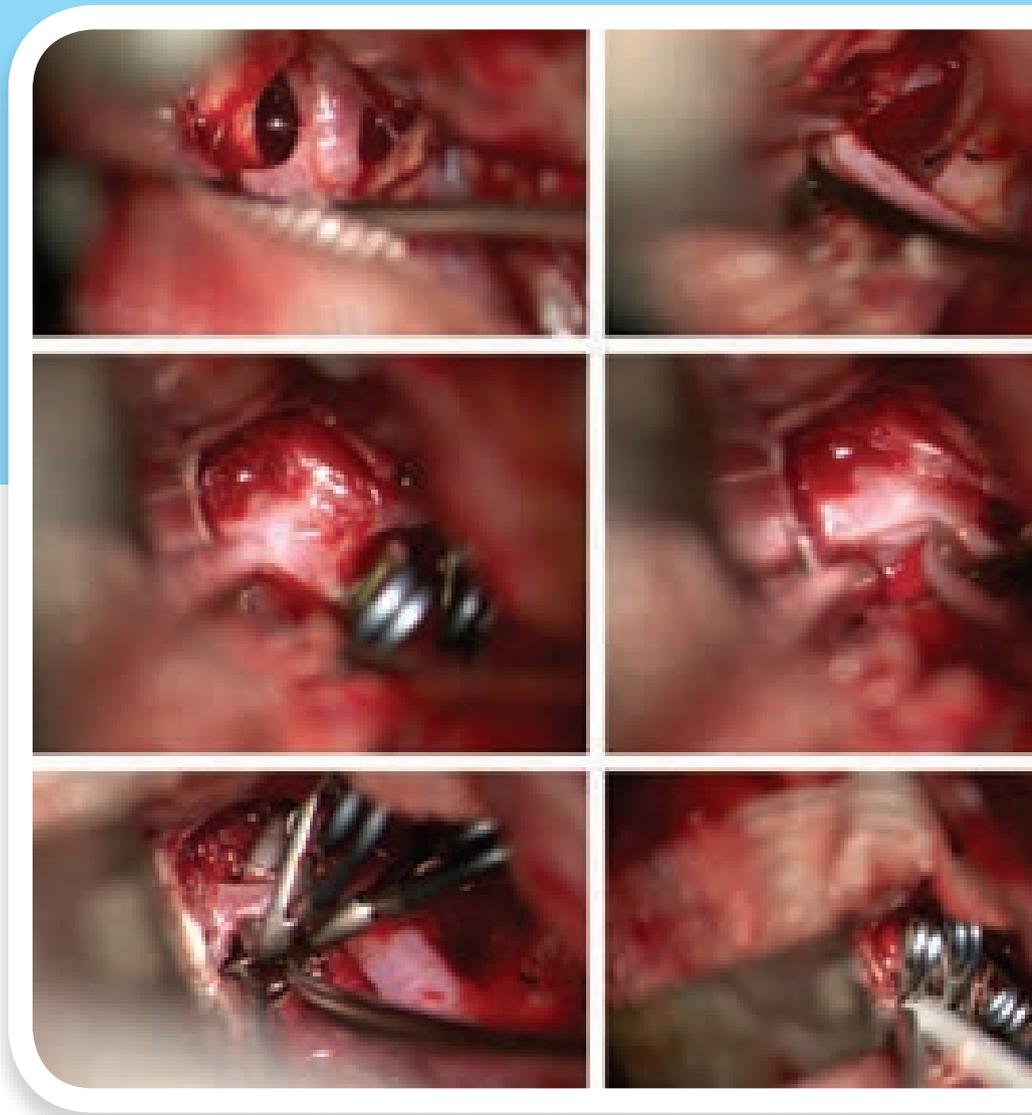


# Brazilian Neurosurgery

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# Epidemiology and Estimated Cost of Surgery for Chronic Subdural Hematoma Conducted by the Unified Health System in Brazil (2008–2016)

## *Epidemiologia e estimativa de custo das cirurgias para hematoma subdural crônico realizadas pelo Sistema Único de Saúde no Brasil (2008–2016)*

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### Abstract

**Introduction** Chronic subdural hematoma (CSH) is one of the most frequent forms of intracranial hemorrhage. It is a collection of encapsulated, well-delimited fluid and/or coagulated blood in several clotting stages located between the dura mater and the arachnoid mater.

**Objective** To describe the epidemiological aspects of CSH described in the database of the Brazilian Unified Health System (SUS, in the Portuguese acronym) regarding admission numbers, hospitalization expenses, health care professional expenses, mortality rate, and death numbers by region from 2008 to the first half of 2016.

**Methods** The present work was performed between August and September 2016 with a review about the epidemiological aspects of CSH in Brazil according to the Informatics Department of the Unified Health System (DATASUS) database, encompassing the period from January 2008 to June 2016, and to scientific papers from the past 10 years which were electronically published at the PubMed, Scielo, and LILACS databases.

**Results** From 2008 to the first half of 2016, the total values were the following: hospital admission authorizations (HAAs). 33,878; hospital expenses, BRL 65,909,429.22; health care professional expenses, BRL 25,158,683.21; deaths, 2,758; and mortality rates ranging from 6.47 to 12.63%.

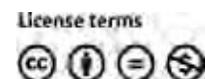
### Keywords

- epidemiology
- chronic subdural hematoma
- cost analysis

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## Resumo

**Conclusion** In spite of the high clinical relevance of CSH, epidemiological studies about this condition are limited. As such, the present paper is an updated approach on CSH, focusing on its epidemiological aspects according to the DATASUS database.

**Introdução** Hematoma subdural crônico (HSDC) é uma das formas mais frequentes de hemorragia intracraniana. É constituído por uma coleção de sangue fluido e/ou coagulado em vários estágios, encapsulada e bem delimitada, localizada entre a dura-máter e a aracnoide-máter.

**Objetivo** Descrever aspectos epidemiológicos do HSDC, coletados no banco de dados do Sistema Único de Saúde (SUS), referentes aos gastos intrahospitalares, remuneração do profissional médico, taxa de mortalidade, e número de óbitos por região, de 2008 ao 1º semestre de 2016.

**Métodos** O presente trabalho foi realizado entre agosto e setembro de 2016, a partir de uma revisão da literatura sobre os aspectos epidemiológicos do HSDC no Brasil, com base no banco de dados do Departamento de Informática do Sistema Único de Saúde (DATASUS), compreendendo o período de janeiro de 2008 a junho de 2016, bem como em artigos científicos dos últimos 10 anos, publicados nos bancos de dados eletrônicos PubMed, Scielo e LILACS.

**Resultados** Dos aspectos analisados no período de 2008 ao 1º semestre de 2016, foram encontrados os seguintes valores totais: autorizações de internação hospitalar (AIHs), 33.878; valor de serviço hospitalar, R\$ 65.909.429,22; valor dos serviços profissionais, R\$ 25.158.683,21; dias de permanência, 304.746; óbitos, 2.758; e taxa de mortalidade, entre 6,47 e 12,63%.

**Conclusão** Embora o HSDC represente um assunto de grande relevância clínica, entende-se que ainda são escassos os estudos epidemiológicos que tratem do tema. Desta forma, o presente artigo traz uma abordagem atualizada para o tema, com enfoque em aspectos epidemiológicos nacionais do HSDC coletados no banco de dados do DATASUS.

## Palavras-chave

- ▶ epidemiologia
- ▶ hematoma subdural crônico
- ▶ análise de gastos

## Introduction

Chronic subdural hematoma (CSH) is one of the most common forms of intracranial hemorrhage. It is a collection of encapsulated, well-delimited fluid and/or coagulated blood in several clotting stages located between the dura-mater and the arachnoid mater.<sup>1,2</sup>

Chronic subdural hematoma is clinically important because, in most cases, its evolution without immediate surgical treatment can lead to high morbidity and mortality. Therefore, early diagnosis and successful surgical drainage are imperative to ensure a complete recovery.<sup>2,3</sup>

Although CSH is a highly frequent and clinically relevant disease, epidemiological data are still limited in Brazil. The present work aims to describe the epidemiological aspects of CSH according to the database of the Brazilian Unified Health System (SUS, in the Portuguese acronym). The following variables were studied from 2008 to the first half of 2016: admission numbers, hospitalization expenses, health care professional expenses, mortality rate, and death numbers.

## Materials and Methods

Information on the surgical treatment of CSH was obtained from code 04.03.01.03.14 between January 2008 and June 2016; this

code is used for issuing hospital admission authorizations (HAAs) at the Informatics Department of the Unified Health System (DATASUS, in the Portuguese acronym) virtual database. The present review evaluates admission numbers, hospitalization expenses, health care professional expenses, mortality rates, and death numbers during the period studied.

Scientific papers from the past 10 years which were electronically published at the PubMed, Scielo and LILACS databases were used in the present review. These papers were retrieved using the following descriptors in Portuguese and in English: *epidemiologia/epidemiology* and *hematoma subdural crônico/chronic subdural hematoma*. Initially, 119 papers were found at PubMed, 3 at Scielo, and 71 at LILACS. However, only 13 papers were used in the present review, since those that did not present epidemiological information on CSH, such as prevalence and incidence, were excluded.

## Results

According to ▶ **Table 1** and ▶ **Fig. 1**, there were 2,389 HAAs in 2008, of which more than half were in the Southeast region (1,284). Meanwhile, the Northern region had the lowest HAA index, with 101 cases. The number of HAAs increased over time, with a total number of 4,885 in 2015, and of 2,453 HAAs in the first half of 2016.

**Table 1** Number of hospital admission authorizations (HAAs) approved per year for the chronic subdural hematoma surgical treatment code

	North	Northeast	Southeast	South	Central-West	Total	Population estimate
2008	101	289	1,284	415	300	2,389	189,600,000
2009	169	479	1,676	515	403	3,242	191,480,630
2010	223	533	1,906	599	471	3,732	190,755,799
2011	193	601	2,060	640	512	4,006	–
2012	207	645	2,127	643	483	4,105	193,946,886
2013	230	747	2,209	722	484	4,392	201,032,714
2014	230	759	2,380	768	537	4,674	202,768,562
2015	213	808	2,540	706	618	4,885	204,450,649
2016*	95	410	1,260	392	296	2,453	206,391,315
Total	1,661	5,271	17,442	5,400	4,104	33,878	–
Average	184.55	585.66	1,938	600	456	–	–

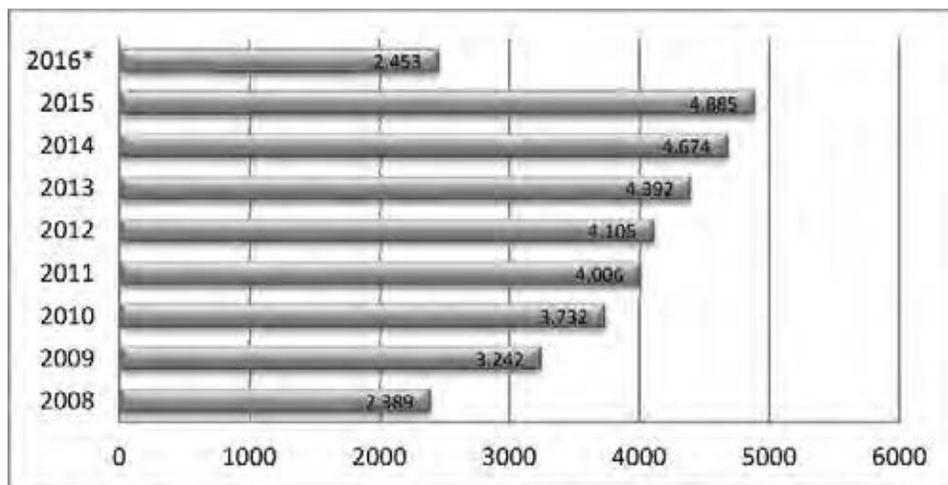
\*Data from 2016 correspond to the first semester. Source: DATASUS/IBGE (<http://tabnet.datasus.gov.br/cgi/tabcgi.exe?sih/cnv/qiuf.def>). Accessed on September 1, 2016.

–**Table 2** shows that hospital expenses also increased over the years. In 2008, the total expense was of BRL 3,803,093.70 and, in 2015, of BRL 10,319,288.77; the highest value was reached in the Southeast region, followed by the South and Central-West regions. In the following years, the Northeast region assumed the 3<sup>rd</sup> position regarding hospital-specific expenses, with BRL 855,176.35 in 2011, and BRL 1,681,269.55 in 2015.

Regarding health care professional expenses, the Southeast region surpassed the other regions, with an amount of BRL 2,121,365.37 in 2015, and of BRL 700,318.46 in 2008. The North region, however, presented lower values compared with the remaining regions, with BRL 188,139.34 in 2015, and BRL 53,762.80 in 2008. Although the expenses with the surgical treatment of CSH were higher in the Southeast region, the highest growth in expenses during the evaluated period occurred at the Northeast region, whose 2015 figures were 4 times higher than those observed in 2008 (–**Table 3**).

Regarding the number of deaths per region, in 2008, the Southeast region presented the highest number, with 104 cases, followed by the South region, with 43, the Northeast region, with 31, the Central-West region, with 20, and the North region, with 2 cases. This proportion was sustained until 2011, when the Northeast region surpassed the South region, with 53 deaths, while the Southeast region continued to present the largest number of deaths, 181. From 2014 to 2015, there was a significant increase in deaths in the Southeast region, with 216 and 218 cases, respectively. It is important to note that the number of deaths mentioned above did not reflect the mortality rate from each region (–**Table 4**).

Regarding the mortality rate (ratio between the number of deaths and the number of approved HAAs, computed as admissions during the period, multiplied by 100), the Northeast region surpassed the other regions in 2008, with 10.73. In 2009, the Southeast came in first place, with 9.19; and the Northeast had the lowest mortality rate, of 6.47. The mortality

**Fig. 1** Number of hospital admission authorizations (HAAs) approved per year for the chronic subdural hematoma surgical treatment code. \*Data from 2016 correspond to the first semester. Source: DATASUS. Accessed on September 1, 2016.

**Table 2** Hospital expenses (in BRL) per year for the chronic subdural hematoma surgical treatment code

	North	Northeast	Southeast	South	Central-West	Total
2008	142,396.23	366,680.83	1,993,721.28	878,814.47	421,480.89	3,803,093.70
2009	288,533.57	605,903.04	2,759,078.36	1,177,276.80	742,875.76	5,573,667.53
2010	399,103.93	689,638.94	3,434,604.02	1,405,519.81	870,073.75	6,798,940.45
2011	356,896.29	855,176.35	3,840,565.12	1,566,255.29	834,296.26	7,453,189.31
2012	421,962.60	1,064,555.36	4,293,906.36	1,580,971.21	811,095.93	8,172,491.46
2013	456,460.13	1,314,112.97	4,435,818.31	1,710,917.57	844,060.34	8,761,369.32
2014	420,430.72	1,419,506.18	4,902,076.14	1,970,913.28	1,102,661.80	9,815,588.12
2015	466,275.23	1,681,269.55	5,271,961.14	1,760,071.79	1,139,711.06	10,319,288.77
2016*	185,630.52	735,221.44	2,686,763.48	1,043,346.61	560,838.51	5,211,800.56
Total	3,137,689.22	8,732,064.66	33,618,494.21	13,094,086.83	7,327,094.30	65,909,429.22
Average	348,632.14	970,229.41	3,735,388.25	1,454,898.54	814,121.59	–

\*Data from 2016 correspond to the first semester. Source: DATASUS (<http://tabnet.datasus.gov.br/cgi/tabcgi.exe?sih/cnv/qiuf.def>). Accessed on September 1, 2016.

**Table 3** Health care professional expenses (in BRL) for the chronic subdural hematoma surgical treatment code

Region	North	Northeast	Southeast	South	Central-West	Total
2008	53,762.80	141,443.90	700,318.46	247,544.28	155,470.16	1,298,539.60
2009	97,828.75	236,906.46	932,017.33	316,638.23	237,877.98	1,821,268.75
2010	131,841.80	269,179.70	1,134,175.59	377,252.52	289,015.45	2,201,465.06
2011	140,023.38	380,987.59	1,471,347.18	482,553.69	358,388.00	2,833,299.84
2012	180,931.30	492,039.69	1,778,718.92	548,813.49	385,851.03	3,386,354.43
2013	196,851.19	569,261.68	1,825,123.26	601,079.45	389,804.15	3,582,119.73
2014	185,939.20	589,062.45	1,981,432.59	657,176.98	452,894.78	3,866,506.00
2015	188,139.34	648,661.78	2,121,365.37	600,818.21	511,468.30	4,070,453.00
2016*	89,461.94	312,593.27	1,059,720.20	350,981.53	285,919.86	2,098,676.80
Total	1,264,779.70	3,640,136.52	13,004,218.90	4,182,858.38	3,066,689.71	25,158,683.21
Average	140,531.08	404,459.61	1,444,913.21	464,762.04	340,743.30	–

\*Data from 2016 correspond to the first semester. Source: DATASUS (<http://tabnet.datasus.gov.br/cgi/tabcgi.exe?sih/cnv/qiuf.def>). Accessed on Sep 1, 2016.

**Table 4** Number of deaths related to the chronic subdural hematoma surgical treatment code

	North	Northeast	Southeast	South	Central West	Total
2008	9	31	104	43	20	207
2009	14	31	154	44	29	272
2010	31	51	151	42	30	305
2011	18	53	181	45	39	336
2012	21	42	173	46	16	298
2013	26	70	174	58	26	354
2014	23	65	216	39	28	371
2015	21	84	218	60	30	413
2016*	12	36	111	27	16	202
Total	175	463	1,482	404	234	2,758
Average	19.44	51.44	164.66	44.88	26	–

\*Data from 2016 correspond to the first semester. Source: DATASUS (<http://tabnet.datasus.gov.br/cgi/tabcgi.exe?sih/cnv/qiuf.def>). Accessed on September 1, 2016.

**Table 5** Mortality rate related to the chronic subdural hematoma surgical treatment code

	North	Northeast	Southeast	South	Central-West	Total
2008	8.91	10.73	8.10	10.36	6.67	44.77
2009	8.28	6.47	9.19	8.54	7.20	39.68
2010	13.70	9.57	7.92	7.01	6.37	44.57
2011	9.33	8.82	8.79	7.03	7.62	41.59
2012	10.14	6.51	8.13	7.15	3.31	35.24
2013	11.30	9.37	7.88	8.03	5.37	41.95
2014	10.00	8.56	9.08	5.08	5.21	37.93
2015	9.86	10.40	8.58	8.50	4.85	42.19
2016*	12.63	8.78	8.81	6.89	5.41	42.52
Total	94.15	79.21	76.48	68.59	52.01	370.44
Average	10.46	8.8	8.49	7.62	5.77	–

\*Data from 2016 correspond to the first semester. Source: DATASUS (<http://tabnet.datasus.gov.br/cgi/tabcgi.exe?sih/cnv/qiuf.def>). Accessed on September 1, 2016.

rate has changed in all regions during these years, so that in 2012, 2013, and 2014, the highest rates were observed in the North region, with 10.14, 11.30, and 10.0, respectively. In 2015, the Northeast region returned to the first place, with 10.40; in 2016, from January to June, the North region had the highest mortality rate, with 12.63 (► **Table 5**).

## Discussion

In 1657, Johannes Breakfast Wepfer found a large cyst filled with blood under the dura mater of a patient who had suffered a stroke. In 1817, Houssard discovered that the cyst previously described was a membrane-covered clot; the histology and formation of this lesion were described by Virchow, in 1857, who also called it internal hemorrhagic pachymeningitis. Next, Trotter defended the theory of trauma in drainage veins for the superior longitudinal sinus of the subdural hemorrhagic cyst, consolidating its traumatic etiology in 1914. In 1925, internal hemorrhagic pachymeningitis was renamed CSH.<sup>1,4,5</sup>

Subdural hematomas can be classified as acute, subacute and chronic, according to the time elapsed after the triggering factor. An acute subdural hematoma occurs within 72 hours after the trauma; a subacute hematoma, between 72 hours and 20 days posttrauma; and a chronic hematoma, 20 days posttrauma. Acute hematomas affect mainly young adults, whereas chronic hematomas affect the elderly population.<sup>1,6</sup>

The risk factors and associated diseases for CSHs include trauma, systemic arterial hypertension, neurological diseases (cerebrovascular conditions, malformations and neoplasms), male gender, higher age (> 50 years old), Caucasian ethnicity, alcoholism, diabetes mellitus, cardiovascular diseases, smoking, cerebrovascular diseases, kidney diseases, epilepsy, blood dyscrasias, lung diseases, psychiatric history, and positive human immunodeficiency virus (HIV) serology.<sup>1,7</sup>

Epidemiologically, CSH is most commonly found in the 7<sup>th</sup> decade of life. A male predominance is observed in ~ 70 to

90% of the cases. Its incidence has increased worldwide due to the increase in the populational life expectancy; moreover, it is believed that, in a few years, CSH treatment will be the most performed neurosurgical procedure, surpassing the resection of primary and metastatic tumors.<sup>2,8,9</sup>

The pathophysiology of CSH is not fully understood. The main theories regarding its emergence include the osmotic theory and the recurrent bleeding theory in encapsulated hematomas. The first one is based on the idea that the liquefaction of the hematoma increases the osmotic pressure and the protein content, with the consequent attraction of adjacent fluids to the cavity through a semipermeable membrane. The second theory, of the recurrent bleeding, is more accepted and states that the blood vessels and the abnormal capsule of the hematoma are more subject to bleeding. It is noteworthy that the outer CSH membrane is rich in vessels, with large capillaries, but lacking smooth muscles in its wall.<sup>7-9</sup> Classically, CSH results from a traumatic lesion of the parasagittal Mittenzweig vessels. The causes of nontraumatic CSHs include arteriovenous malformations, intracranial aneurysms, coagulopathies, cerebral convexity tumors, and meningeal carcinomatosis.<sup>1,3</sup>

Among the signs and symptoms found in patients with these characteristics, headache is present in ~ 80% of the cases, as well as a mentation change that can manifest itself in different degrees: confusion, somnolence, or coma. These patients may still present focal neurological deficits, such as hemiparesis (observed in up to 56% of the cases). Some atypical presentations were cited in selected studies, such as isolated neurological deficits (vertigo, nystagmus, and oculomotor paralysis), and extrapyramidal syndromes with a predominance of parkinsonian symptoms.<sup>2,7,10</sup>

Differential diagnoses include stroke, subarachnoid hemorrhage, and tumors.<sup>2</sup> At a computed tomography (CT) of the skull, CSH presents as a hypodensity. Although less used, a magnetic resonance imaging (MRI) of the skull can also be requested in cases in which the CT has some degree of limitation, as in cases of small-volume CSHs.<sup>1,7,10</sup>

The treatment for CSH may be conservative or surgical. Nonsurgical treatments include absolute rest, and use of steroids, mannitol, and other hypertonic solutions.<sup>1,7</sup> It is worth mentioning that the conservative clinical treatment is not indicated for most cases, since surgery is aimed at preventing clinical worsening, neurological deficits, or even death. Currently, the best option is the surgical treatment, which can often be performed through trepanation or craniotomy, with or without postoperative drainage placement.<sup>1,7</sup>

After the surgery, recurrences are not uncommon; they are related to three factors: the patient (age, gender, ethnicity, bleeding tendency, and involved comorbidities), the pathophysiology of the hematoma, and the surgical process. Possible postoperative complications include acute subdural hematoma, intracranial hypertensive hemorrhage, and hypertensive pneumocephalus. However, postoperative healing and recovery rates are high.<sup>1,6</sup>

Pereira et al<sup>11</sup> report that the annual incidence of CSH in the general population ranges from 1.72 to 7.35 per 100,000 people, according to aging, with a higher value found in the 8<sup>th</sup> decade of life. It is estimated that this incidence will continue to grow steadily as the life expectancy increases.

In Japan, where the elderly population grows faster compared with other countries, the annual incidence of CSH is 20.6 per 100,000 people, with 76.5 per 100,000 people in the 8<sup>th</sup> decade of life, and with 127.1 per 100,000 people in individuals > 80 years old.<sup>12</sup> A study conducted in the United States found an annual incidence rate of 79.6 per 100,000 people in the elderly population. The same study also predicted an increase in the number of cases by 2030, reaching 121.4 per 100,000 people in this specific population, and 17.6 per 100,000 people in other age groups, totaling ~ 60,000 cases per year in the USA.<sup>13</sup>

There is limited data on the general incidence of CSH in Brazil. However, it can be inferred from the present paper that, in 2015, the number of hospitalizations due to CSH corresponded to 0.0023% of the population, or 2.39 hospital admissions per 100,000 inhabitants. It should be remembered, however, that the number of hospitalizations does not represent a reliable incidence in the country. This bias could be explained by the noninclusion of selected patients treated in the private health care system.

According to the DATASUS, from 2008 to the 1<sup>st</sup> semester of 2016, there were 33,878 hospital admissions; the Southeast region had the highest number of hospital admissions, with 17,442 hospitalizations, and an annual average number of 2,325.6. The North region had the lowest number of hospital admissions, 1,661, with an annual average of 221.5. Consequently, the Southeast and North regions, respectively, have the highest and lowest indices of hospital expenses, of health care professional expenses, and of number of deaths. The mortality rate was not affected by this rule, since the numbers were higher in the North region.

During the same period, 33,878 CSH-related hospitalizations generated a total hospital expense of BRL 65,909,429.22, and health care professional expenses of BRL 25,158,683.21. In the South region, the average hospitalization and health care

professional expenses were of BRL 2,424.83 and BRL 774.61, respectively.

The total number of deaths by CSH during the evaluated period was 2,758, with the largest number, 1,482, in the Southeast region, followed by 463 in the Northeast region, 404 in the South region, 234 in the Central-West region, and 175 in the North region. It is worth mentioning that the mortality rate ranged from 6.47 to 12.63 per 100,000 patients during the period studied.

Thus, the obtained data allows inferring that a patient admitted due to CSH generated hospital expenses of BRL 1,945.50, and professional expenses of BRL 742.62. A hospitalization day cost BRL 216.16.

During the preparation of the present paper, the non-comprehensiveness of data from surgeries performed by the private health system and health insurance companies was considered a limitation, since our purpose was to collect information from procedures performed by the SUS. In addition, there is a marked scarcity of Brazilian and worldwide epidemiological contents related to the incidence and prevalence of CSH. Another possible bias of the present study was the probability of multiple and/or erroneous diagnoses inclusion in the results found in the DATASUS database.

## Conclusion

The incidence of CSH has increased in Brazil and worldwide. It is a benign pathology when diagnosed early, since its main approach is surgical treatment. According to the DATASUS database, from 2008 to the 1<sup>st</sup> semester of 2016, there were a total of 33,878 hospital admissions for surgical CSH treatment. The total number of deaths from this disease during the same period was 2,758, and the mortality rate ranged from 6.47 to 12.63%. These admissions totaled hospital expenses of BRL 65,909,429.22, and health care professional services expenses of BRL 25,158,683.21. Although CSH is a clinically relevant disease, there are few epidemiological studies about it. Thus, the present paper approaches the Brazilian epidemiological aspects of CSH.

## Conflicts of Interest

The authors have no conflicts of interest to declare.

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# Epidemiology and Estimated Cost of Surgeries for Carpal Tunnel Syndrome Conducted by the Unified Health System in Brazil (2008–2016)

## *Epidemiologia e estimativa de custo das cirurgias para síndrome do túnel do carpo realizadas pelo Sistema Único de Saúde no Brasil (2008–2016)*

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### Abstract

**Objective** To define the epidemiological aspects and estimated costs of surgeries performed by the Brazilian Unified Health System (SUS, in the Portuguese acronym) for the treatment of carpal tunnel syndrome (CTS) in Brazil between 2008 and 2016.

**Materials and Methods** Documentary study, with data from the Informatics Department of the SUS (DATASUS, in the Portuguese acronym), about the absolute number and incidence of admissions, the total and mean length of stay (in days), the total expenses, and the hospital and professional services expenses in the surgical treatment of CTS.

**Results** During the period studied, there were 82,123 hospitalizations for surgery, with a 62% increase from 2008 to 2015, accompanied by an increase in the values of professional services. The incidence and the absolute number of procedures were higher in the South and Southeast regions. The North region presented the lowest expenses, absolute number, and incidence of admissions, as well as the longer mean length of stay. The mean length of stay decreased from 1.2 days in 2008 to 0.7 day in 2016. The expenses totaled BRL 29,463,148.80 during the period studied. After 2011, professional services became the largest portion of the total expenses. Hospital expenses corresponded to 52.49% of the expenditure in 2008, and to 36.24% in 2015, while professional expenses received the largest investment compared with the total amount expended in 2012.

### Keywords

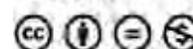
- ▶ epidemiology
- ▶ carpal tunnel syndrome/surgery
- ▶ median nerve
- ▶ health expenditures

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**Conclusion** The present study was not consistent with the international literature regarding epidemiological data and cost estimates. Brazil presented a disparity between absolute numbers and annual incidence of admissions and length of stay. Future researches can assess variables that have influenced these results, as well as contribute to public interventions aimed at the improvement of health care.

## Resumo

**Objetivo** Definir aspectos epidemiológicos e estimativas de custos decorrentes das cirurgias realizadas pelo Sistema Único de Saúde (SUS) para tratamento da síndrome do túnel do carpo no Brasil, entre 2008 e 2016.

**Materiais e Métodos** Estudo documental, com dados do Departamento de Informática do SUS (DATASUS), referentes ao número absoluto e à incidência de internações, dias totais e média de permanência nosocomial, valor total investido, e valor dos serviços hospitalares e profissionais no tratamento cirúrgico da síndrome.

**Resultados** No período estudado, realizaram-se 82.123 internações para cirurgia, com crescimento de 62% de 2008 a 2015, acompanhado de incremento no pagamento aos profissionais. A incidência e o número absoluto de procedimentos sobressaíram-se no Sul e Sudeste, invariavelmente. A região Norte apresentou número absoluto, incidência e valor investido menores, além do maior tempo médio de internação. A permanência hospitalar média reduziu-se de 1,2 dias em 2008 para 0,7 dia em 2016. Aplicaram-se R\$ 29.463.148,80 no período. Após 2011, o valor do serviço profissional se tornou a maior parcela do total. A remuneração hospitalar correspondeu a 52,49% do gasto em 2008 e a 36,24% em 2015, enquanto o gasto com profissionais recebeu o maior investimento em relação ao total de despesas em 2012.

**Conclusão** O presente estudo foi discrepante da literatura internacional no que concerne a dados epidemiológicos e estimativas de custo. No Brasil, notou-se disparidade entre o número absoluto, a incidência anual e o tempo de internação hospitalar desses pacientes. Pesquisas futuras poderão avaliar as variáveis que influenciaram estes resultados, bem como contribuir para intervenções públicas que visem à melhoria da assistência.

## Palavras-chave

- ▶ epidemiologia
- ▶ síndrome do túnel do carpo/cirurgia
- ▶ nervo mediano
- ▶ gastos em saúde

## Introduction

Carpal tunnel syndrome (CTS) is the most common compressive mononeuropathy, and it is characterized by the compression of the median nerve at the level of the wrist.<sup>1-5</sup>

The first description of CTS is attributed to Paget, who reported a case of compression of the median nerve resulting from a distal radius fracture.<sup>6</sup>

The carpal tunnel is an inextensible osteofibrous tunnel defined as the space between the flexor retinaculum (FR), which constitutes the roof, and the carpal canal, which constitutes the fundus. It is delimited at the ulnar border by the hook of the hamate and by the pyramidal and pisiform muscles, and at the radial border by the scaphoid, the trapezium, and the flexor retinaculum muscles. The base is formed by the capsule, and the anterior radiocarpal ligaments cover underlying portions of the scaphoid, the lunate, the capitate, the hamate, the trapezium, and the trapezoid muscles. The median nerve, in turn, is accompanied by the four tendons of the flexor digitorum superficialis (FDS), by the four tendons of the deep flexor muscles of the fingers (FPD), and by the tendon of the flexor longus muscle of the thumb (FPL). The FPL is the most radial element. At the entrance of the tunnel, the median

nerve is dorsal to the tendon of the palmaris longus muscle (PL), between the tendons of the Flexor Carpi Radialis (FCR) and the flexor digitorum superficialis (FDS). In the distal part of the tunnel, the median nerve divides into the following branches: lateral, medial, and recurrent thenar; the former two branches subdivide further. The thenar branch passes through a separate tunnel before reaching the thenar muscles in 56% of the cases.<sup>7</sup>

Typical CTS symptomatology is manifested by pain, hypoesthesia, and paresthesia in the sensory innervation territory of the median nerve and, in some cases, paresis for thumb opponency and abduction, as well as signs of thenar hypotrophy.<sup>8</sup> The risk factors for CTS include diabetes mellitus, alcoholism, amyloidosis, rheumatoid arthritis, infectious synovitis, gout, dermatomyositis, scleroderma, hypothyroidism, long-term hemodialysis, obesity, repeated flexion-extension wrist movements, extended wrist use to move loads, and wrist trauma.<sup>9</sup>

The differential diagnosis should include C6 and C7 vertebrae radiculopathy, proximal medial nerve compressions in the arm (Struthers ligament) or in the forearm (pronator syndrome), and double impact syndrome. Less frequently, upper motor neuron

disease, cervical intraradicular injuries, neuropathies, and syringomyelia can be considered in the differential diagnosis.<sup>8</sup> Although an obvious cause for CTS is not identified, most cases are well-related to heavy manual labor.<sup>10</sup>

The treatment for CTS can be conservative or surgical. Surgery is indicated in cases with electrophysiological evidence of median nerve denervation; acute post-traumatic cases associated with significant local edema; significant functional impairment of the median nerve, evidenced by thenar atrophy and/or severe motor and sensorial changes; conservative treatment failure; or complying with the choice of the patient.<sup>11,12</sup> The frequency of indication for surgical treatment is high, ranging from ~31 to 40% of the diagnosed patients; it is estimated that US\$ 2 billion are spent annually in the United States on the surgical treatment of CTS.<sup>13,14</sup>

The surgical technique for the treatment of CTS underwent great advances. Open surgery results in a bigger surgical scar, increasing the recovery time and the risk of complications. Modern strategies consist of endoscopic and mini-incision methods, which reduce the length of stay, complications, and scarring, resulting in an early return to work.<sup>13</sup> A major limitation of the new techniques, especially the endoscopic technique, is the need for larger hospital equipment, often requiring expensive disposable items, which is not a reality in low-budget regions. Another caveat is the need for endoscopic training, which increases the time required for a proper learning curve. The mini-incision technique tries to combine the lower expense of the open surgeries with the shorter length of stay and reduced scarring of the endoscopic method; however, further studies are needed for its long-term assessment.<sup>15,16</sup>

Given the socioeconomic impact (up to US\$ 30,000 in the lifetime of each United States worker) of this pathology and the best results obtained with surgical treatment (superiority in functional improvement, in addition to early and long-term regression of the symptoms),<sup>17</sup> the present study tried to define the epidemiological aspects and cost estimates resulting from surgeries performed by the Brazilian Unified Health System (SUS, in the Portuguese acronym) for the treatment of CTS in Brazil between 2008 and 2016.

## Materials and Methods

This is a documentary study. Queries in the information system from the Informatics Department of the SUS (DATA-SUS, in the Portuguese acronym) yielded national and regional registries regarding the surgical treatment of CTS from January 2008 to July 2016. The study population corresponds to people submitted to treatment with the procedural code 0403020123 (surgical treatment of compressive syndrome in osteofibrous tunnel at the carpal level).

The variables studied were the absolute number and the annual incidence of hospital admission, the total and mean length of stay (in days), total expenses, and hospital and professional services expenses.

For the proper interpretation of the results, data on population variation over the years was also obtained, using the 2010 demographic census and estimates by the Brazilian

Institute of Geography and Statistics (IBGE, in the Portuguese acronym).

The results were tabulated using the Microsoft Excel (Microsoft Corporation, Redmond, WA, USA) software and were subsequently analyzed.

Next, a bibliographic survey was conducted, searching for Brazilian and international publications in the PubMed, Virtual Health Library (BVS, in the Portuguese acronym), Latin American and Caribbean in Health Sciences (LILACS), National Library of Medicine (MEDLINE), and Scientific Electronic Library Online (SciELO) databases with the following descriptors: *epidemiology*, *carpal tunnel syndrome*, *median nerve* and *costs*. To narrow the search, only articles published from 1998 to 2016 were considered. The following inclusion criteria were used: papers published in indexed national and international journals, written in English and Portuguese. Papers with titles and abstracts that did not fit the research objectives were excluded.

## Results

From January 2008 to July 2016, there were 82,123 hospitalizations for the surgical treatment of CTS at the SUS. From this total, 14.18% were performed in 2014, the year with the highest number of procedures. In all of the years analyzed, the Southeast region presented the highest rates, while the North region remained with the lowest rates (► **Table 1**).

The highest and lowest counts per region occurred in the Southeast region in 2014 and in the North region in 2010, with 5,911 and 72 hospitalizations for surgery, respectively. There was a growth of 62%, rising from 6,830 in 2008 to 11,078 in 2015. All of the regions presented an increasing pattern, except the North region, with oscillation over the years, and the Northeast region, with an initial decrease associated with a discrete growth in 2014 and a sudden peak in 2015 (1,072 procedures).

The annual incidence of hospitalizations for the procedure increased 50.27%, with its apex in 2014, when it reached a rate of 5.46 per 100,000 Brazilian inhabitants. The highest regional incidence was recorded in the South region in 2014 (12.32), and the lowest in the North region in 2010 (0.43). These regions accounted for, respectively, the highest and lowest admission numbers in all of the years studied. The North and Northeast regions were the only ones presenting a decrease in the incidence in the comparison between 2008 and 2015. Like the admission incidence, the growth rate was higher in the South and Central-West regions, and lower in the North and Northeast regions (► **Table 2**).

The total hospitalization time for the surgical treatment of CTS declined nationally over the years (► **Table 3**). Considering the fully reported period (2008–2015), the greatest length of stay occurred in 2008, with 8,520 days, and the lowest occurred in 2015, with 8,177 days. Following the national trend, this absolute value fell in the North, Northeast and Southeast regions; however, it grew significantly in the South and Central-West regions. The mean length of stay in Brazil fell from 1.2 to 0.7 day in 2015. It was higher in the North region in all of the years studied, and lower in the

**Table 1** Number of hospital admissions for the surgical treatment of carpal tunnel syndrome at the SUS by region, 2008–2016

Year	Number of procedures	North region	Northeast region	Southeast region	South region	Central-West region
2008	6,830	113	1,027	3,858	1,411	421
2009	7,920	74	927	4,306	2,082	531
2010	8,577	72	919	4,865	2,128	593
2011	10,030	109	864	5,673	2,695	689
2012	10,097	143	882	5,225	3,045	802
2013	10,166	132	833	5,054	3,237	910
2014	11,646	155	844	5,911	3,576	1,160
2015	11,078	118	1,072	5,807	3,082	999
2016*	5,779	74	680	3,010	1,457	558
Total	82,123	990	8,048	43,709	22,713	6,663

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS, 2016. \*Reports from the first 7 months of 2016.

**Table 2** Annual incidence of surgical treatment of carpal tunnel syndrome at the SUS by region per 100,000 inhabitants, 2008–2015

Year	Annual incidence	North region	Northeast region	Southeast region	South region	Central-West region
2008	3.60	0.74	1.93	4.81	5.13	3.07
2009	4.13	0.48	1.72	5.32	7.51	3.82
2010	4.49	0.43	1.73	6.05	7.77	4.22
2011	5.21	0.67	1.61	7.00	9.77	4.83
2012	5.20	0.87	1.63	6.40	10.97	5.56
2013	5.05	0.77	1.49	5.98	11.24	6.06
2014	5.46	0.89	1.50	6.94	12.32	7.62
2015	5.41	0.67	1.89	6.77	10.54	6.46

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS, 2016. Incidence determined from population data provided by IBGE. Data from 2016 was not included due to possible seasonal incidence variations not analyzed in the present work.

**Table 3** Total length of stay (in days) of carpal tunnel syndrome patients surgically treated at the SUS per region, 2008–2016

Year	Total days	North region	Northeast region	Southeast region	South region	Central-West region
2008	8,520	277	1,620	4,215	1,415	993
2009	7,801	138	1,414	3,858	1,639	752
2010	7,930	198	1,343	4,020	1,524	845
2011	8,321	203	1,222	4,148	1,761	987
2012	8,410	243	1,233	3,654	2,120	1,160
2013	8,370	321	1,134	3,406	2,218	1,291
2014	9,105	245	1,154	3,706	2,347	1,653
2015	8,117	247	1,382	3,073	1,993	1,422
2016*	3,865	85	697	1,419	902	762

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS, 2016. \*Reports from the first 7 months of 2016.

South and Southeast regions, which presented the lowest mean lengths of stay in all of the periods studied. The Southeast region had the lowest rate of 2016 (0.5). The highest mean value was recorded in 2010, in the North region, reaching 2.8 days (►Table 4). Regarding the total

procedural expenditure, ►Table 5 shows that the largest value occurred in 2014, when it reached BRL 4,957,966.85, decreasing to BRL 4,632,777.69 in 2015. This cost is higher than those observed in 2013 and in the previous years. The only year in which there was a cost reduction compared with

**Table 4** Mean length of stay of carpal tunnel syndrome patients surgically treated at the SUS per region, 2008–2016

Year	Mean period (days)	North region	Northeast region	Southeast region	South region	Central-West region
2008	1.2	2.5	1.6	1.1	1.0	2.4
2009	1.0	1.9	1.5	0.9	0.8	1.4
2010	0.9	2.8	1.5	0.8	0.7	1.4
2011	0.8	1.9	1.4	0.7	0.7	1.4
2012	0.8	1.7	1.4	0.7	0.7	1.4
2013	0.8	2.4	1.4	0.7	0.7	1.4
2014	0.8	1.6	1.4	0.6	0.7	1.4
2015	0.7	2.1	1.3	0.5	0.6	1.4
2016*	0.7	1.6	1.1	0.5	0.6	1.4

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS, 2016. \*Reports from the first 7 months of 2016.

**Table 5** Total expenditure (in BRL) of the surgical treatment of carpal tunnel syndrome at the SUS per region, 2008–2016

Year	Total (BRL)	North region	Northeast region	Southeast region	South region	Central-West region
2008	1,956,232.97	32,450.94	293,199.53	1,104,713.59	399,028.41	126,840.50
2009	2,264,984.46	25,316.52	267,189.58	1,229,803.11	631,633.43	152,351.28
2010	2,505,128.36	22,727.17	274,414.41	1,419,187.76	614,466.80	174,332.22
2011	3,330,652.34	36,400.55	296,005.01	1,874,866.80	894,129.29	229,250.69
2012	3,621,110.85	51,397.39	318,348.32	1,859,957.38	1,082,463.07	308,945.08
2013	4,060,832.58	49,872.47	304,416.70	1,872,309.66	1,409,867.96	424,365.79
2014	4,957,966.85	55,386.46	310,609.16	2,453,460.40	1,600,756.42	537,709.41
2015	4,632,777.69	44,931.42	392,388.76	2,387,386.78	1,355,372.79	452,598.94
2016*	2,133,462.70	26,194.35	260,334.74	1,076,805.11	516,313.48	253,815.02

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS, 2016. \*Reports from the first 7 months of 2016.

the previous one was 2015. Over 8 years and 7 months, the expenses totaled BRL 29,463,148.80. Analyzing the entire period studied, the highest expense occurred in the Southeast region, reaching BRL 2,453,460.40 in 2014, equivalent to ~ 49.5% of the national expenditure of that year. There was a 136.8% increase in investment from 2008 to 2015 throughout Brazil. Throughout the years, the Southeast region was superior to the others in terms of applied capital. The Northeast, South and Central-West regions showed a clear investment growth, while the North region, despite modest variations, always maintained the lowest total expenditure, reaching a maximum of BRL 55,386.46 in 2014.

The hospital services expenses, which are components of the total expenditure, oscillated in a similar way over the years, with a maximum expense in 2014 (► **Table 6**). The hospital services expenses accounted for 52.49% of the total invested in 2008, falling progressively to 35.7% in 2014, increasing to 36.24% in 2015, and to 40.58% in the first 7 months of 2016. These proportions have maintained the same percentages in all of the regions in 2008 and in 2015.

The highest amount paid for professional services was in 2014 (BRL 2,373,272.41), which corresponds to 47.5% of the total expenditure in 2008, and it reached a maximum

proportion of 56.77% in 2012. The professional service expenses fell progressively to 47.86% in 2014. In the first 7 months of 2016, this proportion was of 54.52%, indicating a new rise that started in 2015 (48.74%) (► **Table 7**).

## Discussion

Although CTS is the most common neuropathy of the upper limb and its surgery is usually performed by neurosurgeons and orthopedists, it is a condition with few epidemiological data in Brazil and other countries. Data from international studies show a relevant divergence regarding the annual incidence of CTS. A study performed in the Netherlands in 2001 showed an annual incidence of 1.8 per 1,000 inhabitants, predominantly in female patients aged between 45 and 64 years old. In Siena, Italy, from 1991 to 1998, the incidence of CTS was of 329.4 per 100,000 people. In the United Kingdom, a study performed from 1992 to 2001 showed an annual incidence of 139.4 for women and of 67.2 for men per 100,000 inhabitants in the city of Canterbury, and of 83.2 for women and of 48 for men per 100,000 cases in Huddersfield; all of the cases were confirmed by electrophysiological tests. In 2008, 127,268 patients ≥

**Table 6** Total amount (in BRL) paid for hospital services related to the surgical treatment of carpal tunnel syndrome at the SUS per region, 2008–2016

Year	Total (BRL)	North region	Northeast region	Southeast region	South region	Central-West region
2008	1,027,020.31	17,085.56	154,185.39	578,351.61	208,226.80	69,170.95
2009	1,188,365.60	14,709.68	141,461.27	643,205.75	308,421.35	80,567.55
2010	1,283,200.64	12,135.40	142,844.11	724,654.98	312,903.75	90,662.10
2011	1,508,050.76	16,842.17	135,945.82	847,939.35	400,306.52	107,016.90
2012	1,520,242.30	22,291.00	136,633.74	778,419.56	458,155.97	124,742.03
2013	1,545,954.63	22,560.91	130,193.12	753,333.04	497,924.79	141,942.77
2014	1,770,406.62	23,879.47	130,987.62	886,127.41	550,382.92	179,029.75
2015	1,679,020.01	19,058.65	165,658.33	869,743.15	470,898.48	153,661.40
2016*	865,848.20	11,400.75	105,293.13	446,178.44	220,892.10	82,083.78

Source: DATASUS 2016. \*Reports from the first 7 months of 2016.

**Table 7** Total amount (in Brazilian reais) paid for professional services related to the surgical treatment of carpal tunnel syndrome at the SUS per region, 2008–2016

Year	Total (BRL)	North region	Northeast region	Southeast region	South region	Central-West region
2008	929,212.66	15,365.38	139,014.14	526,361.98	190,801.61	57,669.55
2009	1,076,618.86	10,606.84	125,728.31	586,597.36	281,902.62	71,783.73
2010	1,221,927.72	10,591.47	131,570.30	694,532.78	301,563.05	83,670.12
2011	1,822,601.58	19,558.38	160,059.19	1,026,927.45	493,822.77	122,233.79
2012	2,055,957.32	29,106.39	181,714.19	1,065,604.56	617,055.38	161,476.80
2013	2,072,507.25	26,963.94	171,964.05	1,033,671.66	655,507.26	184,497.34
2014	2,373,272.41	31,506.99	172,843.50	1,209,616.92	724,318.67	234,977.33
2015	2,258,446.24	24,134.67	220,473.71	1,186,946.83	624,363.27	202,527.76
2016*	1,163,347.14	14,793.60	138,686.31	611,092.48	289,091.68	108,681.07

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS 2016. \*Reports from the first 7 months of 2016.

20 years old underwent surgery in metropolitan France, with an incidence of 2.7 surgeries per 1,000 inhabitants.<sup>1–5</sup> In a study conducted in the eastern part of the county of Kent in the United Kingdom, from 1992 to 2001, 6,245 patients were diagnosed with CTS by an electroneuromyography test, and, of these, 4,646 underwent surgical treatment, resulting in an incidence of 349 procedures per 100,000 inhabitants, considering the entire regional population in 2001.<sup>4,18</sup> It is estimated that 400 thousand surgeries for CTS correction will be performed annually in the United States, equivalent to an incidence of 125 surgeries per 100,000 inhabitants, considering the population of the country in 2014.<sup>17,19</sup>

There was a total of 82,123 hospitalizations for surgery throughout the period studied in all of the Brazilian territory, with a 62% increase in the number of procedures from 2008 to 2015. The incidence of surgical treatment for CTS varied similarly to the total surgeries rate: from 3.6 per 100,000 people in 2008 to 5.41 per 100,000 people in 2015, equivalent to a 50.27% growth.

The analysis of the aforementioned data reveals that the Brazilian incidence of the surgical treatment for CTS is significantly lower than those observed in developed countries. This

discrepancy could have different explanations: a result of the precarious investment in the public health system; technical difficulties for the health care professionals; lack of reference services; SUS inefficiency, resulting in long waiting periods for the procedures to be performed; preference of the patients for the private system, as well as the noncomputation of the data coming from the private system.

The highest absolute number of hospitalizations was observed in the Southeast region, with a total of 43,709, which represents 52% of the national number. The same cannot be said of the incidence of hospitalizations, which is higher in the South region, with ~ 10.54 per 100,000 inhabitants in 2015. In contrast, the North region had the lowest hospitalization rate, with a total of 8,048, equivalent to 9% of the whole country. It also presents the smallest incidence of surgeries, with ~ 0.67 procedures per 100,000 inhabitants in 2015. It is possible to notice regional variations that can probably be justified by demographic, economic, and social factors; as such, the South and Southeast regions have a higher population density, greater access to high complexity centers, and a greater volume of physicians qualified for the diagnosis and surgical treatment of CTS.

The highest length of stay occurred in 2014, which is consistent with the number of surgeries performed in the period. Nevertheless, the length of stay decreased over the years, which could be associated with the evolution of the surgical technique, the training of the professionals, and with the greater safety for early hospital discharges. Although presenting the highest absolute lengths of stay for CTS treatment, the Southeast and South regions had the lowest mean lengths of stay. On the other hand, the North region, despite having the lowest number of hospitalizations, maintained the highest mean length of stay.

In the period studied, BRL 29,463,148.80 were spent. The highest total national expenditure was registered in 2014, with BRL 4,957,966.85, an 136.8% increase compared with 2008. This fact demonstrates that the increase in the number of surgeries was accompanied by a greater investment in hospitals and health care professionals. It is important to note that, according to the Management System for the Table of Procedures, Medications and Orthoses, Prostheses and Materials (SIGTAP, in the Portuguese acronym) of the SUS, the total expenses per procedure was BRL 280.14 from 2008 to May 2011, when it increased to BRL 347.62, which corresponds to a 24% increase in 3 years. This variation resulted from a 50% increase in the value of professional services (from BRL 134.96 to BRL 202.44 per surgery).

Regarding the cost of the surgical procedure in other countries, it is estimated that open CTS surgery in the province of Ontario, Canada, costs on average CAD 307 per patient. The total cost of Medicare (the health insurance program of the United States for people > 65 years old) was USD 842.<sup>20,21</sup> These data allow us to infer that international investments in the surgical treatment of CTS are superior to the national ones. This can result from a more attractive professional remuneration and from the greater apparatus available for the performance of the procedures.

At the interregional level, the highest expenses occurred in the Southeast region, with a maximum of BRL 2,453,460.40 in 2014, corresponding to 49.5% of the national expenditure in that year. In contrast, the North region always remained with the lowest values throughout the period studied. This can be explained by the lower incidence and by the lower absolute number of surgeries in the North, since the cost per procedure is the same throughout Brazil.

The hospital and professional services expenses make up the total expenditure. Until 2011, the hospital expenses accounted for most of the total cost; then, there was an inversion, a fact justified by the SIGTAP changes already mentioned. As of 2012, the professional and hospital expenses ceased to be the only components of the total expenditure. This can be observed by the incompatibility of the total recorded value in relation to the sum of both services. In 2008, this result was the exact total cost value. The resources not attributed to the 2 services correspond to 1.25% of the total in 2012; 10.91% in 2013; 16.44% in 2014; 15.02% in 2015; and 4.9% in the first 7 months of 2016. As such, there is a 3<sup>rd</sup> variable composing the total expenses as of 2011, but not explained in the information system, perhaps due to a failure in the documentation process.

Despite the reliable data regarding procedures performed by the SUS, the method used in the present study has the bias of not evaluating the patients submitted to clinical treatment, not considering surgeries performed at the private health care network, and the possibility of typing and registration errors. Another bias is the existence of three more codes that can be used by surgeons for the surgical treatment of CTS: peripheral nerve neurolysis (procedural code: 0403020077), peripheral nerve micro-neurolysis (procedural code: 0403020050), and neuropathy with or without microsurgery (procedural code: 0403020115) (→ **Table 8**).

**Table 8** Distribution of the number of procedures with codes used for the surgical treatment of carpal tunnel syndrome patients at the SUS, 2008–2016

Year	Nonfunctional peripheral nerve neurolysis (procedural code: 0403020077)	Peripheral nerve micro-neurolysis (procedural code: 0403020050)	Compressive neuropathy with or without microsurgery (procedural code: 0403020115)	Total
2008	3,123	4,376	5,151	12,650
2009	3,453	5,024	6,015	14,492
2010	3,157	4,874	5,988	14,019
2011	3,049	4,934	6,532	14,515
2012	3,291	5,059	7,139	15,489
2013	3,021	4,821	7,156	14,998
2014	3,134	4,918	7,064	15,116
2015	3,917	6,413	8,508	18,838
2016*	2,267	1,526	2,582	6,375
Total	26,145	40,419	53,553	120,117

Abbreviations: SUS, Brazilian Unified Health System.

Source: DATASUS 2016. \*Reports from the first 7 months of 2016.

## Conclusion

There was an inconsistency between the present study and the international literature regarding epidemiological data and cost estimates for the surgical treatment of CTS. Within the Brazilian territory, there was a disparity between the absolute number of admissions, the annual incidence, and the length of stay of these patients. Expenses with the surgical procedure were lower compared with other studied countries, although Brazilian patients had longer lengths of stay. Over the years, there was a value readjustment, favoring professional services. It is important to emphasize the presence of a noninclusion bias from patients submitted to clinical treatment or who underwent surgeries through the private health care system. Future research can evaluate the variables that influenced the results obtained, as well as, in some way, contribute to public interventions aimed at improving the medical care of these patients.

### Conflicts of Interest

The authors have no conflicts of interest to declare.

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# Epidemiological Profile of Malignant Brain Neoplasms in the Northern Region of Brazil: data from the Cancer Hospital Registry of the Instituto Nacional de Câncer

## *Perfil epidemiológico das neoplasias primárias malignas do encéfalo na região Norte do Brasil: dados dos Registros Hospitalares de Câncer do Instituto Nacional de Câncer*

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### Abstract

**Objective** Tumors of the central nervous system (CNS) are considered rare, with an incidence of 3.4 cases per 100,000 individuals worldwide. Although uncommon, CNS tumors have been gaining epidemiological importance due to their increased incidence and mortality. In Brazil, there is a lack of population research regarding CNS cancer, especially in the Northern region. Thus, the authors aim to trace an epidemiological profile of malignant brain neoplasms in the Northern region from 2001 to 2013.

**Methods** Data were collected from the Cancer Hospital Registry of the Instituto Nacional de Câncer (RHC-INCA, in the Portuguese acronym) and stratified according to origin, gender, age, detailed primary location, and histological type. A total of 742 cases were analyzed. Most of the cases came from inland areas, with a male predominance.

**Results** The most affected age groups were between 0 and 9 years old and between 30 and 49 years old, with an accentuated decrease in incidence starting at the age of 70 years old. The frontal lobe was the most affected area, followed by the temporal and parietal lobes. Astrocytic tumors accounted for 64.3% of cases, followed by embryonal tumors (18.2%), and ependymal tumors (7.4%). Among the astrocytic tumors, astrocytoma, not otherwise specified (NOS), and glioblastoma, NOS corresponded to 82.2% of the cases. Among embryonal tumors, medulloblastoma accounted for 71.9% of the cases.

**Conclusion** More epidemiological studies in this area, especially in the Northern region, are required to identify risk factors and allow prevention and early diagnosis.

### Keywords

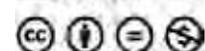
- ▶ brain neoplasms
- ▶ malignant neoplasm
- ▶ epidemiology

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**Resumo**

**Objetivo** Os tumores do sistema nervoso central (SNC) são considerados raros, com incidência de 3,4 casos a cada 100 mil indivíduos no mundo. Apesar de serem considerados raros, os tumores do SNC vêm ganhando importância epidemiológica devido ao aumento de sua incidência e mortalidade. No Brasil, há carência de pesquisas populacionais quando se trata de câncer no SNC, principalmente no Norte do país. Dessa forma, os autores objetivam traçar o perfil epidemiológico das neoplasias encefálicas malignas na região Norte de 2001 a 2013.

**Métodos** Os dados foram coletados dos Registros Hospitalares de Câncer do Instituto Nacional de Câncer (RHC-INCA) e estratificados em: procedência, gênero, faixa etária, localização primária detalhada e tipo histológico. Foram analisados 742 casos. A maioria dos pacientes teve procedência de cidades interioranas, e houve predominância do sexo masculino.

**Resultados** As faixas etárias mais acometidas foram de 0 a 9 anos e de 30 a 49 anos, com decréscimo acentuado a partir dos 70 anos. O lobo frontal foi o mais acometido, seguido pelos lobos temporal e parietal. Os tumores astrocíticos compuseram 64,3% do total, seguido pelos tumores embrionários (18,2%) e ependimários (7,4%). No grupo dos tumores astrocíticos, o astrocitoma, sem outra especificação (SOE), e o glioblastoma, SOE corresponderam a 82,2% dos casos. No grupo dos tumores embrionários, o meduloblastoma correspondeu a 71,9%.

**Conclusão** Ressalta-se a necessidade de mais estudos epidemiológicos nessa área, especialmente na região Norte, com vistas à identificação de fatores de risco, prevenção e diagnóstico precoce na população.

**Palavras-chave**

- ▶ neoplasias encefálicas
- ▶ neoplasia maligna
- ▶ epidemiologia

**Introduction**

Primary tumors of the central nervous system (CNS) are considered rare, with an incidence of 3.4 cases per 100,000 individuals worldwide.<sup>1</sup> However, knowledge about these tumors is still restricted in the medical literature.

Statistics from the United States indicate an incidence of 117,023 malignant cases between 2008 and 2012, with the most frequent brain location at the frontal, temporal, and parietal lobes.<sup>2</sup>

In Brazil, it is estimated that 10,270 new cases of primary CNS neoplasms occurred in 2016, more than a half of these in men. The most affected region is the Southeast, with 4,210 cases. In the Northern region, 420 cases were estimated, mostly in men (54.7%). This type of cancer is the tenth most frequent in the region. It is estimated that the rate of these tumors per 100,000 individuals is of 2.62 cases for men and of 2.21 cases for women.<sup>3</sup> Although rare, CNS neoplasms are among the deadliest, and only half of the diagnosed individuals survive for 1 year after the diagnosis.<sup>4</sup>

In Brazil, there is a lack of populational research regarding CNS cancer, especially in the Northern region. The need for further epidemiological studies on these neoplasms has already been evidenced in systematic reviews and meta-analyses.<sup>5</sup>

Although this is a constantly evolving area in the medical literature, developing countries, such as Brazil, suffer from deficiencies in the screening of suspected cancer cases. With 7 states and a total estimated population of 17,707,783 inhabitants,<sup>6</sup> the Northern region has one of the lowest Human Development Indexes of Brazil, and it is still plagued

by infrastructure shortages, such as lack of hospitals and difficult access to reference hospitals; these factors contribute to deficient records, implying the need for research and studies on the regional epidemiological profile of brain tumors. As such, the authors aim to trace an epidemiological profile of primary brain malignant neoplasms in the Northern region from 2001 to 2013.

**Materials and Methods**

This is a descriptive, longitudinal, retrospective study. The present research complied with the ethical precepts established by CNS 466/12, and its preliminary project was approved by the Nucleus of Research and Extension in Medicine and Research Ethics Committee under the protocol number 07/15. Data collection was performed at the José de Alencar Gomes da Silva database of the Cancer Hospital Registry of the Instituto Nacional de Câncer (RHC-INCA, in the Portuguese acronym).

State health departments submit RHC information to the INCA, who builds a consolidated national base after the elimination of multiplicities. Nine hospitals from the Northern region sent primary CNS neoplasms data to the INCA system during the period analyzed; two hospitals from Pará and Tocantins, and one hospital from each of the remaining states (Acre, Amazonas, Amapá, Roraima, and Rondônia).

Data from patients enrolled in the RHC-INCA database who were diagnosed with primary malignant brain neoplasms in the Northern region between January 2001 and December 2013 were included. Data were stratified for further analysis



**Fig. 1** Distribution by origin of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).

according to origin, gender, age group, detailed primary location, and histological type. To assure data reliability, the authors analyzed the records included in the RHC-INCA database, excluding those from patients who were not diagnosed with primary brain malignancies, not originating from the Northern region, not pertaining to the investigated period, and who were deemed insufficient for the assessment required in the present study, such as incompletely filled or blank forms.

The information obtained was organized in a database and submitted to statistical analysis using Microsoft Office Excel 2016 (Microsoft Corporation, Redmond, WA, USA) and TabWin version 3.6 for Windows (DATASUS, Rio de Janeiro, RJ, Brazil). The chi-squared test was performed to investigate the association between gender or age and histological type. The null hypothesis rejection index was set at 0.05, or 5%.

Microsoft Office Excel 2016 software (Microsoft Corporation) was used to prepare the graphs and tables. Maps were prepared with TabWin software version 3.6 for Windows. This text was written using Microsoft Office Word 2016 (Microsoft Corporation, Redmond, WA, USA).

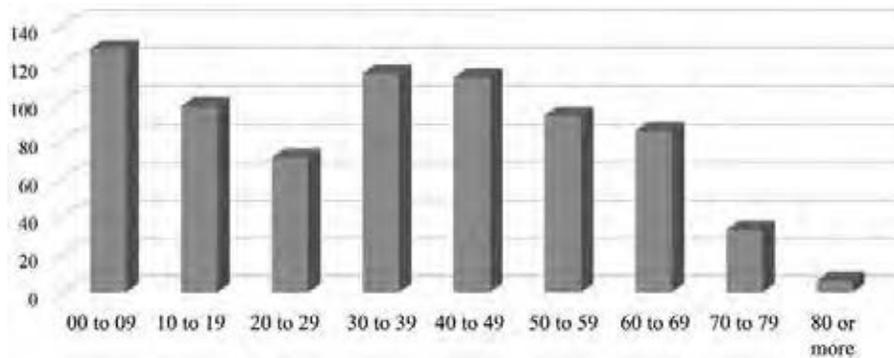
## Results

There were 933 cases in the period studied, of which 191 met the exclusion criteria because their information was insufficient for an adequate analysis. Thus, a total of 742 cases were assessed, of which 344 originated in capital cities (46.4%) and 398 in inland areas (53.6%) (►Fig. 1). There were 428 male cases (57.7%) and 314 female cases (42.3%), with a statistically

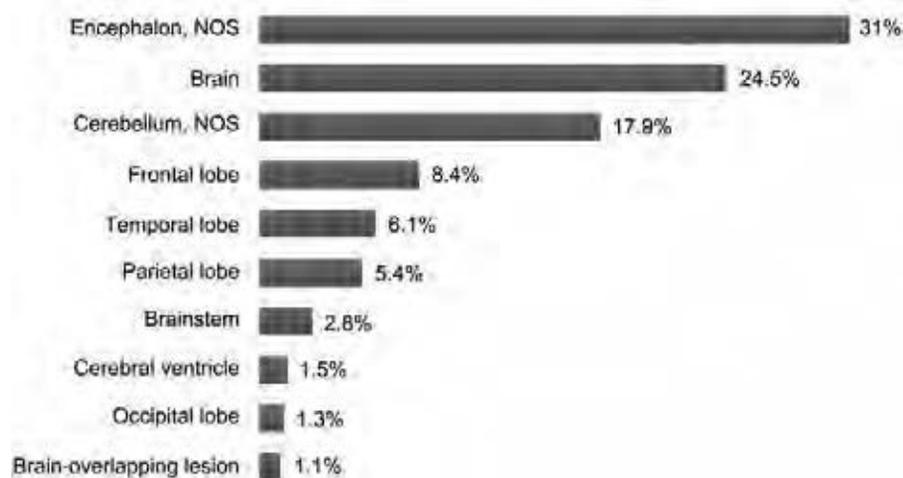
significant difference ( $p = 0.0007$ ) (►Fig. 2). The most affected age groups were between 0 and 9 years old (128 patients), and between 30 and 49 years old (115 patients), followed by between 40 and 49 years old (113 patients), with a marked decrease in incidence starting at the 7<sup>th</sup> decade of life (►Fig. 3). The primary detailed location could not be determined in 55.5% of the cases, being referred only as encephalon, not otherwise specified (NOS), or brain, NOS. However, in these patients, the frontal lobe was the most affected area (8.4%), followed by the temporal lobes (6.1%), and the parietal lobes (5.4%). Neoplasms located in the cerebellum accounted for 17.9% of the cases (►Fig. 4). Astrocytic tumors corresponded to 64.3% of the cases, followed by embryonal tumors (18.2%) and ependymal tumors (7.4%) (►Fig. 5). Among the astrocytic tumors, astrocytoma, NOS (29.8%), and glioblastoma, NOS (52.4%) were the most frequent (►Fig. 6). In this group, the prevalence was higher in males (57.4% compared with 42.6% in females), with a statistically significant difference ( $p = 0.02$ ) (►Fig. 7). The number of affected patients increases with age, especially in those > 30 years old, and it decreases in people > 70 years old (►Fig. 8). Among the embryonal tumors, medulloblastoma, NOS, accounted for 71.9% of the cases, followed by neuroblastoma, NOS (8.9%), and by primitive neuroectodermal tumor, NOS (8.9%) (►Fig. 9). No statistically significant difference between genders was found in this group (►Fig. 7). Approximately 45% of the patients were aged between 0 and 9 years old. The embryonal tumors was the most frequent histological type, among all of the analyzed ones, in this age group. About 70% of all cases occurred in patients < 20 years old (►Fig. 10).



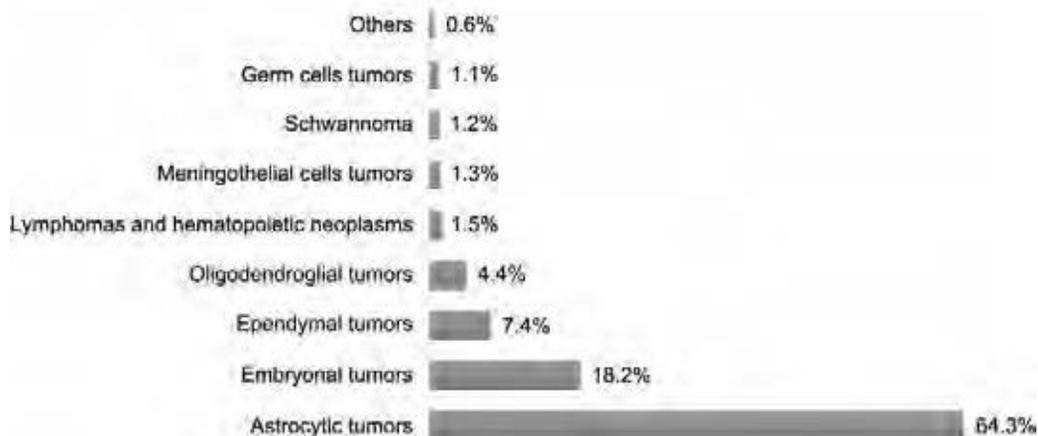
**Fig. 2** Distribution by gender of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).  $p < 0.05$ .



**Fig. 3** Distribution by age group of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).



**Fig. 4** Distribution by detailed primary location of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Abbreviations: NOS, not otherwise specified. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).

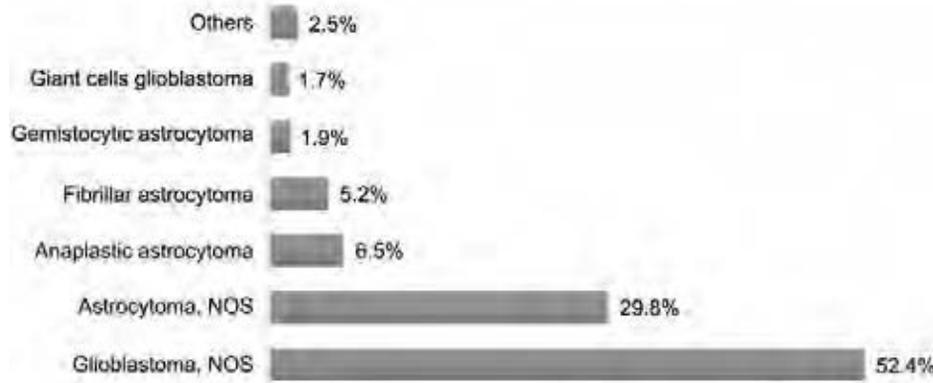


**Fig. 5** Distribution by group or World Health Organization (WHO) histological type of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).

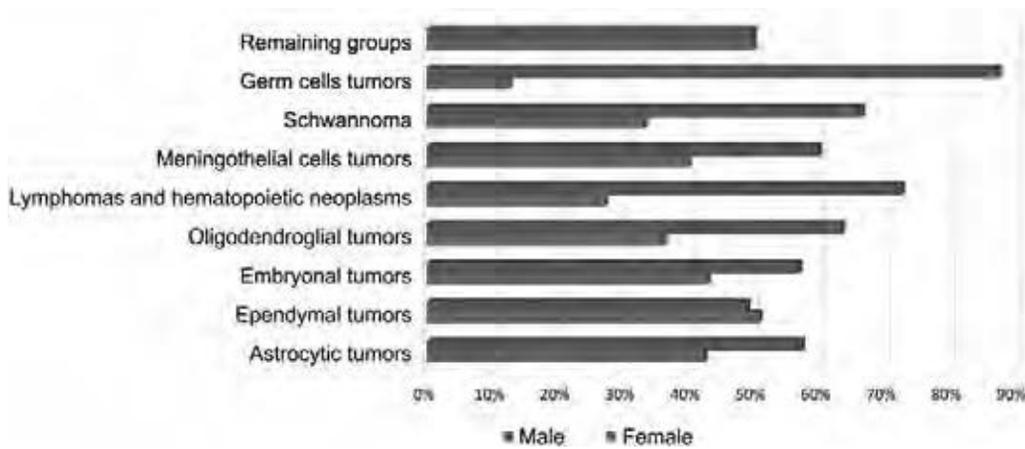
## Discussion

The establishment of effective cancer control measures requires quality information about the prevalence, the char-

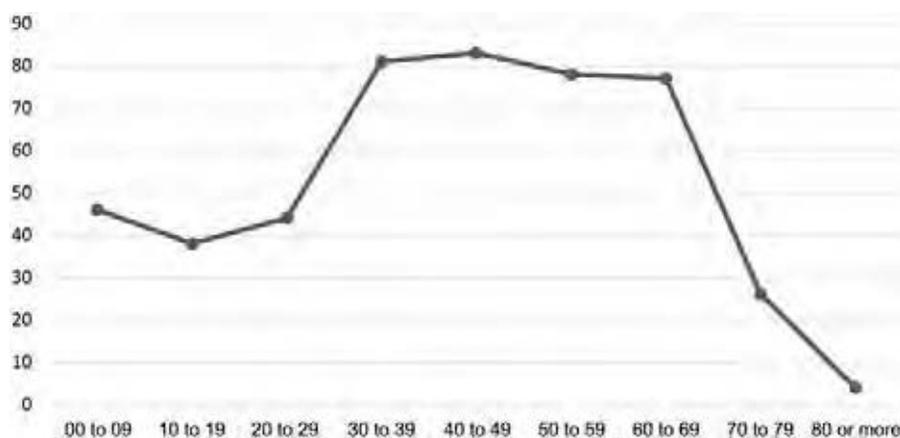
acterization, and the mortality distribution, allowing a better understanding of the disease and its determinant factors, the formulation of causal hypotheses, the assessment of technological advances applied to prevention and treatment, as well



**Fig. 6** Distribution after astrocytic tumors stratification of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Abbreviations: NOS, not otherwise specified. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).



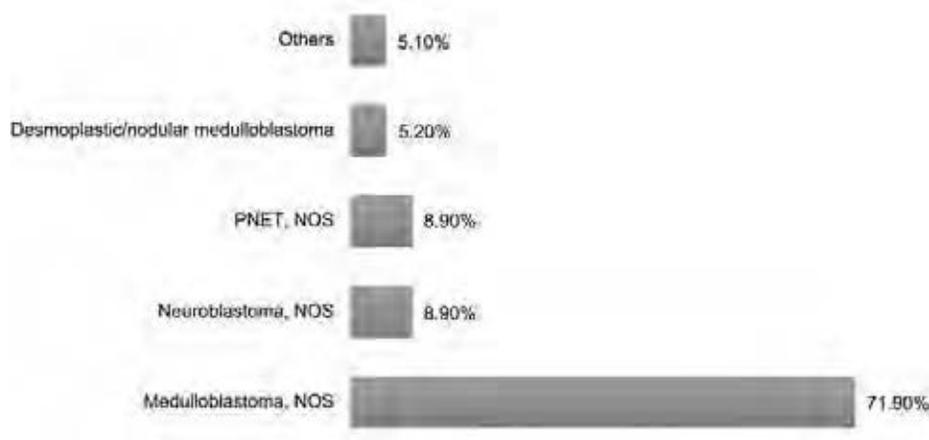
**Fig. 7** Distribution by World Health Organization (WHO) histological type and gender of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA). In astrocytic and germ cells tumors:  $p < 0.05$ .



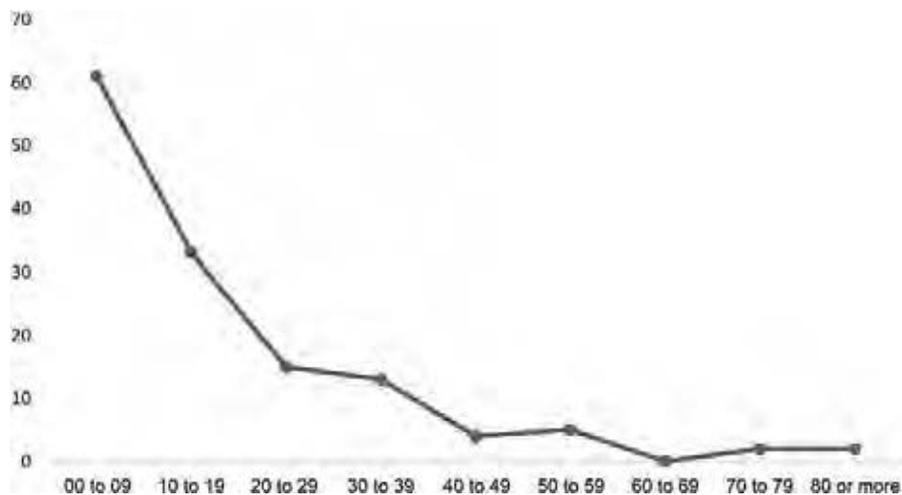
**Fig. 8** Distribution after age group stratification in the group of astrocytic tumors of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).

as an improved effectiveness in health care.<sup>7</sup> This context calls for cancer registries, which consolidate themselves as fundamental pillars of epidemiological cancer surveillance. These registries are essential sources for epidemiological and clinical

research and for the planning and evaluation of cancer control actions.<sup>8</sup> In Brazil, cancer registries are now legally protected, especially since the publication of the ordinance GM/MS No. 3,535/98, establishing mandatory RHC functioning.<sup>9</sup>



**Fig. 9** Distribution after embryonal tumors type stratification of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA from 2001 to 2013. Abbreviations: NOS, not otherwise specified; PNET, Primitive neuroectodermal tumor. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).



**Fig. 10** Distribution after age group stratification in the group of embryonal tumors of central nervous system primary malignant neoplasm cases at the Northern region reported at RHC/INCA after from 2001 to 2013. Source: Cancer Hospital Registry (Registro Hospitalar de Câncer, RHC) – Brazilian National Cancer Institute (Instituto Nacional de Câncer, INCA).

The present study found that 53.6% of the patients came from inland areas. This can be explained by the structure of the Brazilian Unified Health System (SUS, in the Portuguese acronym), based on health care networks subdivided into three levels: primary, secondary, and tertiary; tertiary care, however, concentrates in large cities. Since cancer is a complex disease, requiring the incorporation of specialized teams with higher technological density, services that treat brain tumors patients belong to the tertiary level. As a result, inland patients seek out reference hospitals in capital cities. This is more evident in poorer areas and in places with restricted access to health care services, such as the Northern region.<sup>10</sup>

The present study showed a male predominance in seven of the nine histological groups, with statistically significant differences for astrocytic and germ cells tumors. In the United States, the incidence of primary CNS tumors was higher in women (57.9%) than in men (42.1%). However, male patients are the most affected by malignant neoplasms. In addition, the incidence of tumors located in the brain, in the ventricles, and in the cerebellum was also higher in men.<sup>2</sup>

Other studies have also shown that more male patients were affected.<sup>11–13</sup> A meta-analysis revealed an incidence of 15.8 cases per 100,000 inhabitants in women and of 14.33 in 100,000 inhabitants in men; however, this difference was not statistically significant.<sup>5</sup> Our data, however, disagree with the incidence rates reported by other authors.<sup>14,15</sup> This discrepancy may be explained by the higher number of meningiomas found in these studies, since these tumors (mostly primary and benign) are more frequent in women, and the presence of female hormone receptors in this neoplasm has been demonstrated.<sup>16</sup>

The reason for the higher incidence in men compared with women has yet to be determined; however, genetic, regional, and population differences may play a role. The effects of gender in cancer occur at several levels, that is, cell transformation, tumoral tissue organization, and in the body as a whole, resulting from the influence of immunity and of sex hormones. Some genetic alterations found in glioblastomas, such as neurofibromin loss, *PTEN* and *p53* gene mutations, and platelet-derived growth factor receptor, are more frequent in men.<sup>17</sup>

Tumor location is an important factor for the evaluation and staging of neoplasms, serving as a parameter for the prognosis. The present study found the highest number of cases in the frontal, temporal and parietal lobes. At a North American study, the most frequent location was the frontal lobe, followed by the temporal lobe, overlapping brain lesion, and the parietal lobe.<sup>18</sup>

Another author showed that brain parenchyma tumors were more frequently located in the frontal lobe, followed by the cerebellum, the temporal lobe, and the parietal lobe.<sup>19</sup> Tumoral regional and behavioral differences in each population may explain data disagreements; however, more studies are required. The large number of cerebellar cases can be explained by the frequent presence of medulloblastomas, which have a unique cerebellar location.

In the present study, astrocytomas and glioblastomas were the most frequent histological types. This finding is observed in other literary studies regarding malignant primary CNS neoplasms.<sup>2,20–22</sup>

Europe had a substantial increase in the incidence of astrocytomas and of glioblastomas in patients > 40 years old, with a peak at the 6<sup>th</sup> decade of life.<sup>23</sup> Similar results were also reported elsewhere.<sup>24–26</sup> These studies, however, suggest that the decrease in the incidence starts at 80 years old. In the present study, we have verified that this phenomenon starts in the 7<sup>th</sup> decade of life.

The medical literature still debates why recent epidemiological studies show an increase in primary brain neoplasms, especially malignant ones, in the elderly. The debate is whether there is a real incidence increase, caused by risk factors not yet known, or if the phenomenon reflects the improved diagnostic techniques. The literature agrees on the need for further research. In our study, medulloblastoma, NOS, corresponded for 70% of embryonal tumor cases. This histological group accounts for most primary, high-grade tumors in children and adolescents, with medulloblastoma as the main histological type. The male gender seems to be the most affected.<sup>27,28</sup> Boys are 46% more likely to have a medulloblastoma than girls.<sup>29</sup>

Pilocytic astrocytoma remains the main primary CNS tumor in childhood and adolescence; however, it is mostly benign.<sup>30–32</sup> Thus, although it is not the most frequent tumor in children and adolescents, medulloblastoma remains one of the main brain neoplasms in this age group.

Recent years witnessed an increase in the number of primitive neuroectodermal tumor (PNET) cases. In the present study, this tumor accounted for 8.9% of the embryonal tumors. The increased incidence may be explained by changes in the classification of brain tumors in the 1980s.<sup>33</sup> Until then, medulloblastomas and PNETs were not considered distinct tumors. However, later studies showed that PNETs can reach the suprasellar region and the cerebellum; moreover, they present different molecular characteristics. Children are 4.6 times more likely to present PNETs than adults.<sup>34</sup>

It should be noted that the present study has limitations; Brazilian epidemiological information still needs to be improved. The present study was performed in this context.

Because of the wide geographic coverage, poor health care services, and poor access to hospitals in many places, the presented data may underestimate the actual situation of the Northern region.

Even though cancer treatment institutions are legally required to have RHC systems and to report their data to health departments, deficiencies are likely to occur. In the period analyzed, there were 933 cases registered in RHC-INCA. However, the INCA estimated that, in 2016 alone, there would be 420 cases in the Northern region. As such, the national database seems to be far from the reality. During the data survey for the present study, we found inconsistencies in the data of some patients, such as blank or incorrectly filled forms. Basic information, such as the detailed tumor location, was not reported in 55% of the cases from the present study. Since the RHC is a computerized system, it is possible that data from paper medical records are lost when entered in the RHC database, especially in hospitals with no electronic medical record system. Likewise, it is critical that state health departments submit collected data to the INCA. Difficulties faced by Brazilian researchers conducting an epidemiology study require special attention from health teams and managers, since these data will base the planning of health care actions.

We highlight the need to properly train health care teams in RHC in each hospital to improve its use. Coordination between federal, state, and municipal governments is required to adequately equip cancer-treating hospitals, especially those that treat CNS tumors in the Northern region.

In addition, we suggest further specific epidemiological studies, especially in Brazil, since we lack data to better understand the genesis, the diagnosis, and the prognosis of primary CNS tumors in the country.

## Conclusion

Analyzing the 742 cases, it is possible to conclude that most of the patients came from inland areas and were male. The most affected age groups were between 0 and 9 years old and between 30 and 49 years old, with a marked decrease starting at 70 years old. The frontal lobe was the most affected region, followed by the temporal and parietal lobes. Astrocytic tumors accounted for 64.3% cases, followed by embryonal tumors (18.2%), and ependymal tumors (7.4%). Among the astrocytic tumors, astrocytoma, NOS, and glioblastoma, NOS, corresponded to 82.2% of the cases. Among embryonal tumors, medulloblastoma accounted for 71.9% of the cases.

## Conflicts of Interest

The authors have no conflicts of interest to declare.

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# Hip Flexion Weakness following Transpsaos Interbody Fusion

## *Fraqueza de flexão do quadril após fusão lombar por via transpsaos*

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### Abstract

**Objective** The present work evaluated the motor deficit resulting from the psoas muscle access through the extreme lateral interbody fusion (XLIF) approach.

**Methods** This was a prospective, non-randomized, controlled, single-center study with 60 patients, with a mean age of 61.8 years old. All of the subjects underwent a lateral transpsaos retroperitoneal approach for lumbar interbody fusion with electro-neuromyographic guidance and accessing 1 to 3 lumbar levels (mean level, 1.4; 63% cases in only 1 level; 68% cases included L4-L5). The isometric hip flexion strength in the sitting position was determined bilaterally with a handheld dynamometer (Lafayette Instrument, Lafayette, IN, USA). The mean value of three peak force measurements (N) was calculated. Standardized isometric strength tests were performed before the procedure and at 10 days, 6 weeks, 3 months and 6 months postsurgery.

**Results** Ipsilateral hip flexion was diminished ( $p < 0.001$ ) at the early postoperative period, but reached preoperative values at 6 weeks ( $p > 0.12$ ). The mean hip flexion measures before the procedure and at 10 days, 6 weeks, 3 months and 6 months after surgery were the following, respectively: 13 N; 9.7 N; 13.7 N; 14.4 N; and 16 N (ipsilateral); 13.3 N; 13.4 N; 15.3 N; 15.9 N; and 16.1 N (contralateral). Neither the level nor the number of treated levels had a clear association with thigh symptoms, but hip flexion weakness was the most common symptom.

**Conclusions** Patients in the early postoperative period of transpsaos access presented hip flexion weakness. However, this weakness was transient, and electro-neuromyography use is still imperative in transpsaos access. In addition, patients must be thoroughly educated about hip flexion weakness to prevent falls in the immediate postoperative period.

### Keywords

- ▶ extreme lateral interbody fusion
- ▶ hip flexion
- ▶ spine
- ▶ weakness
- ▶ psoas

### Resumo

**Objetivo** Avaliar o déficit motor decorrente do acesso através do músculo psoas na técnica de fusão intersomática por via extremo-lateral (XLIF, na sigla em inglês).

**Métodos** Estudo prospectivo, não randomizado, controlado, único centro. Total de 60 pacientes com média de 61,8 anos. Todos os participantes passaram por fusão intersomática lombar por acesso lateral retroperitoneal com monitoração eletro-neuromiográfica. Foram operados de 1 a 3 níveis nesses casos (média de 1,4; 63%

eram de apenas um nível; 68% incluíram L4-L5). A força de flexão isométrica do quadril em posição sentada foi determinada bilateralmente com um dinamômetro de mão (Lafayette Instrument, Lafayette, IN, USA). As médias das medidas de 3 picos de força (N) foram calculadas. Testes isométricos padronizados foram realizados antes e em 10 dias, 6 semanas, 3 e 6 meses após a cirurgia.

**Resultados** A força de flexão do quadril no lado ipsilateral diminuiu ( $p < 0,001$ ) no pós-operatório imediato, mas em 6 semanas atingiu os valores pré-operatórios. As médias de pré-operatório e 10 dias, 6 semanas, 3 meses e 6 meses após a cirurgia para flexão de quadril medidas foram, respectivamente: (ipsilateral) 13 N; 9,7 N; 13,7 N; 14,4 N; 16 N; (contralateral) 13,3 N; 13,4 N; 15,3 N; 15,9 N; 16,1 N. Nem o nível nem o número de níveis tratados tiveram clara associação com sintomas na coxa, mas a fraqueza de flexão de quadril foi o sintoma mais encontrado.

**Conclusões** O pós-operatório imediato do acesso transposas apresentou fraqueza de flexão de quadril após a cirurgia. Entretanto, essa ocorrência é transiente, e o uso da eletroneuromiografia é essencial no acesso transposas. Somado a isso, a educação do paciente deve ser amplamente aplicada para alertar sobre a possibilidade de fraqueza de flexão de quadril com o intuito de prevenir quedas no período pós-operatório imediato.

#### Palavras-chave

- ▶ fusão intersomática por via extremo-lateral
- ▶ flexão de quadril
- ▶ coluna
- ▶ fraqueza
- ▶ psoas

## Introduction

Degenerative diseases of the lumbar spine can cause back pain irradiating to the legs that may compromise the health and the well-being of the patient. These diseases range from intervertebral disc degeneration to vertebral canal stenosis.<sup>1</sup> Treatments vary on a case-by-case basis, from conservative therapy in milder conditions to interbody fusion in patients with more severe radiological and clinical features.<sup>2</sup>

Interbody fusion is a surgical treatment for various degenerative diseases of the lumbar spine. Vertebral bodies can be fused through some surgical techniques, including interbody fusion by extreme lateral access via transposas or extreme lateral lumbar interbody fusion (XLIF),<sup>3</sup> which has been widely used and is highly applicable for several pathologies.<sup>3,4</sup> However, like any other surgical approach, this access has disadvantages, such as the risk of injury to the lumbar plexus, which can lead to postoperative motor and sensory deficits in the lower limbs.<sup>3</sup> Considering the inherent risks of this surgical technique, the present study aimed to evaluate the flexion strength of the hip before and after surgical intervention in cases of lumbar spine degeneration.

## Materials and Methods

This was a prospective, single-center, non-randomized study. The present study was approved by the Research Ethics Committee of the Hospital Nove de Julho, São Paulo, SP (66616317.0.0000.5455). A total of 60 patients (39 females), with a mean age of 61.8 years old, were submitted to the XLIF technique by the same spinal team. The inclusion criteria were: patients with degenerative lumbar spine submitted to interbody fusion by the XLIF surgical technique after conservative treatment failure for at least 6 months. One to 3 lumbar levels were instrumented (mean level, 1.37; 63% cases in only 1 level; 68% cases involving L4-L5). Surger-

ies were performed with retroperitoneal lateral access via the transposas approach by senior spinal surgeons.<sup>4</sup>

The hip flexion strength was evaluated in the sitting position with a handheld dynamometer (Lafayette Instrument, Lafayette, IN, USA).<sup>5</sup> Three measurements of peak force (N) were performed, and the means were calculated. These measurements were assessed in pre- and postoperative evaluations at 10 days, 6 weeks, 3 and 6 months postsurgery, both ipsilateral and contralateral to the surgical access. The strength in both sides was compared. Pain on both sides was also assessed using the visual analogue scale (VAS) questionnaire.<sup>6</sup> Descriptive and comparative statistical analyses were performed with the SPSS software (IBM SPSS, Armonk, NY, USA). The comparative statistical analyses between groups were performed using the student t-test and an  $\alpha$  value of 0.05.

## Results

Lateral interbody fusions were performed in 60 patients. The mean age was 61.8 years old (ranging from 22 to 85 years old), and 39 patients were female. Among the 60 cases included in the present study, 38 were single-level arthrodesis, and 41 involved the L4-L5 level (▶ **Table 1**).

A significant 25.4% reduction in the strength of the psoas at the instrumented side was observed between the preoperative measures and those obtained at the 10<sup>th</sup> day visit (13 N and 9.7 N, respectively) (▶ **Fig. 1**). Despite this decrease at the 1<sup>st</sup> postoperative follow-up, the force level returned to baseline 6 weeks after the surgery, and it was sustained until the 3<sup>rd</sup> month. At the end of the study, the hip flexion strength was higher than before the surgery (16 N).

The reduction of the ipsilateral psoas strength at the 1<sup>st</sup> postoperative follow-up was not observed on the contralateral side (13.3 N and 13.4 N, respectively). The strength remained constant until the 10<sup>th</sup> day visit, followed by a

**Table 1** Demographics and surgical data

Cases	60
Male/female ratio (%)	35/65
Mean age	61.8 (22–85)
Surgical time (minutes)	114.3 (30–400)
Blood loss (mL)	166 (50–1,700)
Operated levels (mean)	1.37
1 level	42 (70%)
2 levels	15 (25%)
3 levels	3 (5%)
Postoperative walking (hours)	20 (0.5–120)

20% increase in subsequent visits, reaching 16.1 N 6 months postsurgery.

No differences were observed between the strength of the ipsilateral and of the contralateral sides before the surgery ( $p = 0.71$ ). However, a 38% increase in the hip flexion strength was observed on the contralateral side (13.4 N) compared with the ipsilateral side (9.7 N) 10 days postsurgery, a difference that ceased to exist after the 6-week evaluation until the end of the study, at the 6-month visit ( $p > 0.05$ ).

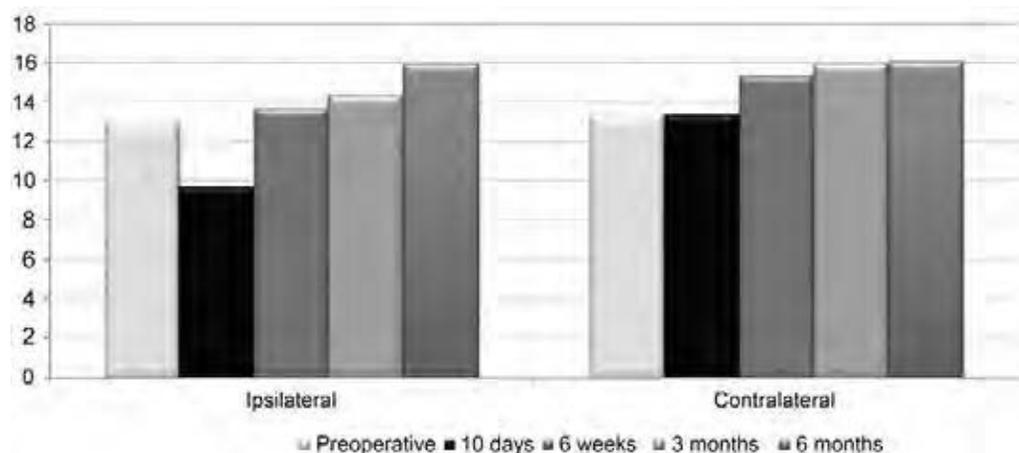
Regarding the pain in the ipsilateral and contralateral sides, it was observed that, even before the surgery, there was already 27.4% more pain on the instrumented side (6.05 and 4.75, respectively). This difference remained until the 3-month evaluation, leaving no distinction between both sides at the 6-month visit (2.77 and 2.57, respectively).

Some adverse effects were observed during the present study. In addition to the reduction in the strength of the psoas, five patients also presented with quadriceps deficit. However, all of the patients regained both psoas and quadriceps strength, with no permanent deficit. No case has progressed to reoperation due to plexopathy. In addition to lumbar XLIF, 1 patient underwent a lateral arthrodesis at the thoracic level, and, in this procedure, the intraoperative bleeding was more pronounced (1,700 mL). However, this blood loss did not cause additional harm to the patient.

## Discussion

Interbody fusion is a widely used surgical approach for the treatment of lumbar spine degenerative diseases.<sup>3</sup> To achieve this goal, extreme transpsoas lateral access has great applicability and biomechanical advantages over other options, mainly for maintaining the integrity of the posterior and anterior longitudinal ligaments, the latter the most important stabilizing ligament of the spine.<sup>7,8</sup> To reach the disc space, XLIF uses the transpsoas approach, crossing between the muscular fibers. This is one of the main spinal stabilizing muscles, and it is transected by a nerve network called the lumbar plexus. To minimize the risks of nerve damage, this technique uses a directional monitoring system that evaluates the conduction of nerve impulses to the extremities, preserving neural integrity. However, even with the navigation tool, this technique still has postoperative side effects. Reduction of hip flexion strength is the main postoperative effect of XLIF. This strength is mainly performed by the psoas muscle, which is transected by dilators for disc space access. Literature studies commonly assess the frequency of this weakness in XLIF patients.<sup>8–12</sup> This phenomenon is usually reported by the patient, and it can often go unnoticed or be overlooked. In addition, only a few papers report strength degree reductions. In a different way, the present work evaluates this parameter in a force scale in N, using measurements obtained with a dynamometer in an objective and systematic form.

In the present study, clinical parameters were evaluated in five different visits. Results showed a significant decrease of ~24% in the hip flexion strength on the ipsilateral side shortly after the surgery, returning to preoperative values 6 weeks after the procedure. Although not assessing psoas strength in the same way as the present study, Sharma et al observed a similar phenomenon of postsurgical hip flexion strength reduction, with all of the patients returning to preoperative strength values at 6 weeks.<sup>8</sup> Similarly, other studies in the literature have also shown that hip flexion weakness is a transient phenomenon.<sup>9,11</sup> This effect is transient since it is not a plexopathy, but a relaxation of the intrinsic innervation of the psoas itself, causing weakness, which is transitory, provided that the muscle is stimulated with regular daily activity.<sup>13</sup> In



**Fig. 1** Ipsilateral and contralateral mean hip flexion forces before and after extreme lateral interbody fusion (XLIF).

addition to this side effect, it was observed that 8.3% of the patients presented with quadriceps weakness after the surgery as an adverse effect, a number similar to those observed in other studies in the literature.<sup>8,9,11,12</sup>

The present study has some limitations. First, since it was conducted at a single research center, the small number of patients diminishes the power and the strength of the study. Second, the lack of data about psoas muscle retraction time from all of the patients is also a limitation, since it is not possible to directly relate this parameter with the presence of postoperative symptoms.

## Conclusion

In summary, the present study brings a quantitative measure in a direct force scale. Thus, it does not depend on the report of the patients regarding the possible side effect of decreased hip flexion strength. This is a common side effect on the ipsilateral side due to the retraction of the psoas muscle, which is, however, transient. The patient should be instructed before surgery about the possibility of this phenomenon, to avoid possible falls—resulting from this loss of strength in the first days postsurgery—that could compromise a better postoperative result. Studies with greater casuistry are necessary to better understand these side effects and the complications resulting from lateral access.

### Conflicts of Interest

The authors have no conflicts of interest to declare.

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# Usage of a Translated Version of Terminologia Anatomica in Brazil: An Examination of the Neurosurgical Literature

## *Aderência à versão traduzida da terminologia anatômica no Brasil: Análise da literatura neurocirúrgica*

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### Abstract

**Introduction** The linguistic factor may have delayed the universal adoption of the International Anatomical Terminology (IAT), which was widespread in Latin and in English only. Independent translations are possible, but they are not devoid of methodological difficulties.

**Objective** To estimate the usage of the translated version of the Terminologia Anatomica in neurosurgical articles in Brazil.

**Method** Consecutive national publications were checked for the correspondence of their anatomical terms to the following categories: IAT – Brazilian version; IAT in Latin; Nomina Anatomica – previous versions; incomplete terms; derivative terms; eponyms; neologisms; and others (misspellings and prosaic terms). The years 2014 and 2015 were chosen for analysis so that included articles were published at least 16 years after the publication of the original IAT (1998) and at least 13 years since the publication of the Brazilian version (2001).

**Results** Out of a total of 183 articles analyzed, 1,132 anatomical terms were identified, referring to 334 different anatomical structures. Most of the structures were described using terms from the Brazilian version of the IAT ( $n = 834$ ; 73.7%). Those that did not belong to or did not derive from any version of the IAT totaled 281 (24.8%). The remaining 17 terms (1.5%) corresponded to words derived or provided for

### Keywords

- ▶ anatomical terminology
- ▶ anatomical nomenclature
- ▶ terminology
- ▶ neuroanatomy
- ▶ anatomy

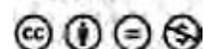
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in the Latin IAT. No association was identified between the number of authors and any category of nomenclature.

**Conclusion** Although most anatomical structures cited in Portuguese were described in accordance with the Brazilian version of the IAT, the degree of adherence was considered moderate given that about one-quarter of the terms escaped this terminology.

## Resumo

**Introdução** O fator linguístico pode ter retardado a adoção universal da terminologia anatômica internacional (TAI), que foi originalmente difundida apenas em latim e em inglês. Traduções independentes são possíveis, mas não desprovidas de dificuldades metodológicas.

**Objetivo** Estimar o uso da versão traduzida da terminologia anatômica em artigos neurocirúrgicos no Brasil.

**Método** Publicações nacionais consecutivas foram verificadas quanto à correspondência de seus termos anatômicos com as seguintes categorias: TAI – versão brasileira; TAI em latim; Nomina Anatomica – versões anteriores; termos incompletos; termos derivados; epônimos; neologismos e outros (erros ortográficos e termos prosaicos). Os anos de 2014 e 2015 foram escolhidos para análise de forma que os artigos incluídos foram publicados no mínimo 16 anos após a publicação do IAT original (1998) e pelo menos 13 anos desde a publicação da versão brasileira (2001).

**Resultados** Dos 183 artigos analisados, 1.132 termos anatômicos foram identificados, referentes a 334 diferentes estruturas anatômicas. A maioria das estruturas foi descrita usando termos da versão brasileira da TAI ( $n = 834$ ; 73,7%). Aqueles que não pertenciam ou derivavam de qualquer versão da TAI totalizaram 281 (24,8%). Os 17 termos restantes (1,5%) correspondiam a palavras derivadas ou previstas na TAI em latim. Nenhuma associação foi identificada entre o número de autores e qualquer categoria de nomenclatura.

**Conclusão** Embora a maioria das estruturas anatômicas citadas em português tenha sido descrita de acordo com a versão brasileira da TAI, o grau de adesão foi considerado moderado, pois cerca de um quarto dos termos escapou dessa terminologia.

## Palavras-chave

- ▶ terminologia anatômica
- ▶ nomenclatura anatômica
- ▶ terminologia
- ▶ neuroanatomia

## Introduction

Anatomical terminology is one of the bases for the proper description of the human body. It is used not only in educational and forensic activities but also in diagnostic and therapeutic procedures.<sup>1</sup> Anatomical terminology was created to standardize language and prevent the same structure from being cited in various ways, which could be a source of controversy or confusion.<sup>2,3</sup>

The first nomenclature was published in 1895 as *Basiliensia Nomina Anatomica*. Seven revisions followed: *Jenaiensia Nomina Anatomica* in 1935, *Parisiensia Nomina Anatomica* in 1955, and five other editions of *Nomina Anatomica* between 1960 and 1989.<sup>4</sup> They were replaced by the *Terminologia Anatomica*, which was prepared by the Federation Committee on Anatomical Terminology, and was approved and made official by the International Federation of Associations of Anatomists in 1998 in Latin and in English.<sup>5,6</sup> This last version was then translated into Portuguese and published by the Anatomical Terminology Commission of the Brazilian Society of Anatomy in 2001.<sup>7,8</sup>

The year of 2018 marks 20 years since the first publication of *Terminologia Anatomica* and is an opportune time to evaluate

how its usage has materialized among researchers and health professionals. Nomenclature modifications impose relatively abrupt changes, but the adhesion of the scientific community is progressive and influenced by the peculiarities of each area of activity. It is expected that this phenomenon does not happen in the exact same way and with the same rhythm among anatomists and practitioners. In contrast, the importance of developing a universal language is well recognized among different subgroups. Therefore, the objective of the present study was to estimate the adherence to the translated International Anatomical Terminology (IAT) by authors of neurosurgical scientific articles in Brazil. A quantitative analysis was performed to identify the terminology standards in the publications that were used to describe the central nervous system, its surrounding structures, and the cranial nerves.

## Method

This was a descriptive study in which the terms used to designate anatomical structures in each publication were checked regarding the Brazilian version of the IAT. If not adhering to the IAT, the terms were classified in alternative

nomenclature categories (described below) for accounting and statistical evaluation.

### Search and Selection Strategy

The journals consulted for analysis were national neurosurgical periodicals publishing articles in Portuguese and indexed in at least one of the following databases: MEDLINE, LILACS, SciELO, DOAJ, or LATINDEX. The years 2014 and 2015 were chosen for analysis so that the articles included were published at least 16 years after the publication of the English and Latin versions of the IAT (1998) and at least 13 years since the publication of the Brazilian version (2001).

All of the scientific articles published during the study period were listed exhaustively for each periodical following direct consultation of the electronic portal of each volume. After the identification, the full text of each document was accessed to verify the following inclusion criteria: published between January 1, 2014, and December 31, 2015, and written in whole or in part in Portuguese. We have excluded publications that did not refer to anatomical structures. In bilingual publications, only the sections in Portuguese (title and abstract) were evaluated.

### Data Collection

The included articles were submitted to a detailed full-text analysis by two independent researchers. Each document received a registration number, which was associated with descriptive data, including year of publication, languages, and number of authors. During the analysis of each document, anatomical structures belonging to the central nervous system (CNS) (including vascularization), its surrounding tissues, or the cranial nerves were identified and listed in a computerized database using Excel software (Microsoft Corp., Redmond, WA, USA). The anatomical terms used by the authors to refer to each structure were classified into one of the following categories: *IAT – Brazilian version*; *IAT – Latin version*; *Anatomical Nomenclature – previous versions*; *incomplete terms* (but in accordance with the IAT); *derivative terms*; *eponyms*; and *neologisms*.

Names that are commonly used but are not based on the anatomical conventions were categorized as *prosaic terms*. These terms, as well as those containing misspellings or not provided for in any of the previous classifications, were allocated to the *others* category. Words denoting pathological formations were not considered anatomical terms and therefore were not accounted for.

Two independent evaluators performed the consecutive categorization. Cases of disagreement were resolved through consensus. In situations in which consensus was not possible, the final category was decided by a vote through the addition of a third evaluator.

### Statistical Analysis

Frequencies and percentages were calculated for categorical variables. Mean and median values were obtained as central tendency measures for continuous and discrete quantitative variables, followed by standard deviations (SDs), confidence intervals (CIs), minimum values, and maximum or inter-

quartile intervals when pertinent. It was defined that a structure mentioned several times using the same term in the same publication would be accounted for as a single entry. Thus, each entry represented the choice of the author per anatomical structure *and* per scientific article.

In addition to the descriptive analysis, terms were grouped according to the year of their publication and to the number of authors of the article ( $\leq 5$  authors or  $> 5$  authors). For the two-tailed comparison of the frequency of a terminology category between two groups, the G-test or the  $\chi^2$  (chi-squared) test for two independent variables was applied. Statistical analysis was performed using BioEstat version 5.0 software (Brazilian Institute of Science and Technology & University of Illinois, Belém, PA, Brazil), adopting a 95% CI and a statistical significance level of  $p < 0.05$ .

## Results

### Number and Characteristics of the Publications

We have identified 217 articles from 2 scientific journals. Four were not included because they were entirely written in another language (English or Spanish). Thirty were analyzed but were excluded because they did not refer to any structure in the CNS, its surrounding tissues, or the cranial nerves.

In the 183 articles included in the study, 1,132 anatomical terms were identified, referring to 335 different structures (**Fig. 1**). Most of the articles had  $\leq 5$  authors ( $n = 141$ ; 77%), ranging from 1 to 13 with mean and median equal to 5.0.

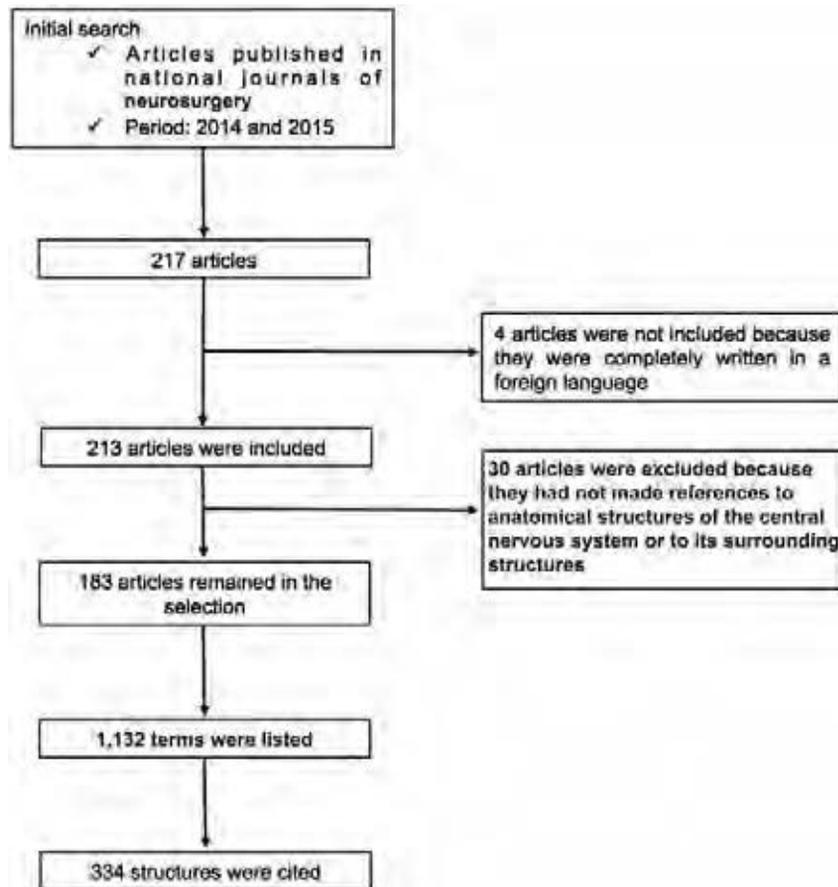
### Terminology Standards

**Table 1** summarizes the classification of terms referring to anatomical structures. Most structures were described with terms present in the Brazilian version of the IAT ( $n = 834$ ; 73.7%). Terms not belonging to or not deriving from the IAT totaled 281 (24.8%). The use of the English version of the IAT was not observed in the sample, which, due to the inclusion criteria, was made of articles written in whole or in part in the Portuguese language. No statistical association was identified between the number of authors and any category of nomenclature. The most commonly used prosaic term was scalp (*couro cabeludo*, 8.5%).

## Discussion

The establishment of terminologies is inseparable from specialized knowledge. However, historical phenomena, local traditions, innovations, and language barriers influence the use of international conventions. In the present study, the adoption of the current Brazilian version of the IAT was estimated quantitatively in neurosurgical publications. A moderate degree of adherence was observed, with about three-quarters of the anatomical terms present or derived from the IAT.

The relative adherence observed in the present study is not a universal or timeless phenomenon. In a Czech publication, Kachlik et al call attention to the fact that obsolete terms are used in clinical practice – some of them almost exclusively by clinicians – even though corresponding terms are



**Fig. 1** Flowchart, selection of scientific articles

already available in the anatomical terminology.<sup>9</sup> In spite of the peculiarities inherent to each country, the temporal nature of the transformations imposed by nomenclature revisions is tangible. Reforms tend to be adopted first by morphologists, teaching professionals, and students.<sup>9,10</sup> Only later do they spread widely among nonmorphologists and individuals with more time since training.

For monitoring, it is important to assess the evolution of adherence among health professionals. Health care activities are associated with habits of spoken and written languages that are specific to each area. Although the clinical terminology is very similar to the IAT, it is not agreed by a standard vocabulary, as is the case of anatomy.<sup>11</sup> One such example is the use of eponyms. Although abolished in the current IAT, they remain ever present in surgical and diagnostic procedures, which reinforces their daily use.

In a survey of the frequency of their use in electronic publications of the *Journal of Neurology of Spain*, Abdala listed 193 eponyms in 46 documents. The most frequent were terms related to the vascular or cerebrospinal fluid system. Expressions such as “Galeno vein” (14%), “Willis polygon” (11.9%), and “cerebral or Sylvian aqueduct” (11.4%) were found mainly in clinical or radiological reports such as those related to computed tomography (CT), magnetic resonance imaging (MRI) or ultrasound (US).<sup>12</sup>

Certain factors impact negatively or delay the adherence to the IAT, such as the difficulty in obtaining copies, the

absence of an accessible electronic database, and the tendency in certain languages or countries to avoid Latin terms.<sup>2,3,10,11,13,14</sup> Some authors consider the linguistic factor to have considerably delayed the universal adoption of the IAT.<sup>14–16</sup> Independent translations are not devoid of problems and of methodological difficulties. Although anatomical nomenclature was classically based in Latin, the previous *Anatomical Nomina* has been translated into several languages. This was not the case for the current terminology, which was widespread in Latin and in English.

Ten years after the publication of the IAT in Latin and in English, Martin et al surveyed 121 members of the American Association of Anatomists related to teaching activities.<sup>14</sup> The participants were consulted by electronic messages that contained 25 sets of expressions considered synonymous or equivalent. Among them were IAT terms, but also older and more clearly established ones. Other questions established the profile of the participants and their familiarity with the IAT. The results showed a preference for terms not belonging to the IAT in 13 of the 25 questions. The preference for the official terms was extremely variable depending on the issue: from 0.8 to 98.4%. Interestingly, more than 76% of the participants did not consider themselves familiar enough with the current terminology, and there was a slight correlation between familiarity and the belief that multiple synonyms are a source of concern in the exercise of teaching activities. The authors

**Table 1** Frequency of nomenclature standards used in 1,132 anatomical terms identified in 217 scientific articles written totally or partially in Portuguese in Brazilian neurosurgical papers

Category	n (%)	Example	Brazilian IAT
IAT – Brazilian version	834 (73.7)	<i>artéria cerebral anterior</i>	<i>artéria cerebral anterior</i> (anterior cerebral artery)
IAT – in Latin	3 (0.3)	<i>septum pellucidum</i>	<i>septo pelúcido</i>
Derivative terms	14 (1.2)	<i>base craniana</i>	<i>base do crânio</i> (skull base)
Incomplete Terms	99 (8.7)	<i>pineal</i>	<i>glândula pineal</i> (pineal gland)
Anatomical Nomenclature – previous versions	91 (8.0)	<i>tronco cerebral</i>	<i>tronco encefálico</i> (brain stem)
Eponyms	20 (1.8)	<i>polígono de Willis</i>	<i>círculo arterial do cérebro</i> (cerebral arterial circle)
Neologisms	12 (1.1)	<i>coluna espinhal</i>	<i>coluna vertebral</i> (spine)
Other	59 (5.2)		
<i>Misspelling</i>	8	<i>calda equina</i>	<i>cauda equina</i> (cauda equina)
<i>Prosaic Terms</i>	51	<i>calota craniana</i>	<i>Calvaria</i> (calvaria)
TOTAL	1,132		

Abbreviation: IAT, International Anatomical Terminology

suggest that the length of teaching experience is related to greater resistance to the new terms.

A similar survey among members of the American Association of Clinical Anatomists was conducted in 2014, more than 15 years after the publication of the IAT.<sup>11</sup> Official terms were preferred in only 13 out of 25 of the issues. Almost 25% of the participants stated that they were unfamiliar with the terminology, and 75% expressed concern about problems related to synonymy.

Some specific cases in Brazilian neurosurgical publications analyzed in the study are worthy of attention. One is the use of prosaic terms that do not belong to the IAT. The term *couro cabeludo* (Brazilian expression for scalp) is widely used in everyday language as well in scientific documents and human morphology books. Such situations lead to discussions on whether the IAT should include routinely used terms. Some authors consider neuroanatomy to be particularly responsible for advances in the description of “new structures” and, consequently, new terms that could be added to the official terminology.<sup>9</sup>

A total absence of prosaic terms describing anatomical structures is probably difficult or even impossible to achieve, given specialty traditions, so that avoiding them completely would seem awkward. This is also the case of incomplete terms, as the omission of long anatomical terms is often used as a strategy to ensure the fluidity and readability of the text.

The 2008 IAT and the Histological Terminology sections dedicated to the central and peripheral nervous systems were recently extensively revised by the Federative International Program for Anatomical Terminology Working Group Neuroanatomy, after which members of the International

Federation of Associations of Anatomists were consulted. These 2 sections were subsequently combined in 2017 in a document entitled Neuroanatomical Terminology.<sup>17–19</sup> Among the main innovations related to the CNS are the implementation of terms for various types of neurons, a better subdivision of infratentorial and supratentorial sub-arachnoid cisterns, a new version of the vascularization section, a more logical presentation of the subdivision between white and gray matter, a subdivision of the forebrain based on embryology and genoarchitecture, the use of new subdivisions of the thalamic nuclei, an update of the subdivisions and cellular types of the cerebral cortex, and an update on long association pathways. Also, the amygdaloid nucleus, the basal forebrain, and the basal nuclei are now listed under the heading *subpallium*.

The present study has limitations. Although an exhaustive evaluation was performed, a coverage period of 2 years does not enable extrapolation of results to the entire volume of national publications and all Brazilian neurosurgeons. Furthermore, neurosurgical articles published in nonspecialized journals were not included in the present analysis.

A single evaluation may omit the temporal and evolutionary character of adherence to the AIT. As the academic community appropriates the new norms and authorships are renewed, figures may change. Pertinent future research should involve longitudinal monitoring and repeated evaluations of national publications. Moreover, there is an increasing tendency to use English in national journals. This phenomenon is partly explained by the efforts of authors and editors to increase the circulation of their documents and to fulfill criteria for indexing in international databases. Thus, it seems

that it will be important to study the adhesion of Brazilian authors to the English version of the IAT in the future.

## Conclusion

According to the results of the present study, most anatomical structures cited in Portuguese in Brazilian neurosurgical journals were described according to the current Brazilian version of the Terminologia Anatomica. The degree of adherence was, however, considered to be moderate, given that approximately one-quarter of the anatomical terms used were not covered by this terminology.

### Conflicts of Interests

The authors have no conflicts of interests to declare.

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# Epidemiology and Estimated Cost of Microsurgical Treatment of Peripheral Nerve Tumor Conducted by the Brazilian Unified Health Care System (2008–2016)

## *Epidemiologia e estimativa de custos do tratamento microcirúrgico dos tumores de nervos periféricos conduzidos pelo Sistema Único de Saúde (2008–2016)*

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### Abstract

**Introduction** Primary nerve tumors correspond to ~ 5% of the soft tissue neoplasms affecting the upper limbs, with benign lesions being more frequent than malignant lesions.

**Objective** To describe the epidemiological data of the microsurgical treatment of peripheral nerve tumors performed by the Brazilian Unified Health System (SUS, in the Portuguese acronym), with the code 0403020131, from 2008 to 2016, regarding the number of annual procedures, hospital and professional expenses, the average duration of hospital stay, and the number of deaths.

**Methods** This is a descriptive epidemiological study whose data were obtained by consulting the database provided by the Health Informatics Department of the Brazilian Ministry of Health (DATASUS, in the Portuguese acronym).

**Results/Discussion** A total of 6,012 procedures were performed during the period studied, with an average of 688 procedures per year. The average hospital stay was of 2.13 days. There was no mortality rate. The average annual cost of the professional was BRL 50,091.45; and the average hospital expenses were BRL 111,887.94.

**Conclusion** The microsurgical treatment of peripheral nerve tumors is a safe surgical procedure with zero mortality rate and short hospital stay. There was no national data on tumors of this nature in the medical literature.

### Keywords

- ▶ epidemiology
- ▶ costs
- ▶ peripheral tumor nerve
- ▶ microsurgery
- ▶ tumor-like lesions

### Resumo

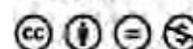
**Introdução** Os tumores primários de nervos periféricos correspondem a ~5% dos tumores de tecidos moles dos membros superiores, dentre os quais os de natureza benigna são os mais frequentemente encontrados.

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**Palavras-chave**

- ▶ epidemiologia
- ▶ custos
- ▶ tumor periférico
- ▶ nervo
- ▶ microcirurgia
- ▶ lesões pseudotumorais

**Objetivo** Descrever os dados epidemiológicos do tratamento microcirúrgico de tumores de nervos periféricos realizados pelo Sistema Único de Saúde (SUS), código 04.03.02.01-31, de 2008 a 2016, referente ao número de procedimentos anuais, às despesas hospitalares e profissionais, ao tempo de internação e ao número de óbitos.

**Métodos** Trata-se de um estudo epidemiológico descritivo cujos dados foram obtidos através do Departamento de Informática do Ministério da Saúde do Brasil (DATASUS).

**Resultados/Discussão** Um total de 6.012 procedimentos foram realizados durante o período estudado, com uma média de 688 procedimentos por ano. A permanência hospitalar média foi de 2,13 dias, com mortalidade nula durante o período do estudo. O custo médio anual com o profissional médico foi de R\$ 50.091,45, e de R\$ 111.887,94 com despesas hospitalares.

**Conclusão** O tratamento microcirúrgico dos tumores nervosos periféricos é um procedimento cirúrgico seguro. Trata-se de um procedimento com taxa de mortalidade nula e curta permanência hospitalar. Notou-se uma ausência de trabalhos nacionais com dados epidemiológicos sobre os tumores desta natureza.

**Introduction**

Peripheral nerves have as predominant supporting elements the connective tissue stroma and Schwann cells. When an axon is involved through multiple spirals by a Schwann cell, it is said to be myelinated. If a Schwann cell involves many axons simultaneously, it constitutes an unmyelinated fiber. Each nerve is composed of the epineurium, a sheath of thick connective tissue, and of the perineurium, a fibrous stroma that surrounds groups of axons forming multiple bundles of nerve fibers.<sup>1</sup>

Among the lesions that can affect the peripheral nerves, primary neural tumors represent ~ 5% of the soft tissue neoplasms of the upper limbs.<sup>2,3</sup> The main peripheral nerve tumors and tumor-like lesions are: traumatic neuromas, Morton neuromas, neural fibrolipomas, nerve sheath ganglions, benign peripheral nerve sheath tumors (PNSTs), schwannomas, neurofibromas, and malignant neoplasms called malignant PNSTs.<sup>1</sup> Benign nerve tumors are much more common than malignant ones, with schwannomas and neurofibromas representing the most frequent neoplasms.<sup>4</sup>

Peripheral nerve tumors can occur in various regions of the body, such as in the flexor regions of the extremities, in the head and cervical regions, in the retroperitoneal space, and in the dorsal roots of the spinal nerves. These regions may have some influence in terms of the technique and approach involved in the surgical treatment and of the prognosis.<sup>1,5</sup>

The objective of the present study is to describe the epidemiological data regarding the number of annual procedures, hospital and professional expenses, the mean length of hospital stay, and the number of deaths of patients admitted by the Brazilian Unified Health System (SUS, in the Portuguese acronym), in the period from 2008 to 2016, using the code of microsurgical treatment of peripheral nerve tumor/neuroma. The present study will also evaluate the impact of inflation on professional spending during the period studied.

**Methodology**

This is a descriptive epidemiological study, whose data were obtained by consulting the database provided by the Health Informatics Department of the Brazilian Ministry of Health (DATASUS, in the Portuguese acronym), at the website (<http://www.datasus.gov.br>), accessed in February 2017. The present study consisted of all cases of patients submitted to treatment with the code: *microsurgical treatment of peripheral nerve/neuroma tumor* (0403020131), from January 2008 to December 2016. It was not necessary to submit the project to the Research Ethics Committee because it is a public domain bank. Subsequently, a bibliographic research was conducted in the PubMed scientific database, searching for Brazilian and foreign publications, with the following descriptors: *peripheral nerves, tumor, schwannomas, and surgery*.

The inclusion criteria used were: indexed journals published in national and international journals, written in English and in Portuguese. The exclusion criteria took into consideration the articles whose titles and abstracts did not fit the research objectives. From the defined strategy, the bibliographic search resulted in a final sample consisting of nine publications indexed at PubMed. The studies were carefully read in their entirety and selected because they strictly met the inclusion criteria and were considered relevant to be part of the proposed study.

To calculate the impact of inflation on the surgical codes during the period studied period, the Brazilian National Extended Consumer Price Index (IPCA, in the Portuguese acronym). The IPCA was chosen because it is an index that covers families with monthly incomes ranging from 1 to 40 minimum wages, whatever the source of income, and who live in the urban areas of the regions of Brazil. For the calculation of the IPCA, the program was available online through the calculating platform of the citizen of the Central Bank of Brazil, where it was possible to post the value to be corrected, specifying the time interval, as well as the index to

be used for the correction of the values in Brazilian Reals (BRL).<sup>6,7</sup>

For the conversion of currency from BRL to US Dollars (USD), the quotation was taken on December 8, 2017,, whose exchange value was of USD 1 = BRL 3.17.

## Results

► **Table 1** shows the total number of hospitalizations for the microsurgical treatment of peripheral nerve/neuroma tumors, which occurred between January 2008 and December 2016, totaling 6,012 procedures, with an annual average of 668 procedures. There was a reduction of 9.76% in the number of treatments performed comparing the years of 2008 and 2016. Most of the treatments occurred in the southeast region (3,421 microsurgeries), and the smallest amount occurred in the northern region (172 microsurgeries), as can be observed in ► **Table 2**.

According to ► **Table 3**, the total cost of hospital services was BRL 1,006,991.50, and the annual average of expenses during the period studied was BRL 111,887.94. The average cost of hospital services ranged from BRL 94,879.01 in 2011, and the highest annual value in 2008 (BRL 150,418.43). And the highest annual value in 2008, reflecting a 36.96%. The total cost of professional services during the period studied

**Table 1** Annual number of hospitalizations in the Brazilian Unified Health System for the microsurgical treatment of peripheral nerve/neuroma tumors between 2008 and 2016

Year	Total hospitalizations
2008	697
2009	663
2010	629
2011	614
2012	709
2013	699
2014	695
2015	677
2016	629
Total	6,012

**Table 2** Distribution of the total number of microsurgical treatment of peripheral nerve/neuroma tumors between 2008 and 2016, by region of Brazil, in the Brazilian Unified Health System

Region	Number of procedures
North	172
Central-west	409
Northeast	760
South	1,250
Southeast	3,421
Total	6,012

was BRL 450,175.06, and the annual average of these expenses was BRL 50,091.45. When evaluating the value of professional services, there was an oscillation of 56.26% between 2010, the year with the lowest cost (BRL 36,401.11), and 2014, the year with the higher value (BRL 56,881.68). The highest total annual value of microsurgical treatment of peripheral nerve/neuroma tumors occurred in 2008 (BRL 203,812.29) with an annual average of BRL 161,907.39.

The mean length of hospital stay is shown in ► **Table 4**. The mean stay between the years analyzed was 2.1 days, with the lowest average in 2014 (1.8 days), and the highest in 2012 (2.6 days). There were no deaths during the period studied.

## Discussion

Schwannomas are the most prevalent peripheral nerve tumors in the upper limbs.<sup>2,3</sup> They are derivatives of the peripheral nerve sheath, develop eccentrically for nerve fibers, being typically encapsulated by the epineurium, so that they do not perform nerve invasion, only compression.<sup>2-4,8</sup> They can be diagnosed at any age, but are most common between the ages of 20 and 50 years old.<sup>4,8</sup> They usually are small, slow-growing, solitary lesions.<sup>4,8</sup> They can occur anywhere in the body; the most affected sites are the head and the neck, the flexor surfaces of the extremities, the mediastinum, and the retroperitoneum.<sup>4</sup> The histopathological characteristic of the schwannoma is a biphasic histopathological pattern with highly organized rotating cell areas and less organized, less cellular areas where the myeloid tissue predominates.<sup>4</sup> Old Schwannoma is the term used for long-standing tumors that are generally relatively large and have undergone degenerative change.<sup>4</sup> Areas of cyst formation, calcification, hyalinization, and hemorrhage may be evident in histological exams.<sup>4</sup> Nuclear atypia has been described and can occur in these tumors based on the degenerative process.<sup>4</sup> Schwannomas are usually not associated with mitotic activity and, therefore, should not be misinterpreted.<sup>4</sup> These tumors rarely undergo malignant transformation.<sup>8</sup>

Neurofibromas arise from Schwann cells and are also composed of a variety of other cells, including neuronal axons, fibroblasts, mast cells, macrophages, perineural cells, and extracellular matrix materials, such as collagen.<sup>5,8,9</sup> They are classified into localized or solitary, presenting in plexiform or diffuse forms,<sup>8,9</sup> with localized being the most common subtype.<sup>4,8</sup> The tumor tissue is closely associated with the primary nerve and is inseparable from the normal nerve.<sup>4,8</sup> The lesions of large nerves may remain confined by the epineurium and, therefore, may sometimes be covered by a true capsule.<sup>4</sup> Most of the lesions originate from small nerves and therefore occur at superficial sites.<sup>4</sup> Deep-seated tumors are seen as neurofibromas from major nerves.<sup>4</sup> Neurofibromas are observed in young patients, mainly between 20 and 30 years old.<sup>4,8</sup> They occur commonly in pictures of neurocutaneous disorders, such as neurofibromatosis type 1, and ~25% of the symptomatic neurofibromas affect the head and neck region.<sup>5</sup> They are commonly

**Table 3** Distribution of costs in Brazilian Reais and in US Dollars involved with microsurgical treatment of peripheral nerve/neuroma tumors between 2008 and 2016, in the Brazilian Unified Health System

Year	Total amount in total in Reais	Total Amount in US Dollars	Value of Hospital Services in Reais	Value of Hospital Services in US Dollars	Value Professional Services in Reais	Value Professional Services in US Dollars
2008	BRL 203,812.29	USD 64,294.09	BRL 150,418.43	USD 47,450.60	BRL 53,393.86	USD 16,843.48
2009	BRL 172,458.93	USD 54,404.44	BRL 127,696.65	USD 40,282.85	BRL 44,762.28	USD 14,120.59
2010	BRL 137,835.52	USD 43,481.23	BRL 101,434.41	USD 31,998.23	BRL 36,401.11	USD 11,483.00
2011	BRL 139,311.68	USD 43,946.90	BRL 94,879.01	USD 29,930.28	BRL 44,432.67	USD 14,016.61
2012	BRL 154,655.21	USD 48,787.13	BRL 101,633.74	USD 32,601.11	BRL 53,021.47	USD 16,726.01
2013	BRL 162,538.04	USD 51,273.82	BRL 108,475.65	USD 34,219.44	BRL 54,062.39	USD 17,054.38
2014	BRL 166,397.68	USD 52,491.38	BRL 109,516.00	USD 34,547.63	BRL 56,881.68	USD 17,943.74
2015	BRL 155,921.61	USD 49,186.62	BRL 103,032.84	USD 32,502.84	BRL 52,888.77	USD 16,684.15
2016	BRL 164,235.60	USD 51,809.33	BRL 109,904.77	USD 34,670.27	BRL 54,330.83	USD 17,139.06
TOTAL	BRL 1,457,166.56	USD 459,673.99	BRL 1,006,991.50	USD 317,662.93	BRL 450,175.06	USD 142,011.33

Abbreviations: BRL, Brazilian Reais; USD, US Dollars.

referred to as painless, well-defined, fusiform or round, slow-growing masses < 5 cm in diameter.<sup>4,8</sup> Histologically, they are composed of elongated cells mixed with undulating bundles of collagen and/or of mucin.<sup>4</sup>

Peripheral nerve tumors are usually asymptomatic, though the symptoms appear when the lesions grow and produce mass effect, being able to generate edema, paresthesia, loss of muscle strength, and pain.<sup>2,5</sup> The Tinel sign is usually present around the tumor.<sup>2</sup> In case of involvement of the vagus nerve, they can cause dysphagia.<sup>5</sup> Even with a positive sign, paresthesias and transverse mobility, the clinical identification is somewhat subjective and, for this reason, nerve tumors are often misdiagnosed because of similarities with other soft tissue tumors such as lipomas, fibromas, ganglions, or xanthomas.<sup>2</sup>

Although neurogenic tumors can be diagnosed on magnetic resonance imaging (MRI), it is difficult to differentiate between schwannomas and neurofibromas.<sup>8</sup> Schwannomas

**Table 4** Mean distribution of the days of stay in the hospital referring to the microsurgical treatment of peripheral nerve/neuroma tumors between 2008 and 2016 in the Brazilian Unified Health System

Year	Average permanence
2008	2.2
2009	2.2
2010	2.1
2011	1.9
2012	2.6
2013	2.2
2014	1.8
2015	2.1
2016	2.1

and neurofibromas share many morphological features in the MRI image.<sup>4</sup> The presence of target signal, fascicular signal, divided fat signal, and continuity with a peripheral nerve points to a neurogenic origin.<sup>8</sup> A dumbbell shape is typical of paraspinal lesions, which may dilate the intervertebral foramen of a spinal nerve.<sup>4</sup> With intermuscular localization, lesions may be surrounded by fat, a feature that can create the "split fat signal" in T1-weighted MRI images oriented on the long axis of the affected limb.<sup>4</sup> In the fluid sequences, the target signal appears with high signal intensity at the periphery and with low signal intensity at the center of the lesion.<sup>4,8</sup> The intralesional architecture with more myxoid material peripherally and fibrous tissue centrally allows the presence of the target signal.<sup>4,8</sup> This radiological sign can be observed in schwannomas, in neurofibromas, and in malignant tumors of the nerve sheath and absent in large masses and in tumors with cystic, hemorrhagic, or necrotic degeneration.<sup>4,8</sup> The fascicular sign describes multiple ringlike structures within the lesion, possibly reflecting the fascicular bundles seen in histopathological examinations.<sup>4,8</sup> A thin capsule can be identified in T2-weighted images, particularly if the tumor is surrounded by fat, which is more frequent in schwannomas.<sup>4,8</sup>

The surgical removal of schwannomas is usually sufficient to resolve the symptoms.<sup>2</sup> This type of procedure, however, can cause morbidity in 15.2% of the patients. As morbidity, it is possible to identify postsurgery the development of new neurological deficits, sensory disturbance, motor weakness, and single wound hematoma. Most of these postsurgery complications are usually temporary.<sup>10,11</sup>

The Brazilian population grew by 14.28 million inhabitants in the period between 2008 and 2016. It is noted that the annual incidence of the peripheral nerve tumors that were surgically treated in 2008 was of 3.63/1 million inhabitants, when the Brazilian population was estimated at 191.8 million, the highest during the studied period, and of 3.05/1

million inhabitants in 2016, when the population was estimated at 206.8 million, which is the lowest incidence in the study. The annual mean incidence of this microsurgical treatment was of 3.35/1 million inhabitants. A total of 6,012 procedures were performed in the period from January 2008 to December 2016.

The mean length of hospital stay varied between 1.8 days in 2014 and 2.6 days in 2012, with a general average of 2.1 days of hospitalization for the accomplishment of the microsurgical treatment. The death rate was zero during the period studied, corroborating the low mortality rate of the surgical treatment of peripheral nerve tumors.

According to the DATASUS (<http://sigtap.datasus.gov.br/tabela-unificada/app/sec/procedimento/exibir/0403020131/02/2014>), the value of transfer to the medical professional for the microsurgical treatment of peripheral nerve in 2008 was BRL 112.00. In the year 2014, after a readjustment of 50%, it became BRL 168.00. The value continues the same since the last adjustment. After correcting the value since 2008 using the IPCA index, the value should be BRL 195.81 as of December 31, 2016, and this shows a reduction of 16%.

It is worth noting that the present study under-reports patients who were submitted to treatment of peripheral nerve tumors from the private sector. Another bias of the present study is the presence of patients submitted to surgical treatment of neuromas due to traumatismos such as those found in amputations and in traumatic injuries of the peripheral nerves of the limbs.

As the epidemiological data present in the DATASUS code *microsurgical treatment of peripheral nerve/ neuroma tumor* includes both surgeries for the treatment of peripheral nerve tumors and tumor-like lesions, this consequently reduces the accuracy of the data presented in the present work.

## Conclusion

The present study reveals that the surgical procedure *microsurgical treatment of peripheral nerve/neuroma tumor* (0403020131) presented zero mortality rate during the period studied, with a relatively short hospital stay. It was

possible to observe an absence of studies with national and international data with epidemiological information on these tumors. The data obtained only reflect the patients coming from the SUS, requiring studies that involve the patients of the private system.

## Conflicts of Interests

The authors have no conflicts of interests to declare.

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# Unveiling the Insular Lobe of Reil: Neurophysiological and Anatomical Features

## *Desvendando a ínsula de Reil: aspectos anatômicos e neurofisiológicos*

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### Abstract

The insular lobe has long been investigated, from its anatomical descriptions to its neurophysiological activity. Located in a central location, the insular lobe participates in several afferent and efferent pathways, forming part of the eloquent and fundamental structures that make up the central core of the brain. The lobe of the insula has participation in language function, such as speech, sensory (e.g., taste), limbic, autonomic (visceral), also forming part of complex associative circuits, including part of the circuits of mirror neurons. Several functional descriptions attributed to the insular lobe have been made in patients suffering from cerebrovascular diseases, as well as in those with epilepsy. Much progress and many descriptions have also been made in patients with tumors. Despite much information already available about the insular lobe, it is likely that much will be discovered in the coming years.

### Keywords

- ▶ insular lobe
- ▶ central core of the brain
- ▶ insular neurophysiology
- ▶ insular neurocircuitry

### Resumo

Há muito tempo já se investiga sobre o lobo da ínsula, desde suas descrições anatômicas até sua atividade neurofisiológica. Situado em localização estratégica, o lobo da ínsula participa de diversas vias aferentes e eferentes, fazendo parte das nobres e vitais estruturas que compõem o bloco central do cérebro. O lobo da ínsula possui participação nas atividades motoras voluntárias, tais como a fala, sensitivas (por exemplo, gustação), límbicas, autonômicas (viscerais), também fazendo parte de circuitos complexos associativos, incluindo parte dos circuitos dos neurônios-espelhos. Diversas descrições funcionais atribuídas ao lobo da ínsula são feitas em pacientes vítimas de acidentes vasculares encefálicos, de neoplasias, assim como em pacientes epiléticos. Apesar de muitas informações já disponíveis acerca do lobo da ínsula, muito ainda deve ser descoberto nos próximos anos.

### Palavras-Chave

- ▶ lobo da ínsula
- ▶ bloco central do cérebro
- ▶ neurofisiologia da ínsula
- ▶ circuito da ínsula

### Introduction

Described for the first time by Johann Christian Reil in 1796,<sup>1</sup> the insular lobe was considered “hidden” by several anat-

mists and physiologists, earning this name for its Latin meaning: *insula* = *island*. Sometimes, it is completely hidden, surrounded by various brain lobes, including the frontal, the temporal, and the parietal operculum. Actually, it composes the most lateral part of the central core of the brain, which we will discuss later in the present paper.

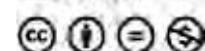
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Macroscopically, the lobe is composed of a central sulcus of the insula, dividing it into two long (posterior) gyri and three short (anterior) gyri, as well as an accessory gyrus in its most ventral portion, described by Brodmann in 1909. Currently, the concept of *central core of the brain* corresponds to several deep regions of the cerebral cortex, present in the two hemispheres, constituting a block between the brainstem and the cerebral cortex. The central core of the brain is composed of the insular surface (insular lobe), the basal ganglia, and the thalamus, thus connecting the brainstem to all of the supratentorial structures, including the extreme, external, and internal capsule. In this topography, the central core of the brain functions as a true station integrating the motor, sensorial, emotional and cognitive information. The understanding of the anatomy and of the physiology of the insula also includes its correlations with these other structures that compose the central core of the brain, elucidating a little more of its neurophysiology.<sup>2,3</sup>

According to studies performed on monkeys, the insular lobe receives afferents from the amygdaloid complex, from the dorsal thalamus, and from various regions of the cerebral lobes, particularly from the auditory areas of the parietal lobes. These afferents predominantly connect to the posterior portions of the insula, whereas the limbic afferents (the entorhinal, the perirhinal, and the posterior orbitofrontal cortexes) and cingulate gyrus connect with the anterior portions of the insular lobe. The anterior portions of the insula have reciprocal efferent pathways to the afferents; however, the same does not occur for the posterior portions of the insular lobe. Current knowledge of the human neurophysiology of the insular lobe is based on the clinical aspects observed in patients with epilepsy, tumors, and sequelae of cerebrovascular diseases that affect this region. Studies have shown that these patients often have a pattern of epilepsy that is extremely resistant to pharmacological treatment, with paresthesia (electric shock sensation, pharyngolaryngeal constriction), dysphonia, and dysarthria. In patients submitted to intraoperative cortical stimulation, stimuli to the anterior portions of the insula induced viscerosensitive symptoms (abdominal and thoracic discomfort as well as nausea), motor and somatosensitive, without specific topographic characteristics, only diffuse and nonspecific sensations.<sup>4,5</sup> More recently, in addition to the symptoms mentioned above, there are descriptions of olfactory-gustatory responses, especially to the sensation of disgust. In general, the somatosensory responses were attributed to the posterior portions of the insular lobe, whereas the viscerosensitive responses were attributed to the anterior portions. Nguyen et al also report, but to a lesser extent, vestibular symptoms and aphasia in those patients with epilepsy refractory to medications.<sup>6</sup> Pollatos et al describe, through neuroimaging, patients with autonomic symptoms, involving, among other things, changes in thermoregulation, also placing the insula in the center of homeostasis regulation.<sup>7</sup>

## Objectives

The authors describe the insular lobe anatomically, highlighting the *central core of the brain* concept, with a

functional correlation of this structure with other functional areas of the central nervous system.

## Methods

Review article using the PubMed (*National Institute of Health Database*), SCIELO, LILACS and Cochrane databases as search tools. The keywords used were: *insular lobe neurophysiology*, *insular lobe*, *insula of Reil*, *insular lobe neuroanatomy*, *central core*, and *insular lobe neurocircuitry*. The most recent articles were selected, considering their citations and their respective impacts.

## Discussion

### Central core of the brain

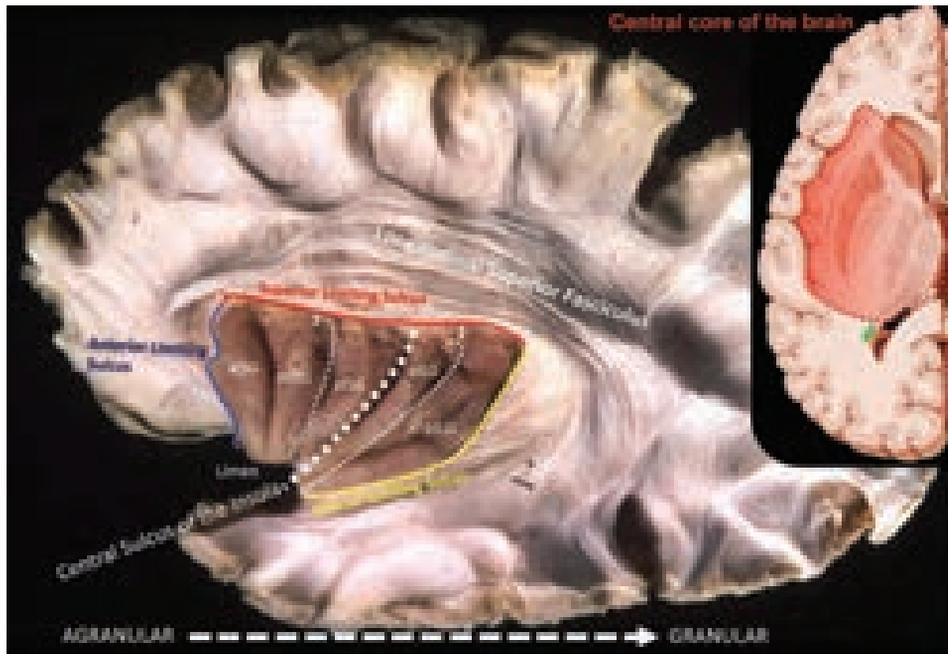
The central core stands as a block on the top of the brainstem, at the morphological center of the supratentorial compartment. This solid block includes, from lateral to medial, the insular surface, the extreme capsule, the claustrum, the external capsule, the putamen, the globus pallidus, the internal capsule, the caudate nucleus, the stria terminalis, the septal region, and the thalamus.<sup>8</sup> Also included in the concept of the central core is the medial portion of the anterior commissure and structures above and posterior to the anterior perforated substance, including the nucleus accumbens (ventral striatum), the Meynert basal nuclei, the ventral amygdalofugal pathways, and the innominate substance, corresponding to the ventral globus pallidus. Superiorly, the central core connects almost to the entire cerebral cortex through the cerebral isthmus and, inferiorly, through white projecting fibers that descend to the brainstem. There is no natural division between the central core and the brainstem.<sup>9</sup>

In the region of the cerebral isthmus, the white fibers arise from the central core radiating to all of the cerebral lobes. The isthmus is composed of the continuation of the extreme, of the external, and of the internal capsule, as well as of the extension of the anterior commissure and of the ventral amygdalofugal pathways. The cerebral isthmus is best visualized deep in the limiting sulci of the insula, where the fibers pass through a narrow space between the limiting sulci of the insula and the lateral ventricle cavity (→ Fig. 1).

The insular surface is the most lateral portion of the central core, seen through the Sylvian fissure, removing the frontal, parietal and temporal operculum. The insular lobe is limited anteriorly, posteriorly and superiorly by the anterior, posterior and superior limiting sulci of the insula.<sup>3,10,11</sup> The third ventricle, the hypothalamus, and the epithalamus are grouped together in the midline, between the central cores of the left and right hemispheres.<sup>8</sup>

### Insular Lobe

The insular surface is the most lateral part of the central core and is observed by splitting the sylvian fissure and retracting the frontotemporoparietal opercula. It is limited by the anterior, superior, and posterior insular limiting sulci (also referred to all together as the circular sulcus of



**Fig. 1** Insular lobe anatomy, and the concept of central core of the brain, with neurological and neurosurgical relevance. 1. Precentral sulcus of the insula; 2. Postcentral sulcus of the insula. Abbreviations: aILG, anterior insular long gyrus; aISG, anterior insular short gyrus; mISG, middle insular short gyrus; pILG, posterior insular long gyrus; pISG, posterior insular short gyrus.

the insula). The limits of the central core were first defined by 3 planes, each drawn from one of the limiting sulci toward the lateral ventricle, marking the anterior, the superior, and the posteroinferior limits. Each of these planes passes through the cerebral isthmus, transecting the connections that link the central core to the other cerebral lobes. The insular surface is the most lateral aspect of the central core and is encircled by the insular limiting sulci. The limen insula is a hook-like structure found at the antero-inferior vertex of the insula and is connected to the anterior limiting sulci (ALS) and to the inferior limiting sulci (ILS). The point where the limen joins the ALS, at the frontal lobe, we refer to as the frontal limen point (FLP) and consider the beginning of the ALS. This sulcus ends at its meeting point with the superior limiting sulcus (SLS), which is referred to as the anterior insular point (AIP), which is also the superior anterior limit of the SLS.<sup>8,9,12</sup> The central sulcus of the insula is visualized on the surface of the insular lobe, dividing the insular lobe into anterior and posterior areas. This sulcus is continuous and runs in the direction of the limen of the insula to the upper limiting sulcus of the insula. The anterior portion of the insula is composed of three short insular gyri most of the time. However, in some specimens, a fourth short gyrus or anterior accessory gyrus of the insula is found. A transverse gyrus of the insula is also present and communicates the anterior lower portion of the lobe with the posterior orbital gyrus and, in some cases, the accessory gyrus lies above this transverse gyrus, running along with the anterior limiting sulcus to the orbitofrontal operculum. The posterior portion of the insular lobe is composed of two long gyri, located posterior to the central sulcus of the insula.<sup>8</sup>

### Deep Structures

After removing the cortical surface of the insular lobe, there will be exposure of the extreme capsule and, more medially, of the claustrum, consisting of a thin layer of gray matter present between the extreme and the external capsules, subdivided into the ventral and dorsal portion of the claustrum. The ventral claustrum runs the depth of the antero-inferior region of the insular surface, surrounded by white matter. Its continuation in the lower sense enters the amygdaloid complex. The dorsal claustrum is located superiorly and posteriorly, and its gray layer is organized in a thin and more compact form.<sup>3,10,11,13,14</sup>

### Insular Vasculature

The insula receives its blood supply predominantly from the M<sub>2</sub> (insular) segment of the middle cerebral artery. The middle cerebral artery, first running under the anterior perforated substance and sending lenticulostriate branches, reaches the limen insula and bends posteriorly over the insular surface. At the limen, or close to it, the middle cerebral artery bifurcates (or trifurcates) into trunks; the inferior trunk is frequently located along the ILS, while the superior one courses in a posterosuperior direction toward the SLS. These trunks, as well as their several main branches, send small arteries into the insular surface to supply its cortex. These small penetrating branches do not seem to reach too deep into the insular subcortical white matter, mostly stopping before the claustrum. The venous drainage of the insular region can be defined by cisternal drainage. The cisternal vessels are composed of the insular veins that drain the insular cortical surface and are directed to the limen insula, where they join the deep sylvian vein or the deep middle cerebral vein to later empty into the

anterior part of the basal vein of Rosenthal. Occasionally, this group terminates at the sphenoparietal or cavernous sinuses.<sup>8,12,14</sup> It is well known that chronic hypertension induces pathological changes in cerebral vessels, resulting in either their occlusion or rupture, which leads to lacunar infarctions or intracerebral hemorrhages, respectively. Some striate arteriovenous malformations have been observed to receive their blood supply from these two groups of arteries: the middle cerebral artery ( $M_2$  segment) and the lateral lenticulostriate artery, which indicates the potential existence of microcommunication.<sup>12</sup>

### Functional Studies of the Insular Lobe

The vast majority of information about insular functions comes from studies following cortical lesions (in animals and humans) or even through cortical electric stimulation. Since the first studies performed by Penfield in 1955, with electric corticostimulation, the insular surface seems to provide viscerosensation and somatosensory symptoms (► **Figs. 2** and **3**).

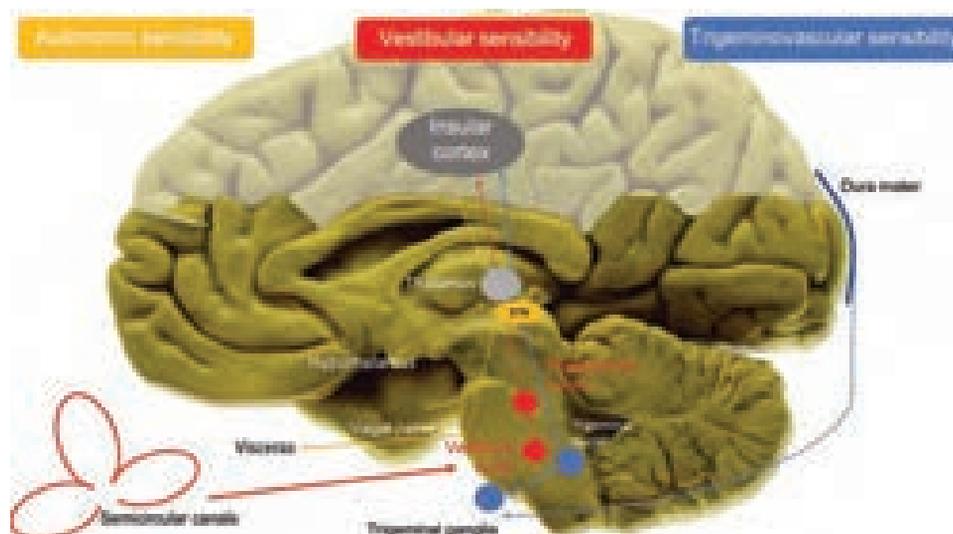
### Thermoception and Nociception

Painful stimuli appear to be triggered by electrical stimuli in the posterosuperior portion of the insular lobe, predominantly in the right hemisphere. In the most posterosuperior portions, the presence of the thermoception associated with somatosensitivity is described. The somatotopic distribution of sensitivity is as follows: upper limbs in the dorsal part, while lower limbs are more ventral. The facial sensitivity is also found in the more ventral portions.<sup>2,15,16</sup> The anterior and lateral spinothalamic pathway, after projecting to the thalamus (posterior ventromedial nucleus), is directed to the postcentral gyrus, and some fibers, such as the spinoreticular pathway (positioned closely to the lateral spinothalamic tract), pass through the posterior cortical surface of the insula and the cingulum after connections to the medullary laminae of the thalamus. The anterior and lateral spinotha-

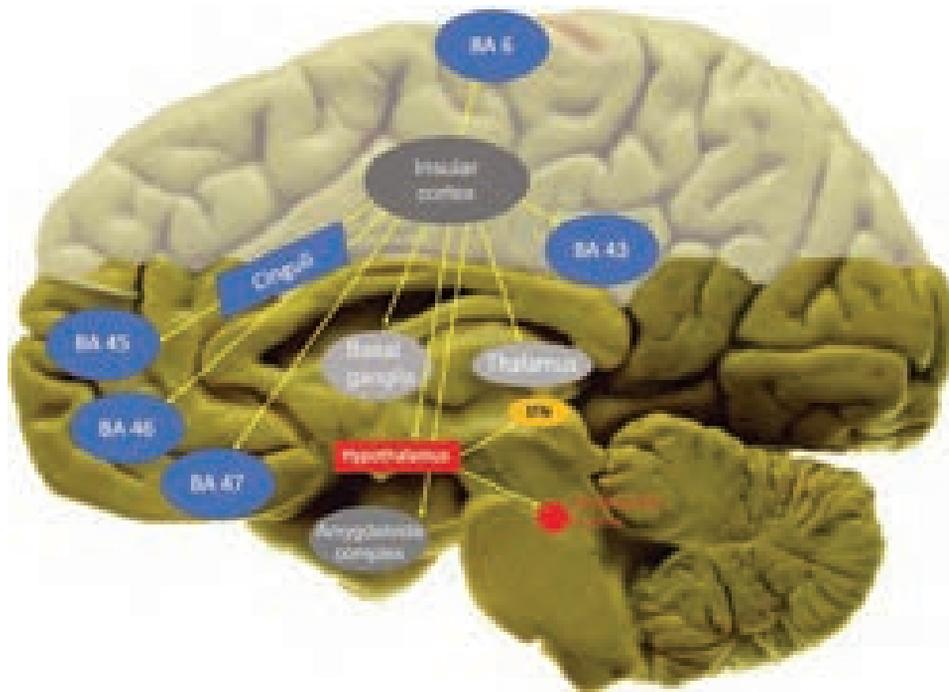
lamic tract is an important projection of the main somatosensory pathway (protopathic touch, nociception, and thermoception), thus differentiating itself from the dorsal column lemniscal pathways responsible for the epicritic and vibratory sensitivity.<sup>16</sup>

### Somatosensitivity

Since 1955, Penfield and Faulk have associated somatosensitivity to the insular lobe. Currently, with functional neuroimaging studies, Eickhoff et al, using studies on cerebral cytoarchitecture, question the actual existence of a specific insular somatosensory area, proposing that this region would consist only of the extension of the parietal operculum (post-central gyrus).<sup>17,18</sup> In a postmortem study in 10 human specimens, Kurth et al separated three cytoarchitecturally distinct area regions in the posterior insular lobe: two granular cortical regions in the posterior dorsal insula, called Ig1 and Ig2, respectively, and a dysgranular cortical area, named Idg1, located in the ventral posterior insula, associated with a great variety of cortical somatosensory electrical stimuli. The presence of the granular cortex is predominantly associated with afferent fibers, associating with somatosensitivity, thermosensitivity, and nociception. Projections of these granular and dysgranular cortices are observed and come from the limbic areas, including the amygdaloid complex and the entorhinal cortex, suggesting the presence of an important limbic-cortical connection in this location, which is associated with motor learning and with the mechanisms of memory consolidation. Thus, the regions of the granular insular cortex represent secondary somatosensory and tertiary areas.<sup>5</sup> Kahane et al<sup>19</sup> describe vestibular phenomena associated with electrical stimuli of the posterior insula, such as dizziness, nausea, vomiting and vertigo. The posterior dorsal region of the insula receives thalamic afferents (upper and lower posterior ventral nuclei of the thalamus), projecting into the parietal insular vestibular cortex, justifying these symptoms.<sup>19</sup>



**Fig. 2** Some afferent pathways related to the insular lobe. The three main systems that deserve attention: autonomic sensitivity pathways; trigeminal sensitivity, and vestibular sensitivity pathways. Abbreviation: STN, solitary tract nucleus. Modified from Borsook D et al.<sup>22</sup>



**Fig. 3** Some efferent pathways related to the insular lobe. Abbreviations: BA, Brodmann Area; STN, solitary tract nucleus. Modified from Borsook D et al.<sup>22</sup>

### Visceroreception

Visceral symptoms are extremely relevant when electrically stimulating the insular cortex. Some authors further propose that the ventral insula could be an extension of the gustatory cortex. Stimuli to these regions give rise to gastrointestinal motor activity, transmitted by vagal nerves bilaterally; once both vague nerves have been damaged, complete suppression of insular gastrointestinal motility control is observed. In addition, the gastrointestinal afferent pathways ascend to the thalamus (medial and parvocellular ventroposterior nuclei), projecting to the granular and dysgranular cortices. Functional neuroimaging studies revealed that after feeding, healthy volunteers had increased metabolic activity in their cortices. Ischemic insular lesions in the regions attributed to visceroreception commonly occur with dysphagia alone, producing evidence for the hypothesis that the solitary tract nucleus (vagal afferent) protrudes mainly to the insular cortex.<sup>2,19,20</sup>

### Taste

The gustatory pathway begins in the chord tympani nerve projecting from the solitary tract nucleus (STN), located in the brainstem, to the hypothalamus and to the ventromedial parvocellular nucleus of the thalamus. It is anatomically accepted that the gustatory area is not part of the primary somatosensitive cortex, where the tongue is included. Taste appears to be situated in the central portion of the insula (being more posterior in humans compared with primates, for example), receiving ventromedial thalamic afferences as well as afferent frontal and parietal operculum.<sup>21</sup> Epileptic seizures involving these regions of the insula commonly occur with unpleasant sensations involving taste, often

described as a “disgust” sensation, also associated with facial expression. However, insular lesions cause gustatory phenomena in the ipsilateral hemitongue to the injured cortex; however, complete deficits (in both hemitongues), with an inability to recognize the food, can be triggered by involvement of the left insular lobe.<sup>2</sup>

### Anterior Part of the Insular Lobe

Initially, we must consider that the central sulcus of the insula does not divide it functionally, only anatomically. The central sulcus of the insula does not coincide with microscopic alterations that justify its functional modification.<sup>22</sup> In animals, the anterior agranular insula seems to have relevant connections with the limbic and paralimbic cortices, especially the connections between the anterior insula and the cingulate gyrus. However, Craig et al<sup>23</sup> propose that the anterior insula would participate in important integrative functions related to self-recognition or interoception (“self-circuit,” like a neural map of body states), attention, emotion, subjective sensations such as joy, sadness, pleasure, and pain beyond their own perception. The anterior part of the insular lobe also seems to be able to codify pleasurable behaviors, justified by their connections to the nucleus accumbens, as part of the reward circuitry.<sup>1</sup> In general, the anterior portion of the insula can be considered as an associative area between homeostatic functions (including autonomic control) and emotional processing.<sup>23</sup> Disconnections between the brainstem and the cortex of the anterior insula, or even between the anterior insula and the claustrum, can severely compromise the level of consciousness.<sup>24</sup> Atypical cytoarchitectural presentations of the anterior insula are observed in individuals diagnosed with autism spectrum disorder (ASD), thus showing the importance

of this anterior region also in social interaction.<sup>25</sup> Thus, the empathy circuit was attributed to the connections between the anterior insula and the medial portion of the cingulate gyrus: individuals considered as being more empathic have a higher density of neurons in this circuit, whereas individuals with ASD, for example, have a marked reduction in the cellularity of this region.<sup>15</sup>

#### Social Behavior and Mirror-neurons Circuit

Empathy is defined as the ability to understand the other, associated with visceral emotional reactions, correlated to the various situations in which the other is living; it is literally the situation of understanding another person by putting oneself in the “skin of the other.” Trying to understand the neural circuitry correlated to empathy, Rizzolatti et al described in 1992 the circuit of mirror-neurons, initially observing primates. Rizzolatti et al reported that when they visualized motor acts performed by their peers, monkeys activated brain circuits, as if they were performing the motor act themselves. Besides the motor aspect, this neurocircuit brought to the neurosciences the idea that socialization depends on one’s own perception of the situation of the other, being able to put oneself in the place of the other. This circuit has great activation only in the fact of imagining the pain that the other could be feeling, anticipating it only by imagination itself.<sup>26,27</sup> As already mentioned, the insula is strategically located in a region where it facilitates the interconnection between the limbic system and the thalamus. The bodily representation itself, that is, the “self-circuit”, is present in the insula, including unpleasant sensations like disgust. The circuits of mirror-neurons also have associative areas in the insular lobe, being intensely activated at the moment of imitation of a movement only initially visualized. Singer et al observed that personally experienced pain sensation activates the insular lobe, as well as the observation of someone experiencing the painful sensation, with only the imagination of the pain sensation.<sup>28,29</sup>

#### Clinical Aspects Related to the Insular Lobe

Several neurological and psychiatric diseases present involvement of the insular lobe, given its behavioral, motivational and cognitive functions. Patients with depression, anxiety and schizophrenia have a significant reduction of gray matter present in the insular lobe. Psychopathy and eating disorders such as anorexia (autoimage disorder) also find correlation with functional insular abnormalities. In neurology, the involvement of facial expression of emotions can be found in Huntington chorea and in multiple sclerosis, whereas in Alzheimer dementia, the impairment of the “self-circuit” becomes more evident.<sup>1</sup> Neuroimaging studies associate dysfunctions of the insular cortex with motivational deficits, as well as with the behavior of addiction. Substance abuse appears to directly activate the insular cortex, thereby modulating its behavioral aspect involved in the reward circuit. Injuries in these regions often occur with anhedonia.<sup>30</sup> Currently, transcranial magnetic stimulation has been performed and maybe will reveal new aspects of the insular neurophysiology.<sup>31,32</sup>

## Conclusions

Topographically located in a strategic position, the insular lobe has several interactions with sensorial modalities, integrating the limbic system to the somatosensitive and the motor systems. Definitely, the anterior insula can be considered an area of tertiary association, participating in a coupled way in the interpretation of information and decision-making, together with the parieto-occipitotemporal areas, the limbic association area, and the prefrontal area (ventromedial, dorsolateral and orbitofrontal). Ischemia affecting the insular lobe may mimic lacunar infarcts, affecting purely motor or sensory functions, often involving functional areas of language, which, when associated with vertigo, nausea, taste disorders and auditory processing, speak strongly in favor of lesions of the insular lobe.<sup>32</sup>

#### Conflicts of Interests

The authors have no conflicts of interests to declare.

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# Deep Brain Stimulation for the Treatment of Juvenile Parkinsonism: Case Report

## *Estimulação cerebral profunda no tratamento do parkinsonismo juvenil: relato de caso*

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### Abstract

Juvenile parkinsonism (JP) is characterized by the clinical manifestation of Parkinson syndrome before the age of 21 years old. This entity is often associated with genetic mutations. After all the possibilities of clinical treatment have been exhausted, surgical treatment is recommended, performed via deep brain stimulation (DBS) in the subthalamic nucleus (STN) or in the internal segment of the globus pallidus (GPi). The present study aimed to report the case of a patient with JP who underwent DBS in the STN with good clinical response. Neuromodulation via DBS is an option for the treatment of JP. However, since this entity is very rare, and even more peculiar when treated surgically, more studies are necessary to evaluate DBS used to control refractory manifestations and levodopa-induced dyskinesia, as well as surgical complications that may occur, aiming to gather more knowledge of the surgical management of JP. Despite the dysarthria after the DBS, the patient presented a satisfactory response regarding the symptoms, corroborated by the Parkinson's Disease Questionnaire (PDQ-39) score, which was 61.19% before the procedure, and decreased to 21.05% 14 months after the DBS.

### Keywords

- ▶ juvenile parkinsonism
- ▶ deep brain stimulation
- ▶ subthalamic nucleus
- ▶ neurosurgery
- ▶ dysarthria

### Resumo

O parkinsonismo juvenil (PJ) é caracterizado pela manifestação clínica da síndrome de Parkinson antes dos 21 anos de idade. Esta entidade está frequentemente associada a mutações genéticas. Depois de esgotadas todas as possibilidades de tratamento clínico, institui-se o tratamento cirúrgico, que é realizado via estimulação cerebral profunda (ECP) no núcleo subtalâmico (NST) ou no globo pálido interno (GPi). O presente trabalho teve como objetivo relatar o caso de um paciente com PJ que foi submetido à ECP no NST com boa resposta clínica. A neuromodulação via ECP apresenta-se como uma opção de tratamento para o PJ. Contudo, em decorrência da raridade desta entidade, que se torna ainda mais peculiar quando abordada cirurgicamente, são necessários mais estudos que avaliem a ECP utilizada para controlar as manifestações refratárias e a discinesia induzida por levodopa, assim

### Palavras-chave

- ▶ parkinsonismo juvenil
- ▶ estimulação cerebral profunda
- ▶ núcleo subtalâmico
- ▶ neurocirurgia
- ▶ disartria

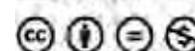
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como as complicações cirúrgicas que podem ocorrer, com o intuito de propiciar maior conhecimento sobre o manejo cirúrgico do PJ. Apesar da disartria apresentada após a ECP, o paciente teve resposta satisfatória em relação aos sintomas, o que foi corroborado pela pontuação na escala Parkinson's Disease Questionnaire (PDQ-39), que era 61,19% antes do procedimento e, 14 meses após a ECP, diminuiu para 21,05%.

## Introduction

Parkinson disease (PD) is a syndrome characterized by at least two of the following symptoms: bradykinesia, rigidity, tremor, and postural changes.<sup>1-3</sup> Idiopathic Parkinson disease is the idiopathic form of this condition, corresponding to between 75 and 80% of the cases.<sup>4</sup> The incidence of PD increases with age, and this syndrome is not common in individuals < 50 years old.<sup>4,5</sup> In cases the aforementioned clinical manifestations develop between the ages of 21 and 40 years old, the syndrome is called early-onset PD (EOPD),<sup>4,6,7</sup> whereas in cases they appear before the age of 21 years old, it is called juvenile parkinsonism (JP).<sup>4,6-8</sup>

Juvenile parkinsonism is a rare condition that is more commonly reported in association with a family history of PD.<sup>4,7</sup> Many patients may have atypical manifestations. Although some cases are typically idiopathic, secondary causes prevail. Therefore, the idiopathic origin is a very rarely described cause.<sup>7,8</sup>

In PD, surgical treatment is an option after the control of symptoms with medication has failed and levodopa-related complications have appeared. Currently, deep brain stimulation (DBS) of different surgical targets is the treatment employed for PD. The present study aimed to report the case of a patient who underwent a surgical treatment using DBS to control JP.

## Case Report

A male patient presented with micrographia at 14 years old and asymmetric resting tremor of upper limb extremities at 19 years old, predominantly on the left side, associated with rigidity and bradykinesia, but he did not have any nonmotor symptoms associated to PD. At 21 years old, he was diagnosed with JP in its primary form, after other secondary and family forms were not confirmed. The patient had no family history of any neurological diseases.

The patient showed a good response to levodopa until he was 30 years old. However, in the following 10 years, he experienced progressive levodopa-induced dyskinesia and an increased frequency of on/off periods, which significantly compromised his quality of life, in spite of the optimized clinical treatment, with 1600 mg/day of levodopa, 4 mg/day of pramipexole, and 400 mg/day of amantadine.

At 40 years old, that is, 19 years after the patient was diagnosed with JP, he underwent DBS with electrodes placed bilaterally in the subthalamic nucleus (STN). Before the surgery, his quality of life was measured using the Parkinson's Disease Questionnaire (PDQ-39), and his score was 61.19%.

Three months after the surgery, the patient presented with dysarthria, which persists until now. Nevertheless, he showed significant improvements on dyskinesia and on/off motor fluctuations, leading to a decrease in the doses of the medications. After the procedure, the clinical treatment was changed, and the patient now takes 600 mg/day of levodopa, 3 mg/day of pramipexole, and 300 mg/day of amantadine. Furthermore, 14 months after the DBS, a global improvement was confirmed based on a decrease of his PDQ-39 score to 21.05%.

## Discussion

Juvenile parkinsonism is an entity described as clinically and pathologically distinct from EOPD,<sup>4,7,8</sup> not only because of the age groups they affect. Different from PD and EOPD, the histopathological finding of Lewy bodies is rare in JP.<sup>4,6,7</sup> This is the reason the term JP is adopted in these cases instead of juvenile PD, in spite of the similar clinical manifestations.<sup>6</sup>

In cases of JP, both the association of the disease with genetic mutations and the presence of a family history are well described.<sup>4,6</sup> It is known that mutations in the parkin gene are among the most common causes of JP.<sup>6,7,9,10</sup> In some studies, alterations in the *PARK2* gene, related to the chromosome 6q25-27, were found to be responsible for autosomal recessive JP in 50% of the family cases analyzed,<sup>10,11</sup> as well as in between 15 and 77% of the sporadic cases in patients ≤ 21 years old.<sup>11,12</sup> The *PARK2* gene is related to the parkin protein, an enzyme associated with the removal of damaged mitochondria, which also promotes the degradation of  $\alpha$ -synuclein.<sup>7,9,10</sup> Two other mutations correlated with JP are found in the chromosome 1p in the *PARK6* and *PARK7* genes, although they may also be involved in other clinical entities in patients > 40 years old.

The patient of the present case report had no family history of motor dysfunction and did not present with atypical manifestations that could suggest other forms of parkinsonism. These findings indicate a sporadic case of JP. The clinical manifestations that corroborate the similarity of the evolution of his condition to that of PD were the presence of bradykinesia, rigidity, and resting tremor, the prospective findings of asymmetry of these manifestations, the progressive nature of the disease, and its good response to levodopa, in association with the exclusion of other diseases. These findings fulfill the clinical diagnostic criteria of PD described in the literature.<sup>13,14</sup> The regular clinical treatment of PD can also be used to treat patients with JP,<sup>4,7</sup> and it was chosen for the patient of the present case report. Nonetheless, the

patient presented with levodopa-induced motor fluctuations and dyskinesia in less than 5 years. In cases of parkinsonism with onset at ages < 40 years old, early motor complications such as on/off periods and severe dyskinesia are observed.<sup>15,16</sup>

In treatment-refractory cases, DBS treatment is the surgical option for patients with PD.<sup>17</sup> The STN, the internal globus pallidus (GPi), and the ventral intermediate thalamic nucleus (VIM) are described as surgical targets in DBS.<sup>17,18</sup> The STN and the GPi have more clinical applications and, comparatively, no differences were found between their efficacy.<sup>19,20</sup> Deep brain stimulation treatment targeting the VIM has been proven to control tremors; however, since it has been considered insufficient to control other motor symptoms, it is not indicated for the treatment of PD.<sup>18,19</sup>

The depletion of dopamine in the nigrostriatal pathway leads to a decrease in the inhibitory activity of striatum units along the indirect pathway, resulting in the inhibition of the external globus pallidus (GPe) and in the consequent disinhibition of the STN, which in association with a decrease in the excitatory influence on striatal units along the direct pathway, strongly disinhibits the GPi,<sup>21,22</sup> having as a final consequence a lower thalamocortical activity.<sup>22</sup> In patients with JP, DBS still needs more systematic studies. In advanced PD, STN hyperactivity is a landmark for neurophysiological mapping. Nonetheless, so far, no studies have detailed whether the same happens in cases of early onset PD.<sup>23</sup>

In the present case, the surgical target was the STN, which can be divided into three parts: dorsolateral, ventromedial, and medial.<sup>24</sup> The dorsolateral portion houses the sensorimotor region, and its precise location is of great importance to achieve the best clinical response regarding the motor symptoms of PD patients and to reduce the adverse effects on cognition and behavior.<sup>25</sup> In the treated patient, several landmarks<sup>26</sup> were used in an integrated way via magnetic resonance imaging (MRI) to locate the dorsolateral portion of the STN and to optimize the implantation of the electrodes.

Although the patient of the present case report presented with early clinical manifestations, he underwent DBS only 20 years after the diagnosis, already in an advanced phase of the disease and with serious motor complications. In the few cases of JP reported in the literature, the eligibility for DBS has been greatly based on the phenotype of the patients and on their responsiveness to levodopa.<sup>27-29</sup> For instance, the treatment of a 14-year-old patient, with several motor fluctuations, using bilateral DBS in the STN, has been described. Satisfactory results were obtained with this treatment for JP, even though he became refractory to levodopa and had complications in less than 5 years.<sup>27</sup> In another case, a 26-year-old JP patient, with onset estimated 8 years before, underwent DBS having GPi as a target. The outcome was satisfactory, since the symptoms were controlled.<sup>28</sup> Advanced PD patients, with disease onset 40 years before, have benefited from DBS in the STN, with a decrease in dyskinesia and motor fluctuations.<sup>29,30</sup>

In spite of the paucity of controlled trials, in some studies it has been reported that DBS can be a good option for the

treatment of JP, because it permits the decrease in the dosage of the medication and improves dyskinesia and on/off periods, as observed in the patient of the present case report.

Aiming to measure the perception of the patient of his quality of life before and after the surgical procedure, the PDQ-39 was employed. It is a self-report questionnaire, composed of 39 items, which encompasses 8 daily life dimensions (mobility: items 1-10; activities of daily living (ADL): items 11-16; emotional well-being: items 17-22; stigma: items 23-26; social support: items 27-29; cognition: items 30-33; communication: items 34-36; bodily discomfort: items 37-39), intended to measure the quality of life of PD patients during the last month, using a 5-point Likert scale (never = 0; occasionally = 1; sometimes = 2; often = 3; always = 4).<sup>31</sup> The highest scores are correlated with a worse quality of life.

Before the surgical procedure, the global percentage of the patient was 61.19% and, 14 months after the DBS, it decreased to 21.05%. These results, comparable to literature data,<sup>32-34</sup> showed a significant decrease in the score, pointing to an improvement in his quality of life. However, the oral communication of this patient was impaired after the DBS, due to dysarthria that occurred as a complication of the procedure. Before the surgery, it was 3.4% of the total score, whereas, 14 months after the DBS, it was 44.5%. In another study, after DBS treatment for PD, 12.8% of the patients had oral communication impairment.<sup>35</sup> Due to the rarity of JP cases, no cases of patients that underwent DBS and had dysarthria, similarly to the case presented here, were found. Further studies should be conducted to follow the long-term repercussions and the efficacy of DBS in JP patients.

## Conclusion

Few cases of DBS in JP patients have been reported. The patient of the present case report had a good response to DBS with the electrodes placed bilaterally in the STN, with a global improvement in his quality of life, even with dysarthria as a surgical complication. The present study evidences that DBS in the STN is an option for treating JP.

## Conflicts of Interests

The authors have no conflicts of interests to declare.

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# Oligodendroglioma with Sarcomatous Transformation: Case Report and Literature Review

## *Oligodendroglioma com transformação sarcomatosa: Caso clínico e revisão da literatura*

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### Abstract

#### Keywords

- ▶ oligodendroglioma
- ▶ oligosarcoma
- ▶ co-deletion 1p/19q
- ▶ spinal metastasis

#### Resumo

#### Palavras-chave

- ▶ oligodendroglioma
- ▶ oligossarcoma
- ▶ co-deleção 1p/19q
- ▶ metástases raquidianas

Oligodendrogliomas are infiltrative tumors of the central nervous system considered to be morphologically stable and to offer a better prognosis. Here, we describe the case of a 36-year-old man with an initial diagnosis of oligodendroglioma, World Health Organization (WHO) grade II, who presented transformation to a sarcomatous form, while maintaining the oligodendroglial component as well as the genetic characteristics of the initial tumor without having undergone any complementary treatments previously. Despite the favorable genetic characteristics, the tumor presented poor response to complementary treatments, and rapid progression, including spinal metastasis.

Oligodendrogliomas são tumores infiltrativos do sistema nervoso central considerados morfológicamente estáveis e com melhor prognóstico. Aqui, descrevemos o caso de um homem de 36 anos com diagnóstico inicial de oligodendroglioma grau II OMS, que apresentou transformação para uma forma sarcomatosa, mantendo as características genéticas do tumor inicial, sem ter sido submetido a tratamento complementar. Apesar das características genéticas favoráveis, o tumor apresentou má resposta aos tratamentos e progressão rápida, incluindo com metastização raquidiana.

### Introduction

Oligodendrogliomas are infiltrative tumors of the central nervous system (CNS). They constitute between 4 and 5% of the primary CNS tumors and ~ 4 to 15% of the glial tumors.<sup>1</sup> Among the glial tumors, oligodendrogliomas have always been considered morphologically stable.<sup>2</sup>

Gliosarcomas are rare primary tumors of the CNS, histologically composed of glial and sarcomatous cells, with the glial component being, in most cases, astrocytic.<sup>3</sup>

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We describe here a case of an oligosarcoma (World Health Organization [WHO] grade III<sup>4</sup>), originating from an oligodendroglioma (WHO grade II<sup>4</sup>), in a patient not previously submitted to complementary treatments.

### Case Report

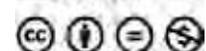
A 36-year-old man presented with an onset of convulsive crisis in 2014. He underwent brain computed tomography (CT) that showed a left frontal intra-axial lesion with calcifications in the interior (▶ **Fig. 1**) and brain magnetic resonance imaging (MRI) that showed a lesion at the same location with hyperintensity T2 and without contrast

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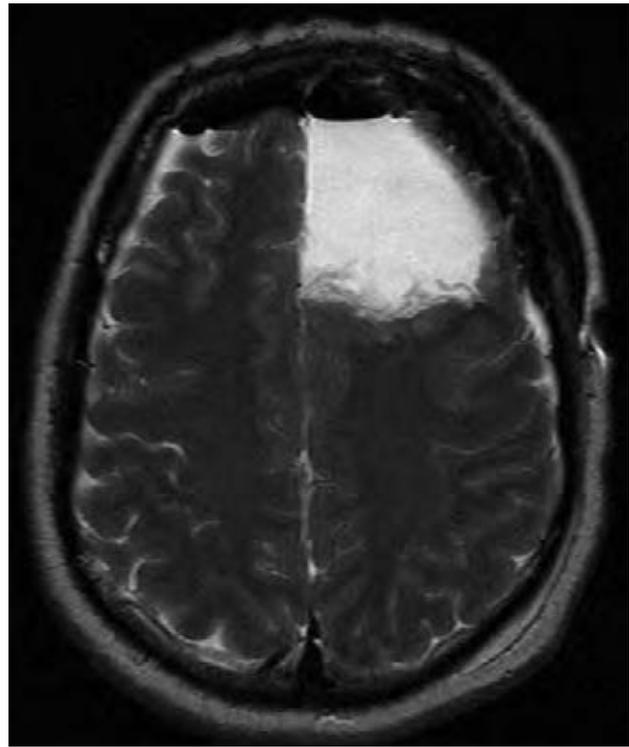
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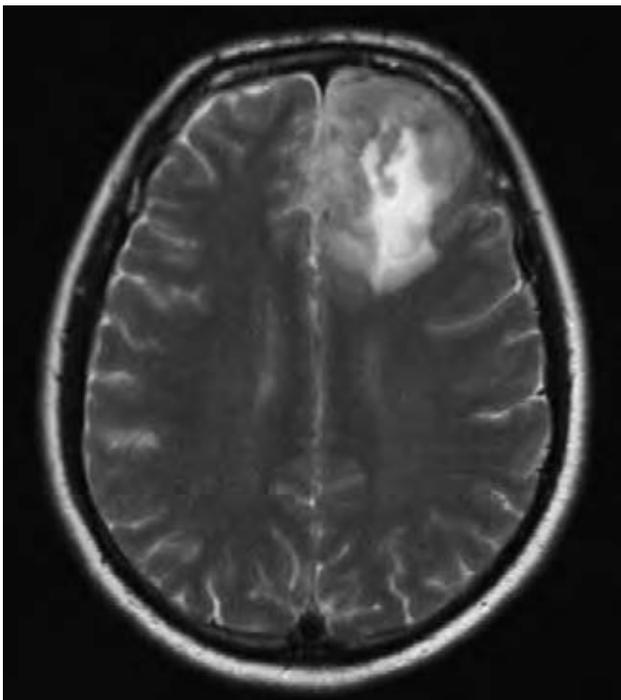


**Fig. 1** Computed tomography scan showed a left frontal lesion with calcification.



**Fig. 3** First surgery postoperative magnetic resonance imaging showed gross total resection of the tumor.

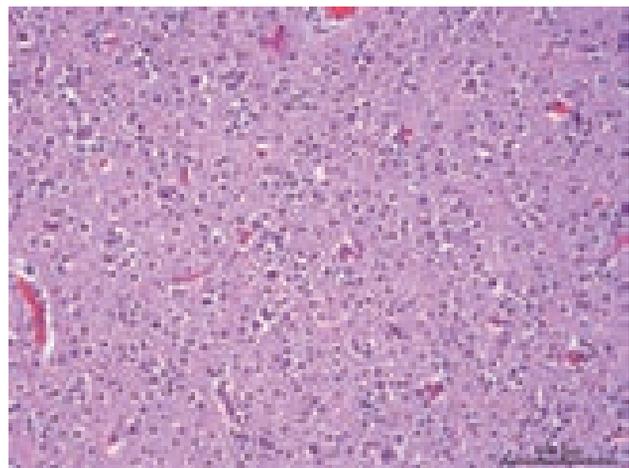
enhancement, suggestive of low-grade glioma (►**Fig. 2**). The patient was submitted to craniotomy and gross total resection (GTR) of the lesion (►**Fig. 3**). Histological analysis showed a tumor with round nuclei cells and perinuclear



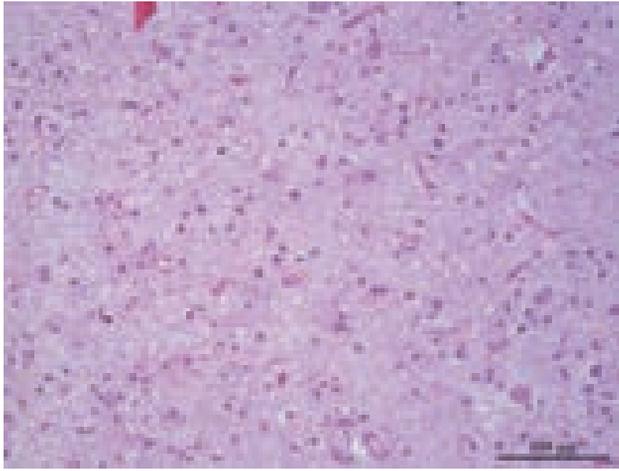
**Fig. 2** T2-weighted magnetic resonance imaging showed a hyperintensity left frontal lesion.

halos, with extensive areas of calcification. It also showed absence of mitoses, necrosis and showed low nuclear proliferation index. The lesion was classified as oligoastrocytoma WHO grade II, according to the WHO tumor classification of 2007<sup>5</sup> (►**Figs. 4 and 5**), and the tumor was positive for the presence of 1p/19q co-deletion. The patient remained under clinical and imaging surveillance, without any complementary treatment.

Two years later, in imaging control, tumor growth was recorded in the posterior and medial portion of the surgical site. An MRI showed characteristics similar to the initial lesion, namely without evidence of contrast enhancement,

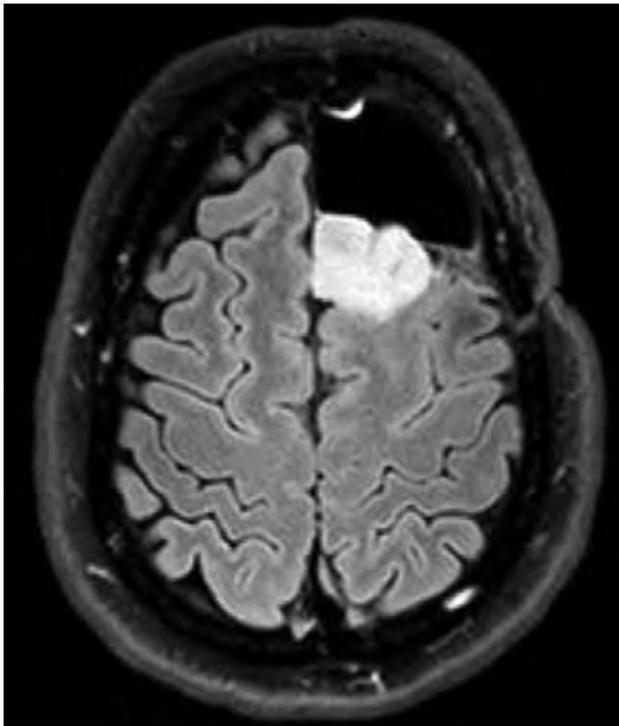


**Fig. 4** HE staining showed round nuclei with a perinuclear halo, and a low proliferation index.



**Fig. 5** HE staining showed round nuclei with a perinuclear halo, and a low proliferation index.

with spontaneous hyperintensity in T2-fluid-attenuated inversion recovery (FLAIR) weighted imaging (►Figs. 6 and 7). The patient underwent a new surgical procedure with GTR of the recurrent tumor (►Fig. 8). The neuropathological characteristics confirmed that it was an oligodendroglioma WHO grade II, isocitrate dehydrogenase (IDH-1) positive (according to the WHO classification of 2016 CNS tumors<sup>4</sup>), maintaining the presence of 1p/19q co-deletion and  $\alpha$  thalassemia/mental retardation syndrome X-linked (ATRX) mutation positive (►Figs. 9 and 10), now with a moderate nuclear proliferation index. The patient was once again referred to clinical and imaging surveillance, not considering



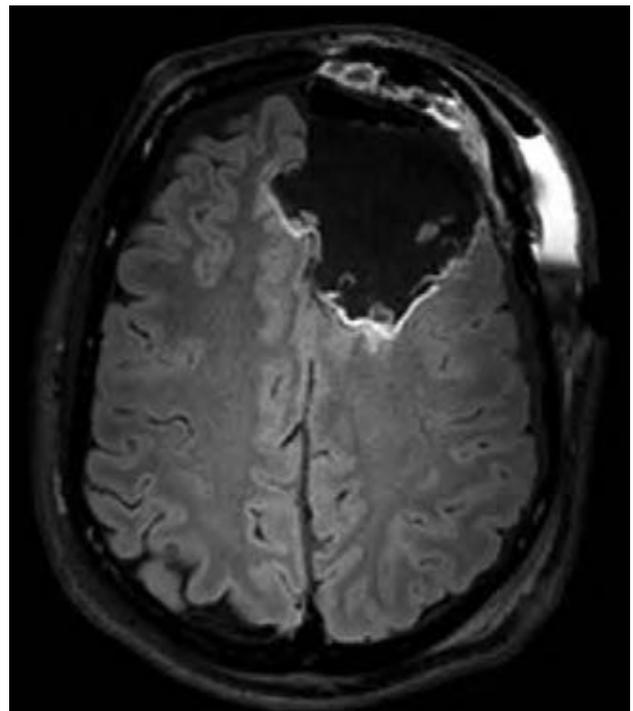
**Fig. 6** T2-fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging showed a tumor recurrence in the posterior and medial walls of the surgical site.



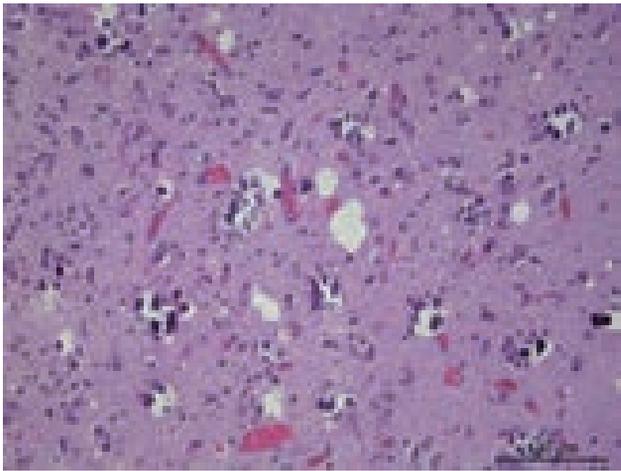
**Fig. 7** T2-fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging showed a tumor recurrence in the posterior and medial walls of the surgical site.

indications for adjuvant treatments, given the clinical, imaging, anatomopathological and degree of tumor removal obtained.

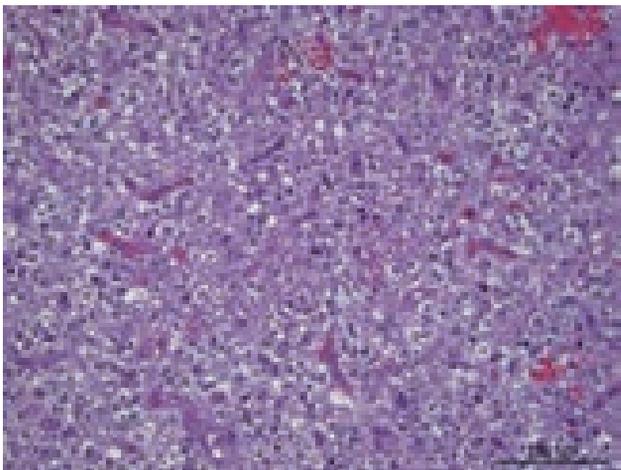
After 12 months, new MRI control showed tumor recurrence, now with a contrast enhancement area, suggesting



**Fig. 8** Second surgery postoperative magnetic resonance imaging showed gross total resection of the tumor.



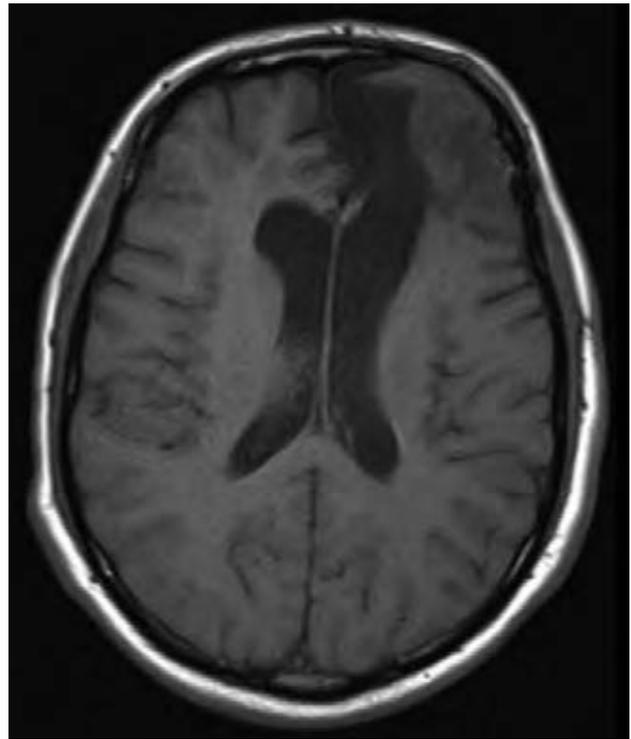
**Fig. 9** Hematoxylin & eosin staining with de same initial type of cells, but with a moderate proliferation index.



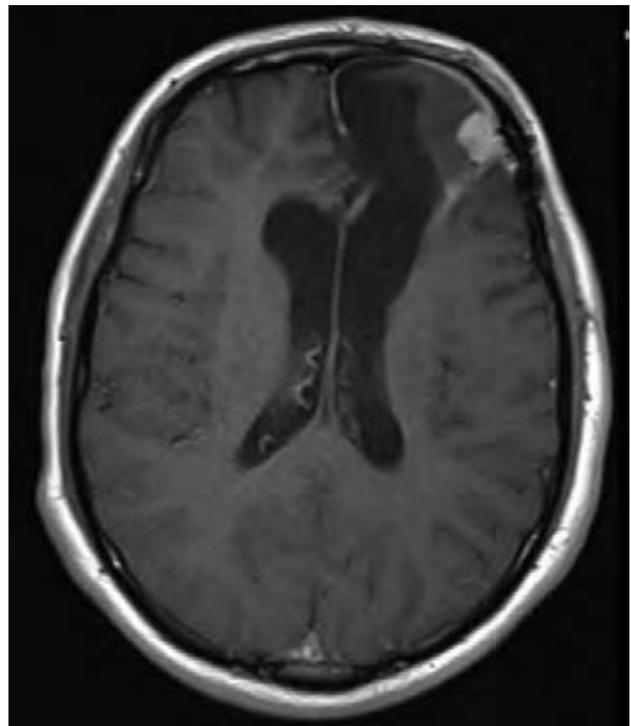
**Fig. 10** Hematoxylin & eosin staining with de same initial type of cells, but with a moderate proliferation index.

possible dedifferentiation (►Figs. 11 and 12). It was operated on with a GTR of the lesion, as evidenced by postoperative brain CT (►Fig. 13). The histological analysis showed a very cellular tumor, with frequent mitoses, areas of extensive necrosis, with rounded nuclei cells, and an evident cytoplasm and a fasciculated aspect. The immunohistochemistry study showed positivity for glial fibrillary acidic protein (GFAP), and the sarcomatous portion was strongly positive for vimentin. It maintained positivity for IDH-1 and ATRX and presented a very high proliferation index. The neuropathological study now showed the occurrence of sarcomatous transformation, maintaining the oligodendroglial component (oligosarcoma WHO grade III<sup>4</sup>), (►Figs. 14–20). With this new histological diagnosis, the patient started complementary treatment according to the Stupp protocol. He completed 30 sessions of radiotherapy (2 Gy/session, totalizing 60 Gy), with concomitant temozolomide 75 mg/m<sup>2</sup> for 7 days/week, for 6 weeks. Following this, the patient underwent 1 cycle of adjuvant temozolomide 150 mg/m<sup>2</sup> for 5 days.

During the 2nd cycle of chemotherapy, the patient presented with a new neurological deficit, a motor apraxia of the



**Fig. 11** T1-weighted magnetic resonance imaging showed a left frontal lesion with contrast enhancement (12).



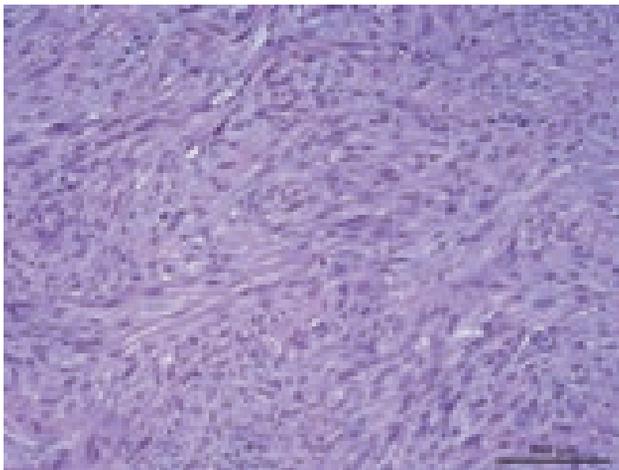
**Fig. 12** T1-weighted magnetic resonance imaging showed a left frontal lesion with contrast enhancement (12).

right upper limb. Another MRI was undertaken, which revealed tumor progression, with a recurrent tumor in the surgical site and still with left hemispheric meningeal dissemination revealing an extra-axial mass with contrast

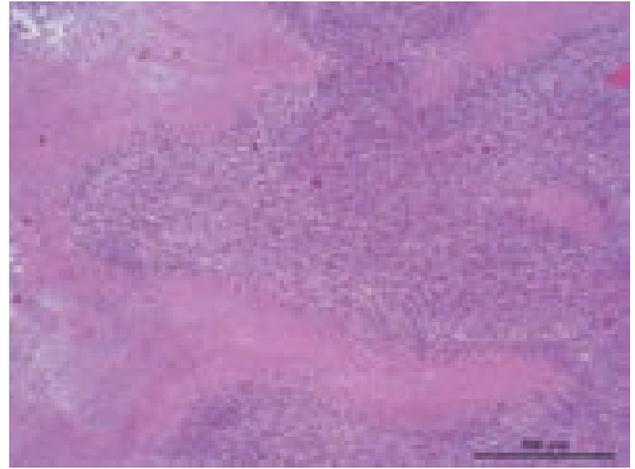


**Fig. 13** Third surgery postoperative computed tomography scan without contrast, showed gross total resection of the tumor.

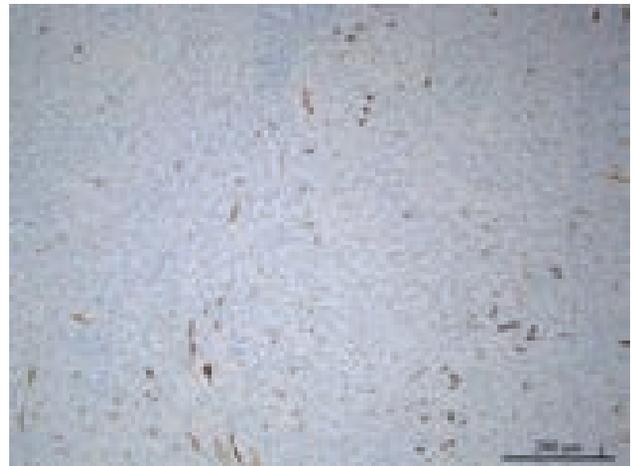
enhancement (► **Figs. 21** and **22**). A new surgical intervention with GTR of the various relapsed lesions, as documented by postoperative MRI (► **Figs. 23** and **24**), was performed. The parietal lesion corresponded to a mass of hard consistency and was extra-axial (► **Figs. 25** and **26**). The neuropathological study of the various lesions was shown to be the same sarcomatous tumor, and complementary treatment with 2nd line chemotherapy (irinotecan with bevacizumab) was initiated. At the end of 2 months of treatment, the patient exhibited severe cervical radiculopathy without relief with



**Fig. 14** Hematoxylin & eosin staining showed hypercellular tumor with round nuclei, with a perinuclear halo, with frequent mitoses and areas of extensive necrosis.



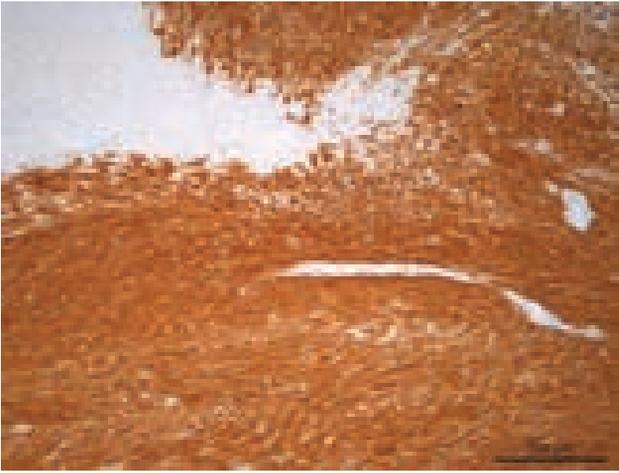
**Fig. 15** Hematoxylin & eosin staining showed hypercellular tumor with round nuclei, with a perinuclear halo, with frequent mitoses and areas of extensive necrosis.



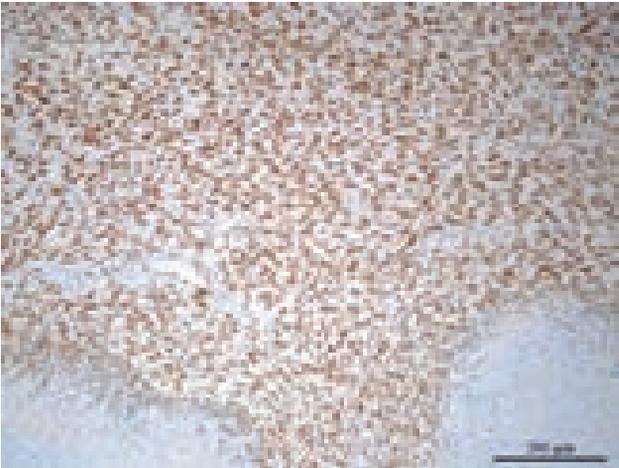
**Fig. 16** Immunohistochemistry showed positive staining for glial fibrillary acidic protein (GFAP).



**Fig. 17** The tumor showed positive staining for vimentin in the sarcomatous portion.



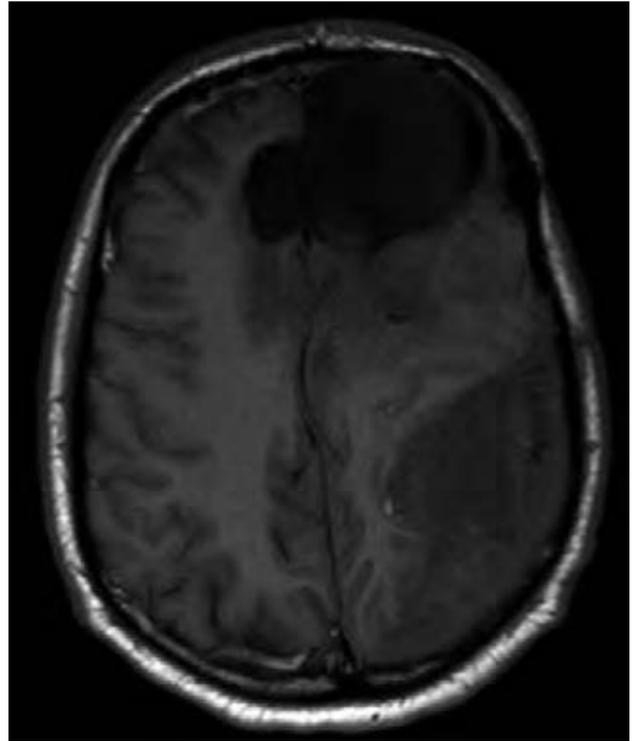
**Fig. 18** The tumor was positive for IDH-1.



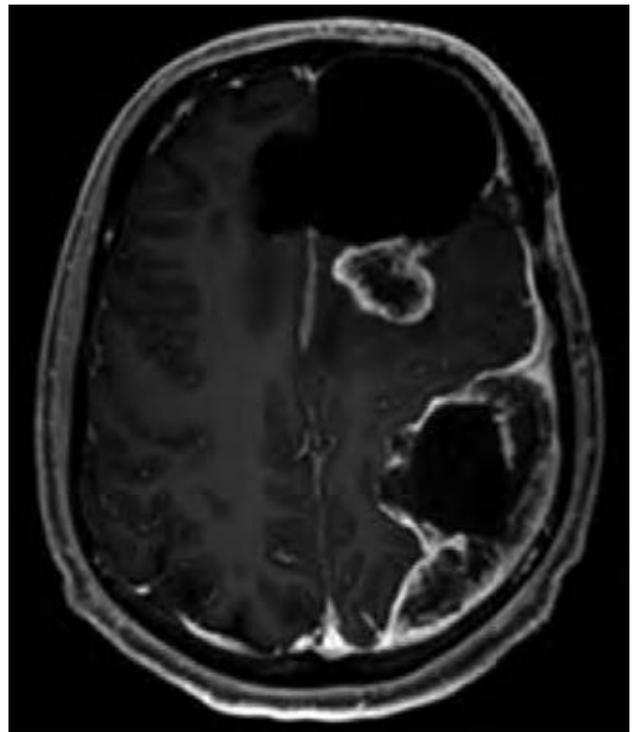
**Fig. 19** The tumor was positive for  $\alpha$  thalassemia/mental retardation syndrome X-linked (ATRX).



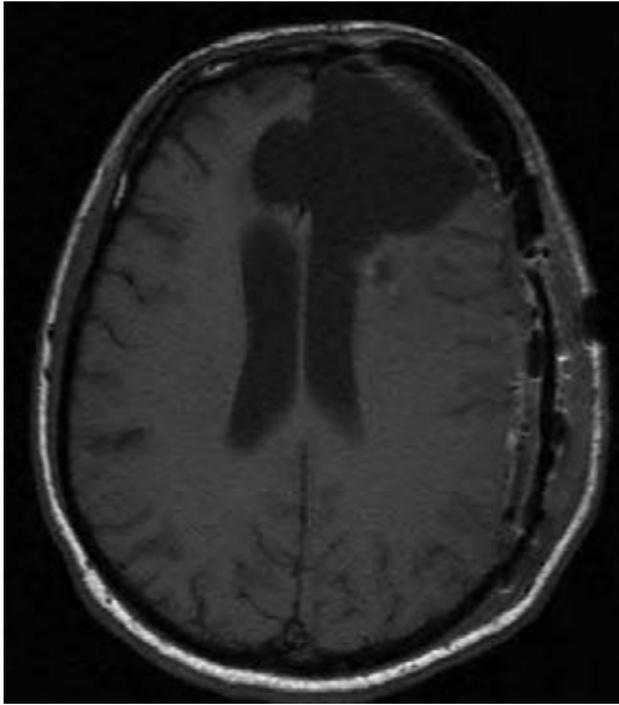
**Fig. 20** The proliferation index was very high.



**Fig. 21** T1-weighted magnetic resonance imaging showed a recurrent tumor in the surgical site and a left hemispheric extra-axial mass with contrast enhancement (22).

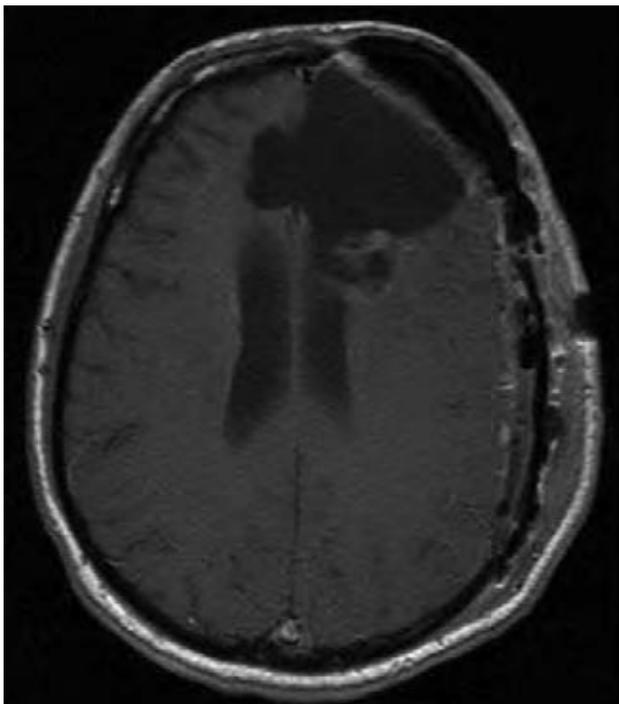


**Fig. 22** T1-weighted magnetic resonance imaging showed a recurrent tumor in the surgical site and a left hemispheric extra-axial mass with contrast enhancement (22).

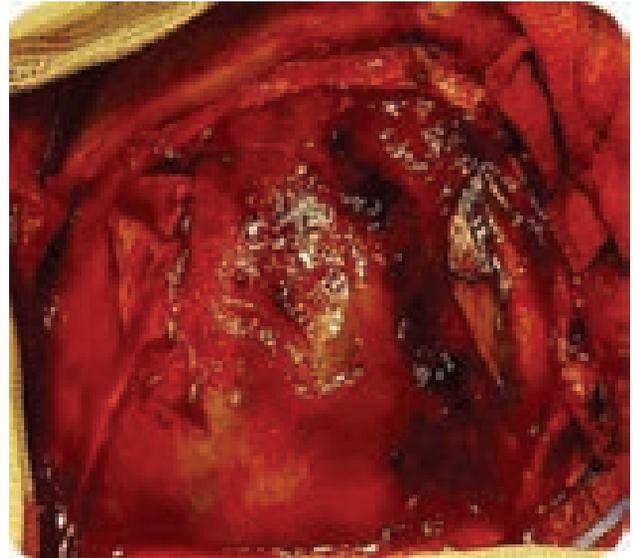


**Fig. 23** Fourth postoperative magnetic resonance imaging showed gross total resection of the tumor.

analgesics, and was submitted to lumbar puncture, which showed the presence of malignant cells in the cerebrospinal fluid. He underwent a neuro-axis MRI, which showed stability of the disease at the brain but extensive spreading to the medulla with intradural and extramedullary lesions at the



**Fig. 24** Fourth postoperative magnetic resonance imaging showed gross total resection of the tumor.



**Fig. 25** Intraoperative images showing a lesion of hard consistency, with plane of separation of adjacent brain parenchyma.

cervical and medullary cone levels (→ **Figs. 27, 28**). He started treatment with 3rd line chemotherapy (lomustina 90 mg/m<sup>2</sup> for 6 days/week, for /6 weeks, and bevacizumab 10 mg/kg for 2 days/week for 2 weeks. The patient also performed 5 sessions of radiotherapy directed to the cervical lesion (5 fractional radiotherapy sessions totaling 20 Gy). Currently, the patient has a 4-year overall survival, with Karnofsky performance status of 60% and eastern cooperative oncology group (ECOG) performance status 2.



**Fig. 26** Intraoperative images showing a lesion of hard consistency, with plane of separation of adjacent brain parenchyma.



**Fig. 27** Cervical T1-weighted magnetic resonance imaging with contrast showed a lesion at C6-C7 level with contrast enhancement.



**Fig. 28** Lumbar T1-weighted magnetic resonance imaging with contrast showed a lesion at T12-L1 level with contrast enhancement.

## Discussion

Isocitrate dehydrogenase-mutated and 1p/19q-positive co-deletion oligodendrogliomas are considered slow-growing tumors with a better prognosis than the other gliomas.<sup>5</sup> The appearance of sarcomatous tumors at sites of oligodendrogliomas resection in patients not undergoing further chemotherapy and/or radiotherapy treatments is very rare.<sup>6</sup> Although in most cases the glial component of the sarcomatous tumors is astrocytic, the literature describes several cases of gliosarcomas in which the glial component is oligodendrocytic.<sup>2,3,6-10</sup>

Here, we describe the case of an oligodendroglioma WHO grade II with 1p/19q co-deletion, IDH-1 and ATRX mutation, with initial GTR, not subjected to complementary treatments, and which was dedifferentiated to the sarcomatous form. The tumor always maintained the same genetic characteristics. Despite the presence of predictive factors of better prognosis, the tumor displayed poor response to radiotherapy and chemotherapy, and even presented spinal metastasis. Although several cases have been described in the literature of oligodendrogliomas with transformation to the sarcomatous form (oligosarcoma), research performed in PubMed and Google Scholar reveals only one case with spinal metastasis.<sup>8</sup>

Most cases with extracranial dissemination are associated with extensive progression of the brain tumor;<sup>11</sup> yet, in this case, the spread occurred with stability of the brain lesions.

This case describes a patient with a low-grade glioma, with most of the predictive factors of better prognosis (age < 40 years, total initial resection and favorable genetics), in whom progression occurred rapidly and with refractoriness to complementary treatments.

## Conclusion

Despite all the good prognostic factors present in this clinical case and the absence of previous adjuvant therapies, the tumor was dedifferentiated to a malignant form, quickly and without any warning signs.

This leads us to conclude that there is a need for further studies that may indicate new prognostic factors, such as imaging, anatomopathological and genetic characteristics that help us understand which tumors will dedifferentiate more quickly and which may respond better to complementary treatments.

## Conflicts of Interest

The authors have no conflicts of interest to declare.

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# Endovascular Treatment in the Subclavian Steal Syndrome

## *Tratamento endovascular na síndrome do roubo da subclávia*

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### Abstract

#### Keywords

- ▶ subclavian steal syndrome
- ▶ subclavian artery stenosis
- ▶ subclavian steal phenomenon
- ▶ cerebrovascular circulation
- ▶ atherosclerosis
- ▶ syncope

When the proximal occlusion or stenosis of the subclavian or of the brachiocephalic artery may require distal arterial filling through reversal flow from the vertebral artery, causing clinically significant blood supply reduction to the brainstem, it is called subclavian steal syndrome (SSS). We report a 54-year-old male patient who presented with multiple episodes of syncope and vascular claudication due to right SSS. He underwent an angioplasty, evolving with complete improvement of the symptoms. We review the clinical presentation, the diagnostic methods, and the treatment options of the disease.

### RESUMO

#### Palavras-chave

- ▶ síndrome do roubo da subclávia
- ▶ estenose da artéria subclávia
- ▶ fenômeno do roubo da subclávia
- ▶ circulação cerebrovascular
- ▶ aterosclerose
- ▶ síncope

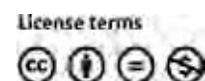
A oclusão ou estenose da artéria subclávia ou do tronco braquiocefálico pode exigir fluxo reverso pela artéria vertebral para seu enchimento distal, causando sintomas devido à redução de fluxo no tronco encefálico. A isso se dá o nome de síndrome do roubo da subclávia. Relatamos o caso de um paciente de 54 anos com múltiplos episódios de síncope e claudicação vascular por síndrome do roubo da subclávia à direita, submetido a angioplastia e evoluindo com melhora completa dos sintomas. Revisamos a apresentação clínica, métodos diagnósticos e opções de tratamento da doença.

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## Introduction

Subclavian steal syndrome (SSS) is a vascular process resulting from an ipsilateral occlusion or from a significant proximal stenosis of the subclavian or braquiocephalic arteries.<sup>1</sup> With pressure impairment in the subclavian artery, blood flows from the contralateral vertebral artery to the basilar artery and descends retrogradely through the ipsilateral vertebral artery, leading to collateral circulation to the upper limb.<sup>1</sup>

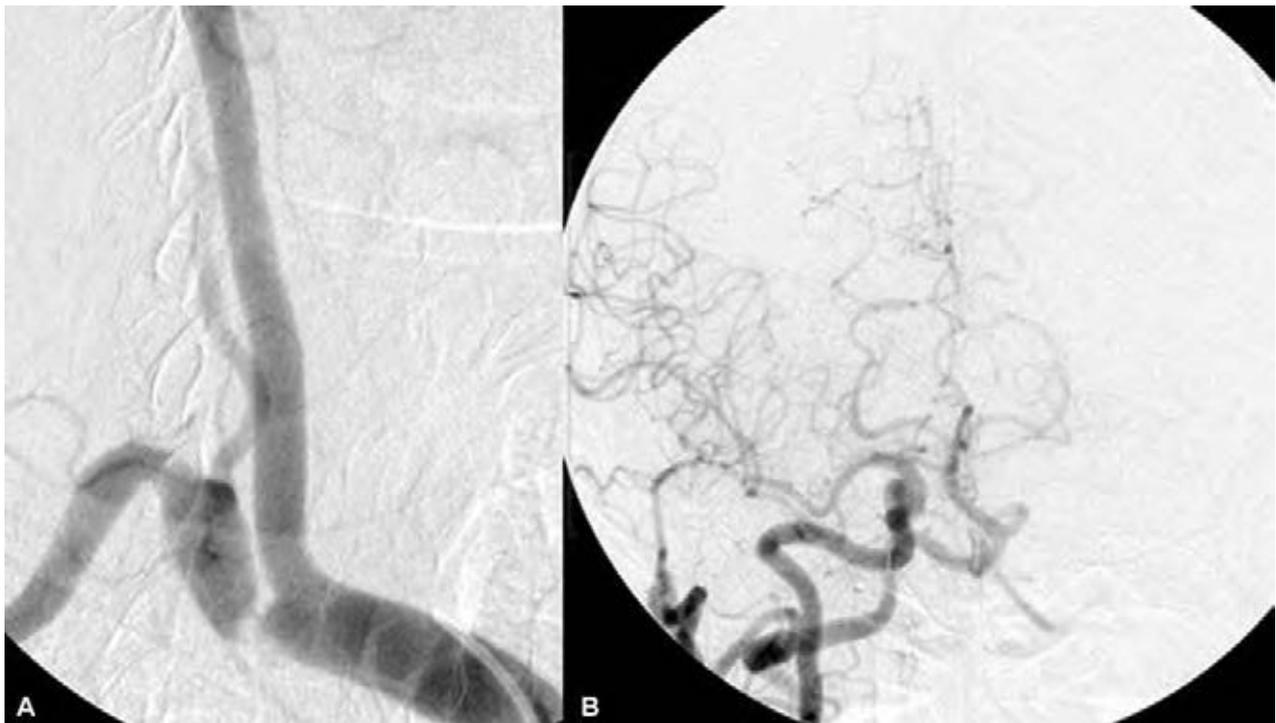
Significant subclavian or braquiocephalic artery stenosis or occlusions, however, are mostly asymptomatic and, therefore, do not require specific therapy other than those directed to the underlying etiology.<sup>2,3</sup> However, when symptoms do occur, the most common manifestations are symptoms of vertebrobasilar insufficiency, such as dizziness and diplopia, or symptoms of upper limb ischemia, such as weakness, paresthesia, numbness, and coldness. These symptoms are often associated with arm exercises.<sup>4-7</sup>

Rarely, however, some patients may provoke exercise syndrome and present with transitory ipsilateral arm claudication, ataxia and/or angina.<sup>8</sup> It is often a differential diagnosis in any patient who has a pulse deficit or a systolic blood pressure difference greater than 20 mmHg between the arms.<sup>8</sup>

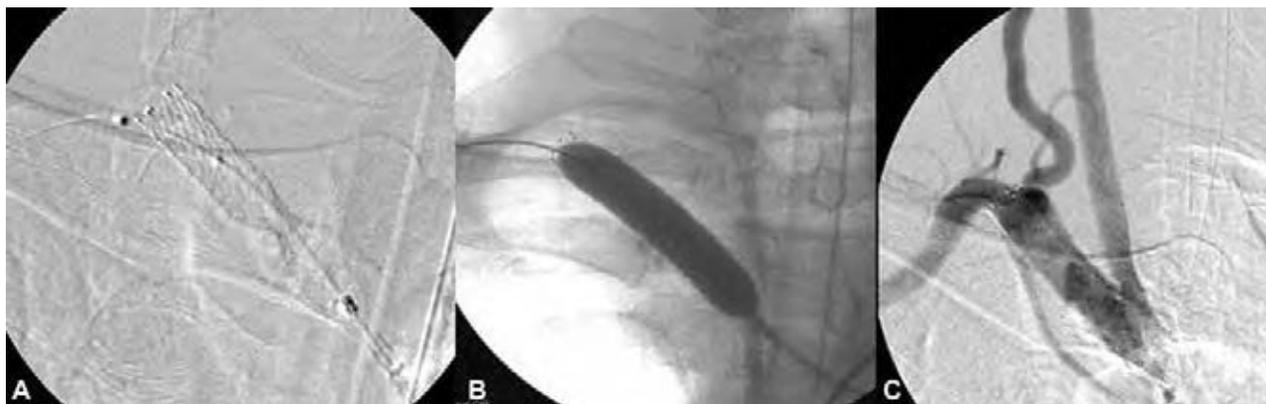
Atherosclerosis is the main cause of occlusive disease involving the subclavian artery, with concomitance of smoking in 78 to 100% of the cases, of coronary artery disease in 27 to 65% of the cases, and of carotid or vertebral arterial disease in one third of the patients.<sup>9-11</sup>

## Case Report

A 59-year-old male presented with multiple episodes of syncope and lower limb vascular claudication during his work (cabinet maker) for ~ 2 years. He had a history of hypertension and smoking and was submitted to bilateral femoral popliteal bypass surgery 1 year before, with total improvement of the claudication of the lower limbs. He had also undergone a bilateral carotid artery angioplasty 6 months earlier due to severe stenosis, but the syncopes remained. The cardiologic investigation and the neurological examination were normal, but blood pressure was 40 mmHg different between his arms. A cervical and intracranial digital subtraction angiography (DSA) showed a severe stenosis of the right subclavian artery, with reverse blood flow in the ipsilateral vertebral artery irrigating the right upper limb, characterizing the SSS (►Fig. 1). Thus, the patient underwent a right subclavian artery angioplasty with stent and balloon (►Fig. 2). The procedure was performed through a right femoral access, under general anesthesia and full intravenous heparinization. A Destination peripheral guiding sheath (Terumo, Tokyo, Japan) was positioned in the brachiocephalic trunk and a Protégé RX stent (Medtronic, Dublin, Ireland) with 10 × 7 mm of diameter and 40 mm of extension was delivered crossing the stenosis. Although the stent produced partial vessel opening, an Armada balloon catheter (Abbott, Chicago, IL, USA) of 9 × 40 mm inflated until 13 Atm achieved definitive stent impingement and normalization of the cervical and of the intracranial blood flow. In the postoperative period, he maintained the use of simvastatin 40 mg, enalapril 20 mg, and



**Fig. 1** A: Right subclavian artery with subocclusive stenosis and a retrograde flow in the vertebral artery. Right anterior oblique view. B: Right common carotid artery and filling of the posterior circulation through extracranial anastomosis. Anteroposterior view.



**Fig. 2** Angioplasty. A: Delivered and opened stent covering the entire stenosis. B: Inflating the balloon. C: Satisfactory final result.

double antiaggregation (aspirin 100 mg and clopidogrel 75 mg) daily. There were no further syncopes or claudication of the arm during 16 months of follow-up, without difference of blood pressure between the arms, despite the patient having not stopped smoking.

## Discussion

### Clinical Presentation

Subclavian steal syndrome is a rare disease, related in ~6% of asymptomatic patients with cervical murmurs.<sup>5</sup>

In the majority of the patients, a subclavian severe stenosis with or without flow reversal in the ipsilateral vertebral artery is clinically asymptomatic.<sup>5,12</sup> It occurs due to the slow progression of the disease and to the development of collateral circulation.<sup>13,14</sup>

However, patients with flow reversal can become symptomatic if the collateral blood supply from the vertebrobasilar circulation cannot accommodate the increased demand, such as during exercise or in the setting of an arteriovenous fistula.<sup>15</sup>

The symptoms of subclavian disease include ipsilateral claudication of the superior extremity (that can be correlated with Takayasu arteritis) and vertebrobasilar insufficiency, such as the syncopes of our patient, or even posterior fossa stroke and myocardial ischemia.<sup>3,5,9,15</sup> Among the clinical manifestations, vertigo or dizziness are the most prevalent.<sup>7</sup> Other possible symptoms include ataxia, tinnitus, drop attacks, visual disturbances, perioral numbness, alternating paresthesia, dysphagia, dysarthria, transient amnesia, or headache.<sup>3,5,9,15</sup>

Complete occlusion is three times more prevalent in the left subclavian artery than in the right subclavian or in the innominate artery.<sup>16</sup>

### Diagnosis

In the physical examination, a discrepancy of > 15 to 20 mmHg in the arm blood pressure (ABP) taken in both upper extremities can be found, beyond pulse impairment in the affected side.<sup>4,12</sup> In a study by Osborn et al,<sup>14</sup> a systolic ABP difference  $\geq 15$  mmHg had a positive and negative predictive value of 100% for SSS, in a total of 59 patients. On the other hand, English et al found in the difference in

systolic ABP  $\geq 20$  mmHg the positive and negative predictive values of 19 and 98%, respectively.<sup>17,18</sup>

Recent studies have shown a linear correlation between the increase of the ABP difference with the occurrence of symptoms.<sup>19,20</sup> Labropoulos et al reported that 38.5% in the group with ABP difference 50 mmHg presented symptoms.<sup>19</sup>

The arterial insufficiency syndrome can lead to skin modification, but only 5% of the patients have cyanosis and trophic skin changes due to severe brachial ischemia or embolism in the distal arterial branches of the upper limbs.<sup>12,21</sup> Also, an audible bruit over the supraclavicular fossa can be heard.<sup>4,12</sup>

The diagnosis can be done with the use of a Doppler ultrasound guided by an experienced professional, with findings such as waveform dampening or monophasic changes.<sup>22</sup> When the Doppler is inconclusive, it is possible to use a more invasive diagnosis test, such as angioresonance or angiotomography, both showing the grade of the stenosis.<sup>12</sup> However, DSA still is the gold standard test due to the dynamic visualization of the retrograde vertebral flow.<sup>22</sup>

### Treatment

A small percentage of patients with SSS really need any therapeutic intervention.<sup>4,19,22</sup> The surgical options, such as bypass grafting, or endovascular treatments involving angioplasty and stenting, are often reserved for serious clinical symptoms.<sup>4</sup>

Asymptomatic patients should not be submitted to interventions,<sup>4,7</sup> since they could prevent future disease with medical therapy, which includes antiplatelet drugs, such as aspirin, and a lipid-lowering statin therapy with a goal of low-density protein (LDL) cholesterol < 100 mg/dL.<sup>4,7,22</sup> Stopping smoking, controlling the blood pressure, and glycated hemoglobin A1C rate < 7% are advised<sup>7</sup> to reduce the morbidity and mortality risk related to atherosclerotic disease, which can lead to ischemic events in the heart and in the brain.<sup>4</sup> Subclavian stenosis is an atherosclerotic disease marker and, as a consequence, cardiological (heart attack) and brain (stroke) events should be warned of the possibility of occurrence.<sup>18,22</sup>

The surgical vascular intervention options include: axillary-axillary bypass, carotid-subclavian bypass, and transposition of the subclavian artery.<sup>22</sup> The carotid-subclavian bypass

showed low risk of mortality and small recurrence rates.<sup>4</sup> Law et al<sup>21</sup> found in their study that transpositions have the highest long-term 5-year patency rate (100%), followed by PTFE (95%) and Dacron grafts (84%), and saphenous vein grafts have the lowest patency rate (65%).<sup>23</sup>

Percutaneous transluminal angioplasty (PTA) has a high rate of patency for subclavian stenosis or occlusion, the latter with a lower rate. Percutaneous transluminal angioplasty has a high rate of good outcome and a low incidence of recurrence, especially in proximal stenosis.<sup>16-18</sup> It is done with local puncture and, therefore, has lower rates of complications; stenting has been considered safe when you compare it with the surgical vascular intervention.<sup>18</sup> In a series of 110 patients treated by PTA with a follow-up of almost 3 years, patients with subclavian stenosis and occlusion had patency rates of 93 and 65%, respectively.<sup>16</sup>

Risks related to PTA are stroke, arterial rupture, stent migration, and reocclusion.<sup>18</sup> Patients who already had an occlusion presented with a high rate of reocclusion (35-50%).<sup>17,18</sup> A regular follow-up for at least 2 years postPTA is recommended.<sup>18</sup>

None of the studies reviewed in the present article clarifies how long a patient with total occlusion is still submitted to an intervention with angioplasty.

Prospective randomized studies are needed to elucidate the fact that several papers analyze subclavian PTA in stenosis and occlusion, but they do not mention whether or not they have clinical and/or angiographic criteria for SSS.

## Conclusions

Subclavian steal syndrome is a rare vascular condition related to cerebrovascular risk factors, with a wide clinical presentation, but normally asymptomatic. Endovascular treatment with stent and balloon, as performed in the related patient, is safe and has good long-term results. Intervention is only indicated in symptomatic patients.

### Conflicts of Interests

The authors have no conflicts of interests to declare.

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# Abdominal Cutaneous Nerve Entrapment Syndrome, Case Report and Diagnostic Evaluation

## *Síndrome de compressão do nervo cutâneo abdominal, relato de caso e avaliação diagnóstica*

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### Abstract

Abdominal cutaneous nerve entrapment is a rarely diagnosed condition that leads to intense neuropathic pain in the anterolateral wall of the abdomen. Generally, it is triggered by some factor implied in the increase of the pressure on the nerve in its passage by the abdominal wall. Its most important differential diagnosis is pain of visceral origin.

### Keywords

- ▶ nerve compression syndrome
- ▶ ultrasonography
- ▶ diagnostic imaging

We present a case in which the clinical findings confirmed on ultrasound and other imaging tests established the diagnosis and in which the noninvasive treatment was effective.

### Resumo

A compressão do nervo cutâneo abdominal é uma condição raramente diagnosticada que causa dor neuropática intensa na parede anterolateral do abdome. Geralmente, ela é iniciada por algum fator implicado no aumento da pressão sobre o nervo na passagem deste pela parede abdominal. Seu diagnóstico diferencial mais importante é a dor de origem visceral.

### Palavras-chave

- ▶ síndromes de compressão nervosa
- ▶ ultrassonografia
- ▶ diagnóstico por imagem

Apresentamos um caso no qual os achados clínicos confirmados por ultrassonografia e por outros exames de imagem firmaram o diagnóstico e no qual o tratamento não invasivo foi efetivo.

## Introduction

Abdominal cutaneous nerve entrapment syndrome (ACNES) is a commonly misdiagnosed condition, its prevalence among patients referred to general practitioners, abdominal surgeons, or pain clinics for chronic abdominal pain ranges from 10 to 30%.<sup>1,2</sup> In an emergency department setting studied by van Assen et al,<sup>3</sup> it was found that ~ 2% of the patients with acute abdominal pain had ACNES. The most

common symptom is a dull or burning pain with a sharp, well-localized component in a restricted area of the abdominal wall. The pain can be chronic or acute. It generally arises from lower intercostal nerves (T8–T11) probably caused by entrapment of the cutaneous abdominal nerve at the lateral border of the rectus abdominis muscle (RAM), specifically where this nerve enters the RAM channel through a fibrous ring. As the nerve passes through the RAM, each nerve and its vessels are surrounded by fat, binding the nerve, arteries and veins into a bundle. Separating the bundle from the muscle, there is the fibrous ring situated about midway between the anterior surface of the

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RAM and its posterior side, providing a smooth environment through which the bundle can slide. This area is believed to be more susceptible to ischemia or to physical compression of the nerve due to the juxtaposition of the soft neurovascular bundle to the firm fibrous ring.<sup>2-10</sup>

We present a case of ACNES seen at the Division of Neurosurgery of the Hospital Universitário Gaffrée e Guinle of the Universidade Federal do Estado do Rio de Janeiro (HUGG – UNIRIO, in the Portuguese acronym), calling attention to the clinical diagnosis, pointing out its imaging characteristics, highlighting the value of high definition linear ultrasound (HDLUS) as a valuable diagnostic method and showing that a noninvasive treatment can be effective.

## Case Report

A Caucasian male, 69 years old, presented with burning pain on the right anterolateral abdominal wall in the region of the T9–T10 dermatomes. The pain had begun 4 months earlier and started after lifting a heavy weight. The patient had diabetes mellitus (DM), systemic arterial hypertension, and was overweight (29.1 kg/m<sup>2</sup>).

A general practitioner, a cardiologist, and a general surgeon successively evaluated the patient. The cardiological tests, the thoracic abdominal and pelvic computed tomography (CT), and the abdominal-pelvic magnetic resonance imaging (MRI) that were requested for investigation were all considered inconclusive.

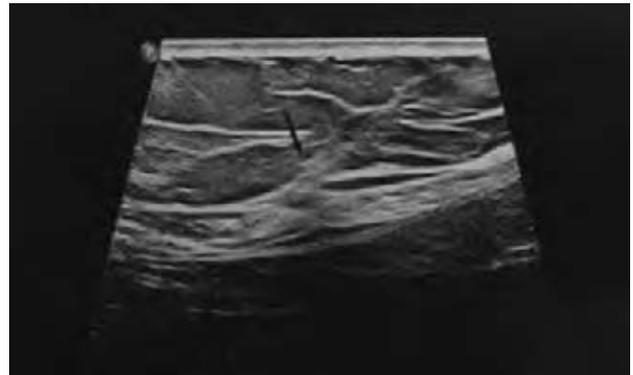
The patient was seen at the HUGG neurosurgery division due to the persistence of pain with neuropathic characteristics. At the neurological evaluation, he reported intense burning pain at the left T9–T10 dermatome area (visual analogue scale [VAS] 8), with no changes at ectoscopy apart from the overweight. The pain was continuous without remission periods and extremely worse during the night. The patient presented with hypoesthesia and discriminative sensibility was reduced compared with the contralateral abdominal wall in the T9–T10 area. The Carnett test was positive. The Valsalva maneuver also enhanced the pain.

As the previous imaging exams of the patient were inconclusive and intercostal nerve compromise was sus-

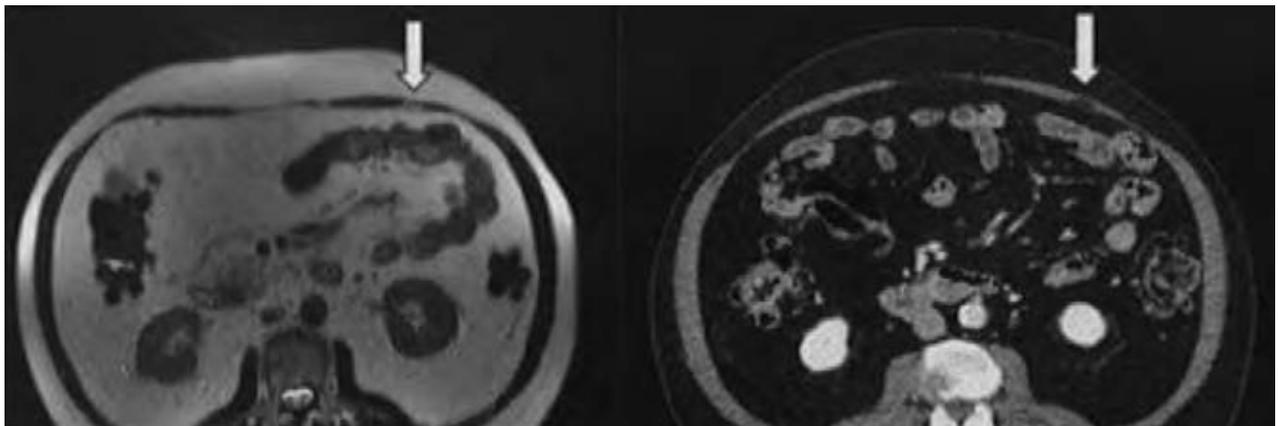
pected, it was performed a HDLUS of the abdominal wall that demonstrated the left RAM and its sheath lesion in a paramedian and supraumbilical topography with cicatricial fibroadipose tissue. The muscle injury was associated with fibrocicatricial thickening of the abdominal cutaneous nerve. Supraumbilical diastasis of the RAM could also be noticed. (►Fig. 1)

Other imaging studies (MRI and CT) were reanalyzed by the radiologist of our institution, after the diagnosis with HDLUS. Some abnormalities were noticed with the reevaluation of the images, such as high signal intensity on the MRI T2 image and decreased density on the CT image around the RAM strain area. (►Fig. 2)

With a clinical and imaging diagnosis of ACNES, the patient was oriented to nutritional evaluation for weight control and pregabalin was administered at a dose of 75 mg 3 times a day in the 1<sup>st</sup> month. The patient was accompanied at the outpatient clinic and, after 2 months, reported pain relief of 80% (VAS dropped from 8 to 3). Three months after the beginning of the treatment, the patient reported a total relief of pain and, after 10 months, he had lost 15kg and the pregabalin administration could be reduced to 75mg/day. The last evaluation was 1 year after the diagnosis, the patient was on physiotherapy, had a normal control of his DM and continued nutrition support. The pain did not return.



**Fig. 1** Ultrasound of abdomen; arrow shows fibrocicatricial thickening of the abdominal cutaneous nerve (black arrow).



**Fig. 2** On the right axial abdominal MRI (T2) and on the left axial abdominal CT; both demonstrate the rectus abdominis strain area (white arrow).

## Discussion

Abdominal cutaneous nerve entrapment syndrome is a commonly misdiagnosed condition that can have visceral pain as differential diagnosis, or the patient might even present with a coexisting visceral condition that does not explain the pain by itself. Therefore, it is important to recognize the clinical signs of ACNES and to look for the imaging characteristics of the lesion.<sup>3-5,9-15</sup>

Typically a unilateral neuropathic pain in the area of the T8-T11 dermatomes with hypoesthesia in this same region, ACNES has an important clinical test, formulated by Carnett,<sup>16</sup> to aid in the diagnosis. This test is based on the presumption that if the pain has a visceral origin, the tensed abdominal muscles guard the underlying structures, reducing tenderness, whereas continued tenderness during muscle contraction implicates the abdominal wall as the source of the pain. In the presented case, the Carnett test was positive and the Valsalva maneuver enhanced the pain, as it can increase the abdominal pressure to the entrapped nerve.<sup>2-6,11,16,17</sup>

The next diagnostic step would be the imaging evaluation. Conventionally, to study the abdomen, physicians can use CT, MRI, and ultrasound (US). Computed tomography is the most commonly used in the emergency room in patients whose first clinical hypothesis is a visceral pain, and that was the case of our patient, who also underwent an MRI because of this suspicion. However, although the images showed indeed high signal intensity on MRI T2 and a decreased density on CT at the RAM strain area (→**Fig. 2**), probably indicating fibrocitrerial tissue, as ACNES was not a clinical suspicion at the moment, these abnormalities were only seen after the reevaluation of the imaging exams in our institution.<sup>12-14,18</sup>

Nonetheless, although we could presume that the best imaging exam for these cases would be MRI, given that the abnormalities can be seen if followed by the clinical suspicion, and that this exam is known to be one of the best to study muscles, we should not forget that it is also one of the most expensive.<sup>18</sup> Therefore, neuromuscular HDLUS (18-5 MHz), used in the present case, is a great contribution in the hands of an experienced examiner, revealing itself even more sensitive to diagnose some neuromuscular lesions than MRI, despite being an examiner-dependent technique. In addition, it is a relatively cheap and fast exam.<sup>19</sup>

Although Kanakarajan et al have already discussed the importance of US on ACNES as a guidance to therapeutic injections, we suggest its use for diagnostic purposes.<sup>8</sup>

With the use of HDLUS on the abdominal wall, it is possible to differentiate fiber discontinuity due to muscle strain of the RAM as decreased echogenicity areas and the fibrocitrerial tissue can be localized as increased echogenicity areas, corroborating the abnormalities found on the MRI and CT exams, imaging methods that demand more time and are more expensive.<sup>18</sup> So, if the clinical evaluation indicates that that problem is on the abdominal wall and not from a visceral origin, we suggest the use of HDLUS as the first imaging modality exam.

Nowadays, it is recognized that a multimodal approach for such cases is paramount, especially in chronic refractory cases. However, some authorities<sup>14,15,20</sup> try to confirm the

diagnosis and use trigger point injection (lidocaine) without mentioning any systemic drug (nonsteroidal anti-inflammatory drugs, weak opioids, antiepileptics, and antidepressants) or other multimodal treatment.<sup>4</sup>

Weight control, physiotherapy, and systemic drug administration following the guidelines for neuropathic pain therapy was the protocol established for the presented case. The result was positive with this noninvasive management. It is important to point out, as the current literature reports, that systemic drugs may be used as a first step or as a complementary treatment for ACNES, considering the result with chronic pain relief. However, these studies do not confirm if the administration of systemic drugs alone can help patients with ACNES.<sup>4</sup>

If the use of systemic drugs fails to decrease the pain, the best option would be lidocaine trigger point injection, preferably ultrasound-guided, not associated with corticosteroids, as this combination is not certain to increase the success of this therapy. As for refractory cases in which multiple trigger point injections associated with a multimodal approach does not have success, the patient might need a more invasive strategy, such as a neurectomy of the anterior cutaneous nerve endings. These two types of management can lead to long-term pain relief in ~ 75% of the patients with ACNES.<sup>4,14,15,20</sup>

Other options described in the literature would be pulsed radio frequency treatment, spinal cord stimulation of the dorsal root ganglion, and intraperitoneal onlay mesh reinforcement. However, these methods still need further studies to confirm their efficacy.<sup>4,21,22</sup>

An important bias of our study is the fact that HDLUS, used in the presented case, is an examiner-dependent method, and that we compare only one case to the current English published literature, which may lead to some disagreements that could be overcome on a larger sample.

## Conclusion

Patients who present on clinical evaluation with unilateral neuropathic pain in the area of the T8-T11 dermatomes, hypoesthesia in this same region should be investigated for ACNES with HDLUS performed by an experienced professional reducing the costs to conclude a correct diagnose, and the noninvasive management might be helpful and effective.

### Note

We declare that the present manuscript has not been previously published in whole or in part or submitted elsewhere for review and we report no conflicts of interest concerning the materials or methods used in the present study or the findings specified in the present paper.

### Conflicts of Interests

The authors have no conflicts of interests to declare.

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# The Unknown Cause of Pott's Puffy Tumor – Importance of Early Diagnose

## *A causa desconhecida de Pott puffy tumour – importância do diagnóstico precoce*

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### Abstract

Pott's puffy tumor (PPT) is characterized by swelling of the glabellar region and osteomyelitis of the frontal bone, owing to a subperiosteal pseudoinflammatory tumor responsible for the detachment of the pericranium from the outer table of the skull. Nowadays, the incidence of PPT is very low, so this entity is frequently underdiagnosed. The late treatment and identification of PPT are strongly associated with intracranial complications, which could jeopardize the life of the patient.

In the literature, PPT is described as a complication of frontal head trauma or of chronic sinusitis. There are a few cases reported in patients with frontal insect bites or in recreational nasal drug users, such as cocaine or methamphetamines.

In the present case report, the authors describe the case of a 40-year-old male who was submitted to a frontal sebaceous cyst surgery. In the postoperative period, he developed an infectious process compatible with PPT. After an extensive review of the literature, no similar cases were identified. Therefore, in the opinion of the authors, sebaceous cyst surgery should be included in the short list of risk factors for the development of PPT.

### Keywords

- ▶ epidural abscess
- ▶ frontal osteomyelitis
- ▶ Pott puffy tumor
- ▶ sebaceous cyst

### Resumo

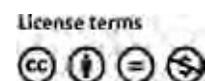
O Pott puffy tumor (PPT, na sigla em inglês) é caracterizado por edema da região glabellar e osteomielite do osso frontal, em consequência de uma lesão tumoral subperiosteal, pseudoinflamatória, que promove a separação espontânea do pericrânio da camada cortical óssea externa do crânio. Atualmente, a incidência de PPT é muito baixa, causa pela qual o subdiagnóstico desta entidade é frequente. O atraso no tratamento desta entidade está fortemente associado a complicações infecciosas intracranianas que colocam em risco a vida do paciente.

Na literatura, o PPT está descrito como complicação de traumatismo craniano frontal ou de sinusite crônica. Existem ainda alguns relatos em pacientes com picadas de inseto na região frontal ou em consumidores de drogas recreativas por via nasal, como cocaína ou metanfetaminas.

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**Palavras-chave**

- ▶ abscesso epidural
- ▶ osteomielite frontal
- ▶ Pott puffy tumor
- ▶ quisto sebáceo

No presente caso clínico, os autores relatam o caso de um homem de 40 anos, submetido a excisão de quisto sebáceo na região frontal que, no pós-operatório, desenvolveu um processo infeccioso compatível com PPT. Após uma extensa revisão de literatura, não foram identificados relatos semelhantes, pelo que se considera que a cirurgia de quisto sebáceo da região glabellar deve ser englobada na pequena lista de fatores de risco para o desenvolvimento de PPT.

**Introduction**

Pott puffy tumor (PPT) is an eponymous for a subperiosteal abscess of the frontal region, generally associated with frontal osteomyelitis. The arising of broad-spectrum antibiotics lowered the incidence of PPT. Although it is a rare entity in the postantibiotic era, clinicians should be alert to this life-threatening entity. Intracranial complications, such as dural sinus thrombosis or subdural abscess, are usually present at the time of the diagnosis, so neurosurgical intervention is often needed.<sup>1</sup> Nowadays, PPT is generally a complication of chronic sinusitis. The most common infectious agents belong to the Streptococci family, being *Staphylococcus aureus* a less frequent causative agent.<sup>2</sup> To the best of our knowledge, there are no cases of PPT as a complication of sebaceous cyst surgery reported in the literature.

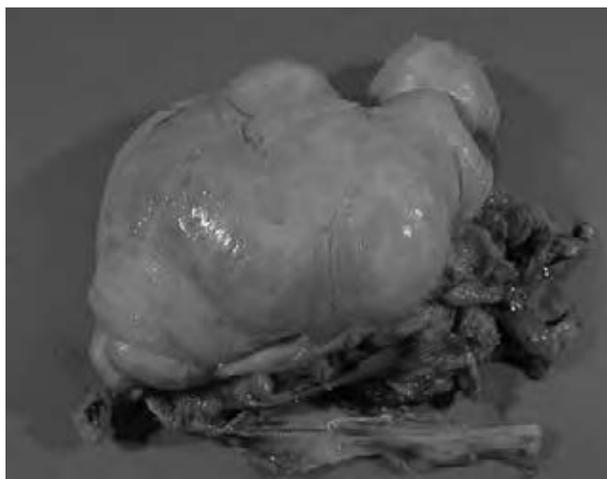
**Case Report**

A 40-year-old male underwent surgery to remove a noninfected and nonpainful lump in the glabellar region with a diameter of 2 cm (►Fig. 1). Prior to the surgery, a soft tissue ultrasound showed a closed sac under the skin filled with serous material. The histological analysis showed a unilocular cyst in the upper dermis composed of a flattened and granular layer of kerathohyalin granules, compatible with sebaceous cyst (►Fig. 2). Two weeks after the procedure, the patient developed a painful swelling in the frontal region with a serous drainage of the surgical wound. Given the

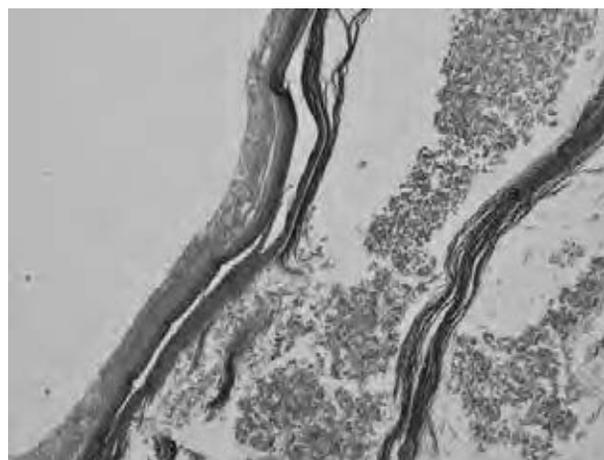
suspicion of postoperative soft tissue infection, the patient was medicated with oral flucloxacillin. Two weeks later, he presented to the emergency department complaining of bilateral frontal headache and maintenance of the frontal swelling, now with fluctuation and inflammatory signs. Tympanic temperature, heart rate, respiratory rate and blood pressure were normal. No signs of meningeal irritation or of neurological deficit were observed in the neurological exam. Blood cell count and biochemical parameters were normal. The patient denied any past history of sinusitis, head trauma, immunosuppression, or of consumption of inhaled drugs.

A contrast-enhanced computed tomography (CT) scan of the head was performed. A frontal subperiosteal abscess with extension to the epidural space (►Figs. 3 and 4) due to frontal bone osteomyelitis (►Figs. 5 and 6) was identified. No signs of frontal sinusitis were observed. The patient underwent emergency surgery. A frontal craniectomy was performed to drain the epidural abscess. No signs of dural infection were observed. The infected bone was replaced by a titanium mesh (►Figs. 7 and 8). On the bacterial cultures, a methicillin-resistant *S. aureus* (MRSA) was identified. After the surgery, the patient underwent intravenous antibiotic therapy with vancomycin and ceftriaxone for 6 weeks, plus metronidazole for 7 days.

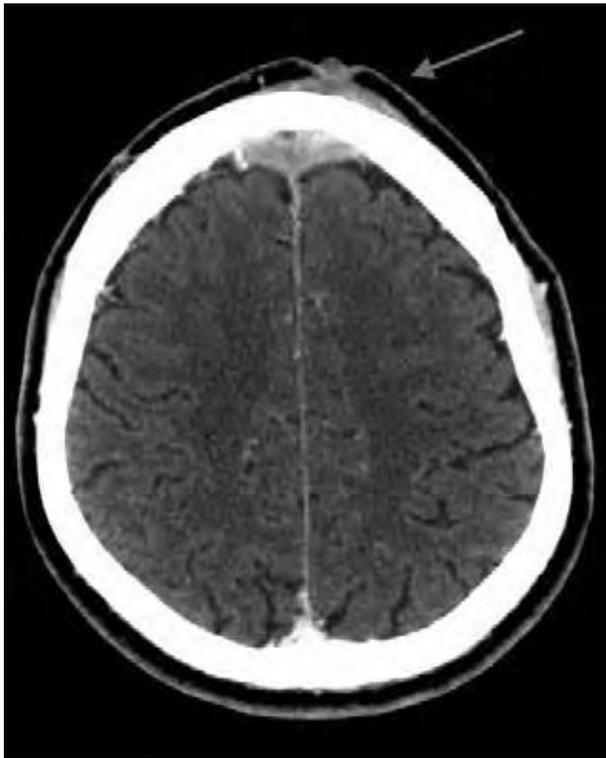
At 1 year postoperatively, the patient continues asymptomatic, with no signs of skin or intracranial infection. The primary cortical areas remain intact, and no focal neurological deficits are observed. The cranioplasty performed presents an excellent aesthetic result. He returned to his daily routine with no restrictions.



**Fig. 1** Macroscopic image of the removed lesion.



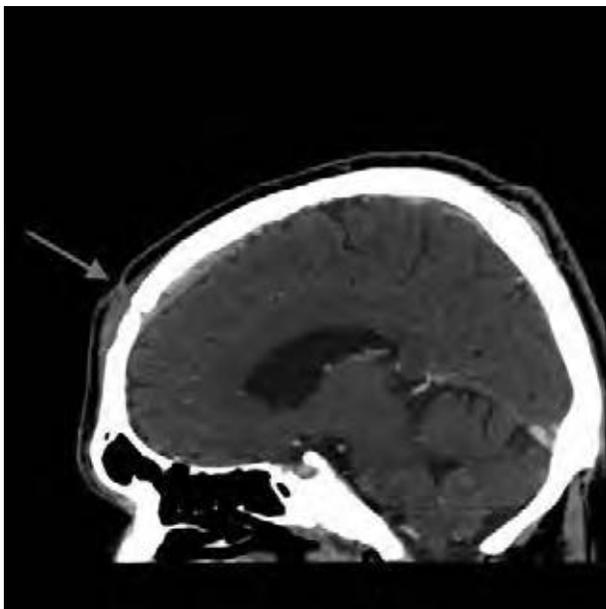
**Fig. 2** Histological features of the removed sebaceous cyst.



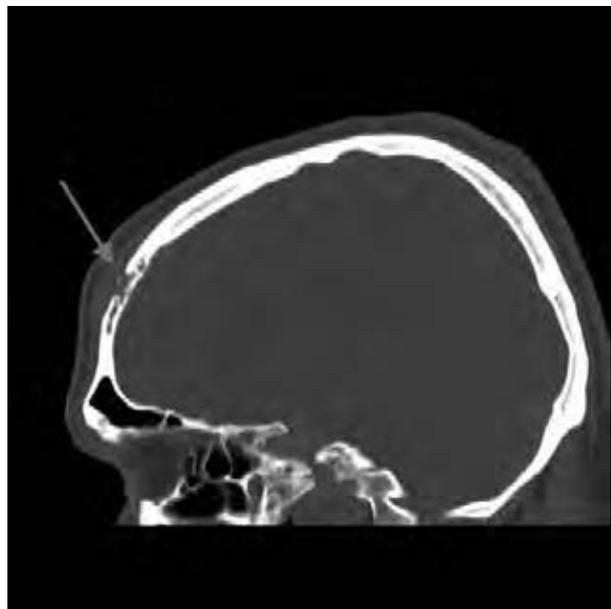
**Fig. 3** Contrast-enhanced computed tomography scan with subperiosteal abscess and epidural empyema – axial view.



**Fig. 5** Computed tomography scan, bone window, showing a frontal osteomyelitis – axial view.



**Fig. 4** Contrast-enhanced computed tomography scan with subperiosteal abscess and epidural empyema – sagittal view.



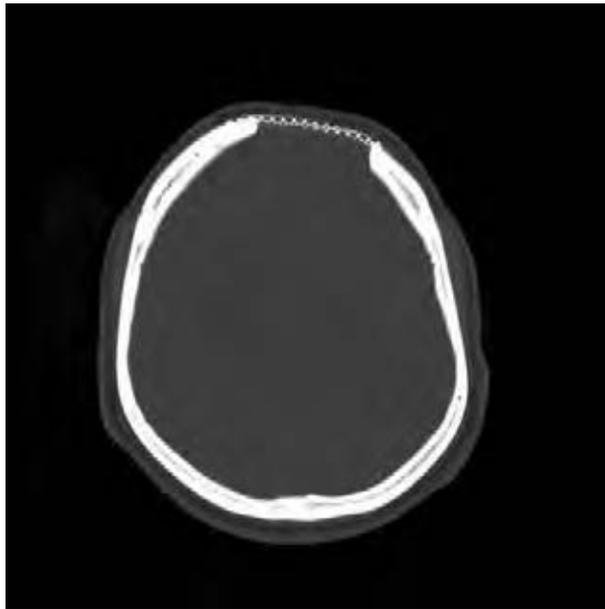
**Fig. 6** Computed tomography scan, bone window, showing a frontal osteomyelitis -- sagittal view.

**Discussion**

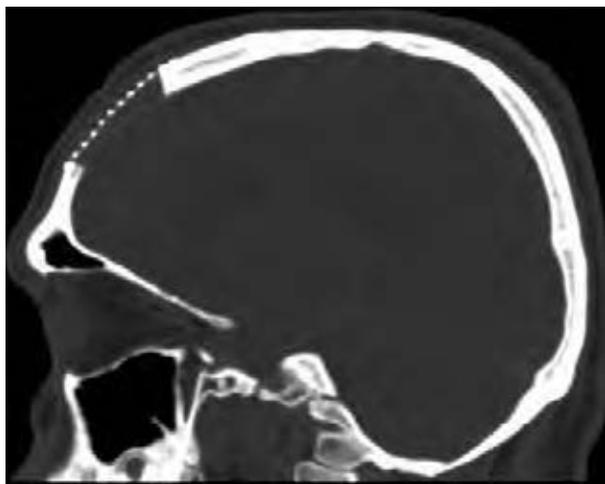
Pott puffy tumor was first described by Sir Percival Pott in 1760 as a complication of frontal head trauma. In 1879, Lannelongue described the first case of PPT related to frontal sinusitis.<sup>3</sup> Although it is rare in the postantibiotic era, PPT continues to emerge. The main etiologies are chronic frontal sinusitis and frontal traumatic brain injury. Few cases of

cocaine and amphetamine consumption and insect bite are also described. Subjects with diabetes mellitus, chronic renal disease, and aplastic anemia have a predisposition for this infection.<sup>4</sup>

To understand the pathophysiology of PPT, it is important to know the anatomy of diploic veins. Diploic veins are large, thin-walled valveless veins present between the inner and



**Fig. 7** Postoperative computed tomography scan showing frontal cranioplasty with a titanium mesh – axial view.



**Fig. 8** Postoperative computed tomography scan showing frontal cranioplasty with a titanium mesh – saggital view.

outer layers of the cortical bone in the skull. They communicate with the meningeal veins and the dural sinus, and with the veins of the pericranium. The four main diploic veins are the frontal, anterior temporal, posterior temporal, and occipital.<sup>5</sup>

Pott puffy tumors emerge more frequently in male adolescents with chronic sinusitis. In these cases, the inflammatory content inside the frontal sinus can destroy its anterior wall promoting the formation of the subperiosteal abscess. A hematogenous spread of the inflammatory content in the frontal sinus can occur through the diploic veins of the bone directly to the dural sinus.<sup>6</sup> Therefore, sinus thrombosis, subdural empyema, and intracerebral abscess are the most common complications. The destruction of the posterior wall of frontal sinus is much slower, so epidural abscess is a less frequent complication.<sup>7</sup>

In the literature, we did not find any case of PPT as a complication of sebaceous cyst removal. Our case portrays a healthy patient who underwent a routine procedure, with no surgical complication reported and with complete cyst excision. In our opinion, cyst fragmentation, partial removal, or wound contamination due to poor aseptic techniques could be the factors responsible for this MRSA infection. The inflammatory content of the cyst passed from the subcutaneous layer to the pericranium, where a subperiosteal abscess was formed.

The spread of the infection to the bone may have occurred through the supraorbital vein. This vein originates in the forehead, runs downwards superficial to the frontalis muscle, and passes through the supraorbital notch, where it receives the frontal diploic vein from a micro foramen in the notch.<sup>5</sup> Therefore, this communication could justify the occurrence of the osteomyelitis in the reported case. Finally, the germ identified in the epidural abscess was a skin commensal, *S. aureus*, indicating that, probably, the infection started in the skin.

## Conclusion

Pott puffy tumor is a life-threatening entity that should be promptly diagnosed. This infection is generally unfamiliar to the majority of the emergency physicians. Pott puffy tumor should not be forgotten, since an early diagnosis is associated with a good outcome and with the complete resolution of the infection. The risk of intracranial infection after sebaceous cyst removal is extremely low and we did not find any case reported in the literature. However, it can occur and could be catastrophic. The existing venous anastomosis between the intracranial and extracranial venous systems promotes an easy propagation of infection from the skin to the meningeal layers and the cerebral parenchyma. Therefore, after cyst removal in the cranial region, the surgical wound must be carefully watched, preventing the onset of soft tissue infections.

## Conflicts of Interests

The authors have no conflicts of interests to declare.

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# Endovascular Treatment of Ruptured Vertebral Artery Dissecting Aneurysm in Fibromuscular Dysplasia

## *Tratamento endovascular de aneurisma dissecante roto de artéria vertebral na displasia fibromuscular*

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### Abstract

#### Keywords

- ▶ fibromuscular dysplasia
- ▶ dissecting aneurysm
- ▶ endovascular procedure
- ▶ carotid stenosis
- ▶ angioplasty
- ▶ postoperative complications

### Resumo

#### Palavras-chave

- ▶ displasia fibromuscular
- ▶ aneurisma dissecante
- ▶ procedimento endovascular
- ▶ estenose carotídea
- ▶ angioplastia
- ▶ complicações pós operatórias

**Background** Fibromuscular dysplasia (FMD) affects predominantly the cervical and renal arteries and may cause the classical angiographic pattern of string-of-beads. The diagnosis is increasing with the advances of imaging techniques.

**Case Report** A 37-year-old man presenting with subarachnoid hemorrhage due to a dissecting aneurysm of the vertebral artery was treated by angioplasty with stent, with good outcome. All of the cervical and renal arteries were diseased and showed dysplasia and/or ectasias.

**Conclusions** There are no guidelines or protocols to treat patients with FMD.

**Introdução** A displasia fibromuscular (DFM) afeta predominantemente as artérias cervicais e renais e pode causar o padrão angiográfico clássico de cordão de contas. O diagnóstico tem aumentado com os avanços das técnicas de imagem.

**Relato de Caso** Homem de 37 anos, apresentando hemorragia subaracnoidea por aneurisma dissecante da artéria vertebral, foi tratado por angioplastia com stent, com bom resultado. Todas as artérias cervicais e renais estavam doentes e apresentavam displasia e / ou ectasias.

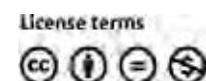
**Conclusões** Não existem diretrizes ou protocolos para o tratamento de pacientes com DFM.

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## Introduction

Fibromuscular dysplasia (FMD) is a group of idiopathic, nonatherosclerotic and noninflammatory diseases of the arterial walls leading to small- and medium-sized artery stenosis.<sup>1,2</sup> Fibromuscular dysplasia can occur in any artery, although it is more prevalent in the renal and cervical arteries.<sup>3</sup> We report a case of FMD presenting with subarachnoid hemorrhage (SAH) due to left vertebral artery (lfVA) dissection, treated successfully by angioplasty.

## Case Report

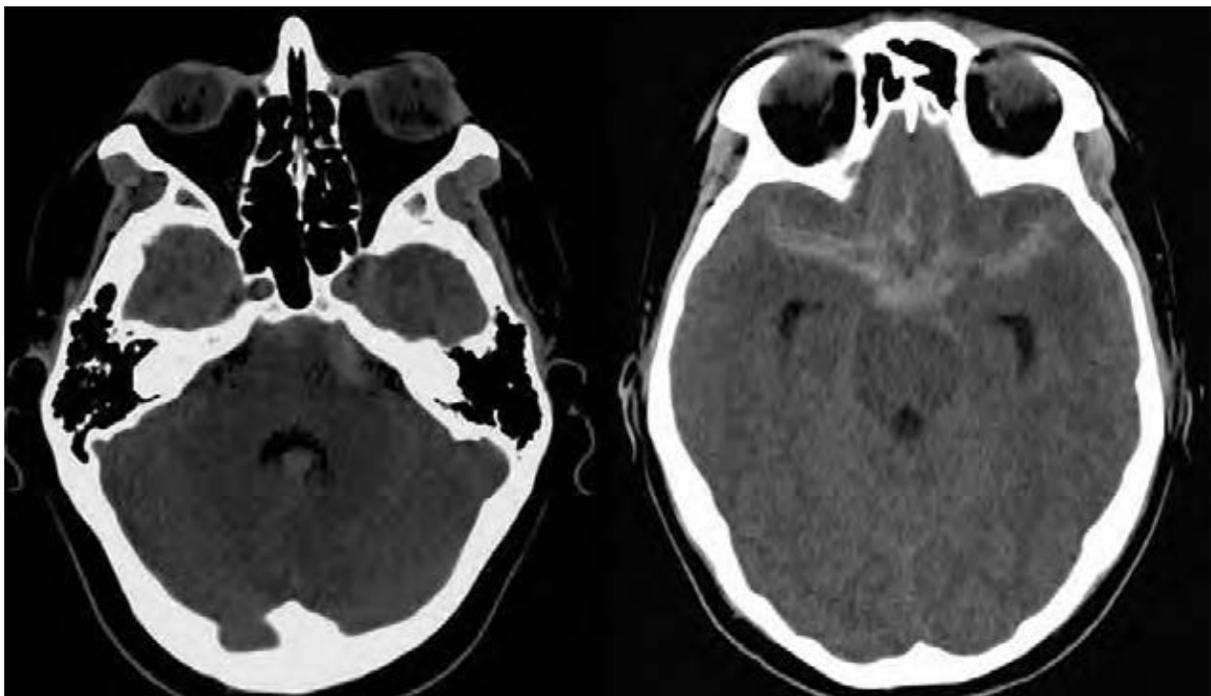
A 37-year-old man presented with sudden headache and vomiting and was admitted 7 days after in the emergency room. He was admitted with mild stiff neck, Glasgow Coma Scale 15, Hunt-Hess I, World Federation of Neurological Societies (WFNS) I. There was a medical history of high blood pressure (HBP) for 12 years controlled with amlodipine, and he denied head trauma, infection of the central nervous system (CNS), addictions, or a family history of cerebrovascular event. A computed tomography (CT) scan and a magnetic resonance imaging (MRI) exam of the head showed a SAH in the skull base cisterns and in the 4<sup>th</sup> ventricle, Fisher IV (►Fig. 1). A digital subtraction angiography (DSA) evidenced two dissecting aneurysms in the V3 segment of the lfVA, one in the right petrous carotid portion, and a “string-of-beads” pattern with stenosis followed by dilatations in both internal carotid arteries (ICAs) (multifocal type), and in both V2 segments, as well as right renal artery ectasia (►Fig. 2), diagnosing FMD. There was clinical worsening requiring mechanical ventilation due to

urinary focus sepsis, non-biliary pancreatitis, and acute renal failure (both contrast-related), bilateral hydronephrosis grade II, and transient thrombocytopenia. After clinical improvement without the need for dialysis, on the 37th day postictus, a transluminal percutaneous angioplasty (TPA) of the dissecting aneurysm was performed under general anesthesia and full intravenous heparinization. The uneventful procedure was performed through the right femoral artery (rtFA), and two telescoped coronary stents covered the dissection (►Fig. 3). The patient was discharged with no neurological deficit, using acetylsalicylic acid (ASA) and clopidogrel. A control DSA 8 months postsurgery showed exclusion of one lfVA aneurysm and decrease of the other one. The patient remained asymptomatic in a 1-year follow-up.

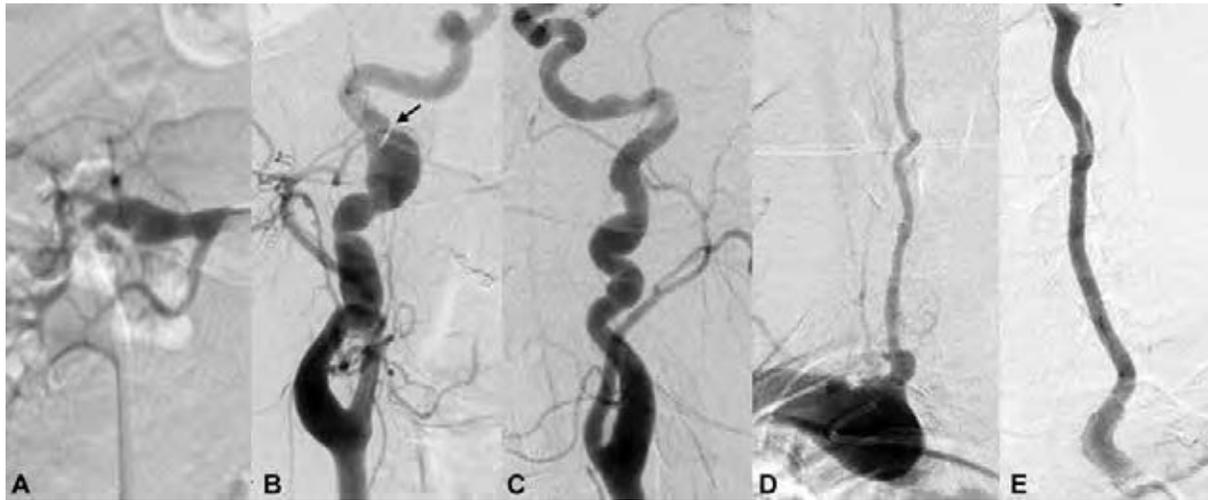
## Discussion

Three main types of renal FMDs were identified according to the arterial wall layer that is mainly affected: intimal (~ 10% of the cases of renal artery FMD), medial (80–90%) and adventitia (< 5%). However, these categories are not mutually exclusive, since the involvement of > 1 layer in the same diseased artery is not uncommon.<sup>4</sup>

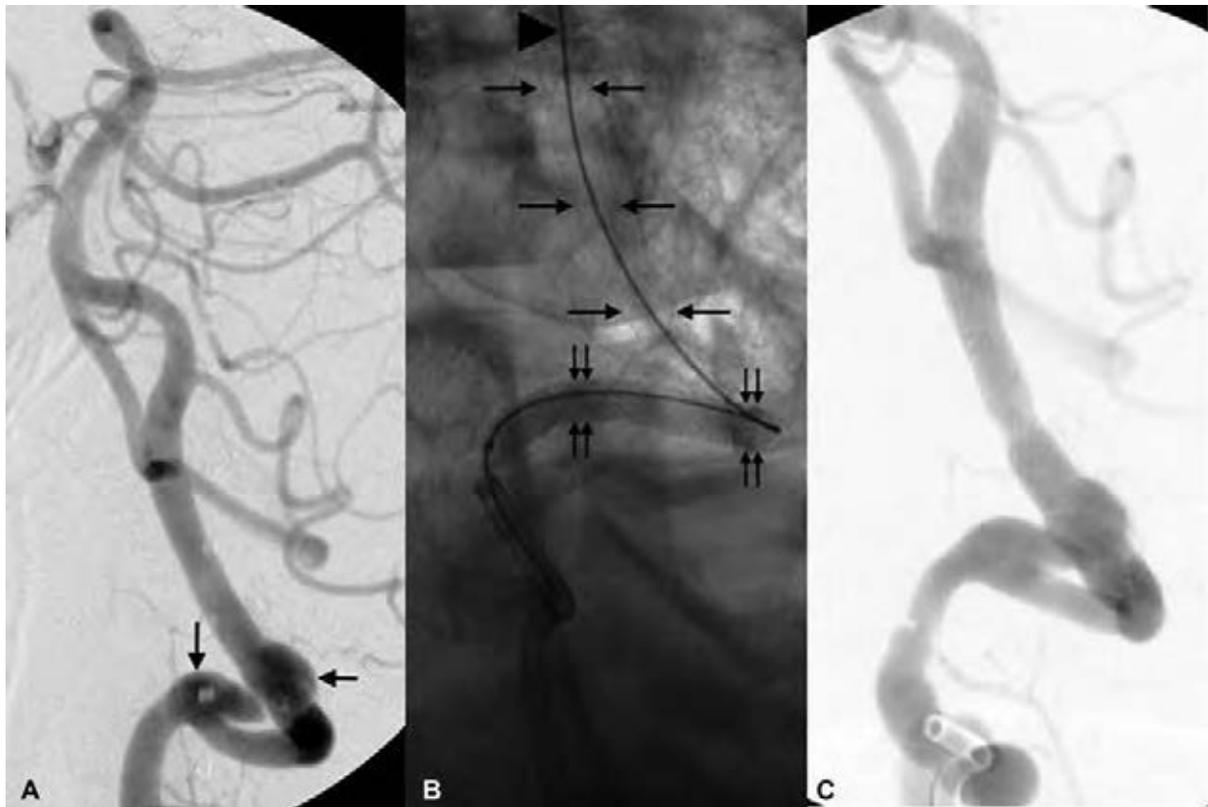
The gold standard for diagnosis is DSA, but this invasive procedure is recommended only for patients who require revascularization during the same procedure.<sup>5</sup> Currently, cervicocephalic FMD is classified angiographically into four subtypes: multifocal, unifocal, tubular, and atypical.<sup>4</sup> Multifocal, with a typical string-of-beads pattern, is the most common angiographic presentation and, histologically, corresponds to medial FMD.<sup>5</sup>



**Fig. 1** Computed tomography scan shows blood in the 4<sup>th</sup> ventricle and subarachnoid hemorrhage in the posterior fossa (A) and in the base cisterns (B).



**Fig. 2** Right renal artery angiogram is with ectasia (A). Cervical angiogram shows dysplastic alterations in the internal carotid artery on the right (B) and on the left (C) as well as in the right (D) and in the left (E) vertebral artery.



**Fig. 3** Left vertebral artery angiogram shows V3–4 segments dissecting aneurysm (arrows) (A); endovascular treatment performed over a microguidewire crossing the dissection (arrowhead); with one angioplasty and stent in V4 (arrows) and another stent in V3 (double arrows) (B), covering all lesions (C).

The clinical presentation varies, depending on the location of the arterial lesions.<sup>3</sup> The most common manifestation is renovascular hypertension secondary to involvement of the renal artery. The FMD of the cervical artery, which mainly affects the ICA, is rarely symptomatic.<sup>6</sup> In addition, FMD is associated with a substantial risk of formation and rupture of aneurysms, as well as of arterial dissection and occlusion,<sup>3</sup> causing a great diversity of signs and symptoms.

Fibromuscular dysplasia is more common in females: Plouin and Kadian-Dodov, in 2016, reported a prevalence of 84%<sup>5</sup> and of 93.5%,<sup>1</sup> respectively; and Lather, in 2017, of 94.1%.<sup>3</sup>

Fibromuscular dysplasia can affect several arterial sites, the most common being the cervical and renal. There are several reports of involvement of the mesenteric, axillary, iliac, hepatic, intracranial and, in some cases, coronary arteries. In a case series review conducted by Persu, the

prevalence of involvement of  $\geq 2$  arterial sites varied from 16 to 28%. For this reason, the investigation of secondary lesions in patients with FMD is recommended, mainly in renal and cervical arteries.<sup>4</sup>

The clinical presentation of FMD is variable, not only by the multiple sites of involvement, but also by the large number of complications. Kadian-Dodov et al performed a study with 921 patients with FMD, in which 384 of them (41.7%) had aneurysms (of the cases) and arterial dissections (25.7%), in addition to 5.8% with simultaneous involvement. In this same study, the most commonly identified aneurysm sites reflected the most frequently affected arterial beds by the FMD: the renal (34%) and extracranial carotid arteries (31%). Intracranial circulation was the third most common site, occurring in 21.5% of all patients with aneurysms and in 4.7% of the 921 patients in the study.<sup>1</sup>

Lather, in 2017, performed an angiographic review of 669 women diagnosed with FMD in the USA, showing a 12.9% prevalence of intracranial aneurysms. There was no difference in the comparison between the different FMD sites: renal involvement with prevalence of 11.9%, cervical with 13.7%, and mixed with 13.2%.<sup>3</sup> If we compare these results with the prevalence in the general population, with rates of 3.2%,<sup>4</sup> the strong relation of the development of intracranial aneurysms with all forms of FMD becomes clear.

Spontaneous cervical artery dissections are a common cause of stroke in young and middle-aged adults and are associated with FMD in  $\sim 15\%$  of the cases,<sup>7</sup> and the most frequent dissections are in the extracranial carotid (63.7%) and vertebral arteries (20.7%)<sup>1</sup>

The management of FMD patients with drug therapy is well established, either with antiplatelet aggregators, anti-coagulants or antihypertensives. Surgical or endovascular treatment is indicated in patients with symptomatic or critical FMD, and may be directed to complications. In renal FMD, options are available in the presence of renovascular hypertension, renal ischemia atrophy, or in the presence of lesions in other sites. In cervical FMD, the indications are more individualized, because of the low possibility of disease progression and, generally, they are directed to the complications. Among the surgical possibilities are the use of coils for aneurysms and percutaneous transluminal angioplasty and stents for dissections or critical FMDs.<sup>8</sup>

Because FMD is a disease of unknown cause, it remains a therapeutic challenge because it does not have a specific

treatment. Currently, the therapy is directed to complications: aneurysms and arterial dissections. Although endovascular or surgical treatment is well-established for symptomatic complications or for those that cause a serious risk to the patient, more studies are needed regarding the treatment of asymptomatic lesions. The follow-up of these patients with DSA, Doppler or AngioCT/MRI is indissoluble, due to the great possibility of progression of the disease and its complications.

## Conclusions

The vascular fragility of patients with FMD of cervical arteries predisposes to dissection, stenosis and occlusion, in addition to being related to intracranial aneurysms. All these vascular malformations may present spontaneous complications, with cerebral hemorrhage or stroke, or secondary to iatrogenesis. Endovascular procedures should be well indicated, and reserved only for the treatment of symptomatic lesions.

## Conflicts of Interests

The authors have no conflicts of interests to declare.

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# A1–A2 Fenestration Tube Technique for Clipping a Ruptured Anterior Communicating Artery Aneurysm

## *A1–A2 Técnica de tubo de fenestração para clipagem de um aneurisma de artéria comunicante anterior roto*

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### Abstract

A fenestration tube is a clipping reconstruction technique that allows the preservation of critical vessels in aneurysm surgery. A patient with a ruptured anterior communicating artery (ACoA) aneurysm with a posterior projection was admitted to our neurosurgery unit. A right dominant A1 with rotation of the A2 fork was observed on preoperative computed tomography angiography (CTA). During surgery, we observed that the recurrent artery of Heubner branched off the A2 just distal to the neck of the aneurysm. Successful clipping was achieved by building an “A1–A2 fenestration tube,” with preservation of the recurrent artery and of the ACoA perforators. Surgical nuances and the advantages of fenestration tubes are discussed.

### Keywords

- ▶ intracranial aneurysm
- ▶ anterior communicating artery
- ▶ microsurgery

### Resumo

O tubo de fenestração é uma técnica de clipagem e reconstrução que permite a preservação de vasos críticos na cirurgia de aneurismas. Um paciente com um aneurisma da artéria comunicante anterior (ACoA) rompido com projeção posterior foi admitido em nosso serviço. A dominante direita A1 com rotação do garfo A2 foi observada na angiografia por tomografia computadorizada (ATC). Durante a cirurgia, a artéria recorrente foi observada ramificando-se a A2 apenas distal ao colo do aneurisma. Uma clipagem bem-sucedida foi alcançada criando um tubo de fenestração “A1–A2,” com preservação da artéria recorrente e dos perfurantes da ACoA. Nuances cirúrgicas e as vantagens dos tubos de fenestração são discutidas.

### Palavras-chave

- ▶ aneurisma intracraniano
- ▶ artéria comunicante anterior
- ▶ microcirurgia

### Introduction

Stacking multiple fenestrated clips creates a fenestration tube.<sup>1</sup> This clipping technique allows reconstruction of complex aneurysms while preserving vascular structures. A critical aspect of anterior communicating artery (ACoA) aneurysm surgery is preservation of the multiple arteries and perforators that are related to the ACoA complex.<sup>2</sup> The recurrent artery of Heubner arises from the proximal A2 segment in most cases

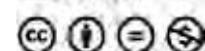
(58%), and its compromise or occlusion is associated with mediobasal striatum infarction and severe neurological deficits.<sup>3</sup> The surgeon must consider A1 dominance pattern,<sup>4</sup> projection of the dome of the aneurysm, ACoA angle,<sup>5</sup> and rotation of the A2 fork<sup>6</sup> when planning treatment strategy.<sup>7</sup> Aneurysms that project posteriorly with medial rotation of the dome of the aneurysm and of the A2 fork are a challenge because the neck of the aneurysm is hidden and perforators may be in the way of a potential clip.

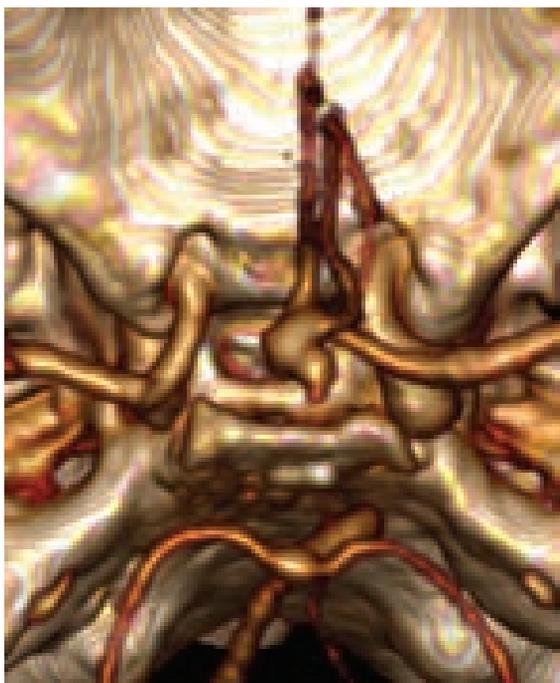
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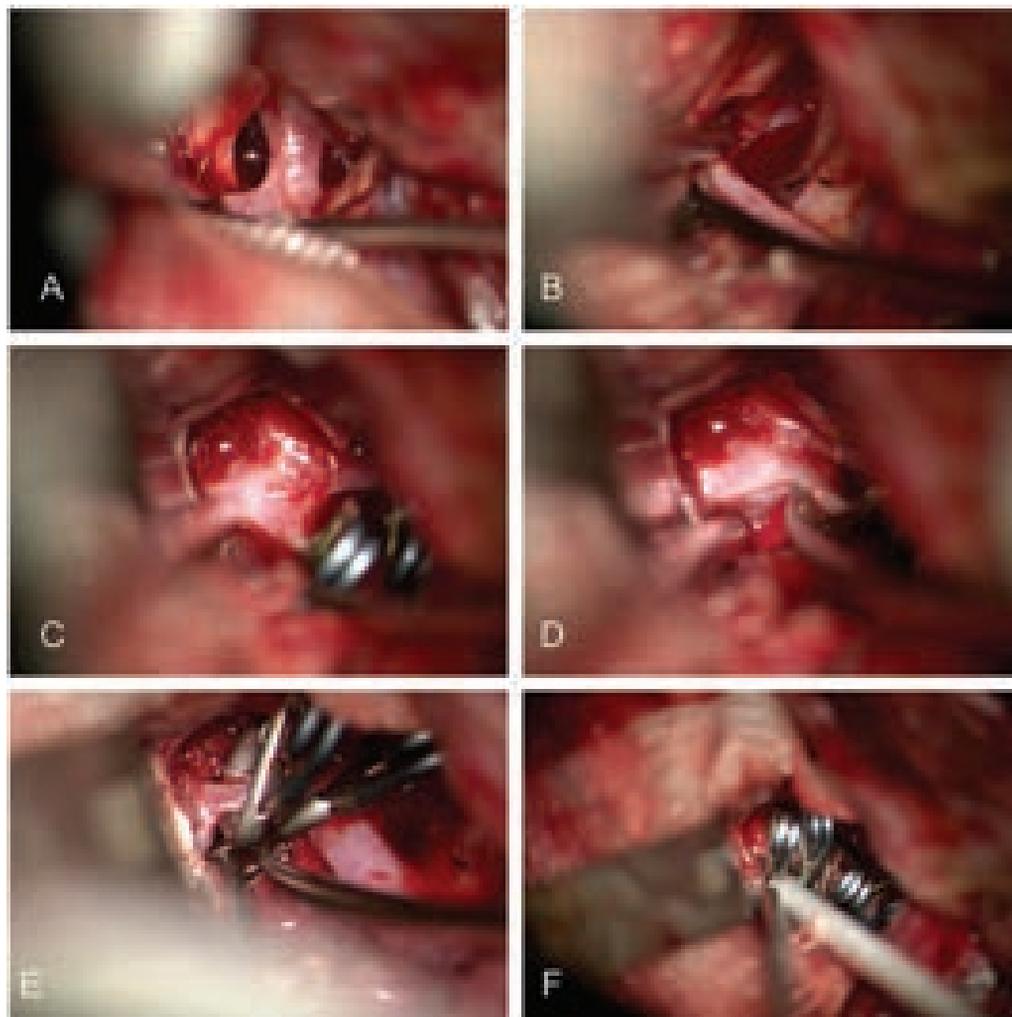
**Fig. 1** Preoperative computed tomography angiography.

## Case

A 69-year-old patient was admitted to our center with subarachnoid hemorrhage (World Federation of Neurosurgical Societies [WFNS] scale of 1). A computed tomography angiography (CTA) revealed a 4.1mm × 9.5mm ruptured ACoA aneurysm with posterior projection, right A1 dominance and rotation of the ACoA complex, with medial shift of the dome of the aneurysm (→**Fig. 1**). Surgical treatment was indicated.

## Surgical Technique

Under general anesthesia, the patient was placed on a Mayfield skull clamp, with 20° of head extension and 30° of contralateral rotation. A pterional approach was performed from the right side, with interfascial dissection of the temporalis muscle. The orbital roof was flattened with a cutting burr, to increase the angle of view. Under the surgical microscope, initial arachnoidal dissection of the optic and carotid cisterns (→**Fig. 2A**) allowed the exposure of the optic nerve and of the internal carotid artery. Blood clot cleansing with saline revealed the carotid bifurcation and ipsilateral



**Fig. 2** Surgical technique.



**Fig. 3** Postoperative angiography.

A1. Following this artery, we identified the optic chiasm and ACoA complex (►**Fig. 2B**). Lamina terminalis fenestration<sup>8</sup> and opening of Lilliequist membrane allowed release of cerebrospinal fluid (CSF). Partial gyrus rectus resection<sup>9</sup> was necessary to expose the bilateral A2. The origin of the recurrent artery of Heubner was localized at the proximal A2. A retractor was placed under the medial frontal lobe with gentle traction. Transitory clipping of both A1 arteries (<5 minutes of total ischemia time) was performed (►**Fig. 2C**), and dissection of the aneurysm neck extended medially to the ipsilateral A1–A2 junction (►**Fig. 2D**). Under direct visualization of the recurrent artery (►**Fig. 2E**), a fenestration tube was completed with two 4 mm length Yaşargil aneurysm fenestrated clips (Aesculap AG & Co., Tuttlingen, Germany) over the A2, and two 6 mm length fenestrated clips over the A1 (►**Fig. 2E**). Microdoppler sonography Mizuho Surgical Probe (Mizuho Inc., Tokyo, Japan) confirmed flow on both A2s, and absence of flow in the aneurysm sac.

A postoperative cerebral angiography confirmed the exclusion of the aneurysm from circulation (►**Fig. 3**), and patency of the arteries of the ACoA complex. The patient had a favorable outcome and was discharged with a Glasgow outcome scale of 5, three weeks after the surgery.

## Discussion

Drake was the first to introduce the use of fenestrated clips<sup>10</sup> in aneurysm surgery in 1969. By stacking multiple clips together, the surgeon can create a fenestration tube. This clipping technique allows treatment of complex aneurysms while preserving critical branches and perforators. In ACoA aneurysm surgery, traditional clipping with straight or curved clips may limit surgical exposure because the head

of the clip and the artery to be protected are in the way of the line of sight of the surgeon; fenestrated clips are located instead “on top” of the artery, so the exposure is preserved, and less dissection of branches from the aneurysm is needed.<sup>11</sup> This way, fenestration tubes allow the surgeon the goal of aneurysm exclusion in a more precise and anatomical fashion. Another advantage of fenestration tubes is the increased closing force of multiple clips that allow treatment of giant atherosclerotic or calcified aneurysms,<sup>12</sup> but great care must be taken to avoid neck tears.

In our case, a right side approach was chosen because the right A1 was dominant. After exposure of the aneurysm, the recurrent artery of Heubner was observed branching just distal to the neck of the aneurysm, at the lateral side of the A2. Fenestrated clips were applied under direct visualization of the recurrent artery, distally from A2 to A1. There were no complications, and exclusion of the aneurysm was possible, maintaining patency in all vessels.

The disadvantages of this technique are the increased number of clips used, and increased clipping time (but less time required to dissect vessels). In cases in which parent artery or branch sacrifice is inevitable, bypass techniques<sup>13</sup> should be considered.

Overall, we believe that fenestration tube is a valuable technique in the surgeon’s armamentarium, and offers advantages over traditional clipping in ACoA aneurysm surgery.

## Conclusion

The A1–A2 fenestration tube is a valuable clipping technique in ACoA aneurysm surgery.

### Conflicts of Interests

The authors have no conflicts of interests to declare.

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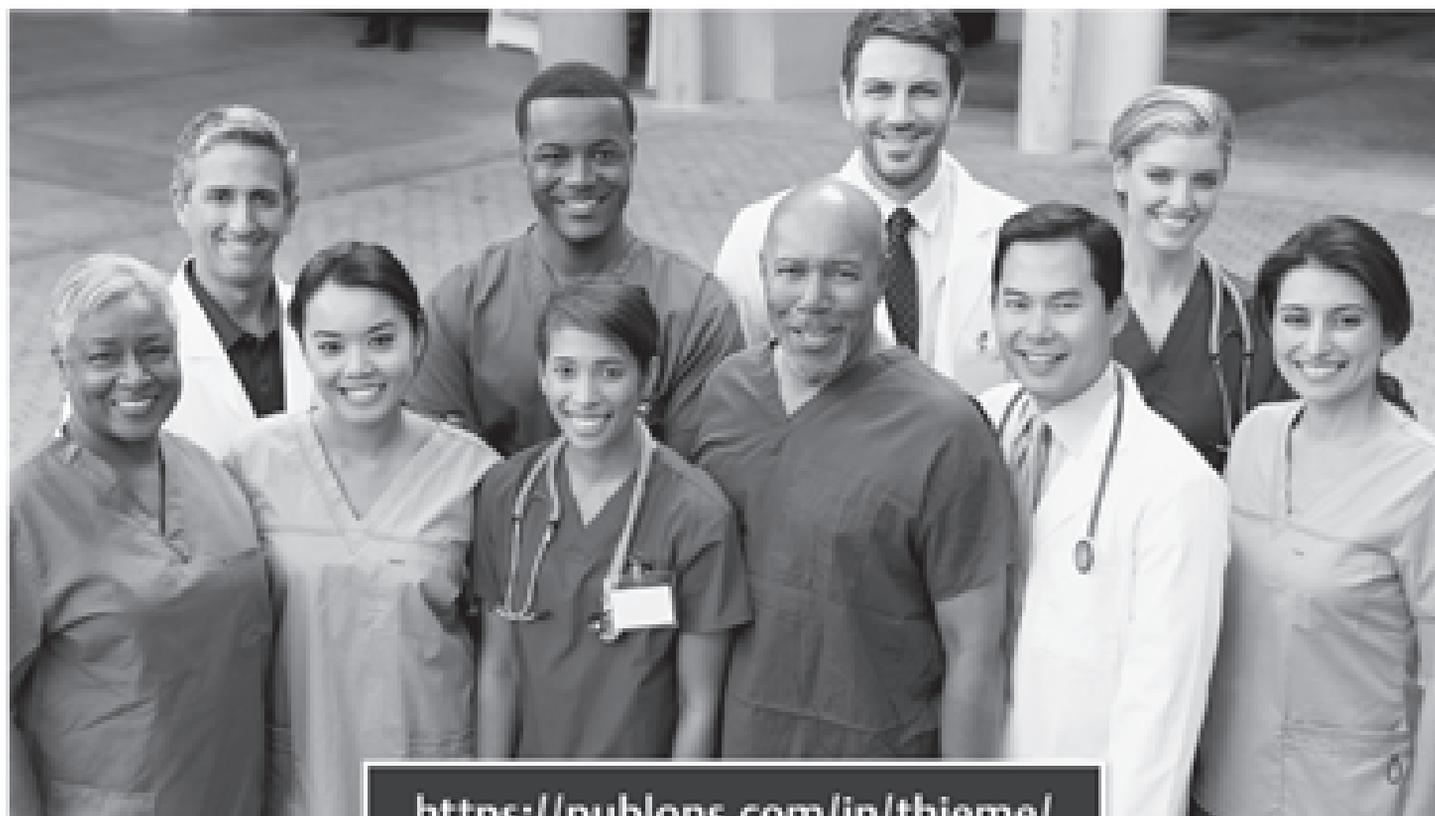
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