8. Lewis GJT, Douglas DM, Reid W, Kennedy Watt J (1967) Cystic adventitial disease of the popliteal artery. BMJ III:411–414
9. Chevrier JL (1962) Un cas de dégénérescence colloïde de l'adventitice de l'artère poplitée. Mem Acad Chir 88:261–265
10. Powis SJA, Morrissey DM, Jones EL (1970) Cystic degeneration of the popliteal artery. Surgery 67:891–894
11. Deutsch AL, Hyde J, Miller SM, Diamond CG, Schanche AF (1985) Cystic adventitial degeneration of the popliteal artery: CT demonstration and directed percutaneous therapy. AJR 145:117–118
12. Ishikawa K, Mishima Y, Kobayashi S (1961) Cystic adventitial disease of the popliteal artery. Angiology 12:357–361
13. Harris JD, Jepson RP (1965) Cystic degeneration of the popliteal artery. Aust NZ J Surg 34:265–269
14. Rizzo RJ, Flinn WR, Yao JS, McCarthy WJ, Vogelzang RL, Pearce WH (1990) Computed tomography for evaluation of arterial disease in the popliteal fossa. J Vasc Surg 11:112–119
15. Patt RH, Lossef SV, Rajan S, Laub G, Carvlin MJ, Barth KH (1990) Optimizing MR angiography of popliteal–tibial arteries. (abstract) Radiology 177:279
16. Crolla RM, Steyling JF, Henippman A, Slootweg PJ, Taams A (1993) A case of cystic adventitial disease of the popliteal artery demonstrated by magnetic resonance imaging. J Vasc Surg 18:1052–1055
17. Bunker SR, Laufen GJ, Hutton JE Jr (1981) Cystic adventitial disease of the popliteal artery. AJR 136:1209–1212

Congenital Absence of the Internal Carotid Artery

Francesco Florio,[1] Silverio Balzano,[1] Michele Nardella,[1] Vincenzo Strizzi,[1] Mario Cammisa,[1] Vincenzo Bozzini,[2] Giuseppe Catapano,[2] Vincenzo D'Angelo[2]

[1]Department of Radiology, Unit of Angiography and Interventional Radiology, ''Casa Sollievo della Sofferenza'' Hospital, Viale dei Cappuccini 1, I-71013 S. Giovanni Rotondo (Foggia), Italy
[2]Department of Neurosurgery, ''Casa Sollievo della Sofferenza'' Hospital, Viale dei Cappuccini 1, I-71013 S. Giovanni Rotondo (Foggia), Italy

Abstract

We report three cases of congenital absence of an internal carotid artery (ICA), diagnosed incidentally by digital subtraction angiography. The analysis of the cases is based on the classification of segmental ICA agenesis proposed by Lasjaunias and Berenstein. Usually the patients with this rare vascular anomaly are asymptomatic; some may have symptoms related to cerebrovascular insufficiency, compression by enlarged intracranial collateral vessels, or complications associated with cerebral aneurysms. Diagnosis of congenital absence of ICA is made by skull base computed tomography (CT) scan, CT and magnetic resonance angiography, and conventional or digital subtraction angiography.

Key words: Agenesis—Internal carotid artery—Cerebrovascular insufficiency—Intracranial aneurysm

Congenital absence of the internal carotid artery (ICA) is a very rare vascular anomaly. Agenesis, aplasia, or hypoplasia are the three conditions responsible for an absent ICA and reflect different developmental failures. This anomaly may be completely asymptomatic and detected occasionally by skull base computed tomography (CT) scan, magnetic resonance imaging (MRI), angiography, or at autopsy or surgical procedure. The symptoms are related to cerebrovascular insufficiency, compression by enlarged intracranial vessels, and associated cerebral aneurysms. Diagnosis is important to explain the clinical and surgical problems related to this condition.

Correspondence to: F. Florio, M.D.

We report three cases of congenital absence of the ICA. Patients 1 and 2 showed absence of one ICA with ipsilateral cerebral circulation supplied by the anterior communicating artery and/or posterior communicating artery. Patient 3 is an example of hypoplasia of the cervical portion of the ICA with the precavernous portion formed by a rete mirabile of vessels through the skull base. We discuss the clinical symptoms, diagnostic methods, and clinical and surgical implications of this condition.

Case Reports

Case 1

A 62-year-old woman was admitted to our hospital for acute onset of severe occipital-nuchal headache and loss of consciousness. The neurological examination revealed nuchal rigidity without focal neurological deficits. CT scan showed subarachnoid hemorrhage. Emergent cerebral angiography demonstrated an anterior communicating artery aneurysm and absence of the left ICA. The omolateral anterior cerebral artery was supplied through the anterior communicating artery; the middle cerebral artery was supplied through a hypertrophic posterior communicating artery arising from the contralateral posterior cerebral artery (Fig. 1). The patient underwent craniotomy and clipping of the aneurysm. The postoperative period was complicated by bilateral middle cerebral artery vasospasm; post-hemorrhagic hydrocephalus was treated with ventriculoperitoneal shunting. The patient has no neurological deficits.

Case 2

A 28-year old woman, treated surgically in 1988 and 1992 for an anaplastic astrocytoma in the right parietal region, was admitted to our hospital for a recurrence of the neoplasm. The neurological examination showed a left hemiparesis. MRI of the brain revealed a recurrence of the tumor with contralateral parietal extension. Surgical treatment was excluded. Digital

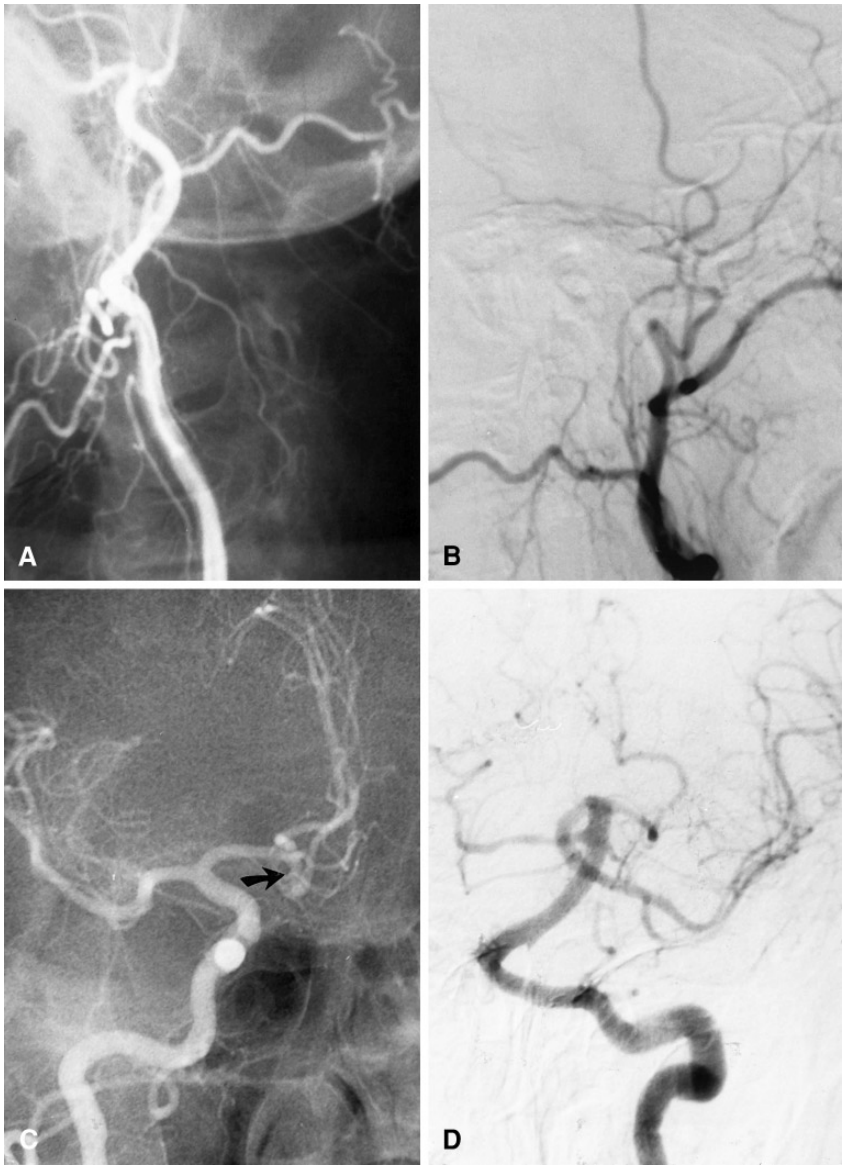


Fig. 1. *Case 1.* Absence of the left internal carotid artery (ICA) with associated anterior communicating artery aneurysm. **A, B** Left common carotid angiogram (oblique and lateral view): absence of left ICA. **C** Right common carotid angiogram (oblique view): opacification of both anterior cerebral arteries and a small anterior communicating artery aneurysm (arrow). **D** Left vertebral angiogram (anteroposterior view): left middle cerebral artery is supplied through a hypertrophic posterior communicating artery arising from the contralateral posterior cerebral artery.

subtraction angiography, performed for intraarterial chemotherapy, demonstrated congenital absence of the left ICA; both homolateral anterior and middle cerebral arteries were supplied through the anterior communicating artery (Fig. 2).

Case 3

A 66-year-old man was admitted to our department for vertigo and loss of consciousness. Neurological examination showed no focal deficits. MRI of the brain showed no pathology. Cerebral angiography revealed a kinking of the right common carotid artery and hypoplasia of the cervical portion of the left ICA. The petrosal portion was filled by a rete mirabile of skull base vessels (Fig. 3). The patient was discharged with medical therapy.

Discussion

Congenital absence of the ICA is a very rare vascular anomaly, with only about 100 cases reported in the literature since the first case described by Tode in 1787. As stated by Padget [1], such an anomaly occurs before 24 days (3-mm stage) of embryogenesis, when the ICA is formed from the terminal segments of the dorsal aorta and the third aortic arch arteries. The three conditions responsible for absent ICA are agenesis, aplasia, and hypoplasia. These are related to different developmental failures [2]. In most cases the anomaly is unilateral. In less than 10% of cases both ICAs are involved. There is a predilection for the left ICA, with a ratio of about 3 : 1.

ICA agenesis can be analyzed following the classification by Lasjaunias and Berenstein [3], based on embryological criteria. The authors describe the segments of the fully developed ICA system in terms of the origins of branches of embryonic vessels. The seven segments are as follows: 1, cervical segment (third aortic arch); 2, ascending intrapetrous segment (between the third and second aortic arches); 3, horizontal petrous segment (between the first and second aortic arches); 4, ascending segment in foramen lacerum (between the first aortic arch and primitive maxillary artery); 5,

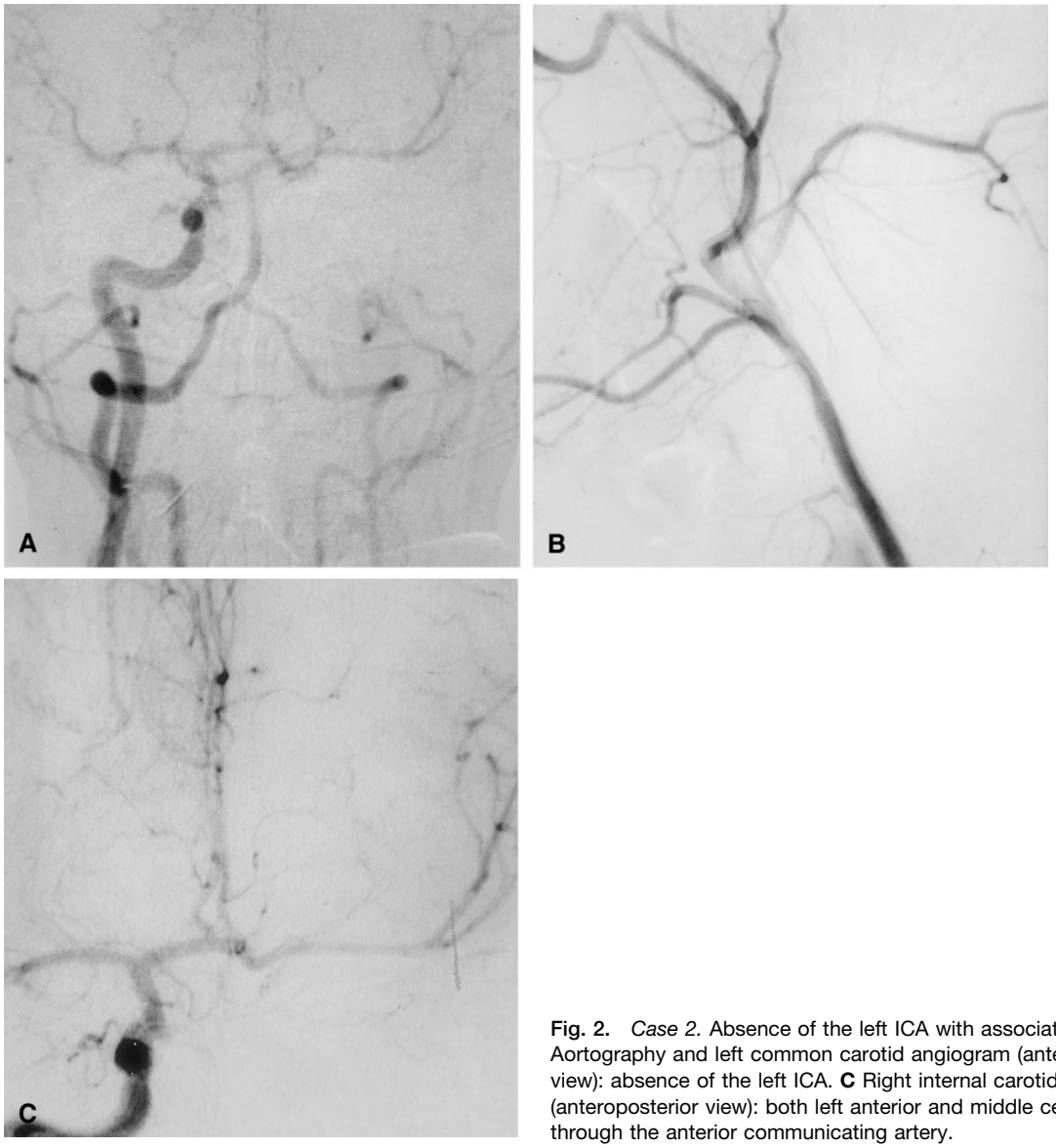


Fig. 2. *Case 2.* Absence of the left ICA with associated astrocytoma. **A, B** Aortography and left common carotid angiogram (anteroposterior and oblique view): absence of the left ICA. **C** Right internal carotid angiogram (anteroposterior view): both left anterior and middle cerebral arteries are supplied through the anterior communicating artery.

horizontal segment of carotid syphon (between the primitive maxillary artery and dorsal ophthalmic artery); 6, clinoid segment (between the dorsal and ventral ophthalmic arteries); 7, terminal segment (between the primitive ophthalmic artery and anterior cerebral artery). These segments appear to be autonomous from the embryological point of view since each may show segmental agenesis. The various types of segmental hypoplasia or agenesis and the resulting collateral pathways can be predicted with this model (Fig. 4). Our cases 1 and 2 show agenesis of the terminal segment with the homolateral cerebral hemisphere supplied via the circle of Willis (Figs. 1, 2). Case 3 is an example of abnormal segment 5 with a rete mirabile between segments 5 and 6 supplied by branches of the internal maxillary artery that correspond to a remnant of the dorsal ophthalmic artery (Fig. 3).

The patient with an absent ICA may be completely asymptomatic due to collateral blood supply to the affected hemisphere. Symptoms may be due to cerebrovascular insufficiency and/or compression by enlarged collateral cerebral vessels. Patients may suffer from recurrent headache, blurred vision, cranial nerves palsy, and hemiparesis. An absent ICA may be diagnosed by conventional or digital subtraction angiography performed for other reasons, such as intracranial hemorrhage due to ruptured aneurysm.

According to the literature an increased occurrence of associated intracranial aneurysms is documented in these patients. The frequency of intracranial aneurysms in patients with a ''normally'' developed ICA is estimated at 2%–4%, while in patients with congenital absence of the ICA the frequency of associated aneurysms is in the 24%–34% range [2, 4–7]. The increased occurrence of intracranial aneurysm formation may be due to a pathological cerebral blood flow pattern related to collateral vessels or to a common congenital defect of the ICA and intracranial arteries. Sporadic cases of absent ICA have been reported associated with cerebral hemiatrophy, Klippel-Trenaunay syndrome, neurofibroma-

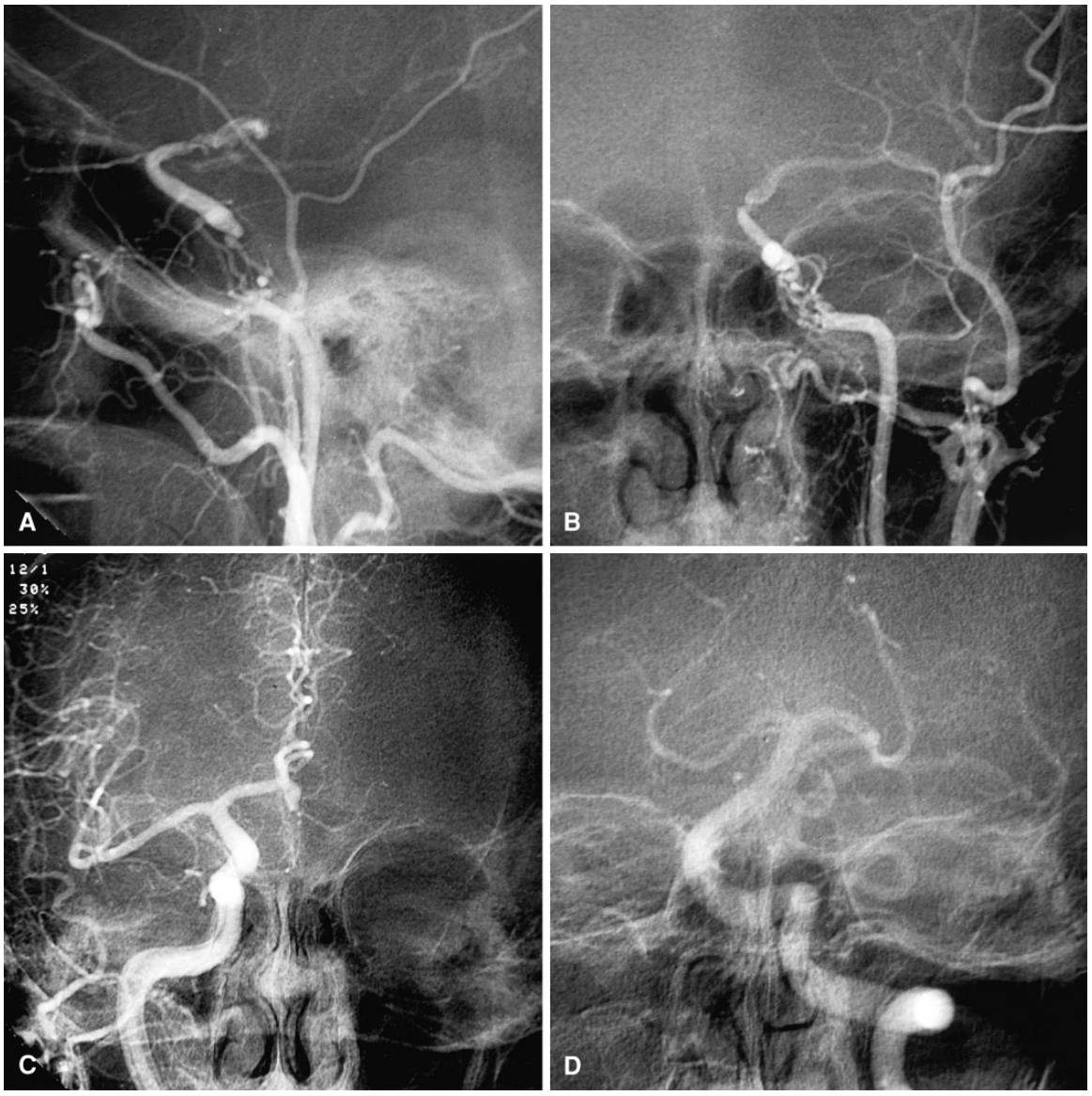


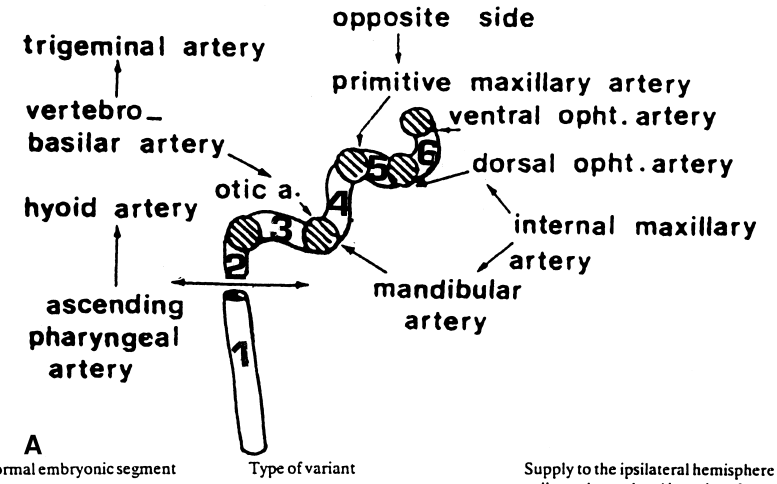
Fig. 3. *Case 3.* Hypoplastic left ICA with ''rete mirabile''. **A, B** Left common carotid angiogram (lateral and anteroposterior view): hypoplasia of the cervical portion of the left ICA while the absent petrosal portion of the ICA is filled by a ''rete mirabile'' of vessels. **C, D** Right common carotid and left vertebral angiogram (anteroposterior view): both left anterior and middle cerebral artery are supplied through the anterior communicating artery and through a collateral vessel arising directly from the posterior communicating artery.

tosis, coarctation of aorta, arachnoid cyst, and symptomatic epilepsy [4, 6, 8–12].

The diagnosis of congenital absence of the ICA can be made by skull base CT scan, MR angiography, conventional or digital subtraction angiography, at surgery or autopsy. The finding on a routine CT scan of an absent or hypoplastic carotid canal is one of the radiological methods of diagnosis of a congenitally anomalous ICA; in these patients, because of the documented high incidence of intracranial aneurysm formation, further evaluation is done with high-resolution skull base CT scan, and CT and MR angiography [13].

Conventional or digital subtraction angiography may be essential to evaluate the intracranial collateral vascular pattern, to confirm the MRI diagnosis of cerebral aneurysm, in difficult cases.

Diagnosis of absent ICA and of the cerebral vascular pattern is important as regards the clinical and surgical problems related to this condition. In these patients only one carotid artery supplies both cerebral hemispheres and thromboembolic neurological diseases can be explained by pathology of the contralateral carotid artery or the vertebrobasilar system. In some surgical procedures, such as carotid endarterectomy or surgery for intracranial aneurysm, there is an increased risk of cerebral ischemic complications that can be identified; in these patients prophylactic surgery such as extra-intracranial carotid by-pass could be considered before performing such procedures. In the



A

Abnormal embryonic segment	Type of variant	Supply to the ipsilateral hemisphere collateral vessel and its point of entry
Third aortic arch (segment No 1 and 2)	– Fenestration of the cervical ICA – Agenesis of the cervical ICA	Ascending pharyngeal artery ("intra tympanic course of the ICA"). Between segment 2 and 3
Dorsal aorta between the first and the second aortic arches (segment No 3)	– Agenesis of the cervical and ascending intra petrous portions of the ICA	Otic or mandibular arteries between segment 3 and 4
Dorsal aorta between the first aortic arch and the primitive maxillary artery (segment No 4)	– Agenesis of the cervical and petrous portion of the ICA	Trigeminal artery or contralateral siphon between segments 4 and 5
Dorsal aorta between the primitive maxillary artery and the dorsal ophthalmic artery (segment No 5)	– Agenesis of the cervical, petrous and cavernous portion of the ICA	Carotid rete mirabile between segment 5 and 6
Dorsal aorta between the dorsal and ventral ophthalmic arteries (segment No 6)	– No carotid visible	External carotid artery (internal maxillary branches) via the ophthalmic artery distal to the 6th segment
Dorsal aorta distal to the segment No 6	– Small carotid ending in the cavernous siphon by a persistent dorsal ophthalmic artery (infero-lateral trunk)	Circle of Willis

B

Fig. 4. Classification of segmental ICA agenesis proposed by Lasjaunias and Berenstein [3]. **A** Segment of the fully developed ICA system based on embryological criteria. **B** Table of the abnormal embryonic segment, type of variant, and supply to the ipsilateral collateral vessel. From Lasjaunias and Berenstein [3], pp 1–32.

transsphenoidal approch to the pituitary gland the surgeon should consider the possibility of encountering an intercavernous vessel in any patient with absence of the ICA. Moreover, these patients should be followed clinically and radiographically because of the documented increased frequency of intracranial aneurysms.

References

1. Padget DH (1948) The development of the cranial arteries in the human embryo. Contrib Embryol Carnegie Inst 32:205–261
2. Quint DJ, Boulos RS, Spera TD (1989) Congenital absence of the cervical and petrous internal carotid artery with intercavernous anastomosis. AJNR 10:435–439
3. Lasjaunias P, Berenstein A (1996) Surgical Neuroangiography, vol 1. Springer, Berlin, Heidelberg, New York
4. Teal JS, Rumbaugh CL, Bergeron RT, Segall HD (1973) Congenital absence of the internal carotid artery associated with cerebral hemiatrophy, absence of the external artery, and persistence of the stapedial artery. AJR 118:534–544
5. Servo A (1977) Agenesis of the left internal carotid artery associated with an aneurysm on the right carotid syphon: Case report. J Neurosurg 46:677–680
6. Afifi AK, Godersky JC, Menezes A, Smoker WR, Bell WE, Jacoby CG (1987) Cerebral hemiatrophy, hypoplasia of the internal carotid artery, and intracranial aneurysm: A rare association occurring in an infant. Arch Neurol 44:232–235
7. Waga S, Okada M, Kojima T (1978) Saccular aneurysm associated with absence of the left cervical carotid arteries. Neurosurgery 3:208–212
8. Goldestein SJ, Lee C, Young AB (1984) Aplasia of the cervical internal carotid artery and malformation of the circle of Willis associated with Klippel-Trenaunay syndrome: Case report. J Neurosurg 61:786–789
9. Chen MC, Liu HM, Huang KM (1994) Agenesis of the internal carotid artery associated with neurofibromatosis type II: Report of a case. AJNR 15:1184–1186
10. Verdeille S, Tonnelier M, Ghossain MA (1988) Agenesis of the left internal carotid artery associated with coarctation of the aorta: Report of an unusual case. Cardiovasc Intervent Radiol 11:325–328
11. Kidooka M, Okada T, Hanada J (1992) Agenesis of the internal carotid artery: Report of a case combined with arachnoid cyst in a child. No To Shinkei 44:371–375
12. Wang PJ, Liu HM, Young C, Lin MY, Shen YZ (1994) Agenesis of internal carotid artery associated with symptomatic partial epilepsy. Epilepsia 35:1337–1341
13. Quint DJ, Silbergleit R, Young WC (1992) Absence of the carotid canals at skull base CT. Radiology 182:477–481