

# A Rare Case of Bilateral Congenital Absence of Internal Carotid Artery

Narra Ramakrishna<sup>1</sup>, Mukkamala Sonia Pavani<sup>2</sup>, Kopuri Ajay Abishek<sup>3</sup>, Yatham Sai Tejaswini<sup>4</sup>,  
Gadde Prathibha<sup>5</sup>

<sup>1</sup>Professor, Department of Radiology, Katuri Medical college, Guntur, Andhra Pradesh, India

<sup>2, 3, 4</sup>3<sup>rd</sup> year Post Graduate, Department of Radiology, Katuri Medical College, Guntur, Andhra Pradesh, India

<sup>5</sup>2<sup>nd</sup> year Post Graduate, Department of Radiology, Katuri Medical College, Guntur, Andhra Pradesh, India

**Abstract:** Congenital absence of the internal carotid artery (ICA) is a rare anomaly. Even more infrequent are cases where the patient has a bilateral absence of the internal carotid arteries. Reported is a case of a 31-year-old male patient who presented with a few days history of severe headache and dizziness, and was incidentally discovered to have a congenital bilateral absence of his internal carotid arteries. The absence of the arteries is not always recognizably symptomatic, with most findings being incidental through imaging studies only. This is because collateral flow allows for sufficient cerebral circulation. However, this condition puts such patients at higher risk for ischemic and hemorrhagic cerebrovascular insults.

**Keywords:** Internal carotid artery, Internal carotid artery agenesis, Magnetic resonance imaging

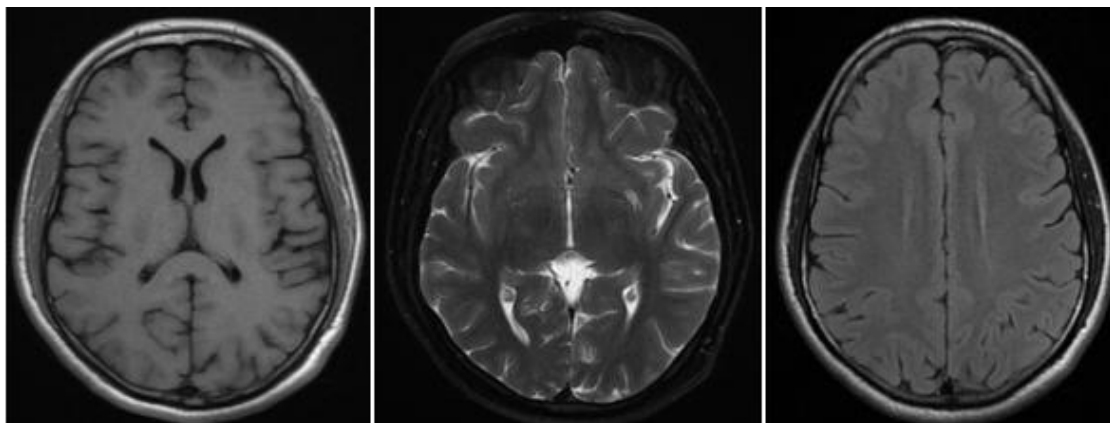
## 1. Introduction

Bilateral congenital absence of the internal carotid arteries (ICA) is a rare congenital defect, with few cases reported in literature. It occurs in less than 0.01% of the population. Absence of the ICA most often presents unilaterally in patients, thereby making instances of bilateral hypoplasia even more rare entity. It encompasses agenesis (no development), aplasia (no development despite the presence of developmental precursors), and hypoplasia (incomplete development). The result of this absence is compensated by collateral blood flow, most commonly from the Circle of Willis. Less commonly, collateral flow is provided via persistent embryonic vessels or from transcranial collaterals originating from the external carotid artery system. Most cases of ICA agenesis are asymptomatic due to sufficient collateral circulation and it is usually an incidental finding on head and the neck imaging by color doppler ultrasonography, computed tomography (CT), or magnetic resonance imaging (MRI). The presented case is a 31-year-old male patient with an incidental finding of bilateral absence of the ICAs.

## 2. Case Presentation

A 31-year-old male patient presented with a few days history of severe headache and dizziness. There was no history of seizures or vomiting. His physical and neurological examinations were normal, and his medical history was unremarkable.

Magnetic resonance imaging (MRI) of the brain was performed, which revealed no evidence of any intracranial space occupying lesion. No signs of acute or chronic ischemic changes were found in the brain parenchyma. However, absence of expected flow voids along the course of the petrous and cavernous segments of the intracranial internal carotid arteries (ICA) was noted. Subsequently, a cervical and intracranial MR angiogram was performed, which revealed non-visualization of bilateral ICAs, prominent bilateral vertebral arteries, and prominent basilar and posterior communicating arteries (PCOMs). The PCOMs were supplying both middle cerebral arteries and the anterior cerebral arteries (ACA) via the circle of Willis.



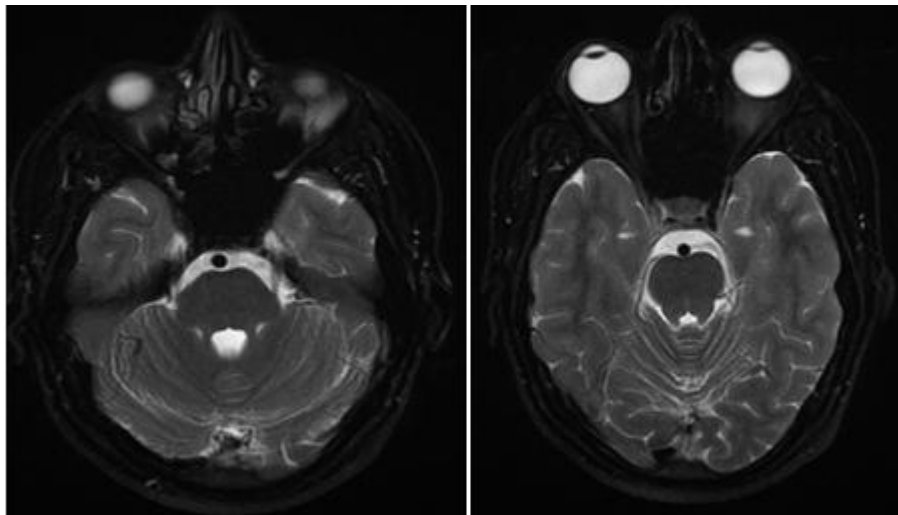
Axial images of the brain, (A) T1 image and (B) T2 image & (C) FLAIR image, which show normal sulcal spaces, cerebrum and basal ganglia.

Note the lack of chronic ischaemic changes.

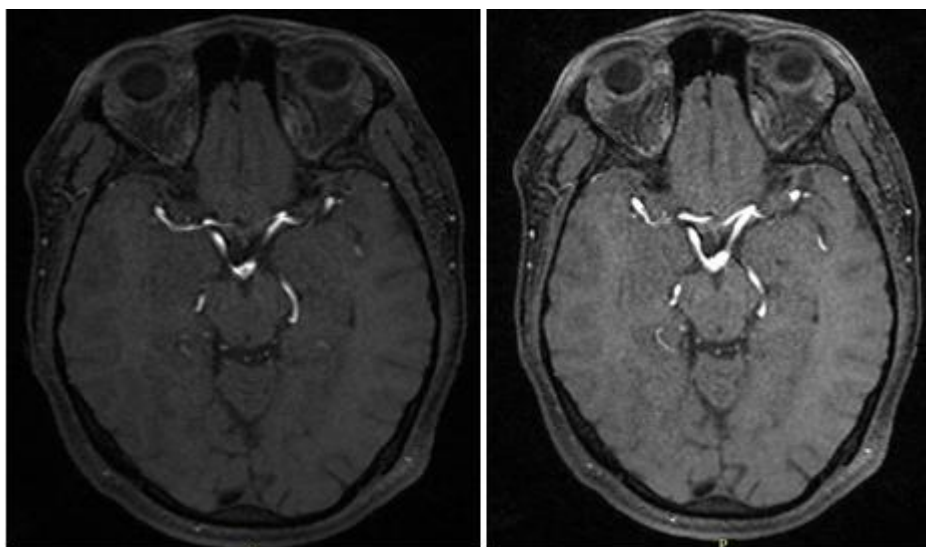
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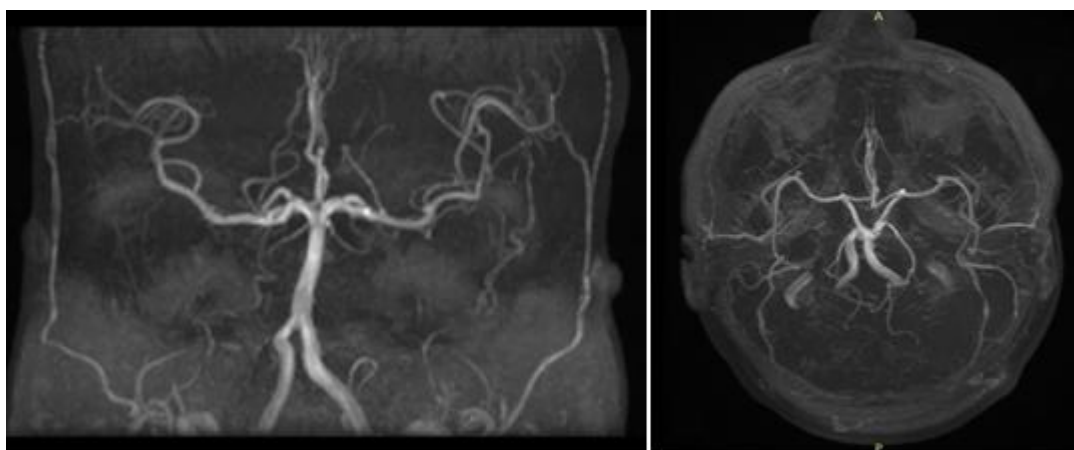
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Axial T2 images of brain depicting non-visualisation of flow void in the expected course of the petrous and cavernous segments of bilateral ICA



Axial 3D time of flight MRI images of the brain showing complete absence of the bilateral internal carotid artery, with the posterior communicating artery supplying the middle cerebral artery on the both sides.



Cervical and intracranial MR angiogram demonstrated a complete absence of the bilateral internal carotid artery, with the posterior communicating artery supplying the middle cerebral artery on the both sides.



### 3. Discussion

Congenital absence of bilateral ICA is very rare congenital anomaly that occurs in less than 0.01% of the population. It includes agenesis (no development), aplasia (no development despite the presence of developmental precursors), and hypoplasia (incomplete development) of the internal carotid arteries. Most cases of ICA agenesis are generally asymptomatic. This occurs because there is a sufficient cerebral circulation supplied by anastomosis in the circle of Willis, intracavernous and external carotid artery anastomosis, besides persistent embryonic arteries. In these cases, the patients are referred for medical assistance because of complications resulting from abnormalities associated with carotid artery agenesis.

Identification and detection of the absence of the ICA may be done via head and neck imaging studies including CT, MRI, carotid Doppler ultrasound or conventional as well as CT/MR angiography.

Lack of the ICA flow void in MRI can be seen in cases of congenital absence of ICA and in cases with a thrombosed ICA; hence the morphology of the carotid canal helps in making the diagnosis. If the agenesis is detected by MRI angiography, it must be confirmed by CT, in an attempt to find hypoplasia or absence of the carotid canal. Generally, the main secondary source of blood supply is the vertebrobasilar system (in cases of bilateral agenesis) or the dominant ICA (in cases of unilateral agenesis or hypoplasia).

Usually, there is enough collateral blood flow to the brain in pediatric patients leading to discovery of this condition in adult population. This is due to inadequate collateral flow

which includes development of atherosclerosis in vessels previously supporting blood flow in the absence of the ICA. Additionally, the development of aneurysms is thought to be slow in children as this issue is not common. There is an increased incidence of aneurysms in these cases compared to general population. Thus, patients with absence of the ICA(s) are at higher risk of aneurysm and should be screened appropriately. In this case report, the patient had not presented with any specific symptoms apart from headache. Other issues are speculated to be associated with abnormalities of the ICA such as cerebrovascular disease.

### 4. Conclusion

The reported bilateral absence of the ICA was discovered incidentally on MRI images of a 31-year-old male patient who initially came with history of headache. Such absences are not common, and oftentimes remain undetected and unexplored due to sufficient collateral blood flow. Origins of such bilateral absence are congenital, with lack of proper channel and vessel development during the fetal period.

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