

THE CAUCASUS JOURNAL OF MEDICAL & PSYCHOLOGICAL SCIENCES よる3よしのし るつのののものし ゆう ちしのさのののし るつのもののようしんつんしょうの КАВКАЗСКИЙ ЖУРНАЛ МЕДИЦИНСКИХ И ПСИХОЛОГИЧЕСКИХ НАУК 高加索医学和心理科学杂志

2023, Volume 1, Number 5-6 eISSN 2720-8788 pISSN 2720-877X

SUPERNUMERARY PHANTOM PAIN: FROM MECHANISMS TO TREATMENT

Burliyat A. Abusueva¹, Kseniia B. Manysheva¹

The article is devoted to the supernumerary phantom pain. It is described as pain in a supernumerary phantom limb. This is a very rare phenomenon with only sporadic and brief descriptions in the literature. However, this syndrome brings great suffering to the patient not only due to the feeling of pain, but also due to the complexity of its description and the difficulty of understanding by the doctor. For simplicity, patients draw a supernumerary limb and mark the location of pain on it. The pathogenesis of supernumerary phantom pain is not completely clear. Therefore, the description of each case is important for clinical practice. The article attempts to summarize the available information about the etiology, pathogenesis, methods of diagnosis and therapy of this syndrome.

pidemiology, etiology, and localization. Supernumerary phantom limb (SPL) is a rare neurological syndrome. SPL refers to the awareness of an illusory extra limb in addition to the real regular limb. Supernumerary phantom pain (SPP) is even rarer phenomenon. Its description is limited to individual clinical cases with only sporadic and brief descriptions in the literature. On average, according to the literature, it occurs in every three patients with SPL [10]. In the described cases, as a rule, it is the result of an ischemic or hemorrhagic stroke [2; 7; 15], less

ABSTRACT ____

often - traumatic injury to the brain or spinal cord [7; 10], brain tumor, multiple sclerosis, epilepsy [8], acute inflammatory demyelinating polyneuropathy [12]. Peripheral nerve and spinal cord lesions may occur SPL. In most of the cases, the SPL appears bilaterally, and it is associated with SPP. The supraspinal lesions include the frontal lobe, temporoparietal lobe, basal ganglia, thalamus, and pons. These lesions may be located either on the right or on the left side, but more frequently, they are found on the right side. The supraspinal lesions are always contralateral to the side of the

KEYWORDS: Supernumerary phantom limb, supernumerary phantom pain, etiopathogenesis, diagnosis, treatment

Cite: Abusueva B.A., Manysheva K.B. Supernumerary phantom pain: from mechanisms to treatment. Cauc J Med & Psychol Sci. 2023; V.1(№5-6): 39-45; doi: 10.61699/cjmps-v1-i5-6-p39-45

¹ Dagestan State Medical University, Department of Nervous Diseases, Medical Genetics and Neurosurgery, Makhachkala, Russian Federation

phantom limb, but SPP is not its obligatory symptom. Supraspinal lesions are less associated with phantom pain due to the absence of pathologic changes on the spinal cord or the peripheral nerves. SPLs caused by the spinal cord or peripheral nerve lesions differed qualitatively from those caused by supraspinal lesions in that they were more associated with pain or other sensory discomfort [8]. It is reported only a few cases occurring in cervical incomplete spinal cord injury (SCI) patients [4].

Phantom sensations accompanying pain is well recognized in patients suffering from SCI, and neuropathic pain syndromes represent one of the most disabling sequels of SCI. Patients with SCI completely lose proprioception, thermoception, and nociception, and under such conditions, they may experience severe pain within the dermatomal boundaries of the deafferent limbs. Neuropathic pain syndromes often persist for many years and are difficult to treat. The sensation of a phantom limb can vary widely, from painless discomfort and uneasiness in the limb to continuous extreme pain. These sensations in SCI often resemble disorders in patients after amputation of limbs [6].

Pathogenesis. The body without an afferent pathway perceives SPL or SPP in addition to the real limbs [3; 8]. Theories about the origin of phantom sensations can be divided into two directions, depending on which mechanisms are more involved in the implementation of the pathological process – peripheral or central. The true mechanisms underlying these phenomena are still unclear. In addition, there are suggestions that they may be the result of a psychological

disorder. It is most likely that both the central and peripheral nervous systems play a role in the formation of SPL, with the central nervous system making a more important contribution [5].

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SPL and phantom limb sensations are considered to be a violation of such normal bodily perceptions. Most often, SPL is the result of brain damage. In this regard, the main theory of its occurrence is associated with the reorganization of the primary somatosensory cortex and the resulting change in the normal scheme of the body. Another theory declares a dissociation between previously formed ideas about sensorimotor representation and a change in the connection between the brain and a paralyzed limb because of a pathological process [4].

The body schema undergoes constant modification through the integration of various sensory inputs and motor outputs. Deafferentation in cerebral lesions causes a perceptual disturbance between the internal representation of the body and sensory inputs from the ascending pathways, leading to dissociation of body images [10].

These illusory phenomena may be caused by a loss of afferent information due to sensory disturbances and an inability to properly integrate multimodal and motor information to create a normal self-location experience. Just as the destruction of sensory roots often leads to the phenomenological experience of an abnormal limb, the complete loss of sensory and proprioceptive sensitivity can reprogram an innate body image into an erroneous one with the creation of an illusory limb. In amputees and SCI patients, the ability to control limb movements is physically or neurologically

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lost. This discrepancy between the motor command of the cortex and the lack of sensory feedback from the peripheral nervous system makes it difficult to distinguish expected movements of real limbs from unexpected movements of illusory limbs. Another mechanism explaining SPL in SCI is the reorganization of subcortical brain structures after SCI [4].

There is an assumption about the mechanism of reverberation of the corticothalamic circuits, which store sensory memories of our internal representations. These representations can be replaced during deafferentation, resulting in the formation of a phantom. At the same time, abnormal activity of the thalamocortical closed loop may underlie phantom phenomena. During the movement of the SPL, an increased signal is formed, depending on the level of blood oxygenation. At the same time, undamaged premotor zones, which translate intention into action, can lead to the formation of a phantom under conditions of deafferentation, which worsens the correction of the discrepancy between expected and real movements [12].

Methods. Functional magnetic resonance imaging (fMRI) is most commonly used to study SPL. SPL movements are associated with increased fMRI responses in motor areas and the bilateral basal ganglia-thalamus [9; 13].

At the same time, the role of the visual system is great in identifying this perceptual discrepancy in conditions of intact polymodal perceptual interaction. It has been shown that in some patients, vision can cancel not only the fact of feeling a phantom limb, but also sensory sensations inherent in phantoms, such as pain [11].

In the absence of brain disease, computed tomography (CT) and MRI scan results, as well as electroencephalography (EEG), may be normal. Neuropsychological assessments usually cannot reveal any deviation from the norm [1; 14].

Virtual reality, robotics, neuroprosthetics and other methods will certainly help to determine the mechanisms of multisensory and cerebral bodily representations [7].

Treatment. Phantom limb pain is a complex condition, and standardized treatment protocols for SPL have not been developed. Phantom limb pain is more difficult to treat than regular pain. One of the important directions in the treatment of such patients is the use of the visual system in identifying this perceptual discrepancy [4].

At the same time, some SPLs are not visible and are not affected by either the visual feedback of real limbs or direct touch on a real limb. However, in some patients, SPL or associated sensory experiences such as pain can be managed with visual feedback and tactile stimulation of a real limb.

The neuropathic component of pain in paralyzed and SPL patients can be reduced with medication, but stimulation has been shown to be effective in reducing the intensity of SPL. Visual-tactile stimulation treatment may be effective in improving SPL intensity in patients. The advantage of this therapeutic tool is that it is convenient to use and carry out in patients with any localization of the pathological process, for example, with an injury to the cervical spine measures

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for the treatment of SPL [4].

The duration of supernumerary phantoms is clinically unpredictable (persistence varies from months to several years). The temporal relationship between neurological recovery and the eventual cessation of supernumerary phantoms does not really indicate a direct causal relationship. Based on the postulates that the development of phantom pain may be based on maladaptive processes in the neuromatrix, various strategies are used to normalize the cortical organization. Visuomotor training has been shown to reduce pain in most patients with SCI. When using this method, it should be borne in mind that its use in some cases can cause pain or non-pain phantom sensations in patients who did not have them before, and also causes aggravation of the existing pain syndrome. Therefore, it should be borne in mind that such methods are not without serious side effects [6].

The main giving hope therapies for phantom limb pain are mirror therapy and virtual visual feedback (VVF). They are effective treatments for pain such as phantom limb pain or type 1 complex regional pain syndrome caused by sensory motor incongruence. In addition, VVF is effective in the treatment of SPP caused by SCI. The use of these therapies results in the compensation of lost somatic information, which may in turn lead to the restoration of motor command of the central nervous system and the unification of the sensory-motor loop in sensory-deprived limbs to alleviate SPP [5].

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Some studies have shown that phantom pain can be alleviated with local anesthesia in some patients, but their use is significantly limited due to the instability of the effect [10].

This article attempts to summarize all known information about supernumerary phantom pain, its most common causes and localization options, known data on the pathogenesis of this syndrome, modern diagnostic methods and its treatment. Of course, as the number of cases of this syndrome increases, such reviews will be supplemented with new information.

References

- Bakheit AM, Roundhill S. Supernumerary phantom limb after stroke. Postgrad Med J. 2005;81(953):e2. doi:10.1136/pgmj.2004.027086
- Bourlon C., Urbanski M., Quentin R., et al. Cortico-thalamic disconnection in a patient with supernumerary phantom limb. Experimental Brain Research. 2017;235(10):3163– 3174. doi: 10.1007/s00221-017-5044-y.
- Brugger P. Supernumerary phantoms: a comment on Grossi, et al.'s (2002). Spare thoughts on spare limbs. Perceptual and Motor Skills. 2003;97(1):3–10. doi: 10.2466/ pms.2003.97.1.3.
- Choi J.Y., Kim H.I., Lee K.C., Han Z.A. Atypical supernumerary phantom limb and phantom limb pain in a patient with spinal cord injury: case report. Annals of Rehabilitation Medicine. 2013;37(6):901–906. doi:

10.5535/arm.2013.37.6.901.

- Cipriani G., Picchi L., Vedovello M., et al. The phantom and the supernumerary phantom limb: historical review and new case. Neuroscience Bulletin. 2011; 27(6):359–365. doi: 10.1007/s12264-011-1737-6.
- Curt A., Yengue C.N., Hilti L.M., Brugger P. Supernumerary phantom limbs in spinal cord injury. Spinal Cord. 2011; 49(5):588–595. doi: 10.1038/sc.2010.143.
- Dieguez S., Lopez C. The bodily self: Insights from clinical and experimental research. Annals of Physical and Rehabilitation Medicine. 2017; 60(3):198–207. doi: 10.1016/j.rehab.2016.04.007.
- Katayama O., Iki H., Sawa S., et al. The effect of virtual visual feedback on supernumerary phantom limb pain in a patient with high cervical cord injury: a single-case design study. Neurocase. 2015; 21(6):786–792. doi: 10.1080/13554794.2015.1011664.
- Khateb A., Simon S.R., Dieguez S., et al. Seeing the phantom: a functional magnetic resonance imaging study of a supernumerary phantom limb. Annals of Neurology. 2009; 65(6):698–705. doi: 10.1002/ ana.21647.
- Kim H.R., Han J.Y., Park Y.H., et al. Supernumerary phantom limb in a patient with basal ganglia hemorrhage – a case report and review of the literature. BMC Neurology. 2017; 17(1):180. doi: 10.1186/ s12883-017-0962-7.

- McGonigle D.J., Hänninen R., Salenius S., et al. Whose arm is it anyway? An fMRI case study of supernumerary phantom limb. Brain. 2002; 125(6):1265–1274. doi: 10.1093/ brain/awf139.
- Melinyshyn A.N., Gofton T.E., Schulz V. Supernumerary phantom limbs in ICU patients with acute inflammatory demyelinating polyneuropathy. Neurology. 2016; 86(18):1726–1728. doi: 10.1212/ WNL.00000000002541.
- Staub F., Bogousslavsky J., Maeder P., et al. Intentional motor phantom limb syndrome. Neurology. 2006; 67(12):2140–2146. doi: 10.1212/01. wnl.0000249135.78905.75.
- Yamada M., Sasahara Y., Seto M., et al. Intentional Supernumerary Motor Phantom Limb after Right Cerebral Stroke: A Case Report. Case Reports in Neurology. 2021; 13(1):251–258. doi: 10.1159/000513302.
- Yoo S.D., Kim D.H., Jeong Y.S., et al. Atypical supernumerary phantom limb and phantom limb pain in two patients with pontine hemorrhage. Journal of Korean Medical Science. 2011; 26(6):844–847. doi: 10.3346/ jkms.2011.26.6.844.

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<u>РЕЗЮМЕ</u>

БОЛЬ В ДОПОЛНИТЕЛЬНОЙ КОНЕЧНОСТИ ПРИ ПСЕВДОПОЛИМЕЛИИ: ОТ МЕХАНИЗМОВ К ЛЕЧЕНИЮ

Бурлият А. Абусуева¹, Ксения Б. Манышева¹

¹Дагестанский государственный медицинский университет, Кафедра нервных болезней, медицинской генетики и нейрохирургии, г. Махачкала, Российская Федерация

Статья посвящена боли в дополнительной конечности при псевдополимелии. Это очень редкое явление, для которого в литературе имеются лишь краткие описания. Однако этот синдром приносит серьезные страдания пациенту не только из-за ощущения боли, но и ввиду сложности его описания и трудности понимания врачом. Для простоты объяснения пациенты обычно рисуют на бумаге дополнительную конечность и отмечают на ней локализацию боли. Патогенез такого варианта боли до конца не ясен, поэтому описание каждого отдельного случая важно для клинической практики. В статье предпринята попытка обобщить имеющиеся сведения об этиологии, патогенезе, методах диагностики и лечении этого синдрома.сти кровотока в ипсилатеральной средней мозговой артерии на 20-35% в случае значительной извитости или перегиба сонной артерии.

Ключевые слова: сверхштатная фантомная конечность, сверхштатная фантомная боль, этиопатогенез, диагностика, терапия

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დამატებითი კიდურის ფანტომური ტკივილი ფსევდოპოლიმელიის დროს: მექანიზმებიდან მკურნალობამდე

ბურლიატი ა. აბუსუევა¹, ქსენია ბ. მანიშევა¹

¹დაღესტნის სახელმწიფო სამედიცინო უნივერსიტეტი, ნერვული დაავადებების, სამედიცინო გენეტიკის და ნეიროქირურგიის კათედრა, მახაჩკალა, რუსეთის ფედერაცია

სტატია შეეხება ფსევდოპოლიმელიის დროს დამატებითი კიდურის ფანტომურ ტკივილს, რაც საკმაოდ იშვიათი ფენომენია, რომლის შესახებაც სამედიცინო ლიტერატურაში მხოლოდ სპორადული და მცირე ინფორმაციის მოძიებაა შესაძლებელი. მიუხედავად იშვიათობისა, ამ სინდრომის არსებობისას პაციენტი იტანჯება არა მხოლოდ ტკივილის შეგრძნების გამო, არამედ ამ შეგრძნების აღწერისა და ექიმისთვის ამის გაგებინების სირთულის გამო. ტკივილის გადმოცემის გასაადვილებლად, პაციენტები, როგორც წესი, ქაღალდზე ხატავენ დამატებით კიდურს და მიუთითებენ ტკივილის ადგილს. ამ ტიპის ტკივილის პათოგენეზი ბოლომდე შესწავლილი არ არის. ამიტომ, თითოეული ინდივიდუალური შემთხვევის აღწერა ძალზე მნიშვნელოვანია კლინიკური პრაქტიკისთვის. სტატიაში ვცდილობთ შევაჯამოთ ფანტომური ტკივილის სინდრომის ეტიოლოგიის, პათოგენეზის, დიაგნოსტიკური მეთოდებისა და მკურნალობის შესახებ არსებული ინფორმაცია.

საკვანძო სიტყვები:ფანტომური კიდური, ფანტომური ტკივილი, ეტიოპათოგენეზი, დიაგნოსტიკა, თერაპია