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Commentary

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The scientific report written in Latin by Carlo Mondini in 1791, titled *The Anatomic Section of a Boy Born Deaf* (1), has been often cited but seldom read. Recently, the *American Journal of Otology* published an English translation by Gordon J. Hartley of Rugby, England, with comments by Peter D. Phelps (2). Originals of Mondini's publication known to exist in the United States are in the collections of the Library of Congress, Washington, D.C., Columbia University, New York City, and the Huntington Memorial Library, San Marino, California.

It is clear from Hartley's translation that the inner ear anomaly described by Mondini consisted of 1) a cochlea of one-and-one-half turns instead of the normal two-and-one-half turns, comprising a normal basal turn and a cystic apex in place of the distal one-and-one-half turns; 2) an enlarged vestibule with normal semicircular canals; and 3) an enlarged vestibular aqueduct containing a dilated endolymphatic sac. Thus, the cochlear anomaly was relatively mild, and would correspond to incomplete partition in the spectrum of congenital cochlear malformation because of an arrest of embryogenesis, as proposed by Jackler et al (3). Arrest at earlier stages in the spectrum include cochlear hypoplasia and common cavity and cochlear aplasia. Even earlier arrests result in otocyst and complete labyrinthine aplasia (Michel's anomaly) (3).

Mondini graphically supported his written description of the anatomic findings with drawings only (1, 2). Alexander is credited with first publishing, in 1904, a histologic section corresponding to the cochlear anomaly described by Mondini (4). In recent decades, multiple authors have published exquisite histologic sections firmly establishing the Mondini anomaly as an entity (5–8). Radiologically, the anomaly was recognized on polytomography by Jensen as early as 1969 (9). Subsequently, he also described a more severe type of deformity consisting of an amorphous cochlear sac continuous with a dilated vestibule, for which he suggested the term ''dysplasia'' (10). Unfortunately, the term ''Mondini dysplasia'' has come to mean virtually any congenital malformation of the osseous labyrinth detectable on radiographic examination (3). Inevitably, seemingly contradictory observations and conclusions about ''Mondini dysplasias'' followed.

For example, dozens of articles have appeared in the literature on the subject of spontaneous CSF fistula and recurrent meningitis associated with an anomaly of the inner ear (11–16). Some authors asserted that such complications occurred in Mondini dysplasia, whereas others insisted that they did not. Who was right? On close reading, Phelps et al (13, 14, 16) clearly restricted the term ''Mondini'' to the anomaly described by Mondini, whereas Olhms et al (15), among others, extended the term to other cochlear anomalies (17). In 1994, Phelps et al showed convincingly that in their patients examined with CT, only those with dysplastic cochleas with a widened basal turn joining a dilated vestibule developed recurrent meningitis, and none of those had true Mondini deformities (16). Using ''Mondini dysplasia'' as an inclusive term would not have permitted differentiation between these two subgroups.

Miyamoto et al performed cochlear implantation on a child with ''Mondini inner ear malformation'' and encountered ''a profuse flow of perilymph which rapidly filled the middle ear'' (18). Silverstein et al likewise implanted a child with ''Mondini deformities,'' but the perilymph flow was ''self-limited and not excessive'' (19). Why were their experiences different? On close inspection of the CT images, Miyamato's patient actually had a markedly dysplastic cochlea without a modiolus, which openly communicated with the internal auditory canal. Conversely, Silverstein's patient had an incomplete cochlear partition with an intact cochlear base, consistent with a true Mondini anomaly. Thus, the degree of perilymph or CSF leak upon cochleotomy could have been predicted by proper classification of the cochlear anomalies. For the purpose of patient selection, surgical planning, and comparison of results in cochlear implantation, most surgeons have since opted for the Jackler classification (20–25).

A third reason for abandoning the use of ''Mondini dysplasia'' as an inclusive term is that not all cochlear anomalies are caused by developmental arrest. Genetic defects, such as those in X-linked progressive mixed hearing loss (26–28) and in familial mixed deafness with branchial-oto-renal syndrome (ear-pits deafness) (29), may result in distinctive cochlear anomalies entirely different from those found in simple arrests in embryogenesis. With the rapid progress in genetic studies and the increasing availability of high-resolution CT and

Address reprint requests to William W.M. Lo, MD, St Vincent Medical Center, 2131 West Third St, Los Angeles, CA 90057.

MR imaging, more specific cochlear anomalies will undoubtedly be identified. The widespread use of ''Mondini dysplasia'' as an inclusive term will only obstruct this progress.

As in the otologic literature, confusion of terminology about Mondini's dysplasia has often occurred in the radiologic literature, even among otherwise superb publications. Often Mondini's description of the anomaly is correctly acknowledged, but CT images of more advanced anomalies are used as illustrations of the ''Mondini anomaly'' (30–33). At the same time, the large vestibular aqueduct has long been recognized by imaging (34) and considered a frequent isolated radiologic finding associated with sensorineural hearing loss (30, 34–37), although a coexisting incomplete partition (the subtlest component of a true Mondini dysplasia) may have been present but overlooked. Lemmerling et al recently found, by careful review of thin-section high-resolution CT, that a shortened modiolus could be seen in all 39 ears previously thought to have only a large vestibular aqueduct, and that incomplete partition between the apical and middle turns was present in the majority of those ears (38). In this issue of the *AJNR,* Davidson et al (page 1435), with MR images of exceptional quality, have further documented that, in fact, isolated enlargement of the endolymphatic duct and sac is rare, and the association of mild cochlear dysplasia, vestibular dilatation, and a large endolymphatic duct and sac, namely the Mondini triad, is common!

The use of eponyms has been justifiably decried (3, 33). Nonetheless, knowing what Mondini did describe is not only important for its historical significance, but also essential for truly understanding the voluminous literature on the subject of inner ear anomalies. Needless to say, the use of the term ''Mondini dysplasia'' to include a variety of cochlear dysplasia must be abandoned. If the term is used, Mondini malformation should refer only to the triad of anomalies that he described. In view of the longstanding confusion in the literature, reports of ''Mondini dysplasias'' should not be cited without careful scrutiny. Descriptive anatomic terms are preferred, and it is gratifying to see that several major radiologic texts on the subject have adopted this approach (32, 33), as have many recent journal articles, including those by Lemmerling et al (38) and Davidson et al.

A classification of arrests of cochlear embryogenesis is essential for prognostic and therapeutic guidance. The system proposed by Jackler (3) is conceptually logical and clinically practical (20–25), but not without minor ambiguity of its own. The term ''hypoplasia,'' used to denote arrested cochlear development at a stage between incomplete partition and common cavity, may misleadingly imply that such cochleas are small in size when in fact they are usually not. Alternatively, certain genetic cochlear anomalies may indeed be small in size but are not caused by sporadic arrest of embryogenesis and may not be fit into this classification. A clarification of the term may be in order.

Phelps restricts the term ''Mondini deformity'' to cochleas with a normal basal turn and a deficient interscalar septum for the distal one-and-one-half turns (ie, a true Mondini deformity, as originally described, and one that corresponds to incomplete partition in the Jackler classification) (16). Phelps uses the term ''cochlear dysplasia'' to denote cochleas with a widened basal turn and a wide communication with a dilated vestibule (an intermediate anomaly that appears to correspond to the Jackler ''hypoplasia''). Thirdly, he calls a primitive cochlear sac continuous with a vestibular sac without bony separation from the arachnoid space simply a ''sac.'' This appears equivalent to the Jackler ''common cavity.''

Fundamental to the accurate evaluation of a labyrinthine anomaly is a careful analysis of each of its components (39). Although the anomaly may also involve other structures, the cochlea generally requires the greatest attention. A full description should include the estimated number of turns, the size of the turns, the integrity of the cochlear base, the length of the modiolus (16, 25, 38, 40), the presence or absence of the round and oval windows, and, on high-resolution MR images, the appearance of the cochlear scalae. Such an approach will ensure the clarity of a scientific communication.

Excellent high-resolution CT and MR imaging, such as that shown respectively by Lemmerling et al (38) and Davidson et al, will greatly facilitate the study of labyrinthine anomalies. Nevertheless, a sound knowledge of the terminology of the anomalies will remain indispensable.

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