impaired during

dystonia. The labial approximations of speech, like those during feeding, were obviously impaired by the dystonic opening of the mouth, occasioning secondary errors of approximation and placement of the oral articulators. The lips struggled to adapt to this circumstance by approximations to the incisors of the maxillary denture.

The dystonia was accentuated by the performance of propositional speech. It could be diminished by coincident writing of the same message. She employed this technique of speech facilitation when speech communication was necessary in spite of current severe dystonia. Dystonic distortion of speech was also diminished when she was reading aloud, or if the speech content was a rote material, such as remembered poetry, or when singing familiar songs. Speech competence was also increased during speech interspersed with laughter and during interjections, as of standard phrases expressing frustration.

Trial of Speech Therapy

The clinical evaluation of speech included 12 months of intensive speech therapy. This was initiated four months after completion of neurochemical studies and pharmacologic therapy trials. In the first six months, weekly sessions were focused upon improvement of articulation, monitoring of speech performance, and voluntary control of dystonia. During the second six months this therapy was supplemented by sensory feedback, reflecting the EMG potentials from the cervical extensor or the frontal muscles after the manner of Frankel (1976). During the first six months of therapy there was noticeable improvement in articulation and in the ability to "pace" speech between the surges of dystonia that the speech occasioned. Little voluntary amelioration of the dystonia was achieved. During the second six months, the subject was able to reduce levels of muscle tension during speech. This control was not generalized outside the

feedback condition and it was not reflected in continued improvement in articulation. Monthly speech probes indicated that consonant articulation errors decreased from 27 to a minimum of 8 per cent, late in the first six months, and later stabilized at 16 per cent.

Speech probes obtained at increasing intervals during the four years following intensive speech therapy demonstrated that this person could maintain control over speech when her awareness levels were high, but overall articulatory competence did not continue at the levels attained during the intensive therapy period. Considerable impairment of speech expression has continued to be a major handicap.

DISCUSSION

This person's motor abnormalities, of dystonia mixed with

tremor, are similar to those described in the tardive dyskinesia following phenothiazine medication in adults (Faurbye et al, 1964; Evens, 1965; Wertheimer, 1965; Bergouignan et al, 1966; Schmidt and Jarcho, 1966; Crane, 1968 and 1973; Crane and Gardner, 1968; Crane and Naranjo, 1971; Chase, 1972; Am. Coll. Neuropsychopharm., 1974). She had little of the minor licking and lip grimacing described as common in phenothiazine-induced dyskinesia, and none of their choreiform motions. The occasion and pattern of her dyskinesia closely resembles three examples of adult onset of "focal dystonia" described by Marsden (1976) in whom blepharospasm and oromandibular dystonia were sequentially manifested. Like those liable to phenothiazine-related dyskinesia, this person is female and advanced in age. In a further resemblance, her dyskinesia has continued in consistent pattern and degree after its changes in the

Figure 5. Pharyngeal swallow during severe dystonia, shown by cineradiography of the meso- and hypopharynx in transverse orientation at 40 frames per second.

The epiglottis is labelled *Epig.*, hyoid is *Hy.*, mandible is *Mand.*, arytenoid mass is *Aryt.*, pharyngeal palate is *Pal.*, and vertebrae C3 and C4.

Frame 1. Prior to swallow effort. The dystonia is indicated by low position of the mandible and retraction of the tongue body superiorward and posteriorward. The valleculae, between the epiglottis and tongue, are filled with barium retained after a previous swallow.

Frame 13. Initiation of swallow. The mandible is elevated toward its normal resting position. The tongue, hyoid and epiglottis are moved anteriorward; the inferior margin of the piriform recess is elevated.

Frame 34. Bolus enters the mesopharynx and begins to spill over the epiglottis. The tongue and hyoid are displaced further anteriorward; the body of the hyoid is out of the frame. The hypopharynx is further elevated.

Frame 37. The epiglottis is moved posteriorward and more barium spills over it. The hypopharynx is further elevated. The laryngeal vestibule is nearly closed by anterior and superiorward displacement of the arytenoid masses.

Frame 39. The body of the tongue is moved posteriorward. The epiglottis is displaced inferiorward; its distal portion approximates the posterior wall. The pharyngo-esophageal segment, at the level of C5, and the cervical esophagus are open.

Frame 44. The tongue is moved further posteriorward and the adjacent posterior wall of the pharynx anteriorward, in the peristaltic wave. The epiglottis is tilted into the hypopharynx and the bolus is in continuity from the mesopharynx into the esophagus; the pharyngo-esophageal segment is normally patent.

Frame 50. The length of the posterior pharyngeal wall is moved forward. The pharyngo-esophageal segment is closed and the peristaltic wave is moving down the esophagus.

Frame 54. The tongue and the posterior pharyngeal wall are returning to the positions in Frame 13. The epiglottis is rising. The laryngeal vestibule is again patent.

Frame 71. Dystonia is resumed. The positions of the mandible, hyoid, tongue, epiglottis and floor of the pharynx are essentially as in Frame 1.

Figure 6.

Sound spectrograms demonstrating interaction of a dystonic preceding utterance with the word "weep". The examples are arranged in a sequence which demonstrates increasing intrusion of the preceding utterance into the word and, accordingly, increasing severity of distortion of the word. See text for further comment.

At the conclusion of the word, the dystonic closure is resumed, but the silence is interrupted by clicks throughout the frequency range.

weeks immediately following withdrawal of the causative medication. We infer, from these similarities, that the neurophysiologic abnormality and anatomic lesion are those of the monamine-containing neuronal systems in the striatal complex (Chase, 1972). Our further specifications of the occasion of the dystonia and of the darting tongue motions, and the specific interactions of these dyskinetic elements with voluntary performances may well be pertinent to phenothiazine-induced dyskinesia as well.

The performances which are differentially distorted by the dystonia are distinguished not only by their voluntary control and their post-infantile calendar of developmental achievement, but also by the role of their pertinent sensory mechanisms. Dystonia of the tongue was increased by contiguous tongue tip stimulation. This is not unexpected, for the sensations generated by voluntary actions are integral and determinant elements of those actions. We extend the definition of volition to include local attention and awareness. In the circumstance of this dystonic person, the motor performances of the tongue are at the mercy of the tongue's sensibilities.

The basic element of this person's dystonia is that of a variably effective mechanism of distortion and interruption interposed upon the corticofugal pathways between the cerebral cortex, which is essentially competent in organization and direction, and upon the essentially competent bulbar sensorimotor mechanisms. The pattern of mouth opening, tongue retrusion, palate elevation and glottic constriction which comprises the dystonia is interpretable as a distortion of the motor background or context of the discriminate details of the voluntary performances of this motor region, particularly the performances of speech. These distortions are motor phenomena which are inhibited incident to the normal coordinations. This subject's motor effort is, to this degree, a failure of inhibition of extrapyramidal elements of cortically governed functions.

Interactions of Dystonia with Modes of Speech

Propositional speech accentuates the dystonia and is, in turn, distorted by it. The dystonia was less occasioned by the subject's speech in other modes, such as interjections of glib or emotionally laden comments, recitation of familiar poetry or singing. Expressive speech in these non-propositional modes is classified as "automatic" by Alajouanine (1956) and Van Lancker (1973), or as "non-intellectual" by Espir and Rose (1970), and is differentially competent in aphasia (Jackson, 1887 and 1889; Alajouanine, 1956; Critchley, 1970; Espir and Rose, 1970; Luria, 1970; Van Lancker, 1973 and 1975; and others). A variety of experimental and clinical evidences indicates that these non-propositional modes of speech are represented in both the non-dominant and dominant cerebral hemisphere (for review see Van Lancker, 1973). An analogous differentiation is described in the aphasics who speak better when reading (Critchley, 1970). In the perspective of the insights derived from the studies of aphasics, the differential impairment of propositional speech in the person of this report affords additional indication that propositional and automatic speech are differently represented in the cerebrum. We would also add that her automatic speech is governed by corticofugal pathways that bypass the dystonia mechanisms.

The corticofugal entity of speech performance includes the gestures of preparation for the intended utterance and also the gestures of return toward open tidal respiration. The gestures of preparation include a "pre-phonatory tuning or pre-setting" of the larynx (Wyke, 1971; Wyke and Kirchner, 1974). The dystonia of this person particularly distorts these gestures of preparation and, incident to this distortion, demonstrates abnormalities of both the activation and the inhibition components of the preparation. The dystonia exaggerates the activity which precedes the intended utterance; the exaggeration initially effects glottal closure, which is abruptly released

as an incongruous vowel sound (Fig. 6A-D). The explanation for the phonation preceding the intended CVC utterance may be quite simple. We propose that the dystonia, accentuated in preparation for speech performance, occludes the larynx during expiratory effort. As the dystonic closure of the larynx is slightly released, phonation is abruptly initiated. The phonation is in a vowel pattern appropriate to the current dystonic contours of the pharynx and mouth. Further inhibition of the dystonia permits modulation of supraglottal contours in preparation for the consonant of the intended CVC utterances and also effects diminution in sound volume. Correspondingly, following the CVC utterance, the vocal folds are dystonically approximated at the time that they would normally be diverging toward their respiration-relevant position; thus a post-utterance phonation or noisy expiration may occur.

These speech distortions in association with dystonia are in some analogy to the tense distortions observed in certain patterns of stuttering. We refer particularly to the form of stuttering, or stammering, in which there is a distortion of voluntary "pre-setting" (Wyke, 1971 and 1974). This distortion has been termed the "anticipatory struggle reaction" (Bloodstein, 1958). This form of stuttering is found in persons who are otherwise normal and also in those who have a lesion in the parietal area of the dominant cerebral cortex (Stremel, 1963; Lenneberg, 1962; Jones, 1966; Rosenbek et al, 1978). The analogies are several. In stuttering, as in the person of this report, the distortion occurs particularly in propositional speech; speech in non-propositional modes is spared (Wyke, 1971, and others).

This person's neuropathy is inferred to be suprabulbar in site. We found no indications of interruption, ablation or abnormal activation of the intrinsic bulbar sensory, motor or coordinative mechanisms. Transverse darting motions of the tongue are uniformly elicitable in infants and have been elicited from mature persons who

have oral sensory losses. They are rarely, if ever, seen in normal neurologically mature persons (Weiffenbach, 1972; Thach, 1973). In the person of this report, the darting motions were elicitable by touch and also occurred spontaneously. Spontaneous darting motions were absent during speech utterances but were frequently seen immediately following the single enunciations employed in our cinema demonstration routines. The darting gestures, particularly the lateralward ones, recurred after the cessation of the speech effort and continued to recur in irregular schedule, interspersed with the abrupt forward thrusts. This schedule of their appearance is interpretable as a phenomenon of release of intrinsic bulbar reflexive mechanisms from suprabulbar control. The appearance of the reflex at a characteristic interval following the speech effort implies a release from the inhibition, essentially an afterdischarge in an otherwise intact bulbar system.

SUMMARY

Observations of tardive dystonia and related distortions of position, feeding and speech in a woman age 67 to 71 years, employing cine- and videophotography, cineradiography and sound recording. The dystonia was manifested in the face, jaw, tongue, palate, larynx and cervical extensor muscles. It was increased by stress and by propositional speech or mature patterns of oral feeding; suckle and swallow were spared. Its interactions with propositional speech afforded useful indications of its severity. These interactions included distortion of the speech preparatory gestures, of phonation and articulation, and of respiration.

These elements are accessible to clinical description and, thus, afford criteria for detection and graduations of severity of tardive dystonia during treatment with medications liable to cause this form of dyskinesia, for study of its mechanisms and also for evaluation of medications employed in trial of its therapy.

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