Endemic cretinism describes a syndrome of developmental abnormalities which geographically coincide with severe iodine deficiency.

Although it is now largely a disease of history to the majority living in the twentieth century, small pockets of the disease are still to be found in the more isolated areas of the world.

The first recorded definition of a cretin appeared in 1754 in Diderot's Encyclopedie which described "an imbecile who is deaf, dumb with a goitre hanging down to the waist" and at that time endemic cretinism was known to exist in Switzerland, Southern France and Northern Italy.

Whether or not it is associated with goitre, endemic cretinism is considered a Public Health problem in itself. At the moment affected populations are being studied in South America, the Himalayas, the Congo, West Irian and New Guinea.

The condition of endemic cretinism varies widely in its presenting features and it has been hard to establish criteria accepted as being diagnostic of the disease. Roughly it may be divided into two main syndromes:

1. Hypothyroid.
2. Neurological.

Both syndromes may occur in the same geographical area but within the present population studies the neurological type of endemic cretin predominates in New Guinea, the Himalayas and South America, the hypothyroid type appears to predominate in the Congo.

The author spent 15 months during 1970/71 with the Anglican Mission at Koinambe, a small village within the Jimi River Valley in the Western Highlands of New Guinea. Endemic cretinism of the neurological type is widely present in this region and its study and treatment by physiotherapy are the subject of this paper.

Koinambe was settled by the Anglican Mission in 1965, and anthropometric data on all children of that village have been regularly recorded including accurate recording of their ages.

The country is mountainous and heavily timbered. The village of Koinambe is accessible by light aircraft but surrounding villages can only be reached on foot.

The people live in small village communities in diffusely scattered family units, often having no central point except an area selected for administration purposes by the European Patrol Officers and Mission personnel.

They are subsistence farmers, living mainly on sweet potato supplemented very occasionally by meat, usually in the form of pork eaten on ceremonial occasions or, more recently, tinned meat and fish. They walk daily to their gardens which may be several thousand feet above or below their villages.

From 1969 to 1971, 51 children from the village of Koinambe itself were studied in detail and 10 were thought to be affected by endemic cretinism. Of the surrounding area, a further 139 children appeared to be affected within a total population of approximately 7,000. However, as the disease covers such a wide spectrum of hearing disability, neuromuscular and intellectual impairment, many mild cases were almost certainly missed and it is possible that very severe cases were hidden by the community.

All children examined showed some evidence of malnutrition, widespread in this area, particularly marked in children from six months to three years, in many cases necessitating admission to hospital.

Charts for this population show a generally low birth weight but between affected and non-affected children there is no significant difference in the pattern of weight gain.

In general the population is small, the average height of all mothers measured being 142 cm (4 ft. 8 ins.). However, with the exception of two cases, the affected children studied appeared to be of normal height for their age. In the two exceptions, the juvenile ratio of upper to lower body segments appeared to have persisted, i.e. short lower limbs in relation to total body length, typical of a child with growth retardation due to thyroid hormone insufficiency.

One small boy of five years was examined whose skeletal proportions were average for a child of two and a half years and whose total height was average for a child of four years.

The most evident difference between normal and affected children was in the rate of motor development. In the normal child a delay in the achievement of physical milestones occurred only in the instance of severe malnutrition and disease. After successful treatment, development proceeded at the normal rate. In the affected children a delay in motor development was constant despite possible overlying conditions.
RESERCH INTO THE CAUSES OF ENDEMIC CRETINISM

In view of the geographical link between iodine deficiency, endemic cretinism and, in most affected areas, endemic goitre, the assumption has been made that endemic cretinism is one of the results of iodine deficiency, maternal and/or foetal hypothyroidism being cited as the cause. However, the identification of the true cause of endemic cretinism is still a vexed medical question and not within the scope of this paper beyond noting that iodine deficiency may well be a significant contributing factor.

Certainly insufficient thyroid hormone during foetal development and the early neonatal period may cause irreversible damage to the central nervous system.

It has been proved that the injection of iodinated poppyseed oil will correct an iodine deficiency for four and a half years. Thus in order to establish a significant link between iodine deficiency and endemic cretinism, a controlled trial was started in the Jimi Valley in 1966. Half the population were injected with iodised oil and half with normal saline.

It was found on follow-up Patrols in the three years following that the symptoms of endemic cretinism in children born to saline injected mothers appeared to be greater than in those born to oil injected mothers.

The Jimi trial was discussed and the results examined. In January 1972, at a symposium on endemic cretinism held by the Institute of Human Biology in Goroka it was decided that although the case for treating goitre with iodised oil had now been well established, the trial had not proved conclusively that iodised oil could prevent endemic cretinism. The criteria used in the Jimi trial were considered sufficient only to diagnose severe cases as the number of mild cases born since the commencement of the trial had almost certainly been underestimated.

A second trial could involve a thorough investigation of only 20 pregnancies with a control group of pregnant women who had received the iodised oil.

It was also suggested that existing cretins be treated with thyroid hormone and the subsequent effect on E.C.G. physical and intellectual growth be assessed.

The thyroid status of the mothers of these children should be examined against a control group and if possible, an investigation made into differences in obstetrical history. This latter study would be a difficult project as almost all mothers in the community deliver at home and only come to hospital; a walk of two to eight hours, if labour is obstructed or the placenta retained.

SYMPTOMS

Hypotonia

In the young child there is a generalised hypotonia causing a marked delay (often two to three years) in the ability to lift the head against gravity on ventral and dorsal suspension, or to control the movement of the head when in the vertical position. Cervical muscle weakness persists and is demonstrated in the older child when he flops backwards as he attempts to look up.

Infants and young children demonstrate the "slip through" sign when lifted from the axilla due to lack of tone in the shoulder girdle muscles.

When the child eventually learns to sit, he does this with the support of his outstretched arms and with kyphosis of the whole spine.

Movements of intention in the upper limb persistently lack precision, and the ability to pick up small objects by opposition of the thumb and index finger is much delayed.

Gait

The pattern of gait and stance is similar to that demonstrated by the Duchenne type of Muscular Dystrophy. In standing the knees are semiflexed and there is some adduction, flexion and internal rotation at the hips with a compensatory lumbar lordosis to bring the centre of gravity far enough forward to maintain balance.

In the early stages the gait is unstable and shuffling and the child has difficulty in negotiating a shallow step. Even as balance improves the features described above persist.

Reflexes

Despite generalised hypotonia, stretch reflexes were hyperactive in all affected children and in most children spasticity eventually developed, more marked in the lower than in the upper limb. The age at which this occurs depends on the severity of the condition in any one child.

In a very severe case, spasticity of the lower limb may develop as early as six months, usually most marked in the hip adductors. Most affected children develop some spastic signs by the age of six years.

There appeared to be a progressive improvement in all children, some to the extent of normal motor function, although the majority suffered residual disability to varying degrees.

Hearing

Hearing in the Jimi child was assessed by observation of his reaction to noise and the spoken command and on the mother's observations of the child at home.

This assessment was obviously inadequate but there was still evidence to suggest that the hearing of many of these children gradually improved, and with hearing, the development of speech.

Vision

Squint was the obvious visual disturbance, although two cases of nystagmus were noted in the Jimi group.

Intellectual Ability

Although it is particularly difficult to assess the intellectual ability of deaf children who are delayed in their motor development and with whom one does not share a common language, there was obvious...
mental impairment in some cases. However, from observations made during play sessions it appeared that the majority of affected children fell within the normal range of intelligence. The only marked difference between the affected child and the normal was in his manipulative ability.

Like the rest of the musculature, the facial muscles of these children are hypotonic, allowing the lower jaw to sag, which, combined with the characteristic squint, gives the child a somewhat imbecilic expression.

**TREATMENT AT KOINAMBE**

In the absence of medical treatment for the already affected children and in an effort to reduce their symptoms, a project of physiotherapy was started at Koinambe in May 1970.

The author made patrols into the surrounding mountains to try to document and examine all the affected children. One hundred and forty-nine children were found whose disabilities were obvious but doubtless, many milder cases were overlooked.

Clinics were held in a pre-determined area. They involved the whole village and the crowded situation was generally unsuitable to establish a detailed assessment of hearing, intellectual and motor ability. The shortcomings of the locale were matched by those of communication, as interrogation of the parents was invariably through an interpreter and many details were missed or suppressed.

Where a case was identified it was suggested to the parents that they carry their child to Koinambe for a programme of intensive physiotherapy. Understandably this suggestion was not well received, since it involved carrying a heavy child, food and firewood for their stay in hospital and for many this meant a journey of two days. It meant also that the mother would be absent from home for some time and as it was she who was largely responsible for the upkeep of the garden, the family she left behind might well go hungry.

The medical status of Koinambe requires description. The hospital falls under the Public Health Department category of "Medical Aid Post" and during 1970/71 was run by one Australian trained nurse with assistance from the author and the wife of the Missionary in charge (also a trained nurse). The rest of the medical "team" was formed by local boys who were taught to apply dressings and give injections.

The main hospital building was described as "permanent" in that it had a concrete floor and tin roof, though the walls were of woven bamboo. It was in this building that medical treatments were administered.

The "ward" was of native materials with an earth floor on which three or four cooking fires burned constantly. This housed the patients, their relatives, food, firewood, piglets and cassowary chicks. The Physiotherapy Department was the grass between the two buildings.

In view of the shortage of trained staff, physiotherapy was limited to those children not yet walking. A normal intake for a period of three weeks was between 16 and 20 children.

Initially, simple equipment was constructed in the form of parallel equipment, a standing table and a walking pusher made from an old pram. As finance became available two more walking pushers were added, rubber balls, a baby bouncer, paddling pool, and several of the kiddicraft constructional toys.

For treatment purposes the children were divided into groups roughly according to neurological age. Group I consisted of children not yet capable of sitting; Group II, children sitting unsupported or with the support of their arms; Group III, those children who were incapable of walking but who readily pulled themselves up to standing and "coasted" round furniture.

Within each group part of the day was spent on the child's own particular problems with the full participation of his mother. The remaining time was occupied by general activities such as play with water, paint, paste and paper and toys appropriate to his age.

It was important to stress to the mother that she was also an integral part of the group as her education in coping with her child formed a particularly vital aspect of the treatment.

These children had to learn consciously those skills which would come naturally to the normal child and the education and stimulation of their children was not a recognised function of these mothers. As there was no knowing when they would be prepared to bring their children to Koinambe for further treatment, or how long they would be prepared to stay once there, a continuation of treatment in the home situation was particularly important.

The aims of treatment were to educate the mother and the rest of the family in the correct stimulation of the child and to show her the most suitable activities for that stage in development; to show the mother how to guide her child's play during therapy sessions; to provide the child with a range of activities consistent with his ability; to encourage mobility and manipulation and thus increase the intensity and duration of sensory stimulation essential for his neurological organisation; to retrace the steps in the normal neurological development and use activities and sensory inputs that have been effective in promoting neurological organisation from early infancy on; and, finally, to provide the child with an adequate diet and to ensure that this was observed at home.

**Treatment of Group I**

It was important to stress to the mothers of this group that the children were not yet ready to be on their feet and that the emphasis at this stage was to improve postural tone, that is, extensor activity, in preparation for sitting. It was also considered important to develop the use of the arms in the initiation of rolling as a means of support. It was felt that the use of the head as the key point in the education of rolling would be dangerous in unskilled hands.

Much of the treatment was therefore carried out with the child in prone, an unpopular position

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amongst these mothers who held the firm belief that the prone position encouraged the development of "crooked legs". The children of this culture, normal and abnormal, are carried for the first two years of life in a string bag suspended from the mother's head, thus much of their day is spent flexed in the supine position.

Once the prone position had been accepted, the mothers were encouraged to facilitate the extensor response of the child by light stroking along the spinal extensors and by stimulating the child to lift his head to watch some movement of hands, leaves, mother's beads or mobile toys. Auditory stimulation was usually ineffective in view of the pathology.

Rolling was encouraged from the supine position and was facilitated by the child reaching with the arms for objects of interest. The movement was assisted at first and repeated many times until achieved actively.

If head control was adequate, but before unsupported sitting was achieved, the child participated in group treatment amongst neurologically older children, supported in an adapted chair or walking frame. This way he was further stimulated by competition to use his arms to reach the object of his choice. Supported, he was able to transfer his weight without the fear of falling and at the same time facilitate equilibrium reactions.

Specific treatment for sitting followed similar lines. The child was supported manually, generally at the hips. Arm and head movement, with gentle weight transference, was encouraged and support gradually withdrawn as this was achieved.

**Treatment of Groups II and III**

Children in this group were encouraged to play in the kneeling and standing positions thus facilitating as far as possible their righting and equilibrium reactions.

Play in the group situation was stressed to encourage interaction between the children, and as it became obvious to them that they were a focus of attention, this in itself was an added and powerful stimulus to activity.

For those already confident in standing with support, parallel bars were constructed. These were of great value and the boys in particular would play between them for long periods kicking a football to one another.

Mothers were shown how to give minimal assistance to their children in the use of the walking pushers. They would enjoy the novelty of pulling this almost as much as the children who, as a result, could then play a purely passive role in the activity.

Sticks were tried but on the whole were unsuccessful. Certainly sticks held less appeal to the children than the brightly coloured toys on wheels and in any case, at this stage the children found crawling the most satisfactory method of locomotion.

Mothers were encouraged to "walk" their children over increasing distances with one or both hands held. This required some instruction. Groups of mothers would set out together on a social walk and soon the child would be suspended by the hands, unnoticed, with toes barely touching the ground.

**Assessment**

With differences in chronological age achievement the development of these children followed the same course through motor perceptual and intellectual levels, as that of the normal child.

It was difficult to decide in this context what constituted the "normal" chronological limit for each stage. European standards such as those laid down by Sheridan, bore little relation to the normal child of the Jini Valley whose cultural and geographical background, nutritional state and psychological influences are so vastly different.

The most practical means of determining progress was to measure the patient against himself and to compare the change in neurological age with the actual time which elapsed since the commencement of treatment. Mary Sheridan's standard from the Developmental Progress of Infants and Young Children was used as a guide and reference.

Despite difficulties, results were encouraging and often well beyond expectation. Of the 75 children treated, who at the start of the programme were unable to walk or to communicate and were unable to sit without arm support or were unable to sit at all, 10 are now walking unaided, 30 are now sitting unsupported and pulling themselves up to standing. The maximum period of treatment for any child was 12 weeks, a period often broken by bouts of malaria, dysentery and other infections.

In many of these children the dramatic response to treatment could well be attributed to the greatly increased environmental stimulation provided by physiotherapy.

A disabled child of this community, although not wholly rejected is left for a large part of the day on his own. A child with severe endemic cretinism would be unable to move or to communicate and would suffer distorted vision and hearing.

It was not surprising that with social interaction and stimulation through physiotherapy that the latent powers of recovery in these children were quickly developed.

An appeal is made for volunteers to continue in this exciting and intensely rewarding field.

**Reference**

SHERIDAN, M. D., *The Developmental Progress of Infants and Young Children*. Modified from Ministry of Health: Reports on Public Health and Medical Subjects No. 102.