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Review article

Bilateral Hypoplasia of the Internal Carotid Artery: A Focused Review

Ljiljana Vasović[1], Ana Mrkaić[2], Miracle Wangsuwana[2], Dimitrije Đorđević[1], Milena Trandafilović[3]

[1]*University of Niš, Faculty of Medicine, Niš, Serbia*
[2]*University of Las Vegas, Las Vegas, United States of America*
[3]*University of Niš, Faculty of Medicine, Department of Anatomy, Niš, Serbia*

SUMMARY

Introduction/Aim. The internal carotid artery (ICA) is a paired artery that primarily supplies the anterior two-thirds of the brain, the pituitary gland, the eye structures, and some other cranial structures. The ICA variations or abnormalities have been more or less described; especially, bilateral ICA hypoplasia (ICAH) was a very rare description that inspired authors to its evaluation.
Methods. The definition of ICAH was based on an artery diameter smaller than 3 mm. Human cases of bilateral ICAH were found by using the corresponding keywords in online bases.
Results. Only 64 cases (28 females, 19 males, and 17 cited cases) were found. Some (ab)normal morphological features such as a narrowing of the ICA beyond a normal initial cervical subpart, the presence of a hypertrophic collateral network, small carotid canals, ICA termination in the ophthalmic artery, an increase of the total flow volume in the vertebral arteries, and a possibility of ICA occlusion and/or cerebral ischemia or hemorrhage at an unusually early age were evidenced. The statistical analysis of cases of known age and gender was performed. A comparison between the main morphological and pathological findings of bilateral hypoplasia and bilateral absence of the ICA was done.
Conclusion. The literature on the ICAH topic provides confirmative and contradictory examples of suggested bilateral ICAH criteria. Some types of cranial hemorrhage and/or aneurysms of the vertebrobasilar system in patients were proven. Paradoxically, aneurysms occurred more frequently in ICAH than in a similar number of cases that described bilateral absence of ICA.

Keywords: internal carotid artery, bilateral hypoplasia, associated pathology, vascular status

Corresponding author:
Milena Trandafilović
e-mail: milena.trandafilovic@medfak.ni.ac.rs

INTRODUCTION

Diameters of arteries, as cited (1), depend on gender, age, body and neck size, and blood pressure. The internal carotid artery (ICA) diameter, as cited (2), increases until the age of 15, maintains the same size during adulthood, and then increases again during old age. On average, ICA caliber is significantly greater in men (5.11 mm) than in women (4.66 mm), although there were no side-to-side differences (3). Omarjee et al. (4) defined ICA hypoplasia (ICAH) as an artery smaller than 3 mm in diameter. It is very well known that many pathological conditions, such as atherosclerosis, neoplasm, arteritis, neurofibromatosis, fibromuscular dysplasia, spasm, increased intracranial pressure, meningitis, and radiation therapy affect the caliber and lead to the ICAH (5). On the other hand, congenital ICAH is a rare finding; only 24 cases of bilateral ICAH were published until 2016, according to Siddiqui and Sobani (6).

Keeping in mind that the first congenital bilateral ICAH-related online title was mentioned in 1957 (7), as cited (8), the purpose of this review was to summarize all cases of bilateral ICAH till 2022 and underline their basic and specific features.

MATERIALS AND METHODS

Definition

The definition of "hypoplasia" (underdevelopment or incomplete development of a tissue or organ) is available online (9).

Study procedure

Keywords such as congenital, bilateral or both sides, hypoplasia, internal carotid artery, and carotid canal were used on online browsers for the recorded ICAH cases.

There were 64 cases of bilateral ICAH reported by the corresponding references on Google Scholar-
https://scholar.google.com/scholar?start=20&q=Congenital+hypoplasia+of+bilateral+internal+carotid+artery&hl=sr&as_sdt=0,5&lookup=0) or
(https://scholar.google.com/scholar?hl=sr&as_sdt=0%2C5&q=congenital+bilateral+hypoplasia+of+carotid+canal&btnG=) and Pub Med
(https://pubmed.ncbi.nlm.nih.gov/?term=Hypoplasia+of+bilateral+internal+carotid+arteries) websites.

The descriptions of the status of cerebral arteries including bilateral ICAH as well as the consequent pathology will be based on the findings of these 64 cases.

This type of study does not require informed consent or Ethics Committee approval.

Statistical analysis

The quantitative findings are expressed in percent and proportion; an additional statistical analysis was performed in SPSS ver. 20.0 (IBM Corp. Released 2011. IBM SPSS Statistics for Windows, Version 20.0. Armonk, NY: IBM Corp.). The Chi-square test was used; a p value less than 0.05 was used as the limit of statistical significance.

RESULTS

General data

There were 64 bilateral ICAH cases, cited as primary or secondary references. The corresponding postnatal periods of all known (49 cases) and/or quoted (17 subjects) cases in the literature are shown in Figure 1. More than a third of the total number of cases of both genders and different ages were reported in Japan (10), France (9), and the USA (6), and there was unrevealed nationality in 8 cases; the remaining 30 cases were detected in 14 other countries. A 16-month-old girl in the USA was the youngest patient (11), while an 87-year-old man in Japan was the oldest (10).

Table 1 presents special characteristics of 28 cases of female gender (5, 6, 11 − 36); Table 2 presents 19 cases of male gender (10, 37 − 53), while Table 3 presents 17 cases of unknown gender and age (4, 7, 54 − 63).

There was no statistically significant difference between males and females ($p = 0.19$), although a ratio of (known) female (28) vs. male (19) cases accounts for 1.47:1.

Age of patients	Bilateral ICA hypoplasia		
	No	Gender	
Newborn (few hours after birth)			
Neonate (≤ 28 days)			
Suckling (≤ 12 months)			
Tot (≤ 3 years	2	2F	
Preschool age (≤ 5 years)			
School age (6–12)	1		1U
Adolescent (13–18)	1	1F	
19–29	3	1F + 2M	
30–39	14	6F + 8M	
40–49	9	5F + 4M	
50–59	11	11F	
60–69	6	2F + 4M	
70–79			
80–89	1	1M	
Unknown gender and/or age	16		16U
Total	**64**	**28F + 19M + 17U**	

Figure 1. *Investigated (64) cases of bilateral internal carotid artery (ICA) hypoplasia shown on the modified map of the world (https://en.wikipedia.org/wiki/File:World_Map_Blank_-_with_blue_sea.svg) and in an overview during the corresponding postnatal periods*

Table 1. *Cases of bilateral internal carotid artery (ICA) hypoplasia in 28 patients of female gender*

No	Patients (age/ gender)	Initial symptoms or reasons of discovery	Outer diameter of both ICAs and/or carotid canals (CCs)	Collateral circulation	Additional arterial variations	Associated pathology	Country [reference]
1	16 mo/F	PHACES syndrome	Diminutive bilateral bony CCs		Hypertrophied left VA; Aplasia of the intradural segment of the right VA; Both VAs were nonunited, and each gave rise PCA; A large calibre of left PCoA; a small calibre of the right PCoA	PHACES syndrome (aortic coarctation/cleft sternum/left maxillary hemangioma)	USA (11)
2	2y/F	Left-sided hemiparesis	Normal sized carotid canals on both sides		Prominent VBS, PCAs and PCoAs	Down syndrome; Cerebral infarction in the right parietal lobe; A past history of the right sided hemiparesis 4 months before.	India (32)
3	17y/F	Following of meningeal hemorrhage				Right PCoA aneurysm	Morocco (18)
4	27y/F	Suspected vasculitis	2 mm at C III vertebra level. Both CCs are small.	Both MCAs and ACAs were filled from the enlarged PCoAs	Hypoplastic left A1 part; ACoA could not be seen; Ectatic and tortuous right VA; Normal course and diameter of the CCAs	Aneurysms of intradural portion of the right VA	Turkey (12)
5	30y/F	Seizures and short-term loss of consciousness just few minutes after childbirth	MRA showing hypoplastic bilateral ICA 1-2 cm distal to their origin: 2.0 mm (right) and 2.6 mm (left). CC: 2.5 mm on the right and 2.8 mm on the left	Bilateral ECAs and VBS enabled collateral circulation		Acute cerebral infarcts of both hemispheres	India (21)
6	30y/F	Acute ischemic stroke	2.8 mm+2.4 mm; Diameter of the carotid canal on the left was smaller (2.7 mm) than that on the right (2.9 mm).		1/25,000 color-coded carotid duplex images		Chinese Taipei (16)
7	30y/F	Tension headaches/ progressive correctable visual loss	Left ICA was smaller than the right one; Left ICA terminated at the base of the skull; Right CC measured approx 4 mm in diameter, whereas the left CC measured only 2 mm in diameter	Both ECAs had numerous collateral vessels but predominately from the MAs which anastomosed with the cavernous ICA bilaterally and the left OphA; The major intracranial blood supply was			USA (28)

				via the VBS through PcoAs;			
8	36y/F	Semicomatose status	Without narrowing of the bony carotid canal on the temporal bone CT	Collateral artery developed from the VA and from the ECA. Anterior circulation was supplied by posterior circulation through the PCoA		Intracerebral and intraventricular hemorrhage	South Korea (23)
9	38y/F	Two spontaneously regressive episodes of amaurosis	2 – 2.5 mm		Bilateral carotid rete mirabile in C3 parts were fed by MAs. Hyperplastic and tortuous VAs, especially on the right side	Pseudoxanthoma elasticum; Congenital narrowing of coronary arteries;	France (33)
10	39y/F	Recurrent attacks of fainting/severe dizziness/ intermittent claudication			Bilateral carotid rete mirabile in C3 parts were fed by multiple small channels of both MAs. Large both ECAs		South Africa (17)
11	42y/F	Progressive mental confusion/ dysphasia/ right-sided paresthesias		Anterior meningeal a. and some branches of the STA filled left ACA and MCA, while MMA, OPhA, STA and OA filled anterior circulation on the right side		A severe stenosis at the left and right OPhAs origin; A pseudo-angiomatous network on both sides	Italy (20)
12	44y/F	Persistent headache/ vomiting	Both CCs are patent	Both OPhA were being filled via hypoplastic ICAs, while both ACAs and MCAs were being filled via BA and PCoAs	.	SAH; Aneurysm of the right AICA origin	Japan (27)
13	45y/F	Treatment of thyotoxicosis			Left carotid rete was supplied by the ICA with some MA contributions; The right rete was filled from the MA; Terminal part of the ICA forms the efferent trunk of the rete	SAH; Thyrotoxicosis	UK (26)
14	46y/F	Amaurosis fugax for about 20 min and persistent visual impairment of the left eye.		Supply of the right MCA and OPhA via opened right PCoA and then the left MCA and OPhA via opened ACoA	Absent left PCoA	Patent foramen ovale	China (36)
15	48y/F	Sudden onset of loss of consciousness/headache /nausea	Both ICAs were hypoplastic from 1 to 2 cm above the carotid bifurcation	Collaterals to the ACAs and MCAs through enlarged PCoAs	Both ICAs terminated as OphAs; Both ACAs and MCAs could not be visualized on the angiograms	Fusiform dilatation of the right PCoA basilar end (suspect aneurysm)	Pakistan (6)

16	50y/F	Coma	Narrowing (2.31 mm) of both ICA 3 cm distal to CCAs bifurcations			Putaminal haemorrhage; Occlusion of both terminal ICAs and PCAs with Moyamoya vessels;	Japan (35)
17	52y/F	SAH			Trans-sellar left-right ICA anastomosis; Carotid rete mirabile was fed by the MA branches	BA tip aneurysm; SAH	UK (22)
18	53y/F	A sudden headache/vomiting/ Unconsciousness an hour before			Right ICA ended as OphA; Right VA, BA and both PCAs were larger than usual		USA (31)
19	54y/F	Occipital headache/right hemifacepa-resthesia/dysarthria /hypertensive peak	Reduced calibre of both CCs	Both ACAs and MCAs were being fill via PCoAs	Caliber of the right and left CCAs were 4.8 mm and 5.4 mm; Enlarged right VA caliber of 7.4 mm, while the left VA was of 6.5 mm)		Brasil (15)
20	55y/F	Left motor weakness			Carotid rete mirabile in the C3 part on both sides via MAs	Pseudoxanthoma elasticum	Japan (13)
21	56y/F	Headache/old silent infarcts	2.8 mm + 2.3 mm (CC: 3.8 and 2.8 mm)		1/25,000 color-coded carotid duplex images		Chinese Taipei (16)
22	56y/F	A loss of con-sciousness followed by fall and slight skull trauma		Both ICAs were hypoplastic from 1–2 cm above the carotid bifurca-tion. A blood supply from the VBS to the carotid system	Enlargement of both VAs (0.4 cm on the right and 0.5 cm on the left), they were larger than CCAs; Left V1 part was tortuous; Right ICA ended as OPhA left ICA ended in OphA and MCA; ACA and MCA could not be visualized	BA tip aneurysm	Italy (14)
23	56y/F	Severe occipital headache/ vomiting	Narrowing of both CCs	Left VA and bilaterally large PCoAs	Right ICA ended in the OPhA; left ICA ended as MCA and OPhA. Hypoplastic and tortuous right VA; dilated left VA. Hypoplastic and tortuous right PICA	Right PICA aneurysm; Diffuse SAH	South Korea (24)
24	56y/F	Sudden onset of loss of consciousness	Narrowing of both CCs		Enlargement of both VAs and PCoAs	BA tip aneurysm	Italy (34)
25	58y/F		Left CC: 2.7 mm; Right CC: 2.4 mm			Left parietal haemorrhagic infarct	France (25)
26	59y/F	Right frontal ischemic lesion of two weeks of evolution	Hypoplasia of both CCs		Right A1 was hypoplastic; High VA flow with velocities of 92 cm/s on the right and 87 cm/s on the left	History of ischemic infarction in the left frontal lobe	Mexico (30)
27	60y/F	Hypertension/ diabetes		VBS and left OPhA		BA aneurysm	France (19)
28	62y/F	Hypertension; Diabetes type 2	Both ICAs were hypoplastic a few		Hypertrophic VBS		Mexico (29)

			millimetres above carotid bifurcation; Hypoplasia of both CCs (right: 2.9 mm; left: 2.8 mm)				

Mo - month; F - female; y - year; VA - vertebral artery; PCA - posterior cerebral artery; PCoA - posterior communicating artery; CCA - common carotid artery; VBS - vertebrobasilar system; C III (vertebra) - the third cervical (vertebra); MCA - middle cerebral artery; ACA - anterior cerebral artery; A1 - pre-communicating part of ACA; ACoA - anterior communicating artery; MRA - magnetic resonance angiography; ECA - external carotid artery; MA - maxillary artery; OPhA - ophthalmic artery; CT - computed tomography; C3 part - cavernous part of ICA; STA - superficial temporal artery; OA - occipital artery; BA - basilar artery; SAH - subarachnoid haemorrhage; AICA - anterior inferior cerebellar artery; V1 part - prevertebral part of VA; PICA - posterior inferior cerebellar artery

Table 2. *Cases of bilateral internal carotid artery (ICA) hypoplasia in 19 patients of male gender*

No	Patients (age/sex)	Initial symptoms or reasons of discovery	Outer diameter of both ICAs and/or carotid canals (CCs)	Collateral circulation	Additional arterial variations	Associated pathology	Country [reference]
1	20y/M	Headache/ vomiting	Hypoplastic both CCs	VBS/PCoAs	Tortuous both PCoAs. Both ICAs terminated as OPhAs.	Left temporo-occipital haemorrhage	Singapore (37)
2	25y/M	Headache	Hypoplastic both CCs	PCoA/ACoA/ Intercavernous anastomosis	Small both CCAs. Intercaver-nous anastomosis		Turkey (38)
3	31y/M	Headache	Bilaterally normal carotid sinus	PCoAs		BA tip aneurysm	Japan (39)
4	31y/M	Progressive weakness of the right upper limb and both lower limbs	Small both CCs		Dilatation of both VAs	Klinefelter syndrome (47, XXY karyotype)	Japan (40)
5	33y/M	Recurrent ischemic episodes referable to left cerebral hemisphere	Bilaterally: ICAs were 2 mm (2 cm beyond the CCA bifurcation). The second abrupt change in each ICA occurred at the Right ICA (midcavernous level) and left ICA (distal cavernous level) became totally occluded.		A radiolucent "cylinder," surrounded by an arterial network (rete), was present bilaterally. ICAs and OPhAs were without connections.	Bilateral ICA hypoplasia had familial appearance (2 from 3 brothers)	USA (41)
6	35y/M	Right sided headaches/left homonymous hemianopsia				Bilateral ICA hypoplasia (brother)	USA (41)
7	35y/M	Headache	Hypoplastic both CCs	ACA/ACoA/MCA territories were supplied by the posterior circulation	Supraclinoid part of both ICAs, ACA and MCA could not be visualized. Enlarged left VA.	Bilobed aneurysm of the BA and left SCA	Turkey (42)
8	36y/M	Transient loss of consciousness. History of fall from a height of 8 feet	Calibre of proximal 1 cm of both ICAs was normal. Diminutive both CCs.	Anterior circulation was supplied by the BA through bilateral dilated and tortuous PCoAs.	Right OPhA probably arose from ACA−ACoA complex, while the left OPhA was not visualized	SAH	India (43)
9	36y/M	Intermittent migraine-like	Hypoplastic both CCs	PCoAs		Hypoplasia of both ICAs	China (44)

		headache 20 years ago				discovered 20 years ago	
10	37y/M	16 days previously he had suddenly collapsed and had been admitted deeply unconscious			Bilateral carotid rete: both ICAs break up into a network at the level of the cavernous sinus; both network communicated with OPhAs and supraclinoid parts of ICAs. Left MA gave off a small branch both to the arterial plexus and to the OPhA.	Small BA tip aneurysm	UK (45)
11	43y/M	Episodes of losing consciousness several years ago	Hypoplastic ICA from the bifurcation to the siphon		Bilateral carotid rete mirabile and bilateral ophthalmic rete. ACA originated from the carotid rete. PCA and MCA arose from the VBS.		USA (46)
12	44y/M	Deep comma			Hypoplasia of ACAs/MCAs ICAs in the neck were not investigated.	Cerebral haemorrhage. Nishimoto's disease.	Romania (47, cited in 41)
13	46y/M	Aphasia/right hemiplegia	Diminution of calibre 1 cm above the CCA bifurcation	Partial revascularization via both VAs			France (48)
14	49y/M	Sudden headache/short-term loss of consciousness				Two aneurysms originating from the right P1 part and left SCA	Turkey (49)
15	60y/M	SAH				Multiple cerebral aneurysms	Japan (50)
16	62y/M	Sudden onset of dizziness	Hypoplastic both CCs		VBS and right PCoA were prominent	Left cerebellar infarct	China (51)
17	64y/M	Vertigo	Hypoplastic both CCs			Right ICA was occluded about 3 cm distal from its origin; left ICA was occluded at the precavernous portion.	Japan (52)
18	67y/M	Episodes of long-standing headache	Hypoplastic both CCs	Anterior circulation was supplied by the hypertrophied right VA through the two PCoAs	Hypetrophic right VA	BA tip and left PCA–PCoA aneurysms	India (53)
19	87y/M	Right giving way	Hypoplastic both CCs		Bilateral narrowing of the CCA. Hypertrophied BA, both VA and PCoAs.		Japan (10)

Y - year; M - male; VBS - vertebrobasilar system; PCoA - posterior communicating artery; OPhA - ophthalmic artery; ACoA - anterior communicating artery; CCA - common carotid artery; BA - basilar artery; VA - vertebral artery; ACA - anterior cerebral artery; MCA - middle cerebral artery; SCA - superior cerebellar artery; SAH - subarachnoid haemorrhage; MA - maxillary artery; P1 - pre-communicating part of the posterior cerebral artery

Table 3. *Cases of bilateral internal carotid artery (ICA) hypoplasia in 17 patients of unknown age and gender*

No	Patients (age/gender)	Initial symptoms or reasons of discovery	Outer diameter of both ICAs	Additional arterial variations	Associated pathology	Country [authors]
1	U				Moya-moya disease	Japan (7, cited in 8)
2	U					U (54, cited in 14)
3	6/U	Citation of one among two cases		Carotid rete		U (55, cited in 14)
4–6	3 cases/U					U (56, cited in 25)
7	U					U (57, cited in 14)
8	U					France (58, cited in 6)
9	U				Aneurysm of PCA	U (59, cited in 14)
10	U					U (60)
11	U					Slovakia (61)
12	U			Carotid rete mirabile		Japan (62)
13	U			Left ICA terminated as OPhA. Persistent right PTA originated from the C3 part and coursed as SCA.	Left PCA–PCoA aneurysm	Japan (63)
14–17	4U/13 patients with PXE				PXE	France (4)

U - unknown; OPhA - ophthalmic artery; PTA - primitive trigeminal artery; C3 part - cavernous part of ICA; SCA - superior cerebellar artery; PCA - posterior cerebral artery; PCoA - posterior communicating artery; ICAH - internal carotid artery hypoplasia; PXE - pseudoxanthoma elasticum

Theories of ICAH appearance

The cause of ICAH has not yet been fully explained (43), although there were some embryological theories discussed. They were related to the failure of the primitive dorsal aorta development and the presence of a normal third primitive aortic arch (PAA3) on one or both sides. That is how the caliber reduction of bilateral ICA from the distal cervical to the cerebral parts was explained (14, 15, 21, 28, 58).

Common carotid artery (CCA) versus ICA(H) anatomy-physiology

The normal average diameter of the CCA and ICA on both sides is 5.9 mm vs. 4.0 mm in women, and 6.5 mm vs. 4.4 mm in men (21).

Some authors reported that the diameter of the ipsilateral CCA with ICAH was smaller in 83% of cases (16). However, a small CCA caliber was herein recorded only in five cases (10.41%) among 48/64 patients with available descriptions of bilateral ICAH (10, 15, 16, 38). Furthermore, Chen et al. (16) showed the characteristic CCA/ICA relations in two female patients of different ages; namely, CCAs calibers were larger in the older woman (56-year old) than in the young woman (30 years) although ICA calibers were hypoplastic and almost of the same size in both women.

Vaghela et al. (32) described the externalization of CCAs due to the waveforms which showed a high resistance flow pattern in the case of bilateral ICAH, while Yaguchi et al. (10) reported a four-fold

increased pulsatility index (PI) in both CCAs and about twice as smaller PI in both vertebral arteries (VAs).

Diagnostic prerequisites and their affirmative or contradictory data

A scheme of bilateral ICAH and the status of some cervical and cerebral arteries in this case are shown in Figure 2.

Kubis et al. (25) presented three morphological criteria required to diagnose bilateral ICAH. These were as follows:

1) Regular narrowing of the ICA beyond a normal initial cervical segment.

There were (available) descriptions of the present ICAH 1 – 2 cm above the bifurcation of the CCA (21, 25, 34, 48) and 3 cm above the CCA bifurcation (52), or at the third cervical vertebra level (12), or throughout its cervical segment in whole (42).

2) The existence of a hypertrophic collateral network

Briganti et al. (14) supplemented the previous criterion by reporting the three paths of collateral circulation:

2.1. Enlarged arteries of the vertebrobasilar system (VBS) and both posterior communicating arteries (PCoAs).

2.2. Branches of the external carotid artery (ECA) and ophthalmic arteries (OPhAs):

There was data that some of the ECA branches were supplementary to the VBS collaterals in two cases (21, 23), or they presented the only collaterals in two cases (20, 28).

2.3. A carotid rete mirabile (CRM) or pseudo-angiomatous anastomoses of the basal ganglia and thalamus.

The CRM—arterial plexus between the maxillary artery (MA) and the cavernous portion of the ICA, followed by a dilation of the OPhA and anastomosis with the MA on both sides, was developed in 10 patients (13, 17, 22, 26, 33, 41, 45, 46, 55, 62). Pseudo-angiomatous network on both sides was described only in one case (20).

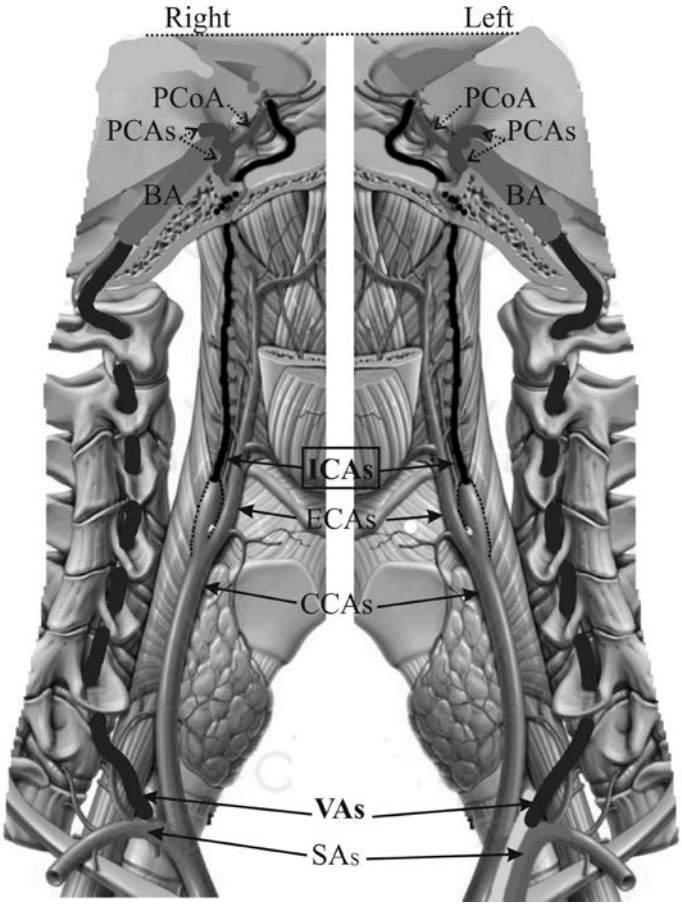

Figure 2. *Bilateral internal carotid artery (ICA) hypoplasia and relation with some arteries in the neck and cranium within modified scheme (https://www.kenhub.com/en/library/anatomy/internal-carotid-artery)*

Collateral circulation was also enabled by the persistent primitive vessels, such as the intercavernous or trans-sellar anastomosis in two cases (22, 38), and the persistent (right) trigeminal artery in one case (63).

3) The same angiographic pattern on a control angiogram (e.g. several months after the initial procedure).

Additional anatomical-physiological-pathological preconditions were as follows:

4) Small or absent carotid canal (CC) (43).

Having in mind that the normal CC diameter ranges from 5.2 to 9.8 mm (25), bilateral hypoplastic CCs were described in 42.55% of cases among 47 patients with ICAH and were more common in patients of male gender. Therefore, the small CC diameter was bilaterally found in 10/19 (52.63%) males (10, 37, 38, 40, 42 − 44, 51 − 53), and in 10/28 (35.71%) females (11, 12, 16, 21, 24, 25, 29, 30, 34). However, it was unilaterally hypoplastic in one case (28), or of the normal size on both sides in three cases (23, 27, 32), or bilaterally hypoplastic in a case of the (right) ICA absence and opposing (left) ICA hypoplasia (64).

5) There is a possibility of occlusion of hypoplastic ICA at the intracranial part, as shown in three reports (34, 35, 61), simultaneous with ICA termination in the OPhA in one of these three cases (34).

6) Bilateral termination of hypoplastic ICA in the OPhA (6, 37, 63), involving the middle cerebral artery on the left side in two cases (14, 24), or unilateral termination in the OPhA (31), or the "filling" of both OPhAs (27) were described in a small number of patients. Some authors described anatomic or physiologic disconnection between hypoplastic ICA and OPhA (36, 41, 43).

7) There was an increase in the total flow volume in bilateral VA, antegrade flow in the ipsilateral OPhA, and increased flow resistance in CCA simultaneously with a mean flow volume of 33 mL/min in both hypoplastic ICAs (16).

8) A foretoken of ICAH relates to the occurrence of cerebral ischemia or hemorrhage at an early age usually, according to Austin and Stears (41). This should not be taken as a rule, since only 2/47 patients (from the infant to the end of the adolescent period) had bilateral ICAH. Bilateral ICAH was discovered upon left-sided hemiparesis and cerebral infarction in the right parietal lobe in a two-year-old girl with Down syndrome (32), and meningeal hemorrhage in a 17-year-old girl (18). However, brain infarction and (meningeal or intracerebral) haemorrhage were more pronounced starting at the age of 30 (16, 21 − 27, 30, 34, 35, 37, 43, 51).

8.1. Investigating the occurrence of headaches in both genders (6/28 females and 8/19 males), this was not a pathological substrate in terms of a foretoken of the cerebrovascular insult and was not statistically significantly more frequent in males ($p = 0.07$).

Additional arterial abnormalities

Previous sections described many arterial variations and abnormalities associated with bilateral ICAH. Here, the focus is on cases with absent arteries, such as:

1. Aplasia of the basilar artery (BA) followed by aplasia of intradural right VA. It was described in a female infant with PHACES syndrome (11).

2. Aplasia of the (left) posterior communicating artery (PCoA). It was associated with the patent foramen ovale in a 46-year-old woman (36).

3. Non-visualization (or aplasia) of the anterior cerebral and middle cerebral arteries and supraclinoid parts of both ICAs (42).

Associated cerebral aneurysms

Single or more than one cerebral aneurysm was found in 34.69% of cases in 49/64 patients (47 cases of known sex plus two cited cases).

Single aneurysms were found in 13/49 cases. The following arteries were targeted: the posterior cerebral artery (PCA) (59), PCoA (6, 18), PCA−PCoA connection (63), intradural VA (12), the posterior inferior cerebellar artery (24), BA (14, 19, 22, 34, 39, 45), and the anterior inferior cerebellar artery (27). Except for one male case (39), and two cases of unknown gender (59, 63), 10/13 cases were of the female gender. Therefore, aneurysms associated with bilateral ICAH in patients of known gender are more frequent in females than in males, although the female-to-male ratio of bilateral ICAH was ~1.5:1.

Two or multiple aneurysms were described in four patients of male gender; two aneurysms developed on the BA and left superior cerebellar artery (SCA) (42), left PCA and SCA (49), and BA and left PCA−PCoA connection (53). Multiple cerebral aneurysms developed in the fourth case, although there was no data on their location (50).

However, the occurrence of the associated cerebral aneurysm or cerebrovascular insult or the absence of pathology was not statistically significant in any of the genders (p = 0.2).

Bilateral ICAH versus bilateral ICA absence (ICAA)

Basic and specific similarities and differences (67, 68) between bilateral ICAH and ICAA (65, 66) cases are shown in Table 4.

Table 4. *Comparison of some general and specific features of the cases of bilateral internal carotid artery (ICA) hypoplasia (ICAH) and previously published cases of bilateral ICA absence (ICAA)*[a]

Number	Parameters	ICAH	ICAA
		General data	
G1	Number of cases /gender	64 (28F+19M+17U)	68 (26F+34M+8U)
G2	The first (available) description	In 1957	In 1913
G3	Youngest – oldest patient	16 months – 87 years	Newborn – 80 years
G4	Number of patients to the period of adolescence	4	25
G5	The countries with the most described cases	Japan/France/USA	USA/Japan
		Embryogenetic base	
E1	Developmental failure	Underdevelopment or incomplete development of tissue or organ (or artery) (9)	Agenesis (the complete failure of development of the ICA including its vestiges (and carotid canal[b])); Aplasia (the complete failure of development of the ICA with the presence of the ICA vestiges) (65)
E2	Hypotheses	The failure of the primitive dorsal aorta development and presence of normal third primitive aortic arch on both sides and the influence of genetic factors.	Interruption of the ICA after a period of its normal growth; An excessive bending of the head; An excessive pressure by amniotic adherences
E3	Association with other congenital disorders	Down syndrome (1); Klinefelter syndrome (1); Patent foramen ovale (1); PHACES syndrome (1); Pseudoxanthoma elasticum (6)	Chiary malformation (1); Chromosome translocation (1); Different cardiac anomalies (4); NOXA1 syndrome (2); PHACES syndrome (4)
		Morphology[c]	
M1	Basic difference	Arterial variation	Arterial abnormality
M2	Status of the vascular source	Hypoplastic common carotid artery (CCA) caliber was recorded only in 5 cases (10.41%).	There was a bilateral absence of the CCA in one case and unilaterally also in one case
M3	Status of the ICA trunk	Regular narrowing of the ICA trunk beyond a normal initial cervical part on both sides.	Bilateral missing of ICA trunk
M4	Status of the carotid canal (CC)	Bilaterally hypoplastic CCs were in 42.55% of the cases; these were more common in men.	Bilaterally absent CCs were found in 61.66% of cases and unilaterally in 3.33% of the cases
M5	Status of the ophthalmic artery (OPhA)	There was bilateral termination of hypoplastic ICA in the OPhA in 5 cases, and unilaterally in one case. There was anatomic or physiologic disconnection between ICA and OPhA in 3 cases.	There were several descriptions of the OPhA origin
M6	Status of the ICA terminal branches	There were data about the hypoplastic caliber of the anterior cerebral (ACA) and	The ACA was bilaterally described (or showed) in 56/60 (known) cases,

		middle cerebral (MCA) arteries only in one case, as well as about their origin from the vertebrobasilar system (VBS) also in one case; Termination of hypoplastic ICA on the left side in the OPhA and MCA was showed in two cases.	except in two cases on the right side, while the MCA was described (or showed) in all cases
M7	Additional arterial variations[d]	Unilateral ectasia of the vertebral artery (VA) (4); Bilateral ectasia of the VA (3); Ectasia of the VBS/posterior cerebral (PCA)/posterior communicating (PCoA) arteries (5); Ectasia of both external carotid arteries (ECAs) (1); Hypoplasia and tortuosity of the right VA and posterior inferior cerebellar artery (PICA) (1)	Origin of the left VA from the aorta (2) or the left-sided arterial duct (1); Hypoplasia of the right VA (2); Duplication of the left VA in the intracranial part (1); Tortuosity of arteries of the VBS (4); Leftward deviation of the basilar artery (BA) (1); Ectasia of the BA (4); Ectasia of the VA (2).
M8	Additional arterial abnormalities[e]	Aplasia of the BA followed by aplasia of intradural right VA in one and left PCoA in other case.	Bridging fenestration into the BA lumen
M9	Persistent primitive carotid-vertebrobasilar anastomoses (CVBAs)	An intercavernous anastomosis (hypothetical segmental remnant of both persistent primitive trigeminal arteries (PPTAs) and the right PPTA were found in three cases.	The PPTA was found in 4 cases and persistent primitive hypoglossal artery (PPHA) in one case
Collateral circulation[‡]			
C1	Definitive arteries	Arteries of the VBS and PCoAs; branches of the ECAs and OPhAs.	Arteries of the VBS and ECAs
C2	Unusual network	A carotid rete mirabile in 10 cases and pseudo-angiomatous anastomosis in one case	Rete mirabile included arteries of the VBS only in one case
C3	Persistent primitive CVBAs	The roles of inter-cavernous anastomosis (in two cases) and the right PPTA (in one case) were proved.	The roles of unilateral PPTA (4 cases), as well as PPHA (proved in one case).
Reasons of discovery			
R1	Headache	It was the first symptom in 14 cases.	It was the first symptom in 19 cases.
R2	Other reasons	There were 7 patients with a loss of consciousness. Other reasons were different and individual.	There were also no specific reasons, such as hemiparesis, dizziness, weakness in the extremities, or diagnostic evaluation of congenital disorders.
Pathology[f]			
P1	Aneurysms (locations)	17/49 (Single and/or multiple): PCoA (2); VA (1); anterior inferior cerebellar a. (1); BA (6); BA+SCA (1); PICA (1); PCA+SCA (1); PCA (1); PCoA-PCA (1); BA + PCoA-PCA (1); multiple cerebral aneurysms (1)	8/60 (VA – 1; PCA – 1; BA – 6)
P2	Other diseases (alphabetical order)	Cerebral hemorrhage (3); Cerebellar infarction (1); Cerebral infarction (4 – single (2) + associated with aneurysm (1) and as an anamnestic data (1)); Fusiform dilatation of the PCoA (1); Intracerebral and intraventricular hemorrhage (1); Occlusion of both ICAs (1); Stenosis of both OPhAs (1); Subarachnoid hemorrhage (3-single finding (1) associated with thyrotoxicosis (1) and aneurysm (1))	Brain compression (1); Cerebellar hemorrhage (2); Cerebral hemorrhage (1); Cerebral infarction (4); Cerebral nodular formations (1); Cranial nerve palsy (1 – single disorder +1 – associated with aneurysm); Malignant process (3); Meningeal hemorrhage (2 – single + 3 – associated with aneurysms); Mental retardation (1); Vertebral artery dissection (1); Vertebral artery stenosis (1)
No explored pathology			
(Un)known cases		27 (11+16)	28 (10+18)

a) Data from the two articles (66, 67)
b) Included opinions of many clinicians, as cited (66).
c) Results are related to the number of patients of the known age/gender aligned in Tables 1-2 and the corresponding cited three cases in Table 3.
d) Divergence in one or more characteristics of an organism or biotype from those typical of or usual for its group (68), or a change in form, position, or condition (https://languages.oup.com/google-dictionary-en/).
e) The quality or state of being abnormal (69), or an abnormal feature, characteristic, or occurrence (https://languages.oup.com/google-dictionary-en/).
f) The congenital disorders were aligned in the row E3.
F - female; M - male; U -unknown.

DISCUSSION

Frequency

Taşar et al. (38) reported that ICAH was bilateral in 40%, right-sided in 40%, and left-sided in 20% of cases in the general population. However, there was only one published bilateral ICAH case among 10,000 cerebral angiograms (26), or two cases in a series of 25,000 color-coded carotid duplex images (16), and a total of 64 cases worldwide in the period from 1957 to 2022 in this review. We found only one paper on this bilateral ICAH in the period from January to November 2023 (70) that was not included in the review for technical reasons.

The influence of embryogenetic factors

Austin and Stears (41) emphasized the influence of genetic factors based on the findings of bilateral ICAH in siblings (brothers). In addition, bilateral ICAH was also discovered in patient(s) with PHACES syndrome (11), Down syndrome (32), pseudoxanthoma elasticum (4, 13, 33), patent foramen ovale (36), and Klinefelter syndrome (40).

Rare congenital arterial malformations, such as Nishimoto's disease (34), or carotid rete mirabile were also associated with bilateral ICAH (13, 17, 22, 26, 33, 41, 45, 46, 55, 62).

Collateral circulation

The collateralization via enlarged VBS and/or PCoAs was described in more than half of the (known) cases (6, 10 − 12, 14, 15, 19, 21, 23, 24, 27, 29, 31, 32, 34, 36, 37, 39, 40, 42 − 44, 48, 51, 53). Increased blood flow through VBS arteries and other carotid system arteries, followed by expanding and consequent thinning of vessels' layers, was probably the reason why they became suitable for the development of aneurysms and/or bleeding.

Age and gender of patients with or without pathology

It is not clear when or why or how bilateral ICAH was initially discovered. Based on the available literature, the authors of this review indicate the following: 1) regarding the first bilateral ICAH description in 1957, Takeuchi and Shimizu (7), as cited (8), investigating Moya-moya disease found bilateral ICAH, or vice versa; 2) regarding the pathology, an evaluation of PHACE syndrome in a 16-month girl (11), and a headache in a 20-year-old man (37), probably contributed to the discovery of bilateral ICAH and its description at an early age and in both genders.

Aneurysms associated with bilateral ICAH in female patients are twice as common in male patients, although the female/male ratio of bilateral ICAH was ~1.5:1. The youngest patient, who was diagnosed with an aneurysm associated with bilateral ICAH, was a 17-year-old girl (18), while the oldest was a 67-year-old man (53). There was no aneurysm associated with bilateral ICAH in 17/49 cases. Single or more than one aneurysm was mainly developed on the arteries of the VBS, except for five cases in which the aneurysm was developed on the PCoA in two cases (5, 18), and the PCA−PCoA connection in two cases (53, 63), as well multiple aneurysms on one (unnamed) cerebral artery or arteries (50).

If we count only 49/64 cases, the percentage of 17 cases with cerebral aneurysms was 34.69%, but if

we take into account all reported cases (64), the percentage was 26.56% and is similar to 24% reported in the study performed by Volpe et al (34).

Bilateral ICAH versus bilateral ICA absence (ICAA)

One of the embryologic hypotheses, as cited (66), indicated that developmental ICA consists of three parts—the root part derived from the PAA3, the intermediate part derived from the dorsal aorta between the PAA3 and PAA1, and the distal part extending from the PAA1 towards the developing brain. This hypothesis of normal ICA development was accepted as an initial basis during the explanation of ICAH (14, 15, 21, 28, 58).

There are some similarities, as well as differences between the cases of bilateral ICAH and ICAA (66, 67), as seen in Table 4. Similarities relate to the number of cases (64 ICAH vs. 68 ICAA), the known number of female cases, countries where both ICAH and ICAA cases were discovered, unsubstantial influence on the CCA status and terminal ICA branches, including predominantly arteries of the VBS and branches of the ECAs in collateral circulation, the consequential appearance of aneurysms on VBS arteries, as well wider spectrum of congenital and other pathological disorders. Differences relate to the substantial meaning of (arterial) hypoplasia and (arterial) absence (aplasia and agenesis); other ones relate to the number of male and unknown cases, time of the first publishing, the number of youngest patients, embryologic hypotheses, the status of the osseous carotid canal, the appearance of unusual arterial network and incidence of aneurysms. Namely, the incidence of aneurysms associated with bilateral ICAH cases is two or two and half times higher compared to the percentage of aneurysms associated with bilaterally absent ICA (13.33%) (66, 67).

LIMITATION

1. Disputable equalization of terms such as aplasia versus hypoplasia (27, 50, 53, 65) or hypoplasia versus occlusion (52) of both ICAs.

2. A lack of data regarding gender and/or age and/or correct terminology in 17 cited cases (as seen in Table 3).

3. Unclear descriptions of the caliber status (mild narrowing vs. segmental narrowing vs. severe stenosis) of one ICA in the presence of hypoplasia of the opposing artery (20).

4. There was a disputable case included in this review where bilateral ICAH was not seen on the posterior maximum intensity projection (MIP) view, while other types of imaging methods were not presented (11).

5. Non-visualization of some arteries on the angiograms does not necessarily mean that they were absent (5, 12, 14, 42, 43).

6. The data on associated pathology including congenital disorders are based on published 50/64 cases (47 cases of known gender and 3 cited cases of unknown gender).

CONCLUSION

Bilateral ICAH is in the true sense of the word one morphological rarity. The importance relates to its association with congenital and acquired pathological disorders, especially cerebral aneurysms which have been found in about 35% of the cases.

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Bilateralna hipoplazija unutrašnje karotidne arterije: fokusirani pregled

Ljiljana Vasović[1], Ana Mrkaić[2], Miracle Wangsuwana[2], Dimitrije Đorđević[1], Milena Trandafilović[3]

[1]*Univerzitet u Nišu, Medicinski fakultet, Niš, Srbija*
[2]*Univezitet u Las Vegasu, Las Vegas, Sjedinjene Američke Države*
[3]*Univerzitet u Nišu, Medicinski fakultet, Katedra za anatomiju, Niš, Srbija*

S A Ž E T A K

Uvod/Cilj. Unutrašnja karotidna arterija (*a. carotis interna* – ACI) jeste parna arterija koja prvenstveno snabdeva prednje dve trećine mozga, hipofizu, strukture oka i neke druge strukture lobanje. Varijacije ili abnormalnosti ACI-ja manje-više su opisane; mimo toga, bilateralna hipoplazija ACI-ja (ACIH) veoma je retko opisivana, što je inspirisalo autore da je ispitaju.
Materijal i metode. Definicija ACIH-a zasnovana je na prečniku arterije manjem od 3 mm. Slučajevi bilateralnog ACIH-a kod ljudi nađeni su korišćenjem odgovarajućih ključnih reči u onlajn bazama podataka.
Rezultati. Pronađena su samo 64 slučaja (28 žena, 19 muškaraca i 17 citiranih slučajeva). Dokazane su neke normalne/abnormalne morfološke karakteristike kao što su suženje ACI-ja izvan normalnog inicijalnog cervikalnog dela, prisustvo hipertrofične kolateralne mreže, mali karotidni kanali, završetak ACI-ja u oftalmičkoj arteriji, povećanje ukupnog protoka u vertebralnim arterijama, a potvrđena je i mogućnost okluzije ACI-ja i/ili cerebralne ishemije ili krvarenja u neuobičajno rano.
Urađena je statistička analiza slučajeva poznate starosti i pola. Urađeno je poređenje glavnih morfoloških i patoloških nalaza bilateralne hipoplazije i bilateralnog odsustva ACI-ja.
Zaključak. Literatura u vezi sa ACIH-om daje potvrdne i kontradiktorne primere predloženih bilateralnih kriterijuma ACIH-a. Dokazani su neki oblici kranijalnog krvarenja i/ili aneurizme vertebrobazilarnog sistema kod bolesnika.
Paradoksalno, aneurizme su se češće javljale u ACIH-u nego u sličnom broju slučajeva koji su dokazali bilateralno odsustvo ACI-ja.

Ključne reči: **unutrašnja karotidna arterija, bilateralna hipoplazija, pridružena patologija, vaskularni status**