Integrating cochlear dead region diagnosis into the hearing instrument fitting process

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Introduction

It is well known that sensory hearing impairment is accompanied by supra-threshold deficits such as degraded frequency and temporal resolution. Until now, there have been few hearing instrument fitting rationales that have relied on these supra-threshold deficits to determine the optimal parameters of a particular hearing instrument fitting. However, recent research suggests that procedures which measure the extent of cochlear dead regions might influence aspects of the optimal prescribed frequency-gain characteristic. Individuals with a cochlear dead region may have different frequency-gain requirements than those with no dead region. New or revised fitting rationales may include an optional formula that can be used when there is evidence for cochlear dead regions (Dillon, 2006). Diagnosing the presence and the extent of a dead region may have important clinical implications for counselling and hearing instrument provision.

PHONAK
hearing systems

The purpose of this Phonak Focus is to provide the hearing healthcare professional with an overview of recent research findings on cochlear dead regions. It concentrates primarily on high-frequency dead regions since high-frequency hearing impairment is by far the most common audiometric configuration in individuals being fitted with a hearing instrument. The content is split into sections that address the following questions:

1. What is a cochlear dead region?
2. Do some hearing-impaired humans have a cochlear dead region?
3. Is there an audiometric pattern associated with cochlear dead regions?
4. Is there a clinical test that can be used to identify cochlear dead regions?
5. What do we know about the prevalence of cochlear dead regions?
6. What are the implications for hearing instrument fitting?
7. Are there any outstanding research questions?

Figure 1
Inner and outer hair cells in the cochlea. They are called 'hair' cells because the stereocilia look like tufts of hair. The inner and outer hair cells have very different functions. The single row of inner hair cells is responsible for changing mechanical vibration to an electrical signal. The outer hair cells have a key role in the 'active' mechanism within the cochlea.
Source: www.fleshandbones.com
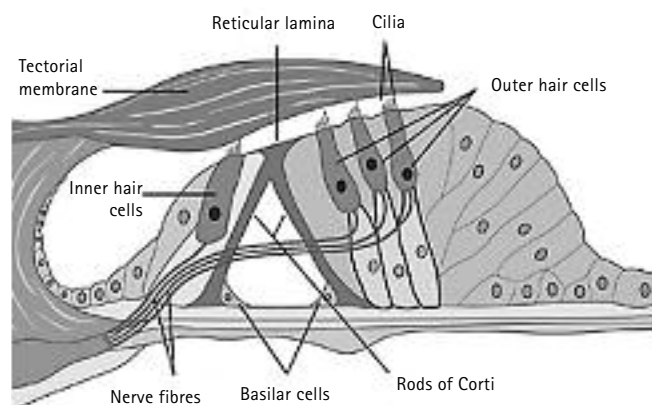
Readers who would like a more detailed account of the concepts, diagnosis and clinical implications of cochlear dead regions are referred to the comprehensive articles by Moore (2001, 2004).

1. What is a cochlear dead region?

The term 'cochlear dead region' (DR) first appeared in the literature about ten years ago (Moore et al., 1996) although the concept of 'gaps' or 'holes' in hearing has been around for a considerable period of time (e.g., Troland, 1929; Gravendeel and Plomp, 1960). Some hearing-impaired individuals have regions of inner hair cells (IHCs) and/or associated neurones that function so poorly, if at all, that they can be considered dead i.e., the mechanical vibration at a particular region of the basilar membrane cannot be transduced into an electrical signal in the auditory nerve (see Figure 1). However, at high presentation levels, a signal producing its maximum vibration in a DR may be detected as a result of a spread of excitation to adjacent regions of the basilar membrane where the IHCs and/or neurones are functioning. This is known as 'off-frequency' or 'off-place' listening. Clinical procedures for the diagnosis of DRs are based on the identification of off-frequency listening. Using an analogy, a DR is somewhat akin to having a piano with a group of broken strings. A heavy hit on the keys may cause adjacent strings to vibrate. In our case, a signal that produces maximum vibration within a DR may still be detected but there may be implications for the way the signal is perceived. This may impact on patient counselling, selection of gain-frequency response, and hearing aid benefit.

There will be some occasions when basilar membrane excitation adjacent to the DR is insufficient for off-frequency detection. For example, take an individual who has non-functioning high-frequency IHCs and/or neurones. Initially, a tone falling in the DR will be detected because of good hearing sensitivity

at the adjacent region on the basilar membrane. Over time, however, the individual may develop a hearing impairment at the adjacent region of the basilar membrane (for example, as a result of natural ageing). Although the high-frequency DR is still present, it may no longer be possible to find evidence for this via off-frequency listening. This is an example where the severity of the hearing impairment may be consistent with a DR although it will not be possible to confirm this via the presence of off-frequency listening.

There are occasions when an individual may have a sick region (i.e., IHCs and/or neurones have impaired function but can respond normally at high presentation levels). This may occur, for example, at the transition between a normal low-frequency region and a dead high-frequency region. A pattern of off-frequency listening for low signal levels and on-frequency listening for high signal presentation levels would be consistent with a sick region (see section four).

Pure tones that fall within a DR are often perceived as sounding distorted or noise-like in quality. However, both normally hearing and hearing-impaired listeners rate some tones as somewhat noise-like, independently of the existence of a DR (Huss et al., 2005). Therefore, subjective reports of noise or distortion can be taken as an indication that a DR may be present but they are not a reliable method of diagnosing a DR.

2. Do some hearing–impaired humans have a cochlear dead region?

Yes, there is evidence in the literature to support the presence of cochlear DRs in some hearing-impaired humans. IHC damage has been confirmed in histological evaluation of temporal bones in humans (Schukneckt and Gacek, 1993; Amatuzzi et al., 2001). Schukneckt and Gacek (1993) showed that hearing impairment in adults was frequently accompanied by loss to IHCs and/or OHCs.

More recently, Amatuzzi et al. (2001) showed that three newborn babies who had been in a neonatal intensive care unit, and had failed a hearing screen using the auditory brainstem response, had a loss of IHCs without accompanying OHC damage when examined histologically[1] (All footnotes are provided on page 15). A further four babies who failed the screen had abnormalities to both IHCs and OHCs. This finding is consistent with a number of animal studies that have reported selective damage to IHCs. In studies using the chinchilla, Harrison (2001) reported extensive IHC degeneration and normal OHCs as a result of both mild chronic hypoxia or treatment with cysplatinin, an oto-toxic anticancer drug. In newborn rats, Mazurek et al. (2003) have also shown that IHCs are more susceptible to hypoxia/ischemia than OHCs. The mechanism underlying the higher susceptibility of IHCs is not well understood but the higher expression of glutamate receptors, the moderate expression of plasma membrane calcium ATPase, the lower glycogen content, and the lower content of mitochondria may all be contributing factors. In summary, there is evidence that cochlear DRs can occur in adults and children with an acquired or congenital hearing impairment.

3. Is there an audiometric pattern associated with dead regions?

No, there is no definitive audiometric pattern associated with DRs but there are certain audiometric features that are more likely to be present. If the OHCs are damaged to the extent that the 'active' process is completely absent, there will be a maximum hearing-impairment of around 60 dB HL. It is also known that the maximum hearing impairment due to IHC damage, before they cease to function altogether, is of the order of 20–30 dB. Therefore:

1. a mild or moderate sensory hearing-impairment may be due to a combination of OHC and IHC damage,

2. a severe sensory hearing-impairment is probably due to a combination of OHC and IHC damage, and

3. a profound hearing-impairment is probably due to total OHC and IHC damage.

The spread of excitation along the basilar membrane usually falls rapidly (at the more apical low frequencies) after it has reached its maximum vibration, as illustrated in Figure 2. If a high frequency tone that falls within a region of non-functioning IHCs is to be detected at a low frequency place on the basilar membrane, then hearing sensitivity at the low frequency place would need to be relatively good because of the rather rapid reduction in excitation. This means that relatively steep audiometric configurations are quite likely to be associated with a DR. However, some hearing-impaired ears do not show a rapid reduction in vibration as the wave of excitation travels along the basilar membrane towards the low frequencies.
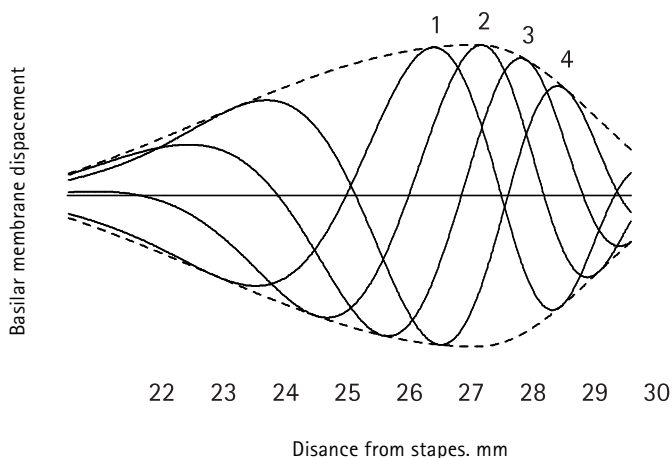
There are reports in the literature of more gentle sloping audiometric configurations being associated with a DR (e.g., Glasberg and Moore, 1986). This may explain why Vinay and Moore (2007b) found that steepness of the audiometric slope was not a reliable predictor of DRs (see section 5). It is not clear if this also applies to cases of congenital hearing impairment where, for example, there may be abnormal patterns of vibration on the basilar membrane due to a malformation within the cochlea. Caution should be used when relying on the audiometric configuration to raise suspicion of DRs in any individual, especially those with a congenital hearing impairment.

4. Is there a clinical test that can be used to identify dead regions?

Since a tone that falls within a DR may be detected at a different place on the basilar membrane, DRs are assumed to occur if a hearing-impaired listener can be shown to be using off-place listening. Two masking techniques have been used for the identification of off-place listening: psychophysical tuning curves (PTCs) and the threshold equalizing noise (TEN) test. Both are based on the assumption that a signal falling within a DR may be detected at a place on the basilar membrane where function is better, despite the amount of vibration being lower than at the peak frequency. In individuals without a DR, a noise at a remote place on the basilar membrane will have little masking effect on the hearing threshold. However, if a DR is present and the tone is being detected at the remote place, the threshold will be elevated by the masker.

The TEN is a broadband noise and it has been developed specifically for assessment of DRs within a clinical environment. The test is based on the measurement of tone thresholds in the presence of ipsilateral TEN. The original version of the TEN produces equal masked thresholds, in decibels sound pressure level, between 0.25

Figure 2
The pattern of activity builds up gradually with distance as it moves from left to right (basal high frequency to apical low frequency) and decays rapidly beyond the point of maximum displacement. Source: Moore (1998).

Basilar membrane displacement

22   23   24   25   26   27   28   29   30

Disance from stapes. mm

and 10 kHz (Moore et al., 2000). A revised version of the test produces equal masked thresholds, in decibels hearing level, between 0.5 and 4 kHz, and this makes it much easier to use in clinical practice (Moore et al., 2004). It is only the more recent version of the TEN test that will be discussed here. Since the TEN masker is not yet a standard option on current clinical audiometers, the TEN has been recorded onto CD[2]. The test requires a two channel audiometer: one channel controls the tones (which may be generated by the audiometer or routed from the CD) and the second channel controls the TEN (which is delivered to the same ear). Normal practice is to measure masked thresholds in the presence of the TEN at the frequencies that are likely to represent the transition from a healthy region to a DR (usually where there is a rapid change in threshold between two adjacent thresholds). Masked thresholds are measured using standard audiometric procedures although Moore et al. (2004) recommends using an ascending step size of 2 dB. Cairns et al. (In Press) have shown that smaller step sizes (down 4 dB and up 2 dB) can improve the reliability of the test. Masked thresholds usually only require one level of TEN which would typically be around 80 dB/ERB[3] (and at least 10 dB above the absolute threshold at the test frequency). A high presentation level is required so that the TEN masker is effective and also to reduce the possibility of labelling a sick region as a DR. Figure 3 summarises TEN test interpretation. If the threshold measured in the TEN is 10 dB or more above the threshold in quiet, and at least 10 dB above the level of the TEN, this is taken as indicative of a DR at the signal frequency (Moore et al., 2000). Meeting the first criterion demonstrates that the TEN masker was effective: meeting the second criterion demonstrates that TEN had a greater masker effect than would be expected from on-frequency listening. If the criteria are met for a DR at all (or most) test frequencies then the results should be treated with caution as greater susceptibility to masking can be

**Figure 3**
Interpretation of the TEN test
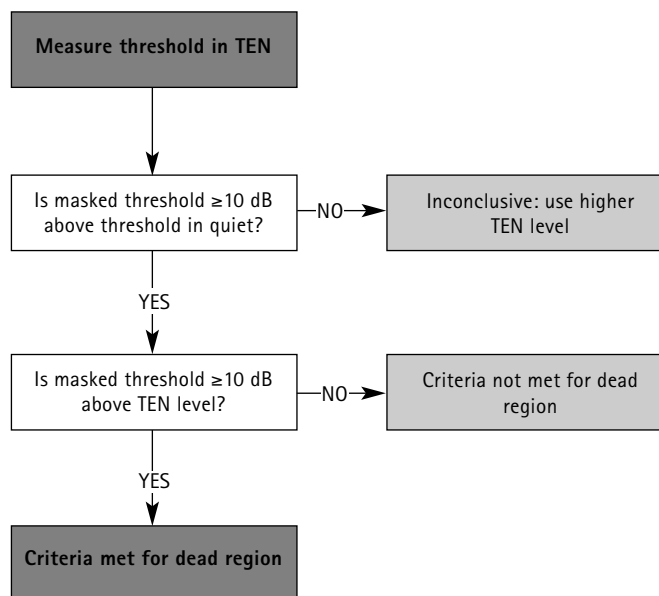
produced by poor processing efficiency in conditions such as auditory neuropathy (Vinay and Moore, 2007a).

Figure 4 shows the hypothetical hearing thresholds for two listeners who are being assessed for a hearing instrument. The audiologist decided to use the TEN test to check for the presence of a DR at the higher frequencies; it is possible that pure tones at 1.5 kHz and higher were detected around the 1 kHz place on the basilar membrane. The TEN was presented at a level of 90 dB/ERB and the audiologist measured the masked hearing threshold at 1, 1.5 and 2 kHz. The pure tone thresholds should be elevated to around 90 dB HL if there is no DR. In order to meet the criteria for a DR the masked thresholds should be elevated to 100 dB HL, or higher. For the individual on the left, the masked thresholds are 90 dB HL at 1, 1.5 and 2 kHz, respectively. Thus, the criteria for a DR are not met at any of these frequencies. For the individual on the

Figure 4

THypothetical hearing thresholds for two listeners who are being assessed for a hearing instrument. Open symbols are hearing thresholds in quiet, filled symbols are hearing thresholds measured in TEN at 90 dB/ERB. For the individual on the left, the criteria for a dead region are not met. For the individual on the right, the criteria for a dead region are met at 1.5 and 2 kHz.
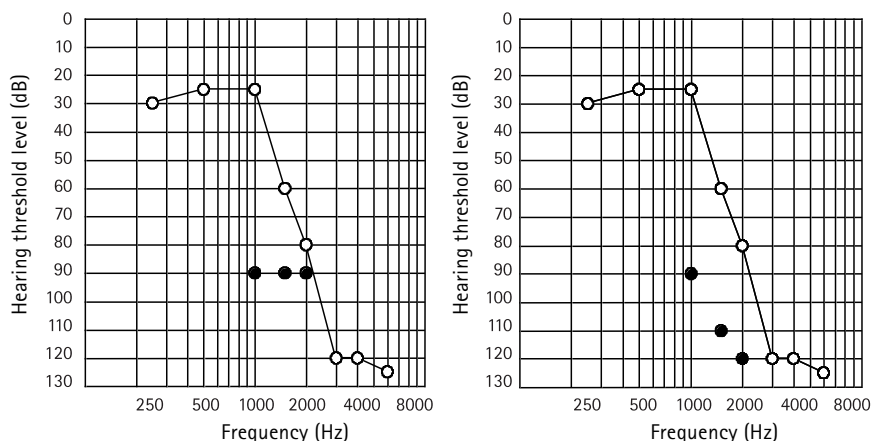
right, the masked thresholds are 90, 110 and 120 dB HL at 1, 1.5 and 2 kHz, respectively. The criteria for a DR are met at 1.5 and 2 kHz. Therefore, pure tones with frequencies of 1.5 kHz and above are being detected by off-frequency listening. The DR appears to commence somewhere between 1 and 1.5 kHz. A more precise estimate of the edge frequency would require measurement of masked thresholds at intermediate frequencies between 1 kHz and 1.5 kHz, but this is probably not necessary for clinical practice (and, in any case, is not possible unless tones are available at less than one half-octave intervals). For the individual on the right, the pure tone audiogram may be thought of as providing an inaccurate measure of high-frequency hearing since there is effectively no hearing above approximately 1.5 kHz. It probably would take the audiologist less than a few minutes to establish the presence of an extensive high-frequency DR in such an individual. For reasons that will be discussed in section six, high-frequency DRs are probably not important for guiding hearing instrument fitting if they commence above 2 kHz.

A small number of studies have investigated the test-retest reliability of the TEN test. Cairns et al. (In Press) carried out a retest within seven days for a group of hearing-impaired adults and a group of hearing-impaired teenagers. A total of 3 (7.5%) and 2 (8%) ears changed category, respectively. Munro et al. (2005) reported that 2 (7.1%) ears of the same subject (that just met the DR criteria) changed category on retest after a period of 12 months. The majority of ears that changed category on retest in both of these studies just met the DR criteria at an isolated frequency. An immediate retest is advisable in such cases. Practical applications and useful guidelines for when and how to use the TEN test are provided by Moore (2001, 2002a, 2004).

For a given amount of energy, a broadband noise such as TEN is perceived louder than a narrowband of noise because it is spread over a number of critical bands. Many studies have reported that some listeners find the TEN to be uncomfortably loud. The loudness can be lowered by reducing the bandwidth of the TEN. The original version of the TEN was band limited between 125-10,000 Hz. Markessis et al. (2006) high-pass filtered the original TEN at 0.5 and 1 kHz with some success. The current version of the TEN is band limited between 354 and 6500 Hz. In theory, there is no reason why narrower bands of noise could not be used. For example, if the edge frequency of a DR is thought to be around 2 kHz, then tones that fall within a DR will be masked by noise centred around 2 kHz. However, this would require a great many separate bands of noise, which potentially complicates the clinical procedure (and it would be hard to know in advance where to centre a narrow band of noise). In any case, this option is not currently available for clinical practice.

The TEN test serves as a useful tool for detecting DRs, but it does not precisely define the edge frequency, although its precision could be improved somewhat by providing

tones at finely spaced frequencies. A solution is to identify the edge frequency using psychophysical tuning curves (PTCs). A PTC shows the level of a narrowband masker required to mask a low level signal, plotted as a function of masker centre frequency. The lowest masker level required to mask the signal defines the tip of the PTC: this is the frequency at which the masker is most effective. In normal hearing listeners the tip of the PTC usually lies close to the signal frequency (Moore, 1978; Moore and Alcantara, 2001). For hearing-impaired listeners without a DR, the tip of the PTC is usually broader but still lies close to the signal frequency (Moore, 1998). In cases where the signal frequency lies within a DR, the tip will be shifted away from the signal frequency (Moore, 1998). The tip of the PTC will be shifted to the frequency which corresponds to the place on the basilar membrane where the signal is being detected. This identifies the edge of the DR. When the tip of the PTC is shifted towards a lower frequency, this indicates a high frequency DR. Conversely, when the tip is shifted to a higher frequency this indicates a low frequency DR. Examples of PTCs are shown in Figure 5.

Since the tip of the PTC corresponds to the edge of the DR, PTCs potentially provide a more accurate method for determining the frequency limits of a DR. Traditional PTC measurement procedures are time consuming to administer, as each PTC requires measurement of many masked thresholds in order to define the frequency at the tip. Therefore, traditional procedures do not lend themselves to clinical situations or for use with listeners who have limited spans of attention such as young children. In addition, traditional PTCs can be affected by the detection of beats and combination tones (Kluk and Moore, 2004, 2005). Recent work on a fast method for measuring a PTC means that it might soon be possible to use these in clinical practice. Several authors have used a fast method for determining PTCs, based on the use of a masker

Figure 5
Examples of psychophysical tuning curves. The right panel is for a listener with a high frequency sensory hearing impairment. The filled symbols show the target and the open symbols show the masker. In these examples, the tip of the tuning curve occurs at the same frequency as the target. The left panel shows a single tuning curve of a listener with a high frequency dead region. The 1.5 kHz signal is most easily masked with a masker around 1 kHz. Source: Moore (2001).
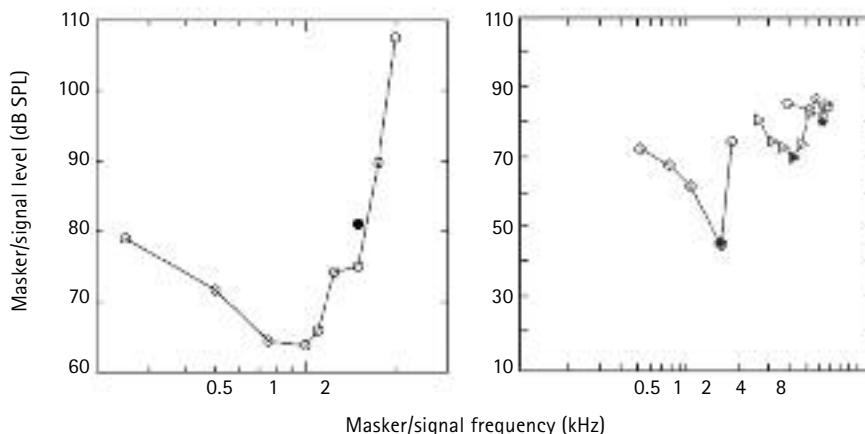
whose centre frequency sweeps across the frequency range using a Békésy-type tracking procedure. Zwicker (1974) used the technique with normal hearing listeners and Summers et al. (2003) used it with hearing-impaired listeners, some of whom had a DR. However, Sek et al. (2005) were the first to systematically evaluate parameters such as rate of change of masker level in order to optimise the procedure for the assessment of DRs in clinical practice. Sek and colleagues demonstrated that the fast-PTC method produces similar results to the traditional PTC measurement procedures. Unfortunately, the approach used by Sek et al. cannot be easily implemented in the clinic because audiometers will not allow an externally generated masker to be controlled adaptively by the listener. In order to make the adaptive technique available clinically, we have implemented the fast-PTC method on a PC fitted with a high quality sound card. The software programme was developed in our laboratory by Richard Baker for use with a Kamplex KC 35 clinical audiometer fitted with TDH 39 headphones[4]. The PC was additionally equipped with an

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Figure 6
A fast-PTC measured from a normal-hearing six year old boy. The masker swept from a low frequency to a high frequency. The tip of the tuning curve lies close to the 1 kHz signal frequency. Unpublished data from Alicja Malicka.
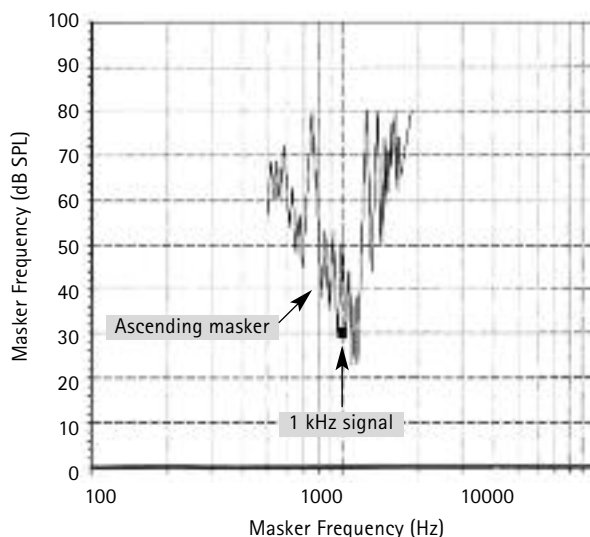
external 24 bit sound card (Edirol UA-5). The attenuation and mixing of the signals were carried out using the audiometer, under computer control via the RS 232 interface, thus maximizing the dynamic range. The main interface of the software enables adjustment of the level and frequency of the signal tone, frequency step size of the masker, masker bandwidth, maximal masker output level (with the limits of the hardware) and direction of the masker sweep. Alicja Malicka and colleagues from our laboratory have investigated the feasibility of measuring fast-PTCs in normal hearing children and also hearing-impaired children with and without a DR. So far, we have been successful at using the technique with children as young as 6 years of age (see Figure 6). The preliminary data from our lab show good agreement between the results obtained using fast-PTCs and the TEN test in children who have undergone extensive testing. This is consistent with the findings of Kluk and Moore (2006), who tested 14 adults with high frequency DRs using the TEN test, fast-PTCs and a forward masking technique and reported

that the edge frequencies obtained from the PTCs were similar and usually close to the values estimated from the TEN test. This is reassuring because Summers et al. (2003) did not find close agreement between PTCs and the results of the TEN test. In 18 ears with steeply sloping high-frequency hearing-impairment, there was agreement in 10 (56%) ears only. Summers and colleagues argued that that the PTCs were more reliable than the TEN test. However, Moore (2004) and Kluk and Moore (2005) argued that some of the PTCs may have been influenced by factors such as beats and combination tones.

5. What do we know about the prevalence of cochlear dead regions?

Most studies reviewed in this section have used the TEN test to identify off-frequency listening. Prevalence data for cochlear DRs in adults with a sensory hearing-impairment have been provided by Vinay and Moore (2007b). They assessed 317 adults (592 ears) who attended an audiology department, generally for the fitting of a hearing aid. A total of 177 (54%) adults or 233 (42%) ears met the criteria for a DR at one or more frequency. It was rare to find evidence for a DR when the hearing threshold was 60 dB HL, or better, although DRs have been observed in individuals with better hearing thresholds when diagnosed using PTCs (e.g., Moore et al., 2000). On the other hand, there were occasions when hearing thresholds were as poor as 85 dB HL without evidence for a DR. Although the presence or absence of a DR at a specific audiometric frequency cannot be reliably determined from the hearing threshold alone, most adults who showed evidence for DRs had a hearing threshold at, or greater than, 65 dB HL. There is a sensitivity/specificity trade-off between separating adults with a DR from those without a DR. Vinay and Moore recommended testing for the presence of DRs when the hearing threshold exceeded 60 dB HL. The ability of hearing threshold data to identify

high-frequency DRs is shown in Table 1 for a cut-off criterion of 60 dB HL and also 70 dB HL. These calculations are based on the data presented by Vinay and Moore (see their Table 1) and assume that the TEN test is perfect at identifying every adult who has a DR. Using a cut-off criterion of 60 dB HL at 2000 Hz as an example, for every 100 patients being fitted with a hearing instrument, all patients having a DR and 29 patients who do not have a DR will meet the criterion for further investigation. If the criterion is changed to 70 dB HL, one patient with a DR will be missed but the number of patients who do not have a DR will be reduced to 15. In a busy clinical environment, there may be some justification in using the latter criterion.

Vinay and Moore also investigated the relationship between the slope of the audiometric configuration and evidence for DRs. The audiometric slope was calculated between the estimated edge frequency and one octave higher. The mean slope of the audiogram was 15-20 dB/octave (depending on the frequency at the edge of the DR) when the TEN test showed evidence for a DR. When there was no evidence for a DR, the slope was 8-15 dB/octave. Since the low frequency side of the travelling wave pattern is usually relatively steep, it is to be expected that there will be a steep slope in the frequency range nearest the start of the DR. Unfortunately, there was considerable variability around the mean slope for both groups. Other studies have also shown considerable overlap between the steepness of the slope of the audiogram and the presence/absence of a DR (Preminger et al., 2005; Aazh and Moore, 2007). Thus, the audiometric threshold or the steepness of the slope of the audiogram does not provide a reliable indication of the presence or absence of a DR.

A number of studies have reported the presence of DRs in selected patient groups. Moore et al. (2000) reported that 68% of adult

Table 1
The ability of pure tone hearing threshold data to identify high-frequency cochlear dead regions in adults. The cut-off criterion is 60 and 70 dB HL in the top and bottom table, respectively. For example, in the top table, a dead region is assumed to be present if the hearing threshold is 60 dB HL, or greater, but absent if the hearing threshold is 55 dB HL, or lower. The performance characteristics were calculated from Vinay and Moore (2007).

60 dB HL			
	Sensitivity	Specificity	Accuracy
500 Hz	24/25 = 96%	431/518 = 83%	(24+431) / (25+518) = 0.84
1000 Hz	36/36 = 100%	362/502 = 72%	(36+362) / (36+502) = 0.74
2000 Hz	100/100 = 100%	249/392 = 64%	(100+249) / (100+392) = 0.71
4000 Hz	132/132 = 100%	119/283 = 42%	(132+119) / (132+283) = 0.60

70 dB HL			
	Sensitivity	Specificity	Accuracy
500 Hz	23/25 = 92%	477/518 = 92%	(23+477) / (25+518) = 0.92
1000 Hz	35/36 = 97%	415/502 = 83%	(35+415) / (36+502) = 0.84
2000 Hz	99/100 = 99%	319/392 = 81%	(99+319) / (100+392) = 0.85
4000 Hz	129/132 = 98%	169/283 = 60%	(129+169) / (132+283) = 0.72

ears showed evidence for DRs; however, these adults were selected because they were likely to have DRs (based on audiometric configuration). Preminger et al. (2005) selected 49 adults having two (or more) hearing thresholds within the range 50 to 80 dB HL and reported that 29% of their adults showed evidence for DRs (6 unilateral, 8 bilateral). They used stricter criteria for identifying a DR than most other studies. Jacob et al. (2006) reported that 92% of ears with a moderate to severe sloping sensorineural showed evidence for DRs. Markessis et al. (2006) selected 35 adults with a moderate-to-severe hearing impairment with a slope of 20 dB/octave over at least one octave from 1 and 8 kHz and reported that over 87% of ears showed evidence for DRs. All thresholds at 4 kHz were greater than 65 dB HL yet only 52% showed evidence for DRs. Aazh and Moore (2007) tested 98 adults with hearing thresholds between 60 and 85 dB HL at 4 kHz and reported that 37% showed evidence

for DRs. Palma et al. (2005) tested one ear each from 28 adults who had at least one hearing threshold better than 60 dB HL and reported evidence for DRs in 25% of ears. Cairns et al. (In Press) tested 20 adults who had hearing thresholds between 41 and 95 dB HL and a difference of at least 20 dB between adjacent audiometric frequencies. They reported evidence for DRs in 22.5% of ears. Cairns et al. also reported the presence of DRs in young people who had a severe-to-profound hearing impairment. They tested 23 ears of 15 teenagers who had at least one hearing threshold better than 80 dB HL and reported that there was evidence for DRs in 13% of ears. In an earlier study using a similar population, Moore et al. (2003) reported evidence for DRs in 63% of ears. The presence of DRs was probably lower in the more recent study by Cairns et al. (2007) for a number of reasons. First, they did not test above 4 kHz where DRs were probably very common. Second, they used a smaller ascending step size of 2 dB: if they had used an ascending step size of 5 dB then the number of ears meeting the criteria would have increased to 48%.

Many of the studies listed above have used pre-selected groups of patients and this probably explains the highly variable occurrence of DRs. The one exception is the study by Vinay and Moore (2007b), who reported that 54% of unselected individuals, referred for fitting of a hearing aid, met the criteria for a DR at one or more frequency in at least one ear. It is not known how many of these individuals had a 'clinically significant' DR. A clinically significant DR is defined here as 'a DR that influences selection of amplification characteristics'. As will be shown in the next section, a high-frequency DR probably only influences the selection of amplification characteristics if it extends down to at least 2 kHz. We reviewed the audiology records of new adult hearing aid referrals at one of our local Audiology Services in South Manchester for the first quarter of 2007. There

were 273 referrals, 242 having a sensorineural hearing impairment and 63 (91 ears) had a high-frequency hearing-impairment of 60 dB HL, or greater, that extended down to at least 2 kHz. Therefore, 26% (i.e., 1 in 4) of adult hearing aid referrals with a sensorineural hearing impairment may have a clinically significant DR. Data are currently being collected by Toal and Munro to identify which of these patients have a clinically significant DR: the number is likely to be much smaller than 1 in 4 since we know from Vinay and Moore (2007b) that only 30% of ears having a threshold of 60 dB HL, or greater, at 2 kHz meet the criteria for DR.

6. What are the implications for hearing instrument fitting?

There is evidence that high-frequency amplification may not always improve speech recognition in adults with a high-frequency hearing-impairment. Some studies have shown no benefit (e.g., Murray and Byrne, 1986) while others have shown a degradation in performance (e.g., Ching et al., 1998). There is little agreement on the degree of loss and/or audiometric configuration that can be used to identify those who will benefit from high-frequency amplification. The lack of benefit may be due, at least in part, to the presence of DRs, although there is some controversy in this regard. A growing number of studies have investigated the benefit of high-frequency amplification in adults with DRs. These studies have used adult listeners and measured speech recognition performance in quiet (Vickers et al., 2001), background noise (Baer et al., 2002) or both (Mackersie et al., 2004).

Studies using speech in quiet
Vickers et al. (2001) compared performance in 18 ears with a high-frequency hearing-impairment. Twelve ears had DRs and six ears did not have DRs. Subjects listened to vowel-consonant-vowel (VCV) nonsense syllables such as /aba/ or /ama/. The VCVs were

presented over earphones and amplified to match the frequency-gain characteristics of the Cambridge prescription formula (Moore and Glasberg, 1998). The listener's performance was then measured after low-pass filtering, i.e., with high frequency amplification removed. Figure 7 shows the outcome from three hypothetical subjects that serve to illustrate the pattern of findings reported by Vickers et al. The scores for subject A improve with increasing cut-off frequency, i.e., the subject benefits from providing high-frequency amplification. This pattern is characteristic of subjects who do not have a DR. Subjects B and C both have a DR commencing around 1 kHz. In both subjects, performance improves up to around one octave above the start of the DR. However, performance above this frequency is different for the two subjects. Subject B did not show any benefit from provision of amplification at the very high frequencies but also did not show any deleterious effects. Most of the DR subjects in the Vickers et al. study showed this pattern of results. However, three (25%) subjects were like Subject C, i.e., the provision of amplification well within the DR had a deleterious effect on performance. One explanation for the divergent pattern at frequencies well above the edge frequency of the DR is that listeners who did not show deterioration in performance did not receive the same restoration in audibility because real-ear gain was limited to a maximum of 50 dB. In summary, the results show that subjects with extensive DRs can extract useful information up to about one octave inside the DR.

Vinay and Moore (In Press) carried out a study that was similar in design to that of Vickers et al. but the listeners had low frequency hearing impairment. There were 19 ears with DRs that commenced from 0.75 kHz or higher and 22 ears without DRs. The ears with DRs did not perform as well as ears without DRs when using broadband amplification. In addition, ears with low frequency DRs benefited from

Figure 7
Speech recognition performance of three hypothetical subjects with amplification and low-pass filtering. Subject A (solid line) does not have a dead region. Subjects B and C (filled circles and open circles, respectively) both have an extensive dead region commencing from around 1 kHz. Both subjects with DRs do not show as much benefit from broadband amplification as the subject without a dead region. For one of the subjects with a DR (subject C), performance deteriorates when amplification extends to the very high frequencies.


— Subject A
● Subject B
○ Subject C
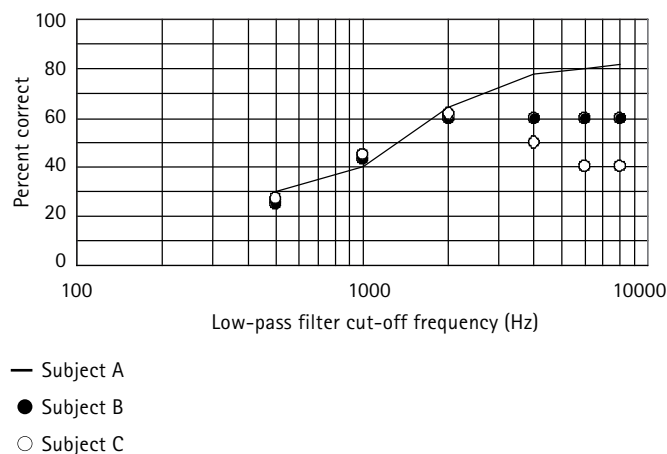
low frequency amplification that extended into the DR by about one octave. However, there was deterioration in their performance when amplification extended well into the DR.

These findings form the basis for the recommendation to limit high-frequency amplification to around 1.7 above the start of the DR (Moore, 2004). This is illustrated in Figure 8 where the edge of the DR is around 1 kHz. The audiogram forms on the left and right shows an extensive low frequency and high frequency DR, respectively (shaded portion). For the low frequency DR, there is little point in providing amplification at frequencies below about 0.6 kHz (1 kHz / 1.7): for the high frequency DR, there is little point amplifying above about 1.7 kHz (1 kHz x 1.7). Of course, if the edge of the high frequency DR commenced around 3 kHz then there would be no need to restrict high-frequency amplification since the bandwidth of most current hearing instruments is unlikely to extend above 5 kHz (3 kHz x 1.7). There is some controversy regarding these findings and

Figure 8
Provision of amplification to subjects with an extensive dead region. The shaded area represents the dead region. The audiogram form on the left shows an extensive low frequency dead region commencing from 1 kHz. The audiogram form on the right shows an extensive high frequency dead region commencing from 1 kHz. Amplification is provided that extends into the dead region by around one octave. For the low frequency dead region, amplification extends down to around 0.5 kHz. For the high frequency dead region, amplification extends up to around 2 kHz.

Extend amplification to around one octave below the start of the low frequency dead region

Extend amplification to around one octave above the start of the high frequency dead region
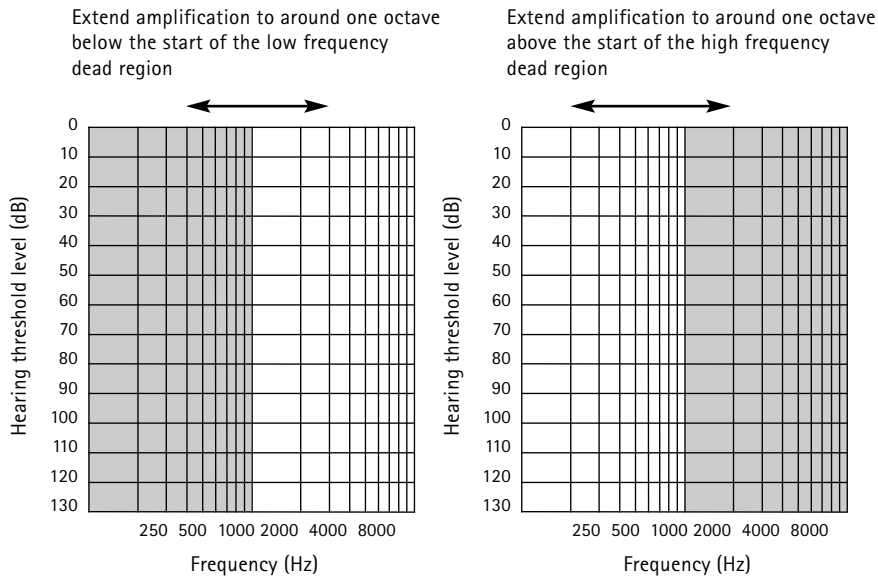
Rankovic (2002) is of the opinion that speech recognition performance can be predicted based on the Articulation Index (AI), regardless of the presence or absence of DRs. However, Moore (2002b) has shown that the incremental benefit of amplifying well above the edge of the DR is not as great as that predicted by the AI.

Vestergaard (2003) compared the effect of low pass filtering of words on 11 ears with DRs and 11 ears with no DRs. Listeners were tested while wearing their hearing aids as fitted by their audiologist. Moore (2004) re-analysed the Vestergaard data so that they could be compared with those of Vickers et al. (2001). Listeners with extensive DRs did not perform as well as subjects without DRs (or DRs restricted to very high frequencies) nor did they show the same incremental benefit with amplification

well inside the DR. Consistent with Vickers et al. listeners with DRs had a more severe hearing impairment than those without DRs; therefore, it is not clear if the difference between groups of listeners is due to the presence of extensive DRs or if there are confounding variable such as severity of hearing impairment.

Mackersie et al. (2004) compared performance in 16 ears with a high-frequency hearing-impairment. Eight ears had DRs and eight ears, matched for audiogramic configuration, did not have DRs. Subjects listened to VCV nonsense syllables in quiet, at 65 dB SPL, while wearing a hearing instrument set to approximate DSL (Cornelisse et al., 1995) frequency-gain targets. The subject's performance was then measured after low-pass filtering. Mackersie et al. reported no difference in performance between the two groups. This contrasts with the results of previous studies. One difference noted by Mackersie and colleagues is that the subjects in their study had a less severe hearing-impairment and less extensive DRs. Therefore, the limited benefit of high frequency amplification when listening to speech in the quiet may be restricted to subjects with extensive DRs.

Studies using speech in noise
Baer et al. (2002) carried out a study that was very similar to that of Vickers et al. (2001) and used many of the same subjects, except that the VCV stimuli were presented in steady speech-shaped noise. There were six ears with DRs and ten ears with no DRs. The noise had the same long-term spectrum as the VCV stimuli. The signal-to-noise ratio (SNR) was selected for each ear so that performance was 10-15% below performance in quiet. In ears without DRs, performance improved with increasing cut-off frequency; however, in ears with DRs, performance generally improved with cut-off frequency up to about 100% above the edge frequency of the DR, but with little further increase.

The study by Mackersie et al. (2004) reported above also measured performance in steady speech-shaped noise at a variety of SNRs. For relatively favourable SNRs, there was no difference in performance between ears with and without DRs. However, for conditions with a less favourable SNR, performance of the DR ears did not show an increase in performance when amplification was extended beyond one octave above the estimated edge frequency of the DR.

As part of a clinical study on DRs, Preminger et al. (2005) demonstrated that hearing instrument users with high-frequency DRs require a more favourable SNR in order to obtain 50% correct on a speech in noise test compared to hearing instrument users with no DRs, despite similar audiograms. The DR patients also reported less benefit from amplification in noise.

Keidser and Dillon (2007) cite a study of Ching et al. (2005) who tested 75 listeners with hearing threshold levels ranging from mild to profound. Speech recognition was measured in quiet and babble noise for sentence material and a consonant test under a variety of filter conditions. The data showed no consistent relationship between speech proficiency and the elevation of hearing threshold in TEN. Currently, full details about this study have yet to be reported. For example, the number of listeners with extensive DRs is not known.

Not all researchers agree that it is necessary to use a separate test to confirm the presence of a DR in severe steeply sloping sensory hearing impairment, claiming that it would not alter hearing instrument management (Summers, 2004). In a small study, it was shown that 10 audiologists would not attempt to provide broadband amplification to individuals with a severe sloping hearing-impairment. Rather, they would provide amplification at the lower frequencies where hearing thresholds were better than 90 dB HL. This appears to agree

closely with the recommendation of Moore (2004) to amplify up to 1.7 above the edge frequency. However, not every subject with a DR has a steeply sloping hearing impairment with thresholds greater than 90 dB HL. Vinay and Moore (2007) reported hearing thresholds that varied from 65 to 125 dB HL at 1.7 above the edge frequency. Therefore, the use of the TEN test to diagnose DRs is recommended.

In summary, the evidence from these adult studies is that: i) there is limited benefit of high-frequency amplification in listeners with extensive DRs when assessed in quiet or noise, and ii) listeners with less extensive DRs may show limited benefit from high-frequency amplification in environments that have poor SNRs.

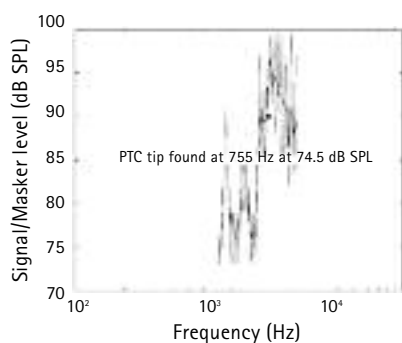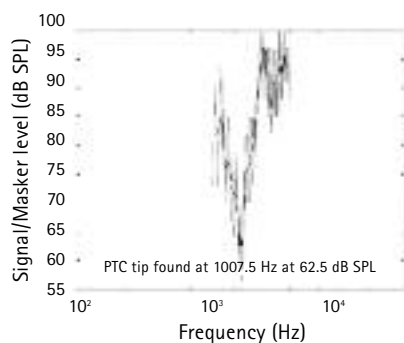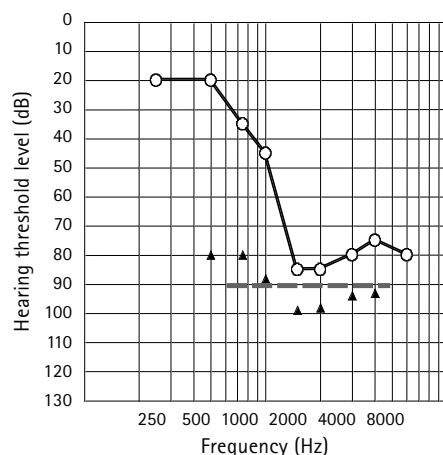## 7. Are there any outstanding research questions?

There are a number of research questions that have yet to be explored in detail and these span the continuum from fundamental to applied research. A few examples of the more clinically relevant questions are given below.

Few studies have investigated DRs in children. It is not known if the presence of DRs in babies and infants has the same implications for hearing instrument fitting as for adults. Currently there is a need to develop test procedures that can be used to identify DRs in babies. An electrophysiological test for the diagnosis of DRs would be a useful addition to the battery of objective hearing threshold techniques that can be used to estimate hearing ability in babies and infants. Preliminary studies in this area have used the cortical auditory evoked potential (CAEP) and the auditory steady state potential (ASSR) (Marriage and Moore, 2006, Kluk et al., 2007).

There are very few studies that have investigated the benefit of high-frequency amplification in children and none, as far as we

Figure 9
The findings from an 8 year child with an extensive high frequency dead region. This ear shows a steep-sloping high-frequency hearing impairment. The masked thresholds (filled triangles) were obtained with TEN at 80 dB/ERB. The TEN test criteria are met at frequencies above 1 kHz. There is no evidence of off-frequency listening on the 1 kHz fast-PTC. However, the tip of the 1.5 kHz fast-PTC is shifted to a lower frequency. Unpublished data collected by Alicja Malicka.

frequencies. It may be possible for infants, who are aided early, to make more use of the 'remapped' information than adults (with an acquired hearing-impairment) because of the greater plasticity in the developing auditory system.

The benefit of high-frequency amplification to children with a DR is one area of research that is being studied in our laboratory. We have used the TEN test and the fast-PTC method to identify DRs in congenitally hearing-impaired 8-12 year olds (Malicka and Munro, in preparation). Figure 9 shows the results for one child with an extensive high-frequency DR. The TEN test criteria were met at frequencies above 1 kHz. The fast-PTCs show evidence of off-frequency listening at 1.5 kHz but not 1 kHz.

We are currently investigating the benefit of high-frequency amplification using VCV stimuli presented in quiet and in noise. The preliminary findings for VCVs in quiet are similar to those reported for adults, i.e., there is little benefit to providing high-frequency amplification that falls well above the edge of an extensive DR. The findings for one child are shown in Figure 10. This child received no additional benefit when amplification was provided more than one octave inside the DR. On the other hand, children with DRs that are limited to the very high frequencies, or to small islands, appear to receive benefit with high-frequency amplification, although our preliminary findings suggest that the mean benefit from broadband amplification may not be as high as for children with no DR who have a similar audiometric configuration. Importantly, we have not observed a decrease in performance with increasing cut-off frequency in any child who has a DR.

An alternative approach for managing extensive high-frequency DRs might be to use frequency compression or transposition. This would mean that information that lies well within a DR can be recoded to lower

are aware, have specifically investigated this in the context of DRs. Based on a review of the literature, Stelmachowicz (2002) and Stelmachowicz et al. (2004) concluded that adult studies should not be used to predict the importance of high-frequency amplification for infants and young children. We know that adults are able to extract some useful information from off-frequency listening as demonstrated by their ability to benefit from amplification up to one octave inside a DR. In addition, Rosen et al. (1999) has demonstrated that normal adult listeners can rather quickly learn to make use of high frequency information that is shifted to lower

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frequencies. The use of frequency compression, in general, has produced mixed findings. Stelmachowicz (2004) points out that there have been few systematic studies that have addressed issues of candidacy, signal processing and parameter optimisation. The limited benefit may also have occurred without clear knowledge of the extent of the DR. There is emerging evidence from the work of Robinson et al. (2007) that there may be some benefit to taking information that falls well within a DR and recoding it to around the boundary of the DR.

Conclusions

There is evidence that DRs can occur in adults and children with an acquired or congenital hearing impairment. It is not possible to identify DRs without the use of test procedures other than the audiogram. One of these, the TEN test, is readily available and has been designed for ease of use within a clinical setting. Additional procedures such as the fast-PTC may also become available in the clinical setting. Approximately 50% of adult hearing aid referrals show evidence of a DR at one or more frequency. DRs are uncommon if the hearing threshold is 60 dB HL, or better. A 'high risk' group for clinically significant DRs would be individuals with an extensive region of hearing impairment of 60 dB HL, or greater (e.g., at all frequencies above 1 kHz). Adults with extensive high-frequency DRs do not appear to obtain the same benefit from broadband amplification as those without DRs. Most adults benefit from amplification that extends into the DR by about one octave. Above one octave, most adults show no further improvement although a subgroup may show a reduction in performance. What little information there is about children with high-frequency DRs suggests that some may not benefit from the provision of amplification well within a high-frequency DR; importantly, none (so far) have shown a reduction in performance.

Figure 10
Performance for a child with an extensive high frequency dead region commencing from around 1.5 kHz (see Figure 9). The percent correct score on the VCV test is plotted as a function of low-pass filter cut-off frequency. Unpublished data collected by Alicja Malicka.
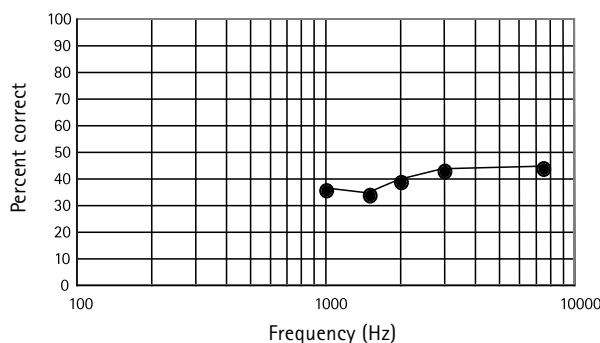
Footnotes
[1] The lack of ABR in the presence of damaged IHCs but normal OHCs is consistent with the umbrella term of "auditory neuropathy".

[2] Information about the test including how to purchase a copy of the TEN CD can be obtained online at www.hearing.psychol.cam.ac.uk

[3] ERB is the average equivalent rectangular bandwidth of the auditory filter as determined for young, normal-hearing listeners at moderate sound levels and its value in Hertz is calculated as 24.7 (4.37F+1) where F is a frequency in kHz. For example, at 1 kHz the ERB is approximately 0.132 kHz (Moore, 2004).

[4] For further details go to http://personalpages.manchester.ac.uk/staff/richardbaker

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