

# Brain Asymmetry: Diagnostic Approach

Asimetría cerebral: enfoque diagnóstico



Alejandro Zuluaga Santamaría<sup>1</sup>  
Sergio Vargas<sup>2</sup>  
Sandra Arango<sup>3</sup>  
Ricardo Uribe<sup>3</sup>



## Key words (MeSH)

Facial asymmetry  
Encephalitis  
Hemimegalencephaly



## Palabras clave (DeCS)

Asimetría facial  
Encefalitis  
Hemimegalencefalia

## Summary

Brain asymmetries are a common finding for radiologists and may be caused by multiple etiologies, including congenital and acquired causes. Congenital causes are conditions that appear during the fetal life, while the acquired causes occur after birth and usually before 2 years of age. In cross-sectional imaging, brain asymmetries are characterized by enlargement or atrophy of all or a part of a cerebral hemisphere. This variability in brain volume may be caused by physiological causes or by non-physiological causes that are secondary to congenital and acquired processes, causing an asymmetry in the hemispheres. In this article we propose a practical diagnostic approach to brain asymmetries based on the evaluation of the cerebral hemisphere and ventricular system size.

## Resumen

Las asimetrías cerebrales son entidades clínicas frecuentes encontradas en la práctica diaria del radiólogo. Son causadas por diversas etiologías que pueden ser agrupadas en congénitas y adquiridas. Las congénitas son aquellas que se presentan por condiciones durante la vida intrauterina, mientras que las adquiridas ocurren después del nacimiento, usualmente antes de los 2 años de edad. En los estudios por imágenes, las asimetrías cerebrales están dadas por aumento o disminución del volumen de la totalidad o de parte de un hemisferio cerebral y esta variabilidad en el volumen cerebral puede ser por causas fisiológicas, también conocidas como normales o por causas no fisiológicas secundarias a procesos congénitos o adquiridos, que dan lugar a desproporción de un hemisferio cerebral con respecto al otro. En este artículo se propone un enfoque diagnóstico práctico para abordar este hallazgo, donde, evaluando el hemisferio cerebral y el tamaño ventricular, se puede desplegar una serie de posibles diagnósticos diferenciales.

## Introduction

When talking about cerebral asymmetry, it must be taken into account that this finding may be a variation of the normality or secondary to some pathological process; the latter may be congenital due to exposure to a noxa before birth, or acquired by exposure, generally, before the two years of life. The assessment of brain volume and ventricular size can guide the possible cause of asymmetry and narrow the range of differential diagnoses. The term cerebral asymmetry also includes physiological asymmetry, hemimegalencephaly and hemiatrophy cerebral.

## Definitions

### *Physiological asymmetry or minimal brain asymmetry*

It corresponds to a variant of normality and has been recognized by anatomists, anthropologists, neurologists and radiologists. It has been found through different diagnostic methods – pneumography, ultrasound, tomography, angiography and magnetic resonance imaging (MRI). This condition is unrelated with any type of brain damage and is also associated with cranial malformations (1). Such a «normal» asymmetry can



<sup>1</sup>Radiologist doctor, CediMed, Las Vegas Clinic. Radiology professor CES university and Universidad Pontificia Bolivariana. Medellín, Colombia.

<sup>2</sup>Neuroradiologist, CediMed, UPB University professor. Medellín, Colombia.

<sup>3</sup>Radiologist resident, CES University. Medellín, Colombia.

compromise a complete hemisphere, one or more lobes or a giral area. The most frequently affected area is the retroinsular cortex, corresponding to the posterior language area (2,3) (Figure 1).

**Non physiological brain asymmetry**

It is a clinico-radiological condition resulting from several congenital or acquired processes, which give rise to a disproportion of the hemisphere with respect to the other. An example of this non-physiological asymmetry is the one that appears during a cerebral infarction in subacute phase (Figure 2).

**Brain hemiatrophy (BHA)**

It is a rare entity (4). The term atrophy connotes an irreversible loss of brain tissue (5). Cerebral hemiatrophy or unilateral cerebral atrophy is the terminal stage of various pathologies that culminate in the atrophy or hypoplasia of a single cerebral hemisphere. Cognitive disorders, changes in behavior, hemiplegia, seizures and emotional deficits are possible functional associations of cerebral hemiatrophy (6,7).

Alper and Dear, in 1939, described that cerebral hemiatrophy can be given for primary or secondary causes. Primary BHA is the true cerebral hemiatrophy or unilateral cerebral hypoplasia, as it actually corresponds to the lack of brain development. In this, damage occurs in utero, with consequent displacement of the structures of the midline to the side of the pathology and associated with a lack of sulcus prominences. These characteristics differentiate the primary BHA from the secondary, a condition in which Identified causes of cerebrovascular type, inflammatory processes or cranial trauma, among others (4,8,9). Possible causes of cerebral hemiatrophy are shown in Table 1.

**Table 1. Possible causes of BHA**

Congenital causes	Acquired causes
Idiopathic	Trauma
Intrauterine vascular trauma	Perinatal intracranial hemorrhage
	Rasmussen encephalitis
	Post-cranial cerebral hemiatrophy, prolonged febrile seizures
	Due to infections: <ul style="list-style-type: none"> <li>• TORCH</li> <li>• HIV</li> </ul>
	Vascular: <ul style="list-style-type: none"> <li>• Dyke Davidoff Masson syndrome</li> <li>• Sturge Weber syndrome</li> <li>• Cerebral cerebellar crossed diaschisis</li> <li>• Commissural diaschisis</li> <li>• Syndrome of hemiplegia, hemiatrophy and hemiconvulsion</li> <li>• Ischemia</li> </ul>
	Neoplasms: <ul style="list-style-type: none"> <li>• Basal ganglia germinoma</li> </ul>
	Radiation
	Facomatosis: <ul style="list-style-type: none"> <li>• Neurofibromatosis</li> </ul>
	Miscellaneous: <ul style="list-style-type: none"> <li>• Linear nevus syndrome</li> <li>• Síndrome de Fishman</li> <li>• Silver-Russell syndrome</li> <li>• Child hemiplegia syndrome</li> <li>• Trephine syndrome</li> <li>• Congenital malformation</li> <li>• Coarctation of the intrauterine aorta</li> <li>• Perinatal hypoxia-anoxia</li> <li>• Mitochondrial encephalopathy</li> <li>• Lactic acidosis</li> <li>• MELES (4,8,10-12)</li> </ul>

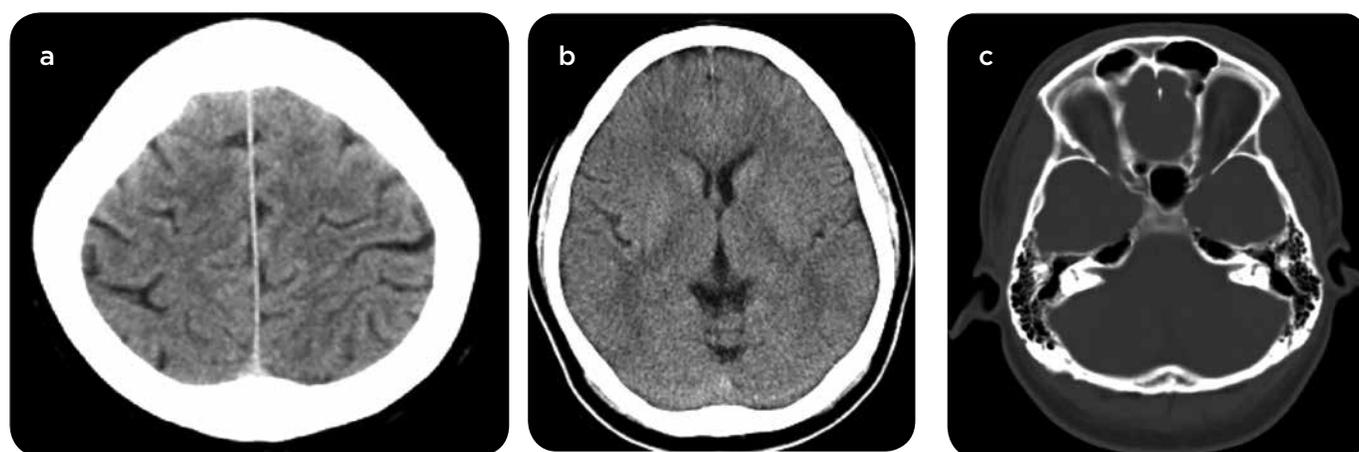


Figure 1. a, b y c) Physiological asymmetry. A 34-year-old woman consulted for headache, with no history of importance. Non-contrasted CT images show a slight asymmetry between the cerebral grooves of both hemispheres and in the frontal horns of the lateral ventricles, with symmetry in the pneumatization of the frontal bones and mastoid cells.

Neuroimaging of CT and MRI plays an important role in the diagnosis of this entity. When we are faced with a patient with cerebral parenchyma atrophy, seizures, hemiparesis, hemiplegia and craniofacial asymmetry, MRI is the modality of choice to evaluate the etiology and extent of the lesion (13). Below is a brief description of some of the causes of BHA:

### ***Dyke Davidoff Masson Syndrome (DDMS)***

In 1933 Dyke, Davidoff and Masson described, on radiographs of the skull and pneumoencephalographs, changes in a series of nine patients characterized clinically by hemiparesis / hemiplegia, seizures, facial asymmetry and mental retardation (8).

DDMS or hemispheric infarction is an atrophy or hypoplasia of a cerebral hemisphere secondary to a vascular alteration as a congenital or acquired ischemic disease, trauma or inflammation which occurs during childhood, with compensatory hypertrophy of the ipsilateral cranial vault (4,8,14).

In the in utero arterial occlusion the compromise of the middle cerebral artery itself, has been described as the most frequent cause. Also, it has been described that DDMS may be secondary to aortic coarctation, a condition that may decrease the flow of the middle cerebral artery in an indirect way (4,8).

In neuroimaging there is a decrease in the size of the cerebral hemisphere, thickening of the ipsilateral cranial vault, increased development of ipsilateral sinuses and contralateral or diffuse cerebellar atrophy (Figure 3).

Changes in the cranial vault only occur when the brain damage occurs before age three and are visible approximately nine months after vascular damage (8). Due to the delayed growth of the cerebral parenchyma, compensatory changes, such as enlargement of the sinuses of predominance in the frontal sinus and increase in the thickness of the diploe with elevation of the major wing of the sphenoid, petrous crest and sphenoidal plane (14).

Treatment of this entity is symptomatic, but in case of incapacitating seizures and hemiplegia, hemispherectomy is indicated with which symptoms are controlled in approximately 85 % of the cases (9).

### ***Sturge-Weber Syndrome***

It is also known as encephalotrigeminal angiomas. This is a congenital abnormality consisting of malformation of the cortical veins of the fetus, resulting in progressive venous occlusion with subsequent chronic venous ischemia.

It has an incidence of 1 case per 20,000-50,000 people (15). It is considered a facomatosis, characterized by facial spots “in wine from Porto” and facial angiomas (16). Neuroimaging shows subcortical calcifications associated with loss of cerebral parenchymal volume, with growth of the cranial vault and paranasal sinuses. The ipsilateral choroid plexus can be enlarged (17,18) (Figure 4).

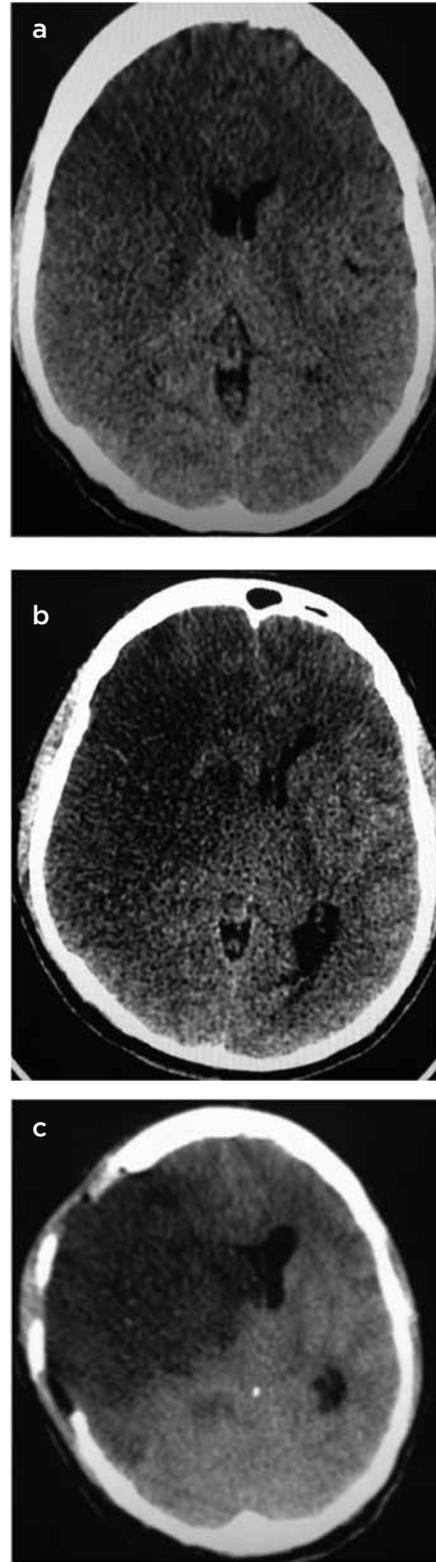


Figure 2. a) Non-physiological asymmetry. Non-contrasted CT scan. Patient with a 6-hour evolution of left hemiparesis. Hypodensity is evident in the right frontotemporal region without significant changes in brain volume or in the ventricles; b) three days after; c) one week later. Cytotoxic edema is observed causing increased right hemispheric cerebral volume and compression of the adjacent ventricle.

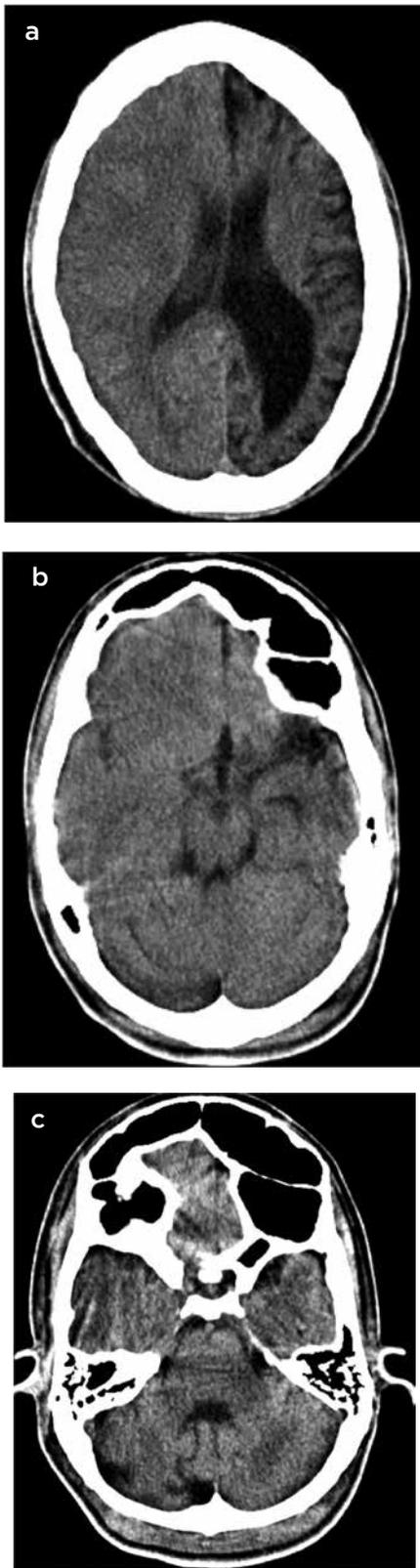


Figure 3. Dyke Davidoff Mansson syndrome. a) Decrease in the left cerebral parenchyma and thickening of the ipsilateral cranial vault. b) Thickening of the left skull with compensatory hypertrophy effect of the ipsilateral sinuses. c) Atrophy of right cerebellar hemisphere.

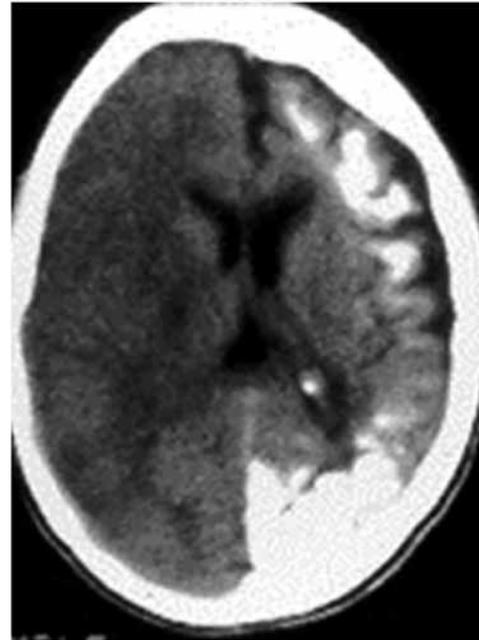


Figure 4. Sturge-Weber syndrome. CT without contrast medium, subcortical calcifications in the left cerebral hemisphere, associated to loss of parenchymal volume on this side and increase in the thickness of the ipsilateral frontal bone. In addition, slight left ventricular dilation.

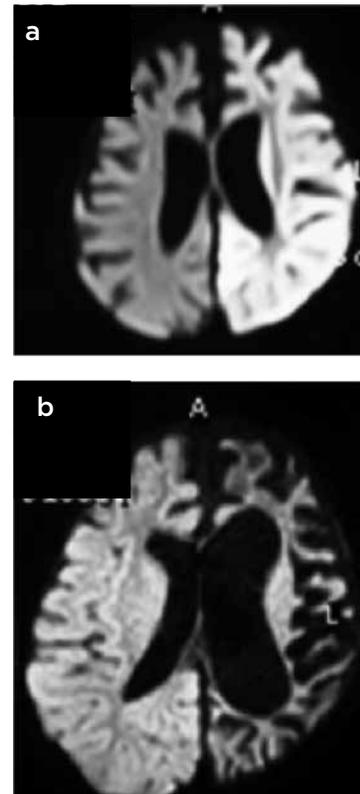


Figure 5. Rasmussen encephalitis. a) MR sequence DWI b1000. Restriction to diffusion in the left cerebral hemisphere is evidenced. b) DWI sequence b1000 of the same patient two years later. There is unilateral cortical atrophy with ventricular dilatation ex vacuo on the same side. There is no restriction on dissemination.

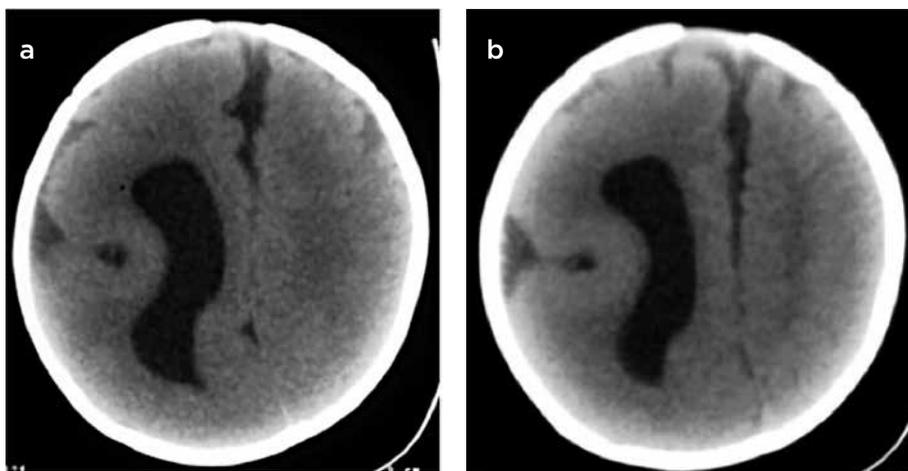


Figure 6. a y b) Hemimegalencephaly. Simple skull CT. 3 years old patient with antecedent of convulsions. Increase in the cerebral volume of the right hemisphere, associated with ipsilateral ventricular dilation and agiria-paquigiria complex, configuration in the form of "eight" of the Silviana crust and "V" shape.

### ***Rasmussen's encephalitis***

It is a focal encephalopathy characterized by being a chronic inflammation pathology of unknown origin, usually affecting one cerebral hemisphere. Eighty-five per cent of cases are in children under 10 years. In general, it is considered an entity of unknown cause, but appears to be related to infection by the Epstein-Barr and cytomegalovirus.

In MRI, in the T1-weighted sequences, unilateral cortical atrophy is observed associated with chronic ventricular dilatation ex vacuo and in T2-weighted sequences, a high signal is identified that compromises the pathological cerebral hemisphere. In the diffusion sequences there exists restriction to the same (in acute phase).

Contrast-enhanced tomography does not show significant enhancement in the compromised cerebral hemisphere (Figure 5).

### ***Herpetic encephalitis***

It is an entity caused by the herpes simplex virus (HSV) type 1. In neonates and infants, the transmission is transplacental, or during childbirth (19,20).

Cerebral hemiatrophy is due to unilateral involvement during initial stages of fetal life, with preference for the limbic system (olfactory tract, temporal lobes, cingulate gyrus and insular cortex). In order from greatest to least, the condition occurs in the inferior temporal lobe, frontal lobe and parietal lobes (21).

MRI is essential for the evaluation of infants with infection because both the cortex and the white matter can be widely compromised (22).

CT is useful for assessing periventricular calcifications, mostly from the gray-white matter union (23).

### ***Hemiegaloencephalia or unilateral megalencephalia***

Described for the first time by Sim in 1835. Its etiology is unknown and is not associated with chromosomal alterations. It presents with alterations in neuronal migration, although it can also be considered a primary disorder of proliferation, in which neurons that are not capable of

forming synaptic connections, are not eliminated and are accumulated.

Unlike other brain dysgenesis, it shows an extreme asymmetry that does not correspond to any of the normal phases of development of the brain.

It is characterized by a hamartomatous growth of part or all of a hemisphere where the compromised hemisphere may have focal or diffuse migration defects with areas of polymicrogyria, paquigiria and heterotopias. Three types of hemimegalencephaly are described: Isolated, syndromic and total:

*Isolated form:* occurs as a sporadic disorder without hemicorporal hypertrophy or cutaneous or systemic affection.

*Syndromic form:* is associated with other diseases and can occur as hemi-hypertrophy of all or part of the ipsilateral body.

*Total hemimegalencephaly:* in which there is also ipsilateral enlargement of half of the brain stem and the cerebellum.

It has no predilection for race or gender and is in 0.1 to 0.3 % of the cases of epilepsy diagnosed during childhood. In 90 % of the patients it is manifested with generalized focal seizures, delayed development, hemiparesis and hemianopsia.

Normocephaly is common in this entity; however, it may show macrocephaly. It is not associated with symptoms of endocranial hypertension. Neuroimaging shows an increase in the size of the lateral ventricle, deep grooves with wide convolutions, contralateral displacement of the sickle, calcifications of the white matter and anomalous venous development. The cortex of the compromised hemisphere may be normal or with polymicrogyria, lysencephaly, agiria/paquigiria or heterotopias (24) (Figure 6).

### ***Trephine syndrome***

Described by Grant in 1939 (25). It is also known as sunken flap syndrome (26,27), and has an incidence of approximately 13 % (26) in patients who are given decompressive craniectomy. It was published in the study by Yang and colleagues where the syndrome was diagnosed in 14 of 108 craniectomy patients (27).

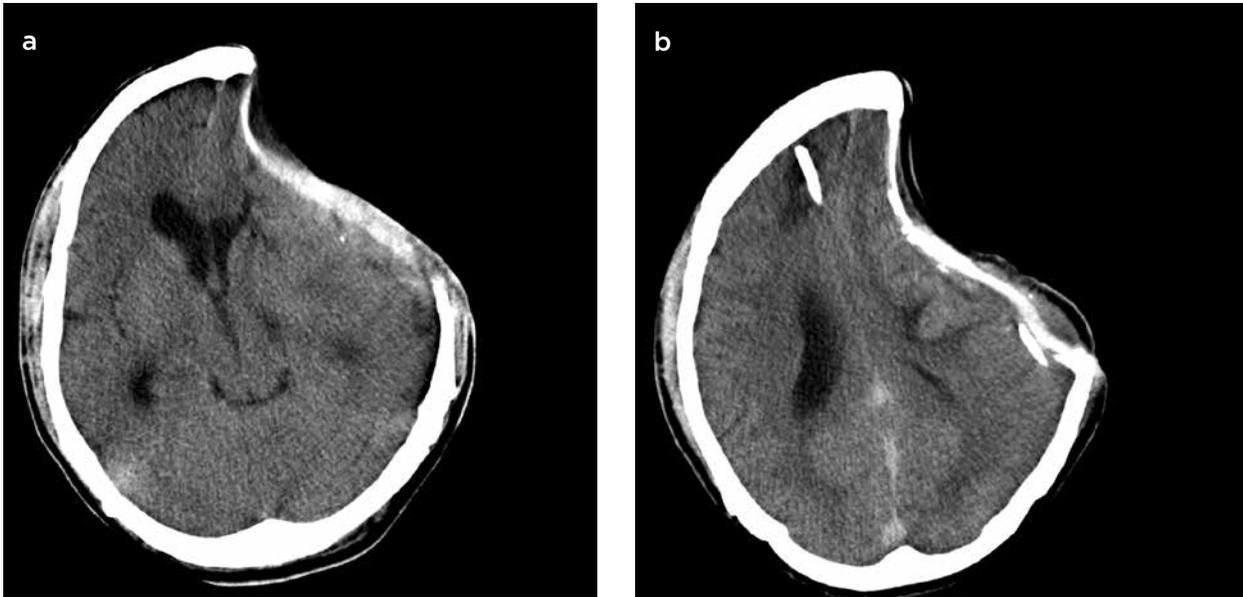


Figure 7. a y b) Trepine syndrome. A 23-year-old male patient, with a history of severe encephalocranial trauma and craniectomy 1 year before. Consultation due to seizures. Simple tomography: left frontal craniectomy with ventriculo-peritoneal shunt that enters by the right frontal lobe. Deviation from the midline to the right with slight collapse of the lateral ventricle and concave deformity of the left frontal and parietal lobe

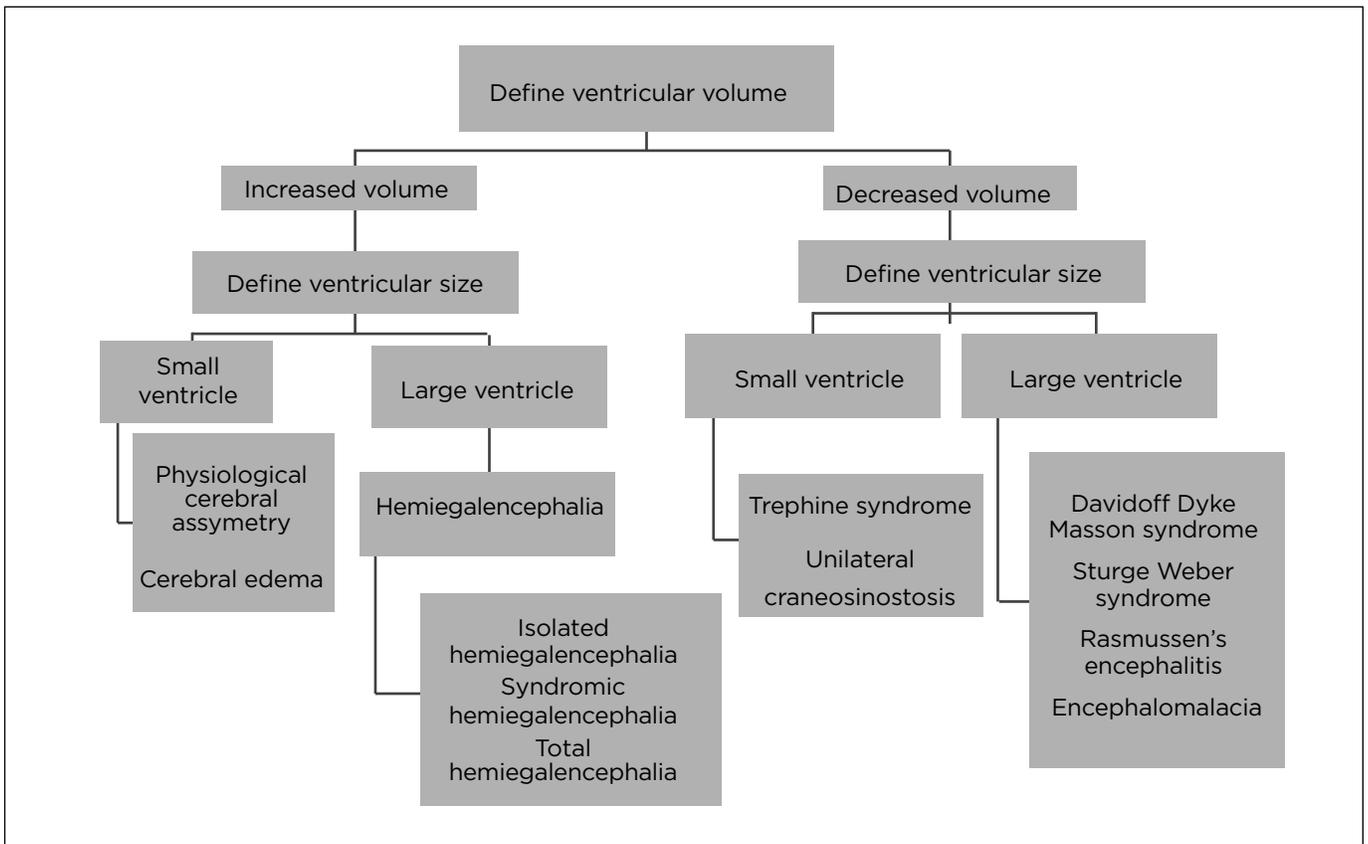


Figure 8. Diagnostic approach algorithm.

This complication occurs in the intermediate and late postoperative period (28 to 188 days, with an average of 88.9 days) (26,27). It is the most common postoperative complication of a craniectomy after the first month of the surgery (27).

Patients with this syndrome often manifest neurological symptoms such as headache, seizures, dizziness, and changes in state of mind (26,27).

The pathophysiology is explained by the exposure of the intracranial content to the atmospheric pressure, which leads to alteration in hydrodynamics of the cerebrospinal fluid of the cerebral perfusion and, finally, to the deformity of the cerebral parenchyma (26,27).

In CT and MRI images the skin flap is depressed in the site of the craniectomy and the adjacent cerebral parenchyma takes shape (26).

It has been reported that dynamic tomography with Xenon and perfusion by tomography are very useful tools for assessing blood flow in these patients (28) (Figure 7).

Its treatment is cranioplasty (25,27), with which there is improvement of the infusion as can be demonstrated by perfusion studies by CT (25).

## Diagnostic algorithm

To make a practical approach when we encounter asymmetries, the following diagnostic algorithm is proposed.

First, one must define what the pathological hemisphere is; second, it is determined whether the hemisphere has increased or decreased in size, and finally, the ventricular size of the abnormal hemisphere is established (Figure 8).

In this way we can define four categories:

- » *Hemisphere with increased volume with small lateral ventricle:* The most frequent etiology in this category is pathologies associated with cerebral edema: trauma, subacute ischemia, intracerebral hemorrhage, primary and metastatic tumors, inflammation or brain infection.
- » *Hemisphere with increased volume with large lateral ventricle:* Hemimegalencephaly is the pathology characteristic of this category.
- » *Hemisphere with decreased volume and small ventricle:* This finding is frequently found in patients with unilateral craniosynostosis and trephine syndrome (sunken flap syndrome).
- » *Hemisphere with decreased volume and large ventricle:* Within this category are associated pathologies to encephalomalacia, trauma and ischemia, among others. Also entities such as Davidoff Dyke Masson syndrome, Sturge-Weber syndrome and Rasmussen's encephalitis.

## Conclusiones

After reviewing the topic it can be concluded:

It is important to know that brain asymmetries are frequent in daily practice.

Brain asymmetries are not always secondary to pathological processes, they may also be a normal finding.

The causes of cerebral asymmetries are multiple, both congenital and acquired.

A Practical algorithm for addressing brain asymmetries can be performed by recognizing the volume of the affected cerebral hemisphere and ventricular size.

## Referencias

1. Shapiro R, Galloway SJ, Shapiro MD. Minimal asymmetry of the brain: A normal variant. *AJR Am J Roentgenol.* 1986;147:753-6.
2. Galaburda AM, LeMay M, Kemper TL, Geschwind N. Right-left asymmetries in the brain. *Science.* 1978;199:852-6.
3. Geschwind N, Levitsky W. Human brain: left-right asymmetries in temporal speech region. *Science.* 1968;161:186-7.
4. Goyal J, Shah V, Rao S, Jindal N. Dyke Davidoff Masson syndrome in children. *J Pediatr Neonatol.* 2009;10(2).
5. Poretis A, Wolfab NI, Boltsgausera E. Differential diagnosis of cerebellar atrophy in childhood. *Eur J Paediatr Neurol.* 2008;12:155-67.
6. Jacoby CG, Go RT, Hahn FJ. Computed tomography in cerebral hemiatrophy. *Am J Roentgenol.* 1977;129:5-9.
7. Siren A, Radyushkin KM, Boretius S, Kammer D. Global brain atrophy after unilateral parietal lesion and its prevention by erythropoietin. *Brain.* 2005;129:480-99.
8. Sharma S, Goyal D, Negi A, Sood RGA, Jhobta A, Surya M. Dike Davidoff Mason Syndrome. *Neuroradiol.* 2006;16:165-6.
9. McMonagle P, Deering F, Berliner Y, Kertesz A. The cognitive profile of posterior cortical atrophy. *Neurology.* 2006;66:331-8.
10. Radzwicki R, Gaillard F. Cerebral hemiatrophy [internet]. 2011 [citado 2016 may. 15]. Disponible en: <http://radiopaedia.org/articles/cerebral-hemiatrophy>
11. Chakravarty A. Crossed cerebral-cerebellar diaschisis: MRI evaluation. *Neurol.* 2002;50:322.
12. Singh JP, Shrimali R, Garg L, Setia V. Radiological quiz-brain. *Indian J Radiol Imaging.* 2002;12:439-40.
13. Atalar MH, Icagasioglu D, Tas F. Cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome) in childhood: Clinicoradiological analysis of 19 cases. *Pediatrics Int.* 2007;49: 70-5.
14. Chridtopher-Rodgman BA, Jay Lombard DO. Dike Davidoff Mason in post-cerebral malaria. *J Child Neurol.* 2011;26(8).
15. Kelley TM, Hatfield LA, Lin DD, Comi AM. Quantitative analysis of cerebral cortical atrophy and correlation with clinical severity in unilateral Sturge-Weber syndrome. *J Child Neurol.* 2005;20:867-70.
16. Omran M, Juibary AG. Sturge-Weber syndrome: Study of eight patients and review of literature. *Int J Neurol.* 2007;7(1).
17. Shen WC, Chen CC, Lee SK, Ho YJ, Lee KR. Magnetic resonance imaging of cerebral hemiatrophy. *J Formos Med Assoc.* 1993;92:995-1000.
18. Zhou J, Li N, Zhou X, Wang J, Ma H, Zhang R. Sturge Weber syndrome. A case report and review of literature. *Chinese Med J.* 2010;123:117-21.
19. Vosskämper M, Schachenmayr W. Cerebral hemiatrophy: a clinicopathological report of two cases with a contribution to pathogenesis and differential diagnosis. *Clin Neuropathol.* 1990;9:244-50.
20. Moritani E. Rasmussen encephalitis. *Neurographics.* 2011;2(1).
21. Dahner W. Radiology review manual (6th ed.). Philadelphia: Wolters Kluwer/Lippincott Williams & Wilkins; 2007.
22. Marisa KB, Glenn AT. Pictorial essay: MRI of cerebral microhaemorrhages. *AJR.* 2007;189:720-5.
23. Moritani E. Rasmussen encephalitis. *Neurographics.* 2011;2(1).
24. Quintana de la Cruz RM, Calvo García M, Fernández Quintero RM, Rozas Rodríguez ML, Jiménez Aragón F, Domínguez Ferreras E. Hemimegalencefalia seram. 2012.
25. Akins PT, Guppy KH. Sinking skin flaps, paradoxical herniation, and external brain tamponade: A review of decompressive craniectomy management. *Neurocrit Care.* 2008;9:269-76.
26. Sinclair A, Scoffings D. Imaging of the postoperative cranium. *Radiographics.* 2010;30:461-82.
27. Yang XF, Wen L, Shen F, Li G, Lou R, Liu WG, et al. Surgical complications secondary to decompressive craniectomy in patients with a head injury: A series of 108 consecutive cases. *Acta Neurochir (Wien).* 2008;150:1241-7.
28. Sakamoto S, Eguchi K, Kiura Y, Arita K, Kurisu K. CT perfusion imaging in the syndrome of the sinking skin flap before and after cranioplasty. *Clin Neurol Neurosurg.* 2006;108:583-5.

## Correspondence

Ricardo Uribe González  
 Universidad CES  
 Diagonal 29d # 9A Sur-150  
 Medellín, Colombia  
[ricardouribegonzalez@gmail.com](mailto:ricardouribegonzalez@gmail.com)

Received for evaluation: May 31, 2016

Accepted for publication: September 17, 2016