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## Case Report

# Right-sided aorta with complete isolation of the left innominate artery

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## ABSTRACT

A right-sided aorta with an isolated left subclavian and left common carotid artery is a rare arch defect. We are presenting the case of a 56-year-old woman who had initially presented with headache and 3 episodes of right-sided facial and extremity numbness. A presumptive diagnosis of transient ischemic attack prompted imaging studies where a computer tomography angiogram showed aberrant vessels.

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## Background

The great vessels are formed from the embryologic aortic arches. Around week 3 of gestation, 6-paired aortic arches, the so-called branchial arch arteries, develop between the ventral and dorsal aortae. The dorsal aorta gives off several intersegmental arteries [1]. The third aortic arch gives eventual rise to the common carotid arteries and the proximal portion of the internal carotid artery. The right fourth arch forms the proximal right subclavian artery [1]. The left subclavian arises from the seventh intersegmental artery whereas the right subclavian arises from a combination of the fourth aortic arch, the right dorsal aorta and the right seventh intersegmental artery [1].

Normal vascular anatomy of the aortic arch features the right brachiocephalic artery branching off of the ascending aorta, followed by the left common carotid and left subclavian artery branching independently. The brachiocephalic artery gives rise to the subclavian and the right common carotid

artery, the latter dividing into the right internal and external carotid arteries. The vertebral arteries are branches of the subclavian and unite to form the single basilar artery, which supplies almost all of the blood to the posterior and inferior surfaces of the brain. The bilateral internal carotid arteries supply the lateral and anteromedial surfaces of the brain.

Aortic arch anomalies are uncommon, and there exist 5 well-recognized groups of variant arch anatomy. These groups are: double aortic arch, right aortic arch with mirror-image branching, right aortic arch with abnormal branching, a left aortic arch with abnormal branching, and a cervical aortic arch [2]. These typical variants have complications, most commonly tracheobronchial or esophageal compression and disturbances of normal blood flow patterns [2]. Rarely, the disturbances of flow can result in symptoms of cerebral ischemia because of reversal of flow through the vertebral-basilar vasculature [3]. Classically, this is known as subclavian steal when this phenomenon is produced secondary to a proximal subclavian artery occlusion [3].

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The aberrant anatomy presented in this case report, which includes a right-sided aorta with complete isolation of the left innominate artery has never been previously documented, and the neurologic sequelae of the variant anatomic anatomy is not commonly seen in other arch anomalies.

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## Case report

A 56-year-old woman with medical history significant for multiple transient ischemic attacks (TIA) affecting the right side of the body and hypertension, presented to a nearby hospital after experiencing a headache and right-sided facial and extremity numbness. Vital signs were stable upon admission. Blood pressure was found to be 99/62 with no significant difference between extremities. Radial pulses were strong bilaterally, with no delay. Physical examination was within normal limits, which included a detailed neurologic examination. Patient was evaluated for a TIA via computer tomography angiography of the head and the neck at the nearby hospital, which revealed an anatomic vascular variant, and thus, a carotid angiogram was recommended by neurology. At this point, the patient was transferred to our facility. Interventional neuroradiology performed a diagnostic carotid and cerebral angiogram for the work-up of the recurrent TIAs affecting the right side of the body.

### Procedure in detail

A 5-French sheath was placed in the right femoral artery under fluoroscopy and was navigated along the aorta, which had no apparent arch. The ascending aorta did not have the brachiocephalic artery; a separate origin for both the right subclavian and the right common carotid artery was present. The aorta descended down the right side instead of the left. The catheter was navigated to the right subclavian artery followed by the right vertebral artery. A cervical and cerebral angiogram was performed.

### Findings

The right vertebral artery angiogram revealed tortuous but otherwise normal V1 through V4 segments. The vertebral-basilar junction was extremely tortuous with a high degree of dolichoectasia noted in the basilar artery. The major blood vessels to the cerebellum and the left posterior cerebral artery were filled with no abnormality appreciated.

Several anatomic anomalies were found. The patient had a right-sided aorta that descended almost immediately with no apparent arch. The brachiocephalic artery was absent. The right common carotid and the right subclavian arteries originated directly off of the aorta. The blood flow from the right vertebral artery was directed into the left vertebral artery in a retrograde fashion resulting in a “steal phenomenon”. In addition, there was no direct connection between the aorta and the left subclavian and left common carotid artery. The left common carotid artery originated from the left vertebral artery. Interestingly, the blood flow from the left vertebral artery fills the left common carotid artery (which is diminutive in nature), with the majority entering the left external carotid

artery. The left internal carotid artery shows a normal but a smaller cervical segment. There is also flow seen entering the petrous cavernous and supraclinoid internal carotid artery segments with minimal flow traversing the left middle cerebral artery. Essentially, the right vertebral artery was responsible for blood flow in the basilar, left vertebral, left common carotid, and left subclavian arteries (See Fig. 1).

A right carotid cerebroangiogram revealed the same vascular anomalies described above. Significant cross flow in the left hemisphere with brisk filling of both the left anterior cerebral arteries and the left middle cerebral arteries and all of their branches was also observed.

The vessels had no specific abnormal findings with an exception at the vertebral-basilar junction as previously described above. No evidence of atherosclerotic disease was identified on the imaging study.

Given the anatomic variations and the abnormal blood flow pattern, the patient likely suffers from an extended subclavian steal-like phenomenon. Activity involving the left arm required additional perfusion, which decreased flow through the circle of Willis. The resulting ischemia would present with right-sided neurologic manifestations. Intervention is currently being contemplated at this time.

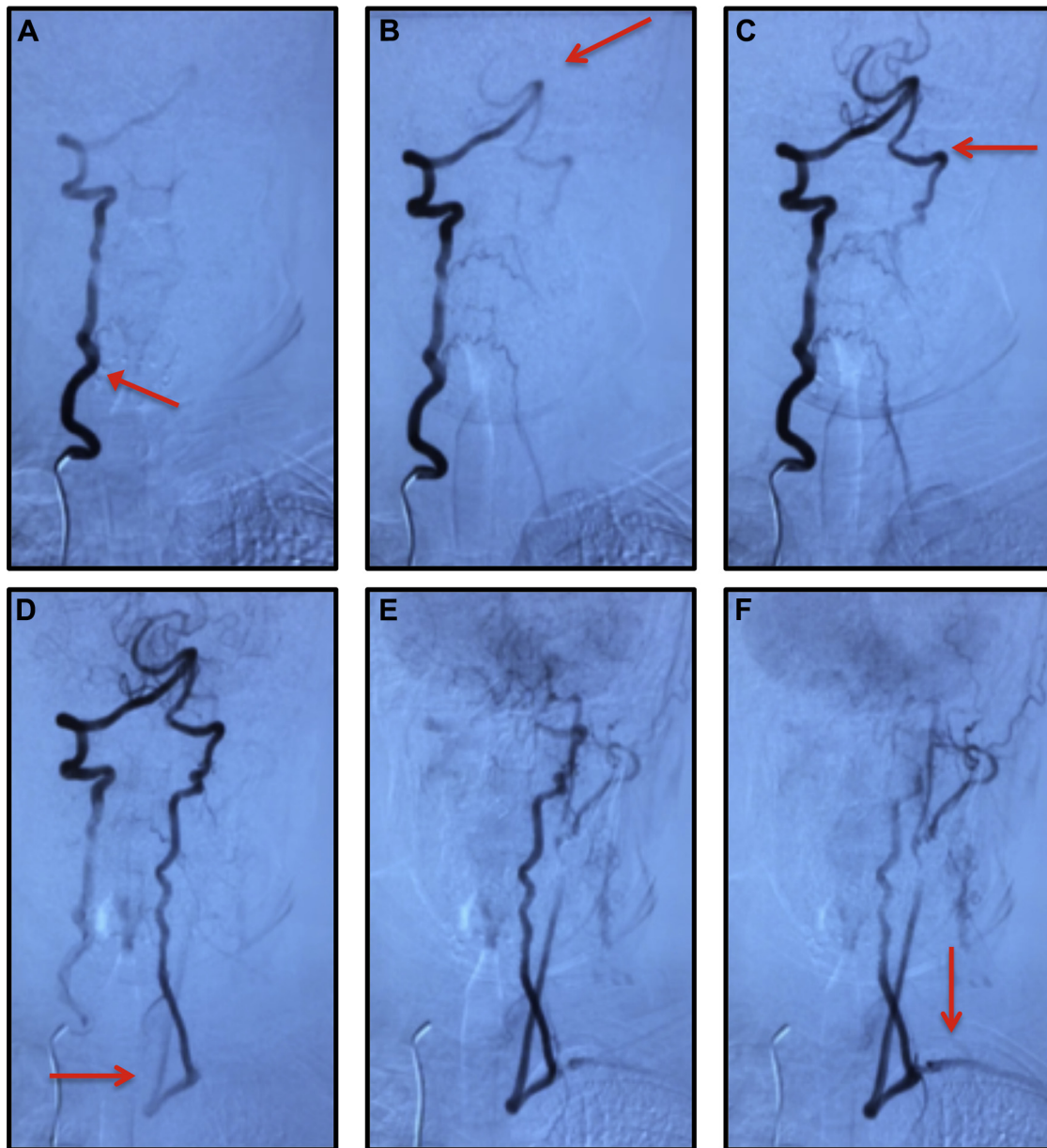
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## Discussion

Our patient presented isolated left subclavian and left common carotid arteries. Therefore, a typical “subclavian steal” [1] scenario is not present, but an extension of the well-reported phenomenon is most likely. The patient also had a right-sided aorta, with no evidence of an arch. In addition, the left common carotid artery originates from the left vertebral artery. The absence of an identifiable arch, adds to the unique radiographic presentation, almost separating itself entirely from similar published vascular anomalies.

A right-sided aortic arch is a rare variant with an incidence theorized to be between 0.05% and 0.1% of the population [4]. Right-sided aortic arches are largely associated with congenital heart defects, the most common of which is the tetralogy of Fallot. There are 3 types of right-sided aortic arch abnormalities: a right aortic arch with mirror-image branching, the aberrant left subclavian artery and with an isolated left subclavian artery [5]. Of these 3 types, the isolated left subclavian artery is the rarest, and is associated with 0.8% of right aortic arches [5].

Normal anatomy has approximately equal blood flow through both the left and right vasculature. With the isolated left subclavian artery, there is no direct connection between the subclavian and the aortic arch; the blood supply to the left upper extremity is thus supplied by a combination of retrograde blood flow patterns from the right-sided vasculature and potential collateral circulation from the aorta. In these individuals, there is a high level of dependency on adequate blood flow through the right vertebral artery, to supply the basilar artery and its respective branches. Given that there is no blood flow coming from the left vertebral artery, blood flows directly from the right vertebral artery into the basilar artery as well as retrogradely through the left vertebral artery and eventually to the left subclavian artery. Increased blood



**Fig. 1 – (A) Introduction of contrast and blood flow in the right vertebral artery (RVA, red arrow). (B) Continued flow through the RVA into the vertebral-basilar junction (red arrow). (C) Flow through the basilar artery (note the tortuous appearance); and the beginning of retrograde flow into the left vertebral artery (LVA, red arrow). (D) Continued retrograde flow through LVA with initial left common carotid artery (red arrow) flow. (E) Flow through the left common carotid artery, with bifurcation into the left internal and external carotid arteries. (F) Flow through the left external and internal carotid arteries. Note the delayed perfusion through the left subclavian artery (red arrow).**

supply demand to the left arm, directs more blood flow through this retrograde pathway, and thus “steals” blood, and precipitates cerebral ischemia.

In most patients with the right-sided arch and isolated subclavian artery, the left common carotid still originates in the aorta, and thus, the branches of the internal and external carotid arteries are adequately supplied. Thus, only the posterior segment of the circle of Willis is normally dependent on the blood flow from the right vertebral artery. However, our patient differs from prior literature on the subject, because instead of having an isolated single left subclavian artery, she

also has an isolated left common carotid artery. There is an increased demand on the system at baseline, and any additional steal results in the signs and symptoms of inadequate cerebral perfusion.

In addition, our patient lacked the normal brachiocephalic artery and instead had the right common carotid and subclavian arising as direct branches from the aorta. To our knowledge, this is the first case reported to have a right-sided aorta lacking a brachiocephalic artery with an isolated left innominate artery without origin to the pulmonary arteries or to a patent ductus arteriosus [6,7].

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## Conclusions

Because of its low incidence, the right-sided aorta with an isolated left innominate artery is rarely considered in a differential diagnoses and will most likely be discovered incidentally through diagnostic imaging. However, it is an extremely rare anatomic defect with important neurologic sequela.

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