

AUDITORY CHANGES ASSOCIATED WITH MULLERIAN ABNORMALITIES

(Received 6 January, 1997)

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ABSTRACT

Objective: A spectrum of auditory changes with mullerian defects has been reported. In a prospective controlled study, we aimed to assess hearing function of women with mullerian abnormalities.

Methods: Thirty women with mullerian defects and 50 women with normal genital tract as a control group were evaluated. Audiologic evaluation was performed to each subject which included pure tone audiometry 250-8000 Hz for air conduction, 500-4000 Hz for bone conduction measurement.

Results: Twenty three of 30 patients manifested a notch at 4 or 6 kHz with a magnitude between 10 to 30 dB. In one patient there was a slight sensorineural hearing loss at 500-1000 Hz. In 15 of 50 subjects in the control group a notch of 5-25 db at 4 or 6 kHz was demonstrated. The incidence of auditory defects was significantly higher in mullerian abnormalities group (80% vs 30%) ($p < 0.01$).

Conclusion: Auditory changes in high frequencies may exist in association with mullerian abnormalities.

Key Words: Mullerian abnormalities, Audiologic changes

INTRODUCTION

Mullerian tract abnormalities result from partial or complete failure of the mullerian ducts to fuse or canalize. This can lead to a variety of developmental defects: unicornuate uterus, septate uterus,

bicornuate uterus and didelphic uterus with or without a septate vagina. Several case reports describe an association of congenital deafness, vaginal agenesis and unilateral renal agenesis (1,2). A spectrum of auditory changes with mullerian defects has been reported (3). In an attempt to investigate the possible auditory defects associated with mullerian tract abnormalities, we performed audiologic evaluation to assess hearing function of women with mullerian defects and compared with controls.

MATERIAL AND METHODS

Patients: Thirty women with mullerian defects underwent audiological evaluation to assess their hearing function. Twenty seven patients had mullerian defects detected on hysterosalpingography and confirmed by laparoscopy, conducted as part of an evaluation for infertility or habitual abortion. Three patients had vaginal septums evident on pelvic examination. A control group of 50 women was chosen from patients who underwent cesarean section in our obstetrics department. Those patients demonstrated a single uterine cavity on operation and normal vagina at pelvic examination. All the patients were otherwise healthy with negative past medical histories.

Audiometric testing: The audiologic tests were performed by a professional audiologist in Industrial Acoustics Company sound treated booth with interacoustic AC 5 audiometer with Telephonics 39 earphones calibrated according to the ISO 389 standards. For immittance measurement interacoustic AZ 7 with AG3 recorder was used. Prior to the testing, patients were examined by an ENT specialist

and otoscopic examination was normal in all subjects. One ear of each patient was randomly chosen. The test included pure tone audiometry 250-8000 Hz for air conduction, 500-4000 Hz for bone conduction measurement.

Speech audiometry was routinely applied. Static compliance, tympanometry and stapedial reflexes were measured in all patients.

Statistical analyses: Differences in ages between the groups were analyzed with Student's t test. Differences in the frequency of audiometric abnormalities were analyzed with Fisher's exact test.

RESULTS

Thirty women with mullerian tract abnormalities and 50 women with normal mullerian ducts were evaluated. The average age was 27.6 ± 4.5 (range 21-36) and 27.2 ± 3.8 (range 22-35) for the experimental and control groups. Mullerian abnormalities included septate uteri (14 patients), bicornuate uteri (10 patients), vaginal septum (3 patients), unicornuate uterus (1 patient), uterus didelphys (2 patients). Bone conduction and electroacoustic immittance tests verified normal middle ear functions in all subjects. Twenty three of 30 patients (76.6%) manifested a notch at 4 or 6 KHz with a magnitude between 10 to 30 dB. Figure 1 demonstrates an audiogram of a patient with bicornuate uterus. In one patient there was a slight sensorineural hearing loss at 500-1000

Hz. In 15 of 50 subjects (30%) in the control group a notch of 5-25 dB at 4 or 6 KHz was demonstrated. Five patients had a notch of 5 dB whereas only 2 patients had a 25 dB notch. The incidence of auditory defects (80% vs 30%) was significantly higher in mullerian abnormalities group. ($P < 0.01$). The defects in audition could not be accounted for by age or occupation related changes.

DISCUSSION

In 1946, Potter (4) described an association of certain facial abnormalities and large, low set ears with proportionally little cartilage, with renal agenesis. Of the 20 neonates she investigated, 17 were male and 3 were female. It is noteworthy that in all 3 of the female, vaginal agenesis was present. Williensen (2) reported the association of the Mayer Rokitansky syndrome with conduction deafness, renal skeletal and facial anomalies. In 1991 Letterie and Vauss (3) reported significantly higher auditory defects in women with mullerian abnormalities when compared with the control group. Thirty three percent of the patients manifested sensorineural defects in the high frequency range.

The results of the present study suggest that a spectrum of auditory changes may exist in association with mullerian abnormalities. Auditory changes were evident in 80% of the patients with mullerian defects, whereas only 30% of the control group demonstrated auditory changes.

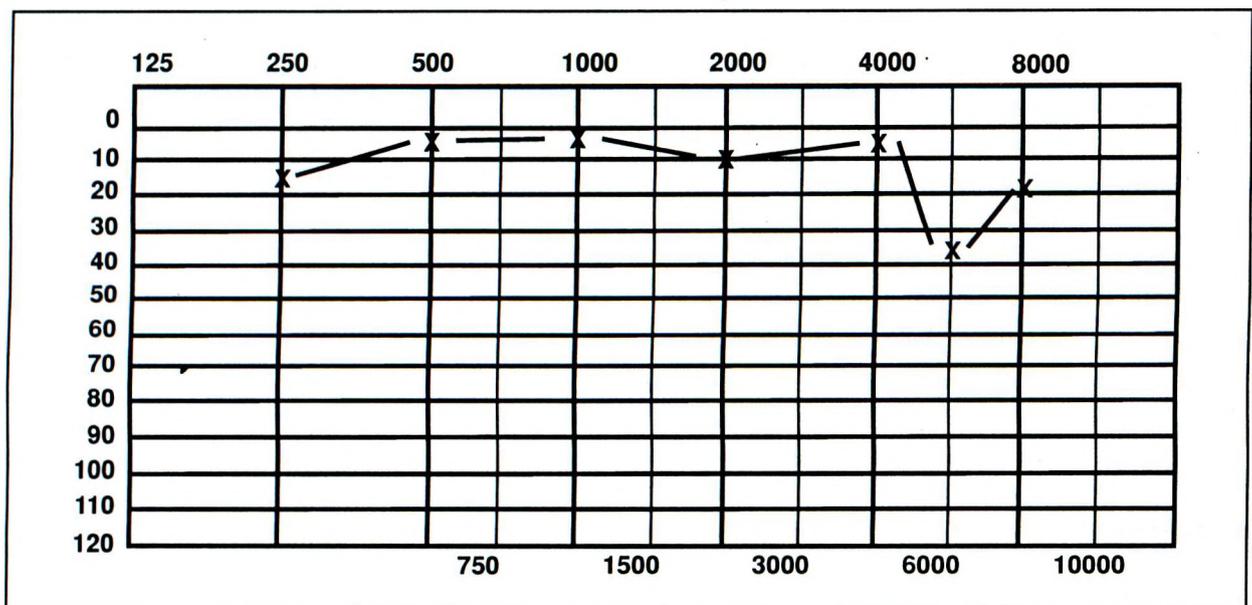


Fig. 1: Left ear audiogram of a bicornuate uterus. There is a significant notch at 6 KHz.

The threshold changes noted in the affected patients were in high frequencies, ranging from 4000 to 6000 Hz, showed a specific configuration which were not related to middle ear pathology. Polygenic and multifactorial inheritance were the previously suggested etiologies of these multisystem defects. Such an inheritance pattern implies that a disorder is caused either by the additive effect of several genes (polygenic) or by the interaction of genetic and environmental factors (multifactorial).

Data from the present study suggest that auditory changes in high frequencies may exist in association with mullerian abnormalities. Such auditory defects may not be clinically evident and may require sensitive audiometric testing to detect. An audilogic evaluation may be performed in patients with mullerian abnormalities. If auditory changes are

detected, periodic audiometric evaluation of these patients will be reasonable, to monitor for possible deterioration in the future.

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