

Technique

# Dorsal root ganglionectomy for the diagnosis of sensory neuropathies. Surgical technique and results

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## Abstract

**Background:** Inflammatory diseases stand out among sensory neuronopathies because, in their active phase, they can be treated with immunosuppressive agents. Immunosuppressive therapy may present severe adverse effects and requires previous inflammatory activity confirmation. Sensory neuronopathies are diagnosed based on clinical and EMG findings. Diagnostic confirmation and identification of inflammatory activity are based on sensory ganglion histopathological examination. We describe the surgical technique used for dorsal root ganglionectomy in patients with clinical/EMG diagnosis of sensory neuronopathies.

**Methods:** The sensory ganglion was obtained from 15 patients through a small T7-T8 hemilaminectomy and foraminotomy to expose the C7 root from its origin to the spinal nerve bifurcation. In 6 patients, the dural cuff supposed to contain the ganglion was resected en bloc; and in 9 patients, the ganglion was obtained through a longitudinal incision of the dural cuff and microsurgical dissection from the ventral and dorsal roots and radicular arteries. All ganglia were histopathologically examined.

**Results:** No ganglion was found in the dural cuff in 2 patients submitted to en bloc removal, and the ganglion was removed in all patients who underwent microsurgical dissection. All but 2 patients that had ganglion examination presented a neuropathy of nerve cell loss, 3 with mononuclear inflammatory infiltrate. These patients underwent immunosuppressive therapy, and 2 of them presented clinical improvement. No surgical complications were observed.

**Conclusions:** Microsurgical dorsal root ganglionectomy for diagnosing inflammatory sensory ganglionopathies was effective and safe. Although safe, en bloc resection of the proximal dural cuff was not effective for this purpose.

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## Keywords:

Dorsal root ganglionectomy; Sensory neuropathy; Surgical technique; Diagnosis

*Abbreviations:* 1st, First; 3rd, Third; 4th, Fourth; 5th, Fifth; 7th, Seventh; 8th, Eighth; C3, Third cervical vertebra; C4, Fourth cervical vertebra; CSF, Cerebrospinal fluid; EMG, Electromyographic; HE, hematoxylin-eosin; NISP, nonmalignant inflammatory sensory polyganglioneuropathies; T6, Sixth thoracic vertebra; T7, Seventh thoracic vertebra; T8, Eighth thoracic vertebra.

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## 1. Introduction

Sensory neuropathies associated or not with an autonomic and with a small or absent motor component may be predominantly secondary to involvement of dorsal root ganglia. Subacute or chronic “polyganglioneuropathies” [23] occurring in patients not using medications or not exposed to industrial toxic agents may be associated with carcinomas, [5,25] with Sjögren syndrome, [1,8-10, 12-14,16,17] or with monoclonal gammopathies, [4] or may be idiopathic. Idiopathic NISPs [23] are ganglionopathies of unknown nature, although there are suggestions that they may depend on an underlying autoimmune process. Information about the pathology of NISP is limited because few reports are available about autopsies or biopsies of the dorsal root ganglion. These findings include a mononuclear inflammatory infiltrate consisting of T cells of the cytotoxic/suppressor type, [9] deposits of immunoglobulin, [4] and reduced number of neurons with proliferation of satellite cells. No effective treatment exists for NISP, but a clinical improvement occurred in a patient with the use of prednisone. Clinical improvement has been reported for another patient treated with plasmapheresis [1]. There are also reports of spontaneous improvements that, paradoxically, are related to electrophysiological worsening. This fact may depend on functional improvement by adaptation of the patient to his sensory ataxia [9]. However, no controlled studies are available in which the efficacy of any treatment was confirmed. Because there are no estimates of its incidence or prevalence among nonhospital populations and because idiopathic NISP seems not to be frequent, a controlled study of this type should be multicentric.

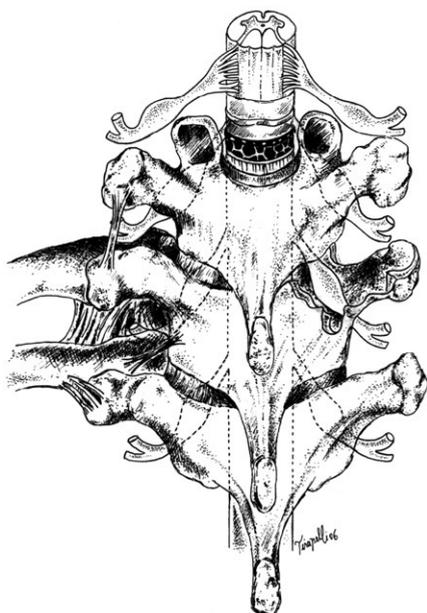


Fig. 1. Schematic view of 2 thoracic vertebrae with the spinal cord and spinal nerves. A small lateral resection of the superior and inferior parts of the adjacent laminae (T7-T8) was performed; and the spinal nerve was exposed laterally, opening the intervertebral foramen underneath the articular facets. The bulging portion of the nerve corresponds to the dorsal root ganglion.

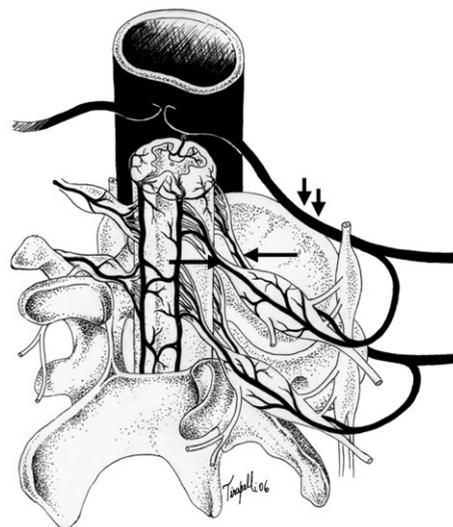


Fig. 2. Schematic view of the vascularization of the spinal cord at the midthoracic level, with emphasis on the formation of the radiculomedullary arteries (arrows) from the spinal branch of the segmental (intercostal) artery (double arrow).

In view of the above considerations and of the fact that sensory ataxia as well as pain may be incapacitating in patients with NISP, the neurologist, based on the pertinent medical literature and with the patient agreement, is ethically authorized to make attempts to treat with plasmapheresis, corticosteroids, and nonsteroidal immunosuppressive drugs. The doses used, the duration of the therapeutic attempt, and the combination of different types of immunosuppressive treatments mainly depend on the presence of an inflammatory process. This is determined by the detection of an inflammatory infiltrate and/or deposits of immunoglobulins in the sensory ganglia. This detection is possible only by a ganglionectomy, which, however, is an invasive procedure requiring general anesthesia. The procedure should be carried out only after the patient has been fully informed about all surgical steps and risks and only if he is willing to submit to immunosuppressive treatment. Few reports of sensory ganglion exeresis are available. Smith [22] and Osgood et al [19] have described techniques for sensory ganglion exeresis in the thoracic and lumbosacral region, respectively, for the treatment of chronic pain. Griffin et al [9] reported exeresis of a sensory ganglion in the thoracic region for histopathological examination but did not describe in detail the technique used.

The objective of this study was to describe the surgical technique used for exeresis of a dorsal root ganglion in patients with NISP diagnosed based on clinical and EMG criteria [23].

## 2. Materials and methods

This study was approved by the Ethical Committee of the Hospital das Clínicas, Ribeirão Preto Medical School, University of São Paulo.

Table 1  
Clinical, EMG, and histopathological findings and surgical technique in patients submitted to dorsal root ganglionectomy

| Case | Sex/Age (y) | Neurological examination  | EMG                                       | Surgical technique                           | Histopathology  |
|------|-------------|---|---|--|---|
| 01   | F/47        | Astasia and abasia for 6.5 y. Generalized sensory ataxia, loss of tendon reflexes, and absence of vibratory sensation   | Sensory neuropathy                        | En bloc resection of the dural cuff          | Marked reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Chronic sensory neuropathy  |
| 02   | F/29        | Gait ataxia for 2 mo. Vibratory sensation loss at the hands and inferior limbs. Light touch anesthesia on legs and feet   | Sensory neuropathy                        | En bloc resection of the dural cuff          | Slight reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Slight chronic sensory neuropathy   |
| 03   | M/49        | Gait ataxia for 1 y. Stocks and gloves anesthetics  | Sensory neuropathy                        | En bloc resection of the dural cuff          | Slight reduction in the ganglionic cells and proliferation of satellite cells. Nodules of Nageotte<br>Slight chronic sensory neuropathy   |
| 04   | M/45        | Chair-bound (intense sensory ataxia) for 8 mo. Pain in the legs. Absence of vibratory sensation and of tendon reflexes  | Sensory neuropathy                        | En bloc resection of the dural cuff          | Marked reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Chronic sensory neuropathy  |
| 05   | M/51        | Gait ataxia for 6 mo. Generalized decrease in the tendon reflexes. Decreased light touch and pain sensibility in the arms and legs. Decreased vibratory sensation in the legs and feet  | Sensory neuropathy                        | En bloc resection of the dural cuff          | No dorsal root ganglion found on the surgical specimen  |
| 06   | M/52        | Paraparetic gait for 1 mo. Generalized decrease in the tendon reflexes. Decreased proprioceptive sensation in the legs and feet   | Axonal, predominantly sensory, neuropathy | En bloc resection of the dural cuff          | No dorsal root ganglion found on the surgical specimen  |
| 07   | M/54        | Ataxia of the hands, transient hypertonia and atrophy in the inferior limbs, sphincteric incontinence, sexual impotence, and loss of recent memory for 6 y. Gait ataxia, loss of tendon reflexes. Positive Romberg sign. Left dysmetria. Global decreased vibratory sensation | Sensory axonal neuropathy                 | Selective dorsal root and ganglion resection | No change in the ganglionic cells<br>No inflammatory infiltrate   |
| 08   | M/38        | Gait ataxia and paresthesias in the inferior limbs for 8 y. Positive Romberg sign. Generalized loss of tendon reflexes 01/27/98—Pulses of methylprednisolone. Important clinical improvement  | Sensory neuropathy                        | Selective dorsal root and ganglion resection | Slight reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Slight inflammatory (lymphocytes) un specific infiltration<br>Slight chronic sensory neuropathy |

|    |      |   |   |  |   |
|----|------|---|---|--|---|
| 09 | F/70 | Numbness in the legs and hands for 3 mo. Difficulty in walking. Global loss of tendon reflex. Left dysmetria and truncal ataxia. Touch and pain sensations decreased below the nipples. Global loss of vibratory sensation. Chair-bound 05/26/99—Pulses of methylprednisolone. Unchanged  | Severe sensory axonal neuropathy              | Selective dorsal root and ganglion resection | Severe reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Moderate lymphomononuclear infiltration<br>Chronic sensory neuronopathy   |
| 10 | F/53 | Paresthesias in the inferior limbs for 20 mo. Dysmetria in the inferior limbs. Positive Romberg sign. Gait ataxia. Decreased light touch and pain in gloves and stocks. Decreased vibratory sensation in the hands, knees, and feet<br>Pulses of methylprednisolone. Clinical improvement | Axonal, predominantly sensory, polyneuropathy | Selective dorsal root and ganglion resection | Slight reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Slight inflammatory (lymphocytes) un specific infiltration<br>Chronic, axonal, predominantly sensory neuronopathy |
| 11 | F/44 | Ascending paresthesias (feet, legs, and hands) for 3 mo. Chair-bound due to intense sensory ataxia. Generalized loss of vibratory sensation and tendon reflexes   | Asymmetric axonal sensory polyneuropathy      | Selective dorsal root and ganglion resection | Slight reduction in the ganglionic cells. Satellite cells proliferation. Nodules of Nageotte<br>Slight chronic sensory neuronopathy   |
| 12 | M/39 | Dysesthesias in the feet and legs for 9 mo and difficulty in walking in the last 4 mo. Gait ataxia. Decreased light touch and pain in gloves and stocks. Loss of vibratory sensation in the feet and decrease in the rest of the body   | Predominantly sensory polyneuropathy          | Selective dorsal root and ganglion resection | Slight reduction in the ganglionic cells. Rare nodules of Nageotte  |
| 13 | F/18 | Pain and burning in the feet and legs for 19 mo. Decreased light touch and pain sensations in stocks. Decreased vibratory sensation in the feet   | Axonal predominantly sensory polyneuropathy   | Selective dorsal root and ganglion resection | No change in the number of ganglionic cells. Only 2 nodules of Nageotte   |
| 14 | M/32 | Numbness in feet and legs for 3 y. Gait with help. Left dysmetria. Decreased light touch and pain sensation in gloves and stocks. Vibratory sensation decreased in the hands and hips and lost in the knees and feet  | Sensory neuronopathy                          | Selective dorsal root and ganglion resection | Slight reduction in the ganglionic cells. Some nodules of Nageotte  |
| 15 | M/48 | Numbness in the feet and legs for 2 y. No objective neurological changes  | Sensory neuropathy                            | Selective dorsal root and ganglion resection | No change in the ganglionic cells<br>Chronic sensory neuropathy   |

F indicates female; M, male.

### 2.1. Surgical technique

The ganglion was obtained from 15 patients with clinical-EMG diagnosis of NISP through a small thoracic hemilaminectomy associated with a foraminotomy [22]. Eight patients were female and 5 were male. Age varied from 18 to 70 years old (mean,  $44.6 \pm 12.31$ ).

A paramedian 4-cm skin incision centered on the spinous apophysis of T7 is made. The right side aponeurosis is incised in a semicircle based on the midline of T6-T8. The paravertebral muscles are displaced from the spinous process and from the dorsal surface of posterior laminae of T6-T8 until the transverse processes are visualized. After a small lateral bone resection of the T6-T7 or T7-T8 laminae, with a small Kerrison rongeur or a high-speed drill, the dural cuff of the T7 or T8 roots is identified and followed through a foraminotomy for exposing the root up to its division in dorsal and ventral branches (Fig. 1). The root emerges from the spinal canal inferiorly to the superior articular facet; the sensory ganglion usually is identified by a local bulging (Fig. 1). Bone resection and further dissection are carried out under a surgical microscope for better visualization and identification of neurovascular structures. The dural cuff is exposed laterally from the dural sac distally to the ganglion.

In the first 6 cases, the dural cuff was tied and cut en bloc proximal (close to its emergence from the dural sac) and distal to the bulging where the ganglion was supposed to be. In the last 9 cases, the dural cuff was exposed further laterally to the bifurcation of the spinal nerve, exposing the intercostal nerve originating from the ventral branch and allowing identification of the radiculomedullary artery originating from the intercostal artery. The radiculomedullary artery with its corresponding veins accompanies the roots (Fig. 2). The dural cuff was longitudinally incised, the dorsal ganglion was identified, the ventral and dorsal roots were split, and the dorsal root was sectioned proximal and distal to the ganglion (Fig. 3). When separation was not possible, usually at the level of the ganglion, the ventral and the dorsal roots were sectioned together. A proximal ligature of the dural cuff was necessary in only one case because of CSF leakage. Isolation of these structures generally causes some bleeding that requires bipolar coagulation. Care should be taken to avoid ganglion or radiculomedullary artery lesions due to coagulation. Usually, small arteries are identified side-by-side with the nerve in the intervertebral foramen; but sometimes, a large branch is present. The surgical wound is closed by suturing the aponeurosis, subcutaneous tissue, and skin.

### 2.2. Histopathological examination

The dorsal ganglia fixed in 10% phosphate-buffered formalin were embedded in paraffin and prepared for light microscopic examination. Ganglia transversal sections  $5 \mu\text{m}$  thick were stained with HE for qualitative analysis of number of neurons and presence of nodules of Nageotte and

mononuclear inflammatory infiltrate, with Masson trichrome for evaluation of amount of connective tissue, and with Congo red for detection of amyloid deposits.

### 3. Results

Table 1 presents a summary of the clinical, EMG, and histopathological findings and the surgical technique used in 15 patients submitted to ganglionectomy. No ganglion was included in the removed segment in 2 of 6 patients submitted to en bloc removal of the dural cuff. In all patients submitted to dural cuff opening, the ganglion could be well identified and removed with the dorsal root or en bloc with the ventral and dorsal roots. No surgical complications or postoperative neurological deficits were observed in the 15 operated patients.

Histopathological examination of ganglia obtained from all but 2 patients showed evidence of neuronopathy with neuronal loss (Fig. 4A-B), characterized by a reduction in neuron density and proliferation of satellite cells forming nodules of Nageotte (Fig. 4C). Evidence of mononuclear inflammatory infiltrate (Fig. 4D) was found in the ganglia of 3 patients, and they were submitted to immunosuppressive

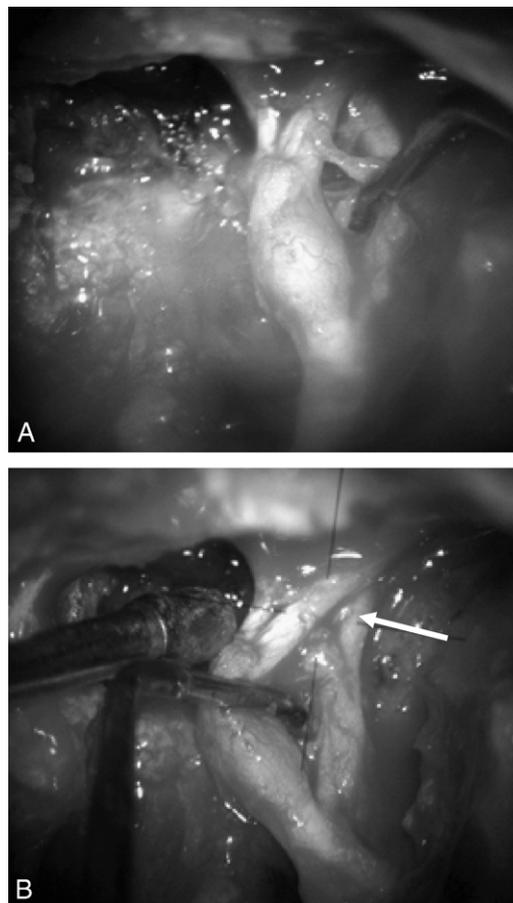


Fig. 3. A: Exposure of a sensory ganglion after opening of the dural sheath under the surgical microscopy. B: The ganglion is being isolated from the anterior root (arrow).

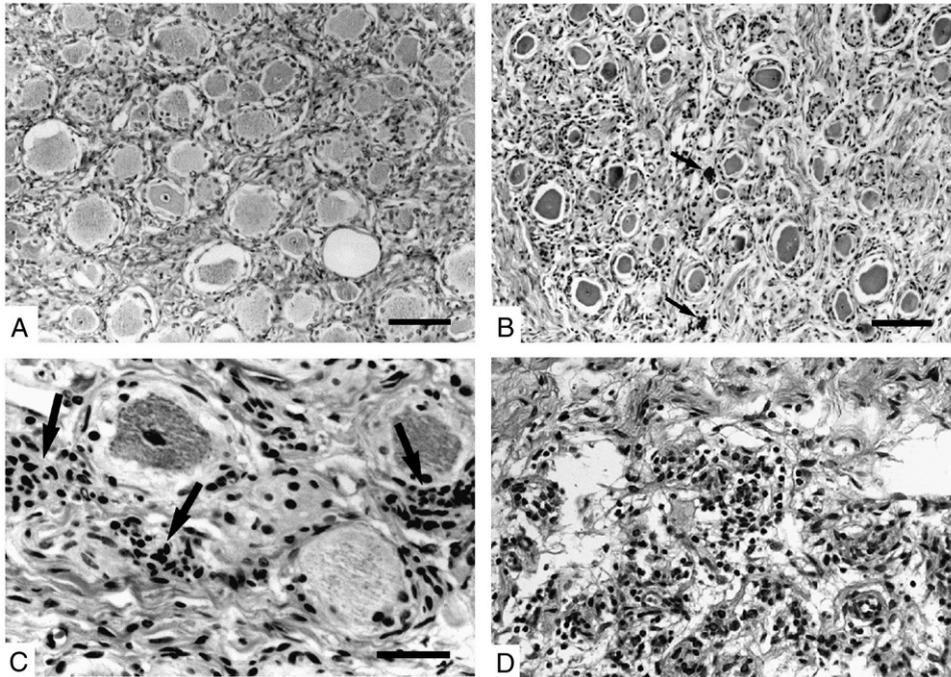


Fig. 4. Microphotography of dorsal root sensory ganglia. A: With normal neurons (control). HE,  $\times 200$ . B: With evidence of neuronal loss and nodules of Nageotte (arrows) indicating neuronal loss. HE,  $\times 200$ . C: A detail of some Nageotte nodules (arrows). HE,  $\times 600$ . D: Mononuclear inflammatory infiltrate (arrows) indicating active disease. HE,  $\times 400$ .

therapy. Two of them had significant improvement of their symptoms. No amyloid deposits were found in the examined ganglia.

#### 4. Discussion

##### 4.1. Clinicopathological comments

Although it was not the aim of this study to analyze the clinicopathological aspects, they deserve comments. No cellular infiltrates or deposits of immunoglobulins were found in the ganglia of our patients. This may be due to the chronicity of our cases (phase without active inflammation), even considering that the disease seemed to be insidious and slowly progressive. Recently, some studies suggested that these ganglionopathies might be associated with antiglycolipid antibodies [9,15,19]. If these antibodies are attacking ganglionic cells at the time of the ganglionectomy, immunoglobulin deposit probably could be found in the ganglion. We also did not find correlation between the ganglionic cell loss and the clinical severity of patients (case 11 was chair-bound; and cases 3, 8, 10, and 12 had an important ataxia with little ganglionic cell loss), suggesting a probable multifocal nature of the NISP.

Effective doses of pharmacological immunosuppressors for at least 3 months are required to evaluate their efficacy. The 3 patients with signs of active illness in the ganglion analysis underwent immunosuppressive treatment, and 2 of them had significant improvement of their symptoms. As there is no marker for disease activity in the NISP and

pharmacological immunosuppression could be of risk, a ganglionectomy for supporting its indication is justifiable. Therefore, we think that ganglion biopsy must be considered before taking the decision of pharmacological immunosuppression. In addition, ganglionectomy will allow pathological and immunopathological studies that may share more light about the knowledge on the NISP.

##### 4.2. Comments on the surgical technique

The need of a dorsal root ganglion for histopathological diagnosis and evaluation of inflammatory activity in NISP gave rise to the question of which ganglion could be resected without neurological deficit. The thoracic region was chosen due to the small depth of the laminae and to the possibility of resecting even one motor root without causing significant neurological deficit. This decision was possible because all patients submitted to biopsy presented involvement of the thoracic dermatomes.

Smith [22] described a technique for resection of thoracic sensory ganglion for treatment of intercostal pain. In this procedure, the dural cuff containing the dorsal ganglion and the sensory and motor roots was removed en bloc using hemilaminectomy and foraminotomy. Ligation of the proximal stump of the dural cuff with a silver clip to prevent CSF fistula was recommended. We used this technique modified in our first patients, performing unilateral exposure of 3 hemilaminae (T6–T8) and foraminotomy to reduce surgical time and to minimize the discomfort resulting from the undermining of paravertebral muscles.

However, in 2 patients, the ganglion was not found in the en bloc removed dural cuff segment.

Osgood et al [19] reported a microsurgical technique for resection of lumbosacral sensory ganglia for treating chronic pain using an incision along the dural sheath proximally and distally to the ganglion, with separation and preservation of the anterior root and usually of the small segmental artery. The distal sheath of the root and of the ganglion was approached through a small resection of the lower facet, as recommended by Scoville [21]. No CSF leakage was observed during these procedures. This fact can be explained because in this region, called a *transition zone* in peripheral nerves, there are trabeculae of the arachnoidal membrane that seal the subarachnoid space [18]. Despite anatomical differences between the root sheaths and their contents in the lumbosacral and thoracic regions, we adapted the Osgood technique for exposure of thoracic dorsal root ganglion in our last patients. Exposure of the dural cuff was performed further laterally using microsurgical techniques to allow adequate identification and preservation of the structures (neural elements and possible large segmental arteries) and to avoid pleural perforations. We could isolate and remove the dorsal root ganglion in some patients; but in other cases, adhesion of the ganglion to the ventral root prevented it, and the ganglion and roots were resected together. Cerebrospinal fluid leakage was not common when the dural sheath of the root was opened; and when necessary, ligature of the dural sheath was done with mononylon to avoid artifacts during computed tomography and magnetic resonance image examinations.

Preservation of the spinal cord circulation is a major concern in this procedure. Irrigation of the spinal cord originates from branches of the vertebral arteries and spinal or root branches of intersegmental arteries, and they are divided into 3 major regions: [6,11,15,20] *superior* or *cervicothoracic* (cervical spinal cord and 1<sup>st</sup>-3<sup>rd</sup> thoracic segments), *intermediate* or *median thoracic* (4<sup>th</sup>-8<sup>th</sup> thoracic segments), and *inferior* or *thoracolumbar* (3 or 4 distal thoracic and lumbar segments).

The *anterior spinal artery* originates from the joining of 2 recurrent branches (*anterior spinal arteries*) of the vertebral arteries and receives the *radicular* or *spinal arteries* derived from segmental vessels [2,3,6,7,11,15,24]. The *spinal arteries* penetrate the intervertebral foramina and divide into a *peripheral branch* to bone case and a *central branch* that originates the *dorsal* and *ventral root arteries* that accompany the ventral and dorsal roots of the respective spinal nerves [7,11,15,20]. The role of these branches in the irrigation of the spinal cord is controversial [11,15]. At the C3-C4 level, the anterior spinal artery receives 1 or 2 branches of the *spinal* or *radiculomedullary arteries* (from the *vertebral arteries*); in the lower cervical part, it receives a branch from the *ascending cervical* or *deep cervical arteries*; and in the upper thoracic region, it receives a branch of the *spinal* or *radiculomedullary artery* (from the 3<sup>rd</sup>, 4<sup>th</sup>, or 7<sup>th</sup> *intercostal arteries*) [6,15,20]. In

the dorsolumbar region, it receives a *spinal branch* (*magna radicular artery* or *Adamkiewics artery*) originated from one of the intersegmental branches of the descending aorta in the last thoracic or in the 1<sup>st</sup> lumbar segments (from 7<sup>th</sup>-12<sup>th</sup> intercostal arteries in 75% and from 5<sup>th</sup>-7<sup>th</sup> intercostals or from the 1<sup>th</sup>-3<sup>rd</sup> lumbar arteries in 25% of cases) and originated on the left side in 75% of cases. This artery is more voluminous than other tributaries of the *anterior spinal artery* and may be responsible for the nutrition of the 2 lower thirds of the spinal cord [6,15,20,24].

Inasmuch as the irrigation comes from more than one source, the vascularization of the spinal cord presents transition zones that may be more vulnerable to lesions [3,26,27]. The upper thoracic segments (T1 through T4) is a transition zone that mainly depends on the radicular branches of the posterior intercostal arteries and secondarily on branches originating from the anterior spinal artery. If one or more of the nearby intercostal vessels are damaged, these segments cannot be maintained by the small branches originating from the anterior spinal artery [26,27]. Our option for the T6-T7 or T7-T8 spaces on the right aimed to avoid transition zone and to reduce the possibility of finding the *magna radicular artery*. Bilateral involvement shown by clinical and EMG findings allowed the choice of the right side of the body. During resection of the dorsal root ganglion, the spinal branch of the corresponding intercostal artery may be coagulated and sectioned, a fact that may impair spinal cord circulation. Some dissections in cadavers have shown that it is possible to detect small arteries along the dural sheath of the spinal nerve that apparently end in the dural leaf toward the peridural space above and below the intervertebral foramen of the thoracic region. Even using the surgical microscope, it may be difficult to properly identify the radiculomedullary artery during surgery. The easy identification of a radiculomedullary artery should serve as an alert because it is probable that its damage could impair the spinal cord circulation. The opening of the dural sheath allows better identification and isolation of the radiculomedullary artery even if the ganglion cannot be separated from the ventral root.

## 5. Conclusions

Ganglionectomy of dorsal root ganglion using microsurgical techniques for diagnosis of the NISPs was effective and safe. Although safe, en bloc resection of the proximal dural cuff was not an effective technique for this purpose.

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## Commentary

Colli et al describe and propose the use of dorsal root ganglionectomy as an important method for diagnosis of sensory neuropathies. This technique is important in view of its clinical applicability. The neurosurgeons have used dorsal root ganglionectomy in different fields, especially for treatment of pain [1-3]. However, the authors in the current article present this unique technique in a new and different field of neurosurgery: for diagnosis. They have thus put forward a safe and effective solution to the diagnostic problem of inflammatory sensory ganglioneuropathies. This method also prevents the adverse effects observed with immunosuppressive therapies. For these reasons, the presented technique is important and interesting. We believe that this method has usefulness not only in neurosurgery practice but in general medicine practice as well.

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The authors have done meaningful analyses of their experience with biopsy of sensory ganglia in cases with the suggestive diagnosis of polyganglioneuropathies. Through their analyses, they describe important adaptations of techniques previously described for dorsal root ganglionectomy to their purpose of biopsy. They used modern microsurgical technique to improve the yield of isolation of the sensory ganglion, thereby enhancing the possibility of histopathological diagnosis. The figures are illustrative and adequate, as is their detailed description of the vascular anatomy of the spinal cord. They describe the anatomy with the intention of avoiding pitfalls of the ganglionectomy technique proposed. This is an important technique to be available to those practicing general neurosurgery and want to collaborate closely with neurologists treating polyneuropathies.

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