Computer Generated Three-Dimensional Reconstruction of the Bony Labyrinth in Mondini’s Dysplasia

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Abstract

The bony labyrinth obtained at necropsy in four cases was studied by a new computer-generated three-dimensional (3-D) system. One case was normal (control) and the other three were histopathologically confirmed cases of Mondini’s dysplasia. In case 1, the cochlea had only 2 turns and the lateral semicircular canal did not make a circle but appeared as a spherical mass projecting from the utricle even though the posterior semicircular canal made a normal circle. In case 2, there were no turns in the cochlea even though the semicircular canals and the vestibule appeared normal. In case 3, the cochlea showed 1 to 1 and 1/2 turns and the semicircular canals were premature showing only bud-like projections. This 3-D imaging system, which utilizes the toggling method, provides a way of obtaining satisfactory images without markers, and the time required to obtain these 3-D images was reduced by using a video camera instead of a digitizer. One of the problems associated with the use of 3-D imaging is the long processing time. We resolved this by inputting the section images with a video camera and by picking up structures using density segmentation instead of tracing with a digitizer.

KEYWORDS: three-dimensional reconstruction, Mondini’s dysplasia, bony labyrinth, cochlea, semicircular canal

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Computer Generated Three-Dimensional Reconstruction of the Bony Labyrinth in Mondini's Dysplasia

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The bony labyrinth obtained at necropsy in four cases was studied by a new computer-generated three-dimensional (3-D) system. One case was normal (control) and the other three were histopathologically confirmed cases of Mondini's dysplasia. In case 1, the cochlea had only 2 turns and the lateral semicircular canal did not make a circle but appeared as a spherical mass projecting from the utricle even though the posterior semicircular canal made a normal circle. In case 2, there were no turns in the cochlea even though the semicircular canals and the vestibule appeared normal. In case 3, the cochlea showed 1 to 1 and 1/2 turns and the semicircular canals were premature showing only bud-like projections. This 3-D imaging system, which utilizes the toggling method, provides a way of obtaining satisfactory images without markers, and the time required to obtain these 3-D images was reduced by using a video camera instead of a digitizer. One of the problems associated with the use of 3-D imaging is the long processing time. We resolved this by inputting the section images with a video camera and by picking up structures using density segmentation instead of tracing with a digitizer.

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Computer-generated three-dimensional (3-D) reconstructions obtained from serial temporal bone sections is a useful method for visualizing the spatial shapes of the inner ear structures. Previous reports in the world literature have described the shape of the bony labyrinth, cochlea, semicircular canals, internal auditory canal, and the vestibular aqueduct and cochlear aqueduct with 3-D imaging from normal (1-5) but not pathologic temporal bones. In this paper, we report the 3-D reconstructions of three cases of Mondini's dysplasia from the Ear Pathology Research Laboratory (EPRL), University of Toronto. The use of the toggling method is described in the production of these images.

Materials and Methods

Serial sections of three temporal bones demonstrating Mondini's dysplasia from the EPRL, University of Toronto were evaluated. All bones were routinely harvested at necropsy, fixed in a 10% formalin solution, decalcified in 5% trichloroacetic acid and embedded in celloidin prior to horizontal serial sectioning at a thickness of 24 μm. Every fifth section was subsequently stained with hematoxylin and eosin (H&E).

Images of the sections were input using the AMICUS system (ISG Technologies, Mississauga, Canada). The AMICUS system restores video camera images and aligns them by the toggling method. The toggling method is used to compare different images quickly by flashing them alternately in rapid succession on the television monitor to align them as closely as possible.

All images were then transferred over a high speed Ethernet link to a 3-D volume investigation workstation, the ICAR 80.8 (ISG Technologies). The host platform utilized a UNIX-based silicon graphics workstation with a high speed proprietary graphic processor consisting of 8 TI 3152 DSP parallel processors, 16 megabytes of memory, and a high resolution (1024 × 1024 pixel) image display format.

This new technology allows the conversion of two-dimensional (2-D) arrays of selected pixels into 3-D voxel-based images. Areas of interest in each section are defined using density segmentation and outline is traced

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around each structure prior to computer-generated reconstruction. The 3-D voxel images can then be interactively displayed and rotated to obtain the preferred presentation of the image. We can select colors of individual structures, and adjust surface illumination to optimize and enhance surface details. The surface of a structure can be ‘cut away’ to reveal the spatial localization of adjacent or hidden components of the entire image. Volumetric information can be obtained for each structure and, finally, photographic slides can be produced from images transmitted across a high speed communications linked by directly printing the images onto photographic paper.

Results

Case 1

This 36-week gestational, 1,880 g male infant with trisomy 13 died four days after birth. Multiple congenital abnormalities were found including a high lumbosacral myelomeningocele and a related Arnold-Chiari malformation. A maternal history was not available. Both temporal bones demonstrated bilateral inner ear malformations compatible with Mondini’s dysplasia.

Histopathological findings. The histopathological findings in both ears were the same. An acellular mastoid filled with primitive embryonal cartilage and hematopoietic elements were noted. In all sections, the

Fig. 1 Midmodiolar sections of one normal case (A) and three cases of Mondini’s dysplasia (B-D). Abbreviations: C, cochlea; IAC, internal auditory canal; V, vestibule; S, saccule; ES, endolymphatic sac; ED, endolymphatic duct; LSC, lateral semicircular canal; PSC, posterior semicircular canal.
stapes appeared intact but was morphologically abnormal. The internal auditory canal (IAC) was abnormally wide and short. Neural elements, presumably of the cochleovestibular and facial nerves, were identified in the IAC surrounded by loose connective tissue and proteinaceous cosinophilic staining debris. The vestibule was relatively enlarged and demonstrated numerous cystic dilatations in the endosteal lining. A large endolymphatic duct was seen entering the vestibule posteriorly. Although the cochlea contained 2 turns, a rudimentary Corti's organ was identified only in the basal coil of the cochlea. The round window membrane appeared relatively normal. The cochlear aqueduct appeared patent (Fig. 1-B).

**Three-dimensional findings.** In the superioanterolateral view, the lateral semicircular canal appeared as a spherical mass projecting from the utricle. The superior semicircular canal appeared normal at the common crus, but was slightly enlarged at its ampullary end. The posterior semicircular canal appeared normal (Fig. 2-C). In the superior view, the cochlea was confirmed to have only 2 turns. The vestibular aqueduct was short and the proximal portion was wide. The cochlear aqueduct appeared funnel shaped (Fig. 2-D).

**Case 3**

This 2 and 1/2-year-old female died from acute sepsis and anoxic encephalopathy. She had been born with numerous congenital anomalies which included a tracheosophageal fistula, patent ductus arteriosus and an anomalous right subclavian artery. Throughout her short life, she had been plagued by recurrent bouts of bronchial pneumonia.

Prior to 1 year of age, her parents had suspected a bilateral profound deafness to be present. Behavioral observation audiometry and cortical evoked response testing at 16 months confirmed a bilateral profound sensorineural hearing loss. Only the left ear was available for histopathology.

**Histopathological findings.** The mastoid cells were observed with numerous rests of hematopoietic elements noted primarily in the petrous apex. The malleus and incus were present, but the stapes and oval window were conspicuously absent from the posterior mesotympanum. The facial nerve was dehiscent in its horizontal segment. The pars superior appeared dysplastic, and demonstrated a number of abnormalities including complete agenesis of the semicircular canals and the corresponding neuroepithelium in the membranous portion. Only a long spherical-shaped vestibule could be recognized demonstrating a presumed primitive utricle receiving innervation from the superior vestibular nerve. Only a single primitive basal coil of the cochlea could be identified. The stria vascularis and endosteal regions appeared hyperplastic with complete absence of Corti's organ and spiral ganglion cells (Fig. 1-D).

**Three-dimensional findings.** In the superioanterolateral view, the cochlea showed 1 to 1 and 1/2 turns. The semicircular canals were premature and showed only bud-like projections (Fig. 2-G). In the superior view, however, the vestibular aqueduct appeared normal and the cochlear aqueduct was longer and wider by comparison (Fig. 2-H).

**Discussion**

In 1791, Mondini originally described a case of congenital deafness which was attributed to an abnormal cochlea that was shortened to 1 and 1/2 turns and lacked a complete intercalar septum (6). Later histopathological studies were important in further documenting a spectrum of congenital inner ear abnormalities involving the bony and membranous labyrinth that has become better known...
Fig. 2  Three-dimensional reconstructions of one normal case (A, B) and three cases of Mondini's dysplasia (C–H). Each case is presented by superoanterolateral (left) and superior (right) views.
as Mondini’s dysplasia (7–8). Although temporal bone histopathology still represents the “gold standard”, polytomography and especially high resolution CT have allowed for ante-mortem diagnosis (9–10). Currently, 3-D CT images and magnetic resonance imaging (MRI) are used to observe the skull, but the slices are too thick to observe the tiny bony labyrinth.

In this study, we input every fifth section cut by 24-µm, but in some cases the surface of 3-D images was not smooth enough to visualize clearly. To obtain better resolution of 3-D images, we must minimize the aberrations in sections and input a greater number of section images into the computer system.

In this paper, we present three previously unreported cases of histopathologically confirmed Mondini’s dysplasia. The cochlea was shortened and the lateral semicircular canal did not make a complete circle in case 1. The cochlea made no turn even though the vestibule and semicircular canal were almost normal in case 2. The cochlea was shortened and all semicircular canals showed only bud-like projections in case 3.

Takagi et al. reported that measurements made by the conventional 2-D graphic reconstruction method can be distorted considerably by variations in the cutting angle of the specimen, but measurements made by use of the 3-D images can be performed regardless of the angle at which the specimen, but measurements made by the use of 3-D sections is the most accurate and useful method to observe the shapes of the inner ear structures because it provides volume, length, and angle from images taken at any angle as well as the histopathological findings. Unfortunately, since markers are indispensable for alignment, there are many sections stored in many laboratories which could not be evaluated using 3-D graphics until now.

We obtained satisfactory 3-D images of Mondini’s dysplasia from the serial sections without markers by using the toggling method. This method is used to compare the images of two sections by flashing them alternately in rapid succession on the display. In this way, we can compare the structures and easily obtain the proper alignment without the use of markers. The toggling speed can be adjusted as needed. Even if the serial sections have markers, we think this method will be also indispensable to reconstruct certain parts of the structures under magnification.

One of the problems associated with the use of 3-D imaging is the long processing time. We resolved this by inputting the section images with a video camera and by picking up structures using density segmentation instead of tracing with a digitizer.

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