HEARING IN CHILDREN WITH FETAL ALCOHOL SPECTRUM DISORDER (FASD)

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ABSTRACT

Background
Alcohol is the most prevalent human teratogen affected by early exposure of the fetus. Although not listed as a major part of the fetal alcohol spectrum disorder (FASD), different texts list hearing loss as a characteristic of the FASD, based on several small studies.

Objective
To characterize hearing in children with FASD, diagnosed in the Motherisk Program in Toronto.

Design
Cross sectional cohort study.

Setting
Academic referral center.

Patients
Children 4-16 years of age that met criteria for FASD, with no other known risk factor for sensorineural hearing loss. A consecutive sample of 41 children (13 girls, mean age 8.9±3 years) was collected. *Intervention:* Physical examination, audiometry and tympanometry. *Outcome measures:* External and middle ear pathology on physical examination, pure tone average (PTA), speech reception threshold (SRT), discrimination and tympanometry. Results were compared to reference values in the normal population. Hearing loss equal or greater than 16dB hearing-level in each frequency tested was considered to be clinically significant.

Results
A total of 5 (11.2%) of children had hearing loss of at least 16dB hearing–level, mostly unilateral. SRT was within the normal range in 40 (98%) of children with FASD and discrimination was normal in all children. None had auricular or external canal dysmorphology. 14.7% of the children had frequent episodes of acute otitis media. Middle ear effusion was detected in 8 ears (9.8%).

Conclusions
The prevalence of mild sensorineural hearing loss in children diagnosed with FASD (16dB hearing-level or greater) was not higher than expected in this age group. However, because children with FASD are academically and behaviorally challenged, early detection of hearing loss and early intervention is warranted.

KeyWords: *Fetal alcohol spectrum disorder, fetal alcohol syndrome, hearing, children, audiology*

Alcohol is the most prevalent human teratogen. Heavy drinking during pregnancy can manifest as Fetal Alcohol Syndrome (FAS), consisting of a triad of pre and/or postnatal growth retardation, central nervous system damage and characteristic facial dysmorphology.[1,2] Brain insult

may affect cognitive function and behavior.[3] The incidence of the full blown syndrome has been estimated at 0.97 cases per 1000 births [4] but the majority of children of alcohol dependent women do not display the full syndrome. The rate of Fetal Alcohol Spectrum Disorder (FASD) in the Western world is estimated at 10 cases per 1000-birth[5] that is almost 1% of the pediatric population.

In animal studies, rat offspring exposed *in utero* to diets containing 17.5% or 35% ethanol-derived calories developed sensorineural or central hearing loss.[6] Hearing loss (HL) greater than 20dB was documented in full-blown FAS.[7-9] In small studies 61% of children with FAS had a conductive hearing loss and 22% have sensorineural or central hearing loss.[9] However, the sampling procedures of these patients were not clear.

Because hearing is a critical factor in speech development and learning abilities, early diagnosis of hearing loss in these children is an important public health issue. Our objective was to quantify the prevalence of hearing impairment in children diagnosed with FASD and to compare them to general population incidence.

METHODS

The study was designed as a cross sectional cohort. The cohort included all available children seen by the Motherisk Clinic in the Hospital for Sick Children, Toronto, Canada, that were diagnosed, according to the Institute of Medicine criteria, by full morphological, psychological and behavioral assessment as having FASD between 2001 and 2004. The age range was 4-16 years. Children are referred to our clinics mostly because of complex neuro behavioral dysfunction coupled with history of problem maternal drinking. No child was referred because of known hearing problems.

The exclusion criteria included: children younger than 4 years old (due to lack of reliability of their audiometric results), children with known sensorineural hearing loss (SNHL) or at risk for SNHL and children that presented stigmata or other findings associated with a syndrome known to include SNHL. The audiological outcome was compared to data regarding hearing loss prevalence of 15% among children 6-9 years of age according to the Third National Health and Nutrition Examination Survey.[10] A questionnaire was completed for each child presented to the clinic. The questionnaire included data on demographics, socioeconomic status, known pregnancy data (detailed alcohol/drug exposure, smoking, intrauterine infection), subjective perception of hearing as described by the child and the parents/care-givers, past and present medical history and events of secretory, acute or chronic otitis media. Physical examination of each child included general and neurological examination, growth assessment, anthropometric measurements, otoscopy, audiometry, and tympanometry.

Audiometry

Each patient was tested in an acoustically treated room and given a pure-tone air-conduction-threshold examination with a Grason-Stadler GSI 61 audiometer, which is calibrated regularly. EAR 3A insert phones were used. Pure tone bone conduction thresholds were tested when air conduction thresholds were poorer than 20 dB HL. The bone conduction thresholds were assessed with mastoid placement of the oscillator and contralateral narrowband noise masking where the child was able to perform the task. Pure tone stimuli were obtained at octave intervals from 250-8000 Hz for air conduction, unless there was 20-dB difference between octaves, whereby the interoctave was tested. Bone conduction thresholds were tested from 250-4000 Hz.

An attempt was made to obtain data in each ear at each frequency before progressing to the next frequency listed. The same priority was given for bone conduction stimuli, with the exception of 8000 Hz. Spondee thresholds and word recognition measurements using monitored live voice and W-22 or PBK word lists depending upon the child's language level were completed. In order to be sensitive, hearing loss greater than 16dB HL in each frequency tested was considered significant [10] Tympanometry was performed on all children (GSI 33 or GSI TympStar tympanometer) to assess middle ear pressure, tympanic membrane compliance, and ear canal volume.

We compared results of children with FASD to normal auditory values by comparison statistics using parametric and non parametric tests according to type of data. The effect of smoking and use of drugs (i.e. marijuana, cocaine etc.) was evaluated by sub-analysis.

RESULTS

Forty-one consecutive children (13 girls and 28 boys) who were assessed psychologically and behaviorally were confirmed to have FASD after full physical and neuropsychological examination. Patients' characteristics including alcohol and drugs of abuse exposure, and physical parameters are delineated in Table 1.

Data regarding the children's ears and hearing including subjective hearing, history of acute otitis media, middle ear effusion, chronic otitis media, or past ventilation tubes were collected in 34 children and is delineated in Table 2.

TABLE 1 Characteristics of 41 Children Confirmed with FASD

		Number / Average ± SD	%
Age		8.9 ± 3.0	
Social Parameters			
	Custody		
	Adoption/Foster home	39	95.1
	Children Aids Society	2	4.9
	Biological parent	0	0
	Years in foster care	5.1 ± 3.2	
	Other drugs of abuse during pregnancy		
	Unclear	25	61.0
	Suspected	3	7.3
	Positive	13	31.7
Physical Parameters			
	Height percentile (N=22)	40.4 ± 26.5	
	Weight percentile (N=22)	44.3 ± 32.2	
	Head circumference percentile (N=26)	38.6 ± 30.7	
	Palpebral fissure length percentile (N=25)	72.4 ± 27.6	
	Philtrum length percentile (N=25)	34.6 ± 16.3	

TABLE 2 Ears and Hearing Characteristics

		Number	%
Subjective Hearing (N=35)			
	Very good	5	14.3
	Good	29	82.9
	Fair	1	2.9
Acute Otitis Media (N=34)			
	Never	22	64.7
	<3 in 6 months	7	20.6
	>3 in 6 months	5	14.7
Present or Past Ventilating Tubes (N=34)			
	No	29	85.3
	Yes	5	14.7
Secretory Otitis Media (N=34)			
	No	26	76.5
	Yes	2	5.9
	Not known	6	17.6
Chronic Otitis Media (N=34)			
	No	33	97.1
	Yes	1	2.9

All 41 children who had confirmed diagnosis of FASD had their hearing and their middle ear status assessed. On physical examination 73 ears out of 82 (91%) were normal while eight (9.8%) had middle ear fluid and one (1.2%) had a perforated tympanic membrane. The average bone and air audiograms are shown in Figure a and b respectively.

FIG. A Average Bone Conduction Audiogram for Children Diagnosed with FASD

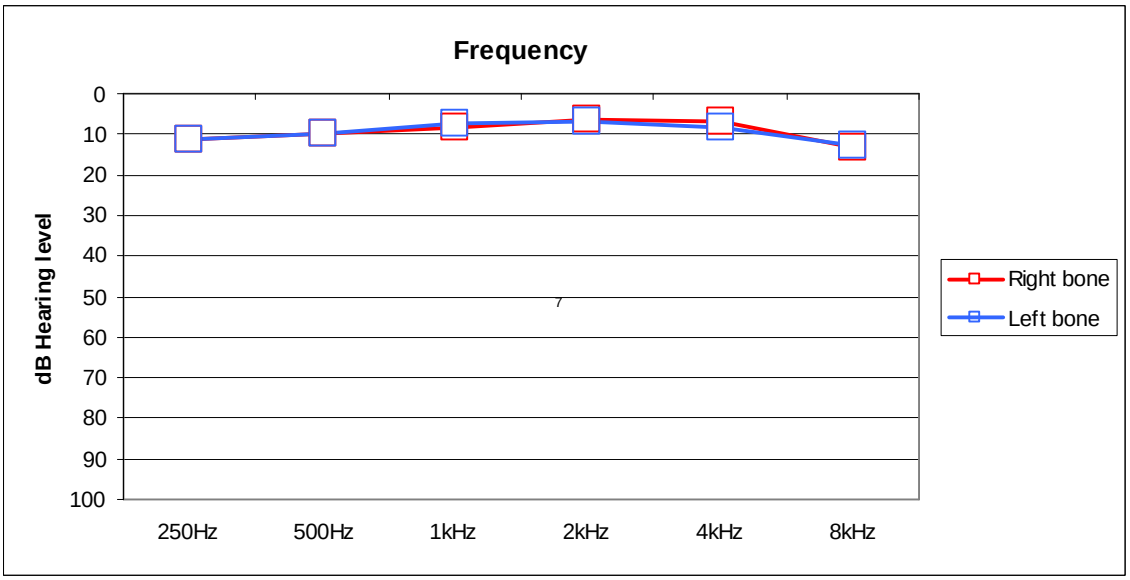
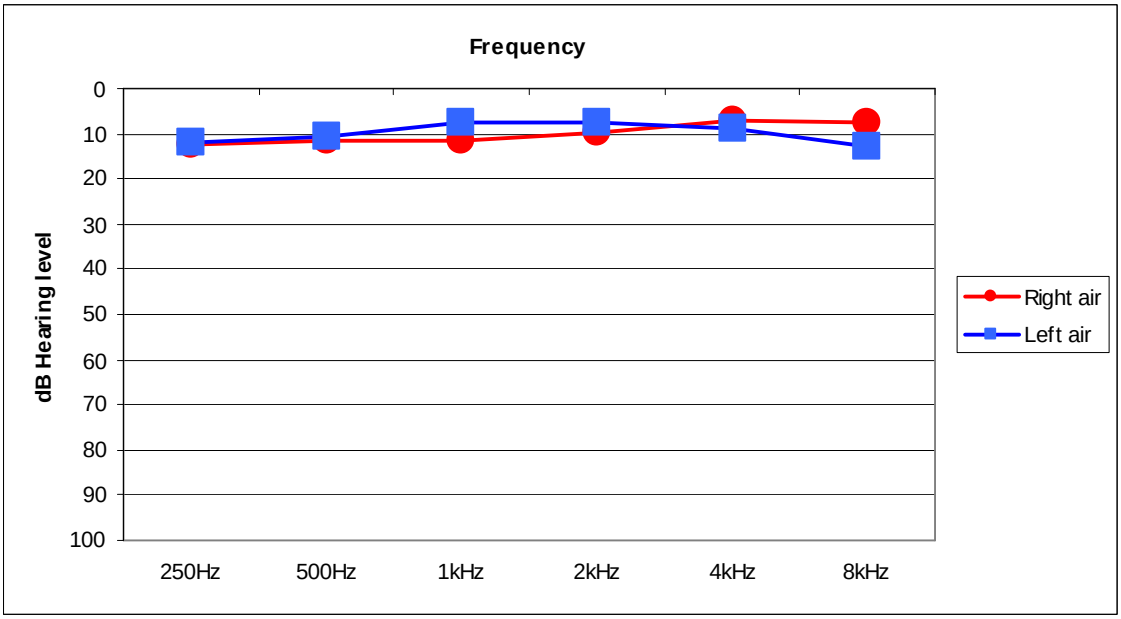


FIG. B Average Air Conduction Audiogram for Children diagnosed with FASD



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The Speech Reception Threshold (SRT) and speech discrimination data are presented in Table 3. Ten ears (12%) were found to have Pure Tone Average (PTA) HL greater then 16 dB HL in air conduction while five ears (6 %) had PTA HL greater then 16 dB in bone conduction, all as mild hearing loss. Comparison of these results showed no difference from the average hearing in a comparable age population which has been reported at around 15%.[10]

TABLE 3 Average Speech Reception Threshold and Speech Discrimination (n=41)

	Right	Left
SRT (dB ± SD)	9.02 ± 7.3	8.4 ± 6.6
Speech Discrimination (% ± SD)	98 ± 3.0	95 ± 15.9

DISCUSSION

Children affected by alcohol during fetal life can potentially present with a wide range of signs, symptoms, and adverse psycho-behavioral effects. The diagnosis of FASD is based on the presence of complex cognitive, behavioral and physical symptomatology among offspring of mothers who drank excessively in pregnancy.[11] Although dysmorphic features may occur in some children[12], the pathognomonic facial features are commonly absent.[13] Likewise, defects in sensorineural pathways such as the visual system[14] and the auditory system reported in association with FAS have not been yet recognized as a major characteristic of FASD.[15]

Three types of hearing disorders have been associated with FASD in several studies: delay in auditory maturation, sensory hearing loss and intermittent conductive hearing loss due to recurrent serous otitis media (RSOM).[16,17]

Church and colleague[6,16,18,9] reported 28% SNHL (10 out of 36 children with FAS) from two clinics. Notably, 12 of those children had also a cleft palate but only 2 had SNHL. Most of the cases with SNHL did not have sub-mucosal or overt cleft palate. These rates are similar to those seen in other children with craniofacial anomalies. Rossing and colleagues[9] reported 7 % incidence of SNHL in a group of 29 patients with FAS.

Conductive hearing loss (CHL) due to RSOM was reported by Church et al in 85% of children with FAS. However, 16 children out of 36 had cleft palate and 14 out of those had persistent SOM. RSOM was also diagnosed in 80% (16 out of 20) of children without cleft palate.[16,18,19] Rossing et al reported a 75% rate of CHL in their group of patients.[9,19] About 80% of children with FAS reported by Church et al had RSOM, exceeding the 12%-20% rate in the general population.[16,19] Clinicians often report anecdotally that auditory and touch/pain sensation (decreased or decreased) are two sensory integration dysfunctions noted in FASD. Our studies were not designed to answer these particular questions, and these will have to be addressed in future studies.

In our study, none of the children diagnosed with FASD had additional craniofacial manifestations or stigmata which may confound the causation of hearing loss. All had hearing levels within the normal range for their age (11.2% in compare to 14.9%). Acute otitis media, middle ear effusion, and chronic otitis media reported as associated with FASD[20] were not more prevalent in our group of patients, as compared to population controls.

As most children with FASD do not exhibit oral cleft, our study suggests that the majority of children with FASD should be expected to have normal hearing. Because these children are often academically and behaviorally challenged, early follow-up for potential hearing loss and intervention may help children with FASD reach their full academic potential.

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