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Radiologic anatomy of segmental agenesis of the internal carotid artery

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Summary: We report six cases of segmental agenesis of the internal carotid a. discovered in adult patients: one case of cervical segmental agenesis (no. 1), one case of cervical and petrosal segmental agenesis (no. 2), two cases of vertical cavernous segmental agenesis (nos. 3 and 4) and two cases of distal segmental agenesis, one unilateral (no. 5) and the other bilateral (no. 6). The collateral pathways observed were: the ascending pharyngeal a. which constitutes an "intratympanic course" of the internal carotid a. (no. 1), an intercarotid anastomosis (no. 2), an arterial network at the base of the skull, the so-called "rete mirabile" (nos. 3 and 4) and the posterior communicating a. (nos. 5 and 6). Recognition of these rare dysgeneses relies upon the following radio-anatomic characteristics: reduced caliber of the a., inconsistent sparing of the carotid body, reduced diameter or even absence of the carotid canal and above all, the presence of collateral pathways. The collateral pathways allow an understanding of the segmental nature of carotid a. development and a distinction between congenital and acquired stenoses.

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Anatomie radiologique des agénésies segmentaires de l'artère carotide interne

Résumé : Six observations d'agénésies segmentaires de l'artère carotide interne, découvertes chez l'adulte, sont rapportées : un cas d'agénésie du segment cervical (n° 1), un cas d'agénésie des segments cervical et pétreux (n° 2), deux cas d'agénésie du segment caverneux vertical (n° 3 et 4) et deux cas d'agénésie distale de l'artère, unilatérale (n° 5) et bilatérale (n° 6). Les voies de suppléance observées sont : l'artère pharyngienne ascendante réalisant un "trajet intratympanique" de l'artère carotide interne (n° 1), une anastomose intercarotidienne (n° 2), un réseau artériel de la base du crâne, dit "réseau admirable" (n° 3 et 4) et l'artère communicante postérieure (n° 5 et 6). La reconnaissance de ces rares dysgénésies repose sur les caractéristiques radio-anatomiques suivantes : réduction de calibre de l'artère, respect inconstant de sa portion bulbaire, réduction des dimensions voire absence du canal carotidien et surtout voies anastomotiques utilisées. Les anastomoses observées permettent en effet de comprendre le caractère segmentaire du développement de l'artère et de distinguer les sténoses congénitales et les sténoses acquises.

Key words: Internal carotid a. – Agenesis – Carotid canal – Rete mirabile – Anastomoses

Among the various forms of dysgenesis of the internal carotid a. (ICA), Lie distinguishes hypoplasia, aplasia, and agenesis, which is the complete absence of an artery [43]. Ageneses of the ICA are considered to be rare, but because they are often clinically silent it is difficult to estimate their actual incidence. Even major dysgenesis such as bilateral absence of the ICAs is not a direct cause of brain damage. Lajaunias' works have well demonstrated that the ICA is formed by the union of distinct segments which have a defined course and whose limits are determined by the origin of the embryonic blood-vessels [37, 38]. Segmental ageneses of the ICA have been described, and correspond to a developmental disorder of one or several segments of the artery. In such cases, arterial blood flow reaches the ICA after having been deviated through persistent embryonic blood-vessels or through the posterior communicating a. The six case reports reviewed in this study serve as the basis of discussion of the radiologic anatomy of agenesis and to highlight the large variety of possible collateral pathways which can develop in response to such developmental disorders of the ICA.

Case reports

Table 1 summarizes the results of the angiographic studies performed in each of the cases.

Table 1. Results of angiograms

Case		Studied vessels	Internal carotid artery		Carotid canal	Collateral pathways	Associated vascular anomalies
			Initial segment	Side			
No. 1	F, 35 years	R + L CCA L VA	--	R	Absence of vertical portion	APhA	Agenesis R A1
No. 2	M, 51 years	R + L CCA R + L VA	--	R	Not visualized (XR)	Intercarotid anastomostic a.	Agenesis R A CCA from IA
No. 3	M, 56 years	R + L CCA R VA	Not dilated	L	Reduced	RM	Agenesis R PCoA Hypoplasia R VA
No. 4	F, 62 years	R CCA R VA	Not dilated	R	Reduced	RM	Non opacification of R A1 and R PCoA R megadolicho-VA
No. 5	F, 52 years	R CCA R + L VA	Dilated	L	Reduced	ACoA PCoA	
No. 6	F, 58 years	R + L CCA L VA	Dilated	R + L	Reduced	PCoA	L mega-VA Persistance of L dOA

LR: Left-Right. *CCA:* Common carotid a. *VA:* Vertebral a. *RM:* Rete mirabile. *A and PCoA:* Anterior and posterior communicating a. *IA:* Innominate a. *XR:* Plain films. *A1:* First segment of the anterior cerebral a. *A PhA:* Ascending pharyngeal a. *dOA:* Dorsal ophthalmic a.

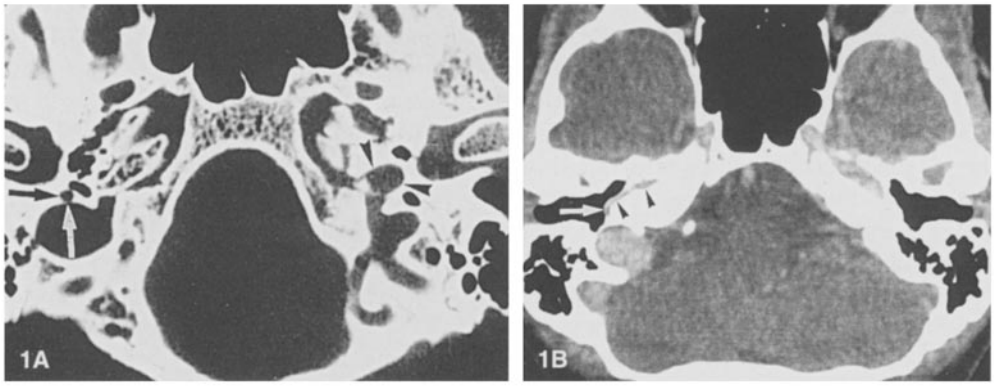


Fig. 1A, B
Agenesis of the cervical segment of the right internal carotid a. (case n° 1). CT scan of the skull base. **A** Axial slice through the lower portion of the petrosa: absence of the extracranial orifice of the right internal carotid canal; enlargement of the tympanic canal (*arrows*). Extracranial orifice of the left carotid canal with a normal diameter (*arrowhead*). **B** Axial slice, after contrast medium injection, through the tympanic cavity: presence of a vascular density structure, located inside the tympanic cavity (*arrow*), continuous with the horizontal portion of the petrosal internal carotid a. (*arrowheads*), corresponding to anastomosis between the inferior tympanic a., which is a branch of the ascending pharyngeal a., and the carotico-tympanic a., which is a branch of the internal carotid a.

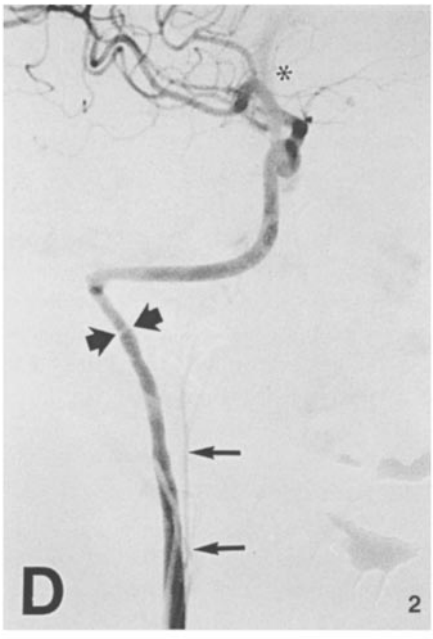


Fig. 2
Agenesis of the cervical segment of the right internal carotid a. (case no. 1). Injection of the common carotid a.: A-P view with head turned to the left. "Carotid flow" through the inferior tympanic, branch of the ascending pharyngeal a.; stenosis of this same a. at the entry point of the inferior tympanic canal (*large arrows*); injection of the mucosal branches of the ascending pharyngeal a. (*arrows*). Note the absence of injection of the precommunicating segment of the anterior cerebral a. (*asterisk*)

Case no° 1 (1991). Agenesis of the cervical segment of the right ICA

A 35-year-old woman, complained of pulsatile tinnitus which increased during physical activity. CT scan of the skull base showed the characteristic elements of an intratympanic aberrant right ICA: absence of the extracranial orifice and the vertical segment of the right carotid canal; enlargement of the inferior tympanic canal, with the presence of an intratympanic structure of vascular density in communication with the horizontal petrosal portion of the ICA (Fig. 1). Angiographic study included injection of both common carotid aa. and the left vertebral a. The diagnosis was confirmed and showed an a. which gave rise to the mucosal branches of the ascending pharyngeal a. (Fig. 2) and whose cervical and vertical petrous portions were more

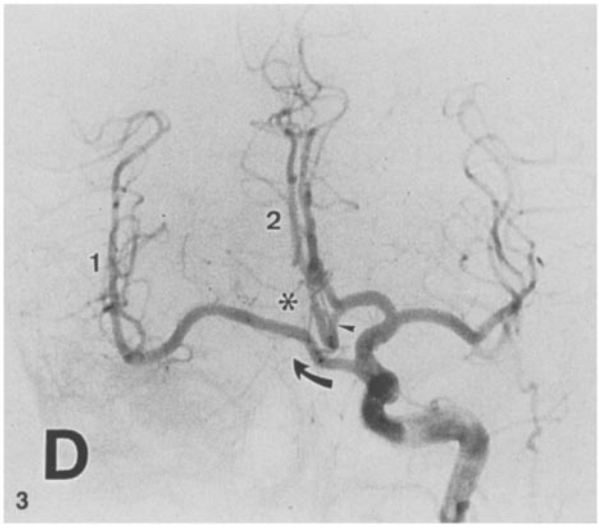


Fig. 3
Agenesis of the cervical and petrosal segments of the right internal carotid a. (case no. 2). Left carotid angiogram, A-P view. Through an intercarotid anastomosis projecting on the sellar region (*curved arrow*), injection of the right carotid flexure and of the right middle cerebral a. (*1*). Agenesis of the precommunicating segment of the right anterior cerebral a. (*asterisk*). Anterior communicating a. (*arrowhead*) allowing injection of the right anterior cerebral a. (*2*)

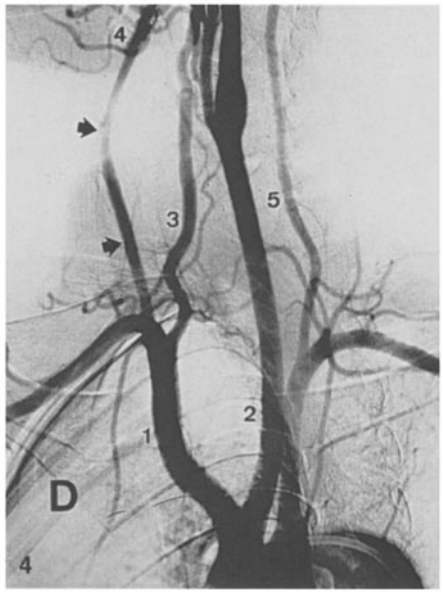
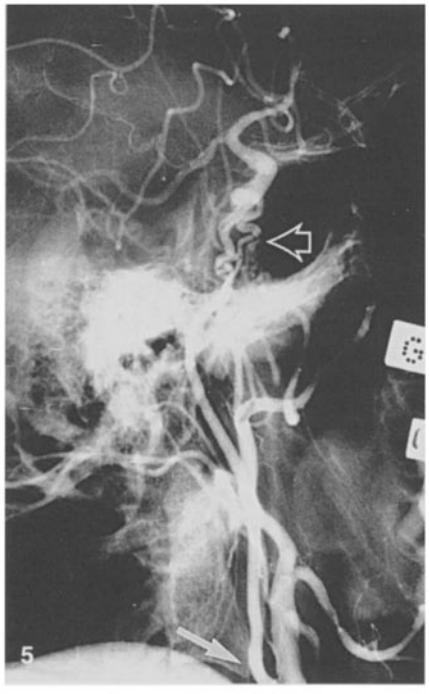


Fig. 4
Agenesis of the cervical and petrosal segments of the right internal carotid a. (case no. 2). Global injection of the aortic arch and its great vessels, left oblique-anterior view. Innominate a. (*1*), giving rise to the left (*2*) and right (*arrows*) common carotid aa. Right common carotid a. terminating in a right external carotid a. (*4*). Right (*3*) and left (5) vertebral aa.

laterally located than usual for the ICA on an A-P view. Furthermore, there was agenesis of the basal horizontal portion of the right anterior cerebral a., also called the precommunicating segment. The left common carotid and internal carotid aa. as well as the left vertebral a. presented no particularities.

Case report no° 2 (1979).
Agenesis of the cervical and petrous segments of the right ICA

A 51-year-old man was hospitalized for investigation of an acute hemiparesis caused by stroke. No blood-flow was found in the right ICA on doppler ultrasound examination of the vessels of the neck. Plain films of the skull did not show the right carotid canal on the A-P view and the sella turcica appeared normal on the lateral view. Angiographic studies included injection of the common carotid a. by direct puncture (Fig. 3) and of the aortic arch through a femoral approach (Fig. 4). Examination results showed the left common carotid a. originating from the innominate a., absence of injection of the cervical and petrosal segments of the right ICA, and the existence of an intercarotid anastomosis located in the coronal plane uniting the carotid flexures of both ICAs. The precommunicating segment of the right anterior cerebral a. was not injected.



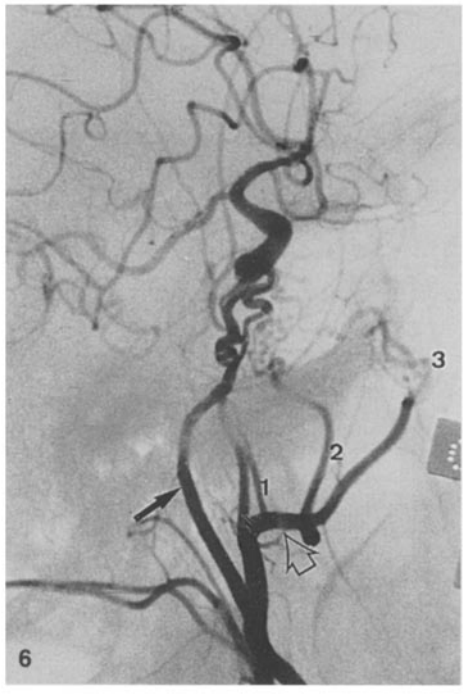


Fig. 5
Agenesis of the vertical cavernous segment of the left internal carotid a. (case no. 3). Injection of the left common carotid a., lateral view. Reduced caliber of the internal carotid a. without carotid body dilatation (*arrow*). Juxtasellar collateral arterial network (*empty arrow*)

Fig. 6
Case no. 3. Injection of the left common carotid a., lateral view, subtracted and shifted films allow stereoscopic view. Internal carotid a. (*arrow*), maxillary a. (*arrowhead*), middle meningeal a. (*1*), accessory meningeal a. (*2*), a. of the foramen rotondum (*3*)

Case report no° 3 (1988).
Agenesis of the vertical cavernous segment of the left ICA

A 56-year-old man was admitted for investigation of a left cerebral hemis-

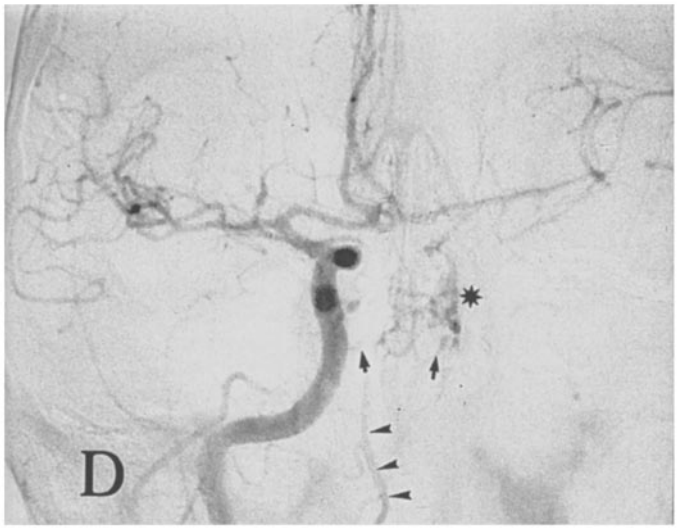
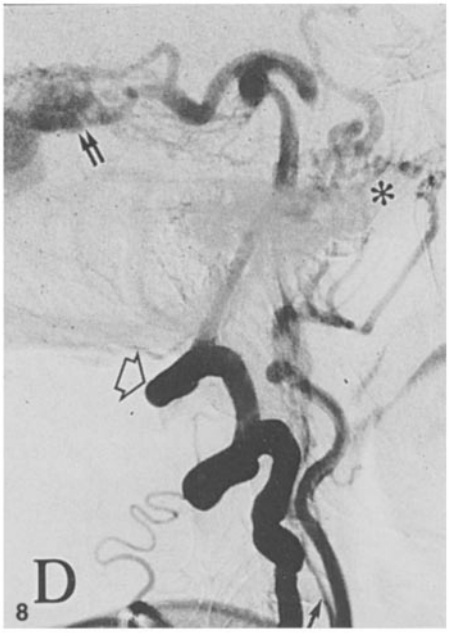


Fig. 7
Case n° 3. Direct brachial a. puncture: A-P view. Injection of the left arterial network (*star*) through the medial clival aa., stemming from the right carotid flexure (*arrows*). Note right vertebral a. hypoplasia (*arrowheads*)



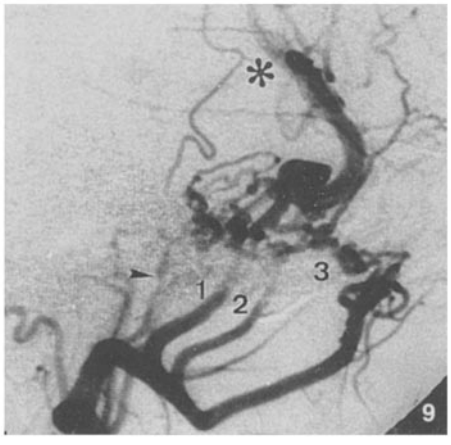


Fig. 9
Case no. 4. Selective injection of the right external carotid a. Juxtasellar arterial network fed by branches of the maxillary and ascending pharyngeal aa. (*arrowhead*). Note the absence of injection of the posterior communicating a. (*asterisk*) and of the anterior cerebral a. Middle meningeal a. (*1*). Accessory meningeal a. (*2*). A. of the foramen rotundum (*3*)

Fig. 8
Agenesis of the vertical cavernous segment of the right internal carotid a. (case no. 4). Direct right brachial a. puncture, lateral view. Reduced caliber of the right internal carotid a. (*arrow*). Right megadolichovertebral a. (*empty arrow*). Arterial network of the skull base fed by branches of the external carotid a. (*star*). Temporo-occipital arteriovenous malformation (*double arrow*)

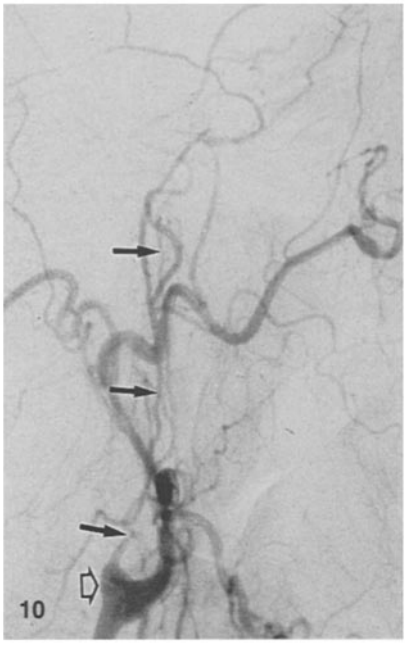
phere tumoral mass. CT study revealed a small left carotid canal. Angiography performed by direct puncture showed a reduced left ICA lumen with no irregularities in its cervical and petrosal course (Fig. 5). As it exited the carotid canal it united with an arterial network fed by branches of the maxillary a. and drained by the distal cavernous portion of the ICA (Fig. 6). The left posterior communicating a. was not injected. The study was completed by injection of the right common carotid and vertebral aa. by puncture of the right brachial a. and showed a hypoplastic right vertebral a., a patent anterior communicating a. and dilated medial clival branches stemming from the right carotid flexure which fed into the arterial network (Fig. 7).

Case report no° 4 (1988). Agenesis of the right vertical cavernous segment of the right ICA

A 62-year-old woman with no prior medical history, was hospitalized for an intraparenchymal cerebral hematoma with intraventricular hemorrhage. The initial angiographic study, performed by puncture of the right brachial a., revealed a right parieto-occipital arteriovenous malformation. The right vertebral a. was dilated and tortuous and the right ICA presented a reduced lumen in its cervical and petrous segments with no irregularities visible between the carotid body and the cervical portion farther downstream (Fig. 8). A juxtasellar arterial network was fed by branches of the maxillary a. which was later confirmed by superselective catheterization (Fig. 9). The right anterior cerebral and posterior communicating aa. were absent. The ophthalmic a., which was poorly injected, originated from the supracavernous segment of the ICA. CT showed a reduced right carotid canal.

Case report no° 5 (1986). Agenesis of the distal segment of the left ICA

A 52-year-old woman was hospitalized for subarachnoid hemorrhage. Angiographic study, performed upon admission, showed irregular luminal stenosis of the basilar a. This stenosis was not found at follow-up studies performed one month later and the diagnosis of basilar a. dissection was made. The left ICA presented an initial normal segment which became tapered and disappeared in the cavernous region (Fig. 10). The vertebro-basilar system partially fed the left middle cerebral a. via the posterior communicating a. The remainder of the supply to the left middle cerebral a. and all of the supply to the left anterior cerebral a. were provided by the right ICA via the anterior communicating a. A final routine angiogram performed 8 years later showed no change in the cranio-cerebral blood supply. CT scan of the skull base shows a marked asymmetry of the carotid canals (Fig. 11).



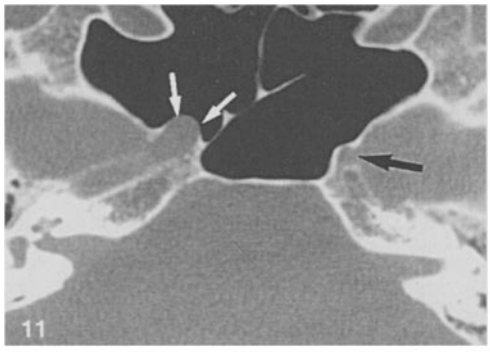


Fig. 10
Distal segmental agenesis of the left internal carotid a. (case no. 5). Injection of the left common carotid a. (*arrow*), lateral view. Apparent narrowing of left internal carotid a. beyond the carotid body showing normal caliber (*empty arrow*)

Fig. 11
Case n° 5. CT exam through the superior portion of the carotid canal. Marked reduction of caliber of the left carotid canal (*single arrow*) in comparison to the right carotid canal (*arrows*)

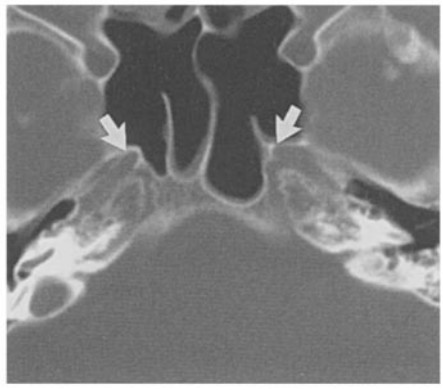
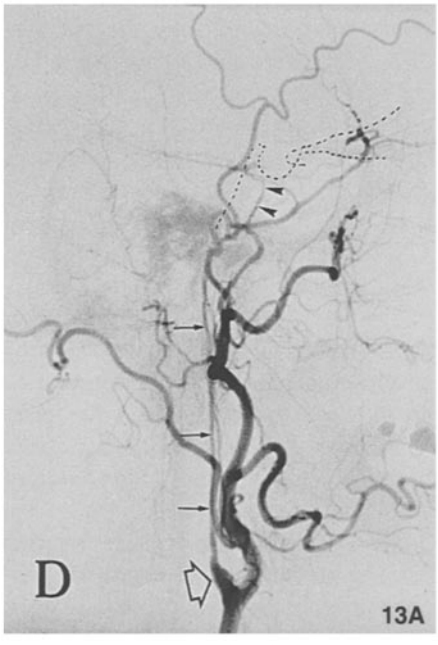
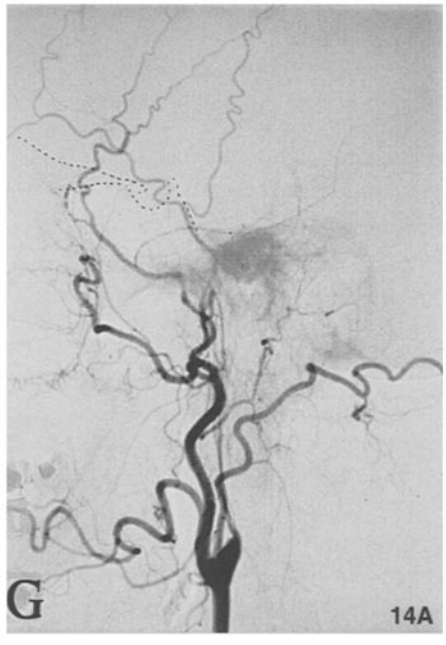


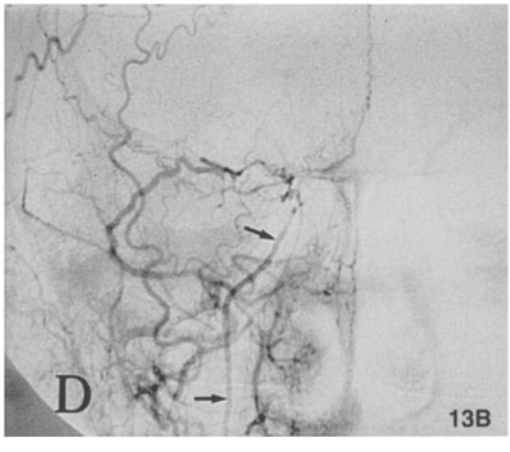
Fig. 12
Bilateral distal agenesis (case no. 6). High-resolution CT view using bone window, axial slice through the horizontal segment of the carotid canal. Reduced caliber of both carotid canals (*arrows*)

Case report no° 6 (1989).
Bilateral agenesis of the distal segments of the ICA

A 58-year-old woman treated for high blood pressure, was admitted for investigation of a parieto-rolandic hematoma which had occured three months earlier. Doppler ultrasound study of the great vessels of the neck diagnosed bilateral occlusion of the internal carotid aa. compensated by the vertebro-basilar system. CT scan showed a reduction in the size of the carotid canals involving the vertical as well as the horizontal portions (Fig. 12). Angiographic studies included injection of both common carotid aa. and the left vertebral a. Both ICAs were normal at their origin and suddenly narrowed and became tapered. In the anterio-posterior view they seemed to be more medially located than is usually the case in their course through the skull base







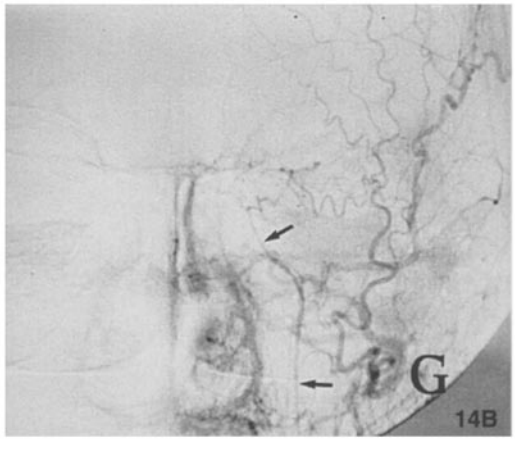


Fig. 13A, B
Case n° 6. Injection of the right common carotid a. **A** Lateral view cervical and petrosal segments of the narrowed internal carotid a. in contrast to the carotid body showing normal caliber (*empty arrow*). Internal carotid a. terminates in cavernous branches (*arrowheads*). **B** A-P view. Narrowed internal carotid a. (*arrows*)

Fig. 14A, B
Case n° 6. Injection of the left common carotid a. Lateral (**A**) and A-P views (**B**). Same appearance as on the right

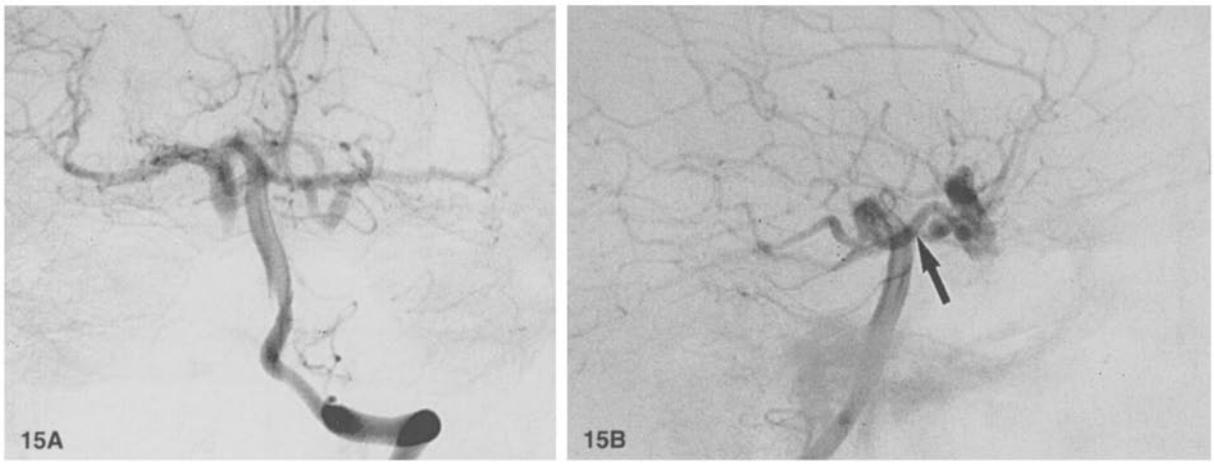


Fig. 15A, B Case n° 6. Injection of the left vertebral a. A-P (**A**) and lateral views (**B**). Dilated and tortuous posterior communicating aa. supplying the carotid territories of the brain

(Figs. 13 and 14). In both cases they terminated in the lower portion of the cavernous sinus. A primitive dorsal ophthalmic a. was present on the left. The left vertebral a. and the basilar a. were both enlarged. The large and serpentine posterior communicating aa. provided blood-flow to both carotid systems (Fig. 15).

Discussion

A. Classification of dysgeneses

Carotid dysgenesis has been classed by Lie into 3 groups: agenesis, aplasia and hypoplasia [43]. In cases of agenesis or complete absence of the blood-vessel, the carotid canal as well as the branches of the external carotid a., derived from the stapedial a. are absent. In case of aplasia, the ICA is present in a vestigial form and both the carotid canal and stapedial branches may be seen. Finally, in hypoplasia, the carotid a. presents a reduced luminal diameter but remains patent throughout its course. The practical interest of this classification is not obvious since the distinction between agenesis and aplasia is difficult and they are consequently frequently confused in the literature. Furthermore, the term "carotid hypoplasia" is sometimes used in the literature to describe a carotid a. of reduced lumen diameter without considering its terminal branch pattern [6, 16, 17, 24, 32, 42, 46, 52, 63]. The diagnosis of hypoplasia of the ICA, however, can only be made if the latter conserves its usual terminal branching pattern. As illustrated by comparative anatomic studies, this usual terminal branching pattern consists of one posterior branch, which becomes the posterior communicating a. and one anterior branch which develops into the anterior choroidal, anterior cerebral and middle cerebral aa. [37]. In some cases of ICA "hypoplasia" the artery is interrupted as in cases n° 5 and 6, before its usual terminal branching pattern [16, 17, 24]. In such cases the dysgenetic process should not be considered as hypoplasia, but should refer to the absence of a distal portion of the ICA [38]. The segmental agenesis of an artery as well as stenosis or acquired occlusion all lead to a reduction in the diameter of the blood-vessel downstream. As an example, an anomaly in the structure of the circle of Willis which leads to a reduction in the usual territory of the ICA may lead to a reduced vessel diameter [23]. A recent MRI study confirms this concept [31]. The ICA was measured in 104 patients. It was 4.62 ± 0.68 mm in the general population and 3.63 ± 0.68 mm in cases of agenesis of the precommunicating segment of the ipsilateral anterior cerebral a.

Data concerning embryologic development provide an understanding of segmental agenesis of the ICA [38, 54]. At the initial stage of embryonic development (embryonic length of 3 mm) the following blood-vessels are found at the cranial extremity: the cranial portion of the dorsal and ventral aorta, united by the aortic arches; the primitive maxillary aa., the dorsal and ventral ophthalmic aa. (Fig. 16). Modifications of this scheme during development include regression of the initial portion of the ventral ophtalmic a. originating from the anterior cerebral a. and of the ventral portions of the two first aortic arches and of the dorsal aorta located below the third aortic arch. The entire carotid system is thereby established: common carotid a., originating from the ventral aorta, located below the third aortic arch, the ventral pharyngeal a. which constitutes the future external carotid a. and which originates from the ventral aorta located above the third aortic arch, and finally the internal carotid a. The latter stems from the third aortic arch (segment 1) and from the dorsal aorta located between the third and second aortic arches (segment 2), the second and first aortic arches (segment 3), the first aortic arch and the primitive maxillary a. (segment 4), the primitive maxillary a. and the dorsal ophthalmic a. (segment 5), the dorsal and primitive ophthalmic aa. which stem from the ventral ophthalmic a. (segment 6), the primitive ophthalmic and the anterior cerebral aa. (segment 7). Figure 17 illustrates the correspondence between these embryonic segments and the mature ICA. The limits of these defined segments correspond to the origins of the following embryonic vessels: hyostapedial a. (2nd aortic arch), mandibular a. (dorsal remnant of the 1st aortic arch), primitive maxillary a. which gives rise to the posterior hypophyseal aa., the dorsal ophthalmic a. which gives rise to intracavernous branches of the ICA also known as the infero-lateral trunk, and finally the ventral ophthalmic a. Any one of these segments, independent from an embryologic view point, may be missing, resulting in segmental agenesis of the ICA [38].

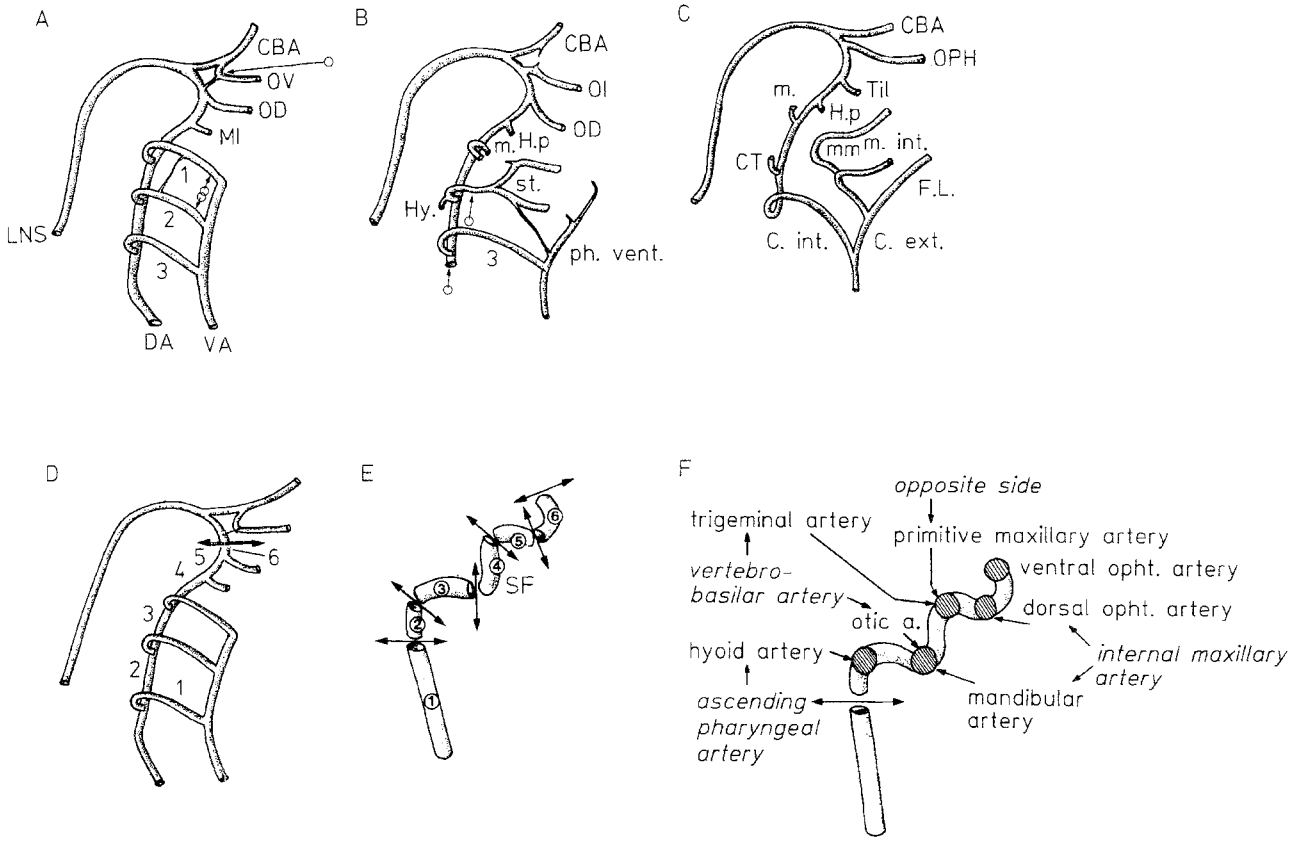


Fig. 16A-F
Successive stages of embryonic vascular development in the cephalic region. *LNS:* longitudinal neural system. *CBA:* anterior cerebral a. *OV:* ventral ophthalmic a. *OD:* dorsal ophthalmic a. *MI:* primitive maxillary a. *OI:* primitive ophthalmic a. *H.p:* postero-inferior hypophyseal a. *m.:* mandibular a. *Hy.:* hyoid a. *ph. vent.:* ventral pharyngeal a. *st.:* stapedial a. *OPH:* definitive ophthalmic a. *Til:* infero-lateral trunk. *mm:* middle meningeal a. *FL:* facio-lingual system. *CT:* carotico-tympanic a. *m. int:* maxillary a. *C. int:* internal carotid a. *C. ext:* external carotid a. *1, 2, 3:* 1st, 2nd and 3rd aortic arches. (With the permission of P. Lasjaunias and A. Santoyo-Vazquez (1984) Anat Clin 6: 133-141)

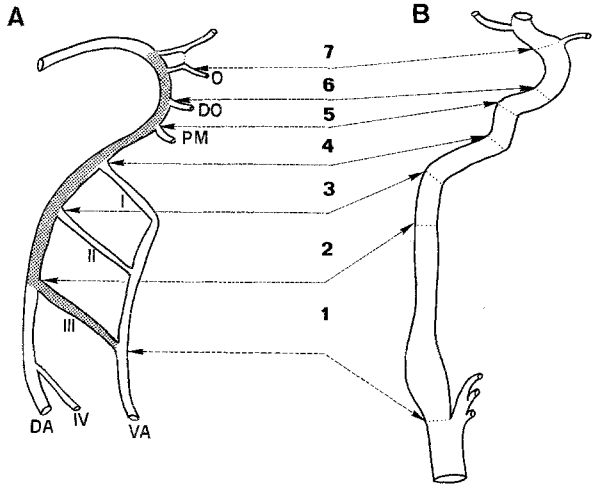


Fig. 17A, B
Diagram of the correspondence between the embryonic (**A**) and definitive (**B**) structure of the internal carotid a. (P Lasjaunias). The different segments of the internal carotid a. appear shaded on the diagram of the embryonic structure and are numbered from *1* to *7*. *O:* definitive ophthalmic a., stemming from the ventral ophthalmic a. *DO:* dorsal ophthalmic a. *PM:* primitive maxillary a., future infero-lateral trunk. *I, II, III, IV:* 1st, 2nd, 3rd and 4th aortic arches. *DA:* dorsal aorta. *VA:* ventral aorta

B. Prevalence and discovery mode of ageneses

The various types of agenesis of the ICA are rare as can be seen from Teal's review of the literature which found only 9 complete and bilateral ageneses and 34 unilateral cases [67]. More recently, Midkiff et al. [47] have estimated the number of unilateral ageneses reported in the English language literature to be less than 80 since the first description by Tode in 1787. The actual frequency of carotid ageneses remains difficult to establish for several reasons: they are often clinically silent; it is difficult to distinguish them from stenotic disease, particularly prior to the appearance of modern imaging techniques; and finally, the dysgenetic pathophysiology is sometimes erroneously interpreted.

Carotid ageneses are essentially discovered in adult patients, pediatric cases being exceptional [1]. The adaptive capacities of the arterial network explains why such variations are most often clinically silent. The circumstances which usually lead to their discovery are stroke [6, 48, 68] or, as in our 6th case report, intracranial hemorrhage. The development of arterial intracranial arterial aneurysms has been attributed to the hemodynamic stress to which the collateral vascular structures are submitted [1, 4, 11, 13, 14, 16, 28, 35, 47, 51, 55, 59, 61, 65, 71] or to the morphologic modifications

of these collaterals as observed in case report n° 6 [20, 32, 60].

C. Mechanism of formation

Histopathologic studies of dysgenetic vessels show subendothelial fibrosis or atheromatosis, which could be mistaken for acquired lesions. A number of arguments, however, are in favour of the congenital nature of this condition. Among the intracranial segmental lesions, none was observed to be related to arteritis or meningitis [24]. The reduced caliber of the arteries involved not only their internal diameter but their external diameter as well [24, 49, 62]. The anomaly may be bilateral and symmetric, or associated with such vascular anomalies as the persistence of a stapedial a. [67], absence of the external carotid a. [67], heart [3] or kidney malformations [27], Apert's syndrome [3] or Von Recklinghausen's disease [12]. Austin and Stears have even introduced the notion of genetic transmission [6] in their observation of bilateral hypoplasia of the ICA involving two brothers.

The mechanisms of formation involved in agenesis of the ICA remains controversial. It may result from either regression at a late stage after an initial period of normal development or, more probably, from an early interruption of development. For Hills and Sament, disturbance of the ICA occurs at an early stage of development, at the 3 to 5 mm stage, as can be illustrated by its association with other major developmental anomalies [27].

D. Radiologic anatomy

The diagnosis of agenesis of the ICA relies exclusively on the results of radiologic studies: high-resolution CT of the skull base to study the carotid canal, and digital subtraction angiography or magnetic resonance angiography to determine the location and extent of the dysgenesis as well as the type of collateral supply.

1. Morphology of the internal carotid a.

a. *Reduced caliber of the internal carotid a.* A reduction in the caliber of the ICA is constantly found in cases of agenesis of its distal segments. In the literature, however, there are no data which define the limiting values between a "normal" carotid a. and a dysgenetic form. Patay and Berky consider hypoplasia to be certain if the internal diameter of the ICA does not exceed 3.5mm along its entire length or along most of its course [56]. This limiting value is in agreement with the data of anatomic and radio-anatomic studies of the carotid canal [34, 40]. Despite these facts, it is difficult to accept this value as an absolute limit as Kane et al. have shown in their study [31]. Furthermore, while this limit is necessary to make the diagnosis of hypoplasia, it is not in itself sufficient to consider an anomaly as being of congenital origin since a reduction of arterial diameter can be observed following an acquired occlusion.

b. *Sparing of the initial portion of the internal carotid a.* Non involvement of the first few cm of the ICA, which corresponds to the carotid body, as in our cases n° 5 and 6 as well as in several reported cases in the literature [10, 22, 42, 46, 53, 63], has been considered by certain authors as characteristic of dysgeneses of the ICA [16, 24]. It is thought either to correspond to a prestenotic dilation or to indicate the embryologic origin of the initial segment of the ICA, which is different from the rest. According to the latter theory, the carotid body, which develops from the 3rd aortic arch, could be spared by the dysgenetic process, which involves the segments of the ICA, derived from the dorsal aorta. Another explanation for the non-involvement of the initial portion of the ICA may be related to the nature of the carotid body itself, whose physiologic function differentiates it from the rest of the ICA. Whatever may be the explanation for the dilatation of the carotid body, its inconsistency does not authorize its use in the diagnosis of carotid dysgenesis.

2. Carotid canal

Differentiating between congenital and acquired stenosis of the ICA is sometimes difficult. The radiologic study of the skull base provides an essential argument in favour of the diagnosis of a congenital condition: the reduced diameter or even complete absence of the carotid canal [21, 25, 58, 59, 66, 74]. Based on the anatomic study of ten petrosae, Leonetti et al. considered the upper and lower limits of the carotid canal to be 4 to 7.5 mm for the vertical portion and 4.5 to 7 mm for the horizontal portion [40]. In another similar study performed using CT scan, these upper and lower limits were found to be 5.2 and 9.8 mm for the diameter of the carotid canal at the junction between the petrous and cavernous portions [34]. Comparison of the two carotid canals may be useful in cases of unilateral involvement. In a CT study comparing a unilateral dysgenetic group of patients to a control group, the ratio of the greater to the lesser diameter was greater than 2 in case of unilateral hypoplasia, while it never exceeded 1.2 in the second group [34].

Complete absence of the canal is found in cases of total agenesis of the internal carotid a. and a reduction in its caliber is found in cases of distal segmental agenesis or hypoplasia. Only the vertical portion of the canal is absent in cases of cervical segmental agenesis of the ICA (case no. 1).

3. Collateral pathways

Depending upon the location of agenesis, either embryonic aa. or the posterior communicating aa. are used as collateral pathways. Lasjaunias has shown that the embryonic blood-vessel located immediately downstream from the missing segment is incapable of providing the collateral blood supply and it is therefore the following vessel which does so [38]. Two other types of collaterals may be exceptionally observed: the collateral network at the skull base or the calvarium [6, 46], which calls upon transarachnoid branches of the meningeal aa., and the pseudo-angiomatous network of the basal ganglia [9, 63]. In this chapter we will analyze only those collateral pathways found in our observations.

a. *"Tympanic" anastomosis.* Agenesis of the cervical segment of the ICA corresponds to absence of the segment which derives from the 3rd aortic arch. As illus-

trated in our case no. 1, the inferior tympanic a., which is a branch of the ascending pharyngeal a., is used as the collateral pathway [37, 50]. The carotid blood flow is deviated through the inferior tympanic a., which feeds into the caroticotympanic a., stemming from the proximal portion of the hyostapedial a. within the tympanic cavity. Beyond this anastomosis, the carotid blood flow resumes its usual course in the horizontal segment of the carotid canal. Proximal agenesis of the ICA and the collateral pathway which develops explains the bone anomalies we observe: absence of the extracranial orifice of the carotid canal and atresia of its vertical portion, enlargement of the inferior tympanic canal, normal appearance of the horizontal segment of the carotid canal except for the disappearance of its posterior cortical wall at the point of entry of the a. [19, 36, 44, 70, 73].

b. *Inter-carotid anastomosis.* In cases of agenesis involving segment 3 of the ICA, which originates from the dorsal aorta located between the 1st and 2nd aortic arches, the collateral pathway uses a branch stemming from the primitive maxillary a. It may correspond either to a trigeminal a. or, as in our case no. 2, an artery uniting the two carotid flexures [2, 15, 18, 26, 28, 29, 33, 55, 64, 69]. These intercavernous communications may have various courses such as transsphenoidal or intrasellar. In the latter case, it may deform the floor of the sella turcica.

c. *"Rete mirabile".* In cases of agenesis of segment 4 of the ICA, the collateral pathway used is a juxtasellar arterial network which feeds into the ICA via the infero-lateral trunk, stemming from the dorsal aorta [38]. Its close resemblance to the "rete mirabile" described in certain animals such as the cat, sheep, ox and pig, have raised the question of its existence in man [5, 13, 30, 41, 49, 53, 57]. Contrary to the arterial network found in animals, which appears late in fetal development and continues after birth, the network found in man is not a developmental anomaly which appears late in fetal life, but rather a well-known system of cavernous anastomoses. This anastomotic system uses branches of the maxillary a. (middle meningeal a., accessory meningeal a. and a. of the foramen rotundum) and the ascending pharyngeal a. [8, 39, 45, 72].

d. *Posterior communicating a.* The use of the posterior communicating aa. as collateral pathways is observed when agenesis involves or extends to the distal segment of the internal carotid a. [1, 7, 10, 16, 24, 25, 32, 46, 52, 60, 66, 61, 65, 71, 74]. The communicating aa. may be dilated and tortuous as in our case n° 3, and may even take on an aneurysmal appearance [20].

Conclusion

Ageneses of the ICA constitute rare dysgeneses which should nevertheless be recognized and distinguished from acquired arterial disease. As shown in our case reports, they are compatible with normal brain development and, in most cases, are not directly responsible for the clinical event which leads to their discovery. This last point highlights the great variety of collateral pathways, the choice of which can be explained by embryologic considerations. The ICA does, in effect, result from the development of different segments. Each of these segments is limited by embryonic vessels which may constitute the collateral pathways of carotid flow in case of agenesis of the ICA.

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