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KWASHIORKOR ON THE NAVAJO INDIAN RESERVATION:

A discussion of the disease as seen throughout the world and a report of three cases in Navajo children

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Kwashiorkor has been called the most prevalent and serious nutritional disturbance and the most important medical problem among children in the world today. It is prevalent among all underprivileged peoples in the tropical and subtropical countries throughout Africa, Asia, the Far East and Central and South America. The first cases reported from the United States involved four Negro children in Louisiana who were older than the usual age for this syndrome (one was twenty years old) and were so heavily infested with roundworms that one of the authors of the report admitted the worms may have caused the protein deficiency state. Jelliffe, in his excellent review, alluded to an occasional case having been seen in New Orleans but he may have been referring to the above mentioned four cases.

The cases to be reported in this paper are more similar to the cases of kwashiorkor seen in other areas of the world and are perhaps the first cases of "typical" kwashiorkor to have been reported from the United States.

The syndrome as described in African children consists of:

1. retardation of growth in the late breast feeding, weaning and post-weaning ages
2. alterations in hair and skin pigmentation
3. hypoproteinemia and edema
4. a variety of dermatoses
5. fatty infiltration, necrosis and fibrosis of the liver
6. gastrointestinal disorders including anorexia, vomiting, diarrhea and steatorrhea
7. irritability and apathy
8. a high mortality rate.

In a series of one thousand cases the usual age was four months to five years with eighty-five per cent of the cases in the seven to thirty-six months age range. Jelliffe divides the cases into those with "pure" kwashiorkor (protein deficiency but a sufficient intake of calories to maintain body fat) and nutritional marasmus (deficiency of calories as well as protein). His criteria for the diagnosis of kwashiorkor are:

The opinions expressed in this article are those of the author and are not to be construed as representing policy of the U. S. P. H. S.

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1. pitting edema
2. low body weight
3. wasted muscles with overlying fat of normal thickness
4. apathy or a "silent, listless inertness".

Often found, but unnecessary for diagnosis, are hair changes (hypochromotrichia, sparseness and easy pluckability), the classical "flaky paint" dermatosis, loose stools and signs of associated vitamin deficiencies, especially ariboflavinosis. The patient with nutritional marasmus has a very low weight, marked wasting of both muscle and subcutaneous fat and no edema or significant apathy. Edema is present in only one form of advanced malnutrition and that is kwashiorkor. Marasmus and nutritional dwarfing are not associated with edema. In marasmus the decreased protoplasmic mass presumably contains a normal proportion of protein.

Children with kwashiorkor are underweight in spite of the edema. The edema involves the feet, hands and genitalia but ascites is rare. The urine contains but a trace of albumin. There is hypoalbuminemia. These features, plus the response to the feeding of protein, lead to the conclusion it is indeed a protein deficiency state.

There is a dyspigmentation of the hair and skin. The hair lacks lustre and becomes pale or almost red. The skin lesions are not characteristic. It has been argued the dermatosis is that of pellagra but the rash involves irritated areas, such as the diaper area, and not exposed areas as in pellagra. There are definite geographic differences (perhaps dependent upon the type of vegetable protein in the diet). For example, in India there are color changes at the periphery of the hair which create a halo effect.

The non-purulent dermatitis usually consists of a dried skin that peels easily, without bleeding, and exposes a pink or white skin underneath (in a dark complexioned person). Lassitude, apathy, photophobia and the picture of dejection and misery characterize the youngsters. There are frequent bouts of diarrhea and usually some degree of anemia which is almost always normocytic or slightly macrocytic.

The literature pertaining to this condition is now large in keeping with its rank as a leading cause of world-wide morbidity. There have been biological investigations of extremely high calibre. Some workers believe the liver elaborates increased amounts of ferritin which acts as an antidiuretic. There is a decrease in pancreatic enzyme activity as in starvation from any cause and this is perhaps the reason for the distressing diarrhea. The anorexia and pancreatic insufficiency lead to further and more complicated malnutrition (superimposed marasmus).

Gomez et al reported the "recovery syndrome" (seen in the Western Hemisphere) in which after successful treatment the liver enlarges with obvious signs of a collateral circulation and there is an increased eosinophile count and a hypertrichosis. In the Spanish-speaking countries of this hemisphere kwashiorkor (which means "red boy" in the Ga language of the Gold Coast) has been called...
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"syndrome pleuricarencial de la infancia". As in Africa and Asia it occurs when mothers' milk becomes inadequate for the needs of growing children and there is no complement of protein of high biologic value.

Not all the serum proteins are depressed to the same degree. Albumin and beta globulin are relatively more depressed than the other globulin fractions which are only slightly altered. Jelliffe and Edozien cite Gitlin's study of radioactive preparations showing the hypoalbuminemia to be due to decreased synthesis. The serum albumin rises to normal with recovery so it would seem the depression is not due to liver damage because the liver is the principal site of synthesis and there is a prompt response to the furnishing of a proper protein in the diet.

The preferred treatment is not parenteral protein as was originally thought but is the furnishing of suitable protein by mouth. Skimmed milk preparations are most widely used and a gastric lavage is often necessary when anorexia is present. Success is usual except in terminal states by using proteins of high biologic value cautiously introduced, then vigorously administered. Recovery occurs in seven to ten days in most series. There is first a loss of weight due to loss of edema fluid then a true gain in body weight occurs.

The following cases were diagnosed and treated at the P.H.S. Indian Hospital, Tuba City, Arizona, between October, 1960, and March, 1961. The environment differs considerably from that of previously reported cases of kwashiorkor in that the altitude is over 5000 feet, the climate is most dry and the temperature ranges from moderate in the summer to below freezing in the winter.

R. A. (008836), a 1½ year old Navajo girl, was brought to the hospital because of swelling, irritability and anorexia of one week's duration. History was inadequate. She had been seen at the clinic previously for pharyngitis and the parents considered her to be well until the onset of edema. She had received "no milk for a long time" and had rarely been given meat. Her diet consisted primarily of tea, soda water and beans.

The child was intensely irritable. She did not appear malnourished. The hair was normal and there was slight edema of the eyelids. Ascites was thought not to be present. There was no dermatosis but there was a pitting edema of the hands and feet. The remainder of the physical examination was normal; the weight was 21 lb., 1 oz.

Initial laboratory study revealed a trace of albuminuria; hgb 9.5 gm., wbc 14,000 with a normal differential count. Total serum protein was 4.8 gm. per 100 ml., the albumin was 2.32 gm. per cent. The urea nitrogen was 8 mg. per 100 ml. and the cholesterol was 164 mg. per 100 ml.

The patient was given 0.6 ml. of a standard polyvitamin mixture per day and was fed whole milk and pureed meat thrice daily. After three days she was described as a "fair eater" and at seven days the weight had fallen to 18 lb., 1 oz. and the edema disappeared. From that point her behavior improved, she became
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quite placid and was described as a “good eater”. Ten days after admission her hair began falling out and continued to do so for the next four days when the process ceased just short of total alopecia. The few remaining hairs were definitely dyspigmented and could be easily and painlessly plucked. The happy, edema-free youngster was discharged on the nineteenth day — the weight being 19 lb., 4 oz.

Urinalyses revealed an occasional trace of albumin. Nine days after admission the serum protein was 4.65 gm. per cent, albumin 2.15 gm. per 100 ml. Fourteen days after admission the thymol turbidity was 9 units (normal under 4) and the G0 transaminase 42 units. The parents demanded release before the follow-up protein determinations could be performed. The Mantoux test was negative as was the chest film.

N. S. (009252), an 11 month old Navajo girl, was brought to the clinic because of swelling and was admitted. She was said to have been well until 4 months before when swelling of the hands, feet and labia was noted. Since that time she had refused to eat well and had lost weight. There had been intermittent vomiting. The edema recurred 1 day prior to admission and according to the mother it made the child appear less starved. She had had skin trouble “for a long time”. It was impossible to obtain a feeding history and quite possibly the child was cared for by siblings as is so often the case in a Navajo group.

The patient resisted examination in a normal fashion and was not apathetic or irritable. The hair was normal. The liver was not palpable and the heart was considered normal. The labia majora were markedly edematous. She had edema of the hands and pitting edema of the feet and lower legs. There was a patch of erythema on the nape of the neck with flaky skin overlying it. A dermatitis behind the ears was more characteristic of pyoderma. There was angular stomatitis and cheilosis. The weight was 15 lb., 13 oz.

Initial laboratory findings were a Hg. of 10 gm., wbc 13,800 with a normal differential. The urinalysis was negative; the total serum protein was 4.5 gm. per cent.

Meat was ordered three times daily and no supplementary vitamins were given for 5 days. The cheilosis and neck lesions remained unchanged; the impetigo was erased by an antibiotic ointment. The cheilosis disappeared after vitamins were ordered.

One week after admission the weight curve had reached its nadir — 13 lb., 11 oz. The skin hung in loose folds and she appeared quite marasmic. At that time she began to eat well, and she became more lively and began to gain weight without the reaccumulation of edema. One week after admission the total serum protein was 5.15 gm. per cent; the albumin fraction was 2.35 gm. Two weeks later the serum protein was 6.88 gm.; the albumin 3.48 gm. The chest film and tuberculin skin tests were normal.

One month after admission the weight was 17 lb. and the patient seemed to be fully recovered. The dermatitis on the neck had persisted, the bacterial and fungal cultures from the skin being negative. A tension pneumothorax with
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Staphylococcal empyema developed and was treated with closed thoracotomy and appropriate antibiotics. She later was sent to another institution for open thoracotomy and upon return was quite well, the skin was clear and the protein determinations at the other hospital were normal.

M. B. (007646), a two year old Navajo girl, was admitted because of sudden swelling. One month before a mild diarrhea began which ceased 2 days prior to admission at which point the edema occurred. The child had been increasingly more listless. For the month during which there was diarrhea the child was fed nothing except “soup” and soda water and occasionally whole milk. The pregnancy and delivery were normal and she had not previously been ill. She had received vitamins until 2 months before and they were restarted a week before admission — a supply having been furnished by missionaries.

The girl was listless, irritable and manifested generalized edema with palpebral and ankle edema being most prominent. There was no pleural fluid or ascites and the liver was not palpable. The most striking finding was an erythematous rash on the gluteal and perineal skin and on the extensor surfaces of the elbows. The rash extended up the back and down the thighs. It did not resemble “diaper rash” as it is commonly seen and the skin was flaky and peeled easily without bleeding. There was cheilosis and glossitis. She was afebrile; the weight was 23 lb.

The initial laboratory study revealed a normal urine, wbc of 28,000, 75% lymphocytes. The total serum protein was 5.0 gm. and the albumin 2.2 gm. per 100 ml. The blood urea nitrogen was 8 mg. per 100 ml., the thymol turbidity 7.7 units and the hg. 13 gm. per 100 ml. Studies of the skin for fungi were negative. Tuberculosis was ruled out.

The patient was given the regular pureed ward diet with whole milk and no vitamin supplement. 50 ml. of lyophilized plasma were administered on each of the first two hospital days. Edema disappeared on the 4th day — the weight then being 22 lb., 11 oz. On the 10th day the weight had fallen to 20 lb., 3 oz. She gained weight steadily after that. Four days after admission there had been no change in the severe dermatitis and vitamins were then ordered. The skin was normal 6 days later. The liver never became palpable. Improvement in behavior and appetite was noted 5 days after admission and by 7 days she was a model patient. Thymol turbidity 3 weeks after admission was 9.0 units.

It is not surprising that kwashiorkor exists among the Navajo people. There is profound poverty combined with the lack of knowledge of what constitutes an optimum diet plus the fact that infant care is often delegated to siblings while the mother tends her sheep. In addition to breast milk the children usually receive the gravies from stews or the water in which beans were cooked. Later the child may be given a piece of fat or fried bread to chew. It is unusual to find a person whose sole momentary occupation is feeding a child — the child may be given spoonfuls of a juice while the mother is eating and in some areas of the reservation a piece of pre-chewed meat may occasionally be given the baby. The diet rarely
approaches an adequate caloric bulk or adequate composition until the child's motor development is such that he can maneuver around the dining area and successfully delve into the pots of food. Blatant malnutrition is common. Proper study of disease in the Navajo people is difficult because of the formidable language barrier, the extreme cultural differences and the inability of the historians to express time in terms of weeks and months. Because of the sparse laboratory figures this report serves only to indicate that the cases here reported are cases of kwashiorkor. Each patient did not exhibit the multiplicity of pathological changes as is usual in Africa but each exhibited more than are necessary to make the diagnosis of kwashiorkor as established by Jelliffe. Where it was possible to perform sufficiently frequent laboratory studies it appeared that the edema disappeared and that there was improvement in the clinical status before the serum albumin had risen to a normal level.

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