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The Beginning of the Kwashiorkor Story in Africa

By

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Dr. Cicely Williams

In 1952 Professor J. F. Brock and Dr. M. Autret reported to the World Health Organization that kwashiorkor "is the most serious and widespread nutritional disorder known to medical and nutritional science". But it was unknown to the textbooks: Manson-Bahr put it in the fourteenth edition of Manson's Tropical Diseases in 1954; Dr. Donald Hunter put it into the ninth edition of Price's Textbook of Medicine in 1956. I hope Dr. Cicely Williams was glad; she wrote the first comprehensive description of the "Deficiency Disease in Infants" in the Gold Coast Annual Report, 1931-32. She described the fatty liver found at two post-mortems, and patients had improved after giving condensed milk. No name was given to the disease; that had to wait until 1935.

A charming description of her life tells the laity something of her pioneer studies in extremely difficult circumstances. Being a lady, and a very gracious lady at that, she arrived by instinct at the correct answer. On the third page of her biography, breathless on arrival from her paediatric appointment at the Queen Elizabeth Hospital for Children in Hackney, to Ghana, in 1929 she saw "twins . . . large bellies . . . peeling skins . . . reddish tinge to their hair" and announced that "It's a mysterious disease . . . it's not in the textbooks". Experts nowadays tell me that I must call it Protein-Energy-Malnutrition (PEM), but never mind; probably they will change again and call it Energy-Protein-Malnutrition (EPM), but I will call it only and forever kwashiorkor, in honour of Cicely, for she was right, and the experts had brought a needless dispute between us for more than a decade.

Since writing the first draft of this article I have been delighted to receive quite unexpectedly a copy of the *Nutrition Reviews*, November, 1973. The entire volume is devoted to honour Dr. Cicely Williams and to mark her eightieth year. Tributes are paid to her by doctors in America. On behalf of those who worked in Africa I would like to add ours. I only met her once for any length of time. One feels at once in the presence of a great person, full of charm and wisdom. In this Review Cicely tells her own story (p. 334-340). Therein I saw for only the second time the admirable first description of the disease published in the Gold Coast Annual Medical Report, 1931-32. We never saw this in East Africa.

Medical missionaries in Kenya

It is difficult to say where a river starts; there are tributaries and springs, and waters under the earth. Before it is too late I wish to pay tribute to the unnamed discoverers of kwashiorkor in Africa. They preceded Cicely and myself by a decade and many are forgotten. Medical missionaries in Kenya first described this disease; I learned from them. Philp (1924-25) in the first volume of Kenya Medical Journal almost certainly described kwashiorkor at the Tumutumu Mission Hospital of the Church of Scotland Mission, and his successor, Dr. R. U. Gillan (1934-35) wrote an excellent description of the disease in that hospital. All the doctors in the Kikuyu Reserve seemed to know about the disease in the early 1930's, and a clinical meeting of the British Medical Association (Kenya Branch) on 13th December, 1933, was devoted to this subject. Dr. A. R. Shaw was soon to publish his article and he demonstrated a case; three doctors, including myself, all stated they had seen similar cases. The condition was regarded as nutritional oedema and ascribed to protein deficiency. Much progress might have been achieved if the Kenya doctors had followed their convictions; unfortunately they

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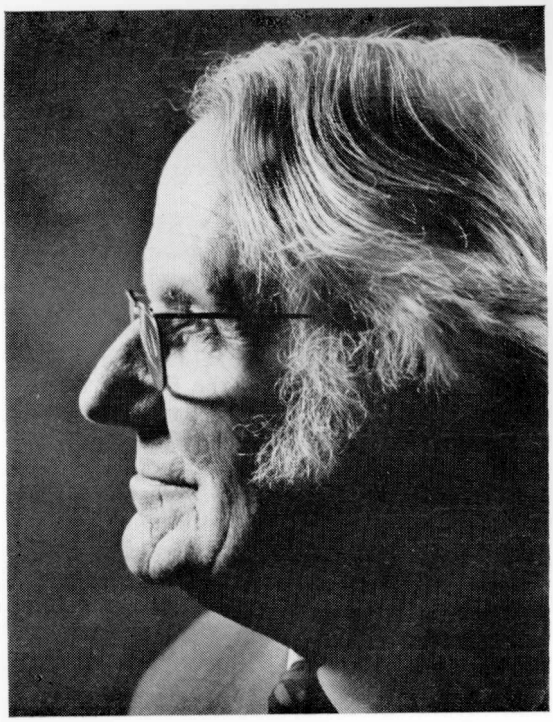
listened to a nutritional expert in London, Dr. H. S. Stannus (1934, 1935), who criticised the excellent description of Dr. Cicely Williams, and stated that she could not recognise a pellagrous dermatosis.

Experts should never be consulted about the status of a new idea in medicine. They are almost always wrong; they have climbed on the steps of their own research to a pinnacle of great eminence; they are hostile to a new idea which may challenge their pre-conceived notions. Stannus was the authority in Britain on pellagra; his papers on the manifestation of this disease in Malawi (Nyasaland) early in the century were classics, and he said that the crazy-pavement dermatosis of kwashiorkor was pellagrous. So did Loewenthal, an aspiring dermatologist in Uganda; so did Sequeira, the doyen of British dermatologists who had retired from the London Hospital to Kenya. The first that we doctors in East Africa heard of Cicely Williams' pioneer descriptions of kwashiorkor in Ghana were the adverse criticisms of Stannus, reported in summary form in the Tropical Diseases Bulletin in 1935 and 1936.



The Author.

My introduction to the problem

My own introduction to kwashiorkor was in 1932 and is told in detail elsewhere. Unduly elated with my membership I had been given the task of training African nursing orderlies who had finished four years of primary education. This was at the old KAR Hospital, Nairobi, with its one water tap, no microscope and no test tubes. Allotted no medical beds I complained and was granted what no one wanted, Ward V, wherein a mixed bag of women and children kept open house with the main dispensary of the hospital. "In sullen mood I went into the corrugated iron building; it was dark, it was full of bed lice, the children were all crying, but many of them had brown hair, although they were African babies, and many of them had swollen legs with oedema. Two or three were in most beds. On walking through the ward at night, lit by one hurricane lantern, one could hear the low moan of the children dying; we did not know why they were dying, but it was kwashiorkor . . . We only knew that when the baby developed black spots on the skin he was going to die . . . The other doctors thought it was worms. So we dosed them of their worms and they went on dying. At that time we were losing about three-quarters of our cases of kwashiorkor". So I started taking notes, and going to post-mortems. But there the trouble started: the muscles were flabby, the liver yellow, the thymus small; often there were ascaris, but not always so. Sometimes a little pneumonia, but nothing to explain death.

The pellagra controversy

Stannus' criticism of Cicely Williams threw my investigations into disarray; but I found a Goan photographer who consented to take good photographs of the dermatosis, in between the numerous wedding engagements, if I took mother and child to his studio. Armed with these photographs of the dermatosis, many clinical notes and several post-mortem sections, I climbed up the steps of the Harley Street consulting room of Dr. Stannus in 1935. He looked long and lovingly at the photographs of the dermatosis, but he refused to look at the sections of skin, liver and other organs. Instead he lent me a reprint of an article about the pathology of pellagra in American negroes and suggested that I would be able to harmonise both descriptions. In deep misgiving I left the palace of the expert, and published my cases of "Infantile pellagra".

This paper was accepted by the Archives of Diseases in Childhood, but one never heard about publication in those days until months later, as air-mail communications had barely started. The first intimation was a cable which was long and its contents were obscure. Something called nicotinic acid had been synthethised in the U.S.A., and the tablets were coming out on the next plane. I remember the day when I unwrapped the big brown bottle that was to cure all my cases of "Infantile pellagra". I treated ten cases; eight died. I could have wept.

It was a tragedy of the first order that after her first pioneer studies of kwashiorkor in Ghana Dr. Cicely Williams was "rusticated" to a smaller unit at Kumasi. Perhaps even there her unorthodox views were not too popular and her unsolicited sudden transfer to Malaya in 1936 came as a great shock. She ceased to contribute to our knowledge of the disease, but she was the mother of kwashiorkor. Mothers are however not above entering into an argument with other members of the family and a lively argument took place in the medical journals (1939-1941) in which she maintained that it was a new clinical entity, with a characteristic dermatosis, and I called it infantile pellagra, then coined the phrase malignant malnutrition as one after another of the B group of vitamins were tried to supplement the obvious deficiencies of nicotinic acid, but I met no success.

Uganda, confusion with congenital syphilis

In 1935 I had been transferred from Kenya to Uganda to help in the teaching of medicine at Mulago Hospital and Makerere University, but was dismayed to find that not only was the disease called congenital syphilis in Uganda, not only had Mulago Hospital been built as a VD clinic, but the whole of the child welfare clinics had been canalised along these lines. There were no fears of overpopulation, instead "the Baganda were in danger of becoming extinct" due to the wholesale deaths from congenital syphilis! Posted to Mbarara in 1935 I attended child welfare clinics almost daily throughout the province. At a dispensary clinic a hundred or so infants would attend each day and many registers were found in which about 90 per cent. of the infants were diagnosed as congenital syphilis and received antisyphilitic treatment. Most of them had scabies, malaria, yaws or kwashiorkor. Next year back at Mulago Hospital I began to realise that the standard accepted teaching of colleagues was that kwashiorkor was not a disease; it was either congenital syphilis, or a muddled mixture of non-specific signs, such as wasting, anaemia and oedema, which might accompany any lingering infections of malaria, helminths or congenital syphilis.

This was to set one at variance with colleagues for fifteen years and agreement was only reached after the report of Brock and Autret in 1952. My own antics did not help the situation. Every few years I would introduce another terminology, another definition. We still continued to lose about 40 per cent. of the children when we gave them milk, or a nutritious diet and all known vitamins. How could this be a deficiency disease? How had the ignorant Africans recognized a disease that we had missed?

Other pioneers in Rhodesia and South Africa

Meanwhile a host of other clinical observers in Africa, such as Gelfand in 1946 in Rhodesia, and Altmann in 1948 in South Africa, were describing the disease; the latter was the first to point out that the old German paediatricians Czerny and Keller (1906) had described *Mehnahrschaden* (damage by cereal flours, a similar condition even if the dermatosis was not as marked a feature. The brilliant histological studies of Professor J. Gillman and his brother, Dr. T. Gillman, culminated in their massive study of malnutrition; alas, they were calling it pellagra in 1948 when they invited me to give some lectures on kwashiorkor at the Medical School, Johannesburg. Night after night I sat up looking at slides with Joe; I have never seen anything so brilliant or so instructive in all my life. He was on fire: organ after organ were examined from conception to the grave and it mattered little, he said, if we called it pellagra, as he did, or kwashiorkor as Cicely did, or malignant malnutrition, as I did; they were all deviated lifetracks and a kaleidoscopic series of changes were occurring in many endocrine glands, especially the adrenal and pituitary, and in the internal organs of liver and pancreas. I have never been so inspired and so perplexed in all my life.

The scientific basis

The fundamental discoveries had to be made by others. Himsworth described the fatty liver of protein deficiency; Brock, after his W.H.O. report in 1952, opened a special unit at Cape Town and made basic studies of the response to protein, calories and the amino-acids.

Excellent research was conducted in the Belgian Congo, West Africa and India. During 1949 in Uganda, Professor J. N. P. Davies described in considerable detail the pathology of kwashiorkor; he described the atrophy of the pancreas as the basic lesion. This explained the inability of the children to digest food; it partially explained the high mortality rate. Unfortunately we did not realise that the pancreatic atrophy soon responds to treatment, so that other pathologists at Mulago Hospital could not confirm his findings while he was away in 1950 in the U.S.A. These were only explained by Dr. Margaret Thompson who published her studies on the pancreatic enzymes of cases in my ward. This was published in 1952 and in 1950 the tide turned, for solid conviction was growing that here was a distinctive clinical-pathological entity, as Cicely had said. It is not proposed to discuss in any detail these scientific achievements which we were able to incorporate in 1952 into our book *Kwashiorkor*. Dr. R. F. A. Dean opened the Medical Research Unit of Research in Infantile Malnutrition at Mulago Hospital in 1951. Excellent work in the West Indies and Central America, likewise India, had taken place in the late 1940's.

Recognition in 1950

The tide turned in 1950. It is a strange experience to work, to sleep and to rise again, then to work, to sleep and to rise again for fifteen years and always to be asking: Is the disease there? I fear I was a nuisance to my colleagues; I was certainly a burden to my wife. There is always this period of confusion, of doubt and hesitancy, with a new idea or a new disease. I had mistranslated kwashiorkor as the "Red Disease" in earlier writings and this attracted a mail from the other side of the Iron Curtain; this did not help. The misunderstandings are now erased; the wounds have healed.

When Medical Headquarters in 1950 heard that Professor Brock was coming to Uganda and Dr. Cicely Williams was also coming, things began to happen. The Governor had me to drinks at Government House and bringing in the sherry himself and speaking of Professor Brock asked "Where will this racket all end?" My Director had me to Medical Headquarters and told me that he felt it understandable that they had re-organised the the Department of Medicine; I was not to be the professor. It had been my research. But to neither question was reply made. When a few months later Brock and Autret reported to the World Health Organization I sent a copy to His Excellency and said that therein lay the answer to his question.

Soon after Professor Brock came to Uganda in 1950, Dr. Cicely Williams also came. There was little to talk about, except that she was right. We would write our book; Jack Davies was coming back from the U.S.A.; Rex Dean was coming from Cambridge. We would call it Kwashiorkor. I was staying on; I was not retiring. Sitting in my drawing room after lunch, with my wife, she said "Hugh, did you know at Geneva, at W.H.O., we called this nutritional disease number one in the world?" Once again I was without words by way of answer. We went out into the garden, I picked some flowers, gave them to her, and she went away.

All this was specially pleasing, for W.H.O. had written to me the year before, asking for a memorandum on kwashiorkor to be considered by the F.A.O./W.H.O. Expert Committee on Nutrition at its first session in Geneva in October, 1949. I wrote the memorandum and sent it to Medical Headquarters, but Dr. M. Autret confirmed at dinner a few days later, with my wife and Professor Brock, that it never reached Geneva. I had not realised this.

Looking back on all these events of some 25 years ago, one wonders why all this proved so irksome at the time. Of course, there had to be uncertainty about the existence of a new nutritional disease, not described in any textbook; it was doubted if it had a distinctive clinical picture, pathology, aetiology and treatment. As late as 1949 research even in Uganda seemed to have shown that it was probably only hookworm anaemia. All this for myself proved one of the most fortunate accidents in my life. The college Department of Medicine went forward under an excellent professor. I was able to give all my time to writing our book on kwashiorkor, published in 1952. Then I had almost eight more years in which to give my mind to the curious prevalence of non-infective disease in Africans. About this I wrote in 1960 and that was the beginning of another story, that of fibre. At the moment we cannot make up our minds if fibre is an even bigger story or merely another "racket".

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Initial Experience with an Endemic Disease Register

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Part 1 THE SYSTEM

Definition and description of the System

The endemic disease register is a simple register of chronic, recurrent or important endemic disease of any aetiology which needs long term follow up. Patients can be divided into three groups following initial assessment and treatment.

(1) Those for whom no further follow up is necessary. They are either dead, cured, or there is no further useful treatment indicated.

(2) Those who require short term follow up to establish full recovery or a definite diagnosis. These patients must eventually enter group (1) or group (3).

(3) Those who require long term follow up for treatment, observation or both. This group forms the basis of the endemic disease register.

The register is designed to achieve maximum efficiency in follow up with maximum convenience for patients in the circumstances of this country; at the same time allowing extraction of important data for planning and for assessment of efficiency.

At the time of preparation of this article (Dec. 1973) the register had been in operation for one year in the Gwelo District (population 100 000) and for two months in the Charter District (population 100 000). At present it only includes African patients dependant on Government and Local Authority Health Services.

The design and management of the system is undertaken by the staff of the Gwelo Hospital who would normally manage the inpatient and outpatient treatment of these diseases, and much of the follow up has been undertaken willingly by the staff of local clinics.

Although different from them in many respects, the system has been derived from the example set by the Tuberculosis, Cancer, and Psychiatric Registers.

The general problems that the system is designed to solve have been previously outlined. (New Thoughts for Old, *C.A.J.M.*, Vol. 19, Nos. 10, 11, 12.)

Description of the system.

On registration an Information Card is completed for each patient. Each card has a serial number with prefix and suffix, which gives information on the disease group, the origin of the patient, and the hospital undertaking the registration.

The Information card is a folded card presenting four faces or pages each of foolscap size.

The first page contains all the basic data concerning that patient including address, social background, investigation results, past and present history, physical findings and diagnosis. As far as possible all information is recorded in a simple standardised fashion.

The second page contains:

(i) spaces for drugs prescribed, arranged in such a manner that it can be seen at a glance which drugs the patient should be receiving; the dose; the date that dose was started and, where applicable, the date the dose was changed or the drug stopped.

(ii) spaces for three simple facts to be recorded opposite the name of the current month, with space for eight years. The three facts to be recorded vary according to the disease, e.g. for diabetes they