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Some Geographical Aspects of Neuro-otology with Particular Reference to the African*

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Summary. The African appears to show a different pattern of neuro-otological disorders than the European. This is probably due to exposure to a different set of geogens. However, bearing in mind the possibility that there may be differences in the basic hearing levels for high frequencies, there are the possibilities of genetic factors operating.

Poorer hearing levels in a Jamaican population were shown to be associated with the neurological and ophthalmological defects that characterize the ataxic type of Jamaican neuropathy, which is possibly identical with the ataxic neuromyelopathy that Money (1959) and Osuntokun (1968) have described in Nigeria. Analysis of the Jamaican audiograms showed a variety of patterns, including the type reported by Osuntokun for a case of ataxic neuromyelopathy in Nigeria.

Judging from the experience gained in the Jamaican studies, it would appear that any worthwhile population audiological study must now encompass neurological, ophthalmological and cardiological examinations.

Résumé. Les Africains semblent montrer un genre de désordres neuro-otologiques différent de celui des Européens. Ceci est sans doute du au fait que ces populations sont placées dans un ensemble différent de facteurs geographiques. Cependant, ne perdant pas de vue la possibilité que peut-être il y a des différences dans les niveaux d'audition de base, en cas de hautes fréquences, il est possible que des facteurs génétiques entrent en jeu.

Des niveaux d'audition amoindrie dans une portion de la population de la Jamaique, se révelerent être associés avec les troubles neurologiques et ophthalmologiques qui caractérisent le type ataxique de "Neuropathie Jamaicaine" qui est peut-être identique à la neuromyelopathie ataxique que Money (1959) et Osuntokun (1968) ont décrit an Nigeria. Une analyse des audiogrammes jamaicains montra une varieté de courbes, y compris celle du type cité par Osuntokun à propos d'un cas de neuromyelopathie ataxique du Nigeria.

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A en juger d'apres l'expérience recue à la suite des études jamaicaines, il apparaitrait que toute étude audiologique valable d'une population devrait comprendre des examens à la fois neurologiques, ophthalmologiques et cardiologiques.

In 1961, Ormerod undertook a preliminary survey of the problem of hearing loss in Africa. Based principally on his findings, Fig. 1 shows the geographical distribution of diseases which are of particular importance in the genesis of hearing loss in the indicated areas. To this map I have added noise-induced hearing loss and otosclerosis to the southern and northern extremities of the continent respectively. Occupational noise-induced hearing loss is related to the degree of industrialization and otosclerosis is a genetically determined conductive hearing loss which is much more prevalent in the white than the black races. Thus, for the moment, these two conditions do not present audiological problems in tropical Africa.

FIG. 1. Map of Africa showing regions where specified conditions are important factors in the occurrence of impairments of hearing in those localities. O.S., ostosclerosis; N.I.H.L., noise-induced hearing loss.

There are a number of scattered reports in the literature relating to hearing and its disorders in tropical Africa. Figure 2 shows the location of particular regions that have been subject to a particular study together with the names of the authors associated with the report. Ormerod emphasized the importance of meningitis as a cause of hearing loss in East Africa, particularly meningococcal in Kenya and pneumococcal in Tanzania, where also,

in the south, an endemic relapsing fever leaves patients with very severe deafness. Drummond's (1968) survey in Malawi indicated that three-quarters of cases of hearing loss was due to chronic suppurative otitis media. Drummond considered that an encephalitis, probably due to an Arbor virus, was important in the aetiology of sensorineural hearing losses in Malawi. In Uganda, both Roland (1960) and Martin (1967) considered that chronic suppurative otitis media was perhaps the commonest cause of hearing loss.

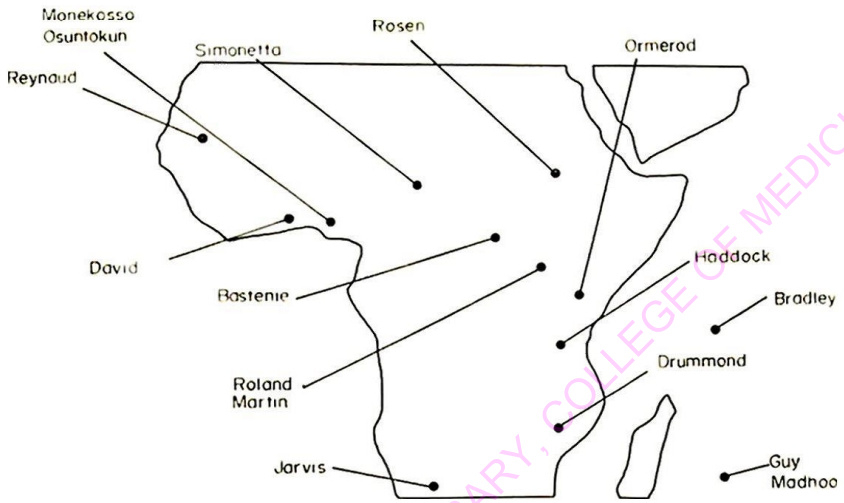


FIG. 2. Map of Tropical Africa showing locations which have been subject to special reports, together with the names of the author, or authors, of these reports (see text).

In Nigeria, both Monekosso (1963) and Osuntokun (1968) have made extensive studies of a particular otoneuro-ophthalmological syndrome, where sensorineural hearing loss is a feature. A similar, if not identical, ataxic neurological syndrome has been reported by Haddock, Ebrahim & Kapur (1962) from Tanzania. Based on his studies in Tanzania, Haddock (1965) further reported that, although benign paroxysmal nystagmus was just as prevalent in Africans as in people of Britain, Ménière's disorder was unknown in that group.

Bastenie and his associates (1962) reported a higher prevalence of endemic cretinism in association with profound deafness in the Uele region of the Congo. David (1964) has reported a village (Adamarobe) in southern Ghana where there is a high prevalence of congenital profound deafness. Finally, if we may include these islands in our discussion on tropical Africa, there have been reports of particular neuro-otological patterns in the Seychelles and in Mauritius. Bradley (1929) reported that a condition termed 'décoquée' occurred in the inhabitants of the outlying islands of the Seychelles. The condition was characterized by soreness and redness of the eyelids, an erythematous rash on the genitalia, an abnormal knee jerk and variable degrees of impairment of hearing and/or vision. Guy & Madhoo (1967) pointed out that the pattern of auditory disorders on Mauritius is so different from that which is known in Europe.

In an attempt to determine basic hearing levels which have been uninfluenced by noise exposure, the hearing of at least four pre-literate peoples have been studied in tropical

Africa. Hearing levels of the Bassari have been reported by Reynaud, Camara & Basteris (1969), those of the Heikum have been reported by Jarvis & van Heerden (1967), those of the Meban (usually referred to as the Mabaan in otological literature) by Rosen and his associates (1962) and those of the Podokwo by Simonetta (1968). The results reported for the Bassari appear to be similar to those reported by Hinchcliffe (1959) for a rural population in southwest Scotland (see Fig. 3). Although the data given by Bergman (1966) indicated that the *absolute* hearing levels of the younger Mebans were poorer than comparable European and North American populations, the data of Rosen and his associates (1962) indicated that there was very little change in hearing levels with age. Unfortunately, in this study, age had to be determined by methods of inference. Nevertheless, the question was raised

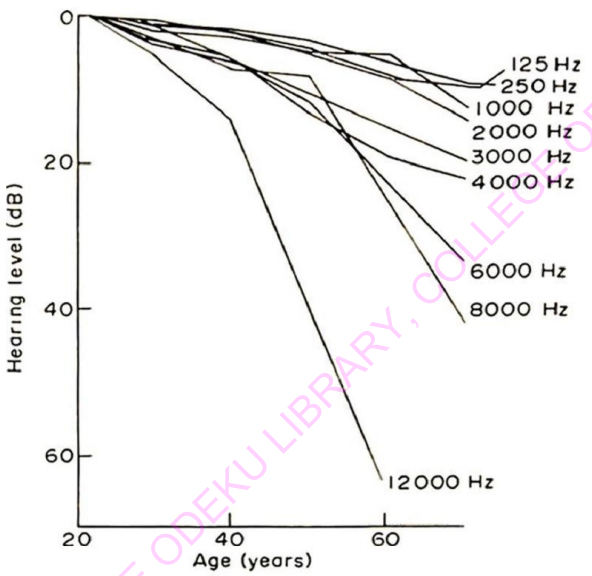


FIG. 3. Median hearing levels as a function of age obtained for a European (south-west Scotland) population. Conductively-impaired ears excluded (after Hinchcliffe, 1959).

whether Africans living in the tropics and away from the so-called civilized areas of the world did not suffer from presbyacusis. We therefore sought to discover another population which was ethnically similar and living under geographical conditions as near as possible to the Mebans, but where the age was known.

JAMAICAN STUDIES

A population conforming to the required criteria was found in rural Jamaica, and a preliminary study of a group of elderly people indicated that their hearing levels were, if anything, poorer than those of comparable populations in Britain (Hinchcliffe, 1964). There was a suspicion that this was because of the high prevalence of an endemic oto-neuro-ophthalmological syndrome, Jamaican neuropathy. It was therefore decided to undertake a comprehensive otological, ophthalmological and neurological survey of a general population in Jamaica. This survey was undertaken in 1966 and the results have

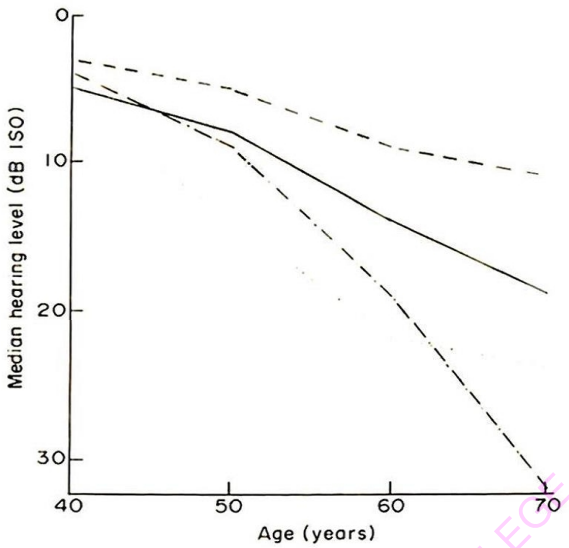


FIG. 4. Median hearing levels as a function of age for women in a Jamaican (August Town) population. Conductively-impaired ears excluded (after Hinchcliffe & Jones, 1968). – – –, 500 Hz; ——, 2000 Hz; · · · ·, 4000 Hz; — · —, 8000 Hz.

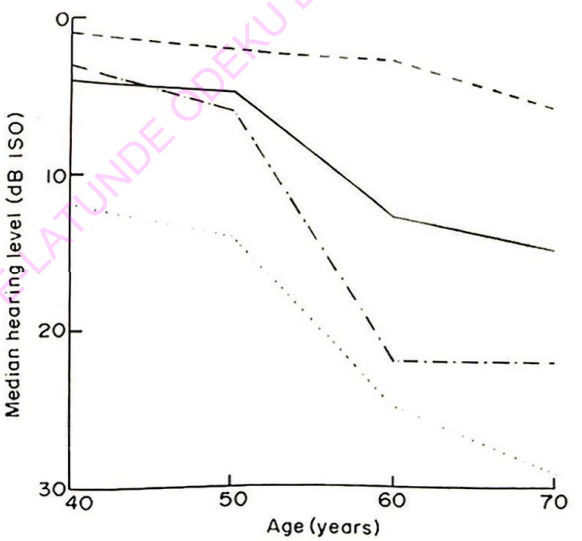


FIG. 5. Median hearing levels as a function of age for males in a Jamaican (August Town) population (after Hinchcliffe & Jones, 1968). – – –, 500 Hz; ——, 2000 Hz; · · · ·, 4000 Hz; — · —, 8000 Hz.

already been reported (Ashcroft *et al.*, 1967; Hinchcliffe & Jones, 1968). Ophthalmologists and other physicians co-operated in the study and, in order to accommodate the requirements of the other investigators, the sample, which was a total population, covered the age range 35–75 years. After the exclusion of conductive hearing losses, the median hearing levels were calculated for each age group and these are shown for female and male ears respectively in Figs. 4 and 5. In contrast to population studies of European populations, this data shows the average male hearing to be better than that for the females. Analysis of the data indicated that this was because there was an association of sensorineural hearing loss with neurological and ophthalmological signs appropriate to Jamaican neuropathy, and this appeared to

TABLE 1. Median hearing levels (dB ISO) at 500–2000 Hz of left ears of females aged 55–64 years in the Jamaican sample

	Frequency		
Sample	500	1000	2000
A	7·6	7·6	14·2
B	7·5	7·4	14·1
C	4·0	5·1	13·1
D	2·5	1·3	10·1

Sample 'A' is the total sample. Sample 'B' comprises the sample after the exclusion of the one ear of Sample 'A' that had a conductive hearing loss. Sample 'C' is after the exclusion of all subjects in Sample 'B' who had certain neurological or ophthalmological signs compatible with a diagnosis of Jamaican neuropathy. Sample 'D' comprises Sample 'C' after the exclusion of subjects with a systolic blood pressure of more than 160 mmHg, a diastolic blood pressure of more than 90 mmHg, or a history of having had 'bad blood' in association with a positive VDRL or RPCF test.

be more prevalent in the women than in the men (Ashcroft *et al.*, 1967). In European populations, it is generally held that the only two factors which influence average hearing levels are, after the exclusion of middle ear disease, noise and ageing. That this was not the case for this Jamaican population is indicated in Table 1. Here, we show the median hearing levels at 500 Hz, 1000 Hz and 2000 Hz for the left ears of women in the 55–64 years age group. Sample 'A' refers to the total sample with no restrictions whatsoever. Sample 'B' comprises those ears which do not give a true Rinne negative response, i.e. this excludes ears (only one, as it happened) with conductive hearing loss. Sample 'C' is Sample 'B' after the exclusion of the data on all subjects with certain neurological or ophthalmological abnormalities. These abnormalities corresponded to the signs which are to be found in

Jamaican neuropathy. The neurological abnormalities included ataxia, impaired joint or vibration sense, absent knee or ankle jerks and extensor plantar responses. The ophthalmological abnormalities included temporal pallor, optic atrophy and visual field defects. Sample 'D' was derived from Sample 'C' by excluding the ears of subjects with a high blood pressure (systolic greater than 160 mmHg, or diastolic greater than 90 mmHg) and those who had probably had syphilis (indicated by a history of 'bad blood' together with positive VDRL and RPCFT tests). This type of pruning of our data produces an appreciable shift in the median hearing levels that were obtained after the exclusion of conductive hearing losses. This raised the question as to whether, in future surveys, we must conduct extensive neurological and ophthalmological examinations of our subjects so that our hearing levels are more valid measures of physiological ageing (together with the effects of noise). Another point of interest is that, after we have excluded all subjects with particular physical signs to arrive at Sample 'D', this reduces the original sample of seventy-five ears to five ears. This therefore raises the question of how prevalent is normality?

TABLE 2. Median hearing levels (dB ISO) of screened ears of 55–64 years age group in August Town

	Frequency (Hz)					
Sex	500	1000	2000	4000	8000	Ears
Female	2·5	1·3	10·1	19·2	8·6	5
Male	−2·5	2·6	13·9	16·7	11·1	2

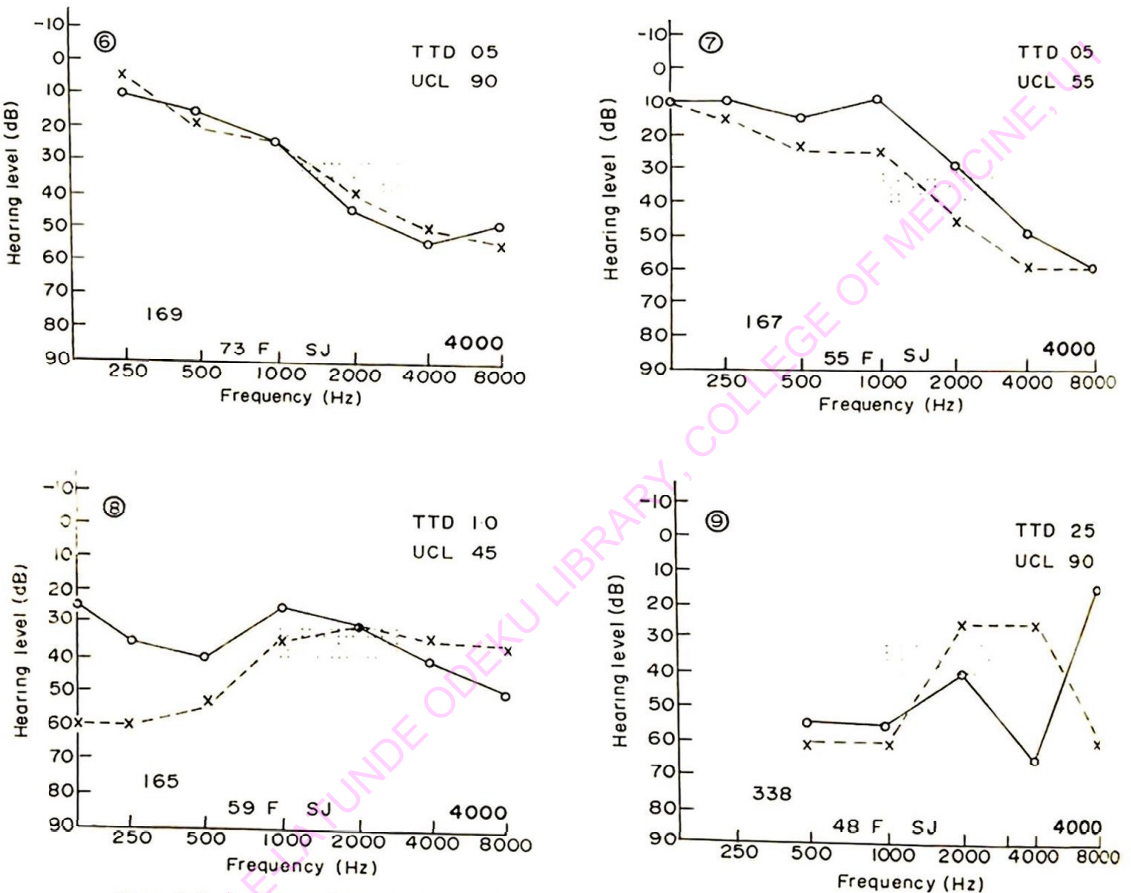
Levels quoted are after the exclusion of those subjects who had a conductive hearing loss, neurological or ophthalmological signs compatible with a diagnosis of Jamaican neuropathy, a blood pressure greater than 160 mmHg systolic or 90 mmHg diastolic, or a history of 'bad blood' in conjunction with a positive VDRL or RPCF test. Note smallness of ultimate sample size and the suggestion that there is now no sex difference in the hearing levels.

Similar pruning of the corresponding male sample left us with two ears. Moreover, after this method of pruning our data, there appears to be no sex difference in thresholds of hearing (Table 2). The hearing levels as a function of frequency for the two sexes intersect with one another. Thus the sex difference observed in the total sample is almost certainly due to the differential susceptibility of the female to one or other general medical disorders which affect hearing, and this we have alluded to previously with respect to the Jamaican neuropathy.

Another point of note is that if we compare the hearing levels of the screened Jamaican ears at 8000 Hz with those for our comparable European population (Fig. 3) we find that the Jamaican hearing levels are better. Table 2 would indicate that the average hearing levels at 8000 Hz for either sex about 60 years of age is of the order of 10 dB HL, whereas we would expect a hearing level of about 25 dB HL for a comparable European sample.

In this 1966 Jamaican survey, the audiometric examination comprised not only determinations of the thresholds of hearing for each ear at frequencies ranging from 250 to 8000 Hz, but also two topodiagnostic tests which were designed to elicit information concerning the

presence or absence of either neuronal or receptor organ lesions. As a measure of TTD (temporary threshold drift) which would indicate threshold fatigue compatible with certain types of neuronal lesions (Stephens & Hinchcliffe, 1968), we used Rosenberg's (1958) modification of Carhart's (1957) 'tone decay' test. As a measure of the loudness recruitment phenomenon, which would indicate a receptor (internal ear) organ lesion, we used Watson's (1944) uncomfortable loudness level (UCL), which is analogous to Hood & Poole's (1966)



FIGS. 6–9. Audiometric results for subjects from a Jamaican survey. ○, Right; ×, left.

FIG. 6. Subject 169 who had a sensorineural hearing loss in association with ophthalmological and neurological signs compatible with Jamaican neuropathy.

FIG. 7. Subject 167 who showed a sensorineural hearing loss together with ophthalmological and neurological signs compatible with Jamaican neuropathy. Note the low uncomfortable loudness level of 55 dB HL.

FIG. 8. Subject 165 who showed a sensorineural hearing loss together with ophthalmological and neurological signs. Again note the low uncomfortable loudness level.

FIG. 9. Subject 338 who showed a sensorineural hearing loss in conjunction with ophthalmological and neurological signs and who was diagnosed as suffering from Jamaican neuropathy. Note the peaked audiometric pattern and the marked threshold fatigue (TTD 25 dB HL). This would indicate both receptor organ and neuronal components in the auditory defect.

loudness discomfort level. The measurements were made in the sequence: hearing threshold determinations, TTD, UCL. Both the TTD and the UCL were determined at 1000 Hz on the poorer hearing ear. Inspection of the patterns of the audiograms of subjects who had a sensorineural hearing loss together with neurological and/or ophthalmological signs which indicated that the subject might have been suffering from Jamaican neuropathy, indicated a variety of patterns. However, two principal types occurred, i.e. (1) a gradually sloping audiogram, and (2) a horizontal audiogram. In addition, there occurred less frequently a type which was characterized with peaks of hearing. Although less common, this latter type is probably more important since it is perhaps pathognomonic of the neuromyelopathic syndrome, the other two patterns being non-specific. Five representative audiograms of individuals who were probably suffering from Jamaican neuropathy are shown in Figs. 6–10 inclusive. The topodiagnostic audiometric measures (TTD and UCL) are shown in the

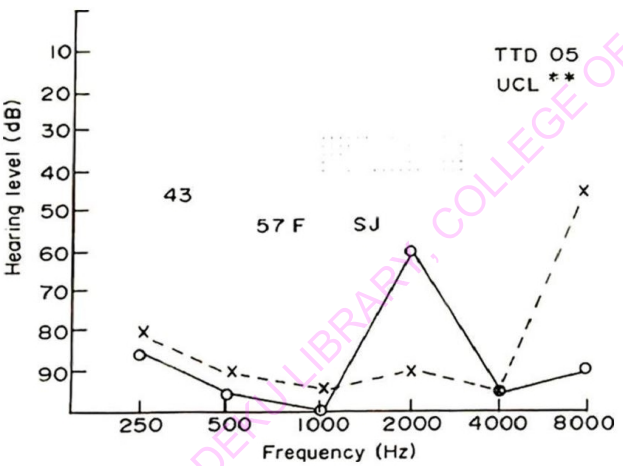


FIG. 10. Audiometric results for subject (43) from a Jamaican survey who had a marked sensorineural hearing loss together with neurological signs and who had been diagnosed as suffering from ataxic Jamaican neuropathy. Again, note the peaked audiometric appearance. ○, Right; ×, left.

top right corner of each audiogram. Each audiogram shows the age and sex of the patient in between the ordinates for the 500 Hz and 1000 Hz frequencies. The survey number of each individual is shown on the left hand side of this data. The stippled area between hearing levels of 30 and 40 dB HL for frequencies between 1000 and 3000 Hz has been referred to as the 'critical area for speech' and, according to whether the threshold of hearing is below or above this area, gives some indication as to whether the hearing loss is, or is not, sufficient to impair the person's ability to hear speech. Figures 6 (subject 169) and 7 (subject 167) are representative of the gradually sloping audiogram, Fig. 8 (subject 165) of the horizontal type, and Figs. 9 (subject 338) and 10 (subject 43) of the 'peaked' type. The type which slopes towards higher frequencies (Figs. 6 and 7) also occurs in presbyacusis and it might be said that, since the first patient (subject 169) is 73 years of age, this represents merely the effects of ageing. However, this patient also exhibited a scotoma, impaired vibration and positional sense, impaired coordination of the lower limbs and Rombergism. The problem of what does

or does not constitute this neuromyelopathic syndrome is, however, a real one, especially with epidemiological considerations in mind. Actually, only two of these five cases so illustrated (Nos 43 and 338), were diagnosed as cases of ataxic Jamaican neuropathy by the consultant neurologist (although not all cases in the survey were seen by him). Nevertheless, ophthalmological and neurological signs in subject 169 would indicate her as suffering from the neuromyelopathic syndrome, and subject 167 showed optic atrophy, scotoma, impaired vibration sense and absent knee and ankle jerks. Subject 165 showed temporal pallor and absent ankle jerks. However, this patient also gave a history of 'bad blood' and both the VDRL and the RPCFT were positive. This brings one to the question as to whether syphilis plays a part in the genesis of Jamaican neuropathy, as Rodgers (1965) claims. It would appear that the term 'Jamaican neuropathy' embraces at least two conditions: an ataxic Jamaican neuropathy and a spastic type (Cruickshank, 1956). It is the former that is characteristically associated with otological and ophthalmological defects, and not the latter, which, histopathologically, has the appearance of a chronic meningomyelitis compatible with a syphilitic aetiology (Montgomery *et al.*, 1964). One should also remember that, in Jamaica, a positive serology is not uncommon but is due to previous infection with *Treponema pertenue* and not *T. pallidum*.

The measures of threshold fatigue and of loudness recruitment did not, as this small sample indicates, show any consistent pattern in the individuals with the sensorineural hearing loss which could be associated with this particular neuromyelopathic syndrome. It is probable that both neuronal and receptor organ lesions occur to varying degrees in patients afflicted with the ataxic Jamaican neuromyelopathy. Although Osuntokun, Monekosso & Wilson (1969) caution us with regard to assuming that the nutritional neuropathy that occurs in prisoners-of-war is identical with the tropical neuromyelopathic syndrome, it is of interest that one of Spillane's (1947) nutritional neuropathies showed localized disintegration of the spiral organ in the basal turn. Montgomery and his associates (1964) report that the stato-acoustic nerves in patients with auditory symptoms also show some demyelination. Since, in the mammalian ear, the spiral organ functions as the acoustic analyser, peaked audiograms (as in subjects 338 and 43) must indicate involvement of this structure in this disorder.

The code '4000' which appears in the bottom right hand corner of the audiograms in Figs. 6–9 inclusive refers to a clinical assessment of the ethnic derivation of the subjects (the measure was not recorded in subject 43). The value '4000' indicates that these subjects were predominantly African in origin, and reflects the general finding for the occurrence of this neuromyelopathic syndrome in Jamaica. This is relevant to Behrman's (1962) suggestion that an African-linked gene, or genes, might be implicated in the aetiology of the tropical amblyopia which might be considered a 'form fruste' of the fully developed neuromyelopathic syndrome. However, because of the high prevalence of African-derived subjects in our sample, we were unable to demonstrate that there was a statistically significant ethnic difference in the prevalence of the condition.

GENERAL DISCUSSION

In view of the clinical similarities, it is tempting to consider that the Jamaican ataxic neuropathy and the Nigerian nutritional neuromyelopathy are identical disorders. Since it has been shown that cyanide-containing cassava is an aetiological factor in the genesis of

Nigerian nutritional neuromyelopathy (Monekosso & Wilson, 1966; Osuntokun *et al.*, 1969), one should consider this possibility with respect to the Jamaican ataxic neuropathy. However, Fox (1964) reported that the typical Jamaican child consumes only 14 g (dried weight) of cassava each week, compared with 140 g of rice and 365 g of white flour. Moreover the serum thiocyanate levels do not appear to be elevated in the Jamaican disorder. Wilson (1969), however, suggests that this may well be due to the fact that the point prevalence epidemiological survey was able to discover only inactive cases of the syndrome, new cases no longer being seen.

Auto-immune studies and measurements of serum vitamin B_{12} provide no evidence that the Jamaican syndrome is either an immunopathy or due to vitamin B_{12} deficiency. Although a number of workers are adamant that the possibility of a virus aetiology can be excluded it would appear that one must still search for a 'slow' virus (Gajdusek, Gibbs & Alpers 1966) as an aetiological factor. 'Slow' viruses may be hormonally and genetically controlled or determined. This could therefore account for some of the sex and ethnic differences which have been observed.

Although Reynaud and his associates (1969) have reported that the hearing levels of the Bassari people in Senegal are comparable to those that were reported for a rural European population (Hinchcliffe, 1959), an analysis of the Jamaican data suggests that the African may have somewhat better hearing levels at high frequencies than the European. This concept receives support from studies by Eagles and his colleagues (1963), by Gavini (1957) and by a subsequent report by Rosen and his colleagues (1964). Further population studies are, however, required to determine whether there are any real genetic differences in hearing level and to determine to what extent hearing levels are influenced by geogens such as cyanogenetic glucoside-containing foodstuffs.

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