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Audiological findings in a case of von Recklinghausen's Multiple Neurofibromatosis.

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Abstract

Bilateral acoustic tumors are reported to be the most common intracranial tumor in adults with von Recklinghauseris disease. The audiological lest findings are reported to be confounded in the presence of bilateral acoustic neurenomas. There)[r]ore, the audiological findings has tobe interpreted vvilh great care. The report of such a confounded audiological test finding in a case of von Recklinghausen' s disease is discussed in this paper.

Introduction

Lindsay et al, (1986) described von Recklinghausen's disease as an inherited condition by autosomal dominant gene transmission, with a positive history of incomplete penetrance or mutation in about 50% of the cases, it is observed that these patients generally are of younger age group in their early twenties. The rate of growth of tumor in this disease is said to be slow. The incidence of von Recklinghausen's disease is 1 in 2000.

Von Recklinghausen's disease is commonly characterized by cutaneous ncurofibromata, cafe - au - lait spots, skelctal abnormality, mental retardation (Gilroy and Meyer, 1979, Walton, 1985, Lindsay et al, 1986, Ramsden, 1987). In this disease, bilateral acoustic tumors arc the most common inlracranial tumor in adults, while astocytomas are the most common in children (Baldwin & Le Master, 1989).

It is also known thai the audiological lest finding gel confounded in the presence of bilateral acoustic neuromas. Therefore, in such cases great care has to be exercised in interpreting the audiological test results. The following is the report of such a confounded audiological test finding

in a case of Von Recklinghausen's disease.

Case **Report**

The subject SGR, male aged 25 years, was specifically referred to the institute by Tata memorial hospital, Bombay, fordelailed audiological investigation, with a complaint of positional vertigo associated with sudden movements of the head. The physical appearance of the patient revealed brown black spots on the face and trunk and over growth of skin. The case did not complain of hearing loss and there was no family history of hearing loss or similar skin manifestations.

The reports of the investigation carried out at the referral hospital suggested negative findings on caloric tests and Rhombcrg's test, normal functioning of **III,** V, VI cranial nerves,and CAT scan showed normal IV ventricle and posterior fossa with widening of Internal auditory meatus(IAM) in both sides by 8 mm and 9 mm in the right and left ears respectively. There was no evidence of bony erosion of I AM.

Investigations

From the complaint and the available investigation reports the client under discussion was suspected to have retrocochlcar pathology with central auditory deficit (CAD) He was examined by ENT specialist, neurologist, speech and language pathologist, clinical psychological and audiologist for the CAD.

The audiological test battery included 1) pure lone audiometry, speech audiometry and other supra threshold tests carried out using the Madsen OB 822 dual channel diagnostic audiometer. 2) Impedance Audiometry carried out with Madsen Z0174 immittance bridge and 3) Auditory Brain stem response carried out with Nicolet Compact IV electrodiagnostic system. All the audiological lest were carried out using standard test procedures and in an acoustically sound treated lest room.

ABR recordings were done by placing silver electrodes at four sites i.e. a common electrode on the forehead, a negative electrode on mastoid bone of each side and a positive electrode at the vertex. Electrical impedance was less than 5 kilo ohms.

Initially, the ABR recordings were obtained by monaural presentation of 2000 clicks of rarefaction polarity with repetition rate of 11.4 clicks / sec through TDH39 earphones at 100 dB nHL. Adequate masking in the non test car was provided while testing each ear. The electrical activity generated as a result of acoustic stimulation were averaged and filtered with a filter setting of 150 Hz to 3000 Hz.

The obtained ABR waves recorded at 100 dB nHL were analyzed to study :

1. Absolute latency of peak I, III & V.
2. Inter peak latency of peaks I - III, III - V, I- V.

And the values were compared with clinical nonns.

Next the ABR waves were recorded at 100 dB nHL with increased repetition rate (57 clicks / sec) and the absolute latency and the morphology of the V peak was compared with the earlier wave form recorded at a click rate of 11.4 click**s / sec.**

Audiological findings

As can be observed from the Fig. 1 pure tone audiogram suggested normal hearing upto 2 KHz and mild to moderate hearing loss at 4 KHz and 8 KHz in the right ear. An air-bone conduction gap (AB gap) of 30 dB was also observed in the right ear.

However, in the left ear mild hearing loss upto 4 KHz and moderate hearing loss could be noticed at 8 KHz.

On speech audiometry, a speech reception threshold (SRT) of 25 dB in right and 20 dB in left ears were recorded. The speech siscrimination scores (SDS) were 95% in the right ear and 100% inthe left ear. Performance - intensity function with phonetically balanced words (PI - PB function) indicated no roll - over effect in any of the cars. The results of pure tone adaptation tests i.e. Tone decay test (TDT) using Olsen & Noffsinger (1974) method & supra threshold adaptation test (STAT) as given by Jerger & Jerger (1975) were found to be negative.

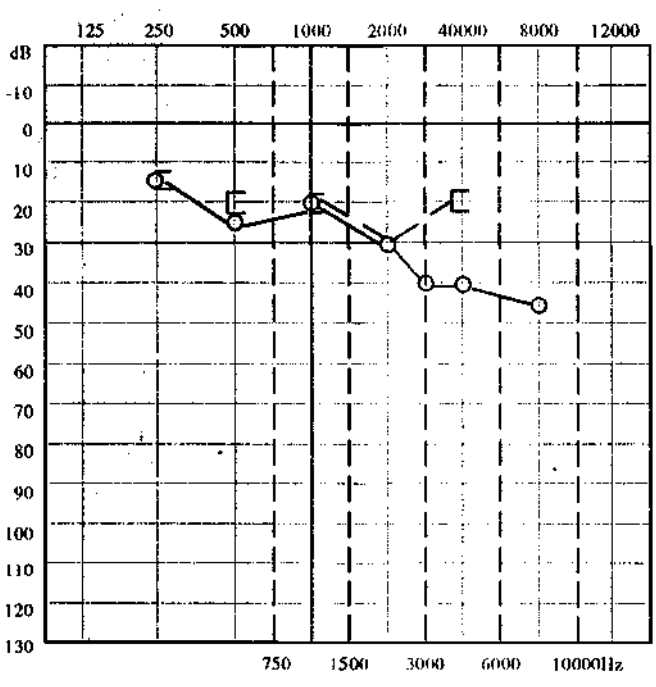


Fig. 1 Audiogram of right ear

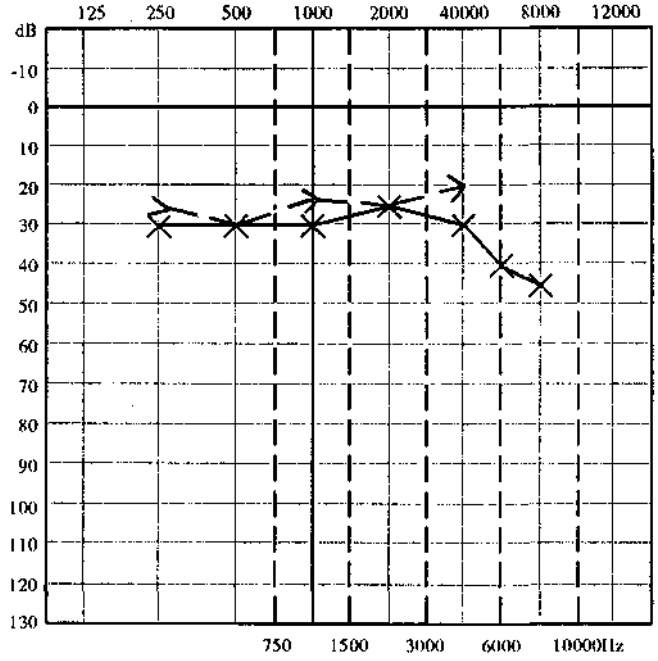


Fig. 2. Audiogram of Left Ear

	SRT	SDS	PI -PB	TDT	STAT
Right ear	25 dB	95%	No Roll over	-Ve	-Vc
Left ear	20 dB	100%	No Roll over	-Vc	-Ve

Table 1:

The impedance audiomelric results indicated A type tympanogram in both the ears, with the static compliance of 0.46 cc in the right car and 0.38 cc in the left ear.

Acoustic reflexes were absent in both ipsilateral and contralatcral recording forboth the ears for pure tone stimulation at 0.5 KHz, 1 KHz, 2 KHz, 4 KHz and wide band noise at the maximum permissible intensity levels.

In ABR the absolute latency of peaks III & V were significantly delayed in the right ear and absolute latencies of peaks III & V were marginally delayed in the left ear at lOOdBnHL. Inter peak latency (IPL) value of III - V and I - V peaks were significantly prolonged in right ear. Similarly in the left ear III - V and I - V IPLs were marginally prolonged and I - III IPL were within normal limits in both the ears.

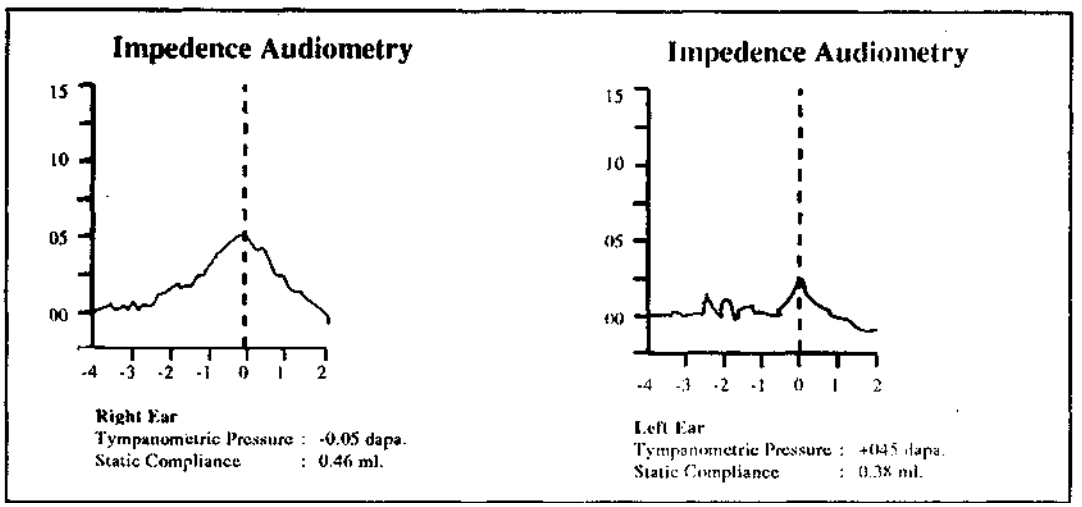
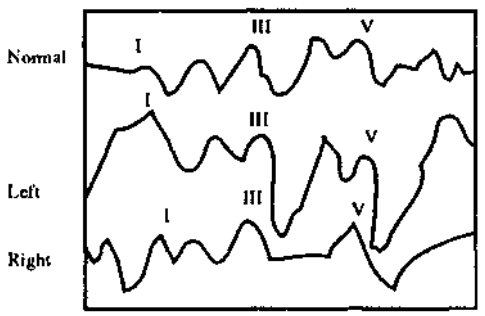


Fig. 3. Tynipanogram of right & left ear



	1	HI	V	I-III	III-V	IV
Normal	1.48	347	530	1.99	1.83	3 80
Ri.	1.58	3.62	5.70	2.04	2.08	412
Subject It.	152	354	560	202	2.06	4.08

Fig. 4. Absolute & Interpeak Latency of **ABR Peaks**

WiLh the increase in Lhe rate of stimulus presentation i.e. from 11.4 clicks/ sees to 57 clicks / sees at 100 dBnHL, the latency of peak V was increased by 0.44 msec in the left car. However, in the right car the wave form was completely degraded

Discussion

The results of the pure tone audiometry, speech audiometry, tympanomclry and the static compliance measurements with negative findings on pure tone adaptation tests (T.D.T. and S.T.A.T.) and the absence of positive roll over, suggested the presence of mild high frequency cochlear hearing loss in left car & moderate high frequency cochlear hearing loss in the right car.

However, the findings of acoustic reflex measurements indicated the absence of (he stapedial reflex in both the cars for pure lone stimulation at all the frequencies and wide band noise at maximum intensity levels. This indicated the possible rclrocochlcar involvement in both the ears, as otherwise, with a mild cochlear hearing loss the stapedial acouslic reflex should

have been present at 70 to 90 dB SL.

Such findings arc not uncommon and is reported to have been seen in confirmed cases of acoustic neuromas by several investigators (Jerger and Jcrger, 1974 b, Johnson, 1977).

Likewise, the presence of high tone loss has also been reported to be seen in many cases of acoustic neuromas by several investigators (Morrison, 1975., Johnson, 1977). Generally it is also common to expect the presence of marked adaptation on pure tone adaptation tests in cases with eight nerve tumor. However, Gibson, (1981) has reported that the typical abnormal adaptation occured in only half of the cases with small eighth nerve tumors. Thus, the negative results on the adaptation tests seen in the client under discussion, do not rule out the possibilities of the presence of small eight nerve tumors.

Further in the client under discussion, the absolute latency of ABR peaks III and V were significantly prolonged in right ear and in the left ear the absolute latency of peaks III and V were marginally prolonged. However the absolute latency of peak I was within normal limits in both the ears. This findings arc in line with similar findings reported by Eggermontet. al., (1980), Hyde and Blair, (1981)., Moller and Moller, (1985) in cases with confirmed acoustic tumors.

The prolongation of the absolute latency of peaks III and V in the client under discussion was further supported by the findings of Jacobson and Morehouse, (1985) who has reported that the peaks III and V latencies were prolonged in a case of von Recklinghausen's neurofibromatosis.

It is also suggested that to compensate for the effect of peripheral hearing loss on ABR results a correction of subtracting 0.2 msec, from the absolute latency values for every 30 dB threshold difference between 1000 Hz and 4000 Hz (Jerger and Mauldin, 1978) or a criteria of adding 0.1 msec, to peak V latency for every 10 dB of hearing loss greater than 50 dB at 4000 Hz (Sellers and Brackmann, 1979) may be applied. However, these corrections did not become applicable as hearing loss at 4000 Hz was less than 50 dB in both the ears and further the difference between the threshold at 1000 Hz and 4000 Hz were less than 30 dB in the client under discussion. Thus, it could be stated that the delay in the absolute latencies of ABR peaks in the client under discussion is more likely to be a retrocochlear manifestation rather than due to the presence of the high frequency peripheral hearing loss.

The presence of bilateral acoustic tumors is further supported by the significant prolongation of the inter peak latency (IPL) values of peaks III - V and I - V in right car and marginal prolongation of the IPL values of peaks III - V and I - V in the left ear in the client under discussion.

Such significant prolongation of IPL values in confirmed cases of eight nerve tumor have been reported by several investigators (Glasscock et. at., 1979.,Eggermont

et. al., 1980, Musieket. al., 1980, Harris and Almquist, 1981., Musiek and Gollegly, 1985). Specially, Jacobson and Morchouse, (1985) have reported prolongation of I - V IPL in a case of von Recklinghauscn's neurofibromatosis.

To further confirm the presence of bilateral acoustic tumor the ABR wave forms were recorded at higher repetition rate (57 clicks / sec) as it is suggested by Musiek and Gollegly, (1985) that the use of high repetition .rate stresses the auditory system and therefore in some cases can uncover abnormality that may not be identified otherwise.

There have been several case reports of eighth nerve or brainstem lesion that have shown significant wave V latency shifts or degradation of wave V morphology at high repetition rates (Weber and Fugikawa, 1977., Yogi and Kaga, 1979).

With increased rate of stimulation, in the client under discussion the wave form morphology was completely degraded in the right car and in the left ear the absolute latency of peak V was shifted by 0.44 msec. But this shift in the absolute latency of peak V was found to be not significant by both Hecox, (1980) and Musiek & Gollegly, (1985)'s criteria.

Conclusion

In the client under discussion the results of the pure tone audiometry, speech audiometry, tympanometry and the static compliance measurements indicated the presence of bilateral high frequency cochlear hearing loss. However, on the other hand the results of the stapedial acoustic rcllex and the Auditory Brainstem Evoked Response tests indicated bilateral rctrocochlcar involvement which was supported by the similar findings as found in the literature. The audiological findings in the client under discussion strongly indicated the possibility of bilateral acoustic neuroma which was not definitely indicated in any of the medical investigations carried out on the client.

Thus, the audiological test battery could be used to uncover the subtle auditory manifestation of small acoustic neuromas in the cases of von Recklinghausen's neurofibromatosis.

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References

Baldwin, R.L. & Le Master, K. Ncurofibromatosis - 2 and Bilateral acoustic neuromas : Distinctions from neurofibromatosis - 1 (von Recklinghausen's disease). 1989. *The American Journal of Otology*, Vol. 10, No. 6, pp439 -442.

Hggermont, J., Don, M & Brackmann, D trictrocochleography and auditory brainstem electric responses in patients with pontine angle tumors. (1980). *Annals of Otology, Rhinology and*

Laryngology, Vol. 89, supplement 75.

Gibson, W.P.R. Diagnosis of acoustic neuroma. Chapter in audiology and audiological medicine (Ed.) Beagley Vol. 1. Oxford University Press, London. (1981).

Gilroy, J and Meyer, J.S. : Medical neurology : 3rd Edition, Macmillan PublishingCo. Inc, N.Y. (1979).

Glasscock, M., Jackson, C, Josey, A., Dickins, J. & Wiet, R

Brainstem evoked response in clinical practice. *Laryngoscope,* (1979). Vol. 89, pp. 1021 - 1034.

Harris, J and Almquist, B. ABR in operatively verified cercbcllo - pontine angle tumors. *Scandinavian Audiology,* (1981). Supplement, 13 pp. 113 - 114.

Hecox, K. (1980) ABR and brainstem involvement. Presented at a symposium on Auditory evoked response in Otology and Audiology, Cambridge, MA, August 8-9.

Hyde, M.L & Blair, R.L. The Auditory brainslcm responses in neurotology perspectives and problems. *Journal of Otolaryngology,* (1981). Vol. 10, pp. 117- 125.

Jacobson, J.T & Morehouse, C.R. Special case studies. Chapter in the Auditory Brainstem response. (Ed.), Jacobson J.T. Taylor and Francis Ltd., London. (1985).

Jerger, J & Jerger, S. Auditory findings in brainstem disorders. *Archives ofOtolaryngology(\974*b). Vol. 99, pp. 342 - 350.

Jerger, J & Jerger, S. Simplified tone decay lest. *Archives of Ololaryngology.* (1975), Vol. 101, pp. 403 - 407.

Jerger, J and Mauldin, L. Prediction of scnsorincural hearing level from the brainstem evoked response. *Archives of Otolaryngology,* (1978). Vol. 103, pp. 181 - 187.

Johnson, E.W. Auditory lest results in 500 cases of acoustic neuroma. *Archives of Otolaryngology,* (1977). Vol. 103, pp. 152 - 158.

Lindsay, K.W. Bone, I & Callander, R. Neurology and ncurosurgery illustrated, Churchill Livingstone Inc, N.Y. (1986).

Moller, M.B. & Moller, A.R. Auditory Brainstem - Evoked response in diagnosis of eighth nerve and brainstem lesions. Chapter in Assessment of central auditory dysfunction. (Ed.), Pinherio and Musiek. Williams and Wilkins, Baltimore, (1985).

Morrison, A.W. Management of sensorineural deafness. London & Boston Butter worths (1975).

Musiek, F.E.&Gollegly, K.M. ABR in Eighth Nerve and Low Brainstem Lesions. Chapter in the Audi0 tory Brainstem response. (Ed.), Jacobson J.T. Tay0 lor and Francis Ltd., London. (1985).

Musiek, F., Sachs, E., Geurkink, N., & Weider. Auditory brainslem response and eighth nerve lesions : A review and presentation of cases. *Ear and Hearing,* (1980). Vol. 1, pp. 279 - 301.

Olson, W.O. & Noffsinger, D. Comparison of one new and three old tests of auditory adaptation. *Archives of Otolaryngology,* (1974). Vol. 99, pp. 94 - 99.

Ramsden, R.T. Acoustic Tumors, Chapter 21 in Scott Brown's ptolaryngology V Edition, Otology, (Ed.), Booth, J.B. Butterworths International Editions, London. (1987).

Rose, D. Brainstem auditory responses 'letters', Mayo clinic Proceedings, (1978). Vol. 53, pp. 132.

Rosenhamer, H. The auditory evoked brainstem electric response in cochlear hearing loss. *Scandinavian Audiology,* (1981). Supplement 13, pp. 83 - 93.

Selters, W.A. and Brackmann, D.E. Brainstem electric response audiomctry acoustic tumor detection. (Ed.), W. House & C. Luetje Acoustic tumors (Vol. 1). Baltimore : University Park Press (1979).

Walton, J. Brain's Diseases of the nervous system. IX Edition, Oxford University Press, N.Y. (1985).

Weber, B and Fuijikawa, S. Brainstem evoked response (BER) audiometry at various stimulus presentation rates. *Journal of the American A udiology Society.* (1977). Vol. 3, pp. 59 - 62.

Yagi, T and Kaga, K. The effect of the click repetition rate on the latency of the auditory evoked brainstem response and its clinical use for a neurological diagnosis. *Archives oj'Olorhinolaryngology.* (1979). Vol. 222, **pp. 91-97.**