# Bilateral hypoplasia of the internal carotid artery and ectasia of the internal jugular vein

Eleni Georgiadi, MS,[a,b,c] Georgia Papavasileiou, MD,[b] *and* George S. Sfyroeras, MD,[a] *Athens, Greece*

## ABSTRACT

Hypoplasia of the internal carotid artery and internal jugular vein ectasia are rare congenital abnormalities, whose diagnosis and treatment are not uniformly described. A 32-year-old neurologically asymptomatic woman with renal failure had a carotid artery duplex ultrasound scan as part of an evaluation for renal transplantation and was found to have bilateral internal carotid artery hypoplasia. Computed tomography angiography confirmed congenital bilateral internal carotid artery stenosis and left internal jugular vein ectasia. She had no neurologic deficits. She underwent antiplatelet treatment. (J Vasc Surg Cases Innov Tech 2023;9:101252.)

**Keyword:** Collateral circulation; Computed tomography angiography; Congenital hypoplasia; Internal carotid artery; Internal jugular vein ectasia

Hypoplasia of the internal carotid artery (HICA) is an extremely rare congenital anomaly caused by incomplete development of the organ and usually noted as a lumen narrowing 1 to 2 cm above its bifurcation. Fewer than 70 cases of HICA have been reported in the literature, and only 22 have been bilateral. The actual number is probably higher, because most cases are asymptomatic. This rare anomaly must be recognized owing to the high mortality of its complications.[1] It is often combined with intracranial aneurysms and, therefore, could complicate planned open or endovascular neurointerventional procedures if undiagnosed.[1] Many diagnostic tools are available, such as ultrasound, magnetic resonance angiography (MRA), and computed tomography angiography (CTA) of the skull base. However, digital subtraction angiography (DSA) is considered the gold standard imaging modality, especially when associated vascular conditions such as aneurysms are present.[2,3] We report the case of a young woman with a history of hemodialysis and asymptomatic bilateral ICA stenosis, which was an incidental finding on CTA. Another noteworthy incidental finding on CTA was left internal jugular vein (IJV) ectasia, which, to the best of our knowledge, has not been previously reported in the literature in association with HICA. The patient provided written informed consent for the report of her case details and imaging studies.

## CASE REPORT

A 32-year-old neurologically asymptomatic woman with a history of renal failure presented to our vascular surgery department for evaluation of bilateral hypoplastic ICAs. She underwent duplex carotid ultrasound when evaluated for kidney transplantation. Carotid duplex ultrasound revealed bilateral type II (intermediate echolucent lesions with small areas of echogenicity) and type III (intermediate echogenic lesions with small areas of echolucency; <25%) carotid plaques extending to the intracranial ICAs with stenosis >50%. In both ICAs, decreased flow velocities were detected (peak systolic velocity, ~40 cm/s; Fig 1). Flow velocities in both common and external carotid arteries were described as normal. Her clinical examination was normal without any neurologic deficits.

Although the patient was asymptomatic, she underwent CTA to assess for associated intracranial aneurysms or other pathologies. CTA showed a normal origin of the common carotid and vertebral arteries; however, both ICAs were significantly hypoplastic (>50% stenosis) 1 cm distal of the carotid bifurcation (Fig 2). From that point on, both cervical and intracranial courses of the ICA were hypoplastic, resulting in hypoplastic anterior and middle cerebral arteries. The CTA revealed a hypoplastic osseous bilateral carotid canal (diameter 4 mm on the right side and 3 mm on the left side; Fig 3). Her external carotid arteries, their branches, posterior cerebral arteries, posterior communicating arteries, and vertebrobasilar system were normal. The imaging findings suggested the diagnosis of congenital bilateral HICA.

Another CTA finding was left IJV fusiform ectasia (diameter, 1.69 cm) extending from the clavicle to the hyoid (Fig 4). She had no history of IJV catheterization. The findings with contrast enhancement of the cerebral venous sinuses were normal. Slight asymmetry was present regarding the venous

From the Department of Vascular Surgery, University of Athens Medical School, Attikon University Hospital[a]; the Second Department of Radiology, University of Athens Medical School, Attikon University Hospital[b]; and the Department of Medicine, National and Kapodistrian University of Athens.[c]

Correspondence: George S. Sfyroeras, MD, Department of Vascular Surgery, University of Athens Medical School, Attikon University Hospital, 1 Rimini St, Athens 12462, Greece (e-mail: gsfyr@yahoo.gr).

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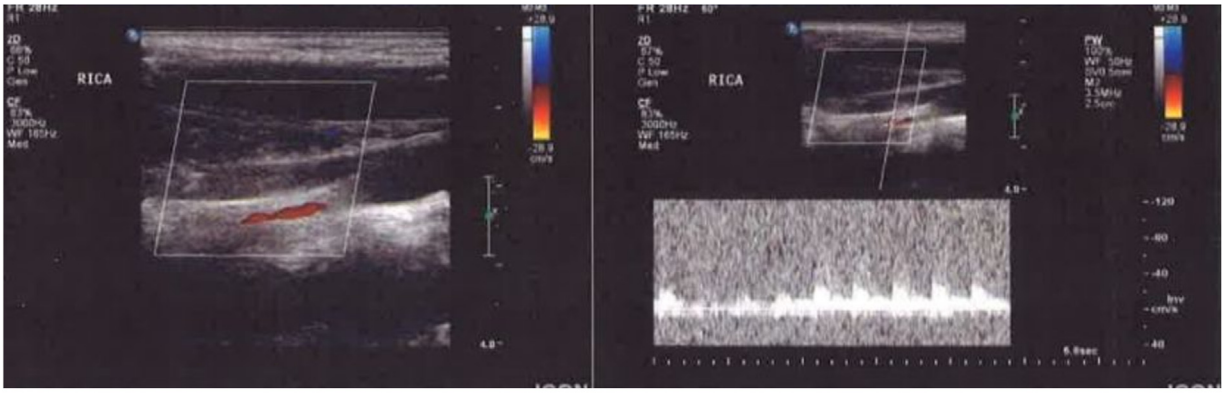


**Fig 1.** Color flow imaging and Doppler spectra of the right internal carotid artery (ICA) demonstrating diffuse luminal narrowing of the left ICA 1 cm distal to the carotid bifurcation (**Left**) with reduced peak systolic flow velocity (~40 cm/s; **Right**).

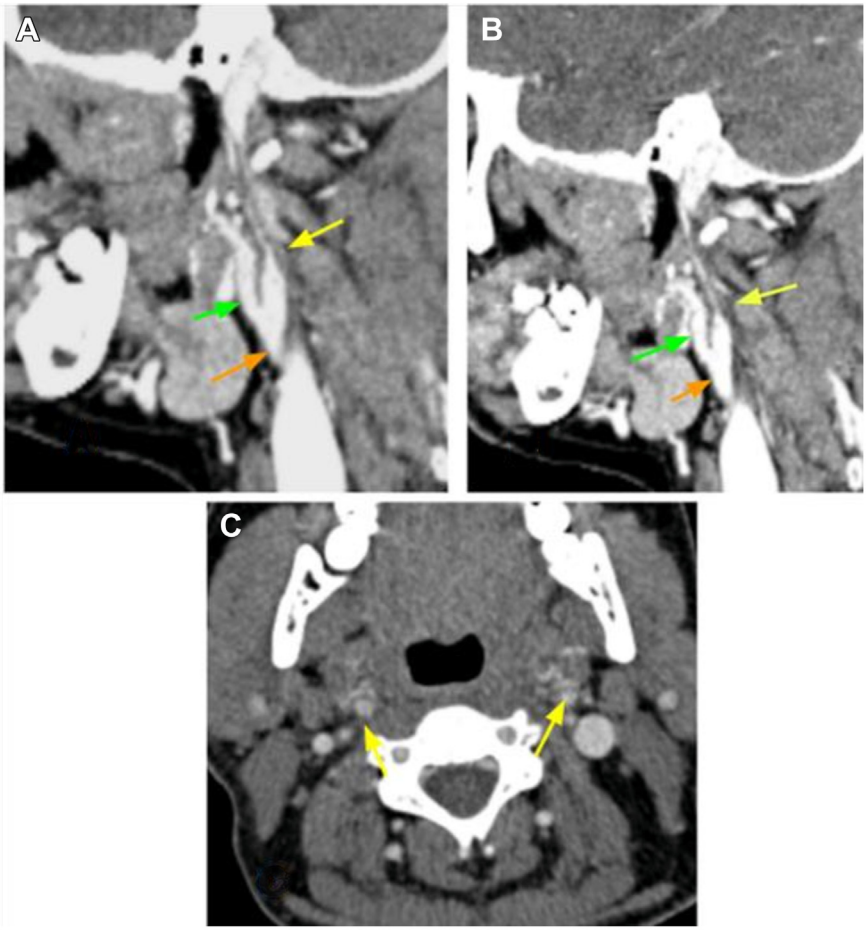


**Fig 2.** Computed tomography angiography (CTA) showing both hypoplastic internal carotid arteries (ICAs). Sagittal CTA showing stenosis in the right ICA (**A**; *yellow arrow*) and left ICA (**B**; *yellow arrow*), which begins above the level of carotid bulb. **C,** Axial CTA showing both hypoplastic ICAs (*yellow arrows*). *Orange arrow* indicates common carotid artery; and *green arrow,* the external carotid artery.

drainage between the two cerebral hemispheres, with the left side predominating. No evidence of bleeding, ischemia, aneurysms, or other vascular intracranial dysplasia was found.

The patient's case was discussed in a meeting with specialized vascular surgeons, interventional radiologists, and neurologists, where it was decided that the most appropriate treatment was antiplatelet therapy.

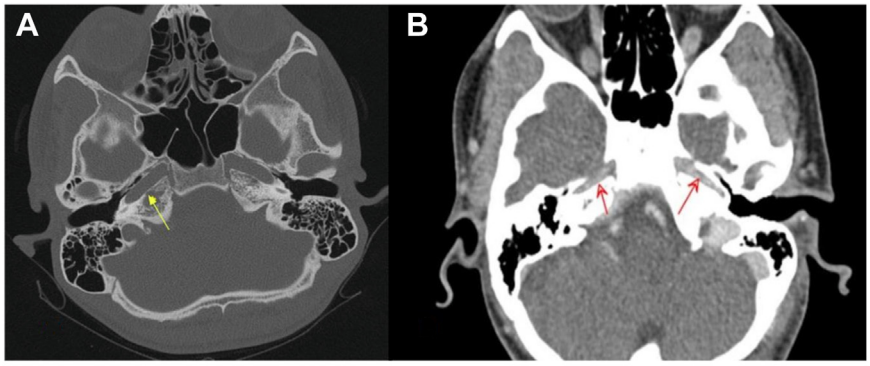


**Fig 3. A,** Axial computed tomography (CT; bone window) of a young adult man showing bilateral normal carotid canals (*yellow arrow* indicates right canal). **B,** Axial computed tomography angiography (CTA) of our patient showing bilateral hypoplastic carotid canals (*red arrows*).

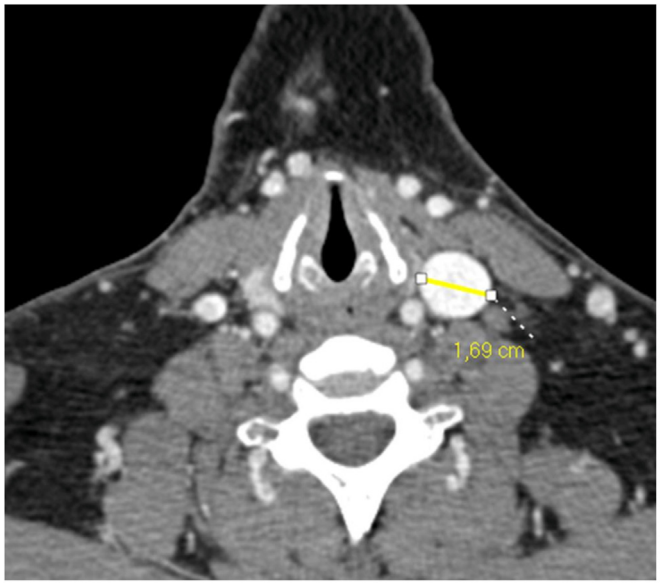


**Fig 4.** Axial computed tomography angiography (CTA) showing left internal jugular vein (IJV) ectasia with a diameter of 1.69 cm.

## LITERATURE REVIEW

We searched for accessible case reports on PubMed using the terms "ICA hypoplasia" and "bilateral." Finally, we included in our review 22 accessible case reports from 1967 to 2021.[3-24] Patients with moyamoya disease were excluded. The inclusion criterion was congenital bilateral HICA in both children and adults.

There were 22 patients (9 women, 9 men, and 4 of undefined gender), with an average age of 41.72 years old. Of the 22 patients, 16 presented with clinical neurologic symptoms, ranging from mild to severe (headache-migraine, 40.9%; loss of consciousness, 27.3%; seizures, 4.5%; vomiting, 9%; dysarthria, 9%; and migraine-like headache with aura, 4.5%). In addition, 4.5% experienced vision loss, hemiplegia, hemianesthesia, ataxia, dizziness, and acute semicomatose state (a state of partial coma, marked by or affected with stupor and disorientation but not a complete coma). Three patients were diagnosed with subarachnoid hemorrhage, two with an acute ischemic attack, and one with intracranial and intraventricular hemorrhage. Six patients had an associated arterial aneurysm, including four cases of a basilar artery aneurysm, two cases of a posterior communicating artery aneurysm, and one case of a posterior cerebral artery aneurysm. The imaging tools were ultrasound (4 patients), CTA (13 patients), MRA (7 patients), and DSA (10 patients).

## DISCUSSION

HICA is an extremely rare congenital anomaly among the structural abnormalities seen in the ICAs, such as agenesis and aplasia, which occur in $<$0.01% of the population.[25] Also, because CTA is not performed in children, HICA is rarely encountered in this age group. In most cases, HICA is discovered incidentally later and is usually asymptomatic; thus, its real prevalence cannot be estimated.[1] It can be either unilateral, which is more common, or bilateral. Some patients with HICA present with neurologic symptoms, including seizures or transient ischemic attack. The most important radiologic finding refers to the carotid canal. In HICA, we usually see a decrease in the diameter of the carotid canal; however, in ICA aplasia, it is completely absent.[1] Narrowing of the bony carotid canal in adulthood does not necessarily mean that the ICA is originally hypoplastic.[26] The plasticity of the bony carotid canal indicates that the canal diameter cannot be used to diagnose HICA.[26] Thus, the bony carotid canal could be narrower even in adulthood, consistent with ICA stenosis. Therefore, HICA cannot be diagnosed by a narrow carotid canal alone.[26] It is believed that HICA is caused by incomplete development of the fetal dorsal aorta, from which normally arises the distal cervical segment of the ICA up to the clinoidal segment. This can also explain that hypoplasia is usually

located 1 to 2 cm above the bifurcation, and the first cervical segment, which develops from the third aortic arch, is normal.[4] The present patient had bilateral HICA found incidentally by CTA. Most cases in the literature were identified by ultrasound, MRA, DSA, CTA, and skull base CT. It is important for physicians to distinguish ICA agenesis from hypoplasia; ICA agenesis can be established from skull base CT when complete absence of the carotid canal is found.[1] According to the literature, the normal diameter of the osseous carotid canal in adults is 5.27 ± 0.62 mm.[27] In HICA cases, it is important to identify the collateral circulation of the brain to predict the neurologic outcome and plan appropriate long-term follow-up. Six types of collateral circulation have been described in HICA patients.[1] In the absence of a unilateral ICA, the anterior communicating artery compensates for the ipsilateral anterior cerebral artery and the enlarged posterior communicating artery for the ipsilateral middle cerebral artery (type A). In type B, the anterior communicating artery supplies the ipsilateral anterior cerebral artery and the middle cerebral artery. In cases of bilateral HICA, the anterior circulation of the ICA blood supply is compensated for by the carotid–vertebrobasilar artery anastomosis of the basilar artery (type C). In cases of unilateral hypoplasia of ICA, blood is supplied to the ipsilateral carotid siphon from the cavernous sinus anastomosis (type D). In type E, the small anterior cerebral artery is supplied by bilateral hypoplasia of the ICA, and the middle cerebral artery is supplied by an expanded posterior communicating artery. Finally, in type F, the distal collateral circulation is provided through the external carotid artery, internal maxillary artery, and skull base anastomosis (ie, the microvascular network of the skull base). Because of the adequate collateral circulation, most HICA cases are asymptomatic. However, many symptoms have also been reported in association with HICA, including headache, transient ischemic attack, seizure, trigeminal neuralgia, oculomotor paralysis, vision loss, symptoms from aneurysm formation, and others.[5] In our patient, the collateral circulation is more compatible with type C. The patient's circulation was well compensated, and she did not have any clinical symptoms. Only a few studies have reported the natural history of HICA and its relationship to ischemic stroke. However, previous studies have suggested a significant correlation between an incomplete circle of Willis and ischemic stroke. In the study by Zhang et al,[28] 25% of the patients developed cerebrovascular events during follow-up, three of whom had no collateral circulation via the circle of Willis and showed cerebral blood flow perfusion defects in the ipsilateral anterior circulation. Thus, the etiology of ischemic events was thought to be due to the insufficient collateral circulation resulting in distal hypoperfusion of the ipsilateral anterior circulation.[28]

Another noteworthy incidental finding in our patient was the unilateral IJV ectasia. Normally, the IJV is at least double in size compared with the common carotid artery, and the normal venous diameter is ~9.1 to 10.2 mm. Considering that IJV catheterization is a common procedure, anatomic variations of the vein might prevent cannulation, and repeated efforts could lead to life-threatening complications.[29] There are a variety of etiologic hypotheses, such as anomalous reduplication of the IJV, compression of the IJV between the head of the clavicle and the cupula of the right lung, superior mediastinal irradiation, trauma, genetic disorders of connective tissue, and so forth.[30] To the best of our knowledge, a combination of HICA and IJV ectasia in the same patient has not been previously reported. Cases have been reported of an acquired carotid artery–IJV fistula, usually due to trauma causing IVJ ectasia[31]; however, our patient had neither a history of trauma nor CTA-related findings. Also, because the ectasia of IVJ was unilateral, in contrast to the bilateral HICA, we concluded that they are both possibly true unrelated congenital anomalies in one patient.

The major causes of mortality for patients with HICA are the associated intracranial vascular events. The most common event is aneurysm formation in the posterior circulation, where hemodynamic stress increases through the collateral circulation. Those with HICA have a 25% to 34% risk of developing intracranial aneurysms compared with a risk of 2% to 4% in the general population.[6] However, aneurysms in the anterior circulation have also been reported. Also, the risk of aneurysm rupture in patients with HICA is higher.[6] Another vascular complication of HICA is acute ischemic attack,[7] especially in patients with an incomplete circle of Willis and inadequate collateral circulation. Also, cerebral artery thrombosis has been reported.[1] Management of these patients with acceptable operative risks is still a challenge and includes aneurysm clipping or endovascular embolism, antiplatelet agents, and long-term follow-up with ultrasound, CTA, MRA, or DSA. Regarding treatment of carotid hypoplasia, antiplatelet therapy was chosen for most patients in our literature review who were asymptomatic and without vascular complications.

## CONCLUSIONS

Bilateral HICA is an extremely rare clinical entity with an increased risk of associated intracranial aneurysms. When suspected, a detailed evaluation of both extracranial and intracranial vessels must be performed. If an intracranial lesion is diagnosed, proper treatment must be applied.

## DISCLOSURES

None.

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