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# Anterior circulation acute ischemic stroke due to vertebral artery ostial stenosis in a patient with congenital internal carotid artery agenesis: a case report

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## Abstract

**Background** Congenital agenesis of internal carotid artery (ICA) is a rare cerebrovascular variation entity. Most cases of congenital ICA agenesis are asymptomatic and discovered incidentally. Congenital ICA agenesis presenting as ischemic stroke is even rare.

**Case presentation** An 80-year-old male patient was admitted to our hospital due to sudden dysarthria and left limb weakness for 3.5 h. Based on emergency physical examination and head computed tomography (CT) scan results, acute ischemic stroke (AIS) of right cerebral hemisphere was suspected. Following intravenous thrombolysis with recombinant tissue plasminogen activator, right congenital agenesis of ICA was confirmed by CT and digital subtraction angiography. Additionally, there was a severe right vertebral artery ostial (VAO) stenosis. After ruling out common causes of AIS such as haematological diseases, arterial dissection, organic heart disease, immunological abnormality and underlying possible malignancies, we hypothesize that the severe stenosis of the right VAO may have contributed to the development of AIS in this case.

**Conclusions** We present a case of right congenital ICA agenesis in which severe stenosis of the right VAO may have played a role in the development of AIS. This case underscores a rare scenario where a lesion in the posterior circulation leads to an infarction in the anterior circulation in the setting of congenital ICA agenesis.

**Keywords** Congenital agenesis of internal carotid artery, Acute ischemic stroke, Vertebral artery ostial stenosis

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## Background

Congenital agenesis of internal carotid artery (ICA), first reported by Tode in 1787 at autopsy [1], is a rare cerebrovascular variation entity with an incidence of less than 0.01% [2]. Although most cases of congenital ICA agenesis are asymptomatic due to well-developed collateral circulations, they can manifest in various ways and may be incidentally discovered. In a large retrospective study, Zhang et al. analyzed the clinical presentations of 64 patients with congenital ICA agenesis. They found developmental delay and subarachnoid hemorrhage like symptoms were most common in patients under 20 years old, whereas transient ischemic attack was the predominant symptom in patients over 40 years old [3]. Congenital ICA agenesis presenting as ischemic stroke is even rare. Herein, we report a case of right congenital ICA agenesis in which severe stenosis of the right vertebral artery ostial (VAO) may have played a role in the development of acute ischemic stroke (AIS). This case highlights a rare scenario where a lesion in the posterior circulation results in an infarction in the anterior circulation under the condition of congenital ICA agenesis. This case is reported according to CARE guidelines.

## Case report

An 80-year-old male patient was admitted to our hospital due to sudden dysarthria and left limb weakness of 3.5 h duration. Upon admission, the patient was afebrile with normal heart rate and normal respiration. Although his blood pressure was recorded at 170/80 mmHg, a family member denied that he has any chronic conditions, including hypertension, coronary heart disease, or diabetes. Emergency physical examination revealed the sleepy patient with slurred specch and right gaze preference. Additionally, he has left sided central facial palsy, accompanied by dense hemiparesis and hemianesthesia on same side. According to the National Institute of Health stroke scale [4], his NIHSS score was 15. The electrocardiogram was normal. Right cerebral hemisphere AIS was considered after head computed tomography (CT) scan (Fig. 1A).

After ruling out any contraindications for thrombolysis, intravenous thrombolysis with recombinant tissue plasminogen activator (r-tPA) at a dosage of 0.9 mg/kg was immediately given as per protocol. The onset-to-needle time (ONT) was 4 h 26 min. Following intravenous thrombolysis, his NIHSS score decreased to 12. A head CT revealed the absence of the right carotid canal (Fig. 1B). Digital subtraction angiography (DSA) showed the following findings: the right ICA was absent, as was the A1 segment of the right anterior cerebral artery (ACA); no remnants were observed in either the proximal or distal segments of the right ICA; the right middle cerebral artery (MCA) was supplied by the enlarged posterior communicating artery (PCoA) and the right ACA was supplied by the anterior communicating artery (ACoA); there was a severe right VAO stenosis (about 80%) and a mild left VAO stenosis (about 45%); and the blood flow in the right MCA and its branches was insufficient compared to the vessels on the left side (Fig. 1C-F). CT and DSA results confirmed the diagnosis of congenital agenesis of the right ICA. Generally, mild stenosis is less likely to result in thrombosis or other adverse events that could lead to acute cerebral infarction. We presume that the right VAO stenosis may have contributed to the development of AIS in this case. To prevent stroke attack again, stenting of the right VA was done. After stenting, the right VAO stenosis was resolved, leading to a significant improvement in the blood flow of the right MCA (Fig. 1G). Tirofiban was administered intravenously at a loading dose of 0.4 μg/kg/min for 30 min, followed by a maintenance dose of 0.1 μg /kg/min for 24 h. The next day, oral administration of amlodipine besylate (5 mg), aspirin (100 mg), clopidogrel (75 mg), and atorvastatin (40 mg) was given to the patient after a head CT scan showed no signs of bleeding. Subsequent brain MRI showed abnormal signals in the right frontal lobe, temporal lobe, insular lobe, and basal ganglia, indicating acute cerebral infarction in the right hemisphere (Fig. 2A-B). A CT angiography (CTA) revealed the absence of the right ICA, along with the absence of the A1 segment of the right ACA (Fig. 2C). Additionally, carotid color ultrasonography showed a smaller right common carotid artery (CCA) (Table 1). The cardiac color ultrasound results were normal. Twelve days after admission, the patient recovered well and discharged from our hospital with a NIHSS score of 3. At the 90-day follow-up, patient had good motor recovery without any noticeable focal deficit and was independent for daily activities with a modified Rankin Scale (mRS) score of 0 [5].

## Discussion

ICA is one of the most stable arteries in human body. Although the exact pathogenesis of congenital ICA agenesis has not been fully elucidated, some scholars believe that this disease is related to the arrested development of ICA during the embryonic stage. Embryologically, the primitive ICA originates from the dorsal aorta and the third aortic arch at around the 3 millimeters (mm) embryonic stage, with complete development of ICA by six weeks. Any disruption in the embryonic development of ICA can lead to developmental anomalies of ICA [6].

Key findings necessary for diagnosing congenital ICA agenesis include the absence of the ICA, the lack of an ipsilateral carotid canal at the skull base, and hypoplasia of the ipsilateral CCA [7]. Traditionally, DSA has been regarded as the gold standard for diagnosing vascular pathologies, including congenital ICA agenesis.

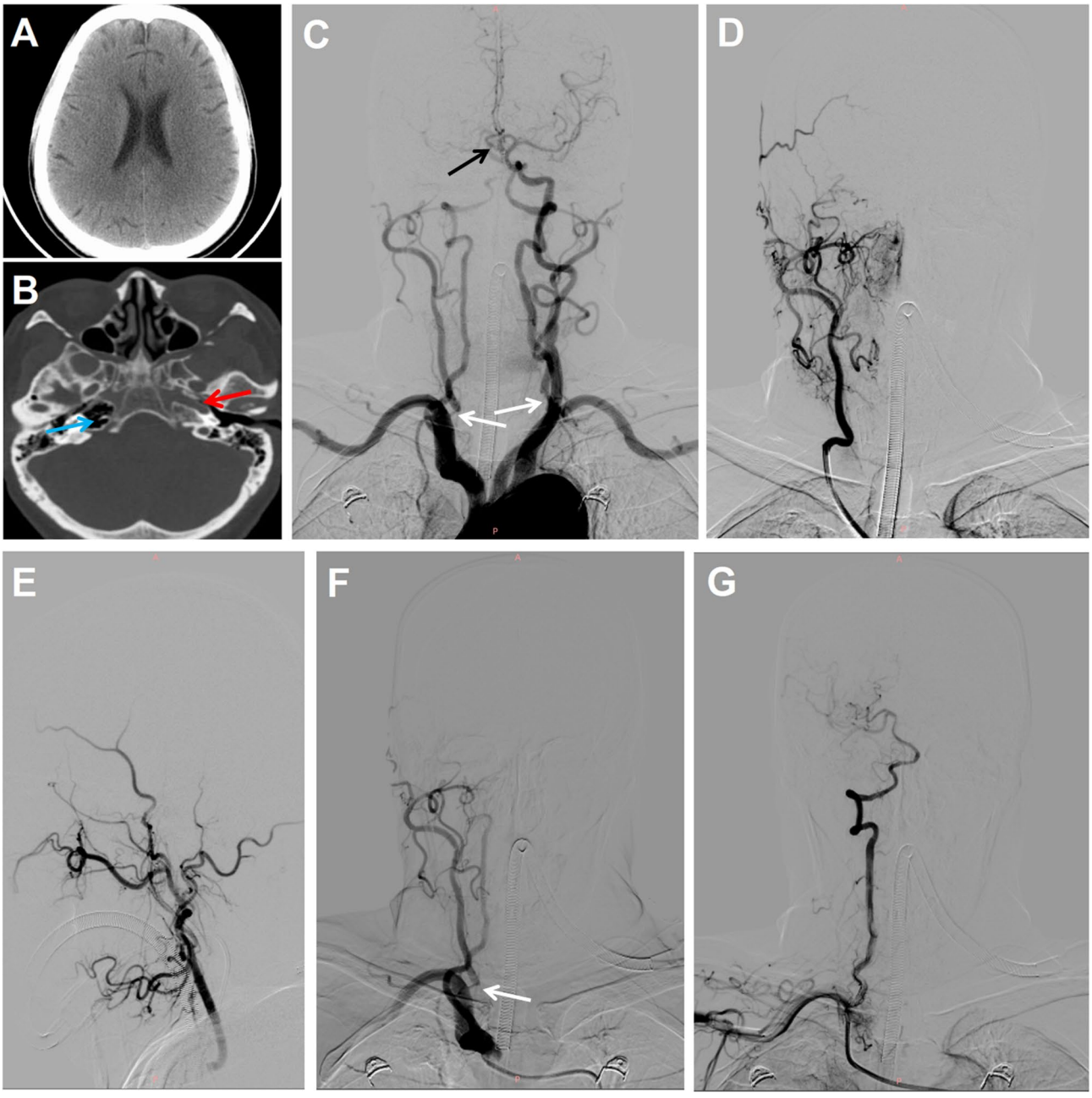


**Fig. 1** (**A**) On admission head CT was almost normal; (**B**) CT scan of the skull base shows the absence of the right carotid canal (blue arrow), while the contralateral was normal (red arrow); (**C**) Aortic arch angiography reveals the following findings: the right ICA was absent, as was the A1 segment of the right ACA; no remnants were observed in either the proximal or distal segments of the right ICA; the right MCA was supplied by the enlarged PCoA and the right ACA was supplied by the ACoA (black arrow points to the general area of the target) ; there was a severe right VAO stenosis and a mild left VAO stenosis (white arrows); blood flow in the right MCA and its branches was insufficient compared to the vessels on the left side; (**D**, front position) and (**E**, lateral position) Right CCA angiography reveals no dysplastic ICA; (**F**) Right brachiocephalic artery angiography shows a right VAO stenosis (white arrows); (**G**) Angiography after vertebral artery stent implantation demonstrates a significant improvement in the blood flow of the right MCA

However, distinguishing ICA agenesis from acquired ICA occlusion can be challenging, as the non-visualization of the ICA in DSA images is a common feature of both conditions. While magnetic resonance angiography (MRA) and CTA provide lower resolution compared to DSA, they are non-invasive imaging modalities that are well-suited for the rapid screening of congenital ICA agenesis. Additionally, Doppler ultrasonography (DUS) serves as another non-invasive and efficient screening tool for congenital ICA agenesis, allowing for dynamic observation of the courses, lumen, and wall structures of the carotid arteries. DUS provides valuable information

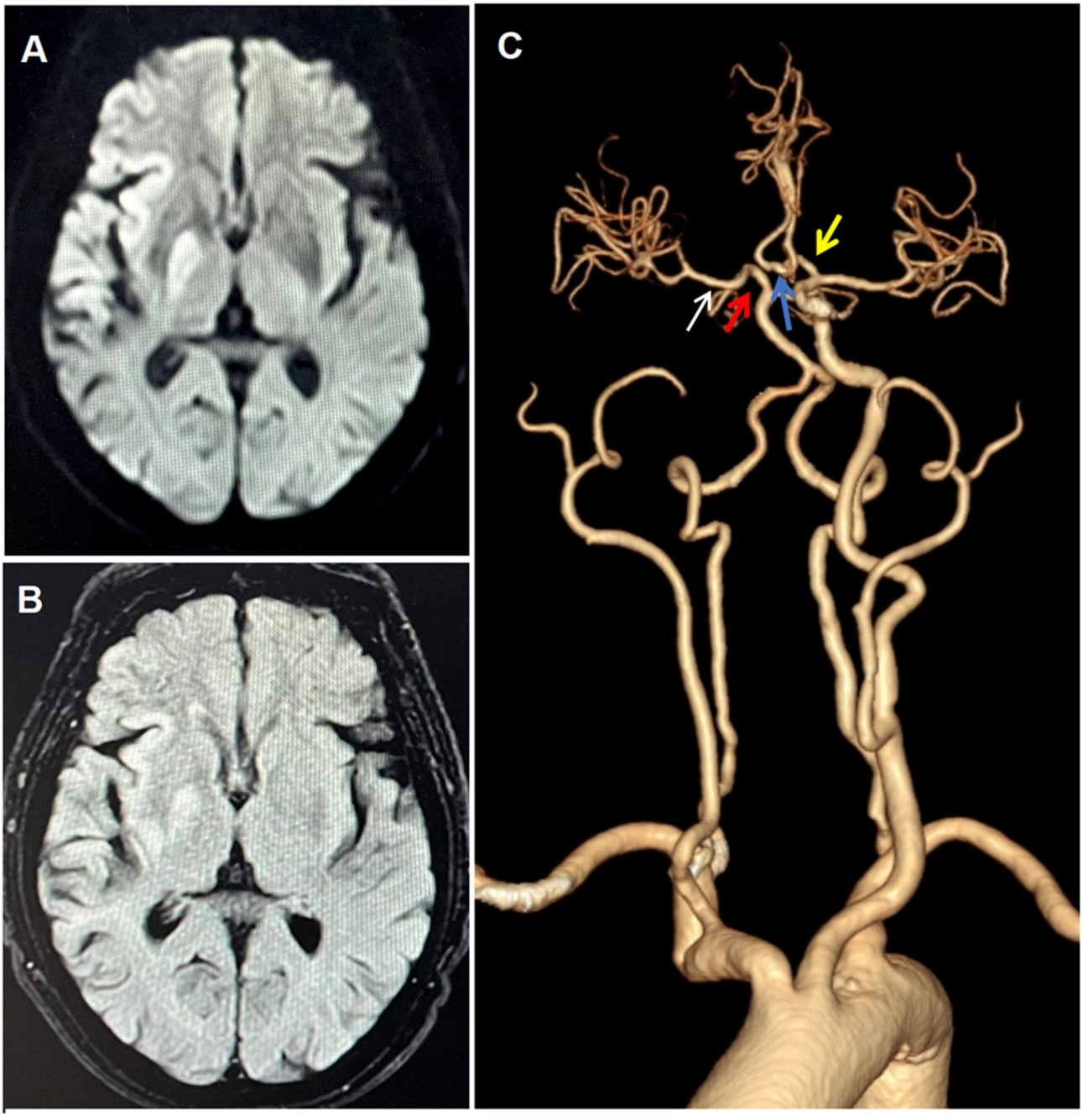


**Fig. 2** Brain MRI (**A**, DWI sequence) and (**B**, Flair sequence) shows diffusion restriction on DWI image with hyperintensity signal on FLAIR image in the right frontal lobe, temporal lobe, insular lobe, and basal ganglia, indicating acute cerebral infarction in the right hemisphere; (**C**) CTA reveals the absence of right ICA and A1 segment of right ACA, the right MCA (white arrow) was supplied by the enlarged PCoA (red arrow) and the right ACA was supplied by the ACoA (blue arrow); yellow arrow indicats the A1 segment of left ACA. DWI, Diffusion-weighted imaging; Flair, Fluid-attenuated inversion recovery

**Table 1** The diameter of different blood vessels determined by the cardiac color ultrasound

| Blood vessels | Left side (mm) | Right side (mm) |
|---|---|---|
| CCA | 7.5 | 4.6 |
| ICA | 6.3 | - |
| ECA | 4.8 | 4.4 |

regarding blood flow velocity and facilitates the assessment of hemodynamic changes, thereby aiding in the diagnosis of congenital ICA agenesis. CT of the skull base offers clear visualization of the bone structure, allowing for the determination of the presence of the carotid canal [2]. Clinically, CT is often used in conjunction with DSA, MRA, CTA, or DUS to achieve a comprehensive diagnosis of congenital ICA agenesis.

Lie et al. summarized six basic patterns of congenital dysgenesis of ICA, including agenesis, aplasia, and hypoplasia. Type A refers to unilateral absence of the ICA, where the ACoA compensates for the ipsilateral ACA and the enlarged PCoA supplies the ipsilateral MCA. Type B also involves unilateral absence of the ICA, where ipsilateral ACA and MCA on the side of ICA absence are supplied by a patent ACoA. Type C describes bilateral agenesis of the ICA, where the bilateral ACAs and MCAs are supplied through carotid-vertebrobasilar anastomoses *via* hypertrophied PCoA. Type D involves unilateral absence of the cervical segment of the ICA, with blood supply to the ipsilateral carotid siphon coming from the cavernous sinus anastomosis. Type E is characterized by hypoplasia of bilateral ICA, where the bilateral diminutive ACAs are supplied by the bilateral hypoplastic ICAs, and the bilateral MCAs are supplied through the posterior cerebral artery *via* the PCoAs. Type F shows collateral flow to the distal ICA *via* anastomoses from distal branches of the ECA system [8]. In the present case, the patient exhibits unilateral absence of the ICA, along with the absence of the A1 segment of the right ACA. Notably, the right ACA is supplied by a patent ACoA, while the ipsilateral MCA is supplied by a hypertrophied PCoA, confirming the diagnosis of Type A congenital ICA agenesis as classified by Lie et al. [8].

**Table 2** Features of congenital ICA agenesis and acquired ICA occlusion

| Conditions | Common features |
|---|---|
| Congenital ICA agenesis | 1) absence of the ICA;<br>2) the absence of ipsilateral carotid canal at the skull base;<br>3) the hypoplasia of ipsilateral CCA |
| ICA occlusion due to atherosclerosis | 1) history of ischemic stroke or current ischemic stroke;<br>2) CT or MRI shows old infarcts in the same vascular territory;<br>3) CTA or DSA shows residual ICA stump;<br>4) CTA or DSA shows non-smooth or plaque-like narrowing of the occluded segment near the heart end;<br>5) carotid ultrasound shows atherosclerotic plaques at the site of occlusion; DSA, CTA or high-resolution MRI shows plaques or calcifications at the occlusion site;<br>6) CT bone window shows the presence of bony canals in the carotid artery. |
| ICA occlusion due to arterial dissection | 1) recent history of neck trauma or massage;<br>2) often accompanied by head and neck pain;<br>3) CTA or DSA shows a residual lumen of the ICA;<br>4) CTA or DSA shows a flame-like or rat's tail appearance of the occluded segment, occasionally shows a double-barreled appearance;<br>5) MRI transverse view clearly shows the cross-section of the vessel wall, with T1 sequence showing a crescent-shaped high signal intramural hematoma, and T2 sequence showing high signal intraluminal flaps. |
| ICA occlusion due to cardiac embolism | 1) history of atrial fibrillation or cardiac valve surgery;<br>2) echocardiography shows atrial thrombus, valve vegetations or cardiac myxoma;<br>3) In cases of paradoxical embolism, echocardiography shows congenital atrial or ventricular septal defects and deep vein thrombosis in the lower limbs. |

Congenital ICA agenesis can be easily misdiagnosed as acquired ICA occlusion [9]. Proper diagnosis requires differentiating ICA agenesis from occlusion caused by various factors, such as atherosclerosis, arterial dissection, and cardiac embolism. Common characteristics of these conditions are summarized in Table 2. Understanding these characteristics will help distinguish congenital ICA agenesis from acquired ICA occlusion. In emergency situations involving surgery for acute cerebral infarction, early identification of ICA agenesis and ICA occlusion can reduce unnecessary surgical procedures and treatment risks.

The occurrence of congenital ICA agenesis presenting as ischemic stroke is rare, with only a few documented cases. For instance, Wang et al. reported a patient with left congenital ICA agenesis who suffered a stroke due to acute cardiogenic embolism of the right MCA [10]. Jennifer et al. described a case of right superficial sylvian ischemic stroke associated with right congenital ICA agenesis [11]. Jianu et al. detailed a case of AIS in the vertebrobasilar territory linked to multiple congenital anomalies of the carotid and vertebral arteries [12]. Gupta et al. presented a case of arterial ischemic stroke in a child with ICA hypoplasia and protein S deficiency [13]. Hou et al. reported a case of left ICA agenesis in a middle aged male associated with ischemic stroke [14]. In our case, the patient had right congenital ICA agenesis and experienced a stroke due to MCA occlusion. Although we were unable to determine the exact etiology of AIS (causes such as haematological diseases, arterial dissection, organic heart disease, immunological abnormality and underlying possible malignancies were ruled out, but the possibility of a special cause stroke or cryptogenic stroke couldn't be excluded), we presume that the presence of right VAO stenosis may have played a crucial role in the cerebral ischemic event observed in our case because the right MCA was supplied by the collateral flow from the VA [15]. According to the Trial of Org 10,172 in Acute Stroke Treatment (TOAST), the stroke in our patient is the large artery atherosclerotic (LAA) type, which is responsible for approximately 15% of stroke [16]. In the situation of Type A congenital ICA agenesis, the MCA is supplied by an enlarged PCoA. Therefore, a thrombus generated at the stenotic origin of the right VA might have lodged into the right MCA *via* the basilar artery-PcoA pathway, resulting in MCA occlusion.

## Conclusions

We present a unique case of right congenital ICA agenesis in which stenosis of the right VAO may have played a role in the onset of AIS. This case highlights a rare scenario where a lesion in the posterior circulation results in an infarction in the anterior circulation under the condition of congenital ICA agenesis.

### Abbreviations

| | |
|---|---|
| ICA | Internal carotid artery |
| AIS | Acute ischemic stroke |
| VA | Vertebral artery |
| VAO | Vertebral artery ostial |
| CCA | Common carotid artery |
| ECA | External carotid artery |
| CT | Computed tomography |
| CTP | CT angiography |
| DSA | Digital subtraction angiography |
| MRA | Magnetic resonance angiography |
| DUS | Doppler ultrasonography |
| MCA | Right middle cerebral artery |
| PCoA | Posterior communicating artery |
| ACA | Anterior cerebral artery |
| ACoA | Anterior communicating artery |
| LAA | Large artery atherosclerotic |
| NIHSS | National Institute of Health stroke scale |
| mRS | Modified Rankin Scale |

## Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12883-024-03917-4.

Supplementary Material 1

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### Author contributions

Conceptualization, J.H. and T.Z.L.; methodology, J.H., T.Z.L. and D.K.L.; software, X.K.W.; investigation, J.H., D.K.L., X.K.W., X.C.L. D.Z., B.S. W, C.G. and J.L.; writing—original draft preparation, J.H. and T.Z.L.; writing—review and editing, T.Z.L.; visualization, J.H.; supervision, T.Z.L.; project administration, J.H.; All authors have read and agreed to the published version of the manuscript.

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### Data availability

No datasets were generated or analysed during the current study.

## Declarations

### Ethics approval and consent to participate

Ethical review and approval were waived for this study due to the retrospective study design.

### Consent for publication

Written informed consent has been obtained from the patient to publish this case.

### Competing interests

The authors declare no competing interests.

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