King-Kopetzky Syndrome? A bio-psychosocial approach to adult ‘APD’

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Abstract

This paper discusses the relationship between King-Kopetzky Syndrome and Auditory Processing Disorder. King-Kopetzky Syndrome is the term applied to adults who present for help in the presence of normal audiometric thresholds. These adults may or may not have an auditory processing disorder and this paper discusses the factors that influence help-seeking and what is known about audiological diagnosis and management.

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Mr Smith has noticed increasing difficulty hearing in group situations. He is concerned that his hearing may be getting worse and seeks help from his Doctor. The doctor refers him to an Otolaryngologist for a medical inspection. No obvious cause or site of hearing disorder is apparent. Mr Smith presents with normal audiometric thresholds. The Otolaryngologist refers him on to the audiologist for further help.

Why do some people complain of hearing difficulties when their hearing thresholds are normal? Adults presenting with disabling hearing difficulties (despite apparently normal hearing thresholds) have been the subject of much research interest and clinical speculation over the years. This is a common occurrence accounting for an estimated 5-10% of all referrals to otolaryngology (Saunders and Haggard, 1989). These cases challenge audiologists to tolerate uncertainty about underpinning causes, best actions for investigation and best treatment options.

This article will discuss how help seeking and symptom detection occur. It will identify the philosophical perspectives that underpin the research to date and examine what this information contributes to our clinical knowledge. It will consider the underlying factors that contribute to an individual seeking help for a hearing problem and what evidence we have about what really helps.

The most commonly applied clinical label for this experience of hearing difficulties with normal audiometric thresholds is ‘Auditory Processing Disorder’, but it has also been referred to as ‘King-Kopetzky Syndrome’.

King Kopetzky Syndrome refers to a wide heterogeneity in presentations, symptoms and underpinning factors. It refers exclusively to adult presentations. Some individuals who present for help may have an auditory impairment but not necessarily (Pryce et al 2010).

Is there a difference between King-Kopetzky syndrome and Auditory Processing Disorder?
There remains ambiguity about whether a disorder identified in children is the root cause of adult presentations with hearing difficulty. Simply because both groups might present for help with normal audiometric thresholds does not mean that the underlying basis can be assumed to be the same. The research literature is dominated by investigations of auditory processing in children, which is then considered to apply to adults as well (AAA, 2010; Moore and Rosen, 2013). Studies focussed on adults only are often overlooked (not least because the APD literature does not reference the King-Kopetzky syndrome literature). The literature that deals exclusively with adult presentations is found under the term ‘King-Kopetzky syndrome’ or ‘Obscure Auditory dysfunction’.

The bio-medical perspective is probably the dominant view held by both clinicians and patients alike (Wade, 2006). If someone is complaining of hearing difficulties, the most likely starting point for the health practitioner is to consider the possible biological causes of such difficulties and check all auditory functions (both peripheral and central) that could have become impaired. Therefore ‘Auditory Processing Disorder’ (AAA, 2010) is the most frequently used clinical label for such patients in audiology in the US and UK. But is this label adequate? In such cases adult patients frequently describe their childhood experience of hearing as normal, without any schooling or other difficulties, though there may be more complicated social factors which influence their decision to check their hearing.

There are implicit philosophical differences in perceived versus objectively observed disorders. It is not inevitable that all presentations with hearing difficulties must be dependent on processing impairment. This corresponds with a dualist separation of mind and body and a belief in the objective presence of an underlying pathology in all cases.

Yet this position is not consistent with what we know about help-seeking behaviours, symptoms perceptions and the way individuals evaluate their health. Dualism is not a helpful model as mind/body interactions are demonstrated in most aspects of health and well being (Balint, 1957). People seek help as a result of multiple factors including impairments in peripheral and central auditory pathways but also as a result of psychological and social factors (Lazarus and Folkman, 1984; Moller –Leimkuhler, 2002; Cramer, 1999 ; Shaw, 2001).

A ‘syndrome’ is defined a set of symptoms that occur together and characterise a particular abnormality in function (http://www.oxforddictionaries.com). This terminology does not pre-judge symptoms as dependent on impairment in processing. For example, a change of lifestyle to a new noisy and stressful listening environment may contribute to someone’s decision to seek help (Pryce, 2003). It may be that their function is not innately impaired but the stress of managing communication in a difficult listening environment has led to concerns about their hearing (Borg and Stephens, 2003). In these cases a diagnosis as having an inherent auditory processing disorder may carry unintended consequences and even
harm... There are increasingly recognised risks of pathologising and over-diagnosing health conditions which may be subject to social and environmental factors (Moyinhan, 2013). Audiologists, in keeping with most other healthcare professionals are under increasing pressure to be mindful of the risk of over diagnosis (Moyinhan et al, 2013). Clinicians are challenged to identify and tease out the scientific evidence from amongst professional preferences and patient desires. As in most areas of audiology it is difficult to see a consensus on one single way to proceed.

Arguably, for adults, it is entirely normal to experience changes in our auditory perception over time (as we age, for example). Our colleagues in medical sociology and health psychology might regard these biological changes as less relevant to clinical presentation. Medical sociologists might consider this as pursuing a socially sanctioned ‘sick role’ (Parsons 1964) which relieves our patient, Mr Smith, of social responsibility, elicits sympathy and justifies communication problems. A health psychologist might consider Mr Smith as responding to a series of cognitions about his hearing performance. He is evaluating it according to individually held beliefs about normal and impaired hearing performance and the consequence of these (Leventhal et al, 1984).

In summary our knowledge about our own health, including our hearing performance, is relative and varies according to our lay beliefs and expectations. We may seek help on the basis of our perceptions of illness and normality, rather than on the basis of objective signs alone (Lazarus and Folkman, 1984).

As Figure 1 illustrates the clinical label King-Kopetzky Syndrome encompasses several causal factors.

Fig. 1 An illustration of the relationship between King-Kopetzky Syndrome and causal factors.
The World Health Organisation (WHO) proposes an internationally agreed framework in which biological, psychological and social factors are deeply interwoven in our experience of health and wellness (WHO, 2001). The risk of adopting a narrow, disease led model of understanding this type of hearing problem is that we fail to recognize the human needs of the patient seeking help. We are in danger of over simplifying a complex problem into a purely biological cause.

King-Kopetzky Syndrome is the proposed term for adults who seek help in the presence of normal audiometric thresholds. They may or may not have an auditory processing disorder. This definition recognizes that for some people the problem is due to anxiety and perception of performance.

Zhao and Stephens (2000) identify seven sub-categories of King-Kopetzky Syndrome:-

1. Middle ear dysfunction
2. Mild cochlear pathology
3. Central/medial olivocochlear efferent system dysfunction
4. Purely psychological problem  
5. Multiple auditory pathologies  
6. Combined auditory dysfunction and psychological problems  
7. Unknown

This heterogenous model of difficulty has in recent years been adopted by the definitions of Auditory Processing Difficulty (BSA, 2011). The British Society of Audiology Special Interest Group in APD now consider acquired APD in adults and children to exist in the presence of or as a result of peripheral hearing loss, including transient and treated hearing loss (BSA 2011); certainly the role of memory and attention is now considered important in ‘developmental’ (or acquired) APD (Moore and Rosen, 2013). Memory, attention and cognitive function can be compromised by numerous factors, including stress (Jeansok et al, 2002). King-Kopetzky syndrome pre dates this revised interpretation of auditory processing disorder and none of the literature is referenced in the position statements on APD (eg. BSA, 2011; AAA, 2010). This impacts the application of guideline to adult cases. There has been relatively little research into the effects of these hearing difficulties (Pryce, 2003; King and Stephens, 1992) or their treatment (Zhao et al, 2008).

**The paradox of help-seeking**

Why do these patients seek help when so many with measurable and disabling hearing losses do not? One of the main curiosities in this area is that these patients seek help with relatively small changes in function. If we accept current estimates, then on average someone will experience hearing difficulties which mask participation in conversation and limit their function for ten to twenty years before they seek help with it (Brink, Van Den et al 1996; Carson 2000; Getty and Hetu, 1994; Kyle Jones and Wood, 1985; Watson and Crowther, 1989). We hear much of the stigma of hearing loss and the barriers to help-seeking (Wallhagen, 2010). Regardless of where a lesion might actually lie, the disorder (if indeed one can be identified) does not accurately predict the perceived limit to function and participation. The answer appears to come from models of illness perception, well understood in health psychology. In order to identify ill health, people are thought to compare their symptoms with prototypical symptom sets (Bishop and Converse, 1986). Illness perceptions also guide cognitive appraisals of the symptom sets (Leventhal, 1990). In other words a person compares their bodily ‘signs’, in this case, how well they appear to hear in a particular environment with their expectation of how well they think they should hear based on their perception of those around them, what they consider to be ‘normal’ for the environment and noise level, people their age and in their position etc. So those who seek help with hearing symptoms do so on the basis of internally held schema about what is ‘normal’.
Where does ‘King-Kopetzky’ come from?

Samuel Kopetzky described the problems of hearing in radio operators returning from the second world war. He conceptualized these cases as having ‘loss of capacity for discriminative listening’ (Kopetzky 1948; Saunders and Haggard, 1989). These individuals had spent many hours endeavouring to decipher enemy signals over radio. They later presented with concerns that their hearing was affected. Kopetzky notes that there was considerable anxiety around this evaluation of their hearing. Air Vice Marshal Peter King developed this work with a fuller description of possible causes in 1954. The term ‘King-Kopetzky syndrome’ was coined by Ron Hinchcliffe in 1992. Hinchcliffe reviewed what was known about such hearing difficulties and concluded that these were manifestations of ‘auditory stress disorder’. He includes the following early description:

‘He was a worried, tense man extremely anxious lest his defect should be the cause of a disaster, particularly when told that his hearing was normal.’ (Hinchcliffe, 1992)

The psychological dimension

One of the difficulties in interpreting Auditory Processing Disorder data are that rarely are help-seekers and non help-seekers compared. Help-seeking is key to the clinical categorisation and diagnosis. Therefore, it is important to consider a comparison between those who do and do not seek help with the same symptoms.

Investigations of King-Kopetzky syndrome have included psychological appraisal. Stephens et al (2003) identified that self-rated participation restriction varied with cognitive appraisal. Likewise cognitive appraisal was important in describing the maintenance of hearing difficulties (Pryce, 2003). The Crown Crisp Experiential Index (Crown and Crisp, 1956) has been used to examine psychoneurotic traits in those with hearing difficulties and normal audiometry and those with chronic pain. This index comprises items measuring levels of anxiety, phobic anxiety, obsessionality, somatic anxiety, depression and hysteria. When compared with control subjects without any reported pathology, more phobic and free floating anxiety were noted in those with hearing difficulties. When compared to chronic pelvic pain the psychoneurotic traits were similar but hearing performance (measured as speech in noise) was worse (Saunders and Haggard 1993). The examination of psychological traits has suggested that this group of patients experience higher rates of anxiety. The direction of causality between anxiety and hearing difficulty is debateable. To aid interpretation, qualitative work has modelled how anxiety is experienced and how it links to hearing difficulties. This suggests that anxiety has a role in maintaining awareness of symptoms by sensitising individuals to particular communication environments or partners. (Pryce, 2003; Pryce, 2006).

Patients describe in detail how the way they interpret their symptoms determines the likelihood of future communication breakdown (Pryce, 2006). The role of symptom
appraisal was examined through a large scale multivariable analysis in which dichotic listening, speech in noise and frequency resolution tests and illness perceptions were examined to identify which factors predicted help-seeking (Pryce et al 2010). This study revealed that the factor that predicted help-seeking was the belief in the consequences of having a hearing difficulty. In other words help-seeking behaviour was better predicted by a psychological factor in the impact of the symptoms than any audiological function or impairment (Pryce et al 2010).

Wider context – how common is a psychological component?

Medically unexplained illness in which psychological components co-exist with physical signs are common in healthcare (Pennebaker, 1982; Salmon, 2000). A review of neurological symptoms found that 26% were medically unexplained (Perkin 1989).

This is not an ‘either/or’ situation. There are frequent examples in the literature where health conditions have a biological underpinning which is exacerbated by emotional response. In particular anxiety has a role in starting and maintaining symptoms. This has been identified in abdominal pain, headaches, back ache and benign palpitations (Mayou, 1992). In otolaryngology such amplification occurs in vertigo (Hallam and Stephens, 1985), globus pharyngis (Dreary et al, 1989) and tinnitus (McKenna et al, 1991).

In a small study detailing the patient perception of the way hearing difficulties in King-Kopetzky syndrome arose emotional distress was highlighted as a factor in both the start and maintenance of symptoms. For some this was a clear incidence of trauma preceding the symptoms i.e. a car accident, divorce, death of a loved one or collapse of a business (Pryce, 2003). Study participants revealed that such traumas were part of their own personal history and interpretation of their symptoms. This study also highlighted that the anxiety experienced during communication would predispose the individual to further difficulty in communication. What is more this ‘vicious circle’ could be reversed when the communication partner was deemed to be understanding of the communication difficulties, the difficulties themselves reduced (Pryce, 2003; Pryce 2006).

The presence of psychological components in the sensitivity to underlying auditory processing disorders mean that for the patient their day to day experience of hearing difficulties varies according to social and psychological factors. Hearing and communication are interactive processes occurring in an ecological context influenced by intra personal, interpersonal social and cultural factors (Borg and Stephens, 2003).

So what helps?

Coping in King-Kopetzky syndrome was examined in detailed qualitative work. This study explored the individual cognitive and behavioural attempts to manage stress cause by hearing symptoms (Pryce, 2006). The process of coping relied on conceptualising the
symptoms and adopting strategies to manage day to day communication. The strategies identified were:

- Concentrating to piece together the communicative message
- Bluffing
- Avoiding communication
- Asking for repetition
- Lip-reading
- Directing speakers to adjust their behaviour
- Changing the environment
- Disclosing their difficulty
- Humour

The strategies are similar to those adopted by people with other forms of hearing loss. What is interesting here is that the clinical encounter could prompt the use of strategies and reduce stress when employing strategies. This was in part because the clinical encounter assisted validation of the symptoms (Pryce & Wainwright, 2008). The characteristics of clinical encounters determined outcomes in either increasing or reducing anxiety about symptoms. In particular encounters that were deemed to be dismissive were unhelpful and raised anxiety. This was typically the case when clinicians sought to provide brief reassurances that test results were normal.

‘they say your hearing is normal and they don’t consider there’s a problem. They’re not interested.’ (participant reported in Pryce and Wainwright, 2008)

One of the most helpful things to emerge from the British Society of Audiology and American Academy of Audiology has been the recognition of Auditory Processing Disorder as a worthwhile and legitimate presentation along with some guidance as to clinical action (BSA, 2011; AAA, 2010). It is hoped that such guidance will encourage and increase helpful and purposeful encounters for patients. However the audiologist should remain vigilant about the potential for testing to cause anxiety and to use test results to tailor further interventions to the individual. The most helpful part of the clinical encounter that patients reported was the sense of being understood and being given an explanation for the difficulties in hearing (Pryce & Wainwright, 2008). Participants reported this assisted both emotion focussed coping and practical uptake of strategies:

‘I realized there were other people that were the same and that actually it wasn’t a really negative thing about me. It was just me....I’ve got more courage now to say “I didn’t hear you because” rather than saying nothing before’ (Participant T3 reported in Pryce & Wainwright 2008)

So what sort of assessment?
The BSA guidance for APD practice (BSA, 2011) proposes a full case history be taken, supplemented with questionnaire measures. One of the challenges is that the questionnaire measures are developed for use with children and do not easily apply to adults. However, a full case history including information about the context in which difficulties occur can be very helpful. It is also useful to directly explore issues around stress in listening (Saunders and Haggard, 1989; Pryce, 2006). The AAA guidance for (C)APD suggests including an evaluation of:

- auditory and/or communication difficulties experienced by the individual
- family history of hearing loss and/or central auditory processing deficits
- medical history, including birth, otologic and neurologic history, general health history, and medications
- speech and language development and behaviors
- educational history and/or work history
- existence of any known comorbid conditions, including cognitive, intellectual, and/or medical disorders
- social development
- linguistic and cultural background
- prior and/or current therapy for any cognitive, linguistic, or sensory disorder or disability. (AAA, 2010)

The guidelines propose that further testing should be offered to those with a history of significant history of otitis media, neurological disease or disorder. Patients presenting with seizure disorders and hyperbilirubinemia should also be considered for testing (AAA, 2010).

Test packages such as the SCAN packages (Keith, 2000; Dawes and Bishop, 2007) are freely available to audiologists. These tests assess ability to detect speech in noise and attend to information for each ear individually (Pryce et al, 2010). However the reliability of tests is still debated as they are subject to learning effects (Domitz & Schow, 2000); and cannot discriminate between pre-cognitive and linguistic factors (Pryce et al, 2010; Moore et al, 2013).

These tests require a clear sense of purpose when applied in the clinic. There application should be in an ethical context where information will be used to benefit an individual in a way that outweighs the associated risks of enhancing anxiety about performance and function further. One way to consider this is to focus efforts on validating and supporting patients to make their choices about hearing management. The act of validation and labelling the condition enhance coping by supporting illness coherence (Pryce, 2006; Pryce...
2003; Pryce et al, 2010). It also has potential to inform both audiologist and patient and facilitate better informed choices about care and management.

Treatment

Borg and Stephens conceptualise the experience of King-Kopetzky syndrome as a disruption between an individual and their environment (Borg and Stephens, 2003). Therefore it is important to examine how strategies can reduce this disruption. Patient behavioural change is required for most rehabilitation in hearing loss and King-Kopetzky syndrome is no different. In this case examining the environment and adapting it where possible is particularly important to improve speech in noise perception. Relatively simple strategies such as turning off background noise where possible can be extremely helpful (Zhao et al, 2008). There is limited evidence that auditory training may have an effect on day to day function (Brouns et al, 2010).

To examine complex interventions such as therapeutic interventions for King-Kopetzky syndrome fully the UK Medical Research Council suggests that we develop clear theoretical understanding of the way interventions might work. In this case the theoretical work has been conducted through qualitative approaches, based on grounded theory. Through this approach there are clear descriptions of how clinical interventions do and do not assist coping as the patient perceives it. Above all the most valuable intervention reported in interview accounts about what aids coping is the act of being listened to and being taken seriously (Pryce & Wainwright 2008; Zhao et al, 2008). This is helpful in developing coping by supporting use of strategies to manage difficult situations and to build confidence in encouraging speakers to slow down, move away from background noise, switch room etc (Pryce, 2006; Pryce and Wainwright 2008). This apparently simple use of counselling and problem solving therapy is effective in other medically unexplained conditions such as fibromyalgia, chronic fatigue syndrome etc (Hassett et al, 2009; ) so it is perhaps not so surprising this works in this instance.

There has been suggestion that auditory training packages might help individuals to function better by re training their discrimination of sound. The theoretical basis for this is not completely clear and findings are mixed (Moore and Rosen, 2013). The practice of repeatedly listening to sound contrasts is believed to drive the development of more efficient neuronal pathways, thereby improving auditory processing (Brouns et al, 2010). However there is a lack of neurophysiological studies to examine this action (Brouns et al, 2010). Moreover there is a dearth of studies demonstrating more than a modest effect size for the benefits of training on task (Brouns et al 2010).

The potential for communication training programmes, which include an educational and behavioural component such as Active Communication Education also deserve further investigation with this population. These programmes work on problem solving and application of strategies to manage difficult situations and have demonstrated effective
improvement in communication function, hearing handicap and well being in adults with hearing loss (Hickson et al, 2007).

It is likely that such approaches will trigger coping adaptations which in turn can affect hearing performance. A study into the effects of mindfulness therapy identified improvements in coping and even reports of improvements in hearing performance suggesting that coping responses can moderate function (Sadlier & Stephens, 2009).

As with other hearing conditions, it makes sense to consider how clinicians can enact shared decision making with this population, especially where treatment is dependent on coping skills and behavioural adaptations. Presenting patients with clear choices in interventions and outlining the advantages and disadvantages of different course of action will be important here (Pryce & Hall, 2014).

**Conclusions**

King-Kopetzky syndrome represents a fascinating experience of mis-hearing based on multiple factors, which vary in emphasis from individual to individual. The role of Auditory Processing Disorder as a sub component to the presentation is interesting and more that is learned about this component will help with better patient counselling and guidance. Evidence from qualitative investigations suggests that the broader, non-biological factors that lead to clinical help-seeking should be considered alongside audiological factors in clinical history taking, counselling and problem solving. This area of work highlights the complexity of the audiological role in addressing psychological and social needs of patients alongside the biological needs. It also highlights the needs for audiologists to engage in counselling and communication training approaches beyond the traditional technical investigations.

It reminds us that healthcare involves the toleration of uncertainties about causation and remediation. Our role is to support the whole person, including providing reassurance about the consequences of not hearing, arming them with practical strategies to repair conversations, informing them about their function and potential and, above all, listening and bearing witness to their experience.

**Learning objective:**

To define King-Kopetzky Syndrome and to consider interventions that can help.

**Question 1:**

People might be labelled as having King-Kopetzky syndrome if
a. They present for help with hearing difficulties when their audiometric thresholds are within normal limits.
b. They complain about noise levels.
c. They have age related hearing loss.
d. They need Therapeutic care.

Question 2:

There is evidence that testing can be helpful because:

a. It helps the clinician to form a plan.
b. It identifies whether there is impairment in auditory function.
c. It provides information for audiologist and patient to share.
d. It proves the patient is right.

Question 3:

What should be included in patient counselling?

a. Advising patients that they may have a disorder
b. Listening and paying attention to the patient’s story
c. Full testing
d. Checking for neurological conditions

Question 4:

Which of these was not a sub group identified by Zhao and Stephens:

a. Purely psychological based problem
b. Unknown problem
c. Cochlear pathology
d. Low mood

Question 5:

The ecological model suggested by Borg and Stephens (2003) suggests:

a. That King-Kopetzky syndrome represents a disruption to individual homeostasis.
b. That King-Kopetzky Syndrome is a form of neuroticism.
c. That it is not possible to have an auditory processing disorder and King-Kopetzky Syndrome.
d. That King-Kopetzky syndrome cannot be treated.

Answers: 1 (a); 2 (c); 3(b);4(d); 5(a)

References


