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CSF Fistula in a Patient with Mondini Deformity: Demonstration by CT Cisternography

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In 1791, Mondini described a membranous and bony dysplasia of the inner ear in a congenitally deaf child [1]. Classically, the Mondini deformity consists of a cochlea containing a normal basal turn and a total of 1.5 rather than 2.5 turns; the middle and apical turns are incorporated into a single sac. Because dysplasia of the labyrinth may be associated with dural dehiscence and a bony defect in the internal auditory canal [2] and with an abnormal stapes footplate [1], a CSF fistula between the posterior fossa and middle ear can occur. In patients with a congenital inner ear anomaly, a CSF fistula has been identified by fluorescein cisternography at surgery [3], and by conventional tomography [4, 5], radionuclide cisternography [6], or cisternal Pantopaque tomography [2, 6, 7] preoperatively. To our knowledge, this is the first report of a CSF fistula of the inner ear demonstrated by CT cisternography.

Case Report

A 2-year-old boy with bilateral deafness and recurrent otitis media was hospitalized at age 20 months and again at age 25 months for pneumococcal meningitis. The child was immunologically competent and had a normal hemoglobin electrophoresis. During the second admission, axial CT scans (1-mm thick) of the temporal bones demonstrated a sclerotic, hypoplastic left inner ear (Fig. 1). The right inner ear (Fig. 2A) consisted of a common vestibulocochlear chamber; the basal turn and promontory were hypoplastic. The vestibule was ectatic. Coronal reconstruction (Fig. 2B) indicated a defect in the fundus of the internal auditory canal. The ossicles, facial nerve canal, and external auditory canal of both ears were normal.

After treatment for the meningitis, the child was reevaluated by CT cisternography. Five milliliters of iohexol 180 mg/dl were introduced into the lumbar subarachnoid space and allowed to flow into the posterior fossa. Direct coronal CT scans (1.5-mm thick) demonstrated contrast material extending through a defect in the lamina cribosa of the right internal auditory canal into the deformed cochlea and vestibule (Fig. 3). The left internal auditory canal showed no leak.

At surgery, a simple mastoidectomy revealed exuberant granulation tissue and inflamed mucosa. A modified tympanomeatal flap was raised and the middle ear was entered. The middle ear space was

filled with polypoid, inflamed mucosa, which obscured virtually all landmarks. This tissue was carefully removed up to the level of the oval window. The mastoid was reentered and an extended facial recess dissection was carried out to define the course of the facial nerve and to allow better access to the oval window region. After removal of diseased mucosa from the oval window, clear fluid was observed to be leaking from the anterior aspect of the oval window. At this time, a stapedectomy was performed. The stapes footplate was small and appeared scalloped around its medial margin. The oval window was normal in size, but was not completely covered by the irregularly marginated stapes footplate. A patch, fashioned from temporalis fascia, was placed over the areas of the round and oval windows. Two small pieces of fat were placed on top of the fascia and the middle ear and mastoid were each packed with Gelfoam. The wound was closed in the standard fashion and a mastoid dressing was applied. Postoperatively, the patient was positioned with his head elevated, placed on bed rest, and treated with acetazolamide. His dressing was removed on the second postoperative day; no further perilymph leak was noted.

Discussion

The Mondini deformity may be found in association with such syndromes as Klippel-Feil, Pendred, and DiGeorge, or it may occur as an isolated abnormality [1]. Familial occurrence has been reported. The dysplasia may be bilateral or unilateral. Acoustic and vestibular dysfunction are variable in severity, and may be static or progressive [1].

Although the Mondini deformity typically denotes a cochlea with a normal basal turn but a deficient total number of turns, variant forms of this dysplasia have been described [8–10]. Becker et al. [10] reported a variant form of the classic Mondini abnormality (pseudo-Mondini deformity), in which the basal turn was dysplastic and a common vestibulocochlear chamber was present. In their series of patients with Mondini dysplasia, Valvassori et al. [11] noted that one half of the abnormal cochleas were reduced in size (dwarf cochlea) rather than reduced in number of turns. In our patient, the small size of the cochlea and semicircular canals in the left ear is consistent with a prenatal insult interfering with growth

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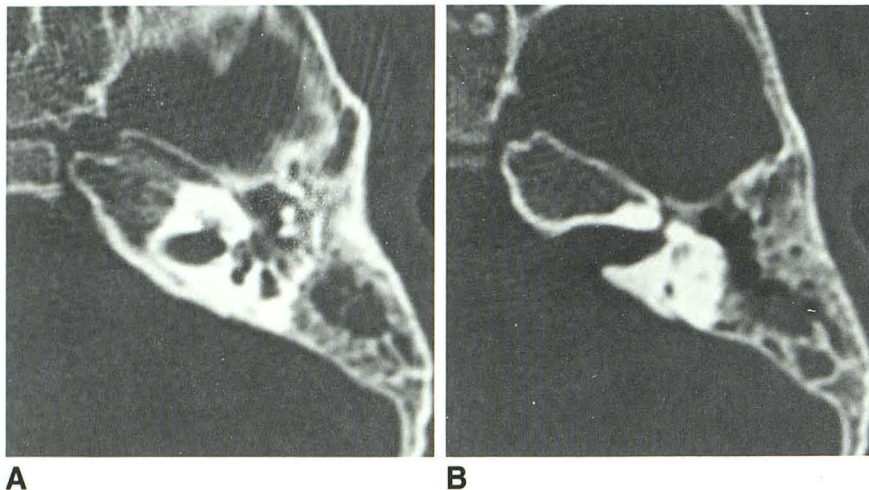


Fig. 1.—Axial CT scan of left ear at level of incudomalleal articulation shows hypoplastic and sclerotic cochlea (A) and vestibule (B). Internal auditory canal has a flared appearance. Note fluid in mastoid air cells, resulting from acute otomastoiditis.

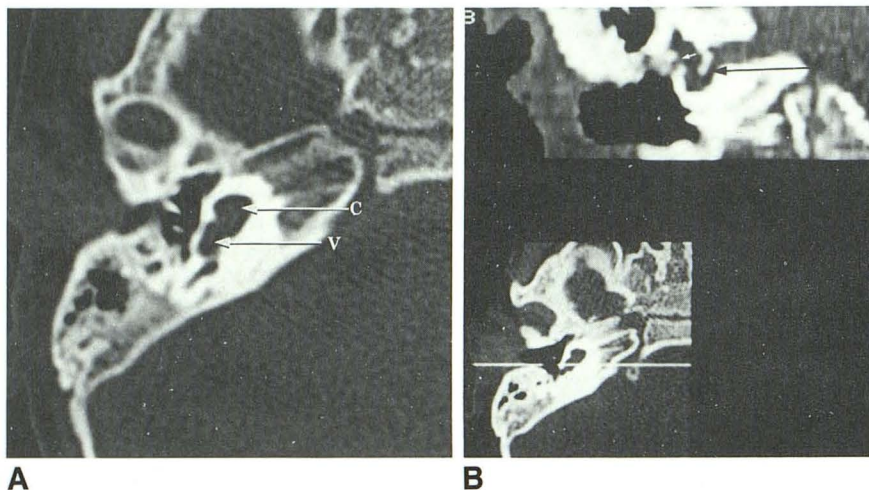


Fig. 2.—A and B, Right ear. Axial CT scan at (A) level of incudostapedial articulation (3 mm inferior to Fig. 1) and reformatted coronal scan (B) demonstrate a common vestibulo (V)-cochlear (C) chamber. Note incomplete fundus of internal auditory meatus with apparent communication to inner ear (black arrow in B). Oval window (white arrow in B) is normal size.

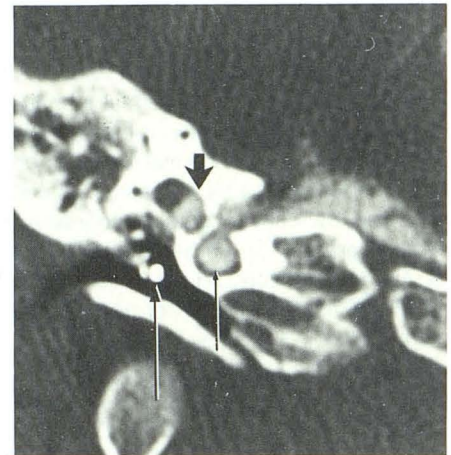


Fig. 3.—Direct coronal CT scan of right ear obtained immediately after lumbar intrathecal instillation of iohexol shows contrast material in internal auditory canal, cochlea (short thin arrow), and vestibule (wide arrow). Hyperdense rounded structure (long thin arrow) in region of tympanic membrane is a pressure-equalizing tube, previously inserted for recurrent otitis media. Note the fluid density in mastoid, caused by otomastoiditis or CSF.

of the inner ear. The marked labyrinthine sclerosis of the left ear may be of congenital origin or might be ascribed to labyrinthitis ossificans, which is a complication of either meningitis or otomastoiditis and cannot be totally excluded ([12] and Mafee MF, personal communication). Mafee et al. [9] reported a patient with bilateral Mondini deformity in whom the right cochlea was small and the semicircular canals were obliterated ("labyrinthitis ossificans and mild form of Mondini anomaly"); the left inner ear consisted of a cystic cochlea and dilated vestibule. The abnormalities in this patient seem identical to the anomalies in our patient. Because malformations of the inner ear may be bilateral but asymmetric, as in our patient, atypical, or due to unrelated causes, anatomic description of each ear is more useful than rigid categorization.

Recurrent pneumococcal meningitis may be due to hematogenous spread from a distant or contiguous site of infection, immune deficiency, or abnormal communication of CSF spaces with mucosal spaces (dermoid sinus, encephalocele, basal skull fracture, and CSF-middle ear fistula). Because 15% of patients will develop partial or complete sensorineural hearing loss after pneumococcal meningitis [13], a high index of suspicion is necessary to investigate the dysfunctional ear as a possible cause, rather than result, of meningitis. Schuknecht [1] suggested that, in Mondini dysplasia, the medial wall of the labyrinth is the most likely site of leakage; histologically, a thin "fragile-appearing" bony shelf may be found between the internal auditory canal and deformed cochlea. The lateral defect, through the oval window, is often associ-

ated with an anomalous stapes. Our patient demonstrated these typical defects in the labyrinth. Other potential sites of leak, such as the cochlear aqueduct, petromastoid canal, or facial canal, are found rarely in the Mondini deformity. Of 33 patients with Mondini anomaly in two series [1, 9], three had recurrent pneumococcal meningitis due to a CSF fistula.

Perilymph fistulas in patients with multiple episodes of pneumococcal meningitis have been demonstrated at surgery with the use of intrathecal fluorescein. Preoperative imaging with ^{111}In -DTPA or $^{99\text{m}}\text{Tc}$ -DTPA cisternography may confirm the presence of CSF otorrhea or rhinorrhea, but lacks spatial resolution and fails to identify precisely the site of leakage. In addition, if inflammation seals the middle ear, radionuclide cisternography may be falsely negative. Conventional tomography after intrathecal Pantopaque administration may accurately demonstrate the site of the fistula, but is associated with the risk of arachnoiditis caused by Pantopaque and with increased radiation dose from conventional tomography [14, 15]. Thin-section CT scans of the temporal bones demonstrate the anatomy of these structures precisely [15, 16]. The combination of contrast cisternography and thin-section CT elegantly identifies the site of a CSF fistula. Once immune deficiency and obvious communication of CSF spaces with the skin or nasopharynx have been excluded in patients with sensorineural hearing loss and pneumococcal meningitis, thin-section CT of the temporal bones is recommended to evaluate the labyrinth. If a congenital anomaly is identified, CT cisternography may precisely display the site of a CSF fistula.

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