

# A NUTRITIONAL DISEASE OF CHILDHOOD ASSOCIATED WITH A MAIZE DIET—AND PELLAGRA

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In a recent issue of the 'Archives of Disease in Childhood,'<sup>1</sup> under the first part of the title given above, Dr. Cecily D. Williams gave a very interesting account of a syndrome found among the native children between one and four years old in the Gold Coast Colony, which she states has not been described before. The syndrome consists 'of oedema, chiefly of the hands and feet, followed by wasting, diarrhoea, irritability, sores, chiefly of the mucous membranes, and desquamation of areas of skin in a manner and distribution which is constant and unique,' and results, it is believed, from an improper diet containing an excess of white maize.

Dr. Williams categorically denies that this affection is pellagrous and has given her reasons. These appear to be based on standard text-book descriptions of the disease which, unhappily not always very comprehensive or correct, have misled her into using fallacious criteria for differential diagnosis. Without having seen these cases in Accra, I am venturing to dispute the correctness of the diagnosis and to suggest that they are cases of pellagra.

The history of these infant patients is that they begin to get irritable, the skin and hair lose their normal colour and glossiness, there are attacks of diarrhoea, slight oedema of the hands, feet and face but the skin is not dry and there is 'no branny desquamation as in pellagra.' Suddenly, at the end of seven to ten days black patches appear on the skin of the extensor surfaces about the ankles, knees, wrists and elbows which spread so that 'soon the legs and forearms, knees and elbows are covered with a sort of crazy pavement of this thickened epidermis.' These thickened crumpled and darkened patches then exfoliate leaving a pink raw surface exposed underneath. 'The patches of desquamation progress up the thigh and may become severe on the buttocks. Very small patches may also be seen on the face, back and elsewhere.' It may be noted that the blackened patches on the skin appear first where there is any focus of irritation or pressure.' In spite of having given a really very excellent description of a pellagrous eruption in a black-skinned native child, Dr. Williams says, 'Their distribution and their character remain distinct from the rash that is typical of pellagra.'

Further on it is stated 'the skin at the corners of the mouth and eyes also begins to peel off, leaving raw areas.' Sores on the mucous membrane in the mouth appear and corneal ulcer and photophobia may be noted.

Diarrhoea becomes persistent and pyrexia is present, irritability is marked; death usually ensues in a month or less in untreated cases. No changes in the reflexes were found.

In the case reports some other features are mentioned, 'the skin all over was atrophic in appearance . . . there were darker almost paved-like patches on the forearms and legs . . . the typical distribution of pellagra was absent, i.e., the dorsum of the hands and feet seemed to escape until the last stages of the disease.' Stomatitis is mentioned, also purulent discharge from the vagina. Casual mention is made of an older child (4 years) who had had a series of improvements with relapses.

Dr. Williams has described what, to workers with an extensive clinical experience of pellagra, must appear as a complete picture of that disease in young children. The present writer over twenty years ago also failed to recognize pellagra when he came across the disease in Central Africa and described<sup>2</sup> the first group of cases to be recognized in that continent south of Egypt.

In the diagnosis of pellagra allusion is commonly made to the triad of symptoms—dermal, gastro-intestinal and nervous, much stress is laid upon the distribution of the skin lesions, and the seasonal recurrence is a point to which prominence is given. The descriptions in text-books are based upon adult cases and reference is rarely made to the skin manifestations as seen in dark races or to a number of other important points.

In the first place the great variability in the course of the disease must be remembered. In a fulminating case, in which continuous diarrhoea is a prominent symptom, death may ensue after a few weeks' illness. The skin lesions might be overlooked and nervous manifestations unrecognized. On the other hand, the disease may run a very chronic course over many years with intermittent symptoms and the case find its way into an asylum for the insane. In many cases the onset is insidious: in others, apparently after the causes have been at work for some long period of time (this side of the question cannot be discussed here) the onset of 'the attack of pellagra' is sudden and the course may be rapid. In children there may be a high mortality, while in infants, if unprotected (they usually are protected) the disease may run a course, well exemplified in the Accra infants, resembling infantile beriberi as recorded by George Bray<sup>3</sup>. The order in which systematic symptoms appear may vary, but in the acute cases gastro-intestinal and dermal symptoms are closely associated, though one may precede the other by some short interval as in Dr. Williams' cases. As in her cases so in pellagra the diarrhoea is a simple diarrhoea intermittent at first. It soon becomes persistent, often unaccompanied by pain, and it may be associated with a tendency to vomiting as seen in the Accra infants.

The loss of gloss and colour of the skin and hair mentioned by Dr. Williams as occurring in sick natives and seen in minor degree in her patients is of course well recognized. In many chronic cases of pellagra it may be well marked. Dr. Williams speaks of 'depigmentation' suggesting that there is actually a removal of the granules of melanin from the skin and hair. This point has nothing specifically to do with pellagra perhaps, but it would be interesting if it could be settled by histological examination. The

appearance may be due merely to a draining away from the skin of blood and other tissue fluids.

The actual skin lesions and their evolution as recorded by Dr. Williams are those of pellagra in a dark-skinned race as has been stated above. They are, I venture to say, absolutely diagnostic. In no other condition is found the lesion as described. The black patches noted by Dr. Williams resemble the skin lesions I described as if the area had been painted over with a layer of paint made of lamp-black which then becomes cracked and wrinkled and peels off leaving a pink, sometimes raw surface beneath.

It must not be forgotten however, that the skin lesions in pellagra are variable. On the one hand, the process may be acute with haemorrhage into the skin and necrosis, or the erythema may be mild and followed by a branny desquamation only. On the face the rash may take on an acneiform character; on the shin the skin may be smooth and shiny having the appearance of having been painted over with collodion, to crack later, characteristically giving rise to the 'crazy pavement' effect, so aptly designated by Dr. Williams.

The distribution of the skin lesions in the West African infants has obviously caused Dr. Williams difficulty. In most text-books the statement is commonly made that the pellagrous rash appears on exposed areas of skin and the reader is led to believe that it can only appear on those parts exposed to sunlight, that in fact the skin is photosensitized and the rash is determined by exposure to the sun's radiations. This is often coupled with another statement to the effect that the skin of the terminal phalanges of the fingers is never affected and that the skin of the palms is likewise exempt.

Sandwith pointed out that the rash was absent from the terminal phalanges of pellagrous Egyptian peasants and demonstrated that this was due to their being hidden from the sun's rays by the handle of their hoes. The statement in regard to the exemption of the phalanges is, however, now repeated by many writers without the explanation, and as such is quite untrue. To say that the palms are never affected in pellagra is also untrue. Certain changes are not uncommon, but they are not the same as those on the dorsum and are overshadowed by the latter.

The pellagrous rash may tend to appear on certain predetermined areas of skin just as happens in the exanthemata, the predisposition being due to certain anatomical and physiological conditions and those areas may correspond to 'exposed areas' and sunlight may commonly be an exciting cause of the eruption, but the truth should be stated in general terms and is to the effect that any part of the skin in pellagra may be affected and that any form of trauma or irritation may determine the site of the lesions. Any form of radiant energy, any chemical stimulus, any form of physical irritation may, it seems probable, act as an exciting factor, very much as in smallpox for example.

Thus are explained all those more unusual localizations of the pellagrous rash and thus are accounted for the localizations in the African children of Dr. Williams as she herself has shown without realizing the significance of her own acute observations. She noted that the rash appeared at points of pressure and irritation and that the distribution was not the same as is

described for adults which after all is what one would expect. The buttocks in some of the infants were affected as may be the case in pellagrous children with diarrhoea, the soiling of the skin being sufficient to determine a pellagrous eruption in this area.

In pellagra other very characteristic signs, as I pointed out many years ago, are the little sodden areas of skin, which appear white in black-skinned races, at the angles of the mouth, at the external canthus of the eye, at the free margin of the prepuce, about the vulva (often associated with a vaginitis) and about the anus. All these, with the exception of the condition about the anus, have been noted by Dr. Williams in her cases. In man a very characteristic change in the skin of the scrotum is often observed. It would be interesting to know if anything of the kind had been seen by Dr. Williams in her male infants.

The stomatitis and ulceration of mucous membrane, again so characteristic of pellagra, were also seen in the Accra infants.

Lastly, in regard to nervous and mental symptoms it may be stated that in pellagra, perhaps the commonest sign referable to the central nervous system is an increased knee-jerk. In a great proportion of cases, where the disease is established, the phenomenon is seen, but just how soon after the beginning of 'an attack' in infancy this change in patella response occurs I do not know. The absence of any alteration in the reflexes in the rapidly fatal cases among the Accra infants in no way invalidates a diagnosis of pellagra. In annually recurring attacks of pellagra in children there may be no nervous symptoms the first years. Some mental change, too, is common in pellagra apart from the gross changes to be witnessed in severer cases. In children the mental responses are naturally limited in certain directions, that seen in infants is one of irritability. In Dr. Williams' cases this was a characteristic symptom. The fever, absence of severe anaemia, leucocytosis and any characteristic lesion at necropsy in the infant cases are, of course, all part of the picture of pellagra.

Dr. Williams has, I believe, described typical cases of infantile pellagra. When in the title of her communication she states that the syndrome she has recorded is associated with a maize diet she suggests that she believes the disease is due to a maize diet. It is an interesting point: the association of pellagra with maize is centuries old, but the proof that a maize dietary plays any part in producing pellagra is still unconvincing.

In conclusion I would like to suggest in regard to treatment that a trial be given to yeast which I have myself found very efficacious. In the Gold Coast Colony there may be some naturally occurring fermentable juice which could be used, one similar to the 'toddy' with which Bray<sup>3</sup> saved generations of infants in the Central Pacific Island of Nauru from death with infantile beriberi. Experiment and research would soon demonstrate the possibility of using such a substance and the conditions and precautions necessary in making an active preparation.

#### REFERENCES.

1. Williams, C. W., *Arch. Dis. Childh.*, Lond., 1933, VIII, 423.
2. Stannus, H. S., *Tr. Roy. Soc. Trop. Med. & Hyg.*, Lond., 1911, V, 112. *Ibid.*, 1913, VII, 32.
3. Bray, G. W., *Tr. Roy. Soc. Trop. Med. & Hyg.*, Lond., 1928, XXII, 9.