Spasmodic Torticollis: A Case Report and Review of Therapies

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Background: Spasmodic torticollis is a movement disorder of the nuchal muscles, characterized by tremor or by tonic posturing of the head in a rotated, twisted, or abnormally flexed or extended position or some combination of these positions. The abnormal posturing of the head allows this disorder to be clinically diagnosed. Psychiatric symptoms frequently accompany or precede the diagnosis of the movement disorder.

Methods: Using the key words "torticollis," "spasmodic torticollis," "therapy," "behavior therapy," "botulinum toxin," MEDLINE was searched from 1989 to 1996 for information on the cause and treatment of spasmodic torticollis.

Results and Conclusions: Therapies include behavior modification, such as biofeedback, hypnosis, or simply training the patient to consciously readjust the position of the head; pharmacotherapy, using a variety of agents, the most commonly prescribed being anticholinergic medications or the botulinum toxin type A; and surgery, which entails selectively denervating the muscles responsible for the abnormal movement or posture of the head. The most effective treatments include surgery and botulinum, with sustained success rates ranging from approximately 60 to 90 percent. (J Am Board Fam Pract 1996;9:435-41.)

Case Report

A 37-year-old woman, well-known to our office for her history of anxiety and phobias, came to the office complaining that her head "wanted to go back." Her symptoms began approximately 6 months earlier with painless "pulsing" in her neck that became worse with stressful situations and physical activity. The symptoms were relieved by relaxation and sleep. She could briefly stop the pulsing by placing her hand on the right posterior aspect of the neck. Her symptoms had progressed to an extension of the neck with spasm, which caused her to lean forward to maintain eye contact with others. She also noted an occasional "eye tic," which seemed to come and go spontaneously. She denied any paresthesias, weakness, dysphasia, visual changes or hearing loss, or bowel or bladder changes. Although she had no family history of specific neurologic problems, the patient reported a maternal aunt who had "facial tics." The patient had a medical history notable for anxiety and several phobias for which she had received psychological counseling; at the time of her examination, she had mild to moderate depressive symptoms.

On physical examination her neck was posteriorly extended, her head was tipped slightly to the left and anteropulsed, and there was a palpable...
spasm and hypertrophy of the left cervical para-
vertebral musculature. She had full range of mo-
tion of the neck in all planes with intact motor
strength, and there were no other motor or sen-
sory deficits. Cerebellar function and reflexes
were normal bilaterally; no other tremor, tic, or
dystonia was observed. Laboratory test results,
including muscle enzymes and serum ceruloplas-
min, were normal.

A diagnosis of spasmodic torticollis was made,
and the patient was prescribed nortriptyline
(Pamelor) for its anticholinergic effect, as well as
for her depression. Nortriptyline reduced but did
not ablate her symptoms after a 2-month trial at
100 to 150 mg each day.

The patient was prescribed in succession bac-
lofen, haloperidol, and clonazepam. She had only
mild improvement on these medications, how-
ever, so she resumed nortriptyline until her con-
tion could be evaluated for botulinum therapy
through a subspecialty clinic.

The patient's first botulinum toxin injection
produced dramatic improvement; although she
had some neck weakness, she preferred that to the
spasm. She continued to receive regular botulinum
toxin injections with ongoing suppression of symp-
toms; at a 2-year follow-up examination, the pa-
tient remained symptom-free until just before her
quarterly botulinum toxin injection was due.

Pathophysiology
Spasmodic torticollis is marked by involuntary
hyperkinesis of neck musculature resulting in ab-
normal head postures or sustained movements of
the head. Idiopathic spasmodic torticollis usually
has an insidious onset that begins in the fourth or
fifth decades of life with no strong sex predomi-
nance.1,2 (Secondary spasm is a self-limited disor-
der and will not be further discussed.)

Few studies address the natural history of spas-
modic torticollis. Physicians who provide care for
their patients for 7 or more years note deteriora-
dation during the first 5 years, a plateau for 5 years,
then slight improvement.1 Spontaneous remis-
sions (partial or complete) have been reported in
up to 60 percent of patients in some series;1 oth-
ers note full remission in 16 percent, with sus-
tained remission for 12 months of 6 to 12 per-
cent.2,3 (Differing definitions of remission and
duration of remission make interpretation of data
difficult.)

The position of the head is described as rotated
(torticollis), extended (retrocollis), flexed (antero-
collis), or inclined to the side (laterocollis). A
combination of muscles is often involved, result-
ing in latero-torticollis or retro-torticollis, and so
forth. Pure retrocollis (6 percent of cases) and
pure anterocollis (3 percent) represent symmetri-
cal involvement of muscles: most cases are asym-
metrical, and the involved hypertrophied muscles
can readily be palpated and contrasted to the con-
tralateral normal musculature. The sternocleido-
mastoid muscle is involved in 75 percent of cases
and the trapezius in 50 percent; other muscles
might include rectus capitis, obliquus inferior, and
splenius capitis.4 Other muscles can become in-
volved: in some cases the spasm generalizes to the
muscles of the shoulder, girdle, trunk, or limbs.5
One author reported up to 13 percent of patients
as having extranuchal dystonia1,4; Jahanshahi et
al5 reviewed several studies of patients with adult-
onset torticollis, and found that extranuchal in-
volveinent ranged from 0 to 80 percent. Others
noted subclinical evidence of a bilateral distur-
bance, most likely within the basal ganglia.6,7

Neck movements can vary from jerky to
smooth.1,5 The patient might have antecedent
symptoms including tremor, writer's cramp, and
blepharospasm.2,4 The torticollis symptoms are
aggravated by standing, walking, or stressful situa-
tions and usually do not occur with sleep. Pain in
the neck and shoulders can accompany spasmodic
torticollis, but it is unusual as a presenting symp-
tom.4 Pain can develop later, however, as the re-
sult of degenerative joint disease of the cervical
spine or as a result of muscle spasm. Patients will
often observe that they can reduce or eliminate
the spasms by a physical stimulus (the geste an-
toniste), such as placing their hands or pillow on
the back of the neck or chin,4,5 as our patient did.

There is no information in the literature on
late or end-stage disease: a MEDLINE search from
1989 to 1996 found no references using key
words “late,” “end-stage,” or “unremitting.”

Antecedents to the Disease
Antecedents to spasmodic torticollis consist of a
variety of psychologic and biologic factors (Table
1). The link between spasmodic torticollis and
psychologic factors appears to be strong. This as-
sociation was evident in the patient we describe.
She had several long-standing phobias (including
tight clothing, bridges, and basements). In addition, she had mixed anxiety and depression.

Rondot et al. noted frequent psychological antecedents in 58 percent of their patients, including schizophrenia and other psychoses, bipolar disorder, major depression, psychiatric hospitalization for any reason, anxiety disorders (very common), cyclothymia, dysthymia, somatoform disorder, and personality disorders.

The controversy about this alleged link between spasmodic torticollis and psychological factors is considerable; some authors find most (up to 80 percent) of their patients have abnormal personalities, whereas others find no major differences.

One group of authors found spasmodic torticollis patients had higher responses on the Yale-Brown Obsessive-Compulsive Scale, and their Beck Depression Inventory scores were also higher than controls. Jahanshahi found 24 percent of patients to be mildly to severely depressed. Duane likewise found the prevalence of depression to be greater than average. Depression scores can improve as symptoms improve, suggesting that depression might actually be a secondary problem related to body image and degree of disability.

Biological antecedents are often found: 5 to 15 percent of patients have a positive family history of a movement disorder. A small percentage of patients have a history of serious head and neck trauma or a long history of neuroleptic drugs, but in most cases the spasmodic torticollis is idiopathic.

Rondot et al. found that 61 percent of patients suggested a discrete event associated with the onset of spasmodic torticollis. In order of frequency, these events included emotional stress, medical problems, vocational upsets, head trauma, a neuroleptic prescription, or a febrile infection.

### Proposed Mechanisms

The pathophysiology of spasmodic torticollis remains unknown across more than 20 years of literature reviewed. There is no central nervous system lesion identifiable with imaging or at autopsy; there are no biochemical markers. Postulated causes range from purely functional to purely organic; others offer slightly more detailed mechanisms, such as "perturbation of the extrapyramidal system," a basal ganglia-related disorder, or a form fruste of idiopathic torsion dystonia. Idiopathic torsion dystonia is a more generalized dystonia with truncal involvement. The location of the human gene for idiopathic torsion dystonia might help to clarify questions about etiology, but most current research focuses on therapy.

### Therapy

Many therapeutic modalities have been explored: psychologic, surgical, and pharmacologic (Table 2). As is typical of any disease of unclear etiology, therapeutic choices span the available spectrum and often have conflictual therapeutic mechanisms.

#### Behavioral Therapy

Behavioral therapies are discussed in older medical literature, usually with few patients, but often with notable successes (Table 3). The current literature has no entries under behavior or psychotherapy in the past 7 years.

### Table 2. Treatment Options for Spasmodic Torticollis.

<table>
<thead>
<tr>
<th>Type</th>
<th>Component</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behavioral</td>
<td>Positive practice</td>
</tr>
<tr>
<td>Pharmacotherapy</td>
<td>Levodopa</td>
</tr>
<tr>
<td>Surgery</td>
<td>Selective peripheral denervation</td>
</tr>
<tr>
<td>Surgical</td>
<td></td>
</tr>
</tbody>
</table>

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Table 1. Antecedents to Spasmodic Torticollis.

<table>
<thead>
<tr>
<th>Type</th>
<th>Characteristic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Psychologic</td>
<td>Psychosis</td>
</tr>
<tr>
<td></td>
<td>Affective disorder (unipolar and bipolar)</td>
</tr>
<tr>
<td></td>
<td>Psychiatric hospitalization for any reason</td>
</tr>
<tr>
<td></td>
<td>Anxiety</td>
</tr>
<tr>
<td></td>
<td>Cyclothymia</td>
</tr>
<tr>
<td></td>
<td>Dysthymia</td>
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<tr>
<td></td>
<td>Somatoform disorder</td>
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<tr>
<td></td>
<td>Personality disorder</td>
</tr>
<tr>
<td>Biologic</td>
<td>Family history of movement disorders</td>
</tr>
<tr>
<td></td>
<td>Head and neck trauma</td>
</tr>
<tr>
<td></td>
<td>Neuroleptic drug use</td>
</tr>
</tbody>
</table>
Spencer et al describe a single-subject study using behavioral therapies that consisted of progressive relaxation, positive practice, and visual feedback. Their patient had significant improvements in all areas, which were maintained at a 2-year follow-up examination.

Agras and Marshall used massed negative practice (ie, repeating the spasmodic positioning), 200 to 400 repetitions of the movement daily, which achieved full resolution of symptoms in 1 of 2 patients. Results persisted for 22 months.

Another single-case study used positive practice (exercising against the spasming muscle groups) in a bedridden woman who had 8 years of spasmodic torticollis symptoms. After 3 months of positive practice, she was able to ambulate unassisted; her therapeutic gains were maintained at a 1-year follow-up examination.

Biofeedback has been used by several authors: Leplow reviewed 184 biofeedback sessions in 10 patients. Considerable improvements occurred during this study; however, they occurred during the instructional phase or very early in the biofeedback training. This finding suggests that cognitive processes and visual feedback (ie, mirrors) might play an important role in the treatment of spasmodic torticollis; the biofeedback might only be of secondary importance.

In another single-case study the author reported gradual resolution during psychoanalysis. Hypnosis is used for both relaxation and specific behavioral-motor changes. Schneiderman et al report a single case that was cured with hypnotherapy: fewer than 3 hours of intervention occurred during 4 sessions, yet the complete resolution of symptoms was maintained through 18 months of follow-up care. Other single-case reports show good symptom control using hypnosis.

**Pharmacotherapy**

Medical treatment consists of a variety of therapeutic agents of different classes (Table 4). Many

<table>
<thead>
<tr>
<th>Drug</th>
<th>Total</th>
<th>Positive Responders</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amantadine</td>
<td>2</td>
<td>1</td>
<td>Sustained improvement after 2 years</td>
</tr>
<tr>
<td>Amantadine and haloperidol</td>
<td>3</td>
<td>3</td>
<td>Superior improvement, no relapse</td>
</tr>
<tr>
<td>Haloperidol</td>
<td>5</td>
<td>1</td>
<td>50% improvement at 4-week follow-up</td>
</tr>
<tr>
<td>Anticholinergic medications</td>
<td>19</td>
<td>10</td>
<td>Patients had positive responses to trihexyphenidyl</td>
</tr>
<tr>
<td>Benzodiazepine medications</td>
<td>7</td>
<td>7</td>
<td>Sustained improvement for 5 months of treatment</td>
</tr>
<tr>
<td>Levodopa</td>
<td>6</td>
<td>0</td>
<td>Very short duration of symptoms, diagnosis accurate</td>
</tr>
<tr>
<td></td>
<td>17</td>
<td>6</td>
<td>3 to 5 months of follow-up</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>0</td>
<td>2 patients had no relief from surgery</td>
</tr>
</tbody>
</table>

Table 4. Oral Pharmacotherapies for Spasmodic Torticollis.

*No studies found in the past 7 years (1989-1996) on MEDLINE.
studies of medical treatment are limited and consist of only 1 to 10 patients. In light of the relatively high rate of spontaneous remission, one must regard these studies with caution.

Levodopa has been used therapeutically by several authors, although others have suggested that levodopa treatment induced spasmodic torticollis. Positive results were reported in all 6 patients in one study; others found favorable responses in 6 of 17 patients, with sustained response in 3 of these.

Amantadine therapy was reviewed in two reports. A case report was described in which a patient had full symptom resolution that was sustained for 2 years of follow-up care; a second study showed no benefit in 9 patients.

A review of anticholinergic therapy investigated in five studies totaling 25 patients reported little to no improvement in 20 of the patients. One study of 100 patients with movement disorders, however, showed a 37 percent response rate among patients with spasmodic torticollis.

Haloperidol similarly had mixed responses: 9 of 27 patients treated with haloperidol alone had favorable results. Haloperidol and amantadine together produced good to excellent outcomes in 11 of 13 patients.

Benzodiazepine medications have also been used to good effect in a few patients. There is very little in the literature regarding benzodiazepines; no references were found searching MEDLINE from 1989 to 1996.

Until the advent of botulinum toxin, many pharmacotherapeutic trials suffered from highly variable responses, faulty research design, clinical heterogeneity, and small sample size. Botulinum toxin type A treatment (Botox injections) has become widely accepted as the most efficacious medical treatment, with more than 60 studies accessed on this topic. Botulinum toxin is a presynaptic inhibitor of acetylcholine release. The local injection of botulinum toxin temporarily denervates the muscle, in a manner similar to the surgical results discussed below. The neck muscles are palpated for hypertrophy, increased tone, and pain: serial local intramuscular injections are administered. Doses of botulinum toxin range from 60 to 240 U per muscle group. Greater efficacy was found at higher doses, with an overall response rate approximating 80 percent (Table 5).

The patient in our case report received about 200 U per session at a cost of $4.05/U of botulinum toxin type A (Botox). She was given an injection every 3 months, and her total quarterly bill was about $1200, which was paid by her health maintenance organization.

Anderson et al suggested botulinum toxin therapy as a first-line treatment and reported pain reduction and symptom improvement in a review of 107 patients receiving botulinum toxin treatments. The median duration of benefit was 9 weeks with a median of four injections over a 15-month period. Adverse effects included transient dysphagia in 44 percent of patients that was severe enough to require hospitalization for hydration in 2 percent of the patients. Other adverse

<table>
<thead>
<tr>
<th>Study</th>
<th>Total</th>
<th>Positive Responders</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anderson et al</td>
<td>107</td>
<td>101</td>
<td>Dose of 200-300 U per muscle, with 7-10 days for maximal response</td>
</tr>
<tr>
<td>Boghen and Flanders</td>
<td>32</td>
<td>24</td>
<td>Only transient side effects, dosing 50-100 U per muscle</td>
</tr>
<tr>
<td>Mezaki et al</td>
<td>51</td>
<td>*</td>
<td>Better responses with 240 U per muscle than lower doses (60-120 U)</td>
</tr>
<tr>
<td>Borodic et al</td>
<td>35</td>
<td>29</td>
<td>Substantial improvement in pain; 24 had improvement in posture and in spasms</td>
</tr>
<tr>
<td>Giladi</td>
<td>108</td>
<td>97</td>
<td>66 improved range of motion, and 97 had alleviation of pain and muscle tension</td>
</tr>
<tr>
<td>Mezaki et al</td>
<td>60</td>
<td>38</td>
<td>Objective improvement; 93% felt better subjectively</td>
</tr>
</tbody>
</table>

*Doses were progressively titrated to achieve response; number of positive responders not indicated in translation.
effects, such as neck weakness, dry mouth, dysphonia, fatigue, and local pain, were observed but did not result in discontinuation of therapy.

Patients can develop antibodies to botulinum toxin, which result in decreased treatment efficacy with time. In a recent review of botulinum toxin, Boghen and Flanders found more objective proof of injection effectiveness by assessing not only pain but posture, tremor, and active range of motion of the neck.

Surgery

Surgery can provide a permanent alternative to the medical treatment of spasmodic torticollis. Surgery is recommended for patients who had initial favorable response to botulinum toxin and then became secondary nonresponders or for those who refuse further botulinum toxin injections. Surgical approaches in the past included such extensive or nonspecific therapies as thalamotomy or bilateral nerve or muscle sectioning. These procedures have been largely replaced by selective peripheral denervation. Selective peripheral denervation has been shown to be effective with few adverse effects. The involved muscles are located through electrical stimulation or nerve block to determine their effect on nuchal movement and are then denervated by cutting the nerve root either within or outside the dura. This operation is known as a rhizotomy; Bertrand more specifically labels it a ramiectomy, a cutting of the posterior (motor) ramus of the nerve. In a review of 260 patients treated with this procedure, he found clinical improvement in 80 percent without serious adverse effects (Table 6).

Conclusion

Spasmodic torticollis is a poorly understood disorder marked by hyperkinesis of the cervical musculature resulting in unwanted sustained head protrusions. Information found on the history and physical examinations are the key to the diagnosis. A wide variety of treatment options are available to help meet the needs of individual patients.

References


