

# Cochlear Dead Regions in Children: Assessment and Management Issues

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## Introduction

Recent research on adults suggests that procedures that measure the extent of cochlear dead regions might influence the optimal prescribed frequency-gain characteristics of a hearing instrument. Some adults with a cochlear dead region may have different frequency-gain requirements than those with no dead region. It is not clear if these findings can be generalized to children although we know, for example, that adult studies should not be used to predict the importance of high-frequency amplification for infants and young children (Stelmachowicz 2002; Stelmachowicz, Pittman, Hoover, Lewis and Moeller 2004). The purpose of this chapter is to provide the hearing healthcare professional with an overview of recent research findings on cochlear dead regions. It extends the recent Phonak Focus article on dead regions (Munro 2007) by including newer published articles as well as describing preliminary data from our ongoing paediatric studies. The chapter concentrates primarily, but not exclusively, 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. 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).

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## What is a Cochlear Dead Region?

The term “cochlear dead region” (DR) first appeared in the literature about ten years ago (Moore, Glasberg and Vickers 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. 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 (see figure 1). 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 instrument benefit.

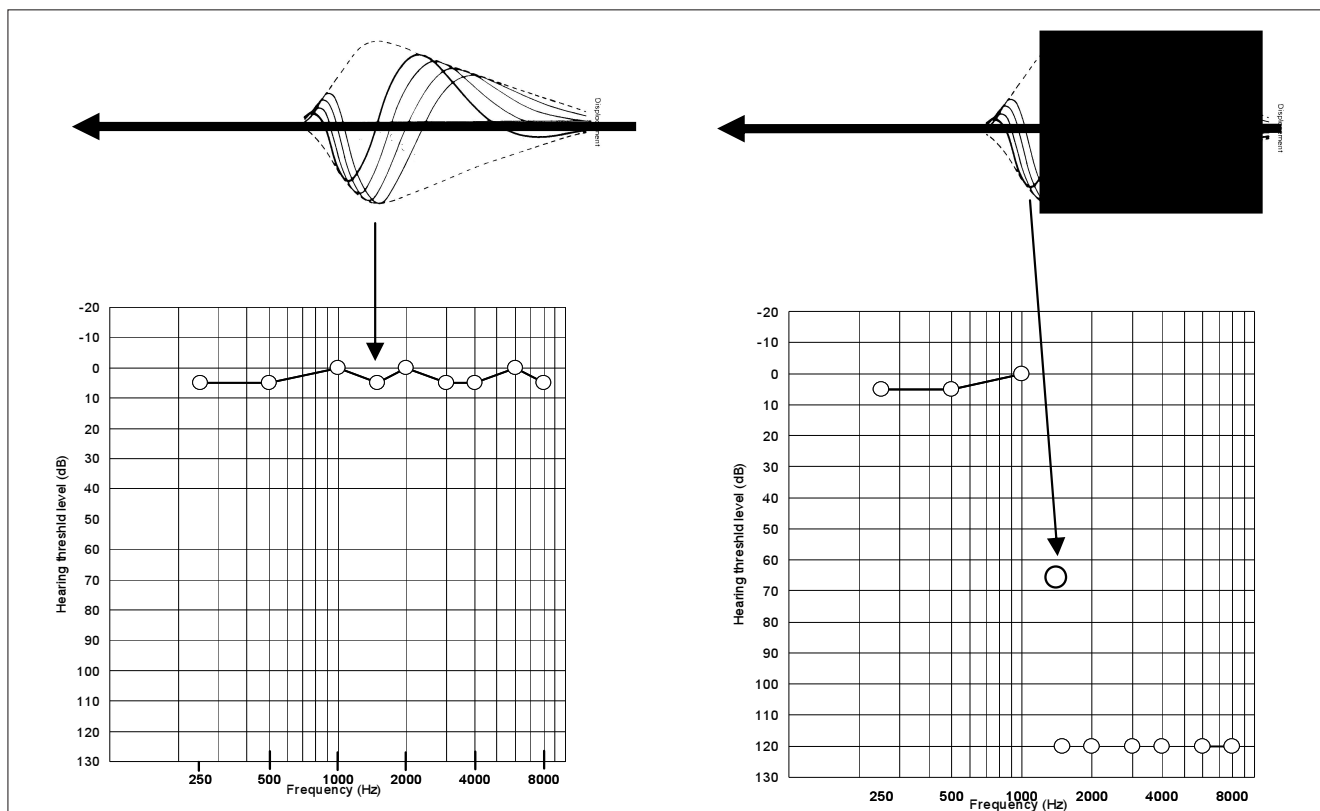
There are occasions when an individual may have a sick region (i.e., IHCs and/or neurones have impaired function but can respond normally at high presentation levels). This may occur, for example, at the transition between a normal low-frequency region and a dead high-frequency region. A pattern of off-frequency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

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 and Moore 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.

There is evidence in the literature to support the presence of DRs in some hearing-impaired humans. IHC damage has been confirmed in histological evaluation of temporal bones in humans. Schuknecht and Gacek (1993) showed that hearing impairment in adults was frequently accompanied by loss to IHCs and/or outer hair cells (OHCs). More recently, AmatuZZi et al. (2001) showed that three newborn babies, who had been in neonatal intensive care units and had failed a hearing screen using the audi-

tory brainstem response, had a loss of IHCs without accompanying OHC damage when examined histologically. (The lack of ABR in the presence of damaged IHCs but normal OHCs is consistent with the umbrella term of “auditory neuropathy”). A further four babies who failed the screen had abnormalities to both IHCs and OHCs.

Therefore, there is evidence that cochlear DRs can occur in adults and children with an acquired or congenital hearing impairment. This 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 and treatment with cisplatin, an ototoxic anti-cancer drug. In newborn rats, Mazurek, Winter, Fuchs, Haupt and Gross (2003) have also shown that IHCs are more susceptible to hypoxia/ischemia than OHCs.



**Figure 1.** The wave of excitation along the basilar membrane for a 1.5 kHz pure tone in a normal hearing listener (left) and a hearing-impaired listener with an extensive high-frequency dead region commencing around 1.5 kHz. The pattern of activity builds up gradually with distance as the wave travels from right to left (basal high frequency to apical low frequency) and decays rapidly beyond the point of maximum displacement. The audiogram form on the left shows normal hearing thresholds. The audiogram on the right is for an individual with a high-frequency cochlear dead region commencing from around 1.5 kHz. Despite the presence of a dead region (and therefore an infinite hearing loss) at 1.5 kHz, the listener is able to detect the signal because the excitation pattern stimulates the IHCs at the 1 kHz place on the basilar membrane i.e., the signal is detected using off-frequency listening.

## Audiometric Pattern and Dead Regions

A review of the literature shows that there is no definitive audiometric pattern associated with DRs; however, 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, the likelihood of IHC damage increases with severity of sensory hearing-impairment. A mild impairment may be due to a combination of OHC and IHC damage, a severe impairment is probably due to a combination of OHC and IHC damage, and a profound impairment is almost certainly 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. 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 (2007a) found that steepness of the audiometric slope was not a reliable predictor of DRs. 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.

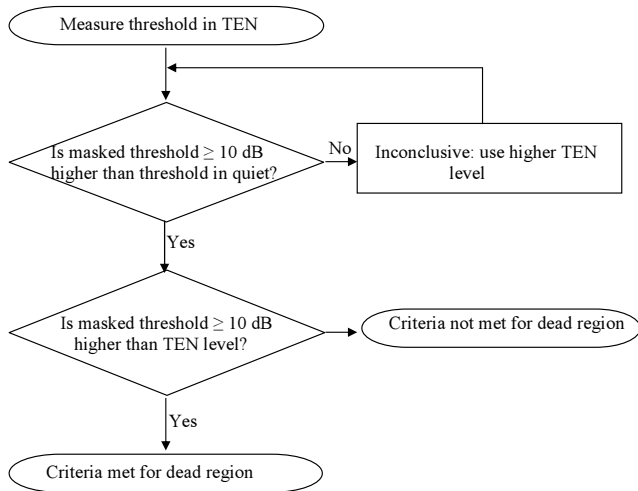
## Identification of Dead Regions

Since a tone that falls within a DR may be detected as 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.

## Threshold Equalizing Noise (TEN) Test

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 and 10 kHz (Moore, Huss, Vickers, Glasberg and Alcantara 2000). A revised version of the test produces equal masked thresholds, in decibels hearing level, between 0.5 and 4 kHz, (referred to as “TEN[HL]” to differentiate it from the original SPL version) and this makes it much easier to use in clinical practice (Moore, Glasberg and Stone 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 (information about the test including how to purchase a copy of the TEN CD can be obtained online at <http://hearing.psychol.cam.ac.uk>). 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). Standard 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) recommend using an ascending step size of 2 dB. Cairns, Frith, Munro and Moore (2007) 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 (and at least 10 dB above the absolute threshold at the test frequency). ERB is the

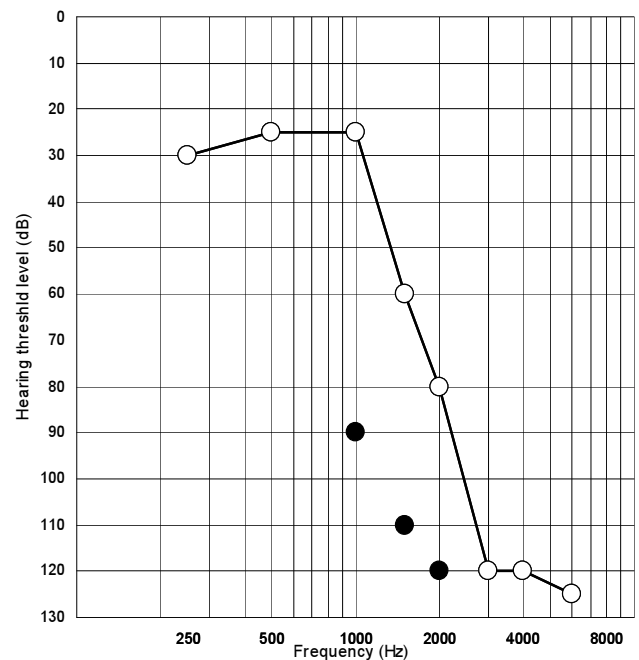
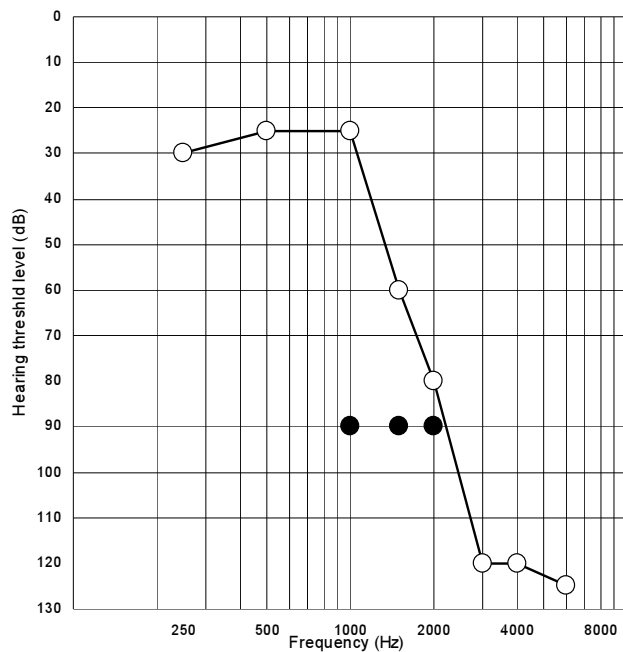


**Figure 2.** Interpretation of the TEN test.

average equivalent rectangular bandwidth of the auditory filter as determined for young, normal-hearing listeners at moderate sound levels. Its value in Hertz is calculated as  $24.7(4.37F+1)$  where  $F$  is frequency in kHz. For example, at 1 kHz the ERB is approximately 0.132 kHz (Moore 2004). 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 dead. Figure 2 summarizes 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 produced by poor processing efficiency in conditions such as auditory neuropathy (Vinay and Moore 2007b).

Figure 3 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.



**Figure 3.** Hypothetical 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.

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 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 would probably 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 later, 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. (2007) carried out a retest within seven days for a group of hearing-impaired adults and a group of hearing-impaired teenagers. A total of three (7.5%) and two (8%) ears changed category in the adults and teenagers, respectively. Munro, Felthouse, Moore and Kapadia (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, Kapadia, Munro and Moore (2006) high-pass filtered the original TEN at 0.5

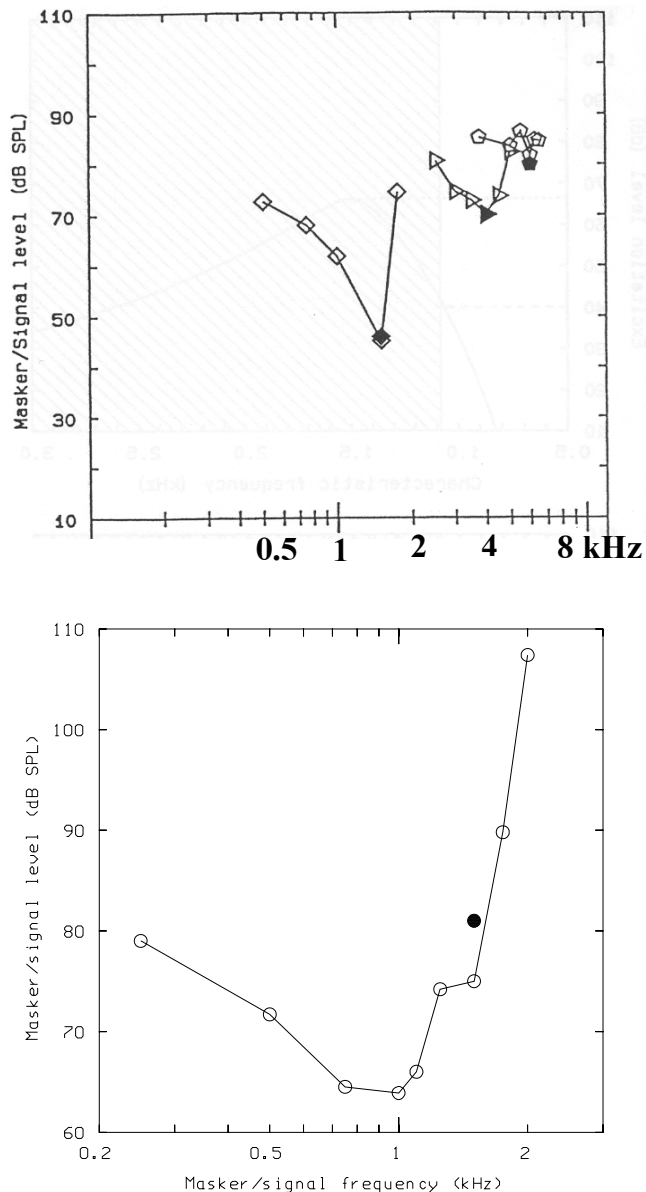
and 1 kHz with some success. The current version of the TEN is band limited between 354 and 6500 Hz. In theory, there is no reason why narrower bands of noise could not be used. For example, if the edge frequency of a DR is thought to be around 2 kHz, then tones that fall within a DR will be masked by 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 center 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).

## Psychophysical Tuning Curves

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 1988). In cases where the signal frequency lies within a DR, the tip will be shifted away from the signal frequency. 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 4.

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



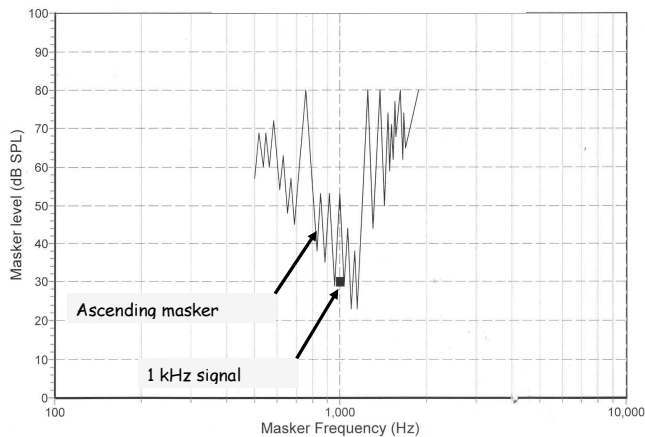
**Figure 4.** Examples of psychophysical tuning curves. The top panel is for a listener with a high-frequency sensory hearing impairment. The filled symbols show the target and the open symbols show the masker. In these examples, the tip of the tuning curve occurs at the same frequency as the target. The bottom 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).

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 whose center 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, Alcantara, Moore, Kluk and Wicher (2005) were the first systematically to evaluate parameters such as rate of change of masker level in order to optimize 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 used by Sek et al. available clinically, we have implemented the fast-PTC method on a PC fitted with a high quality sound card. The software program was developed in our laboratory by Richard Baker for use with a Kamplex KC 35 clinical audiometer fitted with TDH 39 headphones (for further details go to <http://personal-pages.manchester.ac.uk/staff/richardbaker>). The PC was additionally equipped with an 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 (within the limits of the hardware) and direction of the masker sweep.

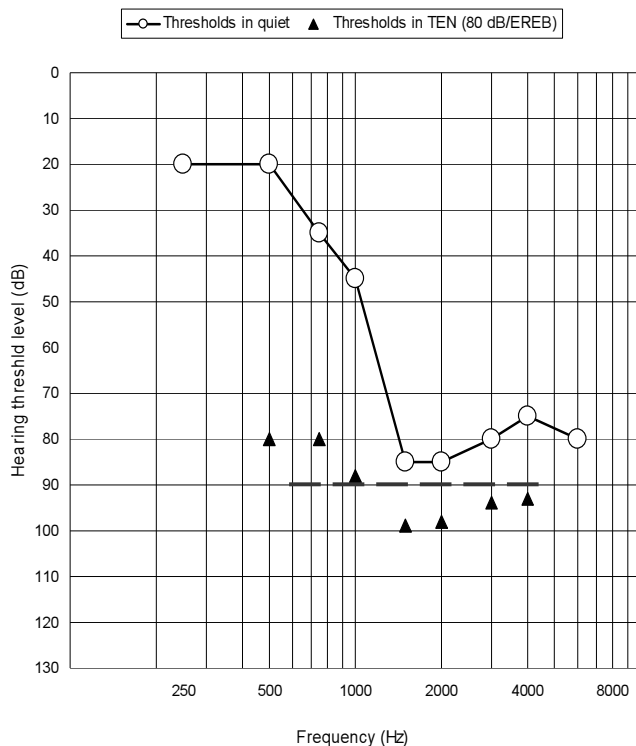
### Fast-PTCs with Children

Diagnosis of DRs may be more difficult with children because psychoacoustic measurements tend to be more variable than with adults. Alicja Malicka and colleagues from our laboratory have investigated the feasibility of measuring fast-PTCs in normal hearing children (Malicka, Munro and Baker submitted) and also hearing-impaired children with and without a DR (Malicka and Munro in preparation). So far, we have been successful at using the technique with children as young as 6 years of age (see figure 5). Subsequently, 12 normal hearing children and a control group of five adults have been tested. The PTCs were measured for 1- and 4-kHz tone signals using ascending and descending masker sweep directions. The results in children show large between-subjects variability. However, there



**Figure 5.** A fast-PTC measured from a normal-hearing 6-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. Data from Malicka, Munro and Baker (submitted).

was no significant difference in the tip estimation between adults and children. The general success rate and test-retest reliability appear adequate for the fast-PTC



**Figure 6.** Data from an 8-year-old 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).

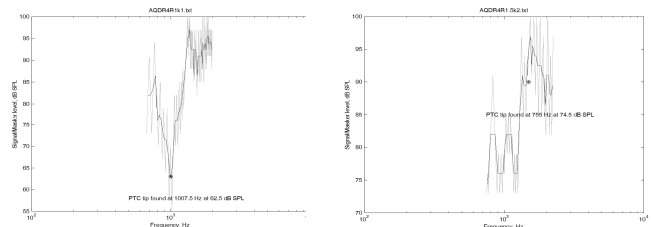
technique to be used with children within the routine clinical setting.

Curran and Munro (2007) have since extended the work of Malicka et al., cited above, by investigating the influence of masker sweep rate, direction of the masker sweep and test-retest reliability in 24 normal hearing children aged 7–10 years.

## Diagnosing DRs in children using fast-PTC and TEN Test

Malicka and Munro (in preparation) compared findings on the TEN (HL) test and fast-PTC in seven hearing impaired children (age 8–12 years). Since children often have elevated masked detection thresholds compared to adults (e.g., Buss, Hall, Grose and Dev 1999), it was not known if the TEN criteria, developed for adults, were also applicable with children. All children were able to complete both test procedures. An example from one child is shown in figure 6. Using the adult criteria, the TEN test showed evidence of DR in nine ears; however the PTC results confirmed the presence of a DR in only six of these ears. Masked thresholds in TEN were elevated, on average, 5.3 dB above the values obtained with adults measured under the same test conditions. This may be due to poorer processing efficiency in children, i.e., reduced ability to extract signal from the noise, or different perceptual criteria used by children. When “15 dB” criteria were used, there was complete agreement between the test findings. Therefore, it may be necessary to use different criteria for interpretation of TEN (HL) test results in children.

Kluk and Moore (2006) 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 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.

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, John, Picton and Moore 2007).

## Prevalence of Cochlear Dead Regions

Prevalence data for cochlear DRs in adults with a sensory hearing-impairment have been provided by Vinay and Moore (2007a). They assessed 317 adults (592 ears) who attended an audiology department, generally for the fitting of a hearing instrument. 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 less, although DRs have been observed in individuals with lower hearing thresholds when diagnosed using PTCs (e.g., Moore et al. 2000). On the other hand, there were occasions when hearing thresholds were as great 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 65 dB HL, or greater.

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. Toal and Munro (2007) reported the prevalence of clinically significant DRs in an adult hearing instrument clinic within the publicly-funded UK National Health Service. Over a four-month period, there were 400 new adult hearing instrument assessments and 62 (15.5%) had high-frequency hearing thresholds of 60 dB HL or greater, extending down

to at least 2 kHz. These adults were investigated for DRs using the TEN test. The test was immediately repeated if the results “just met” the DR criteria and, where possible, a higher TEN level was used. A total of 2.75% (95% CI: 1.57–4.96) of adults met the criteria at one or more frequency. Less than 2% (95% CI: 1.09–2.41) met the criteria for a “clinically significant” DR (5 adults showed evidence of bilateral DRs, and 2 adults showed evidence of a unilateral DR). In summary, the number of new adult hearing instrument referrals with high-frequency hearing thresholds of 60 dB HL or greater was relatively infrequent, and this is reflected in the low percentage of clinically significant DRs. Prevalence data for clinically significant DRs in the pediatric population have yet to be reported.

Vinay and Moore (2007a) 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, Carpenter and Ziegler 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.

Munro (2007) provides a summary of studies have reported the presence of DRs in pre-selected patient groups (Aazh and Moore 2007; Cairns et al. 2007; Jacob, Candido Fernandes, Manfrinato and Iorio 2006; Markessis et al. 2006; Moore et al. 2000; Moore, Killen and Munro 2003; Palma, Bovo, Rescazzi and Prosser 2005 and Preminger et al. 2005). The only studies that have included children or young people are Cairns et al. (2007) and Moore et al. (2003). Cairns et al. tested 23 ears of 15 teenagers with a severe-to-profound hearing impairment who had at least one hearing threshold lower than 80 dB HL. They reported evidence for DRs in three (13%) ears. In an earlier study using a similar population, Moore et al. (2003) reported evidence for DRs in 34 (59.3%) ears. The presence of DRs was probably lower in the more recent study by Cairns et al. 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%.

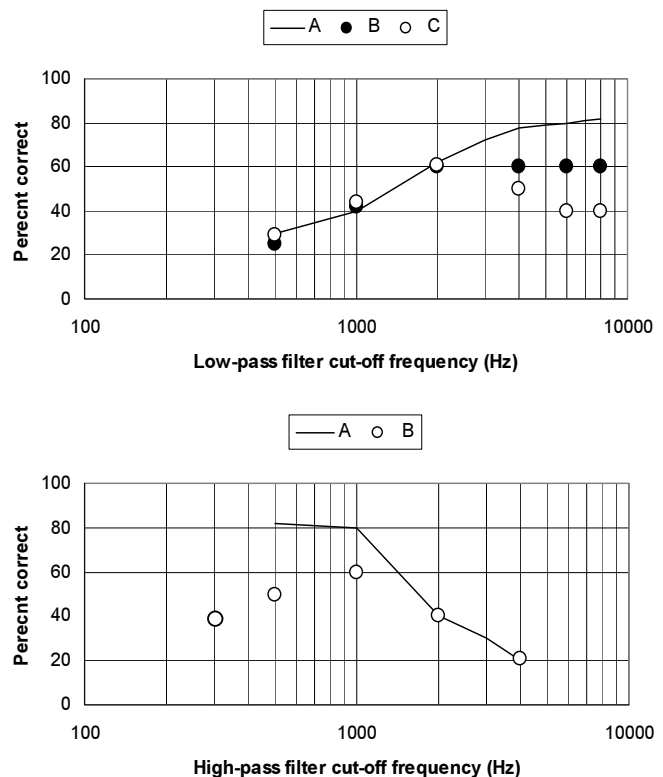
## Implications for Hearing Instrument Fitting in Adults

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, Dillon and Byrne 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, Moore and Baer 2001), background noise (Baer, Moore and Kluk 2002) or both (Mackersie, Crocker and Davis 2004).

## Studies Using Speech in Quiet

Vickers et al. (2001) compared performance in 18 ears with high-frequency hearing-impairments. Twelve ears had DRs and six ears did not have DRs. Subjects listened to vowel-consonant-vowel (VCV) non-sense 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. The top panel in 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 showed a pattern similar to 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.



**Figure 7.** Speech recognition performance of hypothetical subjects with amplification and low-pass filtering (top panel) and high-pass filtering (bottom panel). In the top panel, Subject A (solid line) does not have a dead region but Subjects B and C (filled circles and open circles, respectively) both have an extensive high-frequency 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. In the bottom panel, Subject A (solid line) does not have a dead region; however, Subject B has an extensive low-frequency dead region commencing around 1 kHz. This subject has poorer performance when amplification extends to the very low frequencies.

Vinay and Moore (2007c) carried out a study that was similar in design to that of Vickers et al. (2001), 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 bottom panel in figure 7 shows the outcome from two hypothetical subjects that serve to illustrate the pattern of findings. The scores for subject A improve with decreasing cut-off frequency, i.e., the subject benefits from low-frequency amplification. This pattern is characteristic of subjects who do not have a DR. Subject B has a DR commencing around 1 kHz. Performance improves when amplification is extended a little way into the DR; however, the provision of amplification well within the DR had a deleterious effect on performance. This pattern of finding was present in all subjects with a low-frequency DR.

These findings form the basis for the recommendation to limit amplification within the DR to around 1.7 times the edge frequency (Moore 2004). If there is a low-frequency DR commencing around 1 kHz, there is little point in providing amplification at frequencies below about 0.6 kHz ( $1 \text{ kHz} / 1.7$ ). If there is a high-frequency DR commencing around 1 kHz, there is little point amplifying above about 1.7 kHz ( $1 \text{ kHz} \times 1.7$ ). 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 ( $3 \text{ kHz} \times 1.7$ ). There is some controversy regarding these findings and 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 instruments as fitted by their audiologist. Moore (2004) re-analyzed 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 variables such as severity of hearing impairment.

Mackersie et al. (2004) compared performance in 16 ears with high-frequency hearing impairments. Eight ears had DRs and eight ears, matched for audiometric 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 v4.1 (Cornelisse, Seewald and Jamieson 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 1.7 times 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,

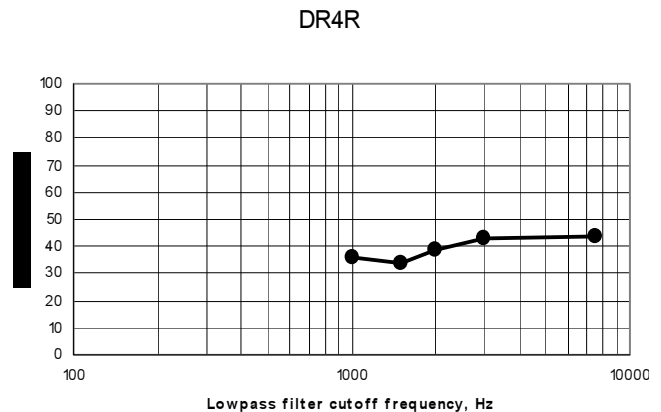
despite similar audiograms. The DR patients also reported less benefit from amplification in noise.

Keidser and Dillon (2007) cite a study of Ching, Dillon, Lockhart, van Wanrooy and Carter (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 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) showed 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 appeared 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 (2007a) 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 high-frequency DRs when assessed in quiet or noise (and some show a deleterious effect); ii) listeners with less extensive high-frequency DRs may show limited benefit from high-frequency amplification in environments that have poor SNRs; and iii) low-frequency amplification is not beneficial, and in most cases shows a deleterious effect, when listeners have a low frequency 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 frequencies. The use of frequency transposition, in general, has produced mixed findings although interesting new developments show considerable promise (see Chapter 13 by Scollie et al. in this volume). There is also emerging evidence from the work of Robinson, Baer and Moore (2007) that there may be some benefit to tak-



**Figure 8.** Performance for a child with an extensive high-frequency dead region commencing from around 1.5 kHz (see Figure 6). The percent correct score on the VCV test is plotted as a function of low-pass filter cut-off frequency. Data from Malicka, Munro and Baer (2008).

ing information that falls well within a DR and recoding it to around the boundary of the DR.

## Implications for Hearing Instrument Fitting in Children

Few studies have investigated the benefit of high-frequency amplification in children, and none, as far as we are aware, have specifically investigated this in the context of DRs. Therefore, the optimal frequency-gain characteristics for children with DRs have yet to be established. 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, Faulkner and Wilkinson (1999) have demonstrated that normal adult listeners can rather quickly learn to make use of high frequency information that is shifted to lower 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.

We are currently investigating the benefit of high-frequency amplification using VCV stimuli presented in quiet and in noise (Malicka, Munro and Baer 2008). 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 8. 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.

## Summary

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 reliably without the use of further test procedures other than the audiogram. One of these further procedures, 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. Adults with extensive high-frequency DRs do not appear to obtain the same benefit from broadband amplification as those without DRs, although there is

some controversy in this regard. A summary of the pediatric studies conducted in our laboratory (and reported in this chapter) is provided in table 1. In our experience, most school-age children are able to perform the fast-PTC measurements; this means we have the potential for a clinical test procedure. Some hearing-impaired children have DRs when assessed using the fast-PTC and the TEN test, although the criteria for detecting DR with the latter may not be the same as with adults. 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. Therefore, we currently recommend using the frequency-gain targets of one of the recognized hearing instrument fitting rationales for all pediatric fittings, irrespective of presence of DR.

## Acknowledgements

The pediatric studies reported in this chapter were funded by Phonak AG, Switzerland, and the National Deaf Children's Society, UK.

**Table 1.** Summary of dead region studies from our laboratory.

Study	Aims	Findings
Cairns, Frith, Munro and Moore (2007)	Repeatability of TEN test in hearing-impaired adults and teenagers	Good repeatability. Advise immediate retest for ears that 'just meet' DR criteria.
Curran and Munro (2007)	Parametric study of fast-PTCs in normal hearing school-age children	No statistically significant difference in tip frequency of PTC between the two masker sweep rates used in the study. Small hysteresis effect related to the direction of the swept masker.
Malicka, Munro and Baker (submitted)	Feasibility of using fast-PTC with normal-hearing school-age children	Children show more variability than adults. The good success rate and reliability are adequate for use with children within the routine clinical setting.
Malicka and Munro (in preparation)	Diagnosing DRs in hearing-impaired school-age children using fast-PTC and TEN test	Some hearing-impaired children have DRs when assessed using the fast-PTC and the TEN test although the criteria for detecting DR with the latter may not be the same as with adults.
Malicka, Munro and Baer (2008)	Effect of amplification on the speech intelligibility of hearing-impaired school-age children with and without DRs	No deleterious impact from amplification with frequency-gain characteristics based on DSL fitting procedure.
Markessis, Kapadia, Munro and Moore (2006)	Modified frequency response of TEN in adults	High-pass filtered TEN used with some success
Moore, Killen and Munro (2003)	Application of the TEN test to hearing-impaired teenagers	Dead regions relatively common when using the SPL version of the test and ascending step sizes of 5 dB (see Cairns et al for HL version of TEN test). Test often inconclusive when severe loss because of inability to tolerate high TEN level or limited by maximum output of audiometer.
Munro, Felthouse, Moore and Kapadia (2005)	Reassessment of DRs in hearing-impaired teenagers	Little evidence of change in interpretation of TEN test when reassessed after 12 months.
Toal and Munro (2007)	Prevalence of DRs in UK adult hearing aid population	Prevalence of extensive dead regions relatively low in a UK adult hearing aid clinic.

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