



A SURGICAL CURE FOR

# Chronic Blushing:

## *Endoscopic Thoracic Sympathectomy*

by Kara Showalter, CST

The sympathectomy procedure was developed in the mid-nineteenth century. It was well known at the time that the sympathetic nervous system affected many body systems, and it was surmised that the autonomic nervous system played a role in how the body regulates many body functions, including response to changes in environment, exercise and emotion.

### HISTORY

**W**illiam Alexander first performed the sympathectomy in 1889, while attempting to treat many conditions, including goiter, epilepsy, glaucoma and severe chest pain. Surgeons were unsuccessful in treating most of these conditions, although they did find a few situations where sympathectomy actually helped. For example, it helped stop intractable chest pain in those suffering from inoperable heart disease. It also put an end to uncontrollable sweating and blushing.<sup>7</sup>

A posterior approach to the procedure was developed in 1908, to provide better access to the nerve tissue in the chest cavity, but it required resection of the ribs, which was very painful. A supraclavicular approach was developed in 1935, to lessen the pain involved with the surgery, but this approach was more prone to damaging important nerves and blood vessels. Because of the risky nature of sympathectomy, the open approach was never a very popular procedure. It fell even further out of favor with the popularization of lobotomy.<sup>7</sup>

Sympathectomy did not become popular again until the 1980s, when an endoscopic version was pioneered by Goren

### LEARNING OBJECTIVES

- ▲ Examine the historical development of ETS
- ▲ Explore the physiology of the sympathetic nervous system
- ▲ Evaluate the indications for a potential ETS candidate
- ▲ Compare and contrast symptoms of Reynaud's Syndrome and erythromelalgia
- ▲ Analyze the ETS procedure, along with the reversal process

Claes and Christer Drott in Sweden. This endoscopic approach decreased recovery time and risks of nerve and vessel damage due to its minimally-invasive nature. Today, the procedure is commonly used to treat hyperhidrosis, facial blushing and some pain disorders affecting the sympathetic nerves.<sup>7</sup>



## PHYSIOLOGY

The autonomic nervous system can be subdivided into two branches that are responsible for controlling involuntary functions of the body, including heart rate, digestion, respiration rate, perspiration and many other basic functions of life. The two branches are the sympathetic nervous system and the parasympathetic nervous system.

These two systems work in opposition to each other to achieve a homeostatic effect. The sympathetic system works to speed up or strengthen a function, while the parasympathetic system works to slow down or weaken a function. The sympathetic nervous system is responsible for increasing heart rate, stimulating sweating, decreasing urine production, dilating the pupils, and reducing intestinal motility. Its main purpose is regulating functions necessary for the “fight or flight” response. The actions of the parasympathetic nervous system can be best summarized as “rest and digest.” It is responsible for decreasing the heart rate, constricting the pupils, increasing intestinal motility and digestion, increasing urine production, and aiding in providing balance and restoring energy.<sup>8</sup>

The sympathetic trunks are a paired bundle of nerve fibers that run from the base of the skull to the coccyx in a downward direction just lateral to the vertebral bodies. The sympathetic trunk is a vital part of the sympathetic division of the autonomic nervous system, as it allows nerve fibers to travel to spinal nerves that are superior and inferior to the one in which they originated. These nerve fibers are called sympathetic chain ganglia, and they are responsible for delivering information to the rest of the body regarding stress situations and the fight or flight response. These sympathetic ganglia are the structures that are destroyed during a sympathectomy.<sup>2</sup>

## INDICATIONS

Endoscopic thoracic sympathectomy (ETS) is performed for a variety of reasons: it is a treatment for idiopathic craniofacial erythema (chronic blushing), hyperhidrosis, and some pain disorders resulting from an overactive sympathetic nervous system.<sup>7</sup>

Idiopathic craniofacial erythema is a medical condition characterized by severe, frequent and uncontrollable reddening of the face, which is often unprovoked. It is unknown why people are afflicted with this condition, but it is definitely the result of an overactive sympathetic nervous system. Chronic blushing is primarily diagnosed by reviewing the patient’s history and by ruling out other conditions that would cause reddening of the skin. Options for conservative treatment for chronic blushing include oral medications and behavioral therapy. Several types of medications are used as treatment for idiopathic craniofacial erythema. Anxiolytics, such as Valium®, are used for anxiety; beta-blockers, which blunt the body’s reaction to stress; or anticholinergic drugs, such as robinol, ditropan, or propanthelin.<sup>3</sup>

Cognitive behavioral therapy has proved to be the most effective noninvasive treatment for chronic blushing. Cognitive behavioral therapy is a type of psychotherapeutic approach based on the idea that our thoughts, not external stimuli, dictate our behaviors. The benefit of this method is that it teaches patients how to deal with dysfunctional emotions in a systematic, goal-oriented process.

Another indication for ETS surgery is hyperhidrosis, or excessive sweating of the palms, face, or axilla. This condition is characterized by abnormally increased perspiration in excess of that required for regulation of body tempera-

concentrations as an antiperspirant and injections of botulinum toxin type A can be used to disable the sweat glands.<sup>4</sup>

Raynaud's Syndrome and complex regional pain syndrome are pain disorders that can be treated with ETS

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surgery. Raynaud's Syndrome is a painful vascular disorder that affects blood flow to the extremities when exposed to cold temperatures. Raynaud's is caused by a hypersensitivity of the sympathetic nervous system causing vasoconstriction of the peripheral blood vessels, leading to tissue hypoxia. Chronic, recurrent cases can lead to atrophy of the skin,

ulcerations of the skin, and ischemic gangrene. Raynaud's diagnosis is made by patient history and physical examination as well as by ruling out other conditions that could cause the vascular symptoms. Warming devices and vasodilators are the main forms of conservative treatment for Raynaud's Syndrome.

Hyperhidrosis is a condition that is inherited as an autosomal dominant genetic trait. Since it is a congenital condition, diagnosis usually results from a family and patient history. Conservative treatments include anticholinergic medications, such as iontophoresis, oxybutynin and robinol. Aluminum chloride can be used topically in high

## ALTERNATIVE TO ETS?

### What is Cognitive-Behavioral Therapy?

The term cognitive-behavioral therapy (CBT) does not represent a distinct therapeutic technique, rather, it serves as a general classification for a variety of therapies in the same family. According to the National Association of Cognitive-Behavioral Therapists, there are several approaches to cognitive-behavioral therapy, including: rational emotive behavior therapy, rational behavior therapy, rational living therapy, cognitive therapy and dialectic behavior therapy.<sup>1</sup>

The foundation of CBT is based on the idea that an individual's *thoughts* cause feelings, behaviors and reactions, not external stimuli, such as other people, situations or events.<sup>1</sup> If this is in fact the case, then theoretically, people are capable of making themselves feel or act better, even if the situation has not changed,

simply by changing the way he or she thinks about the given circumstance.

While other forms of therapy, such as psychoanalysis, can take years, CBT is a much faster process. This has to do, in part, with the differences in the therapist-patient relationship. A cognitive-behavioral therapist's role is to uncover the client's life goals and then figure out how to help the client reach these goals.<sup>1</sup> As stated by the National Association of Cognitive-Behavioral Therapists, "the therapist's role is to listen, teach and encourage, while the [patient]'s role is to express concerns, learn and implement that learning."<sup>1</sup> Put another way, cognitive-behavioral therapists do not tell their patients what their goals *should* be, or what they *should* tolerate. Instead, they *direct* their patients in ways to think and behave in order to obtain what they want.

The ultimate goal of cognitive-behavioral therapy in regard to chronic blushing

is to help stop the reaction by teaching the patient to adjust his or her expectations of social norms. According to researchers, people who blush excessively tend to have unrealistic expectations of how they should behave in social situations. They fear that even a small mistake will be mocked by others, so they become unnecessarily anxious and start to blush. In comparison, babies and small children, who have yet to develop these feelings about social interactions, do not blush at all.<sup>2</sup> By working with a cognitive-behavioral therapist, patients are able to work on changing the way they approach certain circumstances, reducing the likelihood of a flare up.

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Complex regional pain syndrome (CRPS) is a chronic progressive disease characterized by severe pain and swelling of a limb, accompanied by changes in the skin. Pain associated with CRPS is continuous and may be heightened by emotional stress. Moving or touching the affected limb is often intolerable and eventually the joints become stiff from disuse and the bones and muscles atrophy as well. The cause of CRPS is unknown, but is associated with an overactive sympathetic nervous system and diagnosis can be made by performing a series of tests. These tests include thermography, which is a technique used for measuring

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blood flow; radiography, which can detect osteoporosis up to two weeks after the onset of CRPS; and electrodiagnostic testing, which can differentiate between type I and type II CRPS. Conservative treatments for CRPS include physical and occupational therapy combined with pain and anti-inflammatory medications. The physical and occupational therapies are important components to managing CRPS primarily because they desensitize the affected body part and restore range of motion to improve the functionality of the limb.

Surgery is only recommended for severe and disabling cases of these conditions, and the procedure varies for treatment of each disorder. The sympathetic ganglia are identified by the level of the vertebra to which they correspond. For idiopathic craniofacial erythema and facial sweating, the nerve tissue is interrupted at the T2 level. In cases of hyperhidrosis of the palms, nerve tissue is interrupted at the T3 level. For hyperhidrosis of the axilla, the T3, T4, and T5 levels of nerve tissues are all interrupted. In treatment of both Raynaud's syndrome and complex regional pain syndrome, the T2 through T4 nerve ganglia are destroyed. Other variations in the procedure depend on the method used for interrupting the nerve tissues. Most surgeons prefer to clamp the nerves instead of completely resecting them primarily because the clamping method leaves the possibility of easier reversal of the procedure.<sup>6</sup>

## PROCEDURE

Several types of equipment and instrumentation are essential to successfully perform ETS. Necessary endoscopic equipment includes video towers, a fiber-optic light source and endoscopic instrumentation. A 5mm telescopic endoscope and a camera cord and light cord are also very important for performing this procedure. Endoscopic graspers, endoscopic scissors and clip applicators are also required. Soft tissue instrumentation that must be available during ETS surgery includes a hemostat, Adson pick-ups with teeth, a needle holder and suture scissors. Anesthesia will require a double lumen endotracheal tube to deflate the lungs and allow exposure of the surgical site.<sup>6</sup>

In order to prepare for the initial incision, the patient must be positioned, prepped, draped and anesthetized, and some practical considerations about room set-up must be made. The patient will be in the supine position with both arms extended on

padded arm-boards and a safety strap will be applied two inches proximal to the knees. General anesthesia will be utilized for this procedure, with the use of a double lumen endotracheal tube. The prep will begin at mid-chest level and extend from the shoulder, including the axilla, to the iliac crest and down to the table on the affected side. Four folded towels will then be placed around the incision site and a fenestrated drape will be placed on top of the towels. An important consideration regarding room set-up is that ETS is a bilateral procedure. To allow the surgeon an unobstructed view from either side, a video tower should be placed on each side of the patient. An extra set of both prepping and draping supplies should also be opened to allow the transition from one side to the other.<sup>1</sup>

To begin the procedure, the surgeon will direct the anesthesia provider to deflate the patient's lung on the affected side to facilitate exposure of the thoracic cavity. A #11 blade will then be used to make a 5mm incision between the patient's second and third ribs in the axillary plane. A disposable thoracic port is inserted through the incision and a 5mm telescopic endoscope is inserted through the port. The sympathetic chain is identified at the level that will be interrupted (between T2 and T5), and an endoscopic scissor is used to open the pleura. At this point, the nerve will be separated depending on the surgeon's preference of method, either by resecting it with

endoscopic scissors or by clamping it with an endoscopic clip applicator. The port is then removed, and a small thoracic catheter is inserted through the incision, which is then closed. Once the incision is closed, the lung is re-expanded, eliminating all residual pneumothorax through the small thoracic catheter. The catheter is then removed and wound closure is completed. Steri-strips™ and Mastisol® are applied to the closed incision, and the patient's other side is prepped and draped. The same procedure is then repeated on the opposite side. Re-intubation is not required during the transition since a double lumen endotracheal tube is used. A double lumen endotracheal tube allows the anesthetist to deflate and re-inflate either lung to facilitate exposure of the operative site. At the conclusion of the procedure, the patient will be transported to PACU, where a postoperative chest X-ray film will be taken to rule out residual air left in the thoracic cavity.<sup>6</sup>

Patients usually stay in the hospital overnight after ETS surgery, and patients may resume normal activities in about one week. Most patients who undergo ETS have suffered for many years from socially-disabling conditions, so instant relief from these conditions dramatically increases their quality of life, providing a 90-95 percent satisfaction rate. Prognosis for patients suffering from Raynaud's Syn-

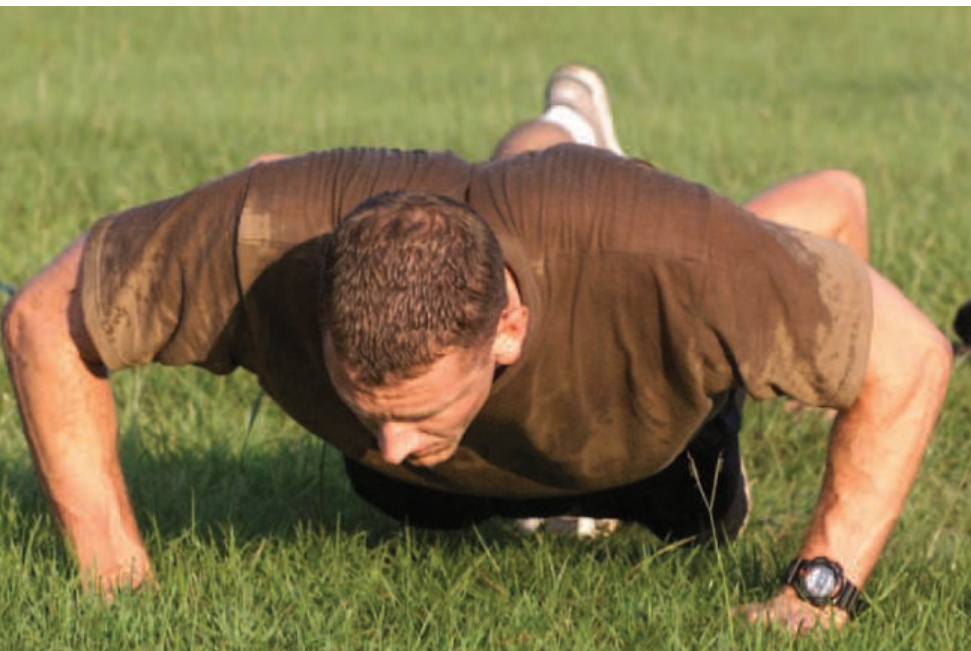
drome, or complex regional pain syndrome, depend on how far the disease has progressed, making the satisfaction rate unpredictable.<sup>5</sup>

Complications associated with ETS include infection, bleeding, respiratory problems, damage to nerves or arteries, compensatory sweating and Horner's Syndrome (Ocu-

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losympthetic Palsy). Compensatory sweating is the most common side effect of ETS surgery, occurring in approximately 50 percent of patients. It is a condition called compensatory hyperhidrosis, in which sweating is shifted from the hands, armpits, face or scalp to the upper and lower back, lower chest, abdomen, buttocks, groin, and backs of the thighs. Compensatory hyperhidrosis is usually mild, and most patients are able to tolerate it without any problems. Rarely, compensatory hyperhidrosis can be very severe, even more so than their original hyperhidrosis. This can cause patients to express regret regarding their sympathectomy and wish to have a reversal.<sup>5</sup>

Horner's Syndrome is the most serious complication of ETS. It results in a slightly smaller pupil and a disfiguring asymmetry of the face due to a slightly drooping upper eye-lid. This complication is caused by damage to the uppermost thoracic nerve node, also called the ganglion stellatum, and can only be reversed by plastic surgery to resect the affected eyelid (blepharoplasty). The risk of this complication depends mainly on the surgeon's familiarity with the procedure. Pneumothorax can also be a significant risk of this procedure. If any air is left in the thoracic cavity, respiratory problems may occur.<sup>5</sup>



## REVERSAL

Occasionally, patients wish to have thoracic sympathectomy reversed. Various reasons for dissatisfaction with the procedure include compensatory hyperhidrosis, obesity and the inability to perform exercise, and a lack of temperature control for the upper body. Resumption of nerve conduction and return to normal sympathetic regulation after a reversal of a thoracic sympathectomy can be a very lengthy process. Initial symptoms of recovery of sympathetic regulation may take six to nine months to show after a reversal. The reversal technique depends on the method used to interrupt the nerve tissues during the original operation.

Various reasons for dissatisfaction with thoracic sympathectomy include compensatory hyperhidrosis, obesity and the inability to perform exercise, and a lack of temperature control for the upper body.

If the surgeon who performed the original operation cut the nerve tissue in order to interrupt it, a nerve graft must be performed to restore the nerve. In this method, the location where the operation was previously performed is prepared by refreshing the edges where the nerve was previously cut. Then a nerve graft is harvested from the ankle region, usually the sural nerve is used, and is connected to the divided edges of the sympathetic chain with a biological gluing agent.

The cutting method is the hardest to reverse. The clamping technique is a much easier version of sympathectomy to reverse, which is why it has become the method of choice for ETS surgeons. In reversal of the clamping method, the clip is simply removed from the nerve and the nerve is allowed to regenerate on its own. This method has shown much quicker improvement in compensatory sweating, and the healing process can be reduced to about three to four months. Both reversal methods are performed endoscopically.<sup>6</sup>

## CONCLUSION

Although it should only be used as a last resort method of treatment due to the possibility of complications, endoscopic thoracic sympathectomy has proven to be an effective treatment for various conditions, including chronic blushing, hyperhidrosis, and pain syndromes associated with an overactive sympathetic nervous system.

## ABOUT THE AUTHOR

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# Man-on-Fire Syndrome

by Tom Borak

Imagine waking up one morning and feeling like your entire body, from head to toe, was on fire. You can't move from your bed, the skin on your toes is cracked and bleeding from the swelling in your feet. There is nothing you can do to dull the sensation of being engulfed in flames.

For a handful of people around the world, this nightmare is a daily reality. They suffer from a rare disease called erythromelalgia (EM), also known as Man-on-Fire Syndrome.

According to the U.S. National Library of Medicine's medical subject headings, a "rare" disease is classified as, "a large group of diseases, which are characterized by a low prevalence in the population. They frequently are associated with problems in diagnosis and treatment."<sup>1</sup> According to a story by ESPN's Steve Cyphers, which follows the journey of Kate Conklin, an EM patient who is training for a triathlon, the disease afflicts fewer than 400 people in the United States.<sup>2</sup>

The name, erythromelalgia, is derived from three Greek words: *erythros* (red), *melos* (extremities) and *algos* (pain).<sup>3</sup> First described in the 1870s, there is still relatively little that is known about this disease, which, due to its rarity, is often misdiagnosed or attributed to a psychological disorder. The symptoms are most common and most severe in the feet and hands. Flare-ups usually occur due to exposure to warmth, and physical activity is often a catalyst. A flare-up will often begin with an itching sensation, which builds into the severe pain and burning symptoms. The pain can be so intense that patients cannot walk.<sup>3</sup> Erythromelalgia is not related to complex regional pain syndrome (CRPS), although it can mimic some of the symptoms.<sup>4</sup>

One troubling aspect of the disease is that most cases are idiopathic, and can manifest at any point in life. Conklin, for example, did not experience symptoms until she was 28-years old.<sup>2</sup> Conversely, remissions are possible, but infrequent.<sup>4</sup> Since there is no known cause, there is no known cure. EM can also be a secondary development to additional medical conditions, including autoimmune, neurologic or blood disorders. Infrequently, EM can develop following an injury or surgical procedure.<sup>4</sup> Some people have the inherited or primary form of EM, and usually have other family members with the disease. Recently, an EM gene was identified, as were several mutations to this gene. Apparently, each affected family carries a different mutation, further complicating the process of finding a cure.<sup>4</sup>

There is evidence that suggests that erythromelalgia shares a common pathophysiology with Raynaud's Syndrome. In some instances, a patient has exhibited both conditions – sometimes simultaneously. Despite this possible relation, there is no evidence to suggest that the ETS procedure can have a positive impact on EM patients.<sup>5</sup>



An erythromelalgia flare-up is indicated on both shins.

It is extremely difficult for those who have not experienced the symptoms to fully comprehend the severity of the pain that these people are in. Conklin once burned herself on the exhaust pipe of a motorcycle, ultimately peeling herself off the bike with a second-degree burn. When asked by her doctor why she did not pull away, she responded, "that's the burning pain that I'm in, so I didn't notice anything different."<sup>2</sup>

Due to the nature of the illness, EM patients cannot wear close-toed shoes or socks. This can be particularly challenging during the winter months, when frostbite is a legitimate concern. Because many patients cannot feel pain unless it is more acute than the constant pain of EM, they are unaware of the damage that is occurring in other parts of their bodies. Many patients instinctively soak the affected parts of their body in cold water as a means to reduce the swelling and the burning sensation. However, according to the The Erythromelalgia Association, it has been demonstrated that icing or soaking can actually result in increased flaring, thus making the symptoms worse. Other problems may include skin tissue and nerve damage, infections, even severe ulcers that can take months to heal. Contrary to outdated medical information, this method of treatment is not advisable.<sup>4</sup> In one extreme case, a patient suffered near-fatal hypothermia related to the constant cooling to control the symptoms.<sup>3</sup>

Research is ongoing to learn more about the causes of erythromelalgia, and to search for a cure. For more information on Kate Conklin, go to ESPN.com and type "Outside the Lines: Burning Desire" into the search bar.

## Resources:

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