Spasmodic Torticollis: A Case Report and Review of Therapies

David L. Smith, MD, and Maria C. DeMario, DO

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.)

Spasmodic torticollis is a clinically diagnosed movement disorder in which many authors describe psychologic accompaniments. The incidence is approximately 1 in 100,000, with either an insidious or an abrupt onset. Abrupt onset is usually secondary disease rather than the subtle onset of idiopathic disease.

Although the disease is uncommon, psychologic and oral pharmacologic therapies are generally part of the family physician’s repertoire.

This case illustrates the diagnosis and treatment of spasmodic torticollis in a patient with historic and current psychologic symptoms.

Methods
We describe a case of a woman with spasmodic torticollis and reviewed the literature published from 1989 to 1996 indexed in MEDLINE using the following key words: “torticollis,” “spasmodic torticollis,” “therapy,” “behavior therapy,” “treatment,” “botulinum toxin.”

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 paravertebral musculature. She had full range of motion of the neck in all planes with intact motor strength, and there were no other motor or sensory deficits. Cerebellar function and reflexes were normal bilaterally; no other tremor, tic, or dystonia was observed. Laboratory test results, including muscle enzymes and serum ceruloplasmin, 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 abate her symptoms after a 2-month trial at 100 to 150 mg each day.

The patient was prescribed in succession baclofen, haloperidol, and clonazepam. She had only mild improvement on these medications, however, so she resumed nortriptyline until her condition 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 symptoms; at a 2-year follow-up examination, the patient 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 abnormal 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 predominance.1,2 (Secondary spasm is a self-limited disorder and will not be further discussed.)

Few studies address the natural history of spasmodic torticollis. Physicians who provide care for their patients for 7 or more years note deterioration during the first 5 years, a plateau for 5 years, then slight improvement.1 Spontaneous remissions (partial or complete) have been reported in up to 60 percent of patients in some series;1 others note full remission in 16 percent, with sustained remission for 12 months of 6 to 12 percent.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 (anterocollis), or inclined to the side (laterocollis). A combination of muscles is often involved, resulting in latero-torticollis or retro-torticollis, and so forth. Pure retrocollis (6 percent of cases) and pure anterocollis (3 percent) represent symmetrical involvement of muscles: most cases are asymmetrical, and the involved hypertrophied muscles can readily be palpated and contrasted to the contralateral normal musculature. The sternocleido­mastoid 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 involved: 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 involvement ranged from 0 to 80 percent. Others noted subclinical evidence of a bilateral disturbance, 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 situations and usually do not occur with sleep. Pain in the neck and shoulders can accompany spasmodic torticollis, but it is unusual as a presenting symptom.4 Pain can develop later, however, as the result 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 antagoniste), 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 association 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 1. Antecedents to Spasmodic Torticollis. |
|-----------------|-----------------|
| Type            | Characteristic  |
| Psychologic     | Psychosis       |
|                 | Affective disorder (unipolar and bipolar) |
|                 | Psychiatric hospitalization for any reason |
|                 | Anxiety         |
|                 | Cyclothymia     |
|                 | Dysthymia       |
|                 | Somatoform disorder |
|                 | Personality disorder |
| Biologic        | Family history of movement disorders |
|                 | Head and neck trauma |
|                 | Neuroleptic drug use |

| Table 2. Treatment Options for Spasmodic Torticollis. |
|-----------------|-----------------|
| Type            | Component       |
| Behavioral therapy | Positive practice |
|                 | Progressive relaxation |
|                 | Massed negative practice |
|                 | Visual feedback  |
|                 | Biofeedback      |
|                 | Hypnosis         |
| Pharmacotherapy | Levodopa         |
|                 | Amantadine       |
|                 | Anticholinergic medications |
|                 | Haloperidol      |
|                 | Benzodiazepine medications |
|                 | Botulinum injections |
| Surgery         | Selective peripheral denervation |
Spencer et al\textsuperscript{16} 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\textsuperscript{17} 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.\textsuperscript{16}

Biofeedback has been used by several authors: Leplow\textsuperscript{18} 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.\textsuperscript{19}

Hypnosis is used for both relaxation and specific behavioral-motor changes. Schneiderman et al\textsuperscript{20} 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.\textsuperscript{21-23}

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

\begin{table}[h]
\centering
\caption{Behavioral Therapies for Spasmodic Torticollis.}
\begin{tabular}{|l|c|l|}
\hline
Therapy & Number of Patients & Outcome \\
\hline
Relaxation, positive practice, visual feedback\textsuperscript{16} & 1 & Remission, maintained at 2-year follow-up \\
Positive practice\textsuperscript{16} & 1 & Full remission through 1-year follow-up \\
Negative practice\textsuperscript{17} & 2 & 1 patient had full remission for more than 22 months \\
Biofeedback\textsuperscript{18} & 10 & Mild to marked improvement in 8 patients, no long-term follow-up \\
Psychoanalysis\textsuperscript{19} & 1 & Complete remission, no specified follow-up \\
Hypnosis\textsuperscript{20} & 1 & Complete remission at 1.5-year follow-up \\
\hline
\end{tabular}
\end{table}

\begin{table}[h]
\centering
\caption{Oral Pharmacotherapies for Spasmodic Torticollis.}
\begin{tabular}{|l|c|c|l|}
\hline
Drug & Patients & Positive Responders & Comments \\
\hline
Amantadine & 2 & 1 & Sustained improvement after 2 years\textsuperscript{24} \\
 & 9 & 0 & Transient improvement in 1 patient\textsuperscript{14} \\
Amantadine and haloperidol & 3 & 3 & 2 patients "excellent," 1 patient "good" response despite failing to respond to surgical denervation\textsuperscript{24} \\
Haloperidol & 6 & 1 & Sustained improvement for 5 months of treatment duration, no relapse on withdrawal of medication\textsuperscript{25} \\
 & 5 & 0 & Amantadine failures\textsuperscript{14} \\
 & 16 & 8 & 50\% improvement at 4-week follow-up\textsuperscript{14} \\
Anticholinergic medications & 19 & 10 & Patients had positive responses to trihexyphenidyl\textsuperscript{26} \\
 & 24 & 4 & Scopolamine or atropine\textsuperscript{14} \\
 & 1 & 1 & 10-year remission after treatment with trihexyphenidyl\textsuperscript{14} \\
Benzodiazepine medications* & 7 & 7 & Very short duration of symptoms, diagnosis accurate\textsuperscript{24} \\
Levodopa* & 6 & 0 & 3 to 5 months of follow-up\textsuperscript{14} \\
 & 17 & 6 & 3 of 6 had long-term remission off medications\textsuperscript{25} \\
 & 3 & 0 & 2 patients had no relief from surgery\textsuperscript{14} \\
\hline
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*No studies found in the past 7 years (1989-1996) on MEDLINE.

438 JABFP Nov.-Dec. 1996 Vol. 9 No. 6
Table 5. Botulinum Toxin Type A Therapy for Spasmodic Torticollis.

<table>
<thead>
<tr>
<th>Study</th>
<th>Total</th>
<th>Positive Responders</th>
<th>Comments</th>
</tr>
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<tbody>
<tr>
<td>Anderson et al12</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 Flanders13</td>
<td>32</td>
<td>24</td>
<td>Only transient side effects, dosing 50-100 U per muscle</td>
</tr>
<tr>
<td>Mezaki et al31</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 al12</td>
<td>35</td>
<td>29</td>
<td>Substantial improvement in pain; 24 had improvement in posture and in spasms</td>
</tr>
<tr>
<td>Giladi33</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 al34</td>
<td>60</td>
<td>38</td>
<td>Objective improvement; 93% felt better subjectively</td>
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</tbody>
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*Doses were progressively titrated to achieve response; number of positive responders not indicated in translation.

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,14 although others27,28 have suggested that levodopa treatment induced spasmodic torticollis. Positive results were reported in all 6 patients in one study14; others found favorable responses in 6 of 17 patients, with sustained response in 3 of these 6.25

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 care24; a second study showed no benefit in 9 patients.14

A review of anticholinergic therapy investigated in five studies totaling 25 patients reported little to no improvement in 20 of the patients.14 One study of 100 patients with movement disorders, however, showed a 37 percent response rate among patients with spasmodic torticollis.26

Haloperidol similarly had mixed responses: 9 of 27 patients treated with haloperidol alone had favorable results.14 Haloperidol and amantadine together produced good to excellent outcomes in 11 of 13 patients.27

Benzodiazepine medications have also been used to good effect in a few patients.29,30 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,12,31 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.12,13 Doses of botulinum toxin range from 60 to 240 U per muscle group. Greater efficacy was found at higher doses,31 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 al12 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
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 ramiexcitomy, 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**


<table>
<thead>
<tr>
<th>Author</th>
<th>Patients</th>
<th>Comments</th>
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<tr>
<td>Davis et al</td>
<td>260</td>
<td>88% total or marked relief (&quot;marked&quot; defined as slight deviation or slight residual movement). Results maintained in 167 patients with &gt; 5-year follow-up, and 64 patients with &gt; 10-year follow-up. Early intensive postoperative physiotherapy for 6 weeks</td>
</tr>
<tr>
<td>Horner et al</td>
<td>41</td>
<td>Mild radiographic swallowing abnormalities preoperatively in 68% of patients. Postoperatively, moderate to severe swallowing abnormalities seen radiographically in 95%, with gradual improvement within 4-24 weeks</td>
</tr>
<tr>
<td>Friedman et al</td>
<td>58</td>
<td>85% had marked improvement; 72% had complete relief of muscle spasms. 45% had mild transient swallowing problems in immediate postoperative period, which diminished over several months</td>
</tr>
<tr>
<td>Braun and Richter</td>
<td>50</td>
<td>76% had marked improvement or disappearance of symptoms, maintained at 25-month average follow-up, with &quot;no major side effects&quot;</td>
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