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 reviewed 13 dystonic patients with various respiratory complaints and described their symptoms and response to botulinum toxin injections.

Results: The male to female ratio was 4:9 and the mean age 58.4 years. 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, dysphagia, pharyngeal dystonia, and other respiratory disorders associated with dystonia.

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 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.

Inflammatory symptoms were not considered in 1 patient who underwent botulinum toxin injection and were not lost to follow up. 67% (7) had improvement in their respiratory distress and 2 (25%) reported no improvement.

Conclusions: 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 dysfunction 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.
- 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 has been included as illustrative cases and can be viewed on the portable computer next to the poster.

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.

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

<table>
<thead>
<tr>
<th>Pt No</th>
<th>Sex</th>
<th>Age at onset</th>
<th>Cranial-cervical dystonia</th>
<th>Diaphragm</th>
<th>Sensory trick for dyspnea</th>
<th>DAD</th>
<th>CDD</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>37</td>
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<td>2</td>
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<tr>
<td>3</td>
<td>M</td>
<td>55</td>
<td></td>
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</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>First author</th>
<th>Year</th>
<th>Number of cases</th>
<th>Diagnosis</th>
<th>Respiratory complaint</th>
<th>Diagnostic tests</th>
<th>Response to treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Szczepanski</td>
<td>2000</td>
<td></td>
<td>Laryngeal dystonia</td>
<td>Good response to</td>
<td>N/A</td>
<td></td>
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<tr>
<td>Braun et al.</td>
<td>2001</td>
<td>5</td>
<td>Laryngeal dystonia</td>
<td>Good response to</td>
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<tr>
<td>Eisenberg et al.</td>
<td>2001</td>
<td>4</td>
<td>Laryngeal dystonia</td>
<td>Good response to</td>
<td>N/A</td>
<td></td>
</tr>
</tbody>
</table>

REFERENCES

- Blitzer et al. 1994
- Braun et al. 2001
- Eisenberg et al. 2001
- Szczepanski et al. 2000

CONCLUSIONS

- 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.