INTRACEREBRAL HEMATOMA CAUSED BY MOYAMOYA DISEASE: A CASE REPORT

Hematoma intracerebral causado por enfermedad de moyamoya: Reporte de caso

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ABSTRACT

Introduction: Moyamoya disease is a chronic occlusive cerebrovascular disease of unknown etiology, characterized by bilateral stenotic and occlusive changes in the terminal portion of the internal carotid, as well as the presence of an abnormal vascular network at the base of the brain. The diagnosis is made with magnetic resonance (MRI) and digital subtraction angiography (DSA), SPECT is useful in the therapeutic decision. The surgical treatment of choice is revascularization.
Clinical Case: A 50-year-old female patient from China, with the Glasgow Coma Scale (GCS) of 9, and a clinical picture of stroke. An admission brain tomography (CT) revealed a right temporal hematoma. Surgical evacuation of the intracerebral hematoma was performed. Cerebral angiography revealed distal stenosis of the internal carotid artery and its branches, being diagnosed with Moyamoya disease. The evolution was favorable, neither a motor deficit nor a decreased level of consciousness (GCS:15) was observed at the time of discharge. A subsequent revascularization surgery was indicated.
Conclusion: Moyamoya disease is a rare cause of intracerebral hematoma but should be suspected in adults of Asian descent. MRI and angiography are the diagnostic methods of choice. Surgical treatment is revascularization, which improves the prognosis.

Keywords: *Moyamoya Disease, Cerebral Hemorrhage, Stroke, Cerebral Angiography (Source: MeSH NLM)*

RESUMEN

Introducción: La enfermedad de Moyamoya es una enfermedad cerebrovascular oclusiva crónica de etiología desconocida, caracterizada por cambios estenóticos y oclusivos bilaterales de la porción terminal de la carótida interna, así como de la presencia de una red vascular anormal en la base del cerebro. El diagnóstico se realiza con resonancia magnética (RMN) y angiografía por substracción digital (ASD), el SPECT es útil en la decisión terapéutica. El tratamiento quirúrgico de elección es la revascularización.
Caso Clínico: Paciente mujer de 50 años, natural de China, con escala de Glasgow (EG) de ingreso de 9 puntos y cuadro clínico de ictus. Una tomografía cerebral (TAC) de ingreso evidenció un hematoma temporal derecho. Se realizó la evacuación quirúrgica del hematoma intracerebral. Una angiografía cerebral evidenció estenosis distal de la arteria carótida interna y sus ramas siendo diagnosticada de enfermedad de Moyamoya. La evolución fue favorable encontrándose al momento del alta sin déficit motor y en EG de 15, por lo que se indicó una posterior cirugía de revascularización.
Conclusión: La enfermedad de Moyamoya es una causa rara de hematoma intracerebral, pero debe sospecharse en adultos de ascendencia asiática. La resonancia magnética y la angiografía son los métodos diagnósticos de elección. El tratamiento quirúrgico es la revascularización la cual mejora el pronóstico.

Palabras Clave*: Enfermedad de Moyamoya, Hemorragia Cerebral, Angiografía Cerebral (Fuente: DeCS Bireme)*

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Moyamoya disease is a chronic occlusive cerebrovascular disease of unknown etiology, characterized by bilateral occlusive and stenotic changes in the terminal portion of the internal carotid artery and an abnormal vascular network at the base of the brain. [1, 2]

Moyamoya vessels occur in various pathologies, for which Mallory et al proposed a classification according to their genetics and etiology, classifying it into 4 types: type 1, classic moyamoya in Asian patients; type 2, moyamoya in white patients or associated with genetic syndromes such as neurofibromatosis or trisomy 21; type 3, moyamoya due to

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autoimmune diseases; type 4, moyamoya associated with vasculitis or atherosclerosis. [1]

The diagnosis of moyamoya angiopathy is based on visualization of the distal stenosis of one or both internal carotids and their branches, in addition to neovascularization. This it can be visualized on MRI and digital subtraction angiography (DSA). In addition, cerebral hemodynamics must be measured for an adequate therapeutic decision and patient follow-up, for which positron emission tomography (PET) or perfusion tomography is used. [4]

Treatment in the acute phase of ischemic and hemorrhagic events is symptomatic, but tissue plasminogen activating factor is contraindicated in ischemia due to the increased risk of hemorrhagic conversion. Antiplatelet therapy is prescribed only when revascularization therapy is postponed due to a recent infarction or mild disease. The final treatment is surgical revascularization. [3, 5]

In 1994, Rosell et al, reported that from 1947 to 1990 at the Almenara Hospital there were 3 cases of moyamoya disease, 2 males and 1 female, with an average age of 28 years, with recurrent subarachnoid hemorrhage and hemiparesis, with typical angiographic image. [7]

In this study, we report 1 case of Moyamoya disease in a patient of Asian origin who presented with an intracerebral hematoma and who was successfully treated surgically in our hospital.

CLINICAL CASE

History and examination: A 50-year-old female patient, a native of China, from Tarapoto, with a history of myomectomy and hypothyroidism, with a history of right hemicranial headache, oppressive and of abrupt onset, followed by sensorial disorder, so after a management initial in local hospital was transferred to this hospital. On physical examination: Soporous patient, who did not obey verbal orders, GCS: 9, without evident motor deficit, isochoric and photoreactive pupils, without cranial nerve deficit. Brain CT without contrast showed a 26cc volume right temporal hematoma in addition to an acute laminar subdural hematoma, a 5mm midline deviation, uncal herniation, perilesional edema, and ventricular invasion (*Figure 1*).

Treatment: A right temporal craniotomy was performed, and intracerebral hematoma was evacuated, without presenting intraoperative complications. Brain CT scan in the immediate postoperative period showed total evacuation of the right temporal hematoma and subdural hematoma. (*Figure 2*)

Clinical evolution: The patient evolved favorably in the postoperative period, being extubated, without presenting a motor deficit, without sensory deficit, the GA was 14 points.

A cerebral panangiography and bilateral external carotid angiography were performed, which showed moderate stenosis of the distal bilateral internal carotid artery (ICA), as well as moderate stenosis of both posterior cerebral arteries, in addition to tortuous collateral vessels dependent on the medial cerebral artery (MCA).), anterior cerebral artery (ACA) and posterior cerebral artery (PCA), without the presence of collaterals of the external carotid artery (ECA), findings compatible with type 2 moyamoya Suzuki disease (*Figure 3*).

A brain MRI with contrast was also performed, which showed hypodensities in both basal nuclei in the SWI sequence, in relation to stasis due to dilated drainage veins; likewise, in the T2 sequence moyamoya vessels were evidenced. (*Figure 4*)

The patient was discharged in good clinical condition, in Glasgow 15 and without motor deficit, with an indication to complete revascularization treatment in her hometown.

DISCUSSION

Moyamoya disease was first described by Takeuchi and Shimizu in 1957. This disease is defined as a cerebrovascular disorder in which there is idiopathic stenosis of the internal carotid artery that generates the formation of aberrant compensatory vessels. On the other hand, Moyamoya syndrome or near-Moyamoya disease is defined as the presence of moyamoya vessels that arise due to therapeutic cranial irradiation or in association with other diseases such

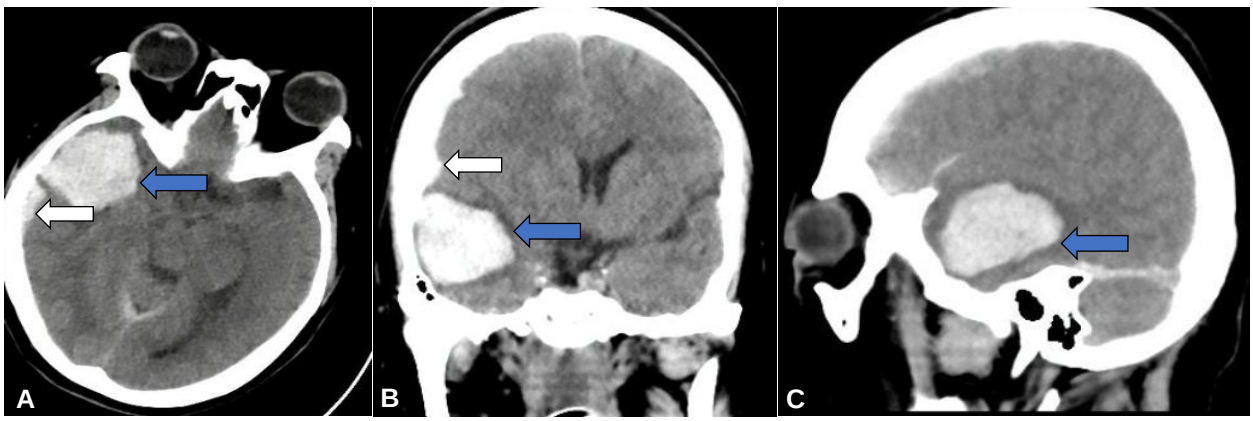

Fig 1. Brain tomography (CT) without contrast in (A) axial view, (B) coronal view and (C) sagittal view, where a right temporal hematoma (blue arrow) with perilesional edema, mass effect and midline deviation is showed; in addition to a laminar subdural hematoma (white arrow).

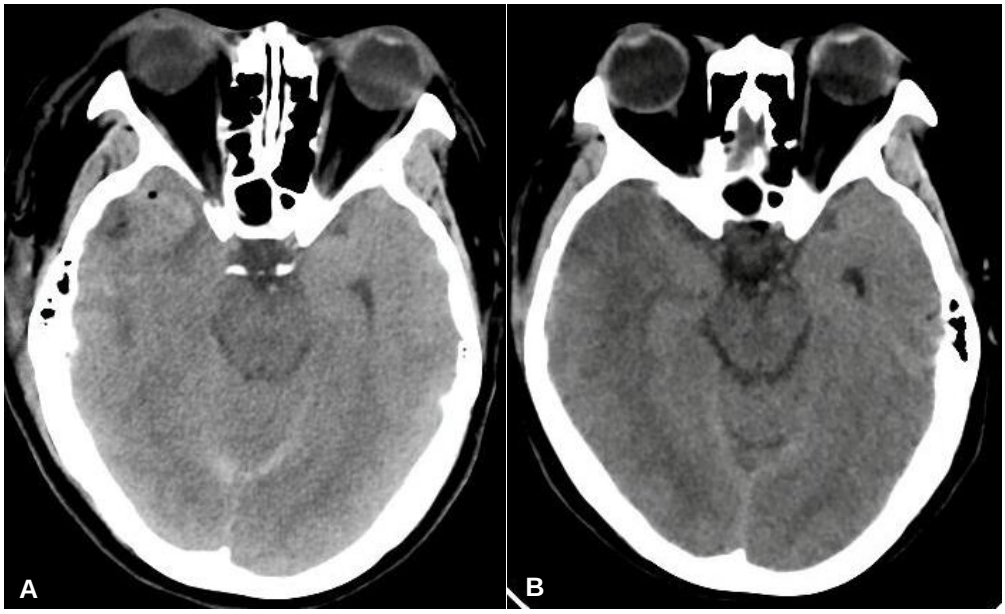

Fig 2. Control cerebral tomography without contrast in axial section: **(A)** Control brain tomography in the immediate postoperative period that shows complete evacuation of the cerebral hematoma without the presence of complications. **(B)** Control brain tomography after the 1st month of surgery showing slight malacia in the right temporal pole, without other significant findings.

as trisomy 21, sickle cell disease, meningitis, autoimmune disease, and neurofibromatosis. [1,2]

The prevalence of Moyamoya syndrome in Japan is estimated to be 0.34 patients per 100,000 inhabitants, constituting only 5.4% of Moyamoya cases.1 The highest prevalence disease is found in Japan and reaches a prevalence of 3.16-10.5 / 100,000 inhabitants and an incidence of 0.35-1.13 / 100,000 / year, with a higher prevalence in women of 2 to 1. It has a bimodal distribution with a first peak in childhood between 5 and 10 years old, and a second peak during the fourth decade of life, as is the case of our patient. Family history can be identified in 10-15% of cases in Japan. [3]

The etiology is unknown, and the disease is characterized mainly by occlusive stenotic changes in the terminal ICA and development of abnormal vessels at the base of the brain. Histopathological studies show hyperplasia of the intimal layer, thinning of the middle layer, and undulation of the internal elastic lamina. The genetic analysis showed that the RNF213 gene in the 17q25 region was involved in the development of the disease in East Asian patients. [2]

The diagnostic criteria are based on criteria obtained from a digital subtraction angiography: 1) stenosis and occlusion of

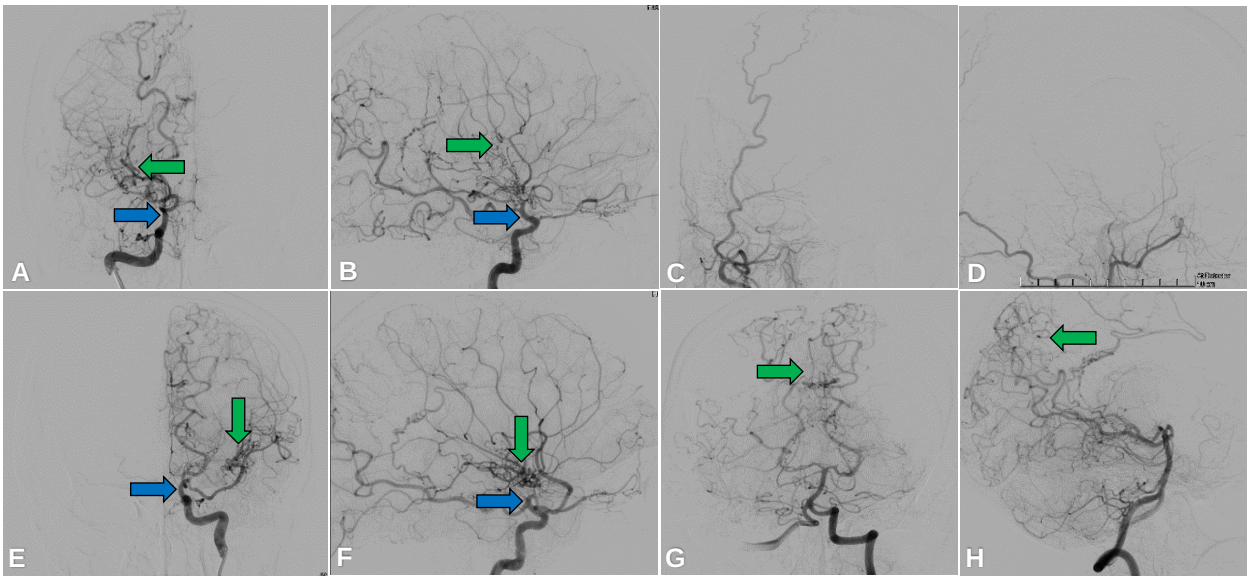

Fig 3. Cerebral panangiography and bilateral external carotid angiography showing stenosis of the distal ICA (blue arrow) and moyamoya vessels (green arrow). **(A)** Angiography of the right ICA in anteroposterior (AP) view. **(B)** Angiography of the right ICA in lateral view. **(C)** Angiography of right ECA in AP view. **(D)** Angiography of the right ECA in lateral view. **(E)** Angiography of the left ICA in AP view. **(F)** Angiography of the left ICA in lateral view. **(G)** Angiography of the left vertebral artery in AP view. **(H)** Angiography of the left vertebral artery in lateral view.

the terminal portion of the ICA or the proximal portions of the ACA and MCA, 2) development of abnormal vascular networks close to the stenotic or occlusive lesions in the arterial phase, 3) bilateral lesion. But MRI criteria can also be used: 1) MRI angiography showing stenosis or occlusion of the terminal portion of the ICA or of the proximal portions of the MCA and ACA, 2) presence of abnormal vascular networks near the stenotic lesion or occlusive, or presence of 2 or more flow voids in the basal ganglia in each hemisphere.[2]

The severity of the disease is classified into 6 progressive stages according to Suzuki: I is a narrowing of the terminal ICA, II is the appearance of deep moyamoya collateral vessels, III is the progression of moyamoya collateral vessels, IV is the appearance of moyamoya vessels transdural collaterals arising from the external carotid artery (ECA), V is the progression of transdural collateral vessels arising from the ECA with reduction of the moyamoya vessels, and finally VI is the occlusion of the ICA and the disappearance of the vessels Moyamoya collaterals.[3] In our case, the patient did not have ECA collaterals, but she did have the recent appearance of moyamoya vessels associated with distal stenosis of both internal carotids, which is why she was classified as Suzuki II.

For diagnosis, in addition to angiography and MRI, hemodynamic evaluations should be performed using semi-quantitative techniques such as PET or SPECT.[4] In SPECT, the injection of I-IMP (IN-isopropyl-p-iodoamphetamine) is used in which detects cerebral blood flow (CBF) before and 15 minutes after intravenous injection of 1000mg of acetazolamide. This makes it possible to measure cerebrovascular reactivity (CVR), which is measured in percentage, subtracting the FSC after acetazolamide and FSC before acetazolamide, all between the FSC before acetazolamide. If the CVR is less than 14% it means that it is decreased. [5]

Czanbanka et al proposed a new system for adults with Moyamoya disease called the Berlin scoring system, where angiographic, resonance and hemodynamic criteria are taken, which predicts postoperative neurological morbidity in Japanese adult patients with this disease. Digital subtraction angiography gives 1 point if there are moyamoya vessels and steno-occlusive lesion, 2 points if there are moyamoya vessels with steno-occlusive lesion and an intracranial anastomosis route, 3 points if there are moyamoya vessels with a steno-occlusive lesion and an extra-intracranial anastomosis route. In the MRI, 1 point is given if there are signs of heart attack, hemorrhage, or brain atrophy, and 0 points if there are none of these. Regarding hemodynamic studies, 2 points are given if there is a theft phenomenon and 0 points if there is not. [5]

In the Berlin grading system, a minimum score of 1 and a maximum of 6 points is given, when adding the numerical values of the 3 variables, finally giving 3 degrees of moyamoya disease: grade I or mild if it gives 1 or 2 points, grade II or moderate if it gives 3 or 4 points, grade III or severe if it gives 5 or 6 points. The grade is measured for each hemisphere, so the patient may have a different grade in each hemisphere.5 When applying the Berlin grade to our patient, she gave 2 points, qualifying as a mild moyamoya disease.

Asymptomatic moyamoya disease is a progressive disease with an annual stroke rate less than or equal to 13.3%, for which the indications for treatment were reviewed by the prospective study AMORE (Asymptomatic Moyamoya Registry). A 10-year follow-up in Japan showed that the use of antiplatelets does not influence the stroke rate in patients with Moyamoya disease. [3]

Revascularization surgery is the most effective treatment for hemorrhagic moyamoya disease, demonstrated by randomized and randomized clinical trials, and it is probably the most effective treatment for ischemic moyamoya disease as well, but clinical trials are still lacking on the latter issue. The primary objective of

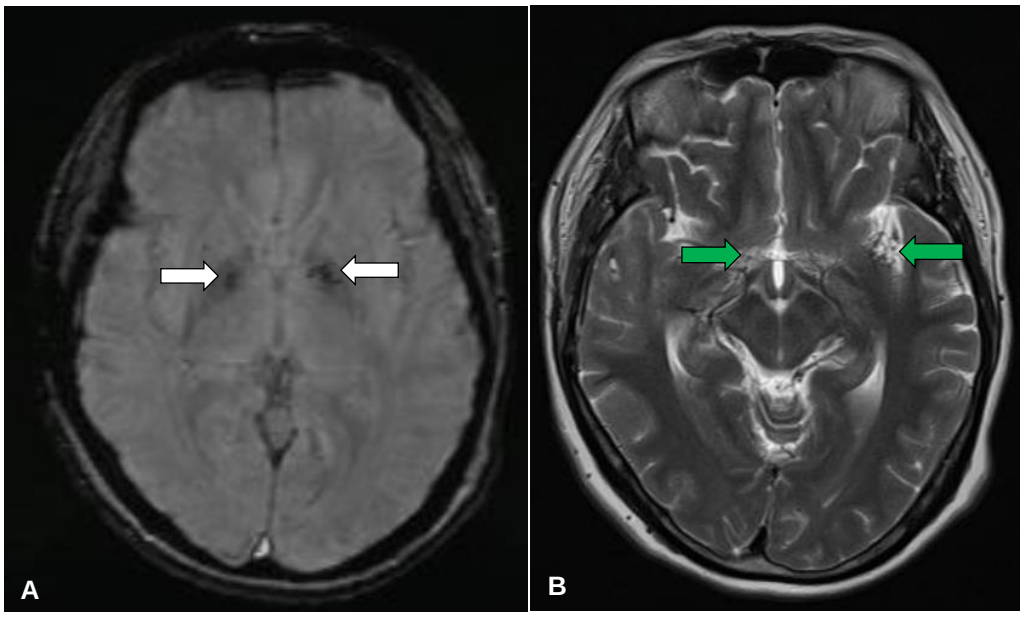

Fig 4. Brain magnetic resonance imaging (MRI) with contrast in axial section: **(A)** SWI sequence showing hypointensities in bilateral basal ganglia (white arrow), corresponding to stasis of dilated draining veins. **(B)** T2 sequence showing moyamoya vessels (green arrow) in the territory of the bilateral anterior cerebral artery and the territory of the left middle cerebral artery.

revascularization is to restore blood supply to stabilize cerebrovascular hemodynamics, thus generating regression of the fragile moyamoya vessels, preventing bleeding. In addition, normalization and / or improvement of cerebral hemodynamics can result in secondary prevention of stroke and improvement of neurological and neurocognitive outcome.[3] Despite the fact that our patient had a Berlin grade I, she had already had a hemorrhagic event, therefore which revascularization surgery is indicated in this patient to reduce associated morbidity and mortality.

In surgical practice, 3 surgical strategies have been applied: indirect revascularization, direct revascularization, and combined revascularization. But there is still no consensus on which is the best revascularization technique. However, current studies indicate that revascularization strategies, including direct bypass, can be applied in moyamoya patients of all ages if technically feasible. [3]

Direct revascularization consists of creating a direct bypass between the superficial temporal artery (STA) and the ACA or the MCA or the PCA, also between the occipital artery and the PCA. Indirect revascularization is based on neovascularization of the cortical surface via angiogenic mechanisms through pedicle grafts, the most common being encephalomyosinangiosis, but there are also variants such as encephaloarteriosinangiosis, encephalomial arteriosinangiosis, encephalodurosynangiosis, encephaloduro arteriosinangioseriosis, encephaloduro-arteriosinangioseriosis, encephalo-angioarteriosinangiosis, encephalo-angioarteriosinangiosis. [3]

A multicenter study in Japan, which was the first prospective study to evaluate the prognosis of asymptomatic patients, evaluated 40 patients, 6 of them underwent revascularization using ATS-MCA anastomosis, and the other 34 underwent conservative follow-up. The 6 patients who underwent surgical treatment did not present cerebrovascular events. Of the other 34, 7 had cerebrovascular events, including 3 transient ischemic attacks, 1 cerebral infarction, and 3 with intracerebral hematoma. Therefore, they conclude that the annual risk of cerebrovascular events was 5.7% and ischemic stroke was 3.2%. [6]

The alteration of cerebral hemodynamics at the time of diagnosis is highly associated with ischemic episodes, a finding that has statistical significance. The disease can progress up to 20% of patients in the 6-year follow-up. Occlusive arterial lesions progressed both in the anterior and posterior circulation, both in unilateral and bilateral lesions, as well as in symptomatic and asymptomatic patients. Multivariate analysis showed that female sex is an independent risk factor for progression. Therefore, asymptomatic disease is not so silent, which is why it is essential to repeat MRI and MRI in a serial manner to detect disease progression, and to be able to perform surgical treatments if necessary. [6]

Our patient decided to return to her hometown to complete her revascularization surgical treatment, so the follow-up was interrupted. This is the fourth case of moyamoya disease in the Neurosurgery service of the Almenara Hospital, adding to the 3 cases reported by Rosell et al. [7] Advances in diagnostic tests, including cerebral hemodynamic studies, allowed the diagnosis, as well as an ideal case management, considering that moyamoya disease is a rare pathology in our population.

CONCLUSION

Moyamoya disease is a rare cause of spontaneous brain hematoma in adult patients but should be suspected in those of Asian descent. The diagnosis is based on the findings of MRI and digital subtraction angiography. Brain hemodynamic studies allow assessing the need for surgical treatment. The surgical treatment of choice is direct, indirect, or combined revascularization, which improves the prognosis of this type of patient.

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Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Authors Contributions
Conception and design: All authors. *Drafting the article:* Vargas. Critically revising the article: Palacios, Saal. *Reviewed submitted version of manuscript:* Vargas. *Approved the final version of the manuscript on behalf of all authors:* Vargas.

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