question "Did any of your doctors at any stage of your illness consider the diagnosis of MS ?". In all patients, antiphospholipid antibody (aPL) status was recorded.
Out of 90 patients with positive aPL tests, 26 (28.8%) recorded a positive response to the question, as compared with 10 (8.4%) of the aPL negative patients [RR: 5.5 (95 CI: 2.8-11) $p < 0.0001$].
The aims of this audit study were two-fold: firstly, to obtain an estimate of the frequency with which patients with SLE and Hughes syndrome had had a differential diagnosis of MS considered. Secondly, to determine whether the MS differential diagnosis was made more frequently in patients with aPL.
This study acknowledges the pitfalls in patient questionnaire surveys, in patient recollection of previous consultation, and in the relevance of aPL. Despite these caveats, we believe that the result of this survey serves to emphasise the importance of the APS as a neurological entity, and as a differential diagnosis from MS.

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References

1. NAVARRETE MG, BREY RL, LEVINE SR: Cerebral disease in the antiphospholipid syndrome. *In* KHAMASHTA MA (Ed.): *Hughes Syndrome, Anti - phospholipid Syndrome*. London, Springer 2000; pp: 43-58.
2. CUADRADO MJ, KHAMASHTA MA, BALLESTEROS A, GODFREY T, SIMON MJ, HUGHES GRV: Can neurologic manifestations of Hughes (antiphospholipid) syndrome be distinguished from Multiple Sclerosis? *Medicine (Baltimore)* 2000; 79: 57-68.

Sensorineural hearing impairment in systemic lupus erythematosus: Sudden or progressive?

Sirs,
Sensorineural hearing loss (SNHL) in systemic lupus erythematosus (SLE) has been described by several authors, though its pathogenesis is still not clear. Sudden onset SNHL is the most frequent SLE related hearing impairment and often represents the consequence of an anticardiolipin antibody (ACL) syndrome involving the inner ear (1).
Despite the prevalence (21.5%) (2) of progressive SNHLin SLE patients at pure tone audiometry, no one ever focused on the cochlear function by using the most recent and sensible techniques, such as transiently evoked otoacoustic emissions (TEOAEs), enabling to evaluate cochlear outer hair cells' function by recording their response to transient acoustic stimulation thorough a probe placed in the external auditory canal (3).
We investigated 30 (27 F; 3 M) SLE consecutive patients (group 1), mean age 41.2 years (range 27-66). A complete rheumatological and audiological assessment was carried out. Mean Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) (4) was 9.12 (range 2-19). Anticardiolipin IgG and IgM antibody (aCL) positivity was found in 6 (20%) subjects at elisa. SLE patients were treated with hydroxychloroquine, non-steroidal anti-inflammatory drugs (NSAIDs), metotrexate (MTX), dehydroepiandrosterone (DHEA), and cyclophosphamide. Conductive hearing impairment was excluded by pure tone audiometry and tympanometry. $TEOAE_s$ were recorded in all 60 ears with ILO88 OAE Analyzer (Otodynamics), and results were compared with those obtained in 30 healthy age-matched controls (group 2). No patient had a history of sudden hearing loss.
TEOAEs average reproducibility was 37.08% (±18.75%) in group 1, and 44.78% (±20.89) in group 2 ($p = 0.031$); the average amplitude was 4.58 (±2.93) decibel Sound Pressure Level (dB SPL) in group 1 and 6.43 (±3.18) dB SPLin controls ($p < 0.001$). In SLE patients, a statistically significant inverse correlation ($r = -0.289$, with a corresponding $p = 0.025$) between duration of disease and TEOAEs amplitudewasfound (Fig. 1). An inverse correlation, though not statistically significant, was found between TEOAEs amplitude ($r = -0.214$, $p = 0.099$) or reproducibility ($r = -0.683$, $p = 0.604$) and SLEDAI. No relation was noticed between audiological parameters and aCL positivity or patients'therapy.
Our data confirm a progressive cochlear impairment in SLE patients, though its pathogenesis is still unclear. Vasculitis processes, fibrosis, and new bone formation in the inner ear may be involved (5). A possible role of antibodies directed against inner ear antigenic epitopes and/or an involvement of aCLin inner ear immunocomplex-mediated vasculitis (6) has been proposed, though our data do not confirm it.
The inverse correlation between duration of disease and TEOAEs amplitude suggests that progressive inner ear impairment is common in SLE patients and represents the consequence of a chronic and progressive damage to the cochlea developing during the disease. This is partially confirmed by the relation, though not statistically significant, between TEOAEs parameters and SLEDAI. Additional prospective studies are needed to elucidate the pathogenetic mechanisms of SLE related cochlear impairment.

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References

1. GREEN L, MILLEREB: Sudden sensorineural hearing loss as a first manifestation of systemic lupus erythematosus: association with anticardiolipin antibodies. *Clin Rheumatol* 2001; 20: 220-2.
2. KASTANIOUDAKIS I, ZIAVRA N, VOULGARI PV, EXARCHAKOS G, SKEVAS A, DROSOS AA: Ear involvement in systemic lupus erythematosus patients: a comparative study. *J Laryngol Otol* 2002; 116: 103-7.
3. KEMP DT, RYAN S, BRAY P: A guide to the effective use of otoacoustic emissions. *Ear Hear* 1990; 11: 93-105.
4. BOMBARDIER C, GLADMAN DD, UROWITZ MB, CARON D, CHANG CH: Derivation of the SLEDAI. A disease activity index for lupus patients. The Committee on Prognosis Studies in SLE. *Arthritis Rheum* 1992; 35: 630-40.
5. YOON TH, PAPARELLA MM, SCHACHERN PA: Systemic vasculitis: a temporal bone histopathologic study. *Laryngoscope* 1989; 99: 600-9.
6. BERRETTINI S, RAVECCA F, BRUSCHINI L, URSINO F, SELLARI-FRANCESCHINI S: Progressive sensorineural hearing loss: immunologic etiology. *Acta Otorhinolaryngol Ital* 1998;18 (Suppl.59):33-41.

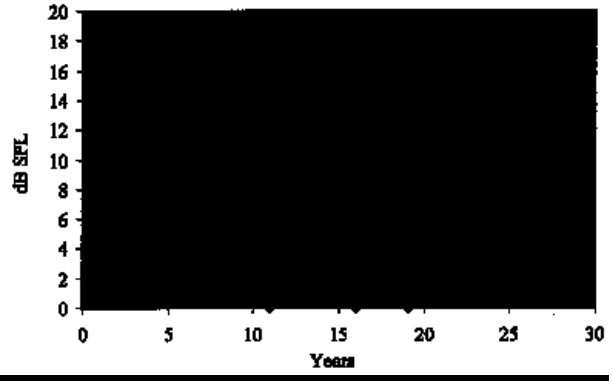

Fig. 1. Correlation between systemic lupus erythematosus duration (horizontal axis) and transiently evoked otoacoustic emission amplitude (vertical axis). The regression line approximates the relation between the variables; dB SPL = decibel Sound Pressure Level.