Persistence of the Stapedial Artery In A First Arch Anomaly: A Case Report

IGNACIO PASCUAL-CASTROVIEJO, M.D.
Madrid, Spain

This is a case report of a three month old girl with a unilateral facial dysplasia in whom the rudiments of external auditory organs were displaced towards the zygomatic region. The carotid arteriography demonstrated a persistent stapedial artery and absence of some branches of the external carotid artery (the middle meningeal artery and the superficial temporal artery). The autopsy study, undertaken at six months of age, confirmed the presence of the vascular anomaly and severe defects in the auditory organ. There was nerve tissue in the facial mass. This is the first reported case in which an association of a unilateral first arch anomaly and persistent stapedial artery has been confirmed by arteriography.

Persistent stapedial artery is a rare vascular anomaly which, like persistent trigeminal and hypoglossal arteries, is the presence during extrauterine life of vascular structures which are present in the early stages of embryonic development. The first case described in the literature was that of Hyrtl (1836). House et al. (1964) undertook a revision and found that only 17 cases had been described until then, all of them being anatomical observations. Since then, a few isolated cases have been reported, with only two being explored and visualised angiographically (Guinto et al., 1972; Teal et al., 1973). We had the opportunity of studying a case in which persistent stapedial artery was associated with unilateral first and second brachial arch syndrome; this being the first reported clinical case confirmed by arteriography.

Case Report

A.I.M.G. The subject was a 3 month old girl born of a normal 40-week pregnancy. The mother affirmed that she had taken no medication nor been exposed to radiation during pregnancy. It was a normal birth, with an Apgar test of 8 at one minute, and a birthweight of 3460 gr. The infant had a malformation on the right side of the face consisting of absence of the external auditory canal and ear, and presence of an irregular, trilobulated mass which was partly soft and partly cartilaginous, 8 x 5 cms. in size, extending from the confluence of the neck with the mandible as far as the temple (Figure 1). There was a moderate right facial paresia. Small appendages were present on the right cheek and in the suprasternal region. There was hypoplasia and forward displacement of the soft palate on the right side. A soft mass the size of an almond occupied the anterolateral portion of the neck. In the first few days after birth there were feeding difficulties, and the child had to be tube-fed. From birth she had respiratory problems owing to the abnormal implantation of the tongue on the right; a tracheotomy had to be performed. The skull radiographs demonstrated a swelling of the external paraorbital frontal zone on the right side, absence of the external auditory canal and displacement of the pars petrosa inwards, with considerable shortening of the right internal auditory canal.

The arteriography of the right carotid ar-

Dr. Pascual-Castroviejo is the Head of the Service of Child Neurology, Hospital "La Paz", Paseo de la Castellana 261, Madrid-34, Spain.

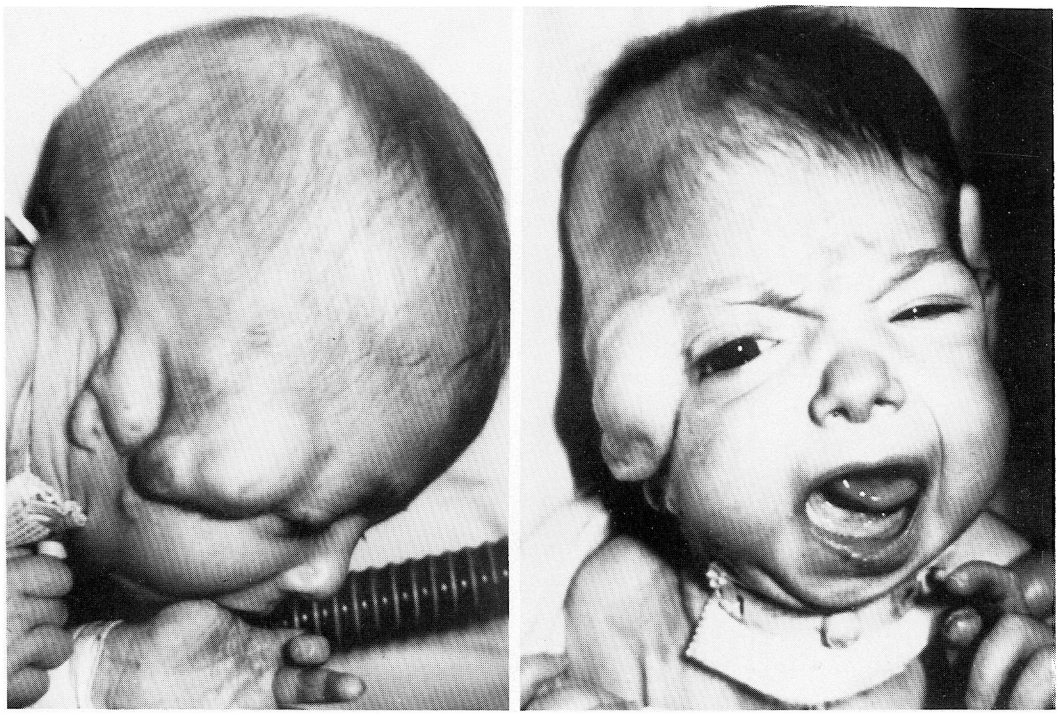


FIGURE 1. Facial aspect of the girl who has absence of the ear and presence of a trilobulated mamelon displaced forward and downwards. Rightsided facial paresia.

tery, studied via the brachial artery, showed several peculiarities, such as the presence of an anomalous artery approximately 2 mm wide which originated in the petrous portion of the internal carotid artery and then went forward and later upwards (Figures 2, 3 and 4) following the internal border of the cranium in the frontal region, giving the impression of forming peripherally the middle meningeal artery. The intracranial branches of the internal carotid artery were of normal aspect and distribution. The external carotid artery, immediately after the carotid bifurcation, gave off the occipital, superior maxillary and superior mandibular arteries, but not the superficial temporal or middle meningeal arteries. The vertebrobasilar system and the left carotid artery appeared to be normal.

When the child was 6 months old she developed a probable viral respiratory infection with pleural effusion, dying three days later. The necropsy study showed absence of the external auditory canal. Cuts of the right temporal bone demonstrated very little development of the squamous portion and a complete alteration of the petrous portion, in which there was only one cavity and this had no recognisable structures except the stapes. This cavity was continuous in its anterior part with an osteocartilaginous appendage situated in the external paraorbital zone which substituted for the zygomatic bone. The outer side of the right orbit and the external part of its floor were missing. There was very little development of the great wing of the sphenoid bone and hypoplasia of the mandibular ramus; the anterior portion of the right malar bone was floating.

An anomalous artery, 2 mm wide, originated in the anterior face of the internal carotid artery in its petrous portion and entered the zone of malformation in the right paraorbital region; the artery was not dissected as far as its terminal branches. There were no abnormalities of the Central Nervous System.

In the microscopic study of the paraorbital facial mass, nerve tissue was observed, with irregularly distributed neurones, the whole being incompletely covered with ependymal

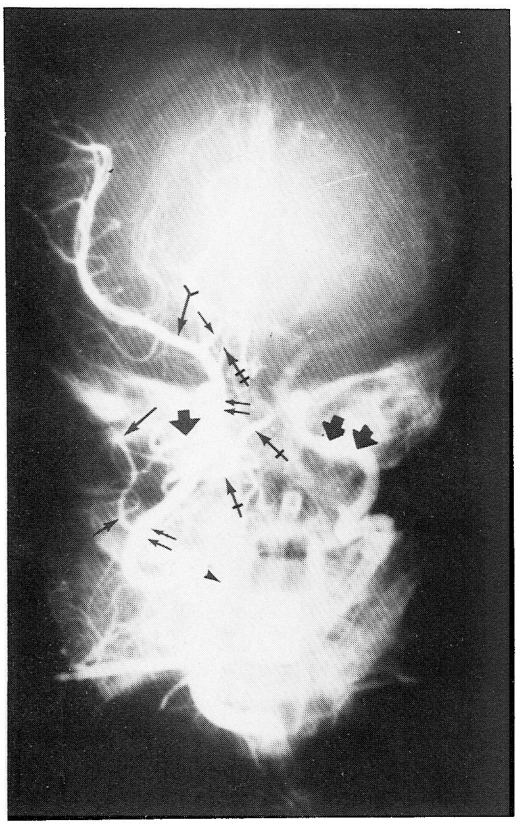


FIGURE 2. Cerebral panangiography, being better visualized the right carotid artery and its branches. The principal vessels contrasted in the picture are: stapedial artery (a wide arrow), internal carotid (two small arrows), facial artery (middle size arrow), occipital artery (→), posterior communicant artery (→), anterior cerebral artery (a small arrow), middle cerebral artery (→), left internal carotid artery (two wide arrows) and right vertebral artery (▶).

epithelium continued excentrically with a thin sheet of cells. There was bilateral bronchopneumonia, purulent pleurisy and bronchoalveolar dysplasia.

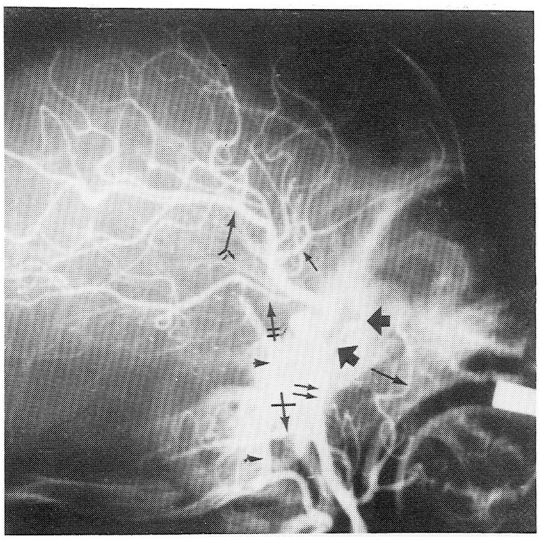
Discussion

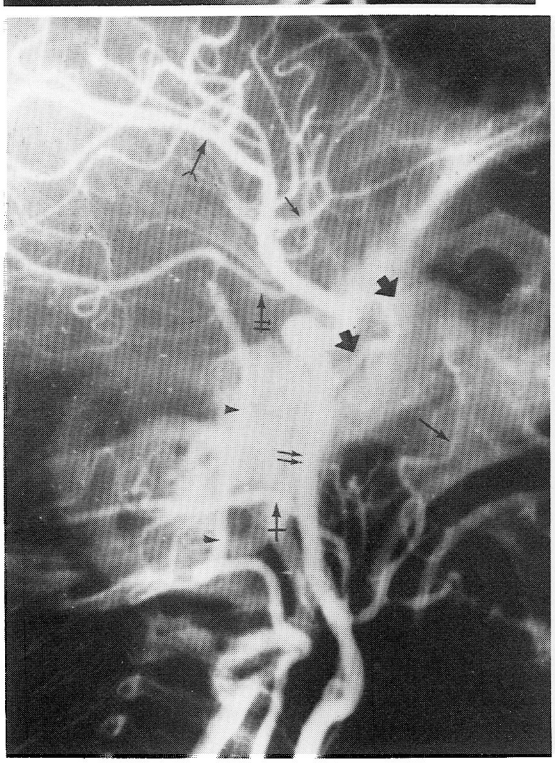
Although it is very difficult to determine the frequency of persistent stapedial artery, it does not appear to be as rare as the small number of cases published would indicate. Steffen (1968) noted 1 case in each 10,000 stapedial operations. Angiographically, the stapedial artery has been rarely described; we only know the cases of Guinto et al. (1972), Teal et al. (1973) and the present case.

According to Padget (1948) the stapedial artery appears in the 14 mm embryo (35 days) as a branch of the hyoid artery very near to its origin, entering the cranial base and leaving there an impression as an annular obturator foramen. The stapedial artery divides into maxilomandibular and supraorbital arteries. The maxilomandibular artery, or dorsal branch, will form the trunk of the middle meningeal artery. The supraorbital artery, or ventral branch, will form the peripheral branches of the future middle meningeal artery. When the embryo is approximately 30 mm long (55–60 days), all the branches of the stapedial artery have usually been annexed by the ophthalmic artery and by the arteries originating in the third pharyngeal arch which branch off the external carotid artery. While these annexations are taking place the hyoid artery and the trunk of the stapedial artery involute, disappearing towards the third month of fetal life (Congdon, 1922; Padget, 1948; Guinto et al., 1972).

Altman (1947) described the stapedial artery as a blood vessel 2 mm in diameter which enters the middle ear cavity and goes along its upper surface towards the posterior part of the promontory where it enters, on the promontory, a bony canal which takes it upwards and backwards. It later goes upwards between the crux and the stapes, entering the facial canal following a slit which is just behind the cochleariform process and continues forward beside the facial nerve for a short distance. Approximately 2 mm behind the geniculate ganglion, the artery leaves the facial canal through a special opening and is then situated between the dura and the internal surface of the middle cranial fossa in front and above.

Presence of the stapedial artery can be associated with aneurysms. Stallings and Mc Cabe (1969) described a case of aneurysm of the internal carotid artery which extended to the middle ear via the stapedial artery. However, we have found no description of cases associating persistent stapedial artery and unilateral facial dysplasia, as in our patient. It seems that the embryonic lesion that causes the mandibular dysostosis takes place in the period when the embryo measures 20–25 mm (48 to 55 days of intrauterine development). As can be observed, both alterations have a pathogenic period between the 5th and 8th weeks, which would seem to suggest that there must be a relationship between them in the





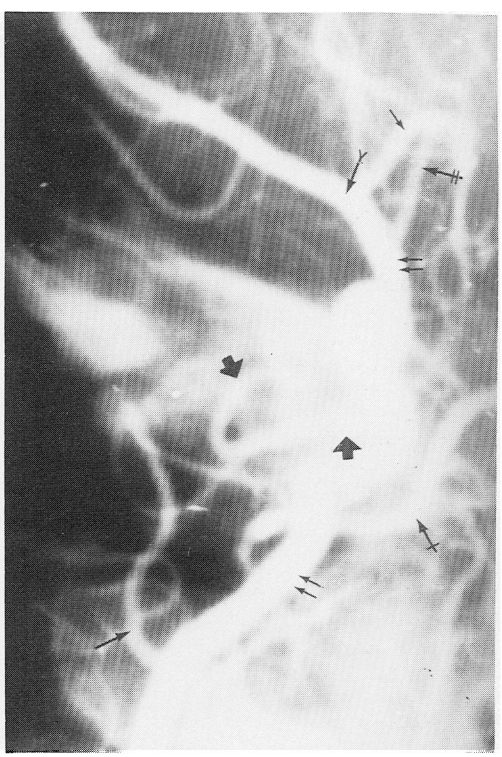


FIGURE 3 and 4. Arteriography of the right carotid artery in lateral and antero-posterior projections. A wide stapedial artery can be observed leaving the petrous portion of the internal carotid artery and going forward and outwards (wide arrow). The arteries are labeled as in the Figure 2.

case of our patient. In this way, Poswillo (1973) reported an experimental study in which a phenocopy of the first and second arch syndrome has been produced. In his opinion the causal mechanism of this syndrome could be the hemorrhage with embryonic hematoma arising from the anastomosis which precedes the formation of the stapedial arterial stem. The severity of the ensuing malformation syndrome was related to the severity of local destruction. It is not, however, usual to find persistent stapedial artery in any type of mandibulofacial dysplasia. In these cases, on the other hand, what frequently occurs is absence of the external carotid artery or one of its branches, especially the occipital branch, as we have been able to observe in a large series of angiographic studies (Palencia Luaces and Pascual-Castroviejo, 1973).

Only twenty previous cases of persistent

stapedial artery have been described. Amongst these, eighteen were discovered during ear surgery for deafness or pathology of the labyrinth in adults, or in autopsy studies when special attention was paid to the cuts of the temporal bone. There are, however, radiological signs of the skull which can suggest persistent stapedial artery; for example, the absence of the foramen spinosum in the roentgenogram of the cranial base. There is a percentage of normal cases in which this foramen is not visible (3% on both sides and 3% unilaterally; Guinto et al., 1972). It would be very difficult to evaluate radiological findings in a patient with severe anomalies. Even carotid angiography shows a persistent stapedial artery only with difficulty because the density of the pars petrosa of the temporal bone prevents good visualisation of this region of the cranial base unless one has a method of substraction. In our patient it was possible to observe the origin and direction of the vascular anomaly because of the reduced bone density in a very young subject. The three cases with angiographic studies (Guinto et al., 1972; Teal et al., 1973; and the present case) have demonstrated lack of some branches of the external carotid artery associated with persistent stapedial artery. One should not systematically associate unilateral facial dysplasia with persistent stapedial artery even if there is total absence of the external carotid artery (Palencia Luaces and Pascual-Castroviejo, 1973).

The etiology of the disease is not clear in our patient because her mother apparently did not take any drug during pregnancy. Poswillo (1973) produced a phenocopy of the first and second arch syndrome in the mouse following the administration of triazene, and in the monkey after maternal ingestion of thalidomide. Persistent stapedial artery was not mentioned in his paper. Prior to the report of Poswillo, teratogenic agents had never been linked with the incidence of first and second brachial arch anomalies. There may be a relation between the pathogenesis of both malformations—persistent stapedial artery and the first and second arch syndrome—but it has not been proven. This paper is only intended to make known an association of both malformations in the same patient. Occasionally, hemifacial microsomia has been reported in patients with chromosomal alterations type Klinefelter syndrome (Kushnick and Colondrillo, 1975; Poonawalla et al., 1980). In a recent report on a large series of cases, (Kaye et al., 1979) indicated a high frequency of positive family histories, suggestive of multifactorial inheritance.

References

Altmann, F., Anomalies of the internal carotid artery and its braches: their embryological and comparative anatomical significance. Report of a new case of persistent stapedial artery in man. *Laryngoscope, 57:* 313–339, 1947.

Calderelli, D. D., Hutchinson, J. C. Jr., and Gould, H. J., Hemifacial microsomia: Priorities and sequence of comprehensive otologic management. *Cleft Palate J., 17:* 111–115, 1980.

Congdon, E. D., Transformation of the aortic-arch system during the development of the human embryo., *Contrib. Embryol., 14:* 47–110, 1922.

Guinto, F. C. Jr., Garrabrant, E. C., and Radoliffe, W. B., Radiology of the persistent stapedial artery., *Radiology, 105:* 365–369, 1972.

Hyrtl, J., Neue Beobachtungen aus dem Gabiete der menschlichern und vergleichenden Anatomie. 3. Ueber mehrere im Menschen vorkommende Analogieen der jenigen Arteria welche Otto bei mehreren Winterschlafern durch den Steigbügel verleufend entdeckte., *Med. Jahrab. Oesterreich Staates, 10:* 457–466, 1836.

Kaye, C. I., Rollnick, B. R., and Prusansky, S., Malformations of the auricle, isolated and in syndromes, IV. Cumulative pedigree data. *Birth Defects. Original Article Series, 15:* 163–169, 1979.

Kushnick, T. and Colondrillo, M., 47 XXXXY patient with hemifacial microsomia., *Clin Genet., 7:* 447–448, 1975.

Lie, T. A., Congenital anomalies of the carotid arteries. Amsterdam, Excerpta Med. Found; Baltimore. Williams and Wilkins, 1968, pag. 98–101.

Padget, D. H., The development of the cranial arteries in the human embryo., Contrib. *Embryol., 32:* 205–261, 1948.

Palencia Luaces, R., and Pascual-Castroviejo, I., Displasias maxilofaciales. *Bol. Med. Hosp. Infant, 30:* 501–528, 1973.

Poonawalla, H. H., Kaye, C. I., Rosenthal, I. M., and Pruzansky, S., Hemifacial microsomia in a patient with Klinefelter syndrome. *Cleft Palate Journal, 17:* 194–196, 1980.

Poswillo, D.,The pathogenesis of the first and second brachial arch syndrome. *Oral Surg., 35:* 302–328, 1973.

Stallings, J. O., and Mc Cabe, B. E., Congenital middle ear aneurysm of the internal carotid., *Arch. Otolaryn., 90:* 39–43, 1969.

Steffen, T. N., Vascular anomalies of the middle ear., *Laryngoscope, 78:* 171–197, 1968.

Teal, J. S., Rumbaugh, C. L., Bergeron, R. T., and Segall, H. D., Congenital absence of the internal carotid artery associated with cerebral hemiatrophy absence of the external carotid artery, and persistence of the stapedial artery., *Am. J. Roentgenol., 118:* 534–545, 1973.