Balancing diagnostic considerations and the benefits of early intervention in the management of Auditory Processing Disorder: A case study report

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The SoundSkills project to develop a scalable model multi-disciplinary specialist auditory processing disorder (APD) clinic is in its fifth year. One of the many issues faced is the lower age limit at which APD can be diagnosed and intervention initiated. At our free monthly APD seminars for parents and professionals we often ask the question “At what age have you heard APD can be diagnosed?” The answer is usually seven years. This is a common belief and practice though some clinics in New Zealand and overseas only see APD cases from age eight. The belief that management of APD should be deferred until age seven is attributed to three factors. Firstly, some early APD tests were only normed on children down to age seven. Secondly, parts of the central auditory nervous system do not fully mature until adolescence or young adulthood, especially in the case of myelination of the corpus callosum which can continue into the early 20s (Yap et al, 2013). Thirdly, and in part because of these maturational factors, behavioural measures on young children show greater variability than behavioural measures on older children; hence behavioural test results on young children have to be interpreted with caution. However, these factors do not impede early intervention for other types of deafness.

Early Diagnosis and Intervention
In fact several APD tests have norms for children younger than seven. One of us, as a graduate student in the same department, was privileged to observe the remarkable work of Dr Susan Jerger on the development of the Pediatric Speech Intelligibility (PSI) dichotic sentences test and its validation with site of lesion studies on children as young as three years with brain lesions, mostly tumours, in the NeuroSensory Centre, Baylor College of Medicine (Jerger, 1987). Stollman et al (2004) describe the development of a central auditory test battery for four to six year old Dutch children. Smart, Purdy and Leman (2012) report the evaluation of two central auditory tests and a phonological awareness assessment tool in six year olds. Hall (2014) states

“Based on application of available procedures, including auditory evoked responses as needed, it’s certainly possible to rule out or strongly suspect APD in children as young as 5 years old. …. FM technology is sometimes very helpful in young children with below-normal performance on APD tests. We do know that early intervention for APD is most effective, and effective management can minimize or eliminate secondary psychosocial and academic problems.”

Table 1 shows the lower limit of age norms for some APD and auditory skill assessment tools. These tools enable assessment of some auditory skills from age three or three years six months upwards with the proviso that it is important to keep in mind the variables and cautions mentioned above. The older the child, the more skills can be assessed. Note that the table shows examples only, not an exhaustive list, of available assessment tools for young children.
Table 1: Age ranges of norms for selected tests and test batteries suitable for assessment of auditory skills in young children

<table>
<thead>
<tr>
<th>TEST</th>
<th>AGE RANGE OF NORMS (Y:M)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pediatric Speech Intelligibility (PSI) Test</td>
<td>3:0 – 6:0</td>
</tr>
<tr>
<td>CELF Pre-School 2</td>
<td>3:0 – 6:11</td>
</tr>
<tr>
<td>Auditory Skills Assessment (ASA)</td>
<td>3:6 – 6:11</td>
</tr>
<tr>
<td>SCAN-3:C</td>
<td>5:0 – 12:11</td>
</tr>
<tr>
<td>Differential Screening Test for Processing</td>
<td>6:0 – 12:11</td>
</tr>
<tr>
<td>Listening in Spatialized Noise – Sentences (LiSN-S)</td>
<td>6:0 – 60:0</td>
</tr>
</tbody>
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Correction of interaural asymmetry evidenced on dichotic tests (amblyaudia) using the Auditory Rehabilitation for Interaural Asymmetry (ARIA) method (Moncrieff and Wertz, 2008) is SoundSkills’ most frequent intervention. Approximately 50% of children with APD have amblyaudia or other dichotic performance deficits (Moncrieff, 2006). Using the Randomised Dichotic Digits Test (Moncrieff and Wilson, 2009) and Dichotic Words Test (Moncrieff, 2011) we currently have norms for diagnosis of amblyaudia from age five (5:0). Moncrieff (2011) reports in regard to children from age five that “despite the variability of results and the interactions of gender and handedness seen with digits testing, children below the age of 7 years can perform ... dichotic listening tests and ... normative data from children within this youngest age group can and should be measured”.

The American Academy of Audiology CAPD Clinical Practice Guidelines (AAA, 2010) provide limited guidance on diagnosis and management of APD in younger children:

“A limited number of behavioral auditory measures have been developed for use with younger children.... Use of measures such as these, coupled with behavioral checklists.... can provide insight into children who may be “at risk” for (C)APD, leading to recommendations for close monitoring of skills, enrichment activities designed to develop and augment auditory skills.... and regular follow-up to determine the appropriate diagnosis as early as possible....

Early identification followed by intensive intervention exploits the brain’s inherent plasticity. Successful treatment outcomes are dependent on stimulation and practice that induce cortical reorganization (and possibly reorganization of the brainstem), which is reflected in behavioral change (i.e., learning)...”.

What is the responsibility of an audiologist faced with the case of a pre-schooler or new entrant in whom peripheral hearing loss and auditory neuropathy have been ruled out, yet there are compelling symptoms of hearing difficulties reported by multiple observers? It goes against all our training and belief in the importance of early intervention to send these children away for two or three years until they turn seven. Animal studies identifying early critical periods for central auditory nervous system (CANS) development (Clause et al, 2014; Penhune and de Villers-Sidani, 2014) and mitigation by auditory training (Kang et al, 2014) support the importance of early intervention for APD.
Audiologists do not require a clearcut diagnosis as a prerequisite for intervention with other types of deafness. In infants, hearing aid fitting can proceed based on limited data such as two estimated audiometric thresholds. Young children with possible central auditory skill deficits but who lack a clearcut diagnosis may benefit from intervention. It is important to note that APD interventions such as mild amplification with remote microphone hearing aids (RMHAs), frequently referred to as “personal FM systems” (Keith and Purdy, 2014), phonological awareness therapy and language therapy are not harmful and indeed may be beneficial to many children with or without APD.

Sometimes children referred to SoundSkills for central auditory evaluation have a long history of otitis media with effusion (OME). Auditory deprivation from recurrent OME has been implicated as a cause of APD (Whitton and Polley, 2012). It is possible, based on evidence from animal research, that some of the children we see may have neuro-anatomical abnormalities in the CANS as a result of auditory deprivation secondary to OME. Some may simply be delayed in their auditory and/or phonemic skills development as a result of the OME. In either case intervention may be warranted. Sometimes the hearing deficits we see are just one facet of more global developmental problems and the comorbidities preclude a definitive diagnosis of APD. Once again the presence or absence of a definitive diagnosis or known cause does not negate the value of intervention.

Our experience has been that few very young children with reports of hearing difficulty in the presence of normal peripheral hearing are referred to our clinic, but when they are the symptoms are usually severe. Our audiological, cognitive and language evaluations usually support the presenting complaints, and sometimes raise the possibility of comorbidities for which additional referral may be recommended or arranged. SoundSkills’ consultant ENT Surgeon serves as a first point of referral when there are possible neurological or ENT concerns, and referral for auditory brainstem response audiometry (ABR) is arranged where acoustic reflex results do not rule out auditory neuropathy spectrum disorder (ANSD) (Berlin et al, 2005).

Suspected diagnosis of APD in children below the usual age range for testing is qualified by SoundSkills as being “at risk for”, “provisional”, or “criteria for diagnosis not met but auditory skill deficits are confirmed”. Follow-up is scheduled to enable more comprehensive and definitive testing when the child is older. Our experience is that we usually confirm the presence of APD in “at risk” children at later evaluations. A provisional diagnosis in young children is helped by independent observation (eg preschool/primary teacher), questionnaire data when possible, and history so that there is a coherent picture of hearing difficulty. We are careful to ensure that parents understand the limitations of evaluation of young children. As with any specialist area of audiology, APD evaluation of young children should only be undertaken by clinicians regularly working in the area, familiar with the tools, and experienced in diagnosing APD.

Care must be taken to ensure fatigue and poor attention do not adversely affect behavioural measurements in children of any age. The ARIA protocol for amblyaudia treatment, for example, involves 20 minutes practice, 20 minutes rest, then another 20 minutes practice in each weekly one hour therapy session (Moncrieff and Wertz, 2008). Our cognitive, language and hearing evaluations are spread over at least two sessions on different days, with breaks as needed during individual sessions. If necessary, children are brought back for additional testing at a separate session at no
extra cost. Our drawing wall, an entire wall on which children can write and draw, makes rest breaks highly popular with our young clients. For the out of town clients who comprise a significant proportion of our caseload, the Parenting Place café and children’s play area downstairs provide a venue for longer breaks if we are scheduling more than the usual number of assessments over the course of a day.

When confronted with our first case of a young child outside of the usual APD assessment age range, a four year old with a psychologist, pre-school teacher and parents all suspecting hearing loss, our team comprising audiologists, a speech language therapist and special education advisers considered that we had an ethical obligation to help the child. Our limited evaluations and parent and teacher questionnaires supported the presenting complaints. We made a diagnosis of “at risk for APD” and decided on a trial intervention with RMHAs. In this case the intervention was successful and we were emboldened to take similar action in other cases.

Case studies
The following two cases from our current caseload, one from within and one from outside Auckland, report the youngest children in whom we have attempted intervention for possible central auditory dysfunction.

Case 1

Name: M
Gender: M
Age at assessment by SoundSkills: 3y 10mo
Referred by: District Health Board (DHB) team; Paediatrician and Speech Language Therapist

History:
In addition to suspected hearing difficulties M is delayed in gross and fine motor development and speech development. He demonstrates learning difficulties at preschool and looks for visual cues when confused. He has difficulty comprehending instructions, difficulty hearing in noise, is upset by
loud environments and is sometimes overwhelmed by the preschool environment. His speech language therapist (SLT) reports that he has “difficulty processing information and understanding spoken language”. M has a history of recurrent OME and grommets. He is or has been under the care of a paediatrician, ENT Surgeon, speech language therapist, occupational therapist, physiotherapist and special education services.

Evaluation results:
Tests of peripheral hearing showed a slight low frequency hearing loss. Tympanometry and otoscopy showed a Type A tympanogram on the right and a patent grommet on the left. M scored at the 4th percentile on the Auditory Skills Assessment (ASA) battery. CHAPS and SIFTER questionnaires (Smoski et al, 1992; Anderson and Matkin, 1996) indicated hearing difficulties in preschool relative to M’s peers.

Conclusions:
Criteria for a diagnosis of APD were not met, but the observation of hearing difficulties was confirmed. M particularly had difficulty hearing in the presence of a competing speaker.

Recommendations:
The management plan recommended

- that the parents attend an APD seminar
- deficit-specific language therapy (referred to Ministry of Education (MoE) speech language therapist (SLT))
- home use of auditory training software under therapist supervision
- a trial with remote microphone hearing aids
- reassessment when M turns 5.

Progress report on intervention:
“The SoundSkills Education Adviser has organised a controlled RMHA trial with measurable goals and observations in preschool in conjunction with the parents and preschool teachers. The preschool teachers have reported that the RMHAs seem to have had a calming influence on M, that he stutters less and has less difficulty finding words when he wears them, that he definitely hears the teachers better with his hearing aids, and that his attention span is better when wearing them. M’s mother reported that the family has noticed a big difference in his ability to understand and interact and friends have commented on it. M’s pronunciation, speech and singing are much clearer. In the car he doesn’t say ‘what’ anymore. M also used his RMHAs at the supermarket and “they were great”. M didn’t join in conversations before and would withdraw but participates now. When his mother reads to him he is more focused, receptive and interactive. He has them in an average of five hours a day at home. He doesn’t like having them in initially but then forgets about them once they are in. His speech language therapist notices a distinct improvement when he wears them for speech therapy. The trial is progressing; post-fitting preschool observations have not been completed, but with the positive indications to date it is likely M will retain the hearing aids.”

Case 2
S
Gender: M
Age at assessment by SoundSkills: 3y 7mo
Referred by: Parents

History:
S’s parents report that S has difficulty comprehending verbal instructions, especially multiple part instructions. He is sensitive to loud noise and can be overwhelmed by loud environments and will remove himself from them. He has difficulty with verbal expression and comprehension, his speech is difficult to understand and his sentence structure can be incomplete. Kindergarten staff have noticed listening and learning difficulties and referred S to MoE for special education support. There is a family history of learning difficulties. S has had previous speech, language and audiological assessments.

Evaluation results:
Peripheral hearing assessment results were normal. On the ASA battery S scored in the “Low” range with a percentile rank of 1. The SIFTER questionnaire completed by S’s kindergarten teacher showed difficulties for all areas probed.

Conclusions:
While it was impossible to separate the effects of language versus auditory delay when interpreting the questionnaire and behavioural observation results, and criteria for a diagnosis of APD were not met, some specific deficits involving auditory skills were confirmed by the ASA results. These included hearing in noise, and mimicry.

Recommendations:
The management plan recommended
- a full language assessment
- that the parents attend an APD seminar
- speech and language therapy (subsequently accepted onto MoE SLT caseload)
- learning support (MoE subsequently requested copy of SoundSkills report for early intervention team and Adviser on Deaf Children)
- home use of auditory training software under therapist supervision
- a trial with RMHAs
- reassessment in 12 months.

RMHA trial outcome:
Over the course of a four month trial (with interruptions) with RMHAs the SoundSkills Education Adviser observed some listening skill improvements but overall the benefits were minimal, in part because the RMHAs were not being used at home and the observed improvements were confined to formal learning situations which comprise only a small part of the preschool day for this child. The RMHA trial was discontinued with a plan to try again when S starts school. Other interventions including speech and language therapy and learning support are in place. Guidance on software-based home therapy and the use of audio books will be offered to the parents.

Discussion
In both cases reported above the parents are pleased with what has been learned from the assessment and the RMHA intervention trial. Mostly they are pleased that someone is providing guidance and intervention to help them cope with their child’s obvious difficulties.

The involvement of a specialist Education Adviser to liaise with the preschool, provide teacher guidance, carry out observations, initiate and monitor a trial with measurable objectives and facilitate a final decision on whether to continue amplification is as critical at preschool level as it is at primary or high school. We observed with these young clients that hearing, listening, comprehension and participation difficulties noted during preschool observation by an experienced observer were similar to hearing, listening, comprehension and participation difficulties observed in older children in class. Because there is little formal teaching or “mat time” in preschool, the potential value of RMHAs for lessons is limited and the value of amplification to an individual child depends more on the degree to which it is helpful in home and family situations. It could be argued that conventional hearing aids may be more practical for some preschool and domestic situations even though they give considerably less signal to noise ratio improvement than RMHAs. The dilemma for an evidence-based practitioner is the poor evidence base for conventional hearing aids for APD versus the excellent evidence base for RMHAs (Keith and Purdy, 2014).

The NZ Government Review of APD services (Esplin and Wright, 2014) emphasises the importance of “whole of life” (ie not just in school) amplification for children with APD in contrast to “a singular primary need only in the classroom”:

“Note all parents interviewed, except one, whose child had a personal FM system reported the importance of their child having access to it outside the classroom. This was for other educational pursuits and also home and wider community life. For some it was a safety issue.”

Intervening without a definitive diagnosis highlights the need for a test of everyday speech perception to verify and quantify auditory skill deficits in typical listening situations and provide a baseline for pre and post intervention measures in children with either suspected or diagnosed APD. We need to be able to evaluate performance on the three parameters that cause most difficulty for children with APD; soft speech, speech in competition, and speech passages of moderate (or greater) length. The University of Queensland Understanding Everyday Speech Test (UQUEST) (Kei et al, 2003) is one of several assessment tools we are evaluating in this regard.

At SoundSkills our diagnostic and intervention methods are evolving constantly. The area of management of APD is less mature than some other areas of audiology and protocols are constantly under review. The recently released New Zealand Government review of APD (Esplin and Wright, 2014) notes the limited consensus on some aspects of APD management internationally. In time, objective tests of central hearing function such as late auditory evoked potentials will assist us in diagnosing APD and language processing difficulties more accurately and at younger ages. For example, Jansson-Verkasalo et al (2004) reported a study in which Mismatch Negativity (MMN) discriminative cortical auditory evoked responses detected deficient phonological representation in four year old children who were preterm and very low birth weight. Moreover, the study showed that the abnormal MMN responses observed at age four predicted object naming difficulty at age
six. For the present however, we lack established objective clinical tools for diagnosing APD in young children (Sharma and Purdy, 2014; Wilson et al, 2013). As APD practitioners we need to provide management for children right now, and thus we are compelled to make challenging clinical decisions and push traditional boundaries more often than is the case in longer established areas of audiology.

References


