

## Bilateral Agenesis of Internal carotid artery: A case report with emphasis on neuroimaging features.

Samah Al Obaidi<sup>1</sup>, Rola Mahmood<sup>2\*</sup>

<sup>1</sup>Consultant radiologist, Department of Radiology, Salmaniya Medical Complex, Bahrain.

<sup>2</sup>Radiology Resident, Department of Radiology, Salmaniya Medical Complex, Bahrain.

### \*Corresponding author

Rola Mahmood, MB, BCh, BAO, Radiology Resident, Department of Radiology, Salmaniya Medical Complex, Bahrain.  
Email: rola.mahmood@gmail.com

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### Abstract

Congenital absence of the internal carotid artery is a rare anomaly, occurring in less than 0.01% of the population [1, 3]. It includes agenesis, aplasia, and hypoplasia of the internal carotid artery [2, 5].

Recognition of these anomalies has important implications during planned surgery, in thromboembolic disease and in the surveillance and detection of associated cerebral aneurysms [2, 3].

It is usually an incidental finding on color Doppler carotid ultra sonography, angiography, computed tomography or magnetic resonance imaging of the head and neck taken for some other indication [2, 6]. Most of the patients are asymptomatic due to the presence of sufficient cerebral circulation supplied by the communicating arteries of the circle of Willis, inter cavernous anastomosis, external carotid artery and by persistent embryologic arteries to the carotid artery territory. Nevertheless, in some cases this anatomic variation may eventually lead to some clinical signs and symptoms [2, 4, 6].

We present a rare case of congenital absence of both internal carotid arteries and its possible implications.

**Keywords:** Internal carotid artery; Agenesis.

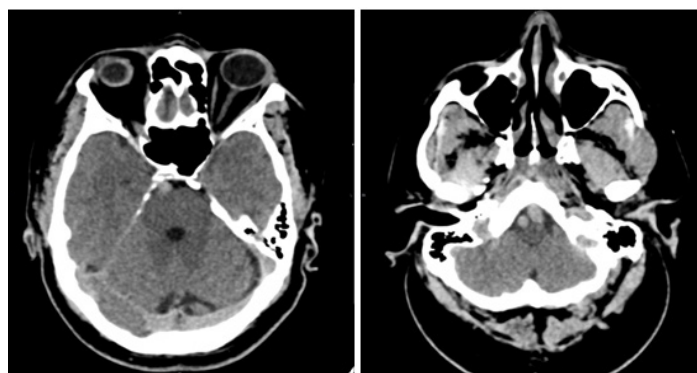
### Case report

A 37-year-old male with no significant past medical history, presented to the ED with a chief complaint of a severe chronic headache of five years duration, not relieved by analgesia. His physical examination was unremarkable.

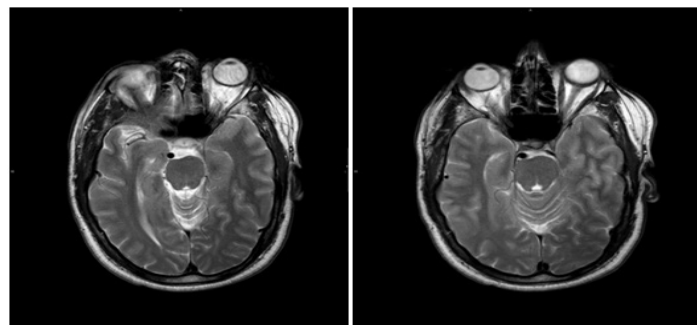
He underwent CT of the brain that showed; dense and prominent posterior circulation of the brain. The cavernous segments of both internal carotid arteries could not be visualized.

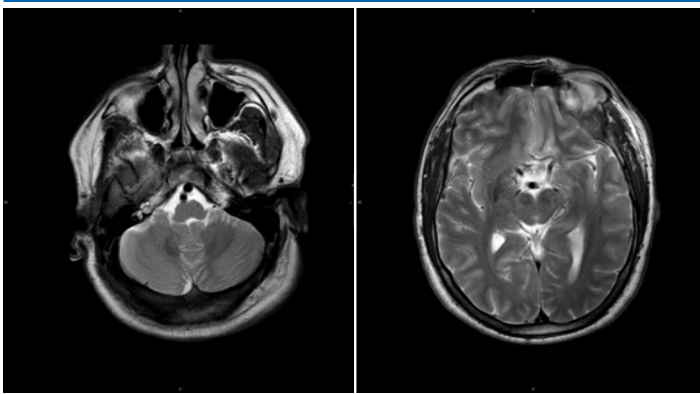
This was followed by a Carotid Doppler ultrasound study that showed narrow common carotid arteries with inability to identify the carotid bifurcation on either side. The vertebral arteries were prominent bilaterally. No collaterals were identified.

The concerning point about the internal carotid arteries was further evaluated by MRA study for the neck vessels which showed atretic internal carotids bilaterally and prominent dilated vertebrobasilar circulation through which the anterior circulation of the brain is supplied. The external carotids were normal.



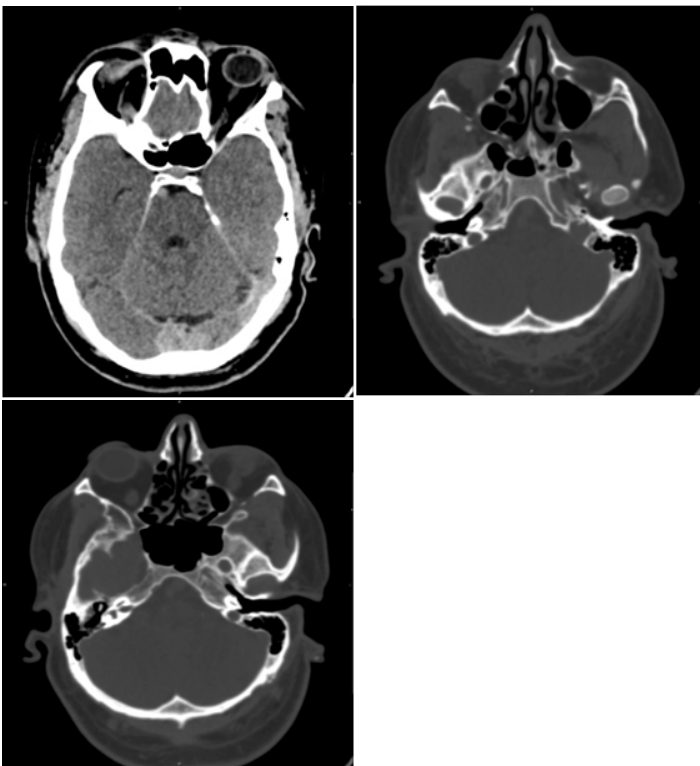
A non-contrast axial CT scan of the brain shows prominent basilar and vertebral arteries with bilaterally absent cavernous segments of the ICA.



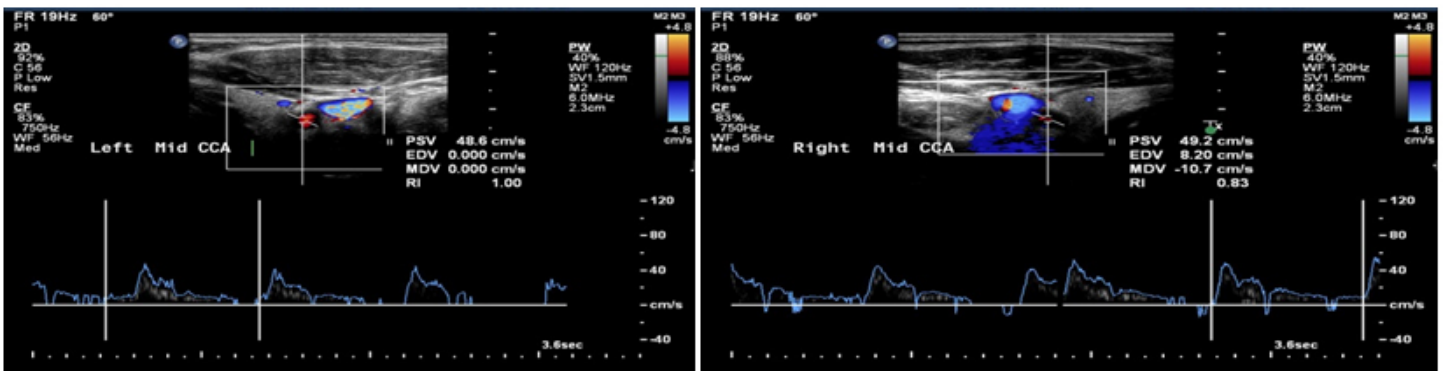


The same changes were noted on These T2 axial images; prominent and patent vertebrobasilar circulation, along with show absent signal voids in the cavernous segments of the ICA bilaterally.

Neck MRA shows absent both ICA with prominent, tortuous and patent vertebrobasilar circulations



A non-contrast axial CT scan of the brain show atretic carotid canals in the skull base bilaterally.



Doppler ultrasound of the carotid system shows narrow and patent common carotid arteries with absent bifurcation up to the level of the mandibular angles.

## Discussion

Absence of internal carotid artery is a rare congenital anomaly, with an incidence of less than 0.01% of the population. It may be unilateral or bilateral, with a predilection for the unilateral variant [1, 2, 5].

Causes of unilateral variant are thought to represent the sequel from a previous insult in utero. Mechanical and hemodynamic stresses placed on the embryo along with comprising effects related to exaggerated folding of the embryo toward one side as well as constriction by amniotic bands are accounted. No explanation for bilateral absence has been rendered in the literature yet [6-8].

Padgett's has proposed that the internal carotid artery originates from the dorsal aorta and the 3rd aortic arch during the early embryonic stage, with the full development of the ICA by 6 weeks [7, 8].

Nevertheless, some controversy regarding the origin of the CCA and ECA is found, some investigators argue that both the proximal ICA and the ECA arise jointly from the 3rd aortic arch and thus agenesis of the ICA is accompanied by absence of the ipsilateral ECA. While others argue that the ECA and CCA can develop normally in the setting of ICA agenesis, as the ECA arises independently from the aortic arch, evidenced by numerous cases of ICA agenesis in the literature with normally developed ECA systems [7, 8].

The majority of the patients remain asymptomatic due to vigorous collateral pathways. However, problems can arise later in life, in particular with progression of an atherosclerotic disease.

Notably, the absence of the carotid artery has also been associated with an increased incidence of cerebral aneurysms [2, 6]. The estimated prevalence of cerebral aneurysms in the general population is 2% - 4%, while it is 24% - 34% in cases of absence of the ICA [1, 3, 5]. This is thought to be related to increased and altered flow dynamics through the collateral vessels [2, 3].

The most common type of collateral flow is through the circle of Willis, through the ACOM and the PCOM, of which the ACOM is reported to be the most frequent site of aneurysm formation in such cases [2, 3].

Less frequently, collateral flow is provided via persistent embryonic vessels or from transcranial collaterals originating from the External carotid artery system [3, 4, 8].

Early recognition of these anomalies maybe of utmost importance particularly in the setting of thromboembolic disease, cerebral aneurysms and during carotid endarterectomy or transsphenoidal hypophyseal surgery [1, 5, 6].

## Conclusion

The diagnosis of internal carotid artery agenesis is usually done on an incidental basis; since most of the patients are asymptomatic. However it can be associated with other potentially serious malformations and disorders [2, 5]. Patients carry a high risk of development of intracranial aneurysms. Therefore, diagnosis of anomalies involving the carotid system should be the signal for rigorous follow-up focused on early identification of potentially fatal changes [2-4].

## References

1. Given II C, Huang-Hellinger k, Bakera M, Chepuri N, Morrisa P (2001) Congenital Absence of the Internal Carotid Artery: Case Reports and Review of the Collateral Circulation, *AJNR* 22: 1953-1959.
2. Shetty A, Radswiki, et al. (2016) Congenital absence of the internal carotid artery.
3. Tasar M, Yetiser S, Tasar A, Ugurel S, Gonul E, (2004) Congenital absence or hypoplasia of the carotid artery: radioclinical issues. *Am J Otolaryngol.* 25: 339-349.
4. Patel SB, Hasmi ZA, Smaroff GC, Cardone JC, Yoon PD (2010) Congenital absence of the left internal carotid artery, *Ann Vasc Surg.* 24: 415.e9-11.
5. Amer S (2015) Rare case of congenital absence of left internal carotid artery, *Ann Indian Acad Neurol.* Jan-Mar. 18: 128-129.
6. Neves W, Kakudate M, Cêntola C, Garzon R, Poçad'Água A, et al. (2008) Agenesis of the internal carotid artery: a case report, *Radiol Bras* 41.
7. Padget DH (1957) The development of the cranial venous system in man, from the viewpoint of comparative anatomy. Philadelphia: Lippincott Williams & Wilkins *Contrib Embryol Carnegie Inst.* 36: 81-140.
8. Oliveira G, SoaresN, Oliveira G, Vale B (2014) Bilateral internal carotid artery agenesis: a case report, *J. vasc. bras.* 13.

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