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Clinical and Laboratory Investigations

Albinism in Nigeria*

A CLINICAL AND SOCIAL STUDY

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SUMMARY

A study of 1000 Nigerian albinos, all of Negro stock, showed various types of albinism with their different modes of transmission—oculocutaneous, ocular and cutaneous. The much higher incidence among the more settled communities in the south, compared with the more nomadic communities in the north, may be related to greater inbreeding tendencies in the south.

The sun and society are hostile to the albinos. Under the tropical sunshine, their melanin-deficient skin develops wrinkles, lentigines, actinic keratoses and epitheliomata from which they may die in early adult life or in middle age. Myopia and other ocular defects retard the progress of many albinos in school and they eventually drop out to seek disastrous menial outdoor occupations.

Registering albinos early in life, assuring their families that albino defects are confined to the skin and eyes, advising on protective clothing and sun-screening agents, correcting myopia, assisting with indoor occupations, and early treatment of actinic keratoses and skin cancer should help many albinos to attain social acceptance and a ripe old age.

Albinos are seen in every racial stock, but they stand out most strikingly among Negroes who are normally darkly pigmented. The existence of albinos in all the ethnic groups in Nigeria has long been recognized. Every major Nigerian language has a name for albinos: *Afin* (Yoruba), *Anyali* (Igbo), *Mbakara-Obot-Ikot* (Efik), *Eyaen* (Bini), *Ugobu* (Idoma), *Zebia* (Hausa). The Efik name is rather derogatory and means literally 'a white man from the bush'. Albinos are taunted from the cradle to the grave with such names as 'D.O.' (District Officer), a reference to the days when District Officers were Europeans.

References have been made to the incidence of albinism in Nigeria based mainly on casual observation (Barnicot, 1952; Clarke, 1958, 1959; Watkins, 1972) but no organized study to establish the real incidence had been undertaken. The present study was inspired by seeing epitheliomata destroying the faces, scalps, necks and shoulders of young albinos in their teens or twenties. Cancer of the skin in Negro albinos has been reported before (Clarke, 1959; Oettle, 1963; Marshall, 1964). The pitiful

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sight of young albinos driven by an uninformed society to menial outdoor work, under the pitiless tropical sun, was an associated reason for taking up this study.

MATERIALS AND METHODS

Starting from albino patients who came to the hospital with cancer of the skin, pre-cancerous keratoses, freckles, wrinkling of the skin or any other skin condition, attempts were made to trace all members of their families who were albinos.

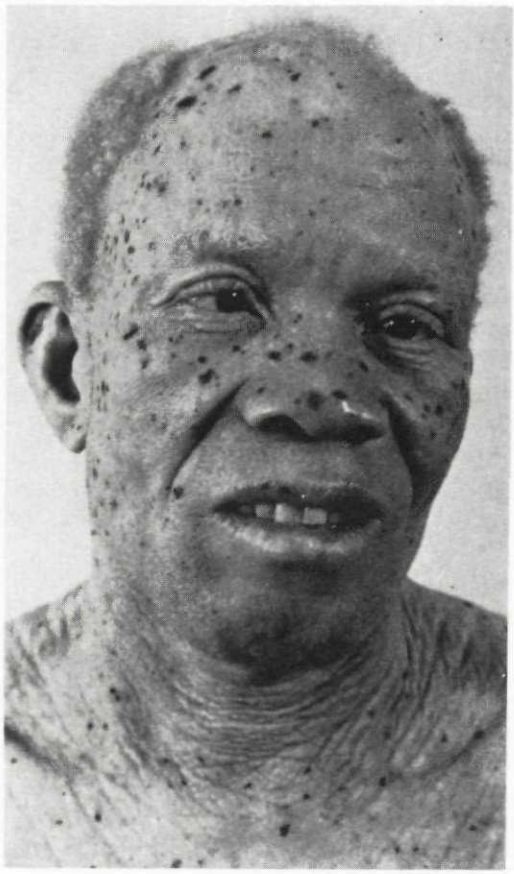

FIGURE 1. Thirty-year-old albino with ephelides and solar elastosis.

Then, through questionnaires and visits to various educational, health, and religious institutions, and to markets, more albinos were traced. The questionnaires explained briefly:

(1) The reason for the study (the delicacy of albino skin and its proneness to actinic damage).

(2) The greatest danger to albinos (skin cancer).

(3) The ultimate aims of the study (care and protection of albino skin, early detection, treatment, and prevention of actinic damage and its complications). The name for albinism was given in each of the major Nigerian languages for easier understanding of the questionnaires.

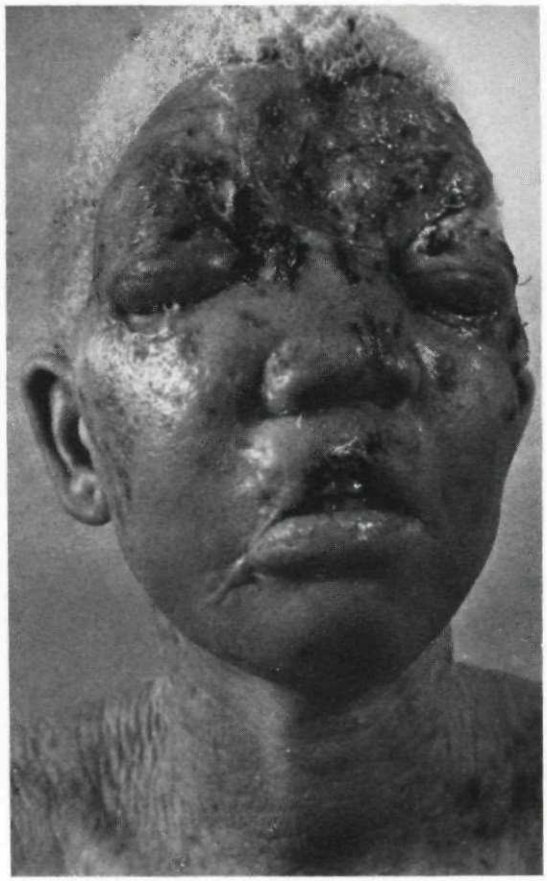

FIGURE 2. Twenty-eight-year-old female albino with squamous cell epithelioma.

A simple information slip on protective clothing (head-gear, clothes, and dark sun-shade glasses) was sent to each albino and, for those at school, an additional letter was sent to the school authorities recommending exemption of albinos from outdoor activities during the hottest and brightest hours of the day. For the albinos in public employment, their deployment indoors where possible was also recommended.

The first phase of the study covered the East Central State (where the author lives and works). The second phase covered all the twelve states in Nigeria. Albinos who came to our clinic with some dermatological problems or in response to our questionnaire were given a full medical examination, and were sent to the ophthalmologist for examination for ocular defects, and correction of amenable defects.

Histopathological examinations of albino skin and of the various lesions associated with actinic damage were carried out.

Factors in the physical and social environment which affect the survival of albinos, and therefore the incidence of albinism in the community, were recorded. Traditional beliefs about causation, and traditional attitudes to albinos were also investigated.

RESULTS

Nine hundred and eighty-nine albinos have been traced so far, 517 in East Central State and 472 in the eleven other states. Most of the albinos examined (95%) were of the oculocutaneous type characterized by absence or a markedly decreased amount of melanin in the skin, hair and eyes. Two per cent were cutaneous, with pale skin and hair but normally pigmented iris and retina. Three per cent were ocular, with pale iris and retina but normally pigmented skin.

Skin. At birth and in early childhood, albino skin has a delightful smooth texture but a sickly yellowish ivory colour. Its delicacy soon becomes obvious. The common bacterial infections of childhood (impetigo contagiosum, folliculitis, furunculosis and cellulitis) all look more severe than in normally pigmented children.

As the child grows and goes outdoors, the early effect of actinic irradiation (if it is not too intense) is a rather pleasant tanning of the exposed parts. But in time, the cumulative effect of years of exposure produces a motley of destructive skin lesions—sunburn, blisters, solar elastosis especially on the neck, arms and other exposed parts, ephelides (freckles), centrofacial lentiginosis, solar keratoses, chronic superficial ulcers and ultimately squamous cell epitheliomata and basal cell epitheliomata. The most exposed parts—forehead, ears, cheeks, nose, neck and shoulders usually suffer most. Sunburn with or without blistering, hypertrophic skin, ephelides and centrofacial lentiginosis are seen in all cases above 10 years of age. No albino over the age of 20 years was free from malignant or premalignant lesions. Keratoses and indolent superficial ulcers were present in 50%. Five of twenty albinos (3 males and 2 females) with squamous-cell epitheliomata treated (surgically) in the past 3 years, have died from extensive epitheliomata eroding the eyes, skull or large blood vessels.

Hair. The hair is a pale yellow colour in childhood and crinkly like normally pigmented hair. Later on, the hair may become deeper yellow, ginger or carrot-coloured. Dyeing it black produces a ridiculous and disappointing result because of the extreme contrast and the poor uptake of the black dye.

Eyes. The cardinal symptoms in oculocutaneous or ocular albinism are photophobia (all cases) and visual difficulties. Examination of the eyes revealed nystagmus in the great majority of cases, head nodding in very severe cases, and strabismus in a few cases.

The nystagmus and head nodding are due to lack of fixation rather than to neurological disorders. Absence or great paucity of pigment was seen in various parts of the eye—the iris, choroid and retina. The irides were pale or light-grey in childhood, becoming translucent or ash-grey in adult life. The fundi were pale or albinotic in all cases and the macula was improperly developed. Refraction tests revealed simple myopia in 90% of the cases and myopic astigmatism or hypermetropia in the rest.

The age distribution of the albinos showed a rapid drop in numbers after the third decade indicating the shortened life span of albinos. This is represented graphically (Fig. 3). Some of the older albinos may, however, have been missed in the questionnaire survey. Among 517 albinos traced in the East Central State, 89% are in the age range 0–30 years while 10% are in the age range 31–60 years. Among 472 albinos traced in the rest of the country, 92·9% are in the age range 0–30 years while 6·3% are in the age range 31–60 years. Among 1000 consecutive non-albino skin patients, 79·5% are in the age range 0–30 years while 20% are in the age range 31–60 years. A population distribution in a developing country (Lucas & Gilles, 1973) (Fig. 4) showed 78·3% of the population in the age range 0–30 years and 20·7% in the age range 31–60 years. This resembled closely the pattern in our non-albino patients.

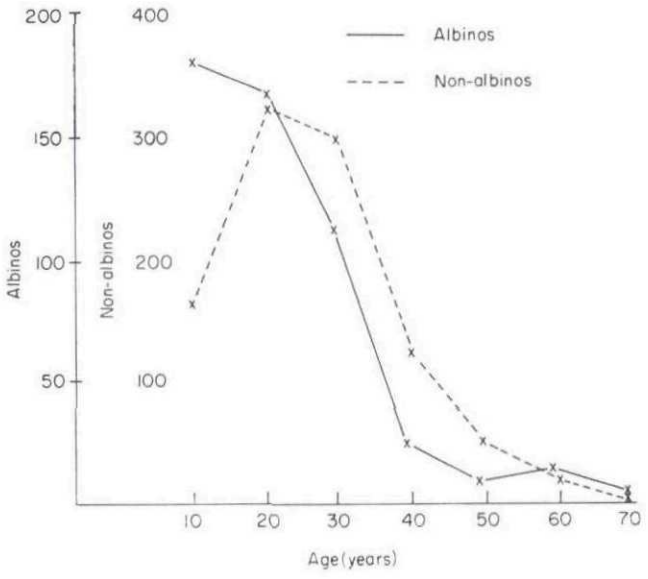

FIGURE 3. Age distribution of albino and non-albino cases.

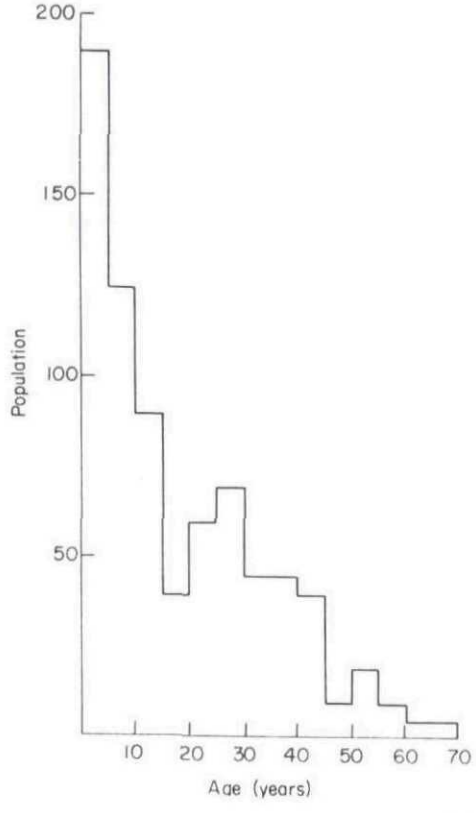

FIGURE 4. Population distribution in a developing country (Lucas & Gilles).

Sex distribution. Fifty-three per cent of the albinos were male and 47% female.

Albinos per family. In the East Central State, 63·3% of the affected families had one albino each, 26% had two albinos each, 7·7% had three albinos each, 2% had four albinos each, and 1% had five albinos each.

DISCUSSION

Nigeria is a vast country (area 357,000 sq. miles) inhabited by over 50 million people of almost exclusively Negro stock.

The map (Fig. 5) shows significant differences between the number of albinos in the six northern states where the population is more nomadic (72 albinos) and the six southern states where the population is more settled (917 albinos).

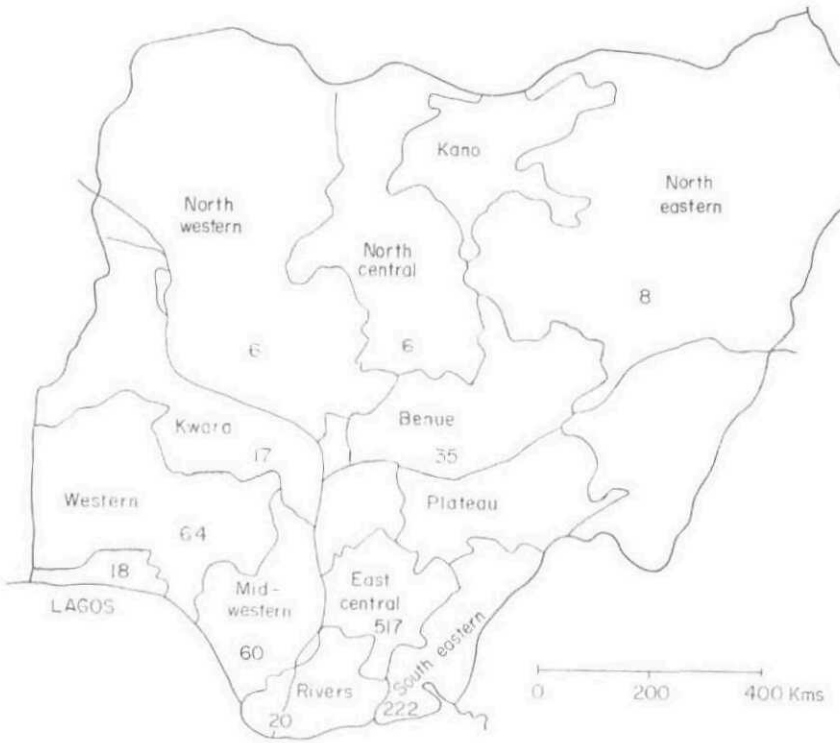

FIGURE 5. Nigeria: Distribution of albinos.

Inbreeding has been recognized as a major factor where there is a high incidence of albinism. The Cuna tribe of Indians on San Blas islands in Panama, inbreeding in isolation, have the highest incidence of albinism in the world—sixty-three per 10,000 (Keeler, 1970). The greater inbreeding tendency among the ethnic groups in Southern Nigeria may account for the higher incidence of albinism among them. Inbreeding, not in the sense of deliberate consanguineous marriages (though some do take place), but in the sense of marriage within the same town or clan, is very common in Southern Nigeria. This inbreeding has been encouraged in order to perpetuate some assumed superior qualities in the

clan, not knowing that it also encouraged the perpetuation of albinism and probably of other recessive genes as well.

Nigeria, lying in the torrid belt from 4° to 14° above the equator, is sunny throughout the year. The role of solar radiation in the development of skin cancer has long been recognized (Unna, 1894; Findlay, 1928; Urbach, 1969). It is no surprise, therefore, that the majority of albinos, lacking the protection which melanin offers, develop skin cancer during their second, third or fourth decades, and succumb to it if no medical help is available.

Traditional attitudes

Nowhere in the traditional views on albinism is it believed to be associated with inbreeding. Rather, weird aetiological theories range from punishment from the gods, to conception during menstruation, or to seeing frightening sights during pregnancy.

Albino children are seldom as treasured as normal children because the inherent delicacy of albinos is well known.

Social life is very difficult for albinos. Neither their relations nor anyone else would agree that most albinos are normal in every respect except for the absence of pigment in their skin, hair and eyes. Albinos are taunted at home and in school. The less enlightened the society, the worse it makes life for the albinos. Many albinos drop out from school partly because of uncorrected refractory errors and partly because of discouragement. Reasonable employment is difficult for them to get, and marriage even more difficult, especially for the girls. The proportion of albino women who are married is very low.

Scope and prospects of the current study

This pilot study has spot-lighted only the tip of the iceberg. The problems of albinism and its study in such a vast and populous country are immense. They include, characteristically slow and incomplete questionnaire returns, ignorance, shyness, despondence, and reluctance to reveal the full extent of family involvement. A complete enumeration of all the albinos has not been achieved even in the East Central State which has been fairly extensively covered. It would therefore be unscientific to give the precise incidence of albinism at this stage. What has been recorded is the number of albinos traced so far. Only an exhaustive enumeration (to be carried out in future) can give the real incidence. As a pointer, among 20,000 skin patients seen in skin clinics over the past 6 years, 102 were albinos, giving an incidence of fifty-one per 10,000 in this group, albeit selected; but, from the figures available in this study, the incidence in the East Central State would be one per 15,000.

CONCLUSION

Albinos need protection against their major enemies—the neglect of an uninformed society, and the hostility of the tropical sun. Registering all albinos early in life, educating the society on the educability of most albinos with correctable ocular defects, and offering them encouragement and guidance through school and employment will help to restore them in society. The damaging effect of the sun can be minimized by protective clothing, sun-screening agents, and indoor occupations.

Regular examination of all albinos for early detection and treatment of the various premalignant and malignant lesions to which they are prone deserves to be included in the current anti-cancer campaign to which the medical world is committed. Finally, when further work has made the diagnosis of the heterozygous (carrier) state easy, then genetic counselling (avoidance of dysgenic marriages) should be advocated since, of the two cardinal variables in the albino-skin cancer association, namely inheritance and sunlight, the former is more readily amenable to control.

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