Sonography of the Fetal Cochlea in the Early Second Trimester of Pregnancy

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Objectives—To examine the feasibility of imaging the fetal cochlea in the early second trimester.

Methods—This study included 42 healthy fetuses of low-risk pregnancies between 14 and 16 weeks. The coronal head sections via sagittal and lateral acoustic windows were used for cochlear visualization. The coronal plane was directed across the thalamus, oriented to the petrous part of the temporal bones. All fetuses were examined by 2-dimensional (2D) transvaginal sonography. Transabdominal scans were added in 11 fetuses at 16 weeks. Three-dimensional (3D) multiplanar reconstruction was used for coronal plane reformatting in cases with inappropriate head positioning for 2D scanning.

Results—Because of temporal bone hypomineralization in the early second trimester, sonographic depiction of the cochlear labyrinth was possible. On coronal imaging, the cochleas showed ringlike echogenic borders containing hypoechoic liquid content. The cochlear modiolus appeared as a central echogenic round spot (“cochlear target sign”). The cochleas were observed caudal to the temporal lobes, separated by the clivus. Shifting of the coronal plane showed progressive coiling of the basal cochlear turn toward the second turn (“cochlear whirlpool sign”). Two-dimensional transvaginal scanning showed at least 1 cochlea in 79% (33 of 42) of the cases. Adding 3D multiplanar reconstruction, at least 1 cochlear target sign was observed in 83% (35 of 42). The cochlear whirlpool sign on at least one side was shown in 57% (24 of 42). The maximal cochlear diameter was measured in fetuses with clear cochlear margin visualization. In this subgroup, the mean gestational age ± SD was 15.8 ± 0.5 weeks, and the mean maximal cochlear diameter was 6.4 ± 0.8 mm.

Conclusions—Prenatal sonographic inner ear depiction was previously considered impossible. Using a unique time window in the early second trimester, we achieved feasible fetal cochlear imaging.

Key Words—cochlear target sign; cochlear whirlpool sign; fetal cochlea; sonography
We present a study aimed at showing the fetal cochlea on prenatal sonography, which to our knowledge has not been reported previously. It was assumed that incomplete ossification of the temporal bones in the early second trimester will enable sufficient ultrasound penetration to achieve sonographic imaging of the fetal cochlea.

Fetal cochleas develop in a unique early accelerated pattern. These internal auditory organs look disproportionally large in a fetal head dissection at the end of first trimester (Figure 1). Jeffery and Spoor investigated fetal cochlear size by postmortem high-resolution MRI. They found that the prenatal labyrinth attains an adult-equivalent size between 17 and 19 weeks’ gestation. Nemzek et al measured the basal cochlear turn diameter in fetal specimens by postmortem computed tomography and MRI. The basal cochlear turn diameter ranged from 5 mm at 14 weeks to 9 mm at 24 weeks. It appears that the fetal cochlea in the early second trimester is large enough for measurement by sonography.

Anatomically, the cochlear labyrinth is located in the medioanterior area of the petrous part of the temporal bone, and the vestibular apparatus occupies its posterior region. The cochlear modiolus axis is directed anteriorly with the basal cochlear turn in its posterior end. This spatial orientation of the cochleas results in their typical coiled appearance on a coronal section through the fetal head (Figure 2B).

Most inner ear malformations arise when formation of the membranous labyrinth is interrupted during the first trimester of pregnancy. The arrested transformation of the otic vesicle into the normal cochlea and vestibule may be either a result of an inborn genetic error or a consequence of teratogenic exposure during the weeks of inner ear organogenesis.

Materials and Methods

This research was conducted as an observational cohort study. Approval for the study was given by the Institutional Review Board (Helsinki Committee protocol), including acquisition of informed consent for sonographic examinations from all patients.

Intending to show the fetal cochlea, we performed 42 sequential fetal scans at 14 to 16 gestational weeks. All patients had singleton low-risk pregnancies with normal fetal anatomy and growth. The examinations were performed with a Voluson 730 Expert ultrasound machine (GE Healthcare, Milwaukee, WI) using RIC 5-9H/OB endovaginal and RAB 4-8L/OB abdominal probes. Based on the cochlear orientation within the petrous part of the temporal bone (Figures 1 and 2B), coronal insonation planes were applied for cochlear imaging.

In the sonographic coronal plane, as in the pathologic dissection, the cochleas were shown in their typical coiled shape (Figure 2A). On the sonogram, the cochlear labyrinth showed ringlike echogenic borders containing hypoechoic liquid content. The cochlear modiolus appeared as a central echogenic round spot. The term proposed for this sonographic appearance was the “cochlear target sign.” The cochleas were observed caudad to the temporal lobes, separated by the clivus in the midline (Figure 2).

For depiction of the cochlear target sign via the anterior fontanel or sagittal suture, the coronal plane was directed across the thalamus, oriented to the petrous part of the temporal bones. On coronal scanning through the mastoid fontanel or the posterior part of the squamous suture, the cochlea enters the plane just posterior to the auricular edge (Figure 3). To minimize temporal bone shadowing, the insonation plane was oriented perpendicular to the mastoid fontanel and posterior part of the squamous suture.

All fetuses were scanned by 2-dimensional (2D) coronal head planes using the endovaginal probe. In addition,
Figure 2. A, Coronal head sonogram of a 16-week fetus. This image was obtained by insonation through the anterior fontanel on a transabdominal scan with the Voluson 730 Expert machine and the RAB 4-8L/OB probe. The cochlear target sign (co) is indicated. B, Corresponding pathologic image. The clivus is marked by the asterisk. In this fetus, the cochlear basal turn diameter was 7 mm, and the skull biparietal diameter was 36 mm, with a ratio of 0.18. See the measurements from the 13-week abortus in Figure 1 for comparison.

Figure 3. A–C, Sequential transvaginal coronal sonograms of the same fetus with 90° rotation. There is a progressive entrance of the cochlear structure (co) into the image during the shift of the insonation plane behind the fetal auricle (asterisk). Cochlear insonation in this case was achieved through the mastoid fontanel. The maximal cochlear diameter is the distance between the arrowheads in C.
In 11 of 42 cases, transabdominal scanning of the fetal head was performed. For the probe facing the lateral fetal head aspect, scanning was performed through the squamous suture and mastoid fontanel. For the probe facing the vertex, the scan was done via the anterior fontanel and sagittal suture.

In the cases with difficult coronal imaging by 2D scanning (usually due to inappropriate fetal head positioning), 3-dimensional (3D) sonography was used to create a proper coronal plane for cochlear depiction. It was achieved by a 3D multiplanar reconstruction display in the 4D View application (GE Healthcare). With this application, the fetal head was orientated in 3 standard orthogonal planes: multiplanar reconstruction plane A for the coronal section, multiplanar reconstruction plane B for the median section, and multiplanar reconstruction plane C for the axial section. In the median section, the fetal head was upwardly positioned at an angle of 20° to 30° between the clivus and vertical axis. This 3D multiplanar reconstruction display setup provides conventional identification of the left and right fetal head sides on axial and coronal sections. Continuing to operate in the median section, the multiplanar reconstruction navigation point was moved to the clivus at the level of the lower vermal edge (Figure 4). This step resulted in depiction of the cochlear target sign in the coronal section. For optimization of 3D multiplanar reconstruction cochlear imaging, the volume acquisition plane was as close to the coronal plane as possible.

Cochlea imaging by the 3D multiplanar reconstruction display was also tested by activation of the static volume contrast imaging mode using a minimal slice thickness (1–2 mm).

**Figure 4.** Three-dimensional approach to cochlear imaging in a 15-week fetus. **A,** The acoustic window of the mastoid fontanel and the squamous suture is shown by the maximal-mode rendering algorithm in the initial acquisition. **B,** Three-dimensional multiplanar reconstruction display of the same volume after standard head orientation (see “Materials and Methods”). The navigation point (red spot) is placed close to the clivus at the level of the inferior vermal margin (yellow dashed line). In this navigation setup, the cochleas (co) are shown in the coronal section.
Performing fine back-and-forth coronal plane shifting, we observed progressive coiling of the basal cochlear turn toward the second turn. We named this dynamic finding the “cochlear whirlpool sign.” This impressive imaging effect was achieved either by delicate probe movement on a 2D B-mode scan or by coronal slicing on the 3D multiplanar reconstruction display. Videos 1 and 2 show the cochlear whirlpool and target signs, respectively.

Our study parameters included the visualization rate for the fetal cochlear target and whirlpool signs in vertex and lateral head approaches. An additional study parameter was the maximal cochlear diameter. The method for measurement of the maximal diameter is shown in Figure 3C. The calipers were placed on the outer echogenic edges of the cochlear target sign. Its widest diameter corresponded to the size of the basal cochlear turn.

Results

Two-dimensional transvaginal coronal fetal head scanning showed at least 1 cochlea in 79% (33 of 42) of all cases. In 6 fetuses, the proper insonation plane was not achieved because of inappropriate head positioning, and in 3 other cases, the resolution was suboptimal because of a deep head location in the scanning plane. When the 3D multiplanar reconstruction display was used for reformatting of the coronal plane, at least 1 cochlear target sign was observed in 83% (35 of 42) of all cases. The cochlear whirlpool sign on at least one side was shown in 57% (24 of 42) of all fetuses. It was possible only by the transvaginal imaging, applying coronal plane shifting along the cochlear axis, as described in “Materials and Methods.”

When the probe faced the lateral fetal head aspect, cochlear scanning was performed through the mastoid fontanel and posterior part of the squamous suture. This lateral head scanning was achieved in all 42 fetuses. By such an approach, the proximal cochlea (close to the probe) was visualized in 83% (35 of 42) of all fetuses by combining the 2D and 3D multiplanar reconstruction imaging modes. Both cochleas were observed in 40% (17 of 42) of all cases. Depiction of the distal cochlea (remote from the probe) was less clear compared to the proximal one.

When the probe was oriented toward the upper head aspect, scanning was performed via the anterior fontanel and sagittal suture. Such an approach was technically feasible during the scans of 31 fetuses. In this subgroup of fetuses, unilateral and bilateral cochlear depictions were achieved in 62% (19 of 31) and 77% (24 of 31) of these cases, respectively. Cochlear images through the anterior fontanel and sagittal suture had lower quality compared to lateral head scanning.

Transabdominal cochlear imaging was less effective and had a lower resolution compared to transvaginal imaging. The cochlear target sign was shown in 54% (6 of 11) of the fetuses in abdominal scanning subgroup. Cochlear visualization was bilateral in most of them (5 of 6).

No cases of cochlear depiction were added by activation of static volume contrast imaging. However, it was our subjective impression that thin-slice static volume contrast imaging provided a more clear perception of the cochlear edges.

Measurement of the maximal cochlear diameter was applied only when clear cochlear margins were visualized. It was feasible in the subgroup of 31 fetuses with clear depiction of the proximal cochlea by the transvaginal lateral head approach (Figure 3C). In this subgroup of the fetuses, the mean gestational age ± SD was 15.8 ± 0.5 weeks, and the mean maximal cochlear diameter was 6.4 ± 0.8 mm.

Discussion

Bilateral severe congenital hearing loss in the human population appears in about 1 per 1000 live births. It is estimated that about half of the affected infants or children have hereditary deafness.5–8 Congenital membranous malformations of the cochlear duct are responsible for approximately 80% of the cases of congenital sensorineural hearing loss.4 In these malformations, the bony architecture of the inner ear is usually normal on computed tomography and MRI. Bony inner ear malformations represent the remaining 20% of congenital sensorineural hearing loss cases. In this group, the malformations can readily be diagnosed by computed tomographic and MRI analysis.9,10

In 1987, Jackler et al14 proposed one of the most widely accepted classifications of inner ear malformations. According to this radiologic classification, there are several distinct vestibulocochlear malformations: (1) complete labyrinthine aplasia, in which there is complete lack of inner ear development (Michel aplasia); (2) cochlear aplasia, characterized by a complete lack of cochlear development; (3) common cavity malformation, in which the cochlea and vestibule form a common cavity without internal architecture; (4) cochlear hypoplasia, in which a small cochlear bud contains a decreased number of turns; and (5) incomplete partition, characterized by a small cochlea with an incomplete or interscalar septum. The last two categories are related to Mondini-Alexander dysplasia in an earlier classification scheme by Omerod.11 Park et al12 reported outcomes of 46 pediatric patients with congenital inner ear malformations. Twenty percent of them had a diagnosis of absence of cochlear structures on radiologic studies of the temporal bones.
The distribution of causes of profound hearing loss in infancy includes environmental etiologies in 50% (viral infection by cytomegalovirus, rubella, measles, herpes simplex virus, neonatal jaundice, neonatal meningitis, prematurity, and ototoxic drugs). The remaining 50% of cases are of genetic origin. Of them, 70% are nonsyndromic genetic disorders (autosomal dominant deafness [DFNA] genes, autosomal recessive deafness [DFNB] genes, X-linked deafness [DFN] genes, and rare cases of mitochondrial inheritance). Abnormal connexin 26 protein is the most common cause of nonsyndromic deafness. Connexin 26 is a gap junction protein in the inner ear cells. Its function is important in potassium concentration regulation. Abnormal connexin 26 causes an improper potassium gradient. It results in the inability of hair cells to generate an action potential. Mutations in the gap junction beta 2 (GJB2) gene result in production of the nonfunctional connexin 26. It is related to about 50% of children with severe to profound nonsyndromic autosomal recessive hearing loss. Syndromic etiologies of congenital hearing loss are responsible for about 30% of cases. There are more than 400 forms of syndromic deafness, including Usher syndrome, Pendred syndrome, Alport syndrome, Waardenburg syndrome, Jervell and Lange-Neilsen syndrome, and branchio-oto-renal syndrome.

Our study represents a new approach to prenatal imaging of the cochlear labyrinth. We used a unique opportunity in the early second trimester when the hypomineralized fetal temporal bone provided an acoustic window for sonography of the cochlea. Because of its rapid growth in the late first and early second trimesters, the fetal cochlea appears as an observable organ in the coronal transpetrosal plane of the fetal head.

We achieved a high feasibility rate of fetal cochlear depiction using coronal head scanning through the mastoid fontanel or squamous suture. Interesting results were retrieved from the subgroup of 31 fetal scans in which the heads were in the appropriate position for coronal insonation and were located in the optimal focal zone of the sonographic plane. In these fetuses, the cochlear target sign (at least on one side) was always observed. This observation is very reassuring because nonvisualization of the cochleas in our study was related to technical imaging obstacles. For this reason, repeated examinations and fetal head manipulation might improve the cochlear depiction rate. The most important limitation of sonographic fetal cochlear imaging is ossification of the temporal bones. It causes acoustic shadowing, which hides cochlear structures in fetal scans at more advanced gestational ages.

On the basis of our results, we hypothesize that early second-trimester cochlear sonography may be of value in the diagnosis of cochlear aplasia. This malformation may appear as a familial autosomal dominant or familial autosomal recessive disorder. It had also been reported with the Klippel-Fiel anomaly, Wildervanck syndrome, thalidomide embryopathy, and anencephaly. A complete lack of inner ear development may be bilateral or unilateral. It may be associated with other abnormalities caused by failure of the otic placode. These include middle ear anomalies, hypoplasia of the petrous bone, and abnormal courses of the transverse sinus and jugular veins.

The much more complicated question is whether our data may be helpful in the diagnosis of the cochlear hypoplasia (Mondini-Alexander dysplasia). As mentioned in the classification of Jackler et al,4 cochlear hypoplasia is characterized by a small cochlear bud with a decreased number of turns. Normal human cochlear coiling consists of 2.5 turns. In a pathologic dissection of the fetal inner ear, complete coiling is observed at 8 to 9 weeks’ gestation. Depiction of all cochlear turns was impossible in our study. In the subgroup of the 31 scans with optimal cochlear visualization, the cochlear whirlpool sign was observed in 77% (24 of 31) of the fetuses. However, we could not count more than 2 cochlear turns, which means that incomplete cochlear rotation is an unreliable finding on cochlear sonography.

The maximal cochlear diameter in our study was related to the size of the basal cochlear turn and showed good correlation with the results of Nemzek et al.3 However, our measurements were not confirmed in a case-control study including fetuses with cochlear hypoplasia. For this reason, the results for the maximal cochlear diameter have limited value in the diagnosis of fetal cochlear hypoplasia.

Summarizing our findings, we can conclude that fetal cochlear imaging in the early second trimester is feasible if high-resolution sonography is used and proper coronal fetal head scanning is performed.

References