Collateral blood flow patterns in patients with unilateral ICA agenesis and cerebral aneurysm

Mohammad A. Jamous, Hani Abdel-Aziz & Farouk Kaisy

Department of Neurosciences, Jordan University of Science and Technology, Irbid, Jordan

Correspondence to: Mohammad A. Jamous, MD., PhD.
Department of Neurosciences, School of Medicine,
Jordan University of Science and Technology,
P.O.BOX 3030, Irbid 22110 JORDAN
FAX: +96279-9773827
EMAIL: mohabujamous@yahoo.com

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Abstract

BACKGROUND: Agenesis of one or both internal carotid arteries (ICA) is usually a benign congenital anomaly. Many patients are diagnosed incidentally and remain asymptomatic, however associated cerebral aneurysms can be life-threatening and result in high morbidity and mortality rates in this population.

MATERIAL & METHODS: Based on the timing of ICA agenesis, during the fetal life, we classified the collateral blood flow pattern into three major types; type I: Collateral blood flow via primitive vessels, type II: Collateral blood flow via ICA branches, and type III: collateral blood flow via branches of the external carotid artery (ECA). The type of collateral blood flow pattern in the reported patients with ICA agenesis and cerebral aneurysm was reviewed in order to determine the relation between the type of collateral blood flow and the development of CAs.

RESULTS: Twenty nine patients with ICA agenesis and CA were reported, of these 27 patients (93%) were found to have type II collateral blood flow, 2 patients (7%) had type I collateral blood flow pattern and none had type III collateral flow pattern. The majority of patients (79%) with CA secondary to ICA agenesis presented with subarachinoid hemorrhage (SAH).

CONCLUSION: Although ICA agenesis is a symptomatic in the majority of cases, it must be recognized because it promotes the development of CA and SAH. Our new classification system for collateral blood flow patterns is easy to use and can predict those at high risk to develop cerebral aneurysms and SAH, and therefore, need annual screening MRA.

ORIGINAL ARTICLE

INTRODUCTION

Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare congenital anomalies, occurring in less than 0.01% of the population [1,2]. The term absence has been chosen to incorporate agenesis, aplasia, and hypoplasia of the ICA. In this setting, the most common type of collateral flow is through the circle of Willis. Less commonly, collateral flow is provided via persistent embryonic vessels or from transcranial collaterals originating from the external carotid artery (ECA) system. Slightly more than 100 cases of congenital absence of the ICA have been reported in the literature [3]. We report

four new cases of absence of the ICA, including one with an associated intercavernous anastomosis.

Agenesis of one or both internal carotid arteries (ICA) is usually asymptomatic in the majority of cases, because adequate collateral supply to the involved ICA vascular territory develops early in life; however some patients are at higher risk to develop CA and fatal SAH. Servo [4] found that 25% of patients with ICA agenesis presented with SAH caused by an aneurysm. Altered hemodynamics produced by absence of the ICA may be responsible for the high frequency of CA.

Here the authors proposed new system to describe the patterns of collateral blood flow in patients with ICA agenesis. The relation between CA and the pattern of collateral blood flow will be examined using our new system.

MATERIAL AND METHODS

The authors classified the type of collateral blood flow in patients with ICA agenesis into three major types. Figure 1 demonstrates schematic illustration of the new classification for the collateral circulation in patients with unilateral ICA agenesis. Type I: Perfusion of the ipsilateral ACA and MCA occurs via primitive vessels. This primitive vessel can be rete mirabilis (Ia), primitive trigeminal artery (Ib), or intercavernous anastomosis (Ic). Type II: Perfusion of the ipsilateral side is provided by collaterals derived from the circle of Willis. This type of collateral blood flow has three patterns; IIa: ACA perfused via the enlarged ACOMA, MCA perfused via the enlarged PCOMA, IIb: both the ACA and MCA are supplied via an enlarged ACOMA, and IIc: ACA and MCA are supplied via both ACOMA and PCOMA. Type III: Perfusion of the affected side is via a transcranial anastomosis from the ECA. The authors reviewed the previously reported cases of ICA agenesis and cerebral aneurysms and used the new classification system to examine the relation between the collateral flow pattern and the occurrence of CAs.

RESULTS

Tables 1 and 2, shows the collateral blood flow patterns in 29 patients with ICA agenesis and CAs (reported from 1957–2003). 27 patients (93%) were found to have type II, 2 (7%) had type I, and none had type III collateral flow pattern. Of the 27 patients with type II collateral flow pattern; 19 patients (70%) had type IIa (ACA perfused via the enlarged ACOMA, MCA perfused via the enlarged PCOMA), 7 (26%) had type IIb (both the ACA and MCA are supplied via an enlarged ACOMA) and one patient had type IIc (ACA and MCA are supplied via both ACOMA and PCOMA) collateral flow pattern. The two patients with CAs secondary to type I collateral flow, were found to have type Ic collateral flow pattern (intercavernous anastomosis).

ACOMA was the most common aneurismal site, it was reported in 21 patients (72%). 5 (17%) had ICA aneurysms, 4 (14%) had basilar tip, and 3 patients (10%) had MCA aneurysm. Twenty three patients (79%) presented with SAH.

DISCUSSION

The internal carotid artery originates during the 4th week of gestation from the 3rd aortic arch. Its distal part divides into anterior and posterior segments that subsequently form the anterior-, middle-, and posterior cerebral arteries. These vessels will form the nidus of the developing circle of Willis [5,3,7]. Although the exact etiology of ICA agenesis has not been established, an insult to the developing embryo may be responsible for this developmental anomaly [8]. The formation of an effective, compensatory collateral system during the early stages of embryogenesis may explain the absence of major neurological deficits in patients with ICA agenesis.

Diagnostic angiograms clearly reveal the pathology and identify the collateral flow to the ipsilateral side in patients with ICA agenesis. In addition, non-invasive imaging modalities such as color Doppler and MRA are useful diagnostic tools [9,10]. However, angiography remains important not only to diagnose ICA agenesis but also to rule out associated vascular pathologies. The absence of the carotid canal on skull-base CT scans confirms the congenital nature of the underlying pathology [11].

Using angiographic findings, Tsuruta and Miyazaki [12] divided patients with unilateral ICA agenesis into 5 groups. However, their classification does not cover all the reported variations in the collateral blood flow. Similarly, the categorization into 6 groups proposed by others [13], who examined the collateral blood flow patterns in patients with unilateral or bilateral ICA agenesis, is not entirely satisfactory.

The type of collateral flow that develops is closely related to the timing of the ICA agenesis [5,6]: if it occurs before the development of the Willis circle, collateral flow will be via primitive vessels. If, on the other hand, it occurs during the development of the circle of Willis, collateral flow is provided via the arteries of the Willis circle. Lastly, if ICA agenesis occurs at a later stage, collateral flow will be provided primarily via the external carotid artery (ECA). Based on these etiological considerations, we propose an alternative system to classify collateral blood flow in patients with ICA agenesis (Figure 3). Our proposed classification is more universal in that it accommodates all reported cases of ICA agenesis while remaining simple to use. The true frequency of different collateral flow pattern in patients with ICA agenesis is difficult to estimate because the majority of patients with ICA agenesis remain asymptomatic.

Although ICA agenesis is usually asymptomatic, it has to be recognized because of its frequent association with CA [1]. The coexistence of CA and this anomaly may support the observation that congenital factors may predispose to CA aneurysm formation, since vascular

Table 1. Reports of patients with unilateral ICA agenesis and cerebral aneurysm.

Series (ref. no.)	Age (yrs), Sex	ICA agenesis side	Type of collateral flow	Aneurysm location	SAH
Legarde, 1957 [12]	42, M	Right	IIb	ACOMA	+
Burmester, 1961 [3]	43, M	Right	IIa	ACOMA	+
	32, F	Left	IIb	ACA	
Moyes, 1969 [14]	37, F	Left	IIa	Basilar	+
Tangachai and Khaoborisut, 1970 [23]	30, F	Left	IIa	MCA	+
Handa, 1971 [7]	28, F	Left	IIb	ICA (Cavernous)	+
Sakurai, 1972 [18]	66, M	Right	IIa	ACOMA	+
Teal, 1973 [24]	66, F	Left	IIa	ACOMA, ICA (Cavernous)	–
Carella, 1975 [4]	19, F	Right	IIa	PCA	+
Servo, 1977 [19]	48, M	Left	IIa	ICA (carotid siphon)	+
Tsuruta and Miyazaki, 1977 [25]	12, F	Left	IIa	ACOMA	+
Waga, 1978 [26]	60, F	Left	IIa	ACOMA	+
Shigemori, 1980 [20]	48, F	Left	IIa	ACOMA	+
Bernini, 1980 [2]	38, F	Right	IIa	ACOMA	+
Kunishio, 1986 [10]	70, M	Left	IIa	ACOMA, MCA, Basilar tip	+
Yoshida, 1988 [27]	67, F	Left	IIa	ACOMA	+
Quint, 1992 [17]	60, F	Right	Ic	ICA (Supraclinoid), ACOMA, MCA	–
	65, F	Left	Ic	ACOMA	+
Nakai, 1992 [16]	27, F	Left	IIb	ACOMA	+
Ide, 1995 [9]	38, F	Right	IIa	PCA	+
Armand, 1996 [1]	NDA	Left	IIb	ACOMA	+
Sugiura, 1997 [22]	65, F	Right	IIa	Basilar tip	+
	67, F	Right	IIa	Basilar trunk	–
Czarnecki, 1998 [5]	45, M	Left	IIb	ACOMA (two)	–
Lee, 2003 [11]	55, M	Right	IIa	ACOMA	+
	19, F	Left	IIa	ACOMA	+
	51, M	Right	IIa	ACOMA	+
	57, F	Left	IIc	ACOMA	–
	50, F	Right	IIb	PCOMA, ICA (Cavernous)	–

SAH; subarachnoid hemorrhage, +; present, –; absent, ACOMA; anterior communicating artery, ACA; anterior cerebralartery, MCA; middle cerebral artery, ICA; internal carotid artery, PCA; posterior cerebral artery, PCOMA; posterior communicating artery, NDA: no data avialable

anomalies tend to occur together [14]. However many authors have suggested that altered hemodynamics produced by absence of the internal carotid artery may be responsible for CA formation in patients with ICA agenesis [12,15,16]. Because the frequency and location of CA in those patients depends on the pattern of collateral blood flow, which is associated with variable degrees of hemodynamic stress, we think that altered hemodynamics is the main factor promoting the formation of CAs in patients with ICA agenesis.

Table 1 and 2 show that none of the reported patients with ICA agenesis and CA had type III collateral flow, in which the ipsilateral side is fed from external carotid artery anastomoses, on the other hand 93% of these patients showed that blood supply to the ipsilateral ICA territory is derived from the normal ICA via the circle of

	Collateral blood flow pattern						
	I(n=2)			II(n=27)			
	Ia	Ib	Ic	IIa	IIb	IIc	III
No. of patients with CA	0	0	2	19	7	1	0
No. of patients with multiple CA	–	–	1	2	2	0	–
No. of patients with CA and SAH	–	–	1	17	5	0	–
Aneurysm location:							
ACOMA	–	–	2	13	5	1	–
MCA	–	–	1	2	0	0	–
ICA	–	–	1	2	2	0	–
BA	–	–	0	4	0	0	–
PCA	–	–	0	1	0	0	–
PCOMA	–	–	0	0	1	0	–
ACA	–	–	0	0	1	0	–

Ia, Ib, Ic, IIa, IIb, IIc, III collateral flow patterns; see figure 3,
CA: cerebral aneurysm SAH: subarachnoid hemorrhage,
ACOMA: anterior communicating artery, MCA: middle cerebral artery, ICA: internal carotid artery, BA: basilar artery, PCA: posterior cerebral artery, PCOMA: posterior communicating artery,
ACA: anterior cerebral artery, –: no CA

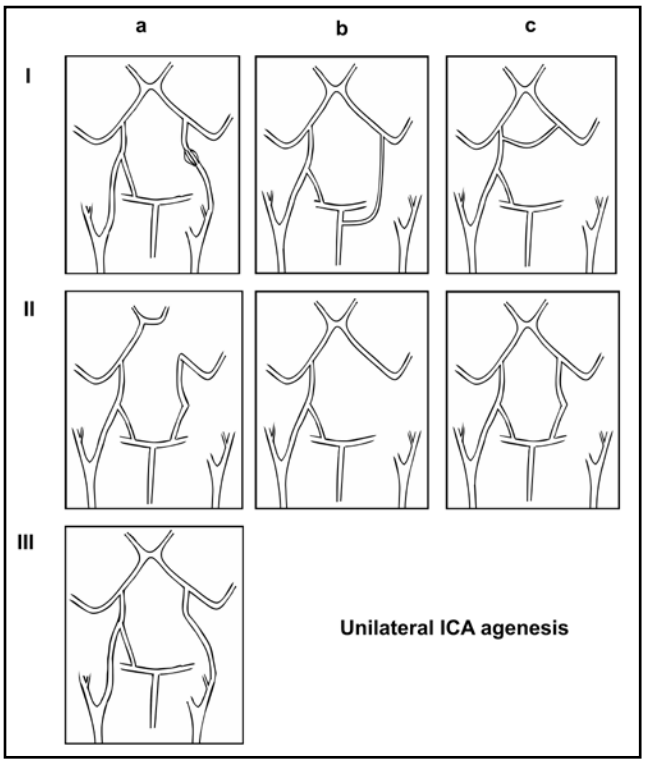

Figure 1. Schematic illustration of our new classification for the collateral circulation in patients with unilateral ICA agenesis.
Type I: Perfusion of the ipsilateral ACA and MCA occurs via primitive vessels.
Ia: ICA perfusion via the rete mirabilis
Ib: ICA perfusion via the primitive trigeminal artery
Ic: ICA perfusion via intercavernous anastomosis.
Type II: Perfusion of the ipsilateral side is provided by collaterals derived from the circle of Willis.
IIa: ACA perfused via the enlarged ACOMA, MCA perfused via the enlarged PCOMA
IIb: both the ACA and MCA are supplied via an enlarged ACOMA
IIc: ACA and MCA are supplied via both ACOMA and PCOMA.
Type III: Perfusion of the affected side is via a transcranial anastomosis from the ECA.

Willis (type II), and 7% of the patients had collateral flow via primitive vessel (type I).

Type IIa (ACA perfused via AcomA, MCA via PcomA) collateral flow pattern was encountered in 19 (70%) and type IIb (both ACA and MCA supplied via enlarged AcomA) was found in 7 (26%) of the 27 patients with type II collateral flow pattern. The patterns of collateral flow in the reported patients with ICA agenesis does not necessarily reflect the true frequencies of these collateral flow patterns in patients with this anomaly, since these are the patients that were selected for angiography because they became symptomatic. Obviously, there is no population- based study ascertaining the incidence of ICA agenesis and patterns of collateral flow in totally asymptomatic patients. Therefore the high frequency of type IIa and IIb collateral flow patterns in the reported cases of ICA agenesis and CA suggests that these collateral flow patterns are associated with high incidence of CAs, which makes it more symptomatic and therefore more prevalent than other collateral flow patterns.

23 (79%) Of the 29 patients with unilateral ICA agenesis and CAs presented with SAH. We posit that the high rupture rate may be attributable to the exposure of the aneurysms to high hemodynamic stress. While 15 (94%) of the 16 patients with ICA agenesis and CA, reported before 1990, presented with SAH, only 8 (62%) of the 13 patients diagnosed after 1990 had SAH. Current widespread of cerebral angiography and MRA may explain the early detection of this malformation before the development of SAH. 82% of the aneurysms in patients with type IIa and IIb collateral flow patterns presented with SAH, while only 33% of the patients with other flow patterns presented with SAH, this indicates that type IIa and IIb collateral flow patterns not only predispose to CA formation, it also promote its rupture.

The high levels of shear stress associated with type IIa and IIb collateral flow patterns, especially in the vessels of the anterior circulation, may explain the high frequency of CAs in these two collateral flow patterns, other flow patterns appear to be more balanced and associated with less hemodynamic stress. The discontinuity of the Willis circle in type IIa and IIb is another factor that may

contribute to the high frequency of CAs; the collateral flow produced by primitive vessel (type I) or ECA (type III) preserve, in one way or another, the continuity of the willis circle, which is not the case in type IIa and IIb collateral flow. Understanding the anatomy of the Willis circle is very important when treating patients with ICA agenesis and CAs; microsurgical or endovascular interference in patients with ICA agenesis must aim at preserving the compensatory collateral circulation that provides blood to the territory of the absent ICA.

Because of the high risk for CA formation in patients with ICA agenesis, some authors recommended annual screening MRA for incidentally diagnosed patients in order to detect early development of CA [9,11]. Based on this report, we conclude that screening MRA is not needed for patients with balanced collateral flow patterns, because the risk for developing CA is very low, on the other hand those with type IIa and IIb collateral flow patterns require annual screening MRA, since these flow patterns appear to be more aneurysmogenic and associated with high risk for SAH.

CONCLUSION

Although ICA agenesis is a symptomatic in the majority of cases it must be recognized because it promotes the development of CA and SAH. Our new classification system for collateral blood flow patterns is easy to use and can predict those at high risk to develop cerebral aneurysms and SAH, and therefore, need annual screening MRA.

Microsurgical and endovascular treatment of cerebral aneurysms in patients with ICA agenesis must not interfere with the compensatory flow pattern to the absent ICA side.

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