Prevalence of Permanent Congenital Hearing Loss

How many babies in the general population are born with permanent hearing loss?

Although it seems like this should be a simple question to answer, widely discrepant figures are frequently cited ranging from ½ to 20 babies per 1,000.

Reports of Screening Programs

One of the best sources of information for estimating the prevalence of permanent congenital hearing loss are the published reports of newborn hearing screening programs who have tried to follow the babies referred from screening programs to determine how many were identified with permanent hearing loss.

Table 1:
Rate Per 1,000 of Permanent Congenital Hearing Loss in Published Reports of UNHS Programs

Location of Program (Time)	Cohort Size	Primary Screening Technique	% of Refers Lost to Follow-up	Prevalence Per 1000 of Hearing Loss*
New Jersey Barsky-Firkser & Sun, 1997 (1/93 - 12/95)	15,749	ABR	41%	3.30
New York Prieve, 2000 (1/96 - 12/96)	27,938	OAE & AABR	23%	1.96
Colorado Mehl & Thomson, 1998 (1/92 - 12/96)	41,976	AABR	52%	2.56
Texas Finitzo, et al., 1998 (1/94 - 6/97)	54,228	OAE	31%	2.15
Hawaii Johnson, et. al, 1997 (1/94 - 6/97)	9,605	OAE	2%	4.15

Source: www.infanthearing.org, *Frequently Asked Questions*

As the table data illustrates, most programs have not been successful in determining the final hearing status of a large percentage of the babies referred from the screening program. In spite of this, these programs are reporting 2-4 babies per 1,000 with permanent congenital hearing loss. If the programs that were unable to determine the hearing status of a large number of babies had been more successful with follow-up, it is likely that their prevalence rates would have been higher. Thus, and estimate of 2-4 babies per 1,000 with permanent congenital hearing loss seems quite reasonable.

Results of Epidemiological Studies

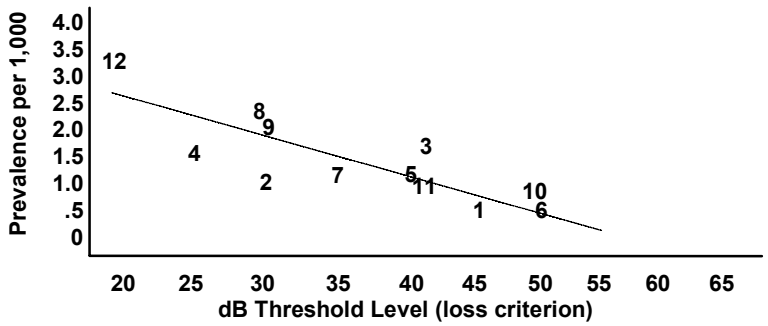
How consistent are the results of these screening programs with epidemiological studies of permanent hearing loss among children? In answering this question it is useful to consider definitional issues related to severity and type of hearing loss, whether the hearing loss is unilateral or bilateral loss, and whether the hearing loss is congenital or late-onset.

Definitional issues

Consider the results (summarized in Figure 1) of 12 studies designed to determine the number of children with bilateral permanent hearing loss in population-based cohorts ranging in size from 10,000 to over 4 million children.

In each study, a large cohort of children who were representative of the general population in that country were assessed for permanent hearing loss when they were 6-12 years old.

Figure 1:
Reported Prevalence Rates of Bilateral Permanent Hearing Loss in Population Based Studies



1. Barr (1980), n = 65,000
2. Downs (1978), n = 10,726
3. Feinmesser et al. (1986), n = 62,000
4. Fitzland (1985), n = 30,890
5. Kankkunen (1982), n = 31,280
6. Martin (1982), n = 4,126,268
7. Parving (1985), n = 82,265
8. Sehlin et al. (1990), n = 63,463
9. Sorri & Rantakallio (1985), n = 11,780
10. Davis & Wood (1992), n = 29,317
11. Fortnum et al. (1996), n = 552,558
12. Watkin et al. (1990), n = 51,250

Not surprisingly, the dashed line shows that the prevalence of bilateral permanent hearing loss is substantially higher when milder hearing losses are included. When children are only counted when they have a bilateral permanent hearing loss greater than 50 dB, prevalence is about 1.0 per thousand. However, when children are included if they have a bilateral permanent hearing loss greater than 30 dB, the prevalence increases to about 2.5 per thousand.

Source: *www.infanthearing.org*

Unilateral hearing loss

The data in the proceeding chart is for children with bilateral permanent hearing loss.

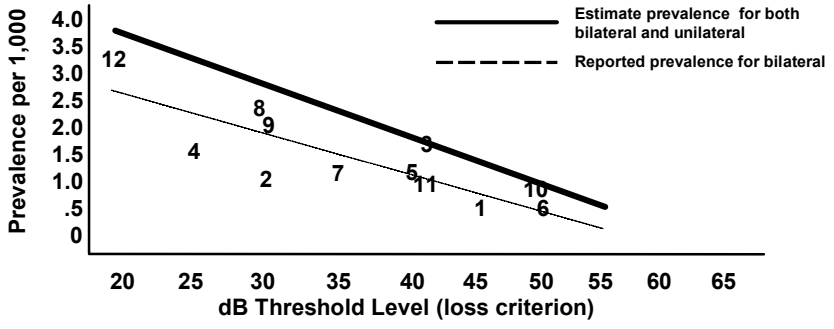
How much would the prevalence increase if children with unilateral permanent hearing loss were included? We can estimate this from the results of research that has evaluated the percentage of all children with permanent hearing loss who have unilateral hearing loss.

Table 2: Percentage of Permanent Hearing Loss That is Unilateral

Author(s) (year)	# of Children with Hearing Loss in Sample	% unilateral
Kinney (1953)	1307	48%
Brookhouser, Worthington, and Kelly (1991)	1829	37%

Using results from these studies to increase the numbers shown in Figure 1 by 40% results in the estimate shown by the solid bold line in Figure 2.

Figure 2:
Estimated Prevalence Rates of Bilateral and Unilateral Permanent Hearing Loss in Population-Based Studies



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Late-onset loss

But how many of the children in the Figure 2 chart have late-onset loss (i.e., were born with normal hearing but acquired a permanent hearing loss by 12 years of age)?

Although we don't have definitive data to answer this question we can make some credible estimates. Based data from a large multi-center longitudinal study reported by Norton (2000), the Joint Committee on Infant Hearing estimated that only about 2% of children with permanent hearing loss by 12 months of age had normal hearing at birth.

This estimate is consistent with anecdotal reports from Coordinators of State newborn hearing screening programs that have been functioning for many years. For example, Colorado, Rhode Island, and Hawaii have been screening almost all of their newborns since the early 1990's. All of these states have reasonably good tracking systems to identify children with hearing loss as they enter school and attempt to determine the status of the newborn hearing screening result for any child identified with hearing loss after the neonatal period. Each of these states report that fewer than 5% of the children with permanent hearing loss at the time they entered school, had passed the newborn hearing screening test. It is important to remember that some of these could have been congenital losses that were missed by the screening.

Conclusion

Based on the consistency of the data from newborn hearing screening programs and the epidemiological studies, we can be confident that 2-4 newborns have permanent congenital hearing loss. If children with bilateral and unilateral hearing loss of 30dB or greater or included, the prevalence will be closer to 4.0 per 1,000. If only bilateral hearing loss of 50 dB or greater are included, the prevalence is significantly lower.

For more information, see www.babyhearing.org or www.boystownhospital.org

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