

Auditory dysfunction and its remediation in individuals with spinocerebellar ataxia



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Spinocerebellar ataxias

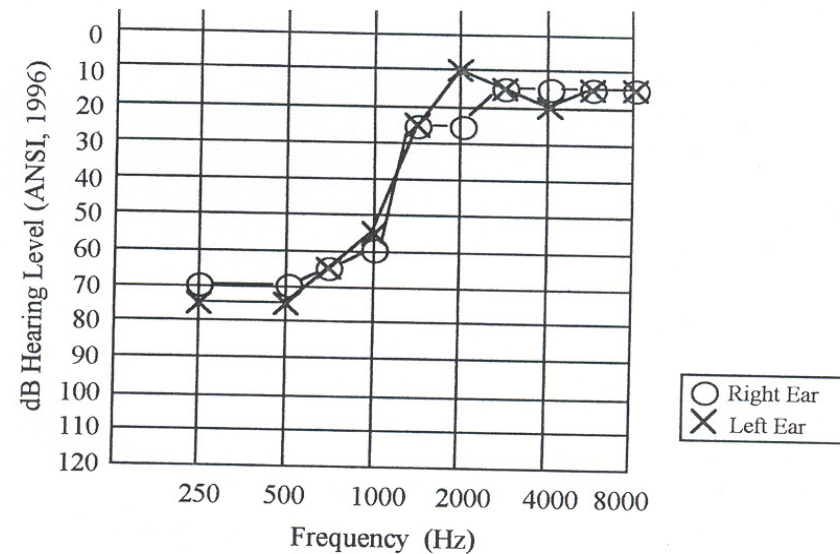
- The spinocerebellar ataxias (SCAs) are a heterogeneous group of neurological disorders caused by varying degrees of degeneration of the cerebellum, brainstem neurons and spinocerebellar tracts.
- We investigated the auditory consequences of SCA types 1, 2 & 6 and explored the capacity of FM systems (remote-microphone listening devices) to alleviate everyday listening/communication difficulties in affected individuals.

Auditory function in ataxia


- Sound detection usually normal (Harding 1981; Durr et al 1996; Rance et al 2008)
- A significant number of patients show evidence of disordered neural conduction in the central auditory pathways
- Electrophysiological testing in such cases typically reveals absent or abnormal ABR (Satya-Murti et al., 1980; Jabbari et al., 1983; Rance et al., 2008) in the presence of normal otoacoustic emissions and cochlear microphonics (Starr et al., 1996; Lopez-Diaz-de-Leon et al., 2003; Rance et al., 2008)
- This result pattern compatible with **Auditory Neuropathy Spectrum Disorder**

Sound detection

- Normal or near-normal audiogram
- In some cases 'untypical' low-frequency hearing loss



Auditory perception

- Impaired speech perception
 - Difficulties with speech perception in noise
 - The ability to perceive rapid changes in auditory signals over time (**temporal resolution**) impaired
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Auditory assessment in ataxia

- ‘Routine’ investigation:
 - **Pure-tone audiogram**

- *Lege artis* investigation:
 - **Speech test**
 - **Speech in noise**
 - **ABR**

Auditory assessment in ataxia

- Psychophysical investigation:
 - **Gap detection**
 - **Amplitude modulation detection**
 - **Frequency discrimination**

Auditory management

- Conventional management approach of fitting of **hearing aids** may be **inappropriate as** amplification is designed to make sounds louder rather than clearer

Auditory management

- **Hearing therapy**
- **FM-systems**
- **Cochlear implantation???**

Auditory management


Future:

- **advances in speech-processing** that make temporal cues more salient

FM system

- FM Systems are wireless assistive hearing devices that enhance the use of hearing aid(s), cochlear implants and also assist people who are hard of hearing but do not wear hearing aids, in particular over distance and in noisy environments.
- They enable sound to be picked up closer to a speaker, sound source or connected directly to the sound source and transmitted to the individual providing greater clarity of speech/sound and a reduction in background noise.

FM system

- FM System is the generic name or term that has been given to Radio Aids. FM is the abbreviation for Frequency Modulation.
 - Frequency Modulation is a wireless transmission method used to transmit the sound.
 - **The main advantage for listeners is that FM transmission is resilient to noise and interference helping to preserve the quality and clarity of the sound transmitted.** Some companies are also using digital transmission.
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FM system

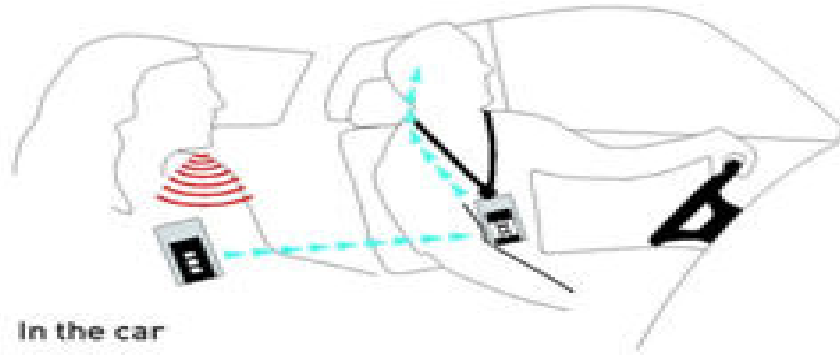


a. iSense micro receiver

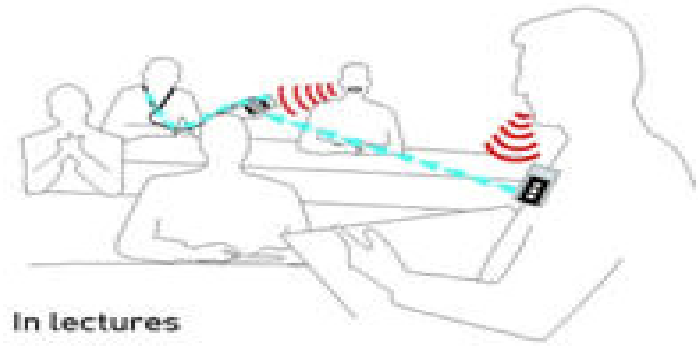


b. Roger inspiro transmitter with iLapel microphone

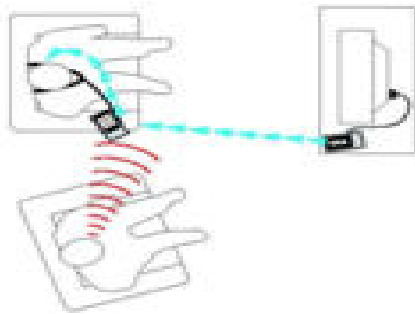
▶ Sound transmission))) Speech



In the car



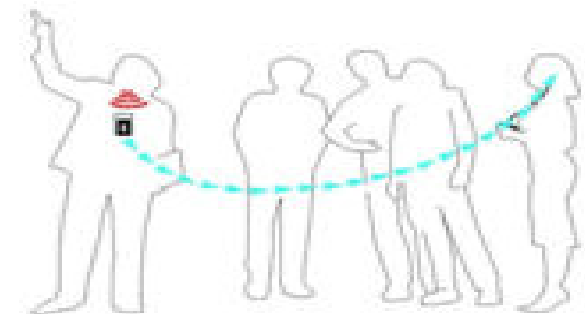
In lectures



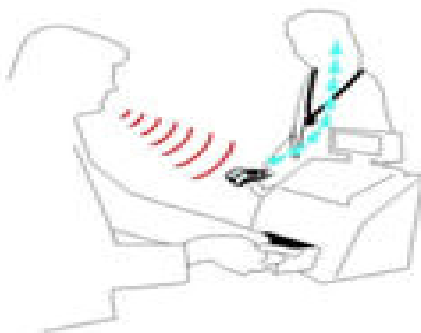
Watching TV



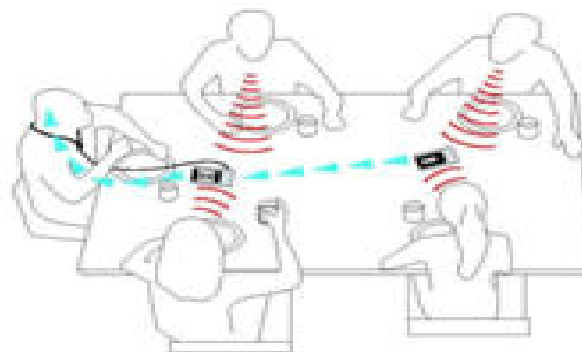
At home



On holiday



Shopping




At the dinner table



Walking


Methods

- 14 adults with SCA (Type 1: N=7; Type 2: N=2; Type 6: N=5) underwent a comprehensive evaluation of peripheral hearing mechanisms (sound detection, cochlear mechanics), auditory neural activity (auditory brainstem response) and functional hearing (monosyllabic speech perception, self-reported communication disability).
 - Findings were compared with data obtained from 130 healthy controls.
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Methods

- Each SCA participant was subsequently fit with an ISense FM listening device and underwent a 6 week take-home trial.

Results

- Sound detection thresholds were within age-corrected norms for 13/14 of the SCA participants.
 - None-the-less, the majority of participants (in each SCA category) showed evidence of severe auditory dysfunction. Twelve individuals presented with abnormal VIIIth nerve/brainstem activity, with absent ABRs, delayed neural conduction or abnormal stimulus-rate sensitivity.
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Results

- Speech discrimination was affected, particularly for signals presented in background noise (0 dBSNR) with 12/14 participants performing at levels below the normal (95% confidence) range.
 - Everyday listening ability was also impaired in most cases.
 - Ten of 14 reported extreme difficulty understanding conversation in background noise and 9/14 felt that their hearing challenges adversely affected communication ability.
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Results

- The use of an FM-listening system did, however, alleviate these difficulties. For 13/14 individuals speech perception scores improved to within the normal range when wearing the device and 11/14 reported **significant improvement** in everyday listening and communication over the course of the 6-week trial.

Conclusions

- Auditory dysfunction is common in SCA1, SCA2 and SCA6, but remote-microphone listening devices appear to be a viable intervention option for many patients.