



Malformazione di CHIARI e SIRINGOMIELIA

diagnosi, trattamento, vita quotidiana

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Cosa chiedere alla neuromodulazione

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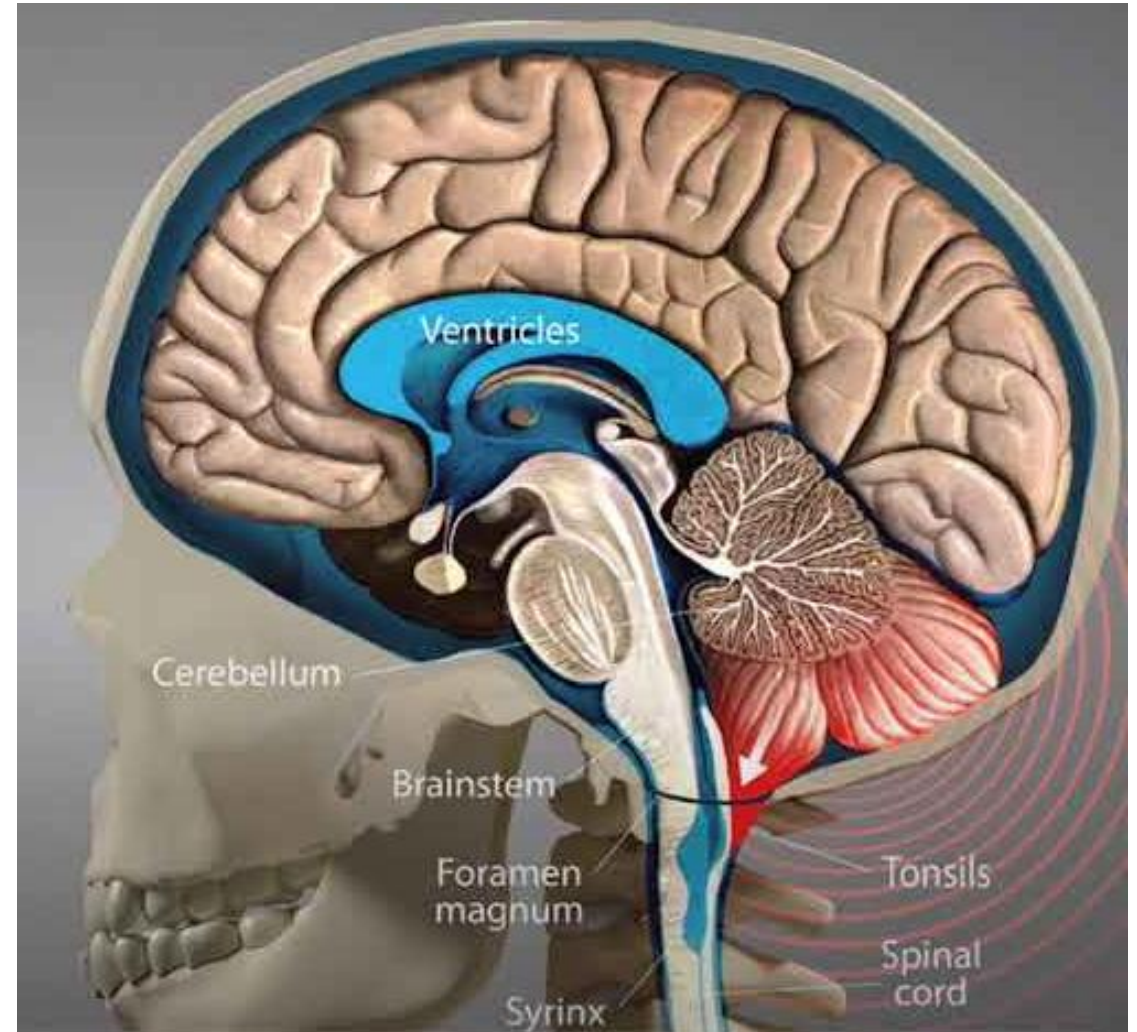
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Chiari Malformation

Chiari malformation is primarily a disorder of the craniovertebral junction characterized by displacement of hindbrain structures, especially the cerebellar tonsils, through the foramen magnum



Chiari Malformation

Type	Key Anatomical Feature	Clinical Associations
Type 0	No tonsillar herniation	Syringomyelia; symptoms may improve after decompression
Type I	Tonsillar herniation ≥ 5 mm below foramen magnum	Headache, syringomyelia; adolescence/adults
Type 1.5	Tonsillar herniation + brainstem descent	More severe Chiari I variant; frequent syrinx
Type II	Herniation of vermis, tonsils, brainstem	Myelomeningocele; hydrocephalus
Type III	Hindbrain herniation into occipital/cervical encephalocele	Severe neurological deficits
Type IV	Cerebellar hypoplasia or aplasia	Rare congenital cerebellar development anomaly

Etiologic Classification of Chiari Malformation

Primary (Developmental)

Posterior fossa
hypoplasia

Hindbrain
overcrowding

Tonsillar herniation

Secondary (Acquired)

CSF
hypovolemia

Hydrocephalus

Mass lesions

Neural Tube Defect– Related

Chiari II

Myelomeningocele

Brainstem
herniation

Syndromic / Complex

Ehlers-Danlos

Basilar invagination

Craniocervical
instability

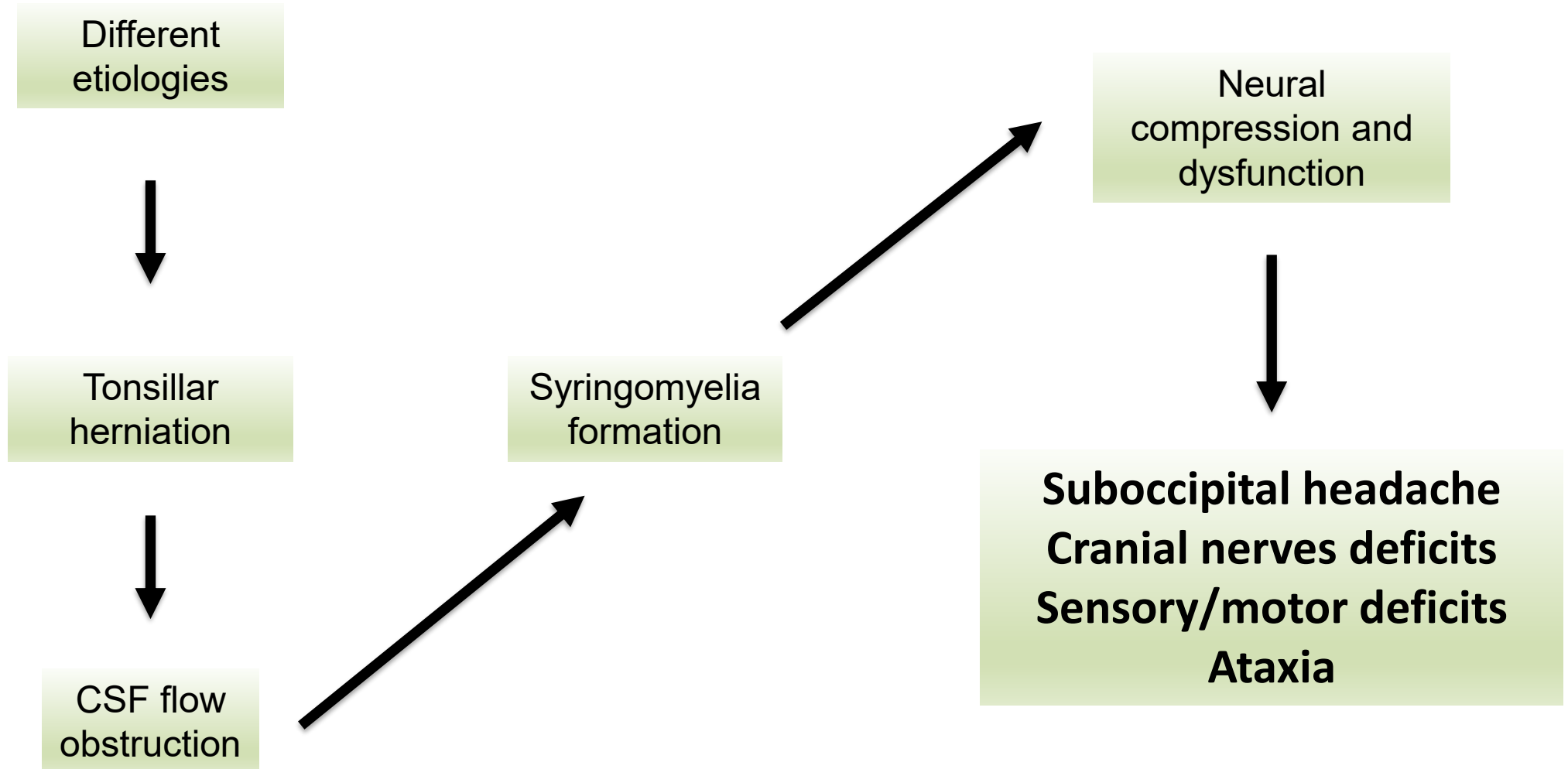
Chiari 0

No tonsillar
herniation

Syringomyelia

CSF flow
abnormality

Chiari Malformation

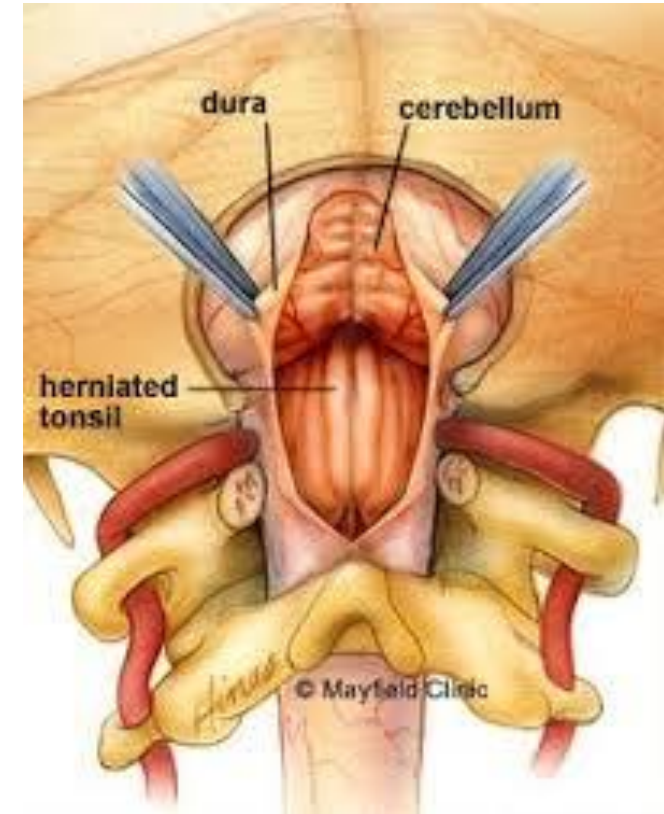


Posterior Fossa Decompression

Standard surgical treatment:

- Suboccipital craniectomy
- C1 laminectomy
- Duraplasty

Goal: restore CSF flow

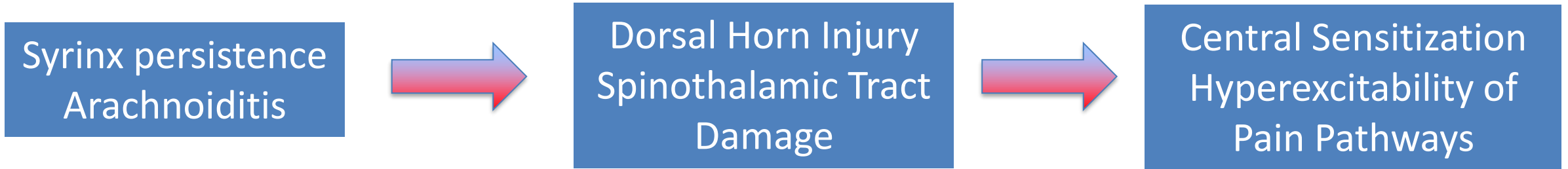


Limitations of Surgery

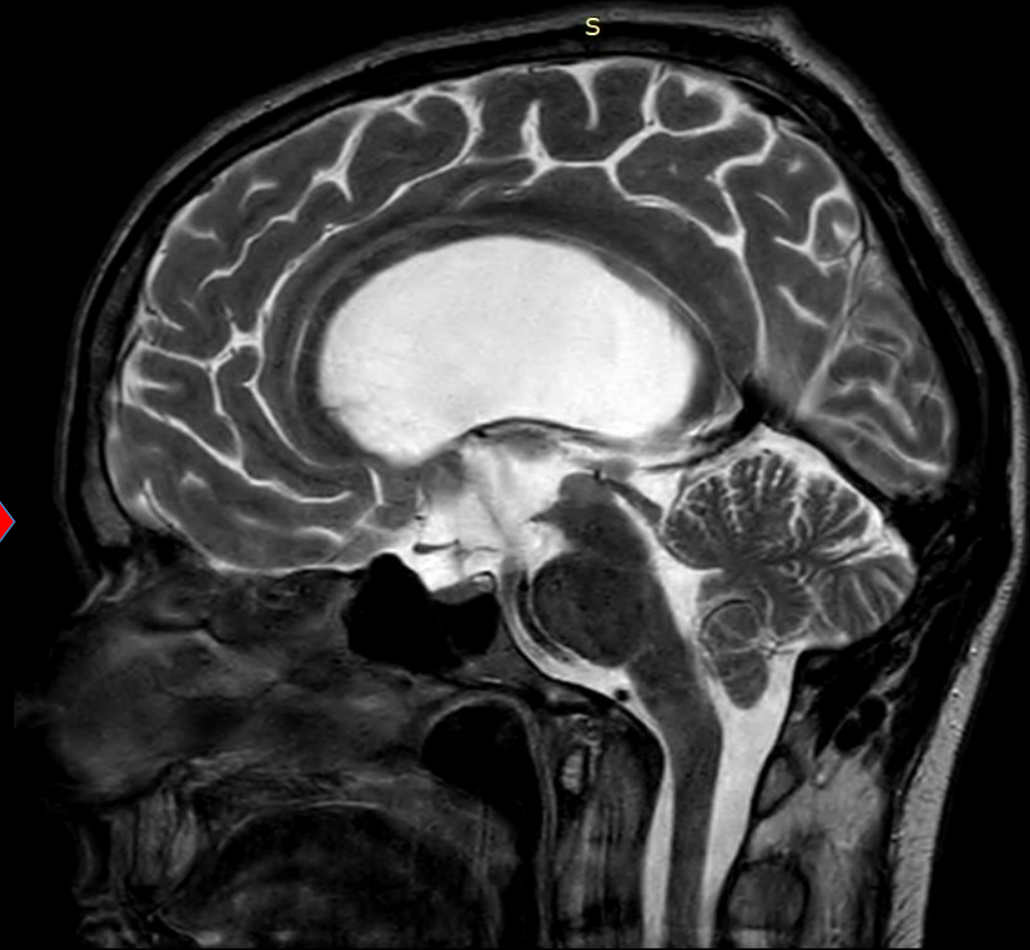
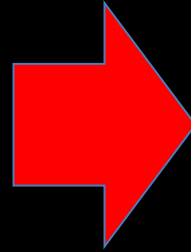
Symptoms may persist despite decompression:

- Headache
- Sensory disturbances
- **Chronic /Neuropathic pain**

Mechanism of Neuropathic Pain in Chiari/Syringomyelia



M 30 year old
Spastic tetraparesis



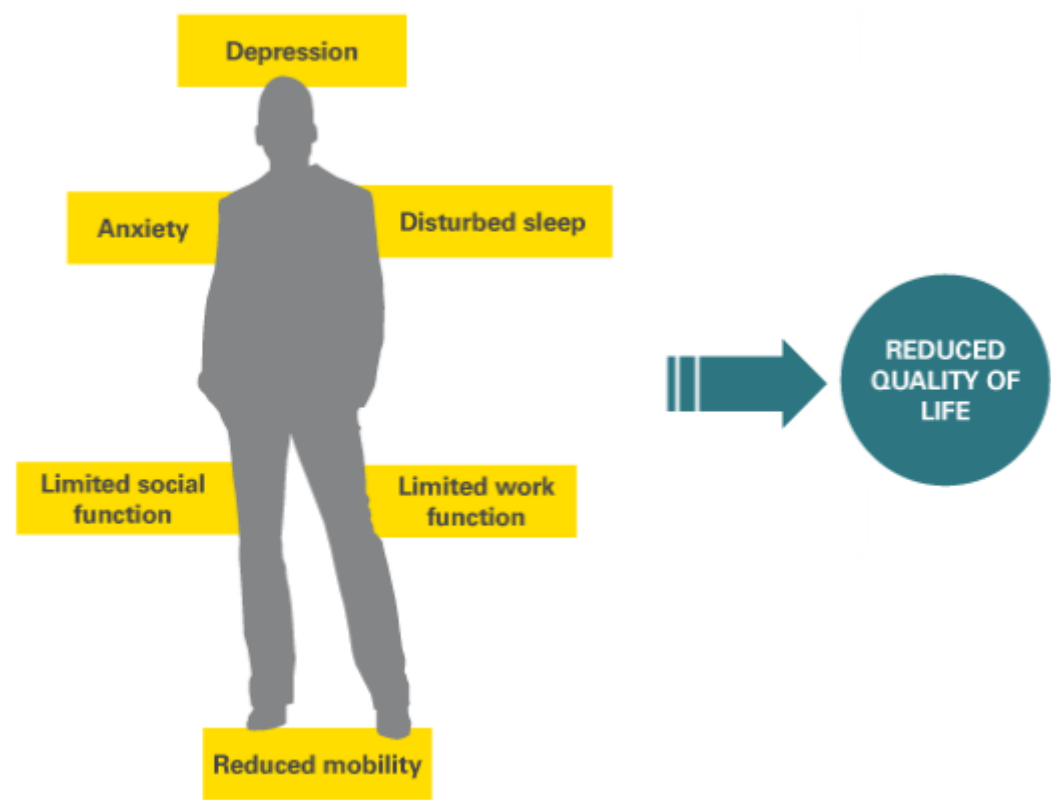
Pain

- Pain is an **unpleasant sensory and emotional experience** associated with, or resembling that associated with, actual or potential tissue damage. Often, pain serves as a symptom **warning of a medical condition or injury**. In these cases, treatment of the underlying medical condition is crucial and may resolve the pain.
- However, **pain may persist despite successful management of the condition** that initially caused it, or because the underlying medical condition cannot be treated successfully.
- **Chronic pain is pain that persists or recurs for longer than 3 months**. Such pain often becomes the sole or **predominant clinical problem** in some patients. As such it may warrant specific diagnostic evaluation, therapy and rehabilitation.
- Chronic pain is a frequent condition, affecting an estimated 20% of people worldwide. It is multifactorial: biological, psychological and social factors contribute to the pain syndrome

Acute pain vs chronic pain

Characteristic	Acute Pain	Chronic Pain
Cause	Generally known	Often unknown
Duration	Short, well-characterized	Persists after healing, ≥3 months
Treatment	Resolution of underlying cause, usually self-limited	Underlying cause and pain disorder; outcome is often pain control, not cure

The burden of chronic pain



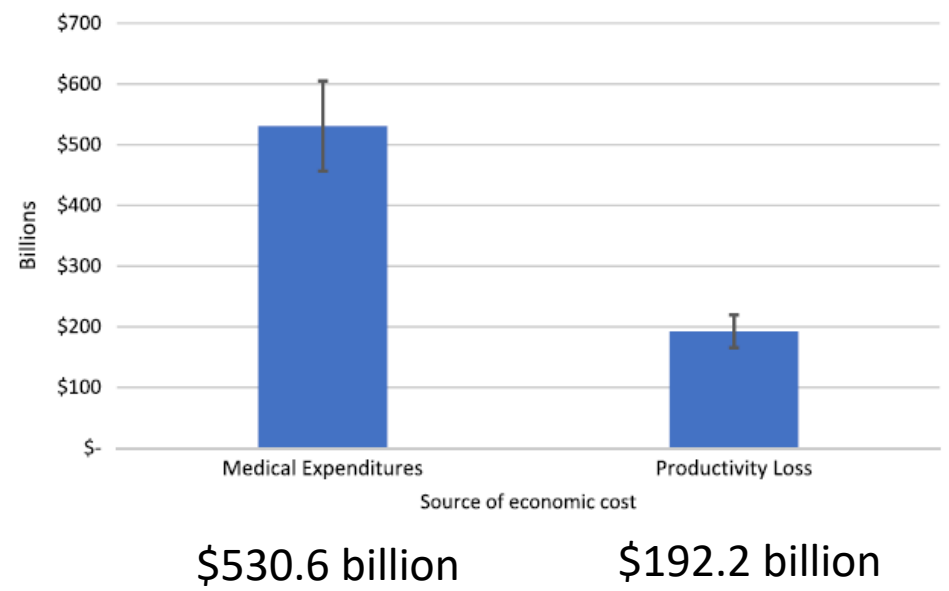
> [Med Care](#). 2025 Sep 1;63(9):679-685. doi: 10.1097/MLR.0000000000002181. Epub 2025 Jul 3.

Economic Costs of Chronic Pain–United States, 2021

Gery P Guy Jr¹, Gabrielle F Miller², Jaswinder K Legha¹, S Michaela Rikard¹, Andrea E Strahan¹, Christina Mikosz¹, Curtis S Florence²

Affiliations + expand
PMID: 40730349 DOI: [10.1097/MLR.0000000000002181](#)

65.8 million persons had chronic non-cancer pain-associated conditions



DN4 – QUESTIONNAIRE

To estimate the probability of neuropathic pain, please answer yes or no for each item of the following four questions.

INTERVIEW OF THE PATIENT

QUESTION 1:

Does the pain have one or more of the following characteristics? YES NO

Burning	☐	☐
Painful cold	☐	☐
Electric shocks	☐	☐

QUESTION 2:

Is the pain associated with one or more of the following symptoms in the same area? YES NO

Score > 4 is indicative of neuropathic pain

EXAMINATION OF THE PATIENT

QUESTION 3:

Is the pain located in an area where the physical examination may reveal one or more of the following characteristics? YES NO

Hypoesthesia to touch	☐	☐
Hypoesthesia to pinprick	☐	☐

QUESTION 4:

In the painful area, can the pain be caused or increased by: YES NO

Brushing?	☐	☐
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YES = 1 point
NO = 0 points

Patient's Score: /10

Neuropathic pain medical treatment

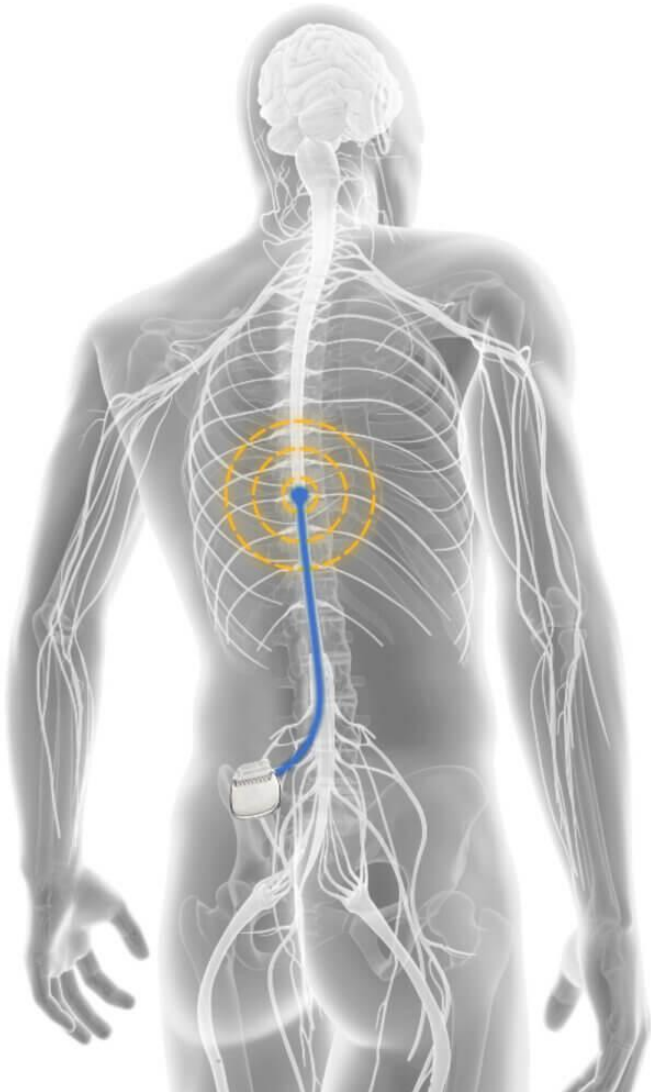
Table 1. Pharmacotherapy for neuropathic pain.

		Drugs	Dose range	Adverse effect
First-line therapy	Gabapentinoids	Gabapentin	150–600 mg/day	Lethargy, vertigo, peripheral swelling, blurred vision
		Pregabalin	300–3600 mg/day	Lethargy, vertigo, peripheral swelling, increased body weight
	Tricyclic antidepressants (TCAs)	Amitriptyline	10–150 mg/day	Anticholinergic effects, QT prolongation (arrhythmia), suicide risk, urinary retention
	Serotonin–norepinephrine reuptake inhibitors (SNRI)	Duloxetine	20–120 mg/day	Nausea, lethargy, constipation, ataxia, dry mouth
Venlafaxine		150–225 mg/day	Nausea, vertigo, lethargy, hyperhidrosis, hypertension	
Second-line therapy	Opioids	Tramadol	25–400 mg/day	Nausea/vomiting, constipation, lethargy, seizures, ataxia
		Tapentadol	50–600 mg/day	Nausea/vomiting, constipation, lethargy, seizures, ataxia
	Topical treatment	Lidocaine	5% patches or gel	Local erythema, itching and rash
		Capsaicin	8% patches	Pain, erythema, itching; rare cases of high blood pressure
Third-line therapy	Strong opioids	Morphine	10–120 mg/day	Nausea, vomiting, constipation, dizziness and lethargy
		Oxycodone	10–120 mg/day	Nausea/vomiting, constipation, lethargy, respiratory control
	Neurotoxin	Botulinum toxin	25–300 U BTX-A 0.9% saline	Pain at injection site

Spinal cord stimulation

Spinal cord stimulation (SCS) is a neuromodulation technique that delivers therapeutic doses of electrical current to the spinal cord

- Adjustable
- Nonlesional
- Reversible



Common SCS indications

Failed back surgery
syndrome
(PSPS type 1)

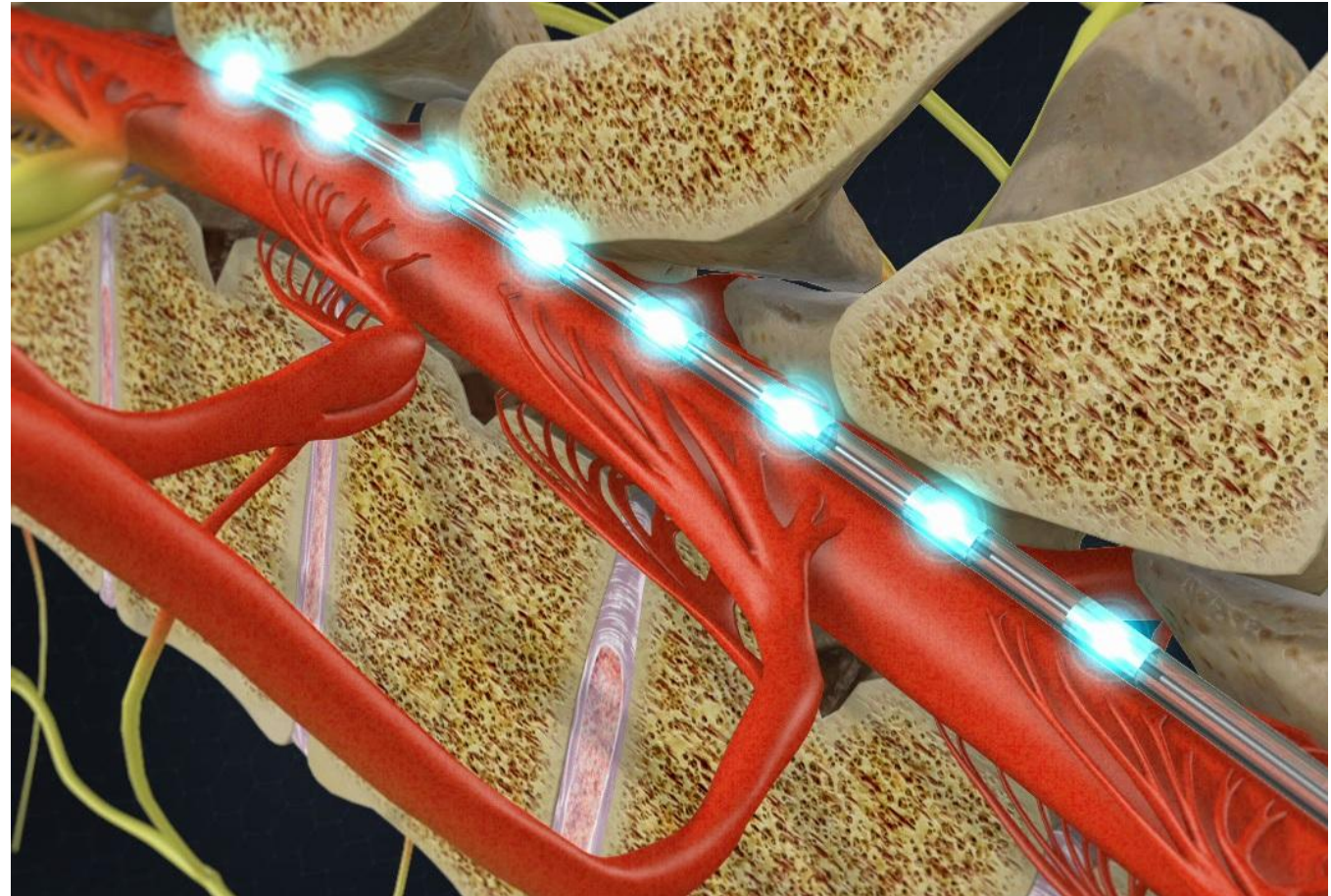
Complex regional
pain syndrome
(CRPS)

Diabetic
neuropathy and
other neuropathies

Angina pectoris
and peripheral
vascular disease

Chronic
radiculopathy

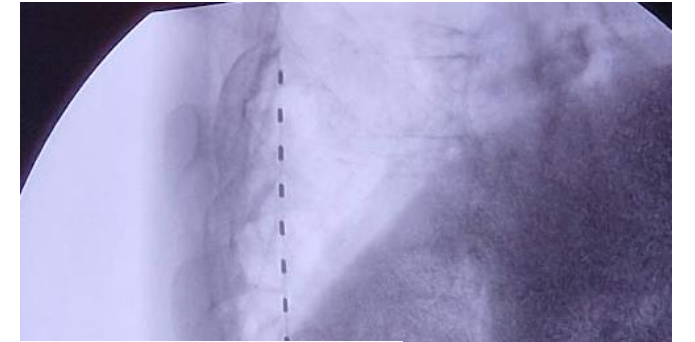
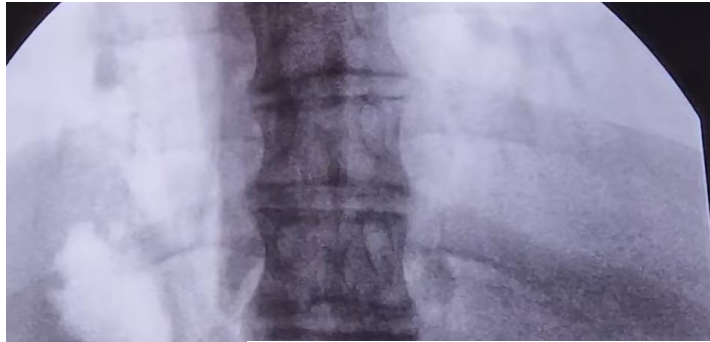
virgin back
(PSPS type II)



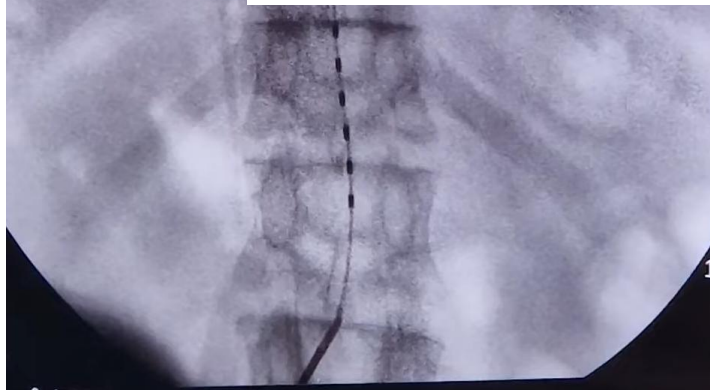
Surgical technique

- Two steps
 - SCS trial (1 month)
 - SCS definitive implantation
- Both in local anesthesia and slight sedation





Intraoperative neurostimulation for paresthesia coverage



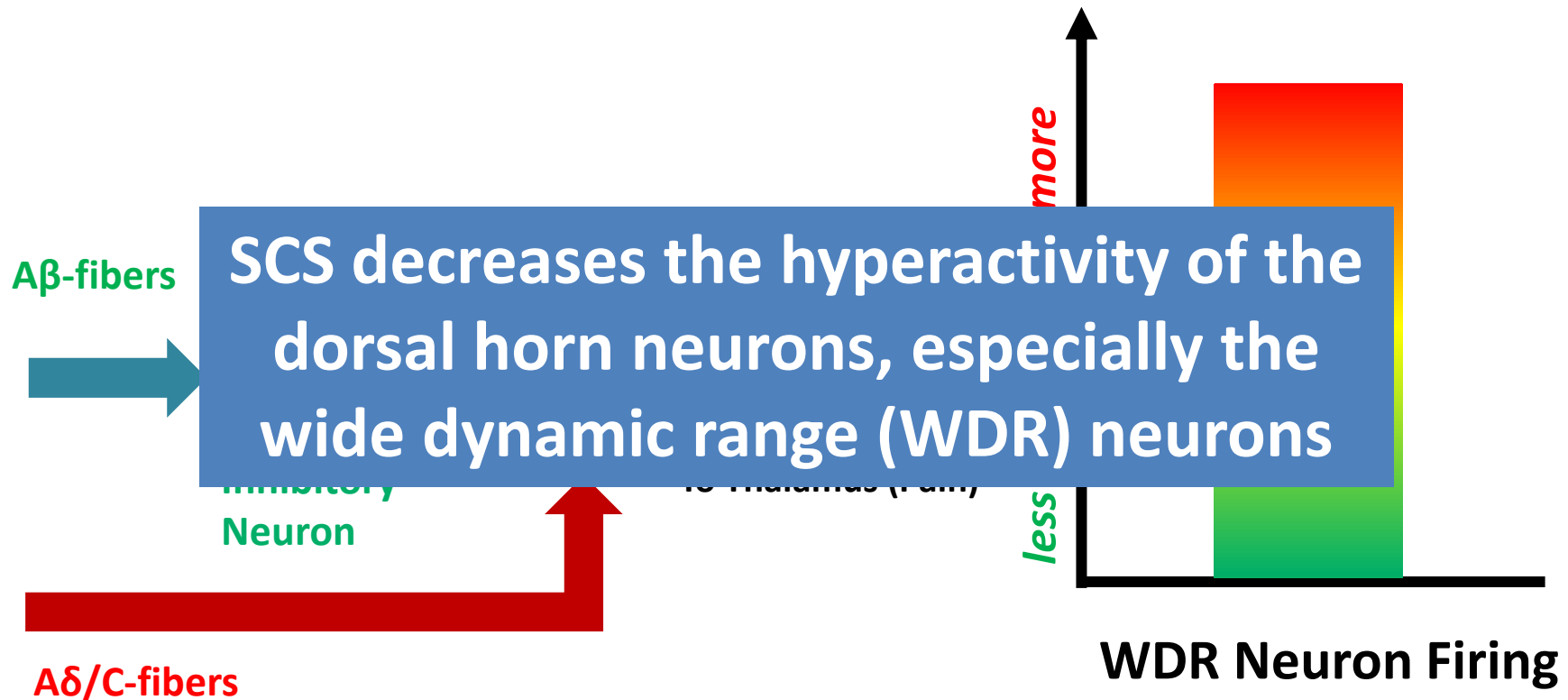
General principles of spinal cord stimulation

Gate control theory of pain (Melzack and Wall)

- In 1959 identification of 2 types of fibers for the transfer of sensory stimuli
- new understanding of pain perception mechanisms
- interaction between the large-diameter fibers that transport tactile, pressure and vibration signals, and the small-diameter fibers associated with pain

Gate control theory of pain (Melzack and Wall)

Stimulation of large-diameter fibers inhibits the response to painful stimuli carried by small-diameter fibers of dorsal column neurons



Mechanisms of spinal cord stimulation

Spinal mechanisms of SCS

involvement of the neurotransmitter GABA and the inhibitory GABAergic interneurons

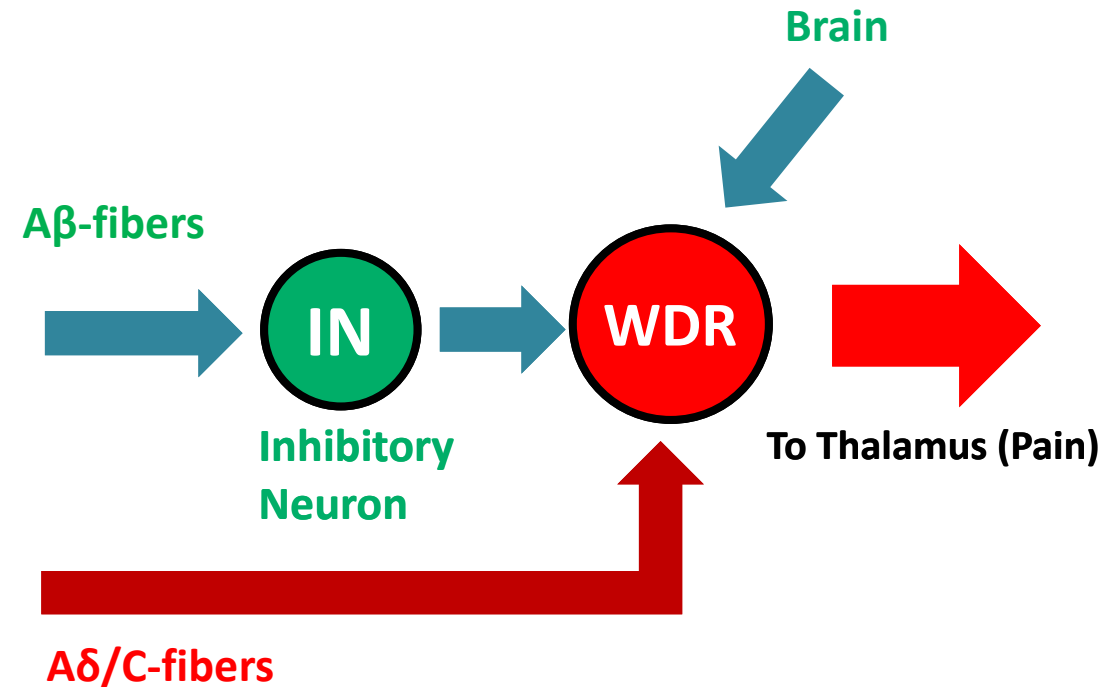
SCS induced GABA release into the extracellular space in the spinal dorsal horn

Supraspinal mechanisms of SCS

The rostral ventromedial medulla (RVM) in the brainstem is involved in the descending modulation of pain via serotonergic input to the dorsal horn

norepinephrine is involved in mediating tonic SCS induced antinociception as well

SCS was shown to increase the synthesis of norepinephrine in the locus coeruleus

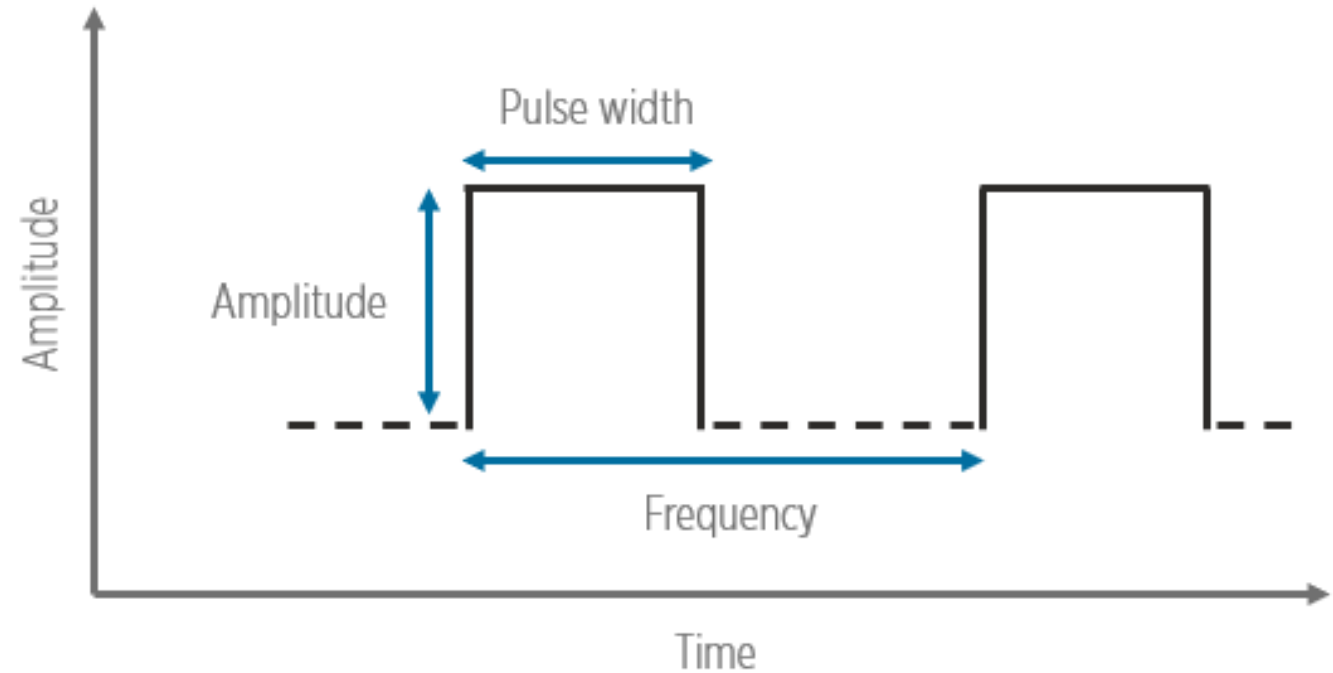


Stimulation parameters

Amplitude: mA

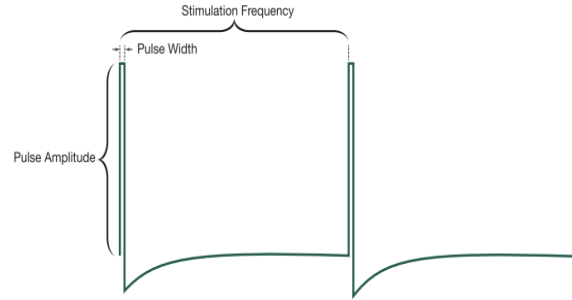
Frequency: Hz

Pulse width: microsec



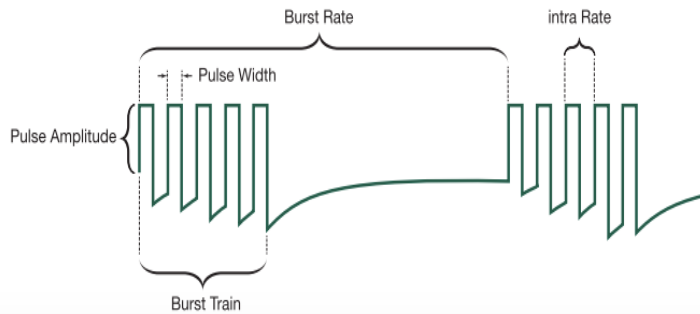
SCS waveforms

Tonic (60-80Hz)



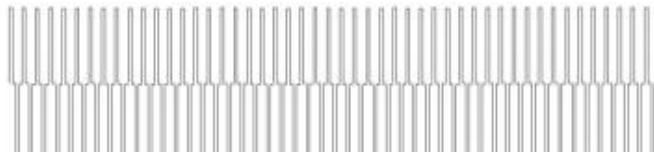
Indirect neural stimulation
through dorsal column

Burst (500Hz)



Indirect neural stimulation
through dorsal column;
modulation of medial and
lateral pathways

High frequency (10KHz)



Direct action on dorsal horn
neurons

- Direct activation of interneurons
- Direct inhibition of projection neurons

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SCS indications

- ▶ PSPS type 1
- ▶ PSPS type 2
- ▶ Complex regional pain syndrome (CRPS)
- ▶ Diabetic neuropathy and other neuropathies
- ▶ Angina pectoris and peripheral vascular disease
- ▶ Chronic radiculopathy
- ▶ Parkinson's disease
- ▶ Spasticity
- ▶ Spinal trauma rehabilitation
- ▶ MS related symptoms
- ▶ Cancer pain

High-Frequency Spinal Cord Stimulation for the Treatment of Spasticity: A Preliminary Case Series

Alessandro Izzo ^{1 2}, Benedetta Burattini ², Renata Martinelli ², Quintino Giorgio D'Alessandris ^{1 2}, Manuela D'Ercole ¹, Maria Filomena Fuggetta ¹, Nicola Montano ^{1 2}

Table 1. Demographic and baseline clinical characteristics of the study cohort. Data are reported as mean ± standard deviation. MS: multiple sclerosis.

Number of patients	6
Sex (M/F)	3/3
Mean age (years)	50.12 ± 9.41
Diagnosis	(1) Relapsing-remitting MS (2) hereditary spastic paraplegia (3) thoracic myelopathy
Disease duration, years	21 ± 18.46
Follow-up (months)	24.32 ± 10.83

Table 2. Clinical outcomes at baseline and follow-up after high-frequency spinal cord stimulation. Values are reported as mean ± SD (median). Statistical comparisons were performed using the Wilcoxon signed-rank test for paired samples.

Parameter	Preoperative	Follow-Up	<i>p</i> -Value	Effect Size <i>d</i> _{Cohen}
VAS	6.16 ± 2.63 (median 7)	3.16 ± 2.71 (median 2)	0.041	1.15
DN4	6.50 ± 3.25 (median 8)	1.83 ± 1.72 (median 2)	0.042	1.71
Ashworth scale	2.66 ± 0.51 (median 3)	2.16 ± 0.75 (median 2)	0.083	0.91
Number of clonuses	6.33 ± 3.83 (median 6)	1.83 ± 2.40 (median 1)	0.065	1.01
Ambulation ability (meters)	21.66 ± 9.83 (median 25)	65.00 ± 73.41 (median 30)	0.179	0.6

Legend: Visual Analog Scale (VAS); Douleur Neuropathique 4 (DN4). *p* < 0.05 significant. Cohen’s *d*: effect size (0.2 small, 0.5 moderate, ≥0.8 large).

Published Neuromodulation Cases in Chiari Malformation

Case	Authors	Age/sex	Cause of syrinx	Range of syrinx	Range of pain	Procedure for lead implantation	Time from surgical intervention to SCS application	Time from onset of pain to SCS application	Improvement of pain	Adverse events	Post-SCS follow-up	Recurrence of pain
1	Campos et al. [11] (2013)	46/F	N/A	C1 ~ T1	Left cervical area from occipital region down to shoulder	Percutaneous	No surgical intervention	4 years	Yes	None	32 months	No
2	Beyaz et al. [12] (2017)	50/F	Congenital (not in detail)	Thoracic to sacral region	From trunk to bilateral lower extremities	Percutaneous	4 previous surgical interventions, 2 years since last surgical intervention	N/A	Yes	None	6 months	No
3	Lu et al. [13] (2022)	68/F	N/A	T1 ~ T8	Left chest, back and upper arm	N/A	No surgical intervention	3 years	Yes	None	N/A	No
4	Lu et al. [12] (2022)	41/F	N/A	Medulla oblongata ~ T3	Headache and back pain accompanied by numbness of right upper limb	N/A	Few years	5 years	Yes	None	12 months	No
5	Schatmeyer et al. [14] (2022)	14/F	Chiari malformation type 1 with syringomyelia	Cervicothoracic region	Left arm	Surgical implantation with laminectomy	N/A	Several months	Yes	None	8 months	No
6	Present case	76/F	Chiari malformation type 1 with syringomyelia	C4 ~ T4	Bilateral finger, upper arm, and chest-back	Percutaneous	3 years	16 years	Yes	None	12 months	No

Conclusions

- Central neuropathic pain can be a disabling complication of Chiari malformation/syringomyelia, often persisting despite adequate surgical decompression
- Always investigate the underlying cause of Chiari malformation/syringomyelia
- SCS represents a consolidated neuromodulation strategy for refractory neuropathic pain and post-surgical pain syndromes
- Current evidence in Chiari malformation/syringomyelia is limited but reported outcomes show:
 - significant pain reduction
 - improvement in quality of life
 - reduction in analgesic use