The Journal of Laryngology and Otology
June 1994, Vol. **108**, pp. 461–465

Historical Article

Ear dysplasia after Mondini

PETER D. PHELPS

Abstract
An historical account of descriptions of malformation of the inner ear.

Key words: Ear deformity, congenital; Deafness

Mondini (1791)

Almost nothing is known about the life of Carlo Mondini except that he was a citizen of Bologna in Northern Italy and published in medieval Latin his operscula (small work) in the quarterly scientific journal of that city (Mondini, 1791). The paper starts by acknowledging the work of previous authorities including a contemporary Italian anatomist Cotunnius (Domenico Cotugno, 1736–1822) who apparently was the first to recognize fluid-filled cavities forming the labyrinth of the inner ear. Mondini then describes to 'his most distinguished colleagues' a dissection performed a few years before on the ears of an eight-year-old boy born deaf who died after being knocked down by a fast-moving wagon. Mondini not only knew the anatomy of the ear but he appears to have been a physician as well as he comments that he had occasion to test the boy's hearing while he was still healthy and therefore was able to confirm that the child was completely deaf. The dissection was very thorough and although there is a full description of the cochlear deformity that bears his name, Mondini seemed more concerned with the grossly widened vestibular aqueduct which he measured ('septum lineas latam' meaning seven lines wide which is equivalent approximately to 15 mm). His text was accompanied by nine line drawings. Two are shown here (see Figure 1 a and b). Moreover the precise descriptions make it quite clear which parts of the petrous temporal bone were abnormal: the vestibular aqueduct grossly abnormal, the vestibule larger than usual, a sac in place of the distal cochlear coils but it is made equally clear that other parts such as the oval and round windows and the basal cochlear coil were normal. Thus there can be no dispute as to what Mondini originally described.

Isben–Mackeprang collection of temporal bones (1824–1837)

The Ibsen–Mackeprang collection of temporal bones was created over a period of 13 years by a professor of anatomy and a physician in Copenhagen. The bones were given to the Institute of Pathology in Copenhagen in 1844. Originally the collection comprised the temporal bones from 55 children who were inmates at the Royal Institute for Deaf Mutes. The collection still exists and is complete apart from nine bones. It was the subject of a temporal bone study (Jensen, 1969). Eighteen of the 101 temporal bones showed evidence of congenital developmental deformity; the Mondini malformation was present in nine and there were other deformities of the semicircular canals and internal auditory meatus (Figure 2).

Cock (1838)

Edward Cock of Guy's Hospital, London (Cock, 1838) described several cases of congenital ear deformity including one with severe dysplasia of the labyrinth such that a sac replaced the cochlea and it had a wide communication with another sac equivalent to a dysplastic vestibule. This would seem to be the first description of the deformity for which there is a significant risk of a cerebrospinal fluid fistula into the middle ear (Phelps, 1986). Cock also described a large vestibular aqueduct in one of the dissected temporal bones.

Thomson (1847)

In the Edinburgh Journal of Medical Science, Allen Thomson (1847) reported three cases of bilateral atresia of the external auditory meatus. Two of these patients also

From the Department of Radiology, Royal National Throat, Nose and Ear Hospital, Gray's Inn Road, London WC1X 8DA.
Accepted for publication: 27 February 1994.

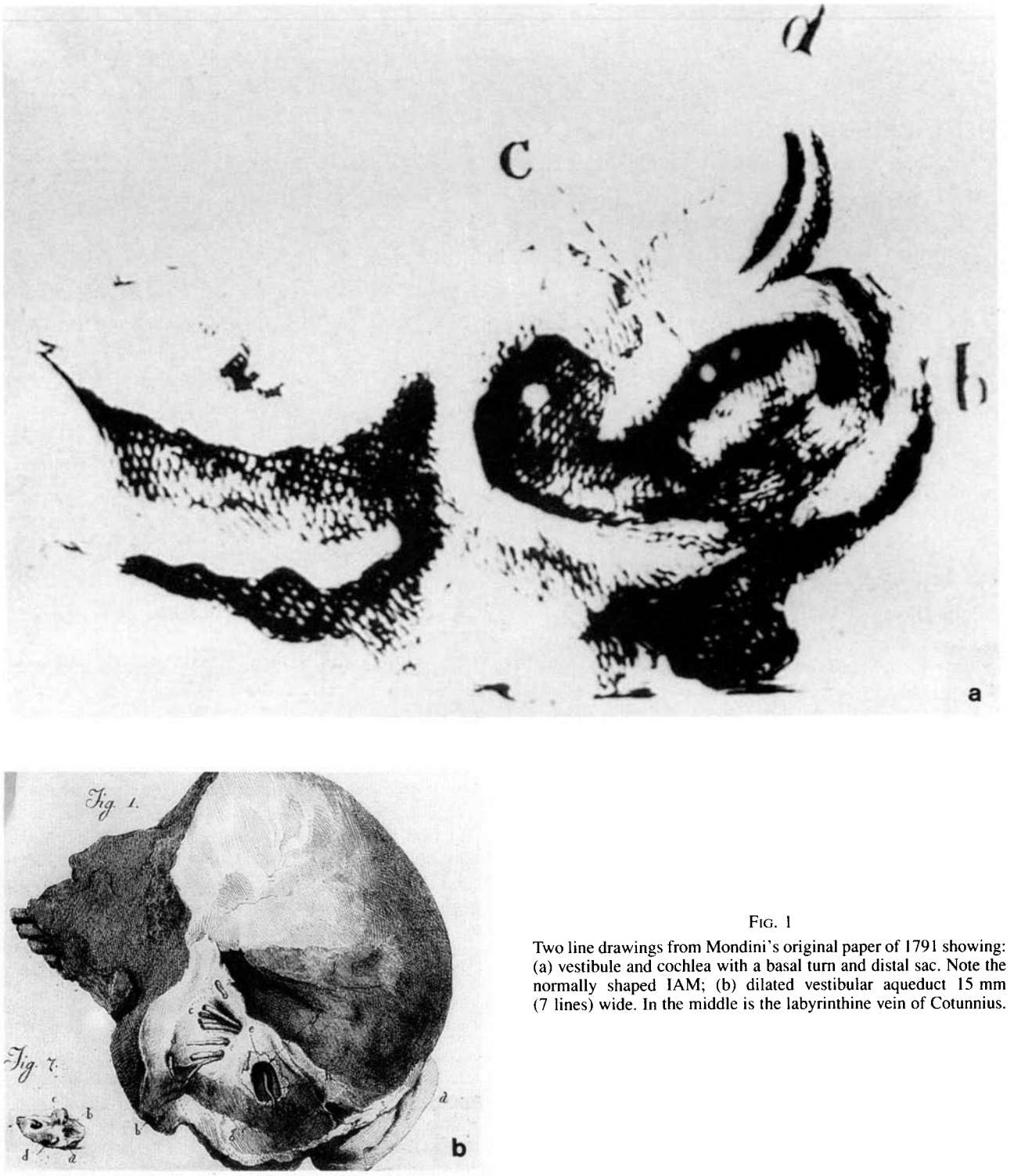

FIG. 1

Two line drawings from Mondini's original paper of 1791 showing: (a) vestibule and cochlea with a basal turn and distal sac. Note the normally shaped IAM; (b) dilated vestibular aqueduct 15 mm (7 lines) wide. In the middle is the labyrinthine vein of Cotunnius.

had deformities of the mandible, malar bones and zygoma. These cases are presumably what would now be described as craniofacial microsomia. In the same journal Toynbee (1847) described a personal dissection of an ear with absence of the external meatus and tympanic ring. The middle ear cavity was small and very narrow with two abnormal ossicles only: a globular malleus and single strut stapes fixed in the oval window (Figure 3). In his subsequent book Toynbee (1860) repeated the descriptions of his dissections of temporal bones from five patients, as well as describing the previous findings and conclusions of Thomson. Unfortunately, of the five sets of temporal bones examined by Toynbee only one set showed definite structural abnormalities of the labyrinth. The superior and posterior semicircular canals were partially absent in one labyrinth but this could have been the result of labyrinthitis obliterans. It is of interest that Toynbee also used the line as a unit of measure.

Michel (1863)

Michel (1863) described complete absence of both inner ear and auditory nerves in a boy of 11 years. Although this exceedingly rare deformity has been named after Michel it had in fact been described previously by Saissy (1819) in a French medical dictionary in 1819. In

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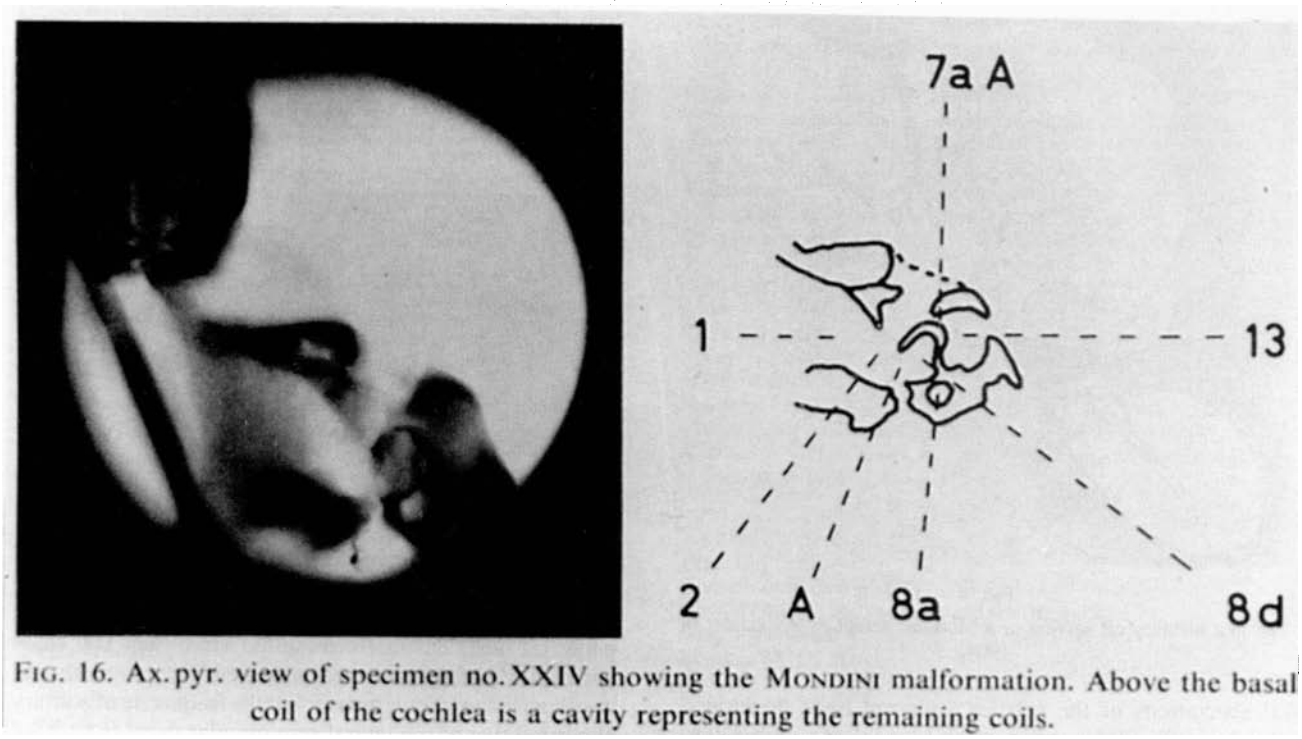

FIG. 16. Ax. pyr. view of specimen no. XXIV showing the MONDINI malformation. Above the basal coil of the cochlea is a cavity representing the remaining coils.

FIG. 2

Pluridirectional tomographical illustration of a temporal bone in the Ibsen–Mackeprang collection (from *Acta Radiologica Supplementum* (1969) **286:** courtesy of Dr Jorgen Jensen).

his paper Michel quotes lesser deformities of the inner ear found by other authors. In particular there is a description of grossly underdeveloped labyrinths represented by a curved tube on one side and a simple sac on the other. This type of primitive otocyst is commoner than the true 'Michel' type.

Mygind (1890)

Dr Holger Mygind of Copenhagen (Mygind, 1890) published an extensive survey of the cases of pathological changes of the temporal bones of deaf persons which had been described up to that date. In 32 of the 118 cases described the deafness was considered to be congenital. The descriptions of all lesions of external, middle and inner ears are listed in tabular form.

ARTICLE II.—*Description of a Congenital Malformation in the Ears of a Child.* By JOSEPH TOYNBEE, F.R.S., Senior Surgeon to the St George's and St James' Dispensary. Read before the Pathological Society of London, January 18, 1847.

The stapes, instead of its two crura, has a process flattened above and below, and about three-fourths of a line in length; to the inner extremity is attached the base, firmly fixed in the fenestra ovalis, while the outer extremity is slightly attenuated and presents no articulating surface. Over the stapes, and having a direction from above downwards and backwards, the portio dura nerve is seen unsurrounded by bone, but in contact with the mucous membrane of the tympanum.

THE TYMPANIC CAVITY, WITH THE EUSTACHIAN TUBE, OPENING INTO ITS ANTERIOR AND INFERIOR PART. THE TWO OSSICLES, DETACHED FROM EACH OTHER, ARE TO THE RIGHT.

FIG. 3

Description and illustration from Toynbee's paper (1847).

Alexander (1904)

Alexander (1904) from Vienna was the first to describe a case of the Mondini malformation histologically (Figure 4). Mondini's name is not mentioned but the findings correspond closely to earlier macroscopic findings and to later histological studies. The 'scala communis' of Alexander's case is equivalent to the 'amplam caveam cavitati ultimi gyri respondentem' of Mondini. This deformity of the cochlea would seem to be the only bony abnormality present in Alexander's case but the dissection showed hypoplasia of the membranous labyrinth in both cochlear and vestibular parts as well as an enlarged saccus endolymphaticus.

Siebenmann (1904)

No attempt at classifying the pathological changes found in the temporal bones from deaf subjects had been made so far but the introduction of modern histological techniques made possible a detailed differentiation. Siebenmann's book on the pathology of deafness (Siebenmann, 1904) lists two types of structural (i.e. bony) abnormality namely those of Michel and Mondini. The lesions in the other types were confined to the membranous labyrinth. Surprisingly this outdated and grossly inadequate classification, although valuable as the first of its kind, is still being quoted (Mafee *et al.*, 1992).

Radiological studies

Greater understanding of the fine anatomy and congeni-

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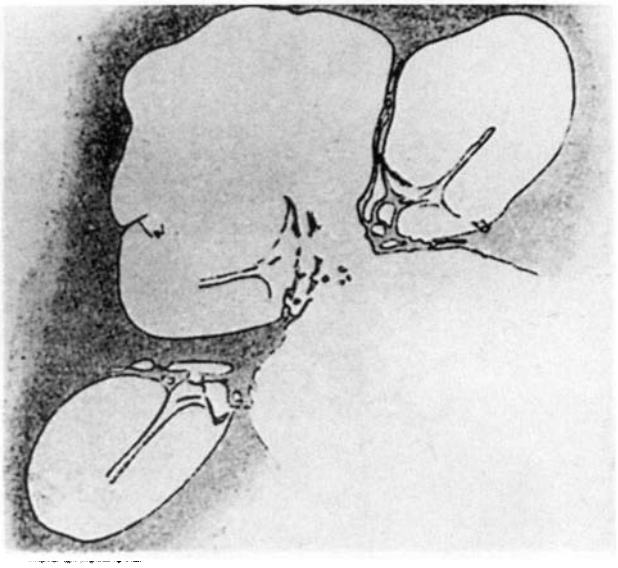


FIG. 4

The first histological section of a Mondini defect by Alexander in 1904.

tal aberrations of the petrous temporal bone developed rapidly in the 20th century after Alexander's description of cochlear malformation but the progress of imaging to assess these deformities was slow. There appear to have been no attempts to describe malformations of the temporal bone as assessed by imaging before the second World War. Most descriptions from the 1930s were concerned with the extent of pneumatization although good plain film views of the labyrinth could be achieved by conventional radiography (Figure 5). However, as with the histological assessment, significant progress depended to a great extent on the development of sectional studies.

The first tomographical assessment of congenital ear malformations seems to have been by Camp and Allen (1940) who examined four patients using a simple attachment to the standard radiographic couch. These patients all had microtia and atresia of the external auditory meatus on one side and were examined by sections in the coronal and sagittal planes. The linear tomograms published are of good quality and not surprisingly give more information

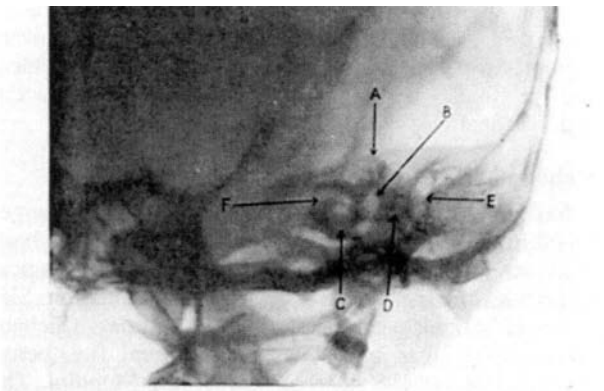
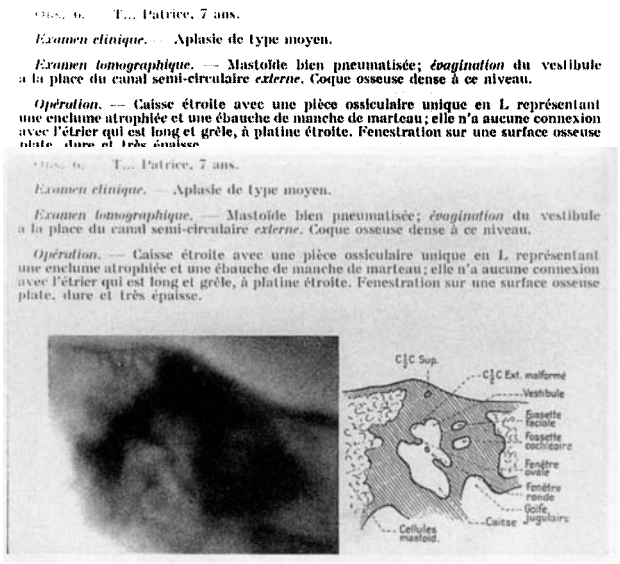


FIG. 212.—Normal mastoid and petrous portions of the temporal bone in the postero-anterior oblique view. The mastoid process is of the small-celled type. A, superior semicircular canal. B, Vestibule. C, Cochlea. D, External semicircular canal. E, Mastoid antrum. F, Internal auditory meatus.

FIG. 5

A Stenvers view of the labyrinth from a prewar radiology textbook. (*A Textbook of X-ray Diagnosis*, by British authors, published in 1939, by H. K. Lewis & Co., London).



the polytome in France was a great help to imaging the inner ear. Ombredanne and Francois (1961) published a paper on inner ear malformations. More than 100 cases with aplasia of the ear were examined. The labyrinth was involved in less than a quarter but the frequency of solitary abnormalities of the lateral semicircular canal is stressed (Figure 6).

Pluridirectional tomography was the imaging investigation of choice (for the next 25 years) to show fine detail in the temporal bone. Developments in computed tomography have subsequently made the polytome obsolete, but imaging studies in the lateral plane (Valvassori and Clemis, 1978) initially with the polytome showed that the large vestibular aqueduct, first described by Mondini in 1791, is probably the commonest congenital structural deformity of the inner ear. Computed tomography is now the imaging investigation of choice for congenital malformations of the ear and can demonstrate bony structures like the crura of the stapes and the cochlear aqueduct which are a fraction of a millimetre in size. Magnetic resonance imaging shows good prospects for assessing soft tissue abnormalities of the inner ear (Figure 7).

A study of the progression of knowledge about malformations of the ear and especially the inner ear over two centuries since Mondini's original operscula would seem appropriate especially as eponyms such as Mondini and

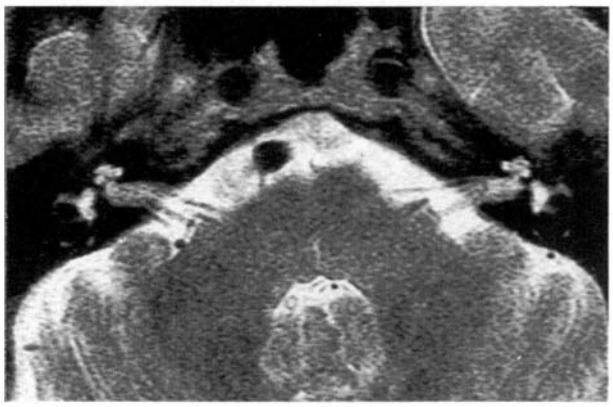


FIG. 7

Axial section through the skull base by magnetic resonance imaging using the fast spin echo technique to show the vestibules, coils of the cochlea as well as the nerves and vessels in the internal auditory meatus.

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Michel deformity are used and, more to the point, misused so frequently. A strong case can be made for keeping to the original descriptions when eponyms are quoted.

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Address for correspondence:
Peter D. Phelps,
Consultant Radiologist,
Royal National Throat, Nose and Ear Hospital,
Gray's Inn Road,
London WC1X 8DA.

Fax: 071–833–5518.