Selective peripheral denervation for spasmodic torticollis: 13-year experience with 155 patients

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Object. Botulinum toxin injections are the best therapeutic option in patients with spasmodic torticollis. Although a small number of patients do not benefit from such therapy, the majority respond well but may develop antibodies to the toxin after repeated applications. In those termed primary nonresponders, no improvement related to botulinum toxin has been shown. In patients in whom no response was shown and those in whom resistance to the therapy developed, selective peripheral denervation is a neurosurgical option.

Methods. Between June 1988 and August 2001, 155 patients underwent selective peripheral denervation. Surgery was performed at a mean of 8.5 years after the onset of symptoms (range 0.5–37 years). The mean age of the patients at the onset of dystonia was 39.7 years (range 17–77 years). For evaluation of results, patients’ responses were assessed. Results were obtained in 140 patients in whom the follow-up period ranged from 3 to 124 months (mean 32.8 months): 18 reported complete relief of their symptoms, 50 significant relief, and 34 moderate relief; 19 noted only minor relief and the remaining 19 no improvement. The results differ substantially when compared with those previously demonstrated in patients who received botulinum toxin injections. Although 80% of the secondary nonresponders were satisfied with the result of surgery, only 62% of the primary nonresponders considered the operation helpful. There were no major side effects. The recurrence rate was 11%.

Conclusions. The injection of botulinum toxin should be the first-choice treatment. If surgery is required, selective peripheral denervation provides the best results and has the fewest side effects compared with all surgical options.

Key Words • selective peripheral denervation • spasmodic torticollis • botulinum toxin • dystonia

The origin of spasmodic torticollis is unknown. Although a few therapeutic options for treatment of symptoms are available, conservative methods such as pharmacotherapy, psychotherapy,18 and physiotherapy are highly unsuccessful, a few symptomatic therapeutic options are available to date. Botulinum toxin A (and recently B) injections are widely accepted as the best therapeutic option; they unify low-risk and promising results.14 A minority of reported patients, however, did not respond to these injections. This group is divided into primary and secondary nonresponders, based on their dominant behavior after botulinum toxin injections. In these cases a more invasive treatment may need to be considered.

Neurosurgical procedures such as stereotactic thalamotomy or cervical rhizotomy have occasionally been successful, but subsequent severe side effects sometimes developed. During the 1980s, Freckmann, et al.,10 and Jho and Jannetta reported small series of 33 and 20 patients, respectively, in whom they performed intradural microvascular decompression of the spinal accessory nerve and the brainstem. The domain of thalamic or subthalamic supramaximal stimulation is the treatment for tremor, although it seems to have some influence on generalized dystonia.22 It is not clear whether it is of value in patients with local cervical dystonia. In 1891, Keen described a peripheral neurosurgical procedure for spasmodic torticollis. His method was refined by Bertrand, et al.,2 who termed it selective peripheral denervation, which has been shown to be highly successful and without major risks. In 1988 we modified their procedure.5 In this study, we present our long-term results with this neurosurgical treatment.

Clinical Material and Methods

Between June 1988 and August 2001 155 patients (74 men and 81 women) underwent surgery at our institution for spasmodic torticollis. Patient age at the onset of symptoms ranged from 17 to 77 years (mean 39.7 years). Selective peripheral denervation was performed a mean of 8.5 years later (range 0.5–37 years). A patient was considered to be a candidate for surgery if conservative methods were unsuccessful and symptoms had endured for at least 1 year. One exception was the third patient enrolled in this study, in whom surgery was undertaken earlier (0.5 years after the onset of symptoms). Since 1990, we have performed surgery exclusively in patients in whom there was no response to botulinum toxin A therapy. These nonresponders can be divided into two subgroups: so-called...
“primary nonresponders” in whom a response to botulinum toxin injections never occurred and “secondary nonresponders” in whom initial response was very good but in whom antibodies to the toxin developed. In the interim, we have treated 47 primary and 71 secondary nonresponders. Recently botulinum toxin B was introduced into the conservative treatment of spasmodic torticollis. Consequently surgery will only be recommended in the future, if the patient does not respond to both types of botulinum toxins.

A computerized tomography or magnetic resonance imaging study of the brain is performed to rule out a space-occupying mass or other intracranial disease entity. The involved muscles are identified by clinical examination and a multichannel electromyographic recording. A simultaneous record obtained in both SCM and splenius capitis muscles is mandatory and, if necessary, in both trapezius muscles as well. The dystonia is documented on video tape. The majority of our patients suffered from a combined involvement of ipsi- or contralateral splenius capitis and SCM muscles.

Surgical Technique

In all cases surgery was performed after induction of general anesthesia with the patient in a semi-sitting position and his/her head fixed in a Mayfield clamp; a right atrial catheter was inserted, precordial or transesophageal Doppler ultrasonography was performed and end-expiratory partial CO₂ pressure recorded. The patient may be placed in a prone position as well. It is worth mentioning, however, that the posterior branches of C-1 and C-2 nerve roots are regularly surrounded by enormous suboccipital venous plexuses. These plexuses tend to bleed heavily when surgery is performed in the prone positioned patient, thus making the identification of the important posterior branches of C-1 and C-2 nearly impossible. Because we have never found evidence of air embolism during this procedure we strongly recommend the semi-sitting position. The different nerve branches have to be identified using monopolar stimulation, and therefore we refrain from administering muscle relaxants.

Surgery is initiated with the denervation of the SCM muscle. We estimate the exit point of the spinal accessory nerve from this muscle in the lateral neck triangle by applying a supramaximal transcutaneous electrical stimulation (2 Hz). Using a 5-cm skin incision on the posterior muscle margin, we first identify the trapezius branch of the spinal accessory nerve and then follow it proximally under amplification. Injury of the greater auricular nerve (crossing the operative field) must be avoided. Once the main trunk of the spinal accessory nerve has been reached, its branch (or branches) to the SCM muscle (Fig. 1 upper) can be detected by electrical stimulation (Radionics RFG-3B and TEC Ganglion Gasseri electrode; stimuli of 0.6 to 2 V and 2-msec duration; Radionics Co., Burlington, MA). Afterward, they are severed and widely resected (Fig. 1 lower). This muscle may be further innervated through anterior branches of the C-1 and C-2 nerve roots or through the recurrent nerves, leaving the trapezius branch of the spinal accessory nerve. These nerves must be resected as well.

In the second step the involved posterior neck muscles are denervated via a separate skin incision. In addition to the trapezius muscle, the autochthonous neck muscles receive their innervation from the posterior Cl–Tl branches. Among these, the Cl–6 nerves are the most important. Selective peripheral denervation of the neck muscles can be conducted superselectively—that is, with any muscle denervated separately (splenius capitis, semispinalis capitis or cervicis, and inferior oblique muscles). Given that botulinum toxin injected into the splenius capitis muscle probably leads to a paralysis of the deeper ipsilateral muscles as well (by passive diffusion of the toxin), we recommend not only the denervation of the involved splenius capitis muscle but also the complete denervation of the autochthonous muscle group on the ipsilateral side. As stated previously, this is accomplished by the complete resection of the posterior Cl–6 nerve branches on the involved sides. Because the trapezius muscle is spared, no craniovertebral instability has to be feared, even if a bilateral neck denervation has to be performed.

The posterior branches can be best identified extraspinally, lateral to the joint facets, leaving the anterior branches (innervating shoulder and arm muscles) intact.

*Fig. 1. Intraoperative photographs. Upper: Main trunk of the left spinal accessory nerve in lateral neck triangle with division into the SCM (asterisk) and the trapezius muscle branches (arrow). Lower: Surgical field after resection of the SCM muscle branch.*
They are approached via a midline skin incision in the neck, running from the external occipital protuberance down to the C-7 spinous process, allowing both the left and the right side to be surgically treated. As in cranial procedures, we routinely do not shave the hair. Having reached the C2–6 spinous processes, the inferior oblique capitis muscle is detached from its origin at the C-2 spinous process. Using microsurgical technique, we then enter the cleavage plane between the semispinalis capitis and cervicis muscles on the involved side. Blunt dissection is performed down to the area lateral to the articular facets until the level of the multifidus muscle is reached. The large posterior branch of C-2 (greater occipital nerve) is easily localized beneath the inferior oblique muscle. The most difficult nerve is the posterior branch of C-1 (suboccipital nerve); however, it is regularly identified 1.5 to 2 cm lateral off the midline above the arch of the atlas and below the VA within the vertebral sulcus (Fig. 3 upper).

The posterior branches of the C3–6 nerves are found on a perpendicular plane between the lateral margin of the semispinalis cervicis muscle and the medial margin of the semispinalis capitis muscle. Whereas the posterior nerve branches C3–4 are easily identified lateral to the facets, and the tiny branches C5–6 are found medially, running on the surface of the semispinalis cervicis muscle (Fig. 3 lower). After resection of all posterior branches of C1–6 we stimulate the entire region lateral to the articular facets, looking for any further muscle contraction. Often, additional posterior branches can be found within the multifidus layer, which also have to be resected. To prevent nerve regeneration, all the proximal and distal nerve stumps are extensively coagulated. After surgery, patients were sent to a rehabilitation institution to undergo physiotherapy for approximately 3 weeks.

A minority of our patients (15 cases) had to undergo surgery twice because their symptoms recurred. Again, the involved muscles were identified using multichannel electromyographic recording. The surgical site was then reopened, and we searched for nerves that might have been missed during the first procedure or for fibers that might have spontaneously regenerated.

**Patient Assessment**

Because most of our patients live far away from our hospital, postoperative results were primarily gathered...
using native-language questionnaires (such as German and Dutch). The overall result was assessed through three different inquiries. First, patients had to specify whether they would have chosen to undergo selective peripheral denervation again to treat their dystonia. Results in all patients who answered “no” were considered as failures. Second, patients had to note the percentage of improvement (from 100% indicating no complaints, to 0% reflecting no improvement, to a “negative improvement” indicating deterioration). Third, they had to score the overall result of surgery (1, very good; 2, good; 3, moderate; 4, minor improvement; 5, no improvement; and 6, deterioration). Interestingly these results were consistent in all cases—that is, a moderate improvement corresponded to a percentage between 40 and 60%.

The second part of the questionnaire was focused on treatment-related adverse side effects, specifically sensory deficit or pain at the back of the head, loss of shoulder strength, ear-related sensory deficit, scar pain, head or neck instability and swallowing problems. The patient had to determine whether the side effect was disturbing. Furthermore, the patient could list any other complaint.

In addition to the mentioned self-assessment, the complex TWSTR Scale score,14 widely used by neurologists performing botulinum toxin injections, was used to assess the effect of surgery. This scale concerns head mobility (≤ 35 points), handicap in daily life (≤ 30 points), and pain (≤ 20 points). A score of 0 would indicate the absence of torticollis and 85 points the most severe torticollis.

**Results**

The overall effect of the selective peripheral denervation was based on results of the aforementioned patient self-assessment tools. To date, 140 patients have responded during a mean follow-up period of 32.8 months (range 3–124 months).

One hundred two (73%) of 140 patients were satisfied with the operation, whereas 38 (27%) regarded the operation as ineffective. Furthermore, 13% noticed a complete relief of their symptoms, 36% a significant, and 24% a moderate improvement of their dystonia. In contrast, 14% (19 patients) reported only a minor and 14% (19) no improvement. Patient age, the different head and neck muscles involved, and the time from onset of symptoms to surgery did not have any influence on outcome.

In contrast, the results differ substantially among those in whom no response to botulinum toxin occurred and those in whom an initial good response ceased to occur. Seventy-one (60%) of 118 patients who had received botulinum toxin before surgery experienced a good initial response. Subsequently antibodies developed, proven either by laboratory examinations or by test injections into the small muscles of the thumb or foot (secondary nonresponders). In 47 (40%) of 118 patients no benefit was derived from botulinum toxin therapy, and the cases were classified as primary nonresponders. Whereas 80% of the secondary nonresponders improved markedly, only 62% of patients in whom a primary nonresponse occurred were satisfied with the result of surgery. In the seven patients with complex generalized dystonia, in whom limbs and trunk were also involved, only one patient (14%) improved slightly after surgery.

The photograph in Fig. 4 left shows a patient with retrocollis preoperatively; Fig. 4 right shows the result 1 year after selective peripheral denervation of the bilateral autochthonous neck muscles.

Our preliminary results basing on the TWSTR Scale do not differ substantially from those obtained using the self-assessment scales. The mean preoperative score was 48 points (range 40–64), which decreased to 33 points postoperatively (range 1–64). Interestingly only patients in whom outcome was rated very good improved markedly from 51 to 10 points. In the other groups the TWSTR Scale score reduction did not differ significantly. In the group with a good outcome, it changed from 46 to 34 points, and in the group with moderate outcome from 53 to 43 points. Surprisingly, a nearly equivalent reduction of
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points occurred in the group with minor improvement (54–45 points), and the group with no improvement (54–46 points). Notably, in two patients who considered the operation a failure significant improvement occurred, as reflected by a reduction of points from 63 to 37 points and from 55 to 29 points, respectively. Because patients generally tend to rate the success of any treatment lower than the examining physician, the results obtained using self-assessment scales normally appear to be worse than those based on so-called objective scores.

To date, we have observed no major complications related to selective peripheral denervation. None of our patients complained of head or neck instability. Because the procedure does not require making contact with the brain or spinal cord, no limp paresis or mental disturbances have to be anticipated. An injury to the VA during the dissection of the posterior branch C-1 is possible, but none has occurred during our procedures. We had to evacuate a postoperative hematoma in the area of the SCM muscle in three patients. Four patients reported a prolonged (≤ 3-month) period of dysphagia of unknown origin; the glossopharyngeal nerve was not affected in these cases. One patient required a temporary tracheostomy after development of a laryngostenosis, probably triggered by a tracheal cuff irritation.

The following minor adverse sequelae were observed: all patients suffered a variable sensory deficit in the area of the greater occipital nerve, which cannot be avoided because the nerve arises from the posterior branch of C-2 (we regularly observed muscular contractions after its stimulation and therefore severed it); three patients (2%) complained of a temporary occipital neuralgia, which spontaneously resolved within 3 months; and some patients experienced a sensory deficit of varying degrees in the region of the greater auricular nerve. During our first 19 procedures we inadvertently sectioned the trapezius branch of the spinal accessory nerve in two patients. In both cases the course of the branch was extremely superficial and had already been affected during the skin incision. It was sutured immediately, and full recovery was demonstrated in both patients. We then began using the surgical microscope at the start of each procedure and consequently did not observe this complication again. In two cases, excessive postoperative scar tissue led to a compression of this branch, causing a paresis of the trapezius muscle. In one case, neurolysis was required. With the help of physiotherapy only a slight weakness (Grade 4/5 paresis) remained in both patients. In two other cases, the tremor component of the dystonia was intensified postoperatively, probably because of the now absent stabilizing effect of the denervated hyperactive muscles.

Symptoms recurred in 11% of the patients. Fifteen patients had to undergo surgery twice. In at least three patients a spontaneous functional motor nerve regeneration after resection of the posterior branches could be proven histologically. In nine additional patients a functional recovery occurred in combination with preexisting nerve fibers, which obviously had been overlooked during the first procedure.

Discussion

Botulinum toxin blocking of neuromuscular transmis-

sion is currently the best treatment available in symptomatic patients with spasmodic torticollis. Additionally, selective peripheral denervation is an appropriate surgical counterpart to the botulinum toxin therapy. As the toxin spreads by diffusion, however, muscles not closely related to the injection site may be inadvertently paralyzed. Therefore, this chemical therapy is not as specific as the operation. These adverse effects do not normally occur after selective peripheral denervation. Additionally, in secondary nonresponders, the desired effects of botulinum toxin can be reached by selective peripheral denervation. Nearly 80% of the patients in this group can be effectively treated without major risks and with good long-term prognosis. Of note, however, if the trapezius muscle is the leading muscle involved in the dystonia, surgery is not recommended because of risk of an omaphrosis.

To date, it is not clear why some patients do not benefit from botulinum toxin injections. Preexisting antibodies may be one explanation, although an unnoticed infection with Clostridium botulinum seems to be very unlikely. An underdosage of the botulinum toxin can be excluded by administering a second higher-dose injection. In our opinion, the involvement of almost all head and neck muscles can be presumed in most of these cases. This is supported by our finding that botulinum toxin had an effect on the dystonia in some patients only if simultaneous untoward side effects such as head instability were obvious. In these cases one might expect, that surgical denervation would not confer much benefit. Surprisingly, 62% of our primary nonresponders were satisfied with the result of surgery. Given that the risk of the procedure is low, surgery is indicated also in primary nonresponders. In patients with complex generalized dystonia, however, the situation is different. Although selective peripheral denervation may improve the cervical dystonia, the procedure induces no greater benefit because the torticollis represents only a small part of the overall symptomatology. Therefore we ceased to perform surgery in this group of patients.

Although selective peripheral denervation is performed in only a few neurosurgical departments worldwide,2,5,8,9 this procedure is responsible for considerable progress in the treatment of spasmodic torticollis. Bertrand and his group1,4,5 are the most experienced surgeons in this field. In their hands, excellent or good results have been demonstrated in 88% of the patients. The results reported by Arce and Russo,2 Davis, et al.,8 and Dieckmann9 have been promising as well. Thus far, no fatality has been reported.

During the 1980s, in the era of increasing suspicion of neurovascular compression syndromes, neurosurgeons such as Freckmann and colleagues9 and Jho and Jannetta3 favored microvascular decompression of the spinal accessory nerve and the brainstem. To date, the theory that vascular compression of these structures is a cause of spasmodic torticollis remains very controversial. Therefore, the success of microvascular decompression as a curative treatment seems to be very unlikely. Freckmann and colleagues, and Jho and Jannetta reported minor instances in 33 and 20 patients, respectively. Aksik1 treated 22 patients and reported an improvement rate of 77%, whereas Jho and Jannetta demonstrated a success of 90%. In contrast, improvement was noted in only 67% of the patients studied by Freckmann and colleagues. Furthermore, they reported one death related to surgery. In the
 aforementioned studies the authors first treated patients in the era before botulinum toxin was introduced. The latest follow-up review was performed primarily by questionnaire (as in parts of the present study). Unfortunately it has not been reported if their patients received botulinum toxin after surgery. Therefore it is uncertain whether the reported improvement was the sole effect of surgery or caused by additional botulinum toxin injections. In contrast, because we have, since 1990, only treated those patients without responses to botulinum toxin our results are clearly based exclusively on the effect of surgery.

Recently, there have been several preliminary reports on the use of pallidal stimulation in patients with generalized dystonia. Islekel, et al., treated one patient and Krauss, et al., treated three patients with spasmodic torticollis. It is widely known that the effect of stimulation tends to decrease within a few years. Thus, although these patients improved immediately after implantation of the electrical device, the long-term effect of deep brain stimulation in patients with local cervical dystonia is not yet known.

Conclusions

Promising therapeutical options were introduced into the treatment of spasmodic torticollis in which chemical and surgical denervation of the involved dystonic muscles is performed. To date, evidence supports the injection of botulinum toxin as the first treatment of choice. If a surgical procedure has to be considered, selective peripheral denervation provides the best results and has the fewest side effects among all surgical options. Surgery alone is recommended in patients in whom there is no response or no longer a response to botulinum toxin A and B therapy.

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References


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