I, 1.—Died aged 82, but had suffered all his life from occasional nose-bleeding and had "spots" on his face and lips.

II, 3.—The second husband of II, 2; suffered from recurrent nose-bleeding and had "spots" on his face like his sons. His brother (II, 5) was similarly affected, but no information could be discovered about the brother's children (III, 5 and 6) nor about his other brothers (II, 6, 7, and 8) and his sister (II, 4).

III, 2.—Aged 60 and married but childless; has suffered from nose-bleeding only rarely throughout his life. He is said to have "spots" on his face just like his brothers.

III, 3.—Examined aged 57, a single man; has suffered from severe nose-bleeding since childhood. The spots on his face appeared during the second decade, but have been more prominent during the last ten years. On examination he was very pallid, with multiple telangiectases of about 2 mm. diameter on the lips, several similar lesions on the tongue and soft palate, a few pinhead-sized telangiectases on the cheeks and ears, and diffuse telangiectasia over the tip of his nose.

III, 4.—Aged 55, also single; was said to have had occasional nose-bleeding all his life and spots on his face like his brother (III, 3).

Summary

Three more families with hereditary haemorrhagic telangiectasia from the West Riding are reported. They illustrate again that epistaxis is usual in this condition, that arteriovenous aneurysm of the lung may be present, and that the defect is inherited as a simple dominant.

We are grateful to Mr. P. R. Allison for so kindly placing his records at our disposal, and to Dr. John Savage, Dr. J. P. Peel, Dr. John Rushton, and Dr. Bennet Smallhorn for allowing us to examine their patients.

REFERENCE

Garland, H. G., and Anning, S. T. (1950). *Brit. J. Derm.* In press.

NUTRITIONAL DYSTROPHY AMONG CHILDREN IN MADRAS

BY

S. T. ACHAR, M.D., F.R.C.P.Canada

Professor of Paediatrics, Madras Medical College

Quite a large number of infants and children with oedema of varying degrees not associated with albuminuria have recently been admitted to the children's wards of the General Hospital and of the Women and Children's Hospital, Madras (South India). A good proportion of these cases had skin changes of a peculiar nature and of varying pattern in different patients and even in the same child (Fig. 1). In some, large areas of skin were discoloured, while in other areas the skin was actually peeling off. Cracks and fissures at the flexures were a frequent feature. Large areas of depigmentation with some hyperpigmented areas were often present.

The term "crazy-pavement skin" or "mosaic skin" serves to describe the majority of skin lesions. An occasional case, however, showed a fine scaling with pigmentation characteristic of pellagrous skin, though the distribution of this scaly lesion was not always characteristic of pellagra. Some cases had keratomalacia, while others showed cheilosis, angular stomatitis, and the orogenital syndrome. The common feature of all was a pronounced debility and apparent low vitality. The child lost its liveliness, looked ill (but not the toxic look of an infectious disease), and seemed to be slipping away from life quietly and rather quickly.

For purposes of this review a series of 78 cases that were admitted to the above-mentioned hospitals in 1948 (March–December) have been considered.

Investigation of a Series of Cases

Age Incidence and Social Status.—One child in the series was 8 months old; the rest were distributed as shown in Table I, the majority being 1 and 2 years. Most came from very poor families, though an occasional one was encountered among the middle and even the rich classes.

Dietetic History and Previous Illness.—Almost all the children had been fed very unsatisfactorily, usually because of the absence of cow's milk and milk products but sometimes from ignorance. Their diet consisted mainly of rice, very little of other cereals, and vegetables—apart from the scanty mother's milk the younger children were able to get. More than half gave a history of dysentery or diarrhoea lasting for a while—often recurrent bouts of diarrhoea rather than a single episode—at some time preceding the onset of oedema. In some instances the lack of liveliness and vigour as well as the oedema appeared to follow an attack of diarrhoea or dysentery.

TABLE I.—*Age Incidence and Economic Background*

Age	No.	Social Status	No.
Under 1 year	4 (5%)	Very poor	40
1 and 2 years	48 (62%)	Poor	31
3 „ 4 „	18 (23%)	Middle	6
5 to 10 „	8 (10%)	Rich	1

Clinical Features.—Apart from the oedema and absence of liveliness in all cases, nearly half of them showed varying types of skin changes, with and without alteration in the colour and texture of the hair. Eye changes indicative of vitamin-A deficiency were fairly prevalent, leading in some cases to blindness, and signs of riboflavin deficiency were present in many. There were surprisingly few with clinical evidence of active rickets or scurvy in spite of the poor dietetic history. The main clinical features are shown in Table II.

Course of Illness

The mortality of the untreated cases is very high and the morbidity among those who survive considerable. There were 12 deaths in this series (mostly in the earlier cases, when the technique of repeated blood and concentrated serum transfusions was being developed). Most of these deaths seemed to be attributable to acute parenchymatous hepatic failure, the child sinking into stupor and dying quickly in this stuporous state. Septic complications were common; otitis media, abscesses, pyelitis,

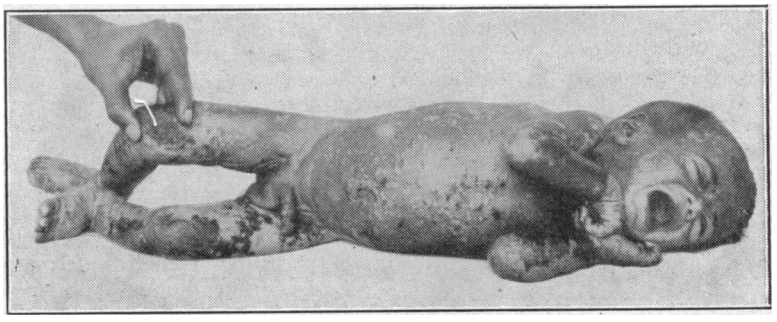


FIG. 1.—Severe dermatoses.

TABLE II.—*Showing the Main Clinical Features in 78 Cases*

Clinical Features	No.
Oedema	78 (100%)
Dermatoses (crazy-pavement or mosaic skin, denuded superficial layers, cracks and fissures at flexures, purpuric areas, depigmented or hyperpigmented areas, scaly skin of pellagra)	36 (47%)
Oral changes (cheilosis, angular stomatitis, glossitis)	35 (45%)
Eye changes (xerosis, keratomalacia, panophthalmitis, corneal opacity)	33 (42%)
Changes in hair (light hair; dry and brittle sparse hair)	40 (51%)
Intestinal infestation with *Ascaris lumbricoides*	70 (90%)
Palpable liver	48 (62%)
Diarrhoea	68 (87%)
Steatorrhoea	8 (10%)
Mucus in stools (in two *C. dysenteriae* was isolated; in one *E. histolytica* was found)	11 (14%)
Previous history of dysentery	28 (36%)
„ „ „ diarrhoea only (no blood or mucus)	22 (28%)

and bronchopneumonia occurred now and then, but in all the 12 fatal cases the child's vitality was at a low ebb on admission, and it seemed to sink into stupor and die quickly without either a febrile reaction due to any septic complication or rapid breathing due to bronchopneumonia. A few children lost their eyesight owing to keratomalacia, especially the earlier cases, in which oral vitamin A was relied upon to combat the keratomalacia and its complications in the eye; but every one of the later cases with the slightest eye change indicative of vitamin-A deficiency was given vitamin A by the intramuscular route, and the pathological process was reversed in many of these. Six of the apparently cured cases were brought back to hospital some time after discharge with a relapse owing to the inadequate diet in their homes.

Treatment

It was found that unmodified cow's milk was badly tolerated by the majority because the food tolerance had broken down owing to previous dysentery or diarrhoea or to the eating of unsuitable food through ignorance or force of circumstances. The problem therefore demanded the greatest paediatric care, and I found that the best results were obtained by giving acidified half-cream milk fortified by casein products, gradually increasing the amount of fat in the milk as the food tolerance improved. Apart from this feeding and the addition of vitamins, with the treatment of infections when present, the main plank in treatment after the initial failures was blood transfusion — repeated if necessary — and/or repeated transfusions of concentrated human serum.

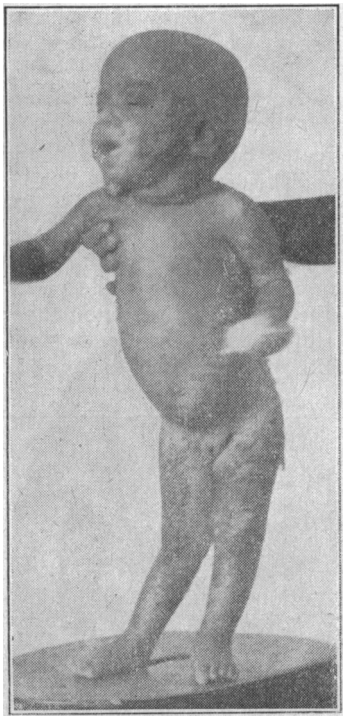


FIG. 2.—Before treatment. Note dermatoses.

This treatment was given because of the disturbed physiology in these children—namely, the low haemoglobin and obvious hypoproteinaemia, and perhaps the inability of the liver to synthesize serum albumin. Suitable veins for repeated transfusions were difficult to find in these oedematous children with skin involvement, but, with experience and the use of the scalp-vein technique where feasible, and the cut-down technique over the saphenous vein at the ankle through the oedematous tissue where the closed methods failed, it was found possible to transfuse repeatedly all those in whom this procedure was indicated. In most of these cases, after the initial blood transfusions (one or often two or three, depending on the haemoglobin level), dried human serum in a concentrated solution was transfused repeatedly.

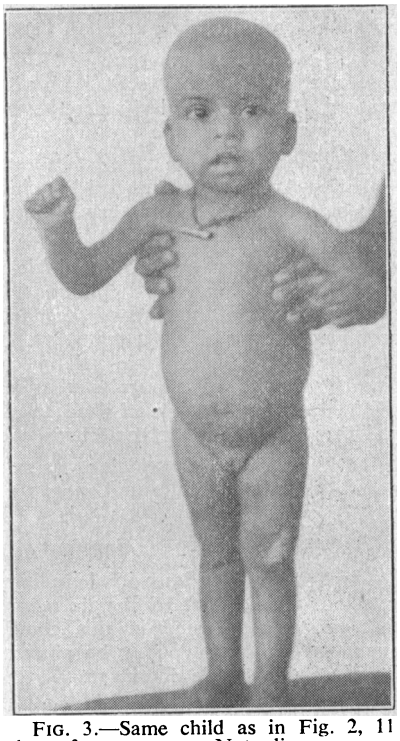


FIG. 3.—Same child as in Fig. 2, 11 days after treatment. Note disappearance of dermatoses.

After a week or two of this therapy there was a dramatic change, the child regaining its liveliness even though the oedema took a long time to disappear. The skin changes receded rapidly (Figs. 2 and 3), whereas under the older therapy of oral high-protein feeding and vitamins the response, if any, was slow and uncertain. Besides this regime, in those cases showing evidence of past or present bacillary dysentery specific treatment was given (sulphonamides followed by mixed dysentery phage). By adopting these methods a much greater percentage of recoveries was obtained in the later cases (only 2 deaths in 50) than in the earlier cases (10 deaths in 28).

Pathological Findings

In most of the fatal cases a piece of the liver was removed at partial necropsy, as permission was not given for post-mortem examination. In two cases, however, it was possible to remove the whole of the liver at partial necropsy. Both livers were large, pale, and obviously fatty. Histological examination of these 12 livers revealed extreme fatty infiltration in nine, fatty infiltration with commencing periportal fibrosis in two, while in one case there was diffuse fibrosis round the lobules.

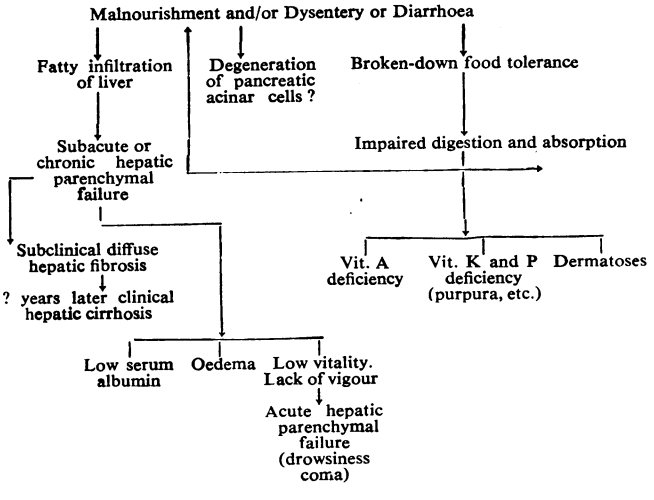
Comment

The skin changes, which were present in about 45% in this series and for which the terms mosaic skin or crazy-pavement skin seem apt in some cases only, cannot be explained easily as pellagrous or as an orogenital syndrome. It is possible that there is some factor in the fats, lack of absorption of which produces the dermatoses. Pottenger (1944) has written on the therapeutic value of a thermolabile factor found in fats, particularly the lecithins, in dermatoses. From the clinical observation that the eye changes do not respond to oral vitamin A, while parenterally the vitamin produces a dramatic change, as well as from the clinical observation that these children do

not tolerate full-cream milk, it is surmised that digestion and absorption of fats and fat-soluble factors is impaired in this condition.

Considerable interest is developing in a similar condition reported from different parts of the world under various names: kwashiorkor (Williams, 1933, 1935, 1940), infantile pellagra (Trowell, 1937, 1940, 1941), malignant malnutrition (Trowell, 1944; Trowell and Muwazi, 1945), and fatty liver disease of infants (Waterlow, 1948). These syndromes seem to be the same disease, and should be described under one common name. In my opinion "infantile pellagra" and "kwashiorkor" are unsuitable, as both the pellagrous condition and the red-boy appearance (in African dialect *kwashiorkor* means "red boy") occur in only a percentage of cases and are neither the essential nor the real trouble. In my series the cases with and without skin changes went downhill in the same manner unless arrested by treatment (which, of course, is not directed to the skin condition in particular). Equally, the term "malignant malnutrition" is inapplicable, as with appropriate treatment the majority recover. "Nutritional dystrophy" seems the best term to employ to describe this condition.

The presence of diarrhoea or a recent history of diarrhoea or dysentery in a majority of my cases is, I think, a finding of considerable importance, but this was not a feature in the series from West Indies reported by Waterlow. The part, if any, played by intestinal infestation with roundworms in the production or maintenance of this condition seems to be a minor one, as marked improvement occurred long before specific treatment was directed towards the infestation. The train of events leading to this condition may be as shown in the accompanying diagram.



It is interesting that diffuse hepatic fibrosis was noted at necropsy in the liver of one child aged only 2 years, who had had diarrhoea for two months and oedema for a fortnight before death. The diet in this instance was, in common with other children in this series, grossly deficient in milk products, the child having been weaned at 3 months on account of the mother's death, and thereafter fed mainly on rice congee with very little milk. The only clinical manifestation of the changes going on in the liver presumably for nearly a year in this case seemed to be a lack of liveliness in the child.

The absence of symptoms and signs of rickets and scurvy in this series, in spite of the diet being grossly deficient in vitamins, was probably due to growth being retarded during the period of gross malnutrition.

Summary

A series of 78 children have been studied. Oedema was present in all of them, dermatoses of various patterns in nearly half, and both eye and mouth changes in less than a half.

The diet was grossly inadequate in cow's milk and milk products in almost all cases.

Diarrhoea past or present was a feature in a large majority.

While in many cases gross signs of vitamin A and B group deficiencies (keratomalacia, angular stomatitis, etc.) were seen, there was, surprisingly, no florid rickets or scurvy.

In the fatal cases the liver showed changes varying from fatty infiltration to diffuse hepatic fibrosis.

Explanation is offered for the common clinical features and skin changes.

Blood and/or concentrated serum transfusions aided by a suitable diet (half-cream acid milk fortified by casein products) proved far superior to other regimes.

I wish to thank my assistants, Drs. S. Damayanti and D. Isac, for their invaluable help, and Lieutenant-Colonel Sangham Lal, Superintendent of the General Hospital, Madras, and Dr. R. K. Thampan, Superintendent of the Women and Children's Hospital, Madras, for permission to publish from their records.

REFERENCES

Pottenger, F. M., jun. (1944). *Sth. med. J.*, **37**, 211.
Trowell, H. C. (1937). *Arch. Dis. Childh.*, **12**, 193.
—— (1940). *Trans. R. Soc. trop. Med. Hyg.*, **34**, 85.
—— (1941). Ibid., **35**, 13.
—— (1944). *Clin. Proc.*, **3**, 381. (Quoted by Trowell and Muwazi, 1945).
—— and Muwazi, E. M. K. (1945). *Trans. R. Soc. trop. Med. Hyg.*, **39**, 229.
Waterlow, J. C. (1948). *Spec. Rep. Ser. med. Res. Coun., Lond.*, No. 263.
Williams, Cecily D. (1933). *Arch. Dis. Childh.*, **8**, 423.
—— (1935). *Lancet*, **2**, 1151.
—— (1940). *Trans. R. Soc. trop. Med. Hyg.*, **34**, 85.

The Teviot Committee on Dentistry recommended the institution of a scheme for the training of oral hygienists on such a scale as would provide an adequate test of their value. After consultation with the Dental Board the Minister of Health has instituted a controlled experimental scheme. Some ex-R.A.F. hygienists will take part, and a limited number of additional hygienists will be trained over a period of some three years. The Minister requested the Standing Dental Advisory Committee to co-operate, and the committee appointed a subcommittee under the chairmanship of Mr. Roper-Hall. Membership of the subcommittee includes nominees of the British Dental Association and of the former Incorporated Dental Society and the Public Dental Service Association, and of the Dental Board, and observers nominated by the Ministries of Health and Education and by the Department of Health for Scotland. Since the work of hygienists is essentially preventive, their training lays special emphasis on the teaching of oral hygiene. Their operative work is limited to the removal of tartar and cleaning of the teeth, and, where directed, the topical application of sodium fluoride. The first course, for 15 pupils, is being held at the Eastman Clinic, and it is proposed that a further course for 15 pupils will start there soon. In the light of experience of these courses other centres may be opened in the provinces. On the completion of training the hygienists will be required to take an examination. Successful candidates will be awarded a certificate of proficiency, and will then be able to accept posts offered by authorities with the Minister's approval in the public dental service, working under the supervision and responsible direction of a registered dental practitioner.