

Hearing in the Fetus: Prenatal Detection of Deafness

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Abstract

A profound hearing loss occurs in approximately 2% of all births. This has marked effect on the development of language, communication and social skills. At present there is no reliable technique for the detection of deafness at birth and confirmation of the diagnosis may not be obtained until two years of age. It is extremely important to diagnose deafness as early as possible, to allow compensatory communication strategies to be instigated and therefore decrease the subsequent negative effects of deafness.

The following paper discusses the development of fetal responsiveness to sound with gestation. Secondly it discusses the possibility of developing a screening tool using the response of the fetus to both light and sound for the prenatal detection of deafness.

Zusammenfassung

Eine ausgeprägte Verminderung des Hörvermögens liegt bei etwa 2% der Neugeborenen vor. Eine solche Beeinträchtigung hat deutliche Auswirkungen auf die Sprachentwicklung, die Kommunikationsmöglichkeiten und die sozialen Fähigkeiten. Zur Zeit gibt es keine verlässliche Technik, Taubheit bei der Geburt festzustellen und diagnostische Sicherheit wird oft erst im Alter von zwei Jahren erreicht. Es ist jedoch äußerst wichtig, die Taubheit so früh wie möglich festzustellen, um kompensatorische Maßnahmen einzuleiten, die die negativen Effekte der Taubheit vermindern sollen.

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Die folgende Arbeit diskutiert die Entwicklung der Antwortbereitschaft auf Geräusche beim Föten während der Schwangerschaft. Dann wird die Möglichkeit diskutiert, ein diagnostisches Mittel zur pränatalen Aufdeckung von Taubheit zu entwickeln, indem man die Reaktion des Föten auf Licht und Geräusch ausnutzt.

Introduction

It is an unfortunate fact of life that approximately 2% of all births are born with a profound hearing loss. This may in fact be an underestimate as many cases are not identified until two years of age. In the past it was thought that the fetus was born deaf and hearing developed during the postnatal period. The question which then needed to be answered was "Can the fetus hear?". It has been recognised since biblical times that the fetus responded to sound.

For behold when the voice of your greeting came to my ears, the babe in my womb leaped for joy.

Luke 1:44

In 1925 Peiper reported increased maternal movement in response to a car horn¹. Since then a wide variety of studies have been carried out to document the hearing abilities of the fetus².

Hearing is the primary sensory modality for speech and language and therefore provides a basis for both social and academic achievement. Hearing loss during childhood has been shown to have a very marked negative effect on the development of speech, language and learning³. As a result of these, severe behavioural and communication problems may arise.

The National Centre of Health Statistics, 1982, estimated a prevalence of 1.63% deafness in children up to the age of fourteen years. The cause of deafness may be divided into three main groups⁴. First, conductive hearing loss which involves the interference of reception of sound by the external ear or transmission from the external ear to the inner ear. Second, sensorineural hearing loss caused by abnormalities of the cochlear hair cells of the auditory nerve. Third, mixed hearing loss when both a conductive and sensorineural hearing loss are present.

Early identification of hearing loss is fundamental to its effective management in infants. Although it may not be possible to correct the hearing loss it is possible to employ many compensatory communication strategies to reduce the negative effects. Often hearing loss is not identified in the neonatal period and confirmation may not be reached until 24 months of age after the critical stages of early language development should have occurred. At present the only screening procedures available observe the behavioural responses to calibrated noise makers, auditory brain stem evoked responses or automated infant hearing screening devices⁵. It is not usually possible to carry these out until six months of age⁵.

The following paper has two aims:

1. to document the hearing abilities of the fetus and its development. By establishing a picture of hearing in the normal fetus it should be possible to detect abnormalities in hearing performance which may be indicative of a hearing problem.
2. to develop a screening tool which can be used during pregnancy to assess whether the hearing of the fetus is impaired. This may be used to screen fetuses of parents who were known carriers of genes affecting hearing or in cases where some environmental insult had affected the fetus.

General Methods

Subjects

Subjects fell into two main categories: first, a group who were part of a larger study known to have a normal pregnancy with no known medical complications or family history of hearing deficits; second, a group who had a deaf parent due to a genetic condition. All mothers were fully informed about the research and consent was obtained. Ethical approval had been previously obtained from the Research Ethical Committee, The Queen's University of Belfast.

Stimuli and Apparatus

The stimulus used was a pure tone sine wave of varying intensities and frequency. The sine wave was produced by a Wavetek model 75 waveform generator. The signal was then fed into a AKG speaker which could be held over the mother's abdomen. The light stimulus was a cold halogen light, giving a narrow beam of light which again could be placed in close proximity to the maternal abdomen.

Procedure

The same initial procedure was adopted for all fetuses in each experiment. Mothers lay in a semi-recumbent position and the fetus was visualised by ultrasound scan using an ATL Ultramark 4 plus scanner with a 3.5 MHz curvilinear scanhead. A routine anomaly scan was carried out to confirm the dating of the pregnancy and to rule out any major abnormality. The position of the head of the fetus was defined and the stimulus applied over this area. The upper body, head, arms and torso were viewed at all times by moving the scanhead as required. The fetus was said to have responded if there was movement visualised of the upper body, arms or head.

Experiment 1

Subjects and Methods

In the following experiment 400 fetuses were examined every 3 weeks from 8 weeks of gestational age. Pure tone sine waves were used to stimulate the fetus and presented to the fetus by means of a 3" speaker held on the mother's abdomen directly over the fetus' head. For each frequency, the stimulus was presented initially at 70 dB for a 2 second period with an inter-stimulus interval of

5 seconds and the response of the fetus observed. If the fetus moved within two seconds of the stimulus onset then it was considered to have "sensed" the stimulus. The dB level was increased by 5 dB for each successive presentation of the tone until the fetus responded.

Results

The graph (Fig. 1) reports the mean intensity level required to elicit a response for each frequency at each age of testing. The results indicate the fetus responds first to 250 Hz frequency and its "hearing" range expands as it develops. Furthermore as gestation proceeds the fetus becomes more sensitive to the stimuli of different frequencies.

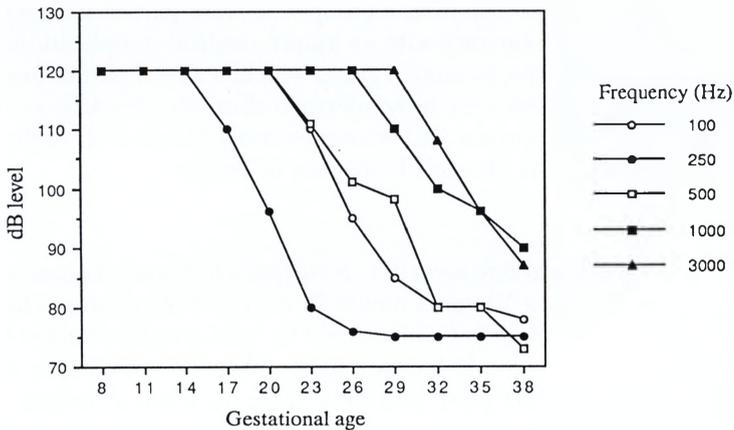


Fig. 1. The mean intensity level in decibels (dB) required to elicit a response from the fetus at each gestational age for pure tone sine waves.

Experiment 2

One of the major problems in attempting to diagnose deafness antenatally is the high false positive rate due to the non-responsiveness of the fetus to sound. For example, in a recent experiment 100 fetuses (34–37 weeks gestational age) were stimulated with a 250 Hz pure tone sine wave at 100 dB for 5 seconds on two occasions separated by 5 minutes.

On the first trial, 23 of the 100 fetuses failed to respond, this showed some improvement on the second trial although 14 still showed no response. Obviously not all these fetuses are deaf (postnatal observations confirmed this) which suggests that a single exposure to a sound is not specific enough to discriminate deafness from other causes of non-responsiveness, e.g. inappropriate behavioural state, sleep.

In order to overcome the problems of non-responsiveness we have used a combination of visual and auditory stimuli, using the fetus' tendency to move

when presented with either a light or sound directly on the abdomen over the head.

Subjects and Method

404 fetuses made up the total experimental group which was then divided. Group 1 consisted of 4 fetuses with parents suffering from hereditary deafness. Group 2 consisted of 400 fetuses from normal, uncomplicated pregnancies with no family history of congenital deafness. The fetus was visualised on ultrasound scan and the mother presented with a 250 Hz tone at 105 dB, duration 2 seconds. If the fetus moves then the sound test is repeated to ensure that this was not a coincidental occurrence. If no movement is observed, then the light (a halogen cold light) is shone on the abdomen directly above the fetus' head. If the fetus responds to the light then the sound stimulus is re-administered. If no response to the sound is observed then the light is applied for a second time. If the fetus responds to the light but not the sound then a possible diagnosis of deafness is entered. The test would then be repeated a week later. If no response to either light or sound is observed then the test would be repeated one week later.

Results

Using this technique, four fetuses with parents suffering from hereditary deafness have been examined. Three were found to respond to the sound and light on each occasion and were confirmed after birth to possess normal hearing, the fourth responded to the light but not to the sound on any occasion and was subsequently confirmed after birth as being congenitally deaf. Furthermore, one fetus from a study group of 400 from otherwise normal pregnancies were found not to respond to the sound on any occasion but each time did respond to the light. On further investigation after birth this fetus were found to be deaf. All the other fetuses were proven to have normal hearing.

Discussion

This paper concludes that although previous studies have stated that the fetus cannot "hear" until 24 weeks of gestation⁶ responsiveness to sound is found in these experiments at 16 weeks of gestation. Responsiveness is first exhibited to frequencies of 250–500 Hz. The results indicate that even though the anatomical development of the ear is not complete at this gestation the fetus is able to perceive and respond to external auditory stimuli earlier than thought. The range of hearing before birth appears wider than for the newborn². Using the combination of stimulation of the fetus with both sound and light overcomes the unresponsiveness we have observed to sound alone, this technique may be used in the antenatal detection of deafness. If detection of deafness could successfully be carried out in the prenatal period it would enable compensatory communication strategies to be employed which could greatly reduce any deficit which result from the hearing loss. The earlier the deafness can be detected the maximum benefit may be obtained.

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