Transcardiac cerebral angiography in a child

Technical note

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The authors present a case in which transfemoral venous, transcardiac cerebral angiography was performed. In this 5-year-old girl with PHACE syndrome, both transfemoral and transbrachial arterial routes could not be used due to aortic interruption and aneurysmal dilation and small looping of the proximal portion of bilateral subclavian arteries. A 5-F balloon-tipped double-lumen catheter was advanced to the right atrium of the heart from the femoral vein. The catheter was then advanced to the left atrium through the patent foramen ovale and was further advanced to the left ventricle and then to the ascending aorta. The balloon catheter was exchanged for a 4-F catheter. Bilateral common carotid angiography was performed without difficulty. This transcardiac approach is useful in the unusual situation in which both femoral and brachial arterial routes are not available.
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Key Words • cerebral angiography • foramen ovale • transcardiac approach • surgical technique

In general, catheter cerebral angiography is performed through the femoral, brachial, or radial artery. In the pediatric population, the transfemoral arterial route is commonly used except in the neonatal period when the umbilical route is theoretically an alternative.[1,7] The transcardiac route is also available in the neonatal period when the foramen ovale is patent.[7] We present a unique case in which transcardiac catheter cerebral angiography was performed via the femoral venous route in a 5-year-old girl with PHACE syndrome when it was not possible to use either the transfemoral or transbrachial arterial route. PHACE is a coined acronym consisting of the initial letters of the following symptoms: posterior fossa cyst, hemangioma of the face, anomaly of the cerebral arteries, coarctation of the aorta, and eye anomaly.[4,8] In this syndrome, anomaly of the cerebral arteries, such as stenosis or occlusion and agenesis or hypogenesis of the cerebral arteries, and persistent embryological arteries are often observed.

Abbreviations used in this paper: CTA = CT angiography; ICA = internal carotid artery; MRA = MR angiography.

Case Report

History and Presentation. This 5-year-old girl was delivered by cesarian section after 34 weeks 5 days of gestation. Her birth weight was 2736 g. She had a flat left facial hemangioma at birth. Ultrasonography, CTA, and left retrograde radial angiography showed interruption of the aorta and an abnormal collateral vessel branching off the aortic arch and extending to the descending aorta. An MRI study showed a posterior fossa (Dandy-Walker) cyst. She was diagnosed with PHACE syndrome. The facial hemangioma increased in size rapidly after Day 7 and began to compress the patient's airway. Steroid treatment was started and was effective. A cyst-peritoneal shunt was placed on Day 28 for treatment of the Dandy-Walker cyst. The interruption of the aorta and the abnormal collateral vessel were conservatively managed with serial follow-up imaging. A follow-up CTA performed when the patient was 3 years old revealed an aortic aneurysm. Except for shunt revision at the age of 9 months, the patient's clinical course was uneventful.

At the age of 5 years, CTA showed enlargement of an

aortic aneurysm that was initially observed when the patient was 3 years old and marked dilation of the abnormal collateral vessel as well as stenosis of the left cervical ICA (Fig. 1). A CTA study of the brain also showed complex cerebral arterial anomalies, such as persistent trigeminal artery on the left and segmental agenesis of the basilar artery distal to the trigeminal artery (Fig. 2). The left vertebral artery—both extracranial and intracranial segments—was absent. The level of the left carotid bifurcation was extremely low, near the site where the abnormally dilated collateral vessel branched off the aortic arch.

Indications for Catheter Angiography. Catheter angiography and pressure monitoring were required for surgical repair of the interruption and aneurysm of the aortic arch as well as possible treatment of carotid stenosis. The indications for transcardiac cerebral angiography were as follows: 1) presence of the interruption of the aorta and aortic aneurysm as well as the abnormally dilated collateral vessel, which precluded a transfemoral arterial approach as well as a left transbrachial arterial approach; 2) a small looping of the proximal subclavian arteries bilaterally (Fig. 1 right), which precluded a brachial arterial approach; 3) the low position of the left carotid bifurcation near the abnormal collateral vessel branching off the aortic arch, which precluded the direct puncture of the left common carotid artery; 4) failure of CTA to precisely show stenosis of the left cervical ICA and to show the hemodynamics of the anomalous cerebral arteries.

An echocardiogram showed no cardiac anomalies (including interatrial septal defect). Although this examination did not reveal the patent foramen ovale before the angiographic procedure, the transcardiac approach was selected because perforation of the septum with a transseptal needle was considered possible (detailed in *Discussion*) in the event of failure to cross the septum. Thus, whether the foramen ovale was patent or not, a transcardiac approach was considered possible.

The Transcardiac Procedure. Under general anesthesia, 5-F short sheaths (6 cm in length) were inserted into the right femoral artery and vein. Pressure monitoring and cardiac/aortic catheter angiography were performed by the pediatric cardiologist. The angiographic procedures for this cardiac and aortic evaluation are not described in this paper; only the procedures required for the transcardiac cerebral angiography are described. The femoral arterial route was used for pressure monitoring and aortography. The tip of a 5-F pigtail catheter was placed at the superior level of the descending aorta distal to the interruption and abnormal collateral vessel.

A 5-F balloon-tipped double-lumen catheter (a wedge-pressure catheter with a dull curve at its tip, Arrow International) was advanced from the right femoral vein to the right atrium; it was then further advanced to the left atrium through the patent foramen ovale without a guidewire while the balloon was deflated. There was no resistance transmitted to the catheter when the catheter crossed the foramen ovale. The balloon catheter with fully inflated balloon was further advanced to the left ventricle (Fig. 3A). With the balloon still inflated, further advancement of the catheter to the ascending aorta was facilitated by a 0.035-inch, tip-deflector guidewire (Cook, Inc.) inserted within a guidewire lumen of the balloon catheter to turn the catheter tip up toward the aortic valve in the left ventricle (Fig. 3B). When the catheter tip reached the ascending aorta, the balloon was deflated. The balloon catheter was exchanged to a 4-F catheter (115 cm, Cerulean G, Medikit,) using a long guidewire (260 cm, Radifocus, Terumo) (Fig. 3C). During the catheter
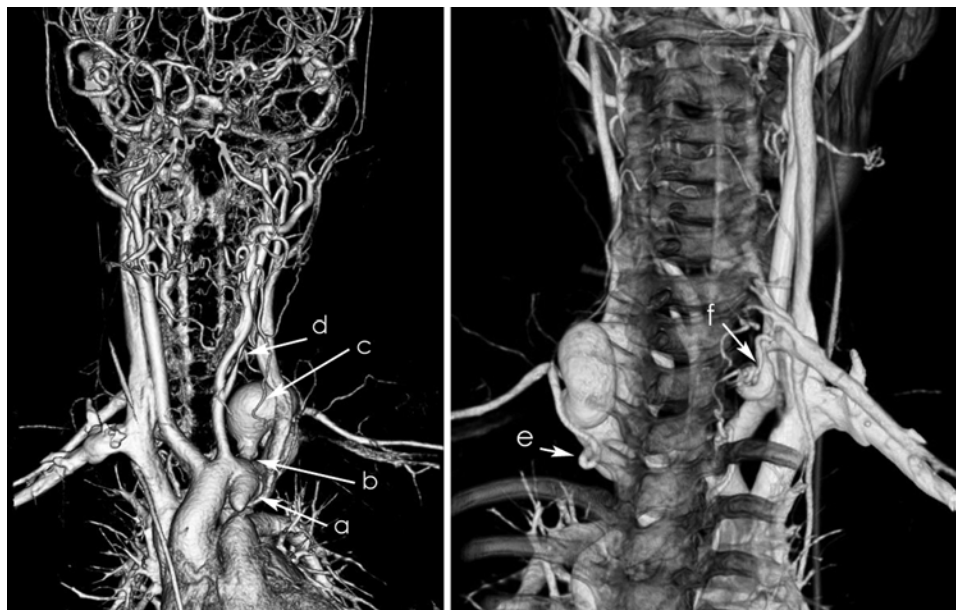

Fɪɢ. 1. Anteroposterior **(left)** and posteroanterior **(right)** view CT angiograms of the aortic arch and brachiocephalic vessels showing aneurysmal dilation (a), interruption (b), and an anomalously dilated collateral vessel (c) feeding into the descending aorta as well as stenosis of left ICA (d). The location of the left carotid bifurcation is extremely low, near the abnormal vessel. There is a small loop at the proximal portion of the subclavian arteries bilaterally (e and f).
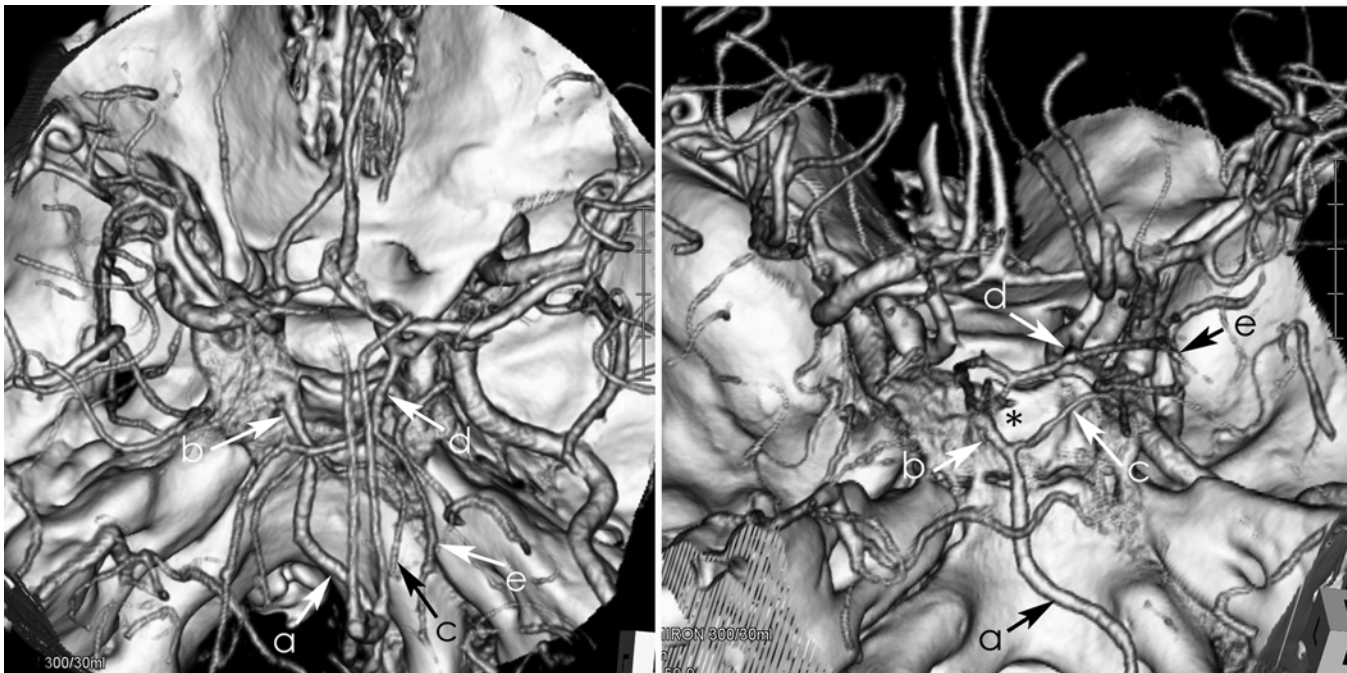
Fig. 2. Vertex **(left)** and posteroanterior **(right)** view CT angiograms of the cerebral vessels revealing connection of the right vertebral artery (a) to the left persistent trigeminal artery (b) and right superior cerebellar artery (c). The right posterior communicating artery (d) is connected to both posterior cerebral arteries and the left superior cerebellar artery. The right posterior cerebral artery is indicated by "e." The left vertebral artery is not demonstrated. The basilar artery distal to the trigeminal artery is also not visualized *(asterisk)*.

exchange, the tip of the guidewire was introduced into the left external carotid artery using the brachiocephalic angiogram for guidance and taking care to avoid inadvertent insertion of the guidewire into the left ICA and aortic arch. Bilateral common carotid angiography was performed by the neurointerventionalist without any difficulty using a regular 0.035-inch guidewire (150 cm, Radifocus, Terumo) (Fig. 4). At the end of the procedure, the catheter was slowly retrieved from the aorta through the heart without a guidewire. During these procedures, no serious arrhythmias were observed.

Findings and Subsequent Management. With transcardiac cerebral angiography, detailed anatomical characteristics of the cerebral vasculature, cerebral hemodynamics, and bilateral stenosis of the cervical ICAs could be evaluated. The short segmental stenosis of the right ICA had not been detected on CTA. The blood flow in the left persistent trigeminal artery was from the basilar artery to the left intracranial ICA, which was not apparent from CTA. There were no ophthalmic collateral vessels from the external to ICAs bilaterally. No adverse effect was observed related to these procedures. Surgical repair
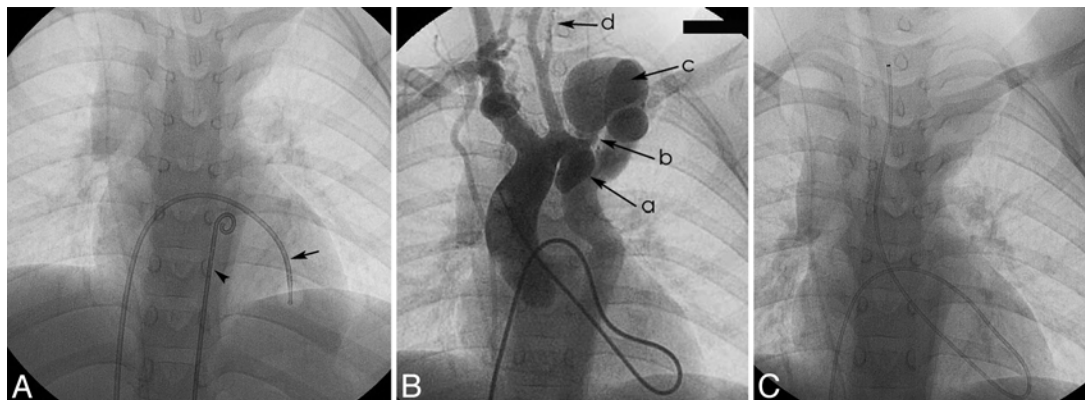

Fig. 3. Tranfemoral venous, transcardiac catheterization. **A:** Plain radiograph showing the balloon-tipped double-lumen catheter *(arrow)* advanced to the left ventricle. The pigtail catheter *(arrowhead)* is in the descending aorta. **B:** Aortogram obtained after injection through the balloon catheter showing the aneurysm (a), interruption of the aorta (b), the dilated collateral vessel feeding into the descending aorta (c), and stenosis of the left ICA (d). **C:** Plain radiograph showing a 4-F catheter coursing the transcardiac route after catheter exchange.
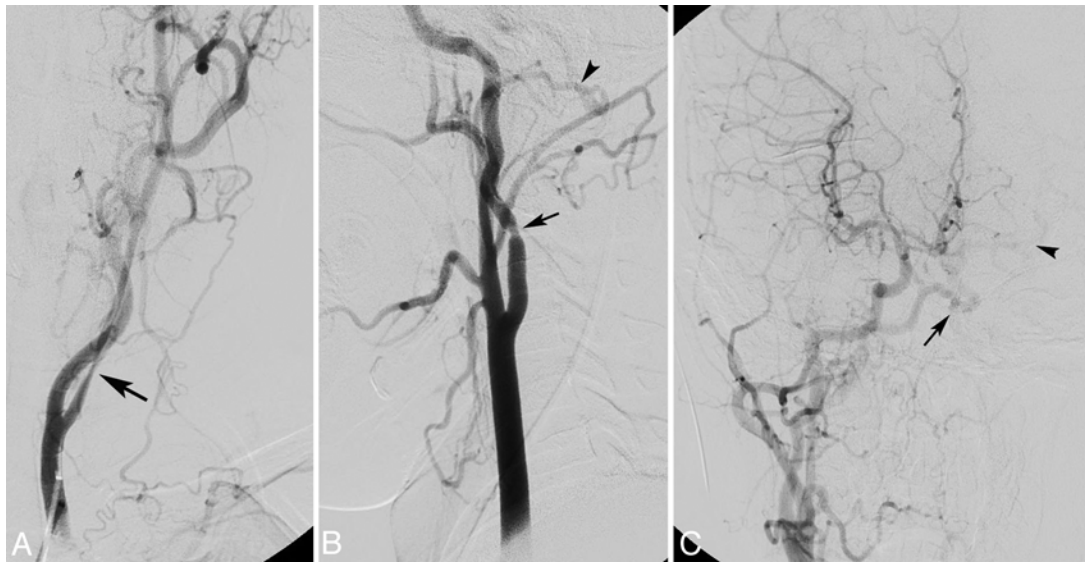
FIG. 4. Transcardiac cerebral angiography.   A: Transcardiac left carotid angiogram (anteroposterior view) showing stenosis of left ICA *(arrow)*.   B: Transcardiac right carotid angiogram (lateral view) showing short segmental stenosis of right ICA *(arrow)*. The right intracranial vertebral artery *(arrowhead)* is supplied by the collateral vessels from the neuromeningeal branch of right ascending pharyngeal artery and right occipital artery.   C: Transcardiac right carotid angiography (anteroposterior view, late arterial phase) showing the left persistent trigeminal artery *(arrow)* and left middle cerebral artery *(arrowhead)*. Blood flow in trigeminal artery is directed from the basilar artery to the ICA.

of the aortic aneurysm was scheduled. The carotid stenoses were managed medically with planned CTA and MRA follow-up.

Discussion

Transfemoral arterial routes are usually used for cerebral angiography in patients of all age groups, and the transbrachial or transradial arterial approach is also commonly used for diagnostic and interventional angiography in adults. The transcardiac approach described in this article is possible for the patient with brain arteriovenous shunts in the neonatal period, especially when a transfemoral arterial approach is not possible due to the small size of the femoral arteries. The umbilical venous route is also useful in the first several days after birth.[7] It is not known whether this approach is possible in the older pediatric patients or even in adults.

The foramen ovale of the atrial septum is bordered by the limbus of the fossa ovalis (septum secundum) and guarded by the valve (floor) of the fossa ovalis (septum primum). In most individuals the foramen ovale closes at birth, but in 25%–33% of the general population it remains patent.[5,9] Patent foramen ovale is of increasing clinical interest since it is related to stroke due to right-to-left embolism, commonly referred to as paradoxical embolism.[6] In an autopsy of 965 normal hearts, the prevalence of a probe-patent foramen ovale (defined as a foramen ovale that can be crossed by a probe) was 30%, 25%, and 20% for age groups 1–29 years, 30–79 years, and 80 years and older, respectively, with no sex-based difference in prevalence.[5] The size of the patent foramen ovale ranged from 1 to 20 mm in diameter (mean 4.9 mm). Thus, there is a 20%–30% chance of the catheter crossing from the right atrium to the left atrium through a patent foramen ovale, even in adults.

It is a common procedure to puncture the atrial septum (fossa ovale) with a standard transseptal needle or radiofrequency transseptal needle and cross the catheter through the perforated atrial septum to the left atrium,[2,10,12] especially in ablation for atrial fibrillation today. This method is called the "Brockenbrough method." The perforated septum closes eventually without any adverse effects. Therefore, in the rare situation that requires transcardiac catheterization for the cerebral arteries, this approach is theoretically possible in adults even if the foramen ovale is closed.

Possible complications of this approach include arrhythmias and cardiac injury during the catheter manipulation. Although arrhythmia is commonly observed during catheter manipulation in the heart, it can be managed by reduction of catheter tension or irritation to the cardiac wall and by changing the position of the catheter tip. When these procedures do not effectively control arrhythmias, we remove the catheter from the heart. When cardiac procedures are performed by experienced pediatric cardiologists, these complications may be minimal or may be entirely avoided.

Transcardiac cerebral access from the femoral vein is possible and useful in the rare situations in which the usual catheter approaches via the femoral or brachial arterial route are not possible as in our case. Cases in which it might be useful include those involving neonates whose femoral arteries are so small that transfemoral angiography is not possible and those involving patients of any age whose femoral and brachial access routes have steno-occlusive changes that preclude catheter introduction. In either situation, cooperation of pediatric cardiologists who are accustomed to the transseptal procedures is essential for this approach.

Finally, a brief discussion of the pathogenesis of mul-

tiple anomalies in PHACE syndrome may help to clarify the necessity of this complicated transcardiac approach. Vascular anomalies in this syndrome include agenesis, hypogenesis, elongation, coiling, looping, steno-occlusive changes, moyamoya phenomena, and persistent carotid-basilar anastomoses, especially a persistent trigeminal artery. Although the pathogenesis of multiple anomalies in PHACE syndrome is not well understood, the heterogeneity is partially explained by an insult to the cephalic neural crest cells, which migrate from the rhombomeres to the branchial arches in the early embryonic period.[11] These neural crest cells contribute to the formation of the outer layers of the forebrain vessels and the brachiocephalic vessels in the metameric fashion.[3] Indeed, the first 3 branchial arches contribute to the formation of the cerebral and carotid arteries, and the fourth branchial arch is related to the formation of the adult aortic arch. Unknown insults in the early embryogenesis may cause multiple vascular anomalies, typically observed in PHACE syndrome.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Komiyama. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Komiyama, Ishiguro, Terada. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Komiyama.

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