PERSISTENT STAPEDIAL ARTERY

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SINCE Hyrtl (1836) first described the persistence of the embryonic stapedial artery fewer than 20 cases have been reported. It has never been described in a thalidomide child and since a great deal of reconstructive surgery is required for these children it is felt that the following case will be of interest. There is no way to confirm the presence of the artery *in vivo*; on the other hand its persistence does not seem to involve pathological consequences except to make stapes surgery more difficult. Its discovery is always accidental although we may presume that it is present more frequently than is revealed by the few reports.

Case Report

A.D., a female child, was born on May 26th, 1960, the first of two children, to a mother aged 33 at an estimated 40 weeks' gestation after a pregnancy complicated by hydramnios, pre-eclampsia and the administration of thalidomide from the 6th to 14th week. The birth weight was 8 lb. 1 oz. and the child's abnormalities were noted at birth.

The upper limbs were rudimentary; three digits were placed on stumps of upper and lower arms. The lower limbs were normal. The left eye was small, closed and opaque; the disc showed a congenital coloboma and there was an oculomotor paralysis. Over the central part of the forehead there was the classical hæmangiomatous birth mark. There was a left facial palsy and the left side of the face was smaller than the right. The external ears were rudimentary. The child thrived well but motor milestones were a little retarded due to a spastic diplegia. Her intelligence is estimated to be only a little subnormal.

She was referred to us in April, 1964, not yet saying any words but definitely babbling. Examination showed the right ear to be rudimentary with a very narrow external meatus and a normal drum. The left ear was also rudimentary but its meatus tapered off to a narrow pit. The upper respiratory tract showed no abnormality.

She localized sound well and equally in all quadrants and response was obtained to suprathreshold intensities with low and high frequency sounds. Free field audiometry showed her to have a 60 db. loss in both ears at 250 and 500 c.p.s., a 60 db. loss on the left at 1,000 c.p.s. and 80 db. loss on the right. No response was obtained above this frequency. E.E.G. audiometry showed a startle response however at 2,000 c.p.s. with a 90 db. intensity.

Tomograms of the temporal bone under sedation showed a right inner ear to be well developed. The right middle ear cavity was large and well formed and

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the ossicular mass was in the normal position. On the left side the inner ear was normally developed but the middle-ear cavity was very small and no ossicular mass could be identified.

In August, 1964, the right ear was explored *via* a Heerman's incision. The drum was small and normal and the meatal wall was widened with a drill. When the drum was reflected the malleus was found to be joined to the posterior wall by a thick bar of bone and to the anterior wall by a thinner piece. These bars joined the short process of the malleus and were removed. The facial nerve was very superficial and was exposed in its vertical course and there was no bony covering to it in the middle ear. There was a malformed incus consisting only of a body which tapered to a sharp point joined to the roof of the middle ear; the long process was rudimentary. The incus was joined to the head of the malleus by a bony ankylosis and was removed.

The stapes was represented by an anterior crus with a bar of bone about one-third of the width of the footplate going posteriorly from this across the footplate. From the floor of the middle ear anteriorly came a large vessel which coursed uncovered over the promontory to reach the medial side of the anterior crus. At the lower side of the footplate it divided into two branches which anastomosed over the facial nerve. It was not possible to trace it farther.

Removal of the footplate was difficult and it required a great deal of drilling before it was entirely removed. When it was removed the oval window niche was a very deep hole and the condition of the membranous utricle was regarded with suspicion. A fat and wire graft was inserted, the wire being crimped round the handle of the malleus. The drum was repositioned and the widened meatus packed. Convalescence was interrupted by a left basal pneumonia which settled with penicillin.

Hearing tests since the operation including E.E.G. audiometry revealed no marked improvement in hearing.

Discussion

The stapedial artery persists completely in quite a number of animals and partially in others. It is persistent in rodents and in the guinea-pig it passes between the crura of the stapes together with a bony barb. Hyrtl (1836) first called attention to a vessel found in some hibernating animals running across the obturator foramen of the stapes. Gradenigo (1887) found in cat embryos that the stapedial artery branched off from the internal carotid artery together with the primitive hyoid artery in a hyo-stapedial trunk; the primitive hyoid artery soon disappears while the stapedial artery persists until the third month.

In man it has been described in the normal, in a case of multiple deformities (Kelemen, 1958), in anencephaly (Altmann, 1957) and in Paget's disease (Altmann, 1947).

Hyrtl (1845), Alexander (1899) and Brock (1922) all suggested that the origin of the stapedial artery was from the internal carotid.

In his classical study on the artery in 1947, Altmann described the artery as coming from very close to the internal carotid artery. It passes

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into a bony canal on the promontory and runs upwards and backwards going in between the crura of the stapes being covered on the outside by mucous membrane and adherent to the posterior surface of the anterior crus. It enters the facial canal at a dehiscence just behind the cochleariform process and extends a short distance forwards together with the facial nerve. 2 mm. behind the geniculate ganglion it leaves the facial canal through a small opening and proceeds between the dura and inner surface of the middle fossa forwards and upwards. Before leaving the facial canal it gives off two branches—one runs forwards together with the greater petrosal nerve and one backward and downward with the facial nerve to anastomose with the stylomastoid branch.

Kelemen (1958) described it as being first seen in the neighbourhood of the eustachian tube forming its canal by deep excavation of the cochlear capsule near the top and winding in a canal in the direction of the floor of the tympanum.

The intracranial ramification of the anomalous artery was studied by Alexander (1899) and Adachi (1928). In Alexander's case, the vessel divided into a medial branch which supplied the inner surface of the greater wing of the sphenoid and anastomosed with orbital vessels and a lateral branch which substituted the middle meningeal artery. In Adachi's case the anastomosis with the orbital vessels was very wide, otherwise the ramification was the same.

In most cases described, the artery was enclosed in a bony tunnel in the promontory, but in the above case and those of Zuckerkandl (1873), Brock (1927) and Adachi (1928) it was not.

Three arteries are produced from the first aortic arch:

(a) the supraorbital, later to become the middle meningeal.
(b) the infraorbital representing the maxillary.
(c) the mandibular or inferior dental.

With the disappearance of the first arch these three vessels are all maintained by the stapedial artery, which is usually a short lived vessel in the human embryo passing through the stapes supplying structures derived from the posterior end of the second visceral arch and affixing itself to the stem of the supraorbital artery. When the external carotid artery develops, it takes over these three vessels again and the stapedial artery disappears. In animals in which the stapedial artery persists, it shows an upper branch which gives origin to the middle meningeal artery and anastomoses with the ophthalmic artery in the orbit, and an inferior branch which develops into the internal maxillary artery.

In 1942, Cauldwell and Anson stated that the stapes first appears in a 7 mm. ($4\frac{1}{2}$ week) embryo as a condensation of mesenchyme at the second visceral arch. In the 10 mm. stage it is pierced by a relatively large stapedial artery and its presence determines the annular shape of the bone.

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Strickland, Hanson and Anson (1962) stated that the process whereby the stapedial artery gains a central position in the stapedial blastema is extremely difficult to determine due to the early occurrence of this inter-relationship and to the rapidity with which it takes place. The fact that an adjacent artery is co-existent with a solid blastema in the early stages may be taken as evidence favouring the theory that the blastema folds around the artery rather than that the artery pierces the blastema in a direct manner. Previous to this it was thought that the vessel was the primary force for the moulding of the shape of the stapes. From the above we see that it is not the artery which forms the stapes but it is the mesenchyme of the second arch which forms a folding ring around the artery. As the stapes develops, the obturator foramen becomes less circular and also the crural arch on the obturator surface is excavated to form a trough. As Hough (1963) states, it would be difficult to imagine that the stapedial artery would produce these characteristics.

Some argument arises as to the significance of a persistent stapedial artery. Because an artery in the adult has a similar position with respect to the stapes that an embryonic vessel had to the developing ossicle, it does not warrant the conclusion that the embryonic artery persisted and directly participated in the formation of the adult artery. Many authors have noted the vessel which travels from above the oval window at the facial canal down to the mucosal network on the promontory. According to Nager and Nager (1953) it arises within the facial canal directly from the descending branch of the superficial petrosal branch of the middle meningeal artery that gives origin to the artery to the posterior crus of the stapes. The course of this artery is, however, opposite to that of the stapedial artery since it ends in the promontorial plexus.

In 1955, McKenzie and Craig put forward a theory of causation of the Treacher Collins syndrome and also of facial agenesia. They postulated that absence of the embryonic stapedial artery caused by some accident around the sixth week, immediately after formation of the primitive face, will give rise to maldevelopment of the first arch vessels. It is interesting to note that the left side of the face in our described case was smaller than the right or operated side.

Suspicion that the consumption of thalidomide by pregnant mothers was a cause of dysmelia was first voiced by Lenz (1962). According to him the critical period for thalidomide damage is between the 37th and 50th day after the L.M.P. It is not our intention here to consider the defects caused by thalidomide, but in 1964, Jørgensen, Kristensen and Buch described aplasia of the inner ear due to thalidomide. The depth of the vestibule, the appearance of the utricle and the poor hearing result in the described case make us think that the inner ear must have been abnormal in this case. A similar anatomical state of a very thick footplate, a narrow vestibule and an aplastic utricle has been seen by us to exist in a temporal

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bone acquired from a deaf mute with the Klippel Feil syndrome. The rest of the inner ear was grossly abnormal and will be described at a later date.

Summary

The finding of a persistent stapedial artery in a case of thalidomide deafness is reported. The relative rarity of such a finding is pointed out and the literature is reviewed. The origin, course and termination of the artery is described as is the embryology. The relationship of the artery to the shape of the stapes, the normal vasculature of the middle ear, and to facial agenesia is discussed.

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