



Dystonic respiratory dysregulation and other breathing disorders associated with dystonia.

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ABSTRACT

Introduction: Respiratory difficulties are sometimes reported by patients with cranial-cervical dystonia (Mehanna R, Jankovic J. Respiratory problems in neurologic movement disorders. Parkinsonism Relat Disord. 2010;16:628-38). This dystonia-related respiratory disorder, however, has not been well characterized.

Methods: We review 13 dystonic patients with various respiratory complaints and describe their symptoms and response to botulinum toxin injections.

Results: The male to female ratio was 4/9 and the mean age 56.84. Cranial-cervical dystonia was the most frequent diagnosis affecting 9 (69%) patients. The cause of dyspnea was an upper airway dystonic obstruction in 9 (69%) patients, diaphragmatic dysfunction in one (8%) and a combination of both in the remaining 3 (23%) patients. Emergent tracheostomy was required in 2 (15%) patients. Among the 5 patients who underwent laryngoscopy, one had an adductor spasmodic dysphonia and 4 had normal vocal cord motion. Pulmonary function tests were performed in 5 patients and were normal. Videofluoroscopy was performed in 4 patients, showed decreased diaphragmatic movement in 2 and was normal in the 2 others. Among the 8 patient who received botulinum toxin injections and were not lost to follow up, 6 (75%) reported improvement in their respiratory distress and 2 (25%) reported no improvement.

Conclusion: In some patients with dystonia, gasping, stridor, interrupted flow of speech, paradoxical breathing, dyspnea on exertion, and other respiratory symptoms suggest involvement of the upper airways, diaphragm, or both as a result of "dystonic respiratory dysregulation". Botulinum toxin injections may be at least partially beneficial in some patients. This series of patients draws attention to respiratory distress as a potentially serious, even life-threatening, complication of dystonia.

BACKGROUND

- Dystonia may cause respiratory problems when it involves the upper airways or the diaphragm.
- Except for the well-recognized respiratory problems sometimes associated with laryngeal dystonia (spasmodic dysphonia), there is limited information regarding breathing abnormalities in patients with other forms of dystonia.
- We have reviewed 13 dystonic patients with various respiratory complaints and attempted to characterize their symptoms and respiratory response to treatment including botulinum toxin (BoNT) injections.

RESULTS

- The clinical features are summarized in **Table 1**.
- The **video** of patients 1 to 3 have been included as illustrative cases and can be viewed on the portable computer next to the poster.

DISCUSSION

- Typically, in patients with cranial-cervical dystonia, the involuntary contractions of the posterior pharyngeal muscles and upper respiratory airways are associated with a paradoxical contraction of the vocalis muscles, particularly during inspiration, and the vocal cord adduction causes stridor or gasping.

Table 1
Demographic and Clinical Data on Patients with Dystonia-associated Respiratory Symptoms.

Pt No	Sex	Age at onset	Dystonic symptoms	Diag	Sensory trick for dyspnea	UAD	CDD
1	F	57	Bleph, facial grimacing, mixed jaw-opening and jaw-closing, dysphagia, platysma contraction.	CCD	Massaging the pectoralis	+	+
2	M	62	Hoarse and strained voice interrupting speech.	SD	None reported	+	
3	F	58	Bleph, bruxism, platysma contraction, pharyngeal dystonia and SD.	CCD	Neck extension or touching both cheeks	+	
4	F	54	Bleph, paranasal contractions, jaw opening, left torticollis and laterocollis, dysphagia, hoarse voice, humming.	CCD	Singing	+	+
5	F	64	Bleph, masseter, submental complex contraction, platysma contraction, right torticollis, anterocollis, dysphagia, dysarthria.	CCD	None reported	+	
6	F	55	Bleph, bruxism, submental complex contraction, retrocollis.	CCD	None reported	+	+
7	F	74	Dyspnea only during speech. Abdominal contractions during speech associated with the dyspnea.	RD	Pressure below ribs, laying down, pressure on walker with arms		+
8	M	49	Right hand's writer's cramp, strained voice with voiceless pauses.	WC and SD	None reported	+	
9	F	37	Bleph, anterocollis.	CCD	None reported	+	
10	M	60	Bleph, paranasal contraction, lip pursing, jaw opening, submental, anterocollis, right laterocollis, left torticollis.	CCD	None reported	+	
11	F	46	Platysma and post neck muscles contraction, head pulling forward with chin pulling up, left laterocollis, strained voice.	CCD	None reported	+	
12	M	72	Bleph, dystonic head tremor.	CCD	Touching forehead or singing	+	
13	F	51	Orolingual stereotypies, moderate retrocollis, tongue and jaw withdrawal.	T dyst	None reported	+	

Legend: add: adductor, Bleph: blepharospasm, CCD: Cranio-cervical dystonia, CDD: Chest or diaphragmatic dysfunction, Pt: patient, RD: respiratory dysphonia, Resp: respiratory, SD: spasmodic dysphonia, T dyst: Tardive dystonia, UAD: upper airway dysfunction, WC: writer's cramp.

- Patients with dystonia may exhibit contractions of the respiratory muscles (including chest and diaphragm).
- As a result of excessive and desynchronized contractions of the upper airways, chest or diaphragm muscles, the patient experiences dyspnea at rest and especially with exertion.
- The term "dystonic respiratory dysregulation" includes all the various respiratory complaints associated with dystonia.
- Table 2** summarizes the literature on respiratory problems associated with dystonia

CONCLUSIONS

- Respiratory complaints are described in dystonic patients, especially in those with cranial-cervical dystonia and presumably arise from involvement of the upper airways, chest and diaphragm.
- "Dystonic respiratory dysregulation" is proposed to include gasping, stridor, interrupted speech, paradoxical breathing, dyspnea at rest and on exertion, and other nonspecific difficulties with breathing in patients with dystonia.

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Table 2:
Literature reported cases of dystonia-associated respiratory problems.

First author/year	Number of cases	Diagnoses	Respiratory complaint	Diagnostic tests	Response to treatment
Laqueny et al, 1995	3	Cranial and cervical dystonia	Loud breathing, breathing arrest or deep inspirations braking the speech	EMG	N/A
Braun et al., 1995	26	4 with focal dystonia, 22 with generalized or multifocal dystonia		Video fluoroscopy, pulmonary function test	N/A
Papapetropoulos et al., 2007	1	Laryngeal dystonia that evolved into an oromandibular dystonia			Good response to BoNT
Fève et al., 1993	1	Tardive laryngeal dyskinesia		Laryngoscopy	Good response to BoNT
Fève et al., 1993	10	Tardive laryngeal dyskinesia		Laryngeal endoscopy. Percutaneous EMG of the thyroarytenoid muscles. 2 had VF.	1 patient was treated with BoNT with good response. He was then treated with TBZ with moderate improvement.
Christodoulou and Kalaitzi, 2005	2	Anti-psychotic induced acute laryngeal dystonia		Unsure	Unsure
Tsai et al., 2008	1	Tardive laryngeal dystonia		None	Good response to change in neuroleptics
Zwirner et al., 1997	3	Spasmodic laryngeal dyspnea		Laryngoscopy	Good response to BoNT
Blitzer and Brin, 1991	200	Laryngeal dystonia, all types		Laryngoscopy	Good response to BoNT
Blitzer et al., 1988	110	Laryngeal dystonia, all types.		Laryngoscopy	Good response to BoNT in the 34 injected patients.
Marion et al., 1992	6	Laryngeal dystonia	Stridor	Laryngoscopy and EMG	Good response to BoNT
Vetrugno et al., 2007	3	MSA and sleep related laryngeal dystonia	Nocturnal stridor	Laryngoscopy and EMG	N/A
Lew et al., 1994	1	Lubag syndrome (X-linked dystonia-parkinsonism) with adductor laryngeal breathing dystonia	Stridor	Laryngoscopy	Worsening aspiration with BoNT.
Grill, 2004	1	Striatonigral degeneration with cranial dystonia	Stopped breathing and "turned blue"		Good response to BoNT
Kato et al., 2007	1	PD with cranial dystonia	Hypoxic spells		N/A
Grillone et al., 1994	7	Adductor laryngeal breathing dystonia	Stridor	Laryngoscopy	Good response to BoNT
Kirton and Riopelle, 2001	1	Cranio-cervical dystonia secondary to cerebral anoxic injury.	Intermittent respiratory distress		Partial response to TBZ
Hamzei et al., 2003	1	MSA with cranial and laryngeal dystonia	Stridor	Laryngoscopy	Good response to BoNT

Legend: BoNT: Botulinum toxin, EMG: electromyography, MSA: multiple system atrophy, N/A: not applicable, PD: Parkinson's disease, TBZ: tetrabenazine.