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The tropical amblyopia syndrome (or tropical nutritional amblyopia) in the Mid-Western State of Nigeria

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Summary

One hundred and seven patients from the Mid-Western State of Nigeria with the tropical amblyopia syndrome are reported. Ninety-five patients (88·8%) had the amblyopia syndrome mainly; twelve patients (11·2%) had amblyopia and other manifestations of the tropical ataxic neuropathy.

The young, aged 10–20 years, represented by sixty-six patients (61·8%), are predominantly affected by the uncomplicated syndrome.

Thirteen patients (12·1%) showed muco-cutaneous evidence of avitaminosis.

Fifty patients (46·7%) had temporal pallor of the optic discs, bilaterally, another fifty (46·7%) had normal discs, two (2·8%) showed pink discs.

Generalized field constriction is the common field defect, though central or centro-caecal scotoma can occur. Red/green defect was present in seven patients.

It is a disease of the poor or those living on a basic monotonous diet consisting mainly of cassava (mannihot) and its derivatives.

It is a significant cause of defective vision in Mid-Western Nigeria.

Résumé

On a trouvé 107 patients dans l'état du centre ouest du Nigeria qui ont la maladie tropicale amblyopia syndrome. 95 patients (88·8% avaient surtout, l'amblyopia syndrome, 12 patients (11·2%) avaient l'amblyopia et d'autres manifestations des maladies tropicales de type ataxique neuropathie.

Les jeunes entre 10 et 20 ans, représentés par 66 patients (61·8%), sont principalement atteints par le syndrome sans complication.

13 patients (12·1%) ont montré cutanéo-muqueuses dues au manque de vitamines. 50 patients (46·7%) avaient une temporal pallor des disques optiques, bilatèraux, les autres 50 (46·7%) avaient des disques normaux, 2 (2·8%) ont montré des disques roses.

L'angle de vision général est limité: c'est le défaut général. La centro-caecal scotoma peut arriver, rouge/verte défaut était présent chez 7 patients.

C'est une maladie des pauvres ou de ceux qui vivent dans une carence alimentaire consistant principalement du manioc (manihot) et ses dérivatifs.

C'est une cause evidente de vision défectueuse dans l'état du centre Ouest du Nigeria.

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The tropical amblyopia syndrome is part of a wider neurological complex, the tropical ataxic neuropathy which is characterized by bilateral optic atrophy, myelopathy, bilateral perceptive deafness and polyneuropathy. Strachan in 1897, first described the amblyopia in Jamaicans. Moore (1930, 1932, 1933a, b) in Nigeria described cases of retrobulbar neuritis associated with muco-cutaneous lesions. He later attributed these to the high intake of cassava derivatives. Clarke (1935) described cases of 'pellagra' and retrobulbar neuritis among Southern Nigerians and concluded that cyanide was the toxic agent. The cyanide source was thought to be cassava.

No previous on-the-spot survey of the tropical ataxic neuropathy or tropical amblyopia syndrome has been carried out in the Mid-Western State of Nigeria.

In this paper, the ophthalmic and other neurological findings in 107 patients with the tropical amblyopia syndrome or tropical ataxic neuropathy are reported.

Patients and methods

Two thousand school children aged 9–17 years based in Benin, Warri and Irrua were examined for defective vision. Day and boarding students were included.

A review of hospital records over a 5-year period, (1966–1971) was made. During this time the Eye Clinic, Benin City, was the only Eye Clinic in the State, the other Clinic being 230 miles away in Ibadan.

Routine visual acuity for distance and near was done on all the students and where visual acuity was less than 6/6, a refraction was done to exclude refractive errors. A fundus examination after mydriasis and neurological examination were carried out on cases whose vision could not be improved by glasses and in whom there were no other ocular diseases or opacities of the media to explain the defect. Cases of macula degeneration or chronic simple glaucoma were also excluded by fundus examination, tonometry (Schiotz or applanation) and Bjerrum screen perimetry in appropriate cases.

Where vision was apparently normal, i.e. 6/6 or 6/5 and yet patients complained of defective vision, special care was taken to exclude macula degeneration or field defects from chronic simple glaucoma, or optic and chiasmal lesions. The Friedmann visual field analyser was used to investigate macula function and central field defects, and a Lister perimeter for peripheral fields. Ishihara test charts were used to determine any colour defects.

All cases had a skull and optic canal X-ray to exclude other pathology. Serum proteins, blood serology, haemogram, genotype and urine for sugar, protein and cells and stool examination for pathogens were routine for all patients.

Cases with visual acuity less than 6/24 had a C.S.F. examination for pressure, cytology, chemistry and serology. Twenty-two patients were examined.

Plasma thiocyanate was determined by the Aldridge (1944) method on deproteinized samples.

Results

Age and sex distribution

In 107 patients comprising fifty-six females and fifty-one males, all were diagnosed either as manifesting the tropical amblyopia syndrome or tropical ataxic neuropathy. This represents 0·178% of all patients seen for eye disease during the period under review.

Ninety-five patients (88·8%) had amblyopia only; twelve patients (11·2%) had amblyopia and neuropathy.

There was no predilection for either sex.

The 10–20 year age group were predominantly affected.

Socio-economic groups

Most of the patients were low-income earners. The students came from poor homes and the soldiers, although well fed in the army, had developed symptoms before enlistment during the Nigerian civil war.

Classification on a divisional basis

Thirty-two point seven per cent of the total number of patients come from Benin division which contains 17% of the total population. This may not be significant in view of the fact that the Eye Clinic is based in Benin City.

Diet and occupation

All the patients had as their main dietary staple, cassava (manihot) or its derivatives and ate it at least twice daily. The hydrocyanic content of cassava or its derivatives varies from 0·5 mg/kg to 160 mg/kg depending on the preparation, species of cassava, the soil, humidity and the time of year during which the roots are harvested (Oyenuga & Amazigo, 1957). Purupuru, the cassava preparation commonly eaten in parts of the Western State of Nigeria and

TABLE 1. No. of patients seen age and sex distribution

Age	0–9	10–15	16–20	21–25	26–30	31–35	36–40	41–45	46–50	51–56	60 and 70	Total
Male	0	20	17	7	6	2	1	1	—	—	1	55
Female	0	19	10	11	7	4	—	1	—	—		52

TABLE 2. Socio-Economic groups

Occupation	No.
Students	
Day 35	50
Boarding 15	
Teachers	4
House-wives	10
Rural Development Officer	1
Soldiers	13
Clerk/Self employed (under 300 per annum)	17
Traders	3
Salesman	1
Farmers/Gari graters	4
Unemployed	4
	107

known to contain 4–6 mg of hydrocyanic acid per 100 g of dry weight (Osuntokun, 1968) is not eaten in the Mid-Western State of Nigeria.

Very little first-class protein was consumed.

None of the patients in the series was a smoker and only one patient drank alcohol in moderate quantities.

Eight patients (7·4%) among whom were three housewives and a student were engaged in cassava growing, grating or frying.

A literate housewife developed symptoms of defective vision and painful feet especially at night after spending a two-month period assisting her mother in cassava processing, in an area largely engaged in the cassava industry. Another boy aged fifteen, after spending 5 years helping his father on a cassava plantation and eating cassava products three times a day had severe visual impairment (visual acuity: right 6/60; left 6/60) and severe truncal ataxia. His visual fields showed a generalized. central field depression with centro caecal and quadrantic scotomata (Fig. 1, b).

Two female patients gave a history of sudden visual impairment accompanied by pain on ocular movement during a period when they had to flee into the bush to avoid the ravages of the civil war (1967–70). Both had bilateral optic atrophy and concentric constriction of their visual fields. Neither was ataxic. Plasma thiocyanate was raised in both of them. These two probably represent a 'forme fruste' of the tropical ataxic neuropathy.

Intermittency of symptoms was demonstrated in a girl aged 21, who was blind from bilateral optic atrophy, deaf and had severe sensory lower limb ataxia. At one stage, she developed a transient paresis of the left arm lasting 48 h. Plasma cyanide and thiocynate were raised. Both fell on admission to the wards. She was a non-smoker.

TABLE 3. Classification of patients on a 'divisional basis'

Division	Total state population, 2·5 million % of state population	Number of patients seen	% of total no. of patients
Benin East / Benin West	17%	35	32·7
Warri	6%	22	20·5
Western / Eastern Urhobo	20%	5	4·7
Ishan	11%	21	19·7
Asaba	7%	6	5·6
Agbor	5%	5	4·7
Ivbiosakon	5%	5	4·7
Western Ijaw	9%	4	3·7
Isoko	5%	1	0·9
Aboh	7%	3	2·8
Etsako	4%	0	0
Akoko-Edo	4%	0	0
Total	100%	107	100%

Population figures: Federal Republic of Nigeria Census, 1963.

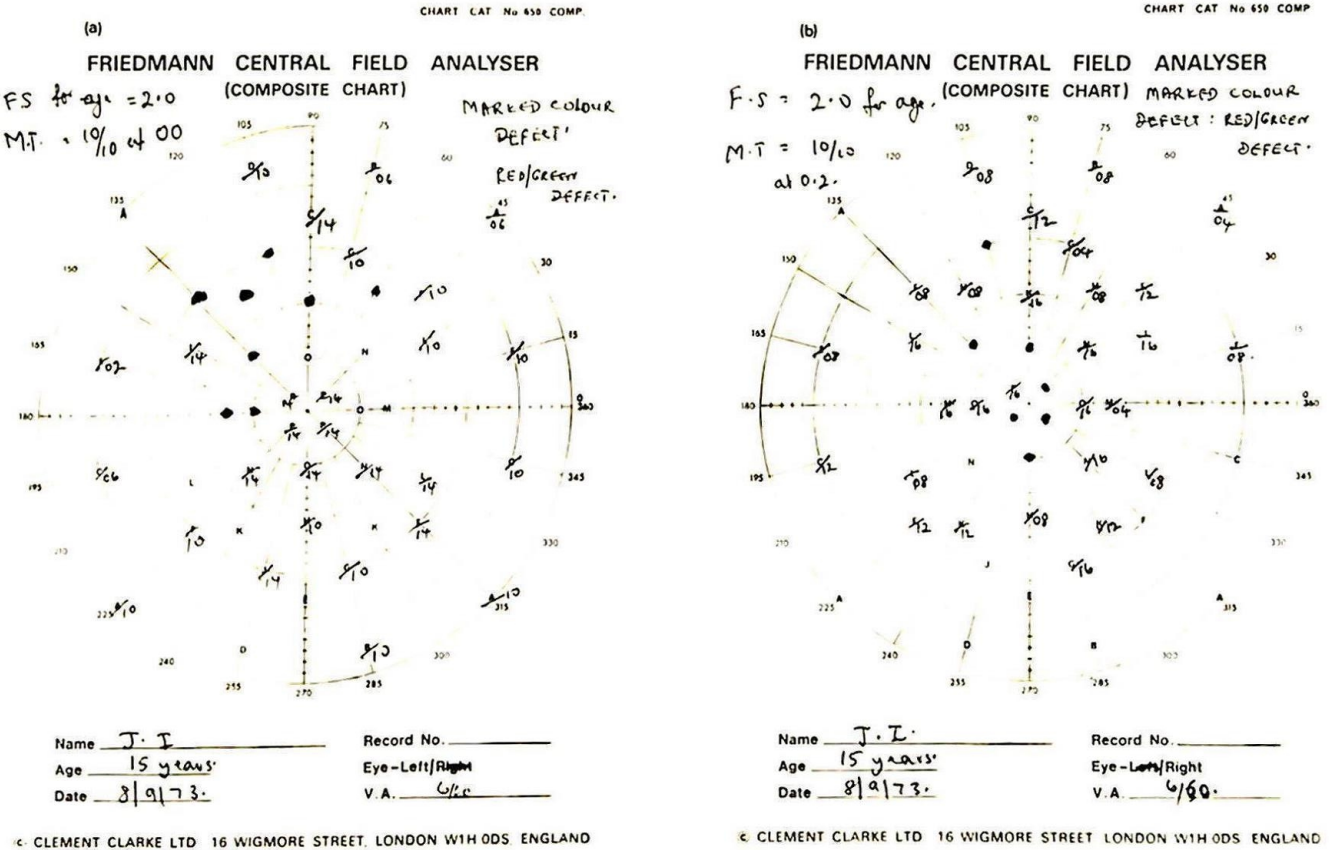

FIG. 1. (a) Upper quadrantic scotomata demonstrated. (b) Paracentral and upper quadrantic scotomata.

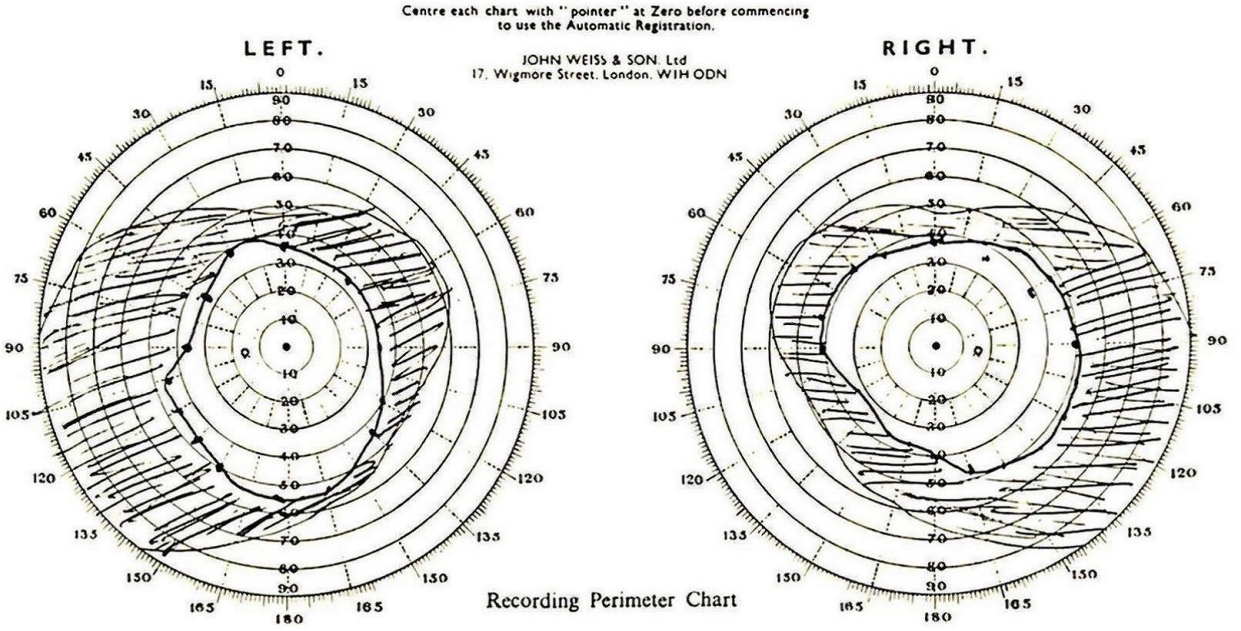

FIG. 2. Generalized field constriction.

Another patient complained of intermittent visual blurring.

Inability to see things in the distance or read the writing on the blackboard in the case of students was the commonest complaint (40·2%). Other symptoms included sudden visual loss (two patients), tinnitus (two patients), deafness (four patients), tingling sensation in feet (two patients), painful feet at night (one patient), ataxia (two patients), transient weakness in one arm (one patient). The average duration of symptoms before seeking medical aid was 2 years.

Muco-cutaneous signs of 'avitaminosis' (follicular hyperkeratosis, angular stomatitis, glassitis) were present in thirteen patients (12·1%).

Blindness was present in six patients (5·6%), (58·8%) had visual acuity ranging between 6/24 and 6/60, 32·7% between 6/12 and 6/18, 2·9% between 6/6 and 6/9.

Discs

Fifty patients (46·7%) had normal optic discs. Fifty others (46·7%) had bitemporal pallor of their optic discs, while five patients (4·67%) had bilateral optic atrophy. Two patients had very 'pink discs'.

Visual fields

Concentric peripheral field constriction was the commonest field defect observed in eighteen of the patients whose fields were investigated [illegible]. On confrontation screen testing, two patients had centro-caecal scotomata. Two other patients investigated with a Friedmann visual field analyser had central field depression with centro-caecal and other quadrantic field defects.

Seven patients showed red/green blindness on Ishihara colour testing.

Neurological examination

Sensory and motor disturbances were mainly confined to the lower limbs. 3·7% had superficial sensory loss (touch, pain), 13·1% had posterior column sensory loss (vibration and joint position sense).

Lower limb ataxia (heel-shin and toeing the line) was present in 13·8%, while finger-nose ataxia was present in one patient only.

Fourteen patients (13·1%) had truncal ataxia.

The cerebro-spinal fluid was normal in the twenty-two cases examined.

No significant abnormality was present in the urine or stool.

Discussion

Various reports (Scott, 1918; Wright, 1928; Moore, 1930, 1932, 1933a, b); Clarke, 1935; Oluwole, 1935; Dagazon, 1956; Cruickshank, 1956; Monekosso & Ashby, 1963; Quere *et al.*, 1967; Osuntokun & Osuntokun, 1971) have confirmed the distribution of the tropical amblyopia syndrome in communities of low standard of nutrition and in prisoners of war camps (Spillane, 1947; Montgomery *et al.*, 1964).

Monekosso & Wilson (1966), Osuntokun (1968, 1969) have since demonstrated the role of chronic cyanide intoxication in the aetiology of the Nigerian ataxic neuropathy and tropical amblyopia syndrome. A similar aetiology has been advanced for the tropical ataxic neuropathy among Tanzanians (Makene & Wilson, 1972).

The eight patients (7·4%) in this series engaged in cassava growing or processing lend support to the observation that cassava processing may be an occupational hazard.

All the patients ate cassava products at least twice daily and in three patients investigated the plasma thiocyanate levels were raised. Only one of the patients in the series drank alcohol occasionally. Alternative sources of thiocyanate; milk, vegetables and beer were consumed rarely and in very small quantities. None of them was a smoker. All the patients were relatively poor.

Six patients with early visual symptoms and normal discs showed marked improvement when they followed advice on change in diet and were treated with hydroxocobalamin, confirming reports of reversibility of the condition in the early stages. No beneficial effect was, however, achieved in a therapeutic trial with hydroxocobalamin, cysteine and riboflavin tables (Osuntokun, 1973). The dietary change may, have been the principal factor in the improvement observed in the six patients.

The tropical amblyopia syndrome has been observed among adults (Scott, 1918; St. John, 1936; Dekking, 1947; Money, 1958; Osuntokun & Osuntokun, 1971). Some have considered it a disease primarily of the young (Moore, 1934; Whitborne, 1947; Monekosso & Ashby, 1963;

Quere *et al.*, 1967). In this study, the highest incidence was in the 10–20 years age group, a finding partly influenced by the fact that students are more likely to notice early warning signs of defective vision than others, e.g. illiterate farmers and housewives, partly by the method of selection.

The disc findings are intriguing. Fifty-five patients (51·4%) had either optic atrophy or temporal pallor of the optic discs. Fifty patients (46·7%) had normal discs, and two patients (1·9%) had pink discs. It is the young adolescent presenting with defective vision, normal macula and discs that poses a problem in diagnosis. With no refractive error present, a relevant dietary history, muco-cutaneous signs, polyneuropathy or myelopathy are invaluable diagnostic pointers. By the time optic disc pallor is marked, heel-shin ataxia or difficulty in toeing the line may be present.

Temporal pallor in this syndrome needs to be distinguished from temporal pallor of the normal or glaucomatous disc and macula degeneration in the Nigerian. Both may occur at a relatively early age, 11-35 years (Ayanru, in preparation). Chronic simple glaucoma occurring at 15–36 years has been described in Nigerians (Kodilinye, 1966).

Total blindness may result from the tropical amblyopia syndrome as in six of these patients.

Although multiple sclerosis is virtually unknown in the Nigerian, intermittency of symptoms with remission as in two of the patients in this series is highly suggestive of the demyelinating nature of the condition (Williams & Osuntokun, 1969).

Eighteen of the twenty patients whose visual fields were investigated had concentric peripheral field constriction, the commonest abnormality in the Nigerian (Osuntokun & Osuntokun, 1971). Quere *et al.*, in Senegal (1967) observed centro-caecal scotomata in 115 patients—peripheral fields were all normal, a finding suggestive of a different aetiological agent. In tobacco amblyopia, however, where cyanide is the toxic factor (as in the tropical ataxic neuropathy) the typical field defects are relative or centro-caecal scotoma, findings also observed in some of the patients with the tropical amblyopia syndrome. Accumulation of cyanide in the vitreous with damage to the unprotected peripheral nerve elements and macula (Makenzie & Philips, 1968; Lancet, editorial, 1969), combined in some cases with isolated retrobulbar neuritis explains the different fields obtained in various patients with the tropical amblyopia syndrome.

That the Mid-Western State of Nigeria is an endemic focus for the tropical ataxic neuropathy (Osuntokun, 1971) and tropical amblyopia syndrome (Osuntokun & Osuntokun, 1971) is confirmed by this study.

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