

The Spasmodic Torticollis Handbook

*A Guide to Treatment
and Rehabilitation*

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What Is Spasmodic Torticollis?

BASICS

You are reading this book presumably because you suffer from tremor, abnormal unwanted movements, or crooked posture of your head and neck. This is an uncommon condition, and you may have been suspected of having a number of underlying causes for it. A number of different medical conditions may manifest as an abnormal head and neck posture, and the term *torticollis* is loosely applied to many of them. In fact, *torticollis* simply means “twisted neck.” This book deals specifically with the neurologic condition known as *spasmodic torticollis* (ST), also known as *cervical dystonia*. Other causes of abnormal posture will be briefly discussed to differentiate them from ST.

Gathering information is the first step in managing any chronic disorder. By reading this book, you will learn about factors that may cause this condition, how this condition manifests itself, how it progresses, and medications that are used to treat it, as well as exercises that may help you to maintain flexibility and control the pain that may accompany ST. Also included are some tips from other people suffering from ST. This book will serve as a reference for you to share with your doctor and family.

For purposes of the discussion in this book, we provide the following definition: Spasmodic torticollis is a neurologic disorder that results in an involuntary turning or twisting of the head and neck, causing them to assume a forced abnormal posture. It is difficult for the person with ST to voluntarily move his or her head back to a normal straight position. In order to give you a thorough understanding of your disorder, we will first examine the above definition in detail and establish a basic vocabulary.

Neurologic disorders are those that affect the nervous system: the brain, the spinal cord, and the nerves that extend out of the

spinal cord to the rest of the body (Figure 1). *Sensory nerves* are those that bring sensory information from the body to the spinal cord to be relayed to the brain (a few sensory nerves, such as those from the ears and eyes, deliver information directly to the brain). *Motor nerves* are those that send information from the brain directly or via the spinal cord to muscles of the body. It is through the motor nerves that muscles are made to contract and cause movement in a particular body part. In general, the brain and spinal cord are referred to as the *central nervous system* (CNS); the motor and sensory nerves are referred to as the *peripheral nervous system*.

The brain contains a number of different types of cells. Those that receive, relay, or send information in the form of electrical and chemical signals are called *neurons*. Certain areas of the brain contain groups of neurons whose main activity is to send, control, or

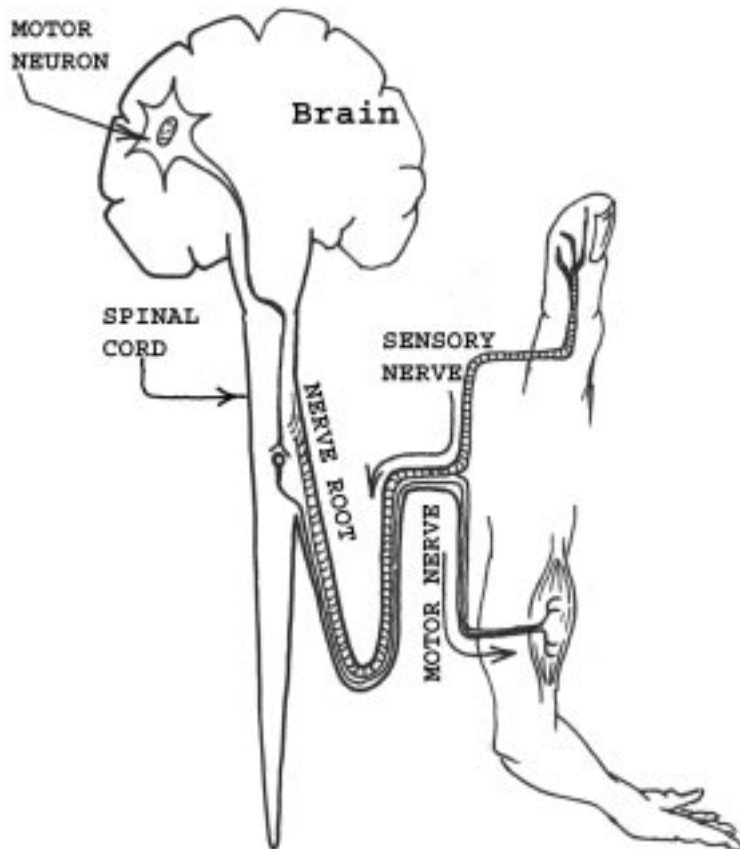


Figure 1 *The nervous system.*

modify information that is sent out through the motor nerves to the muscles. These brain areas, their neurons, and their signal pathways through the spinal cord and motor nerves to the muscles are collectively known as the *motor system*.

Neurologic disorders that affect mainly the motor system are known as *movement disorders*. Movement disorders affect the body's movement and can result in increased stiffness of a limb or other body part due to an increase in the normal resting tone of muscles (rigidity), a slowing down of the normal speed of movements, or an incoordination or loss of dexterity in movements. Some movement disorders may involve an excess of unwanted or difficult-to-control movements, such as tremors or abnormal contorted postures in affected body parts. *Parkinson's disease* is one of the most well known movement disorders. Its symptoms include a slowing down of movements, tremors, and rigidity. Movement disorders that involve mainly an increase in muscle tone and that cause sustained abnormal postures in affected body parts are called *dystonias*. If the affected body part also has tremor, writhing movements, or other uncontrollable motion, the disorder may also be called a *dyskinesia*.

ST is thus a dystonia that mainly affects the muscles of the neck. It may also be termed a dyskinesia if there is abnormal neck motion. The word "spasmodic" indicates abnormal contractions, or spasms, of various muscles that bring about the contorted position. The primary abnormality in ST is not in the neck, however. It is currently believed that the underlying problem is in the motor system of the CNS, specifically in a part of the motor system called the *basal ganglia*. Abnormal signals from the brain cause the head and neck to assume an involuntary contorted posture. The word *involuntary* should be stressed here. This means that the affected individual is not voluntarily, or volitionally, moving his or her head into the abnormal posture. He or she may have to exert voluntary effort to try to bring the head back to normal position, but when that effort is stopped, his or her head will return to the abnormal position. The neck is also known by its Latin name, *cervix*, and thus another term for ST is *cervical dystonia*.

THE SYMPTOMS AND CLINICAL COURSE

Now that you have an understanding of the definition of ST, we will go over the signs and symptoms of this disorder in more detail.

ST may begin at any age, but appears most often between the ages 25 to 55 years. It affects men and women about equally, and has no predilection for any particular race or ethnic group. The disorder usually develops gradually. Neck discomfort, mild pain, and a feeling of stiffness may be the earliest symptoms. Muscle contractions may produce subtle jerking movements of the head that resemble a tremor. These early manifestations may not be present all of the time. They may occur only when the individual is fatigued, such as at the end of the day; or may appear transiently during or after physical stress or exercise; or during anxiety or emotional stress. There may be a subtle tilt to the head, but individuals at this stage are seldom aware of their torticollis as a distinct medical disorder. They usually believe they are experiencing “muscle tension,” and treat themselves with over-the-counter pain medications, massage, rest, and sometimes chiropractic care.

When the symptoms persist, they may see their primary physician. Unless the signs of the disorder are obvious at this point, they may then, appropriately, receive conservative treatments with prescription pain medications, muscle relaxants, and possibly referral to a physical therapist or chiropractor. Sometimes, in mild cases, these conservative measures may be all that are required to keep the symptoms at a reasonably tolerable level, even if a definitive diagnosis of ST has not been made. In many cases, however, the disorder progresses over months, or even a few years. The symptoms become more continuous and persistent. Head and neck position becomes more contorted and the abnormal posture is obvious to the patient and to others. Pain and stiffness may increase, and pain medications or muscle relaxants may no longer be affective. It becomes increasingly difficult to voluntarily move the head back to neutral position. Head tremor may increase, resembling a nodding “yes-yes” or a shaking “no-no” motion. Occasionally, the dystonia can spread to adjacent body parts such as the shoulders, arms, hands, or the back and spine. In very rare cases, severe dystonia can affect the entire body.

Depending on the severity that the disorder eventually reaches, the twisted posture and pain begin to interfere with a person’s normal functioning. An affected person might have trouble shaving, combing his or her hair, or applying makeup, especially if there is significant tremor. It may become more difficult to read, watch television, or drive. Job performance may be impaired. Social interac-

tions may also suffer. This may be due to the individual's own self-conscious embarrassment. It can also be due to aversion or avoidance by strangers, coworkers, or acquaintances; the individual may be assumed by others to have a psychiatric disorder.

Occasionally, especially early in the course of the disease, symptoms of ST will spontaneously remit. In a few cases, remission may be complete and permanent. In the majority of cases, however, the symptoms will reappear at a later date, and the ST becomes a chronic permanent disorder. The general observation has been made that the greater the length of time that symptoms have been present, the more likely that the condition will be permanent. The ST almost never resolves if symptoms have been present for more than one or two years. Its slow progression usually reaches a plateau after about five years, at which point symptoms become stable.

In the chronic stage, the severity of symptoms varies from day to day. Symptoms tend to worsen with fatigue, heavy exercise, and stress, while sleep and relaxation tend to reduce them. Many people find relief by lying on their back, although this is not true for everyone. The neck movements always stop during sleep and, for some people, they do not reappear for a period of time after awakening. This is referred to as the "honeymoon period," and can last from a few minutes to a few hours. The length of the honeymoon period tends to shrink as time goes by. Neck pain may become the predominant symptom.



CASE 1

Sam first began to notice head shaking when he was 40 years of age. In fact, his family first noticed he was shaking his head as if in disagreement several months before he realized something was wrong. They noticed that his head shook off and on, especially when he was driving. His coworkers also noticed this and wondered why he shook his head "no" so often. Sam did not realize his head was shaking until one day he noticed he was looking to the right all the time. It became more difficult to watch television unless his head was supported on the back of his favorite recliner. He wasn't able to read the paper at the breakfast table and had to retreat back to his recliner to do so. He began to have difficulty driving and tended to rest his head on the headrest in order to look straight ahead. He went to his family doctor, who told Sam that he

did not know what was wrong and prescribed a course of physical therapy. Sam completed the physical therapy despite feeling worse after the more rigorous exercise, and was no better. Although the head turning was uncomfortable and caused him problems, he decided that there wasn't anything else to do for this condition and resolved to continue the exercises and live with it. Gradually, his neck returned to the neutral position and he remained symptom free for the next 20 years. Then, while he was walking for exercise, Sam noticed his head again pulling to the right. This time, his family doctor sent him to a neurologist specializing in movement disorders, who diagnosed Sam as having ST.

GESTE ANTAGONISTE: THE “SENSORY TRICK”

Many people with ST develop a tendency to place their hand on the side of their neck or place their fingers against their chin or cheek. For some reason, a light sensory stimulus on the skin of the head or neck dampens the tremors and reduces the severity of the twisting, making it easier to volitionally hold the head in a neutral position. This is known as a “sensory trick” or *geste antagoniste*. Most patients discover this for themselves without needing to be taught. They tend to use this method when reading, watching television, or eating. Unfortunately, the effect of the *geste antagoniste* is temporary, and full symptoms return as soon as the stimulus is removed. The sensory trick has different levels of effect for different individuals, and not everyone can benefit from it. The exact location of the area to be used and the way in which it is pressed or touched also differs. Some people find relief by putting a hand on the back of their neck. This may also be considered a *geste antagoniste*, although its mechanism of benefit may be different, having more to do with supporting the head; they may also find relief by lying down on their back. If you have not yet found an effective sensory trick, it may take some experimentation to find one that is effective for you.

SOCIAL EMBARRASSMENT

ST is a very noticeable medical disorder. When we think of those chronic neurologic illnesses that are perceived by our patients as creating a social stigma, few are as immediately obvious to casual observers as ST. Epilepsy shows no outward signs unless the patient

has an attack while being observed. Even Parkinson's disease may not be noticeable in its early stages, until tremor becomes more pronounced. Even then, Parkinson's disease has only a mild to moderate effect on the face, head, and neck. Among neurologic disorders, only paraplegia, with the patient using a wheelchair, is as obvious to all observers. However, paraplegia is rarely perceived as having a psychiatric component, and it is recognizable to most laypersons as the result (usually) of some physical injury. Wheelchair users are more familiar to laypeople, especially given the extensive media coverage of celebrity spinal cord injury during the 1990s. Parkinson's disease also has received public attention of late. Due to this awareness and the current age of "political correctness," strangers tend to be, or at least appear to be, fairly at ease around persons with these disorders.

The situation is not so easy for people with ST, whose disorder remains rare and obscure. Our face, head, and neck are the most visible parts of our bodies, and it is with these that we do most of our communicating. Not only our verbal output, but also our facial expressions and head movements, are continuously used—overtly and subtly—to interact with other people. Spasmodic torticollis disrupts this communication in a very obvious way, especially if the dystonia also involves facial muscles. It is impossible to hide. Add to this the fact that people with ST are often perceived as having an underlying psychiatric condition, and you can see that this disorder can severely impair social interactions.

Our patients who are diagnosed with ST share many of the responses of patients diagnosed with any chronic disease. They wonder how the disorder will affect their employment or professional lives, and how it will affect their personal and sexual relationships. Self-consciousness and social embarrassment can cause patients to become socially disengaged, give up public activities, and may become as disabling as the physical symptoms of the disease. Social embarrassment is one of the "hidden symptoms" of ST. Some patients become clinically depressed, although this condition is not as common with ST as with Parkinson's disease or stroke.

In our years of treating patients with disabling neurologic conditions, including paraplegia, Parkinson's disease, and stroke, we have seen these common initial reactions to becoming disabled or diagnosed with an incurable condition. We have found, however, that no matter how severe the initial reactions, patients seem to win out over

feelings of despair. The course generally takes one to two years from the time of occurrence or diagnosis. Those who persevere through this phase become surprisingly well adapted to their new state of health and ability. Social embarrassment diminishes greatly and, even if it persists, we rarely see patients who allow it to impede them in pursuing an active life professionally, socially, or professionally. Many look back on the initial phase of their illness and wonder why their despair or embarrassment was so great at the time.

What advice do we have for those newly diagnosed with ST? Simply this. Persevere. Be patient but persistent as you go through the medical system. Specialists experienced in treating your condition may be few in your location, but once under their management, you will find significant relief with current treatment options. Unlike some neurologic disorders, ST is not an inexorably disabling condition. Your pain and your outward symptoms will almost certainly improve with medical management, and your self-confidence and outlook will do likewise.