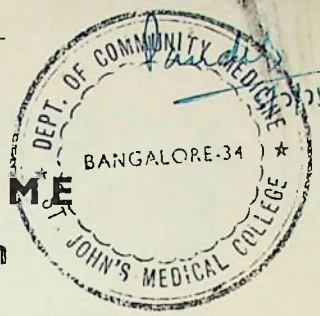

HANDIGODU SYNDROME

A Peculiar Orthopaedic Problem

In the State of Karnataka (old Mysore State) in South India a peculiar orthopaedic problem, hitherto unreported, has come to light in the last two months. The illness tends to affect the people in the Malnad area of the State which is situated along the Western Ghats and mostly at an altitude of 2000-3000 feet above sea level with an annual average rainfall of 80-120 inches. This region is thick in forest wealth and certain areas have classical tropical rain forest. Most of the people in this region are agricultural labourers and the main products include rice, arecanut (*Areca catechu*), sandal wood, timber and in some areas coffee. The temparature varies from the upper nineties (Farenheit) in summer to the lower seventies in winter. The humidity is quite high.

The State of Karnataka has an area of about 120,000 sq. miles and its population according to the latest census in 1971 is about 29 million. As in the rest of the country about 85% of the population are rural. About 85% of the population are Hindus. Amongst Hindus there is a group of people who are referred to as Harijans. They are mainly agricultural labourers performing many of the manual tasks. Though living in the same village, their living quarters are separate from those of the main community. They live in fairly clean surroundings and the source of water supply to their colony is from a well, which is different from the main well for the rest of the population. Some of them own cows, but they tend to sell rather than consume the milk. It is worth emphasizing that the villages are surrounded by lush vegetation which is hardly few feet from the residential quarters of these individuals. None of the villages have an efficient drainage system, but still the surroundings are kept clean, since the village folk tend to go into the forest for their excretory activities.

Joint family system is still very much in vogue in the villages. The family income is made up of that from not only the father, but also other men and women folk, and sometimes even children.

It is interesting that the same area in Malnad has been of considerable medical interest in the last two decades because of the discovery of Kyasanur Forest Disease. This is a haemorrhagic encephalitis caused by the KFD virus, which is serologically akin to the Russian Spring Summer encephalitic group of viruses. The main reservoir of the infection are monkeys in the forest, wherein an epizootic generally tends to precede an epidemic. Transmission to man is by tick bite and hence KFD virus is an arthropod-borne virus infection. Annually anywhere from 30–180 or even 200 cases are recorded even today in this area and in whom KFD virus has been isolated from the blood. This background information is felt to be useful in considering the new syndrome.

In India, administratively each State consists of several districts and these in turn have several taluks. The first cases were reported from Handigodu Village in Sagar Taluk of Shimoga District. In January 1975, 4 patients from Handigodu village were sent to the local hospital stating that they were unable to walk and within the next week there were about 30 cases, all of them from the same village. Initially, these were thought to be paralytic in type and hence a neurologist was asked to take a team and examine these cases. Accordingly, after a preliminary study, a team consisting of neurologists and an orthopaedic surgeon went to Sagar Town (5 miles from Handigpdu Village) and examined as many cases as possible along with controls. During the course of the examination, it became obvious that these patients were not suffering from any primary neurological disorder and that the problem was mainly in the field of orthopaedics. The clinical and investigatory findings of this illness, which appears to be rather unique, form the subject of this report. It is suggested that since such cases do not appear to have been described in the literature before, this illness may be termed as Handigodu Syndrome after the village of that name wherein they were reported first.

About 130 cases have been recorded so far from Sagar Taluk alone in the last 3 months and 20 additional cases from the neighbouring Thirthahalli Taluk of Shimoga District. An additional 60 cases have been reported in the last six weeks from the adjoining district of Malnad – viz., Chikkamagalur. Cases reported so far are all from the rural population.

This report is based on 45 patients, all of them from Sagar Taluk and who form the nucleus of the first few cases reported. This study, it must be emphasived, is based only on those admitted to Sagar Hospital and is by no means an epidemiological study in its strict sense.

For purposes of comparison a group of thirteen controls living in the same environment as the patients were available for study. The controls could not be matched for age and sex. However, this draw-back is being rectified, in that attempts are being made to collect more data to evolve matched controls. Two controls had a severe degree of anaemia and another one was suffering from acute bronchitis. The remaining controls had no symptoms or signs of any disease. It must be emphasized that *none of the controls had a clinical picture of the main disease under consideration*. Eight patients (62%) of the controls were harijans. Seven of the control group had cases of Handigodu Syndrome in their family : 4 of these were harijans and other 3 non-harijans.

Dietetic History

The staple diet of both the patients and the control group, all of whom were mainly agricultural labourers and belong to low socio-economic strata of society (daily per capita income about 20 US cents), was basically the same. Rice

was the staple diet which was consumed 2-3 times a day along with tamarind (*Tamarindus indica*) soup. Monkey fruit (*Artocarpus lakoocha*) soup was also prepared equally by the patients and the control groups. While no detailed dietary survey was made, available information did not show any significant difference in the frequency of consumption of pulses, milk and milk products, eggs, fish, meat or vegetables in the patient population compared to the controls. Red gram (*Cajanus cajan*) soup and small bits of dried fish were consumed twice a week. Green leafy vegetables were eaten only once or twice a month. Eggs or meat were taken hardly 3 or 4 times an year, and milk and milk products practically never. The diet would thus appear to be grossly deficient in proteins and calcium in both the groups. The most commonly consumed vegetable was tuber-ordinary yam (*Typhonium trilobatum*). It is well-known that this product has a very high cyanide content. However, during preparation this is cut into small pieces and soaked for a considerable time in water during which process the cyanide tends to get washed away. Kesari dhal (*Lathyrus sativus*) was not consciously eaten and even then never formed a staple part of the diet. It must, however, be stated that considering the wild flora in the region abutting on the backyard of the house, as it were, one can never be assured of the type of articles that are consumed in the raw state. It is reported that most of the village folk, particularly the harijan community, have a habit of consuming illicit country liquor, which is shared along with women folk and sometime the children also. It is said that the country liquor is a brew made by fermenting jaggery (unrefined sugar) with ammonium sulphate and discarded lead sheets from car batteries. However, reliable information on the frequency of consumption of illicit liquor is notoriously difficult to obtain.

History

This series was made up of 18 males and 27 females (40% and 60% respectively). Just under two-thirds of the patients were in the first two decades of life and just under a quarter were aged below 10 years. The youngest patient was aged 5 and the oldest 45 years. A note of caution is necessary, particularly with regard to the older age groups, in that recorded dates of birth are practically unknown in this group of people, Seventy percent of the patients were Harijans. Sixty percent were from Handigodu village and another 16% from Adderi village, harijans from the former and mainly non-harijans from the latter in the hospital sample. These 45 patients came from 21 families (family means those who live under the same roof and share a common kitchen), all except six of them having more than one case per family. History of similar illness in the previous generations was denied.

The age at onset of symptoms varied from 4-41 years, over three-quarters being in the first two decades. The duration of symptoms, as reported by the patients, ranged from 3 weeks to over 4 years. About a quarter and one-fifth of

the group had a duration of symptoms varying between six months to one year and one year to one and half years respectively. Seven patients (16%) gave a history of fever either at or within one month before the onset of their other symptoms, which was insidious in 44 of the 45 subjects. The lone exception was one subject, in whom the peak disability was reached within 48 hours.

The main symptom was pain. It was often referred to as a sudden catching type of pain either in one of the joints of the lower limbs or the back and which continued to progress in intensity. This type of pain affected the hips first in 50% of the patients, while in the rest it was mainly the knee joints which were affected first. Pain in the knees was complained of at some stage or other of the illness by all the patients in the group, followed by pain in the hips (82%) ankles (33%) and shoulders (22%). Nine individuals (20%) had low back-ache and stray individuals complained of pain in the elbows or the wrists. Thirteen subjects gave a history of swelling of the joints. The knee joints were affected in all the instances, while in 5 of these, the ankles were also involved. There was no positive correlation between a history of febrile episode at the onset on the one hand and a history of swelling of the joints on the other.

Apart from the febrile episode there were no other antecedent symptoms complained of by the patients.

Physical Examination

The general body build of these patients did not differ significantly from that of the controls. In general, they were shorter and weighed less compared to the more affluent sections in this part of the country. However, the age and sex adjusted heights and weights did not show any significant difference in the patient group compared to the controls. Mucocutaneous signs of vitamin deficiency were an exception rather than the rule (only 3 cases) and were mostly those of vitamin A deficiency in the form of xerosis of the conjunctivae. Mucocutaneous signs of vitamin B complex deficiency were present in only one instance. Anaemia was noted clinically in both the patients and control groups with no significant difference between themselves (2 patients with anaemia in the control group being excluded for this analysis).

Detailed general and neurological assessment was within normal limits.

The findings on clinical examination were confined essentially to the bones and joints. Though all patients complained of pain in the joints, clinical examination showed objective evidence of involvement of the joints in only 32 patients (71%). The most frequent involvement was in the hip joints (30 patients ; 67%), followed by the knee joints (10 patients ; 22%), and shoulders (3 patients ; 7%) ; ankles or wrists were involved in one instance each. Spinal movements were uniformly normal and painless in all the patients, low backache in some notwithstanding.

The findings on examination of the joints were arbitrarily graded as normal, mild, moderate and severe, the criteria for which were as follows :—

Normal	:	Full range of movements without pain.
Mild	:	75% or more of normal range of movement possible with or without pain on movement of the joint.
Moderate	:	50 $\angle$ 75% of normal range of movement possible with or without pain on movement of the joint, with fixed deformities of joints.
Severe	:	$\angle$ 50% of normal range of movement with or without pain on movement of the joint, with fixed deformities of joints.

In general the frequency and severity of involvement of the hip joints were more compared to the rest, the knee joints coming way behind as second in the list. There was severe involvement of the hips in 9 instances (20%), but that of the knees in only 2 patients (4%). Severe involvement of other joints was not noted in this series. Objective findings in the hip joints were significantly more frequent when the symptoms were beyond six months in duration. In view of the small numbers, for purposes of statistical analysis, age at onset was arbitrarily divided as below 14 years and 14 years and above ; similarly duration of symptoms were grouped as below 6 months and 6 months and above. Chi square type of analysis or the 't' test was resorted to throughout and significance expressed at 95% level of confidence.

Eleven out of 45 patients (24%) had definite evidence of swelling of their joints, 9 of these involving the knee joints and one each the ankle and wrist. There was no correlation between age at onset, duration of symptoms or fever at onset on the one hand and the swelling of joints as complained of by the patients and/or elicited by physical examination on the other. Objective evidence of spasm of the flexors and adductors of the hip joints and flexors of the knee joints was noted in 25 (57%), 24 (53%) and 13 (33%) instances respectively. There was a significant positive correlation between the duration of symptoms and spasm of the muscles referred to above. Thinning of quadriceps muscles, particularly vastus medialis, was noted in 12 out of 42 (28.5%) instances. Unfortunately, in 3 instances information on this aspect was not recorded. This thinning was almost always mild. This could well be a result of disuse since there was no corresponding demonstrable weakness of the muscle, but there was no correlation between the presence of objective evidence of involvement of the knee on the one hand and thinning of the quadriceps on the other. Likewise, there was no correlation between the age at onset, duration of symptoms or fever at onset on the one hand and thinning of the quadriceps on the other.

Fixed deformities were noted in the hip joints in 19 instances (42%) and of the knees in 2 patients (4%). There was again a significant positive correlation between the presence of fixed deformities in the hip and the duration of symptoms.

It may be pertinent to point out that though all patients complained of pain in the knees, objective involvement of the knees was present in only 10 instances (22%). It is very likely that the pain in the knees was more often due to its being referred pain from the hips.

Investigations

Four patients refused haemotalogical investigations. Non-significant differences between the patients and the controls included the following: haemoglobin, peripheral eosinophilia, ESR, random blood sugar, blood urea, serum electrolytes (Na, K, Mg), total serum proteins, total serum Ca, serum inorganic P and serum alkaline phosphatase. Those instances wherein the values were beyond the normal range are indicated below.

An attempt was also made to look for any difference in the ranges for values of various estimations. Haemoglobin of less than 13 gms/100 ml was noted in 27% of the patients and 36% of controls - not significant. Though an absolute eosinophil count could not be carried out, admittedly a rough estimate of the same was arrived at from the total and differential leucocyte counts. Peripheral eosinophilia of over 1000 cells per cubic mm was noted in 73% of patients and 38% of controls - not significant. Toxic granules in polymorphonuclear leucocytes were significantly more frequent (54% as against 12%) in the controls compared to the patients. Erythrocyte sedimentation rate (Westergren method) was over 20 mm at the end of one hour in 52% of the patients and 60% of controls, the difference being statistically not significant. Serum electrophoretic fractionation was normal in 75% and 85% of the patient and control groups. The abnormalities noted in both the groups were very mild and did not show any significant difference between themselves. Eighty-five per cent of the patients and 60% of the controls showed hypocalcemia (below 9 mg/100 ml) with no significant difference within themselves.

Tests of liver function like serum bilurin, Vanden Bergh reaction and thymol turbidity were normal in all except one patient, who showed evidence of mild liver dysfunction. These were normal in all the controls. Serum alkaline phosphatase was above the normal range (5–13 K.A. units) in all the subjects in the patient and control groups. The values ranged between 16-26 K.A. units in the patients and 16-25 KA units in the controls.

Urinalysis, available in all showed mild albuminuria in 3 patients, but none of the controls. Blood VDRL was positive in a dilution of 1 in 8 or above in 3 patients, two of them being husband and wife, but none in controls.

C-reactive protein was positive in 21 out of 41 instances (51%) while the Rose-Waaler test, which was available in an equal number, was positive in 14 instances (35%). C-reactive protein was significantly more frequently positive in those with

peripheral eosinophil count below 1000 than those above. Otherwise, there was no correlation amongst the eosinophil count, ESR, C-reactive protein and Rose-Waaler test among themselves.

Cerebrospinal fluid was obtained from the lumbar theca in 10 instances and the findings normal in 9. CSF VDRL was negative in all. One sample was the result of a traumatic tap. Electrophoretic fractionation of the CSF was done in 8 instances with completely normal findings. Lumbar puncture was not done in the controls.

Electrocardiogram was available in 44 patients and all the 13 controls. In the former group 6 patients, 4 of them aged below 10 years and two aged 12 and 15 years respectively, showed inverted T waves with peaking in V_1 through V_3 or even V_4 leads associated with ST segment changes, suggestive of a possible ischemic cardio-myopathy. The EKG was normal in all the 13 controls.

Electromyographic studies, available in 36 patients and 7 controls, were within normal limits in all. Motor nerve conduction velocity was determined in 34 patients and 7 controls. The findings were in the normal range and were not significantly different between the two groups.

Radiology

Considering that the patients were admitted in a peripheral hospital with extremely limited radiological facilities, one could not carry out as complete a radiological examination as one would have wished. Four patients refused any type of radiological investigation. In all but one amongst the rest, the following radiological examinations were carried out:

1. X-ray of the chest AP or PA view.
2. X-ray of the pelvis and the hip joints AP view.
3. X-ray of the lumbar spine AP and lateral views.
4. X-ray of the knee joints AP and lateral views.
5. X-ray of the wrists AP view.

One patient allowed only X-ray of the chest, hip, knee and wrist. X-ray of the shoulders as seen in the chest films were available in 37 instances. Four patients had X-rays of the ankles, since they had pain referrable to that region.

X-rays of the chest were normal in all but one, who showed evidence of bronchiectasis in the right lung. X-rays of the bones, especially of the limbs, tended to show varying degrees of demineralisation of the bones, in some instances fairly severe, and varying in intensity from one bone to another in the same individual.

The most characteristic, significant and diagnostic radiological features were seen in X-rays of the pelvis, wherein the components of the hip joint were chiefly involved. The changes in the hip joints are described below. Premature

closure, of varying degrees, of the epiphyseal lines of the femoral head, greater and lesser trochanters was a constant feature in all patients aged below 16 years. Reduction in cartilage space of the joint, irregularity of the articulating surfaces, sub-chondral bony sclerosis, cystic areas surrounded by sclerotic bone - these were seen individually in over three-quarters of the patients. Height of the femoral head appeared diminished in about 70% of the patients. Other changes, but less common, were marginal lipping even in young adults, Coxa vara, features suggestive of epiphyseolysis of varying grades and widening of the symphysis pubis. Nine patients showed radiological changes akin to Perthe's disease consisting of mushrooming and flattening of the femoral heads. In extreme cases of hip joint disease, there was complete loss of articular space, destruction of the normal shape of the femoral head, which in some instances was shifted from its normal relationship with the acetabulum.

X-rays of the knee joints showed qualitative changes similar to those in the hips, but of varying intensity. They consisted of varying degrees of premature closure of epiphyseal lines of the bones of the joints in patients below the age of 16 years and diminution of cartilage space, irregularities of joint surface, cystic areas in the articulating ends and osteophytosis. Excavations and marginal sclerosis were seen in the patellae in nearly three-quarters of the cases, while one patient showed changes in the knee joints akin to Perthe's disease of the hips.

X-rays of the lumbar spine showed evidence of interference with ossification of the apophyseal nucleus of the vertebral bodies - platy-spondylia and in older age groups, marginal lipping. These were noted in nearly three-quarters of the cases. X-rays of the ankle joints, available in 4, showed flattening of the body of the talus in two. In the wrists, in addition to varying degrees of diminished articular cartilage space, the other changes were of the nature of osteochondritic lesions, particularly in the lunate bone. The most pronounced feature of the X-rays of the shoulder was irregularities of the articular surface and cystic changes, particularly in the head of the humerus. It is interesting that three subjects showed evidence of diaphyseal aclasis involving either the femur or humerus or both.

For the purpose of this study, the radiological findings in the joints (spine excluded) were graded as normal, mild, moderate, severe and gross, the criteria for which are enumerated below:

Normal	:	None of the below
Mild	:	Narrowing of joint spaces
Moderate	:	Narrowing of joint spaces and minimal bone changes - cystic areas, sclerosis, osteophytic lipping.
Severe	:	Gross degree of diminution of joint spaces and/or irregular articular surfaces and marginal sclerosis.
Gross	:	Complete loss of joint space and/or destruction of the articular ends with or without displacement.

Distribution of severity of the radiological lesions in the various joints are given below :

	Hips	*Knees*	*Ankles*	*Wrists*	*Shoulders*
Normal	0	3	1	13	21
Mild	6	10	1	11	7
Moderate	5	18	1	14	7
Severe	21	9	1	2	2
Gross	9	1	0	0	0
Total ...	41	41	4	40	37

It was uniformly observed that the radiological changes were always well in advance of the clinical signs and symptoms. Similary, though clinical examination of the spine was normal, radiological changes of the lumbar vertebrae were fairly marked.

Three of the controls aged 15, 20 and 26 years, showed certain radiological changes in the hip joints similar to those observed in the patients. These occurred in the form of reduction of joint space, irregularity of the articular surface, subchondral bony sclerosis, cystic areas, osteophytosis and appearance suggestive of epiphyseolysis and in one instance akin to Perthe's disease. Similar changes, but of lesser degree, were seen in the knee joints in all these 3 instances, who also showed excavations and marginal sclerosis in the patellae. Appearance of platyspondylia of the lumbar spine was noted in two of these above 3 controls. The other 10 control subjects did not show any radiological changes similar to those observed in the various patients. X-ray of the chest in one control showed pneumonic consolidation.

Activities of Daily Living

The disability of the patients was arbitrarily graded in terms of pain as a symptom, findings on examination of the joints and activities of daily living. The last was graded as full, mild, moderate, severe and gross, the criteria for which are given below :

Full : Can walk any distance and do any work to which the patient is normally accustomed. (N=5)

Mild : Walking restricted because of pain, but can walk a shorter distance at normal pace without support and can squat, get up or bend without help and freely. (N=11)

Moderate : Walking restricted and pace slower, but without support and/or can squat, get up and bend without support but with difficulty. (N=17)

Severe : Walking restricted and needs support for walking and/or needs help for squatting, getting up or bending. (N=7)

Gross : Cannot walk except a few paces, that too with support and/or cannot stand. (N=5)

Disability

Absent	:	None of the below (N = 0)
Mild	:	Pain present; examination of joints show mild abnormalities; ADL full or mild. (N = 17)
Moderate	:	Pain present; moderate abnormalities in the joint on examination; ADL mild or moderate. (N = 16)
Severe	:	Pain present; severe abnormalities on examination of the joints; ADL severe or gross. (N =12)

In general there was a positive correlation between the degree of involvement of ADL and disability on the one hand and duration of symptoms, frequency and severity of changes on examination of the hip joint, presence of flexor and/or adductor spasms of the hips and flexor spasm of the knees.

Treatment

Patients whose disability had been rated as mild have been advised analgesics and physiotherapy to the involved joints along with short-wave diathermy. Cases coming under the category of moderate disability are being given, in addition, prednisolone in declining doses for a period of six weeks. Since this therapeutic regime has just been started, it is too early to assess the results.

Cases listed as having severe disability have been chosen for surgical correction. Indeed four of them have had orthopaedic corrective procedures, which consisted of release of soft tissue contractures at the hip joints in two patients, and the flexors released from their attachment. Hip joint capsule was released from its superior attachment so as to get complete correction of the flexion at the hip, confirmed by Thomas' Test on the operating table. When the hip joint was opened to release the contracture, the opportunity was utilised to take out specimens of capsule, synovial membrane, articular cartilage, and the metaphyseal bone along with the deeper cancellous bone for histopathological as well as virological studies. Incidentally, the gluteus medius muscle was also taken for biopsy. Subsequent to the operative procedure, these patients were put on skin traction - to relieve the pain and to retain the correction obtained.

Examination of the exposed hip joint revealed a greyish pale synovial membrane and glistening white articular cartilage, both on the acetabular side, as well as on the femoral head. Synovial fluid was thin and clear and was not significantly increased in quantity.

One patient had biopsy of the structures of the knee joint in addition to aspiration of the knee joint fluid. The synovial membrane did not appear congested nor was it thickened. The articular cartilage appeared white and glistening. In another patient the periosteum over the lower end of the shaft of the femur along with a block of bone consisting of the metaphysis, epiphyseal line

and part of the adjoining epiphysis was taken out. These were normal macroscopically. Biopsy of the vastus medialis was also carried out in 3 of the 4 cases.

CSF from 4 patients, synovial fluid from the left knee joint in one instance and biopsy material from cartilage, synovial membrane, femur and its periosteum along with gluteus medius and vastus medialis from one patient each were submitted for bacteriological culture for aerobic and anaerobic organisms and fungi, but with negative results. Culture for M. tuberculosis was also negative.

Sera from 41 patients and all the controls, spinal fluid from 10 patients, synovial fluid from the left knee joint in one instance and biopsy material including cartilage, bone, muscle and synovial membrane from 4 subjects have been sent to Virus Research Centre, Poona, for virological studies with special reference to arboviruses, and the results awaited. Similarly, the biopsy material from 4 patients who were operated upon have been sent for histopathological examination to the Indian Registry of Pathology, New Delhi, and the results awaited.

Future Work

More controls are in the process of being collected, so as to match them with the patient population with respect to age and sex. Follow-up evaluation of these subjects will be carried out as far as possible, but the great limiting factor is the distance of nearly 220 miles between Sagar Town and Bangalore. More cases in the severe grade of disability would be submitted for orthopaedic corrective procedures, during which process biopsies of the appropriate tissues will be obtained.

Attempts are being made to examine the cooked food from the patients and controls for the presence of any toxic products, particularly cyanogens and nitrile compounds. Samples of urine would be collected from as many patients and controls as possible and analysed for porphobilinogen, thiocyanates and nitrile compounds.

Addendum :—Since writing up this report, the total series is now made up of 52 patients and 51 controls matched for age, sex, community and environment. Three more patients have been operated. Data are being analysed.

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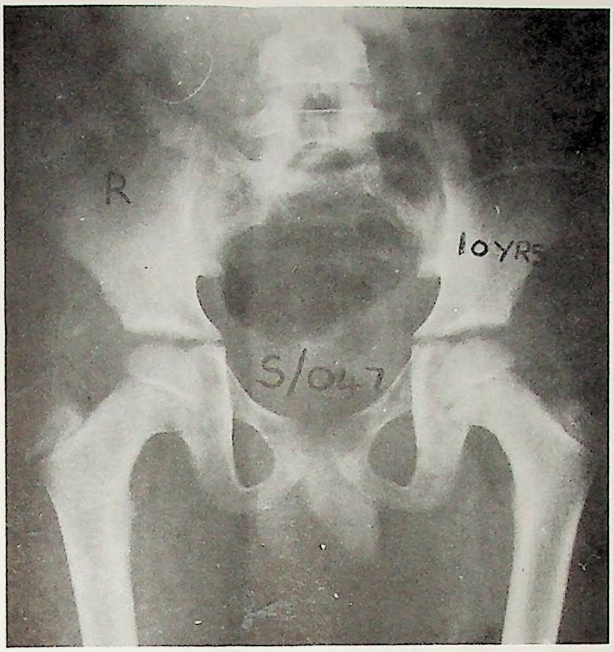

Fig 1. Normal hip joints

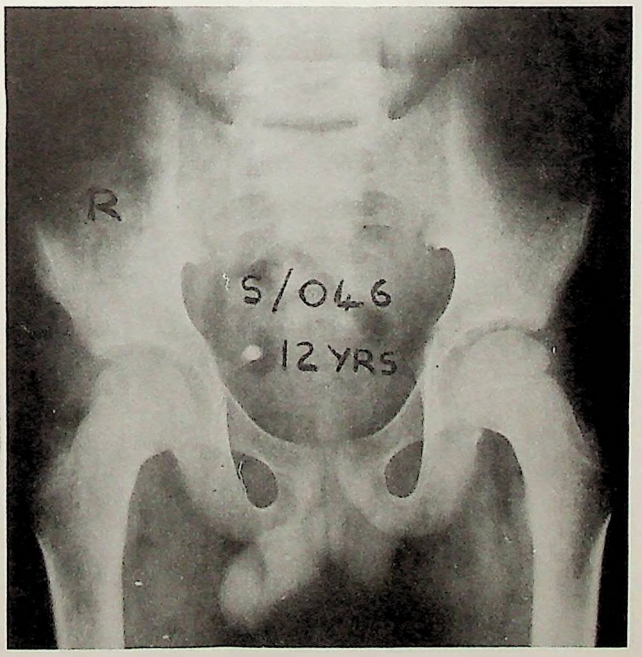

Fig 2. Normal hip joints

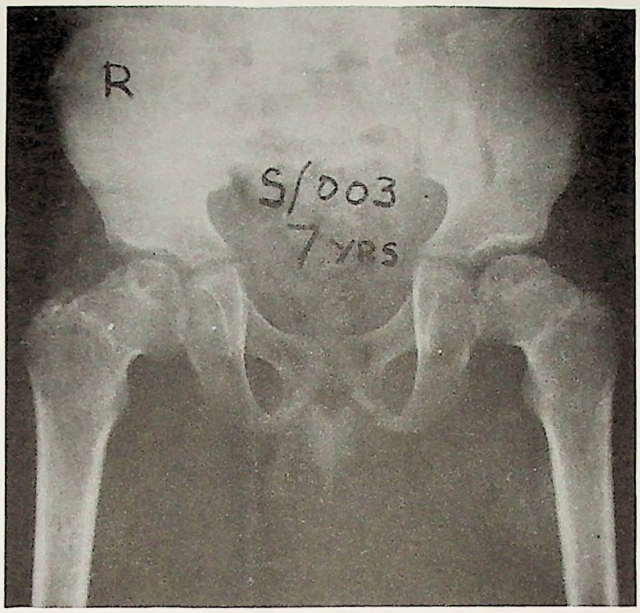

Fig 3. Hip joints - Moderate changes. Note premature closure of the epiphyseal lines, narrowed joint space and cystic areas with marginal sclerosis

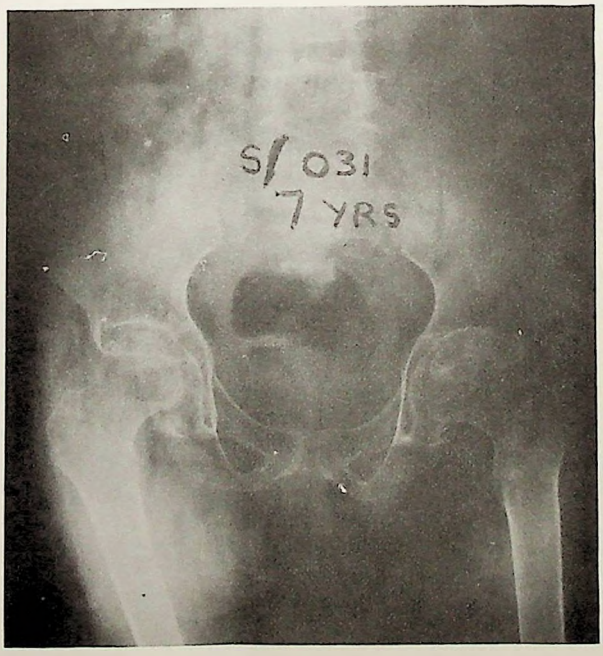

Fig 4. Hip joints - Severe changes. Note premature closure of the epiphyseal lines, gross reduction of the joint space and irregular articulating surfaces

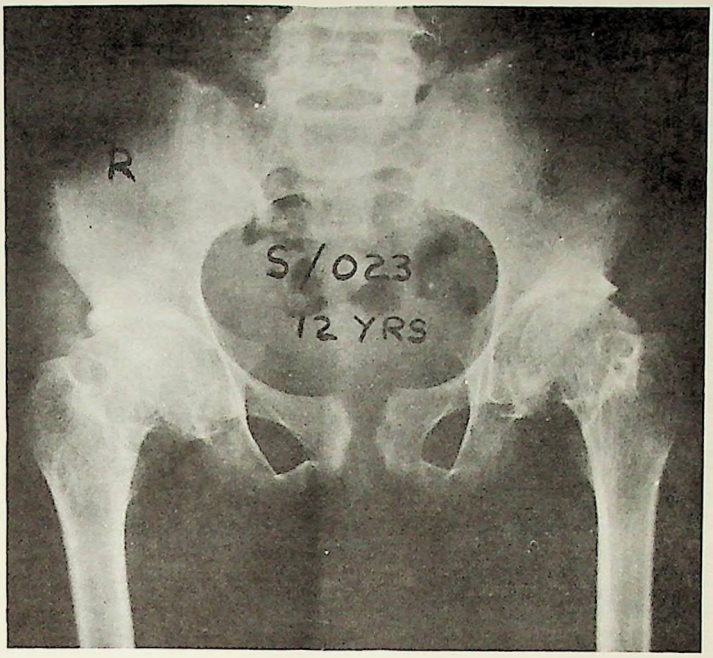

Fig 5. Hip joints - Severe changes. Same as in Fig 4 plus widening of symphysis pubis and marginal osteophytosis at the hip joints

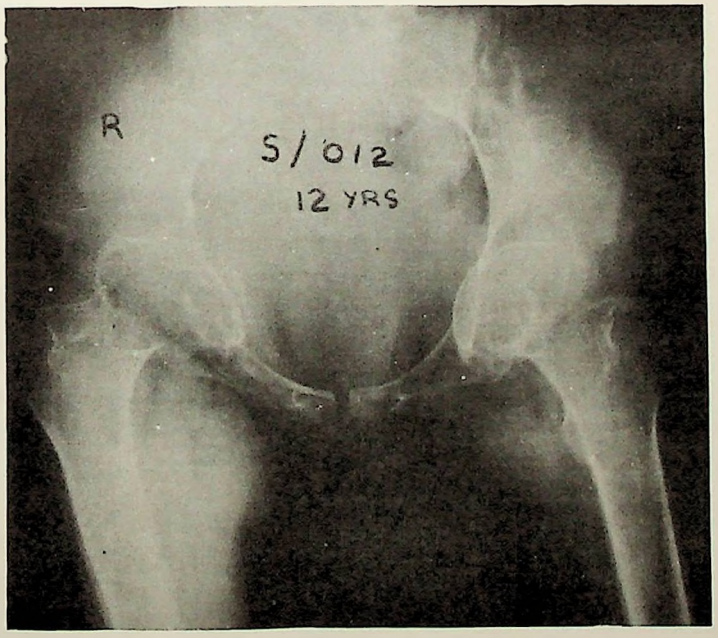

Fig 6. Hip joints - Gross changes Note the complete loss of joint space and deepening of the acetabulae

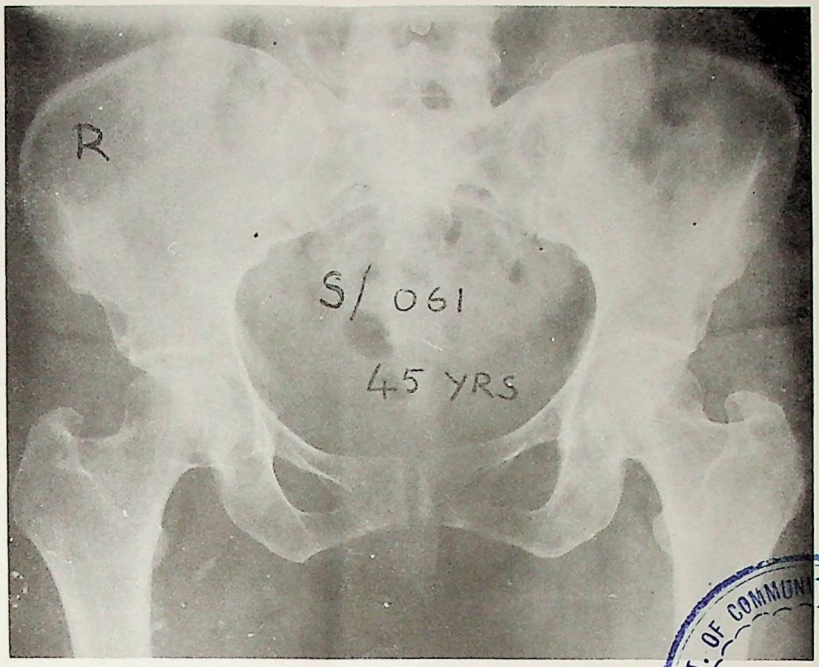

Fig 7. Normal hip joints, but in an adult. Right thigh shows cigar shaped calcification, suggestive of cysticercosis - an incidental finding

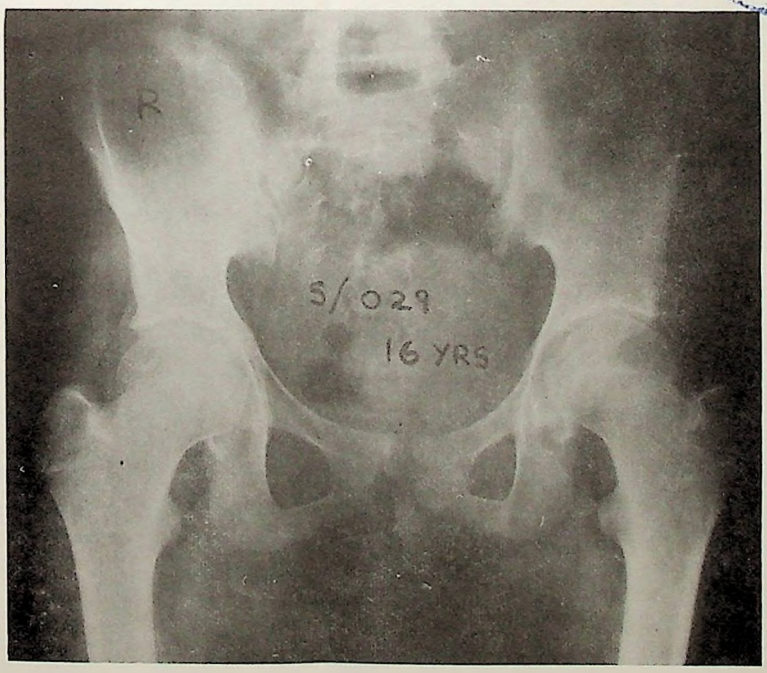

Fig 8. Mild change - Narrowing of joint space

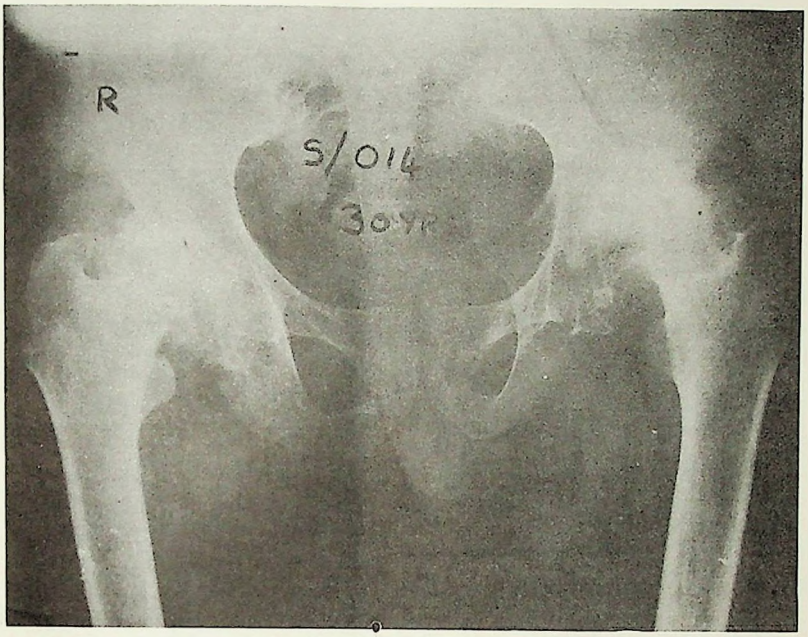

Fig 9. Hip joints - Gross changes. As in Fig 6, but in an adult.
Note the gross destruction of the left hip joint with subluxation.

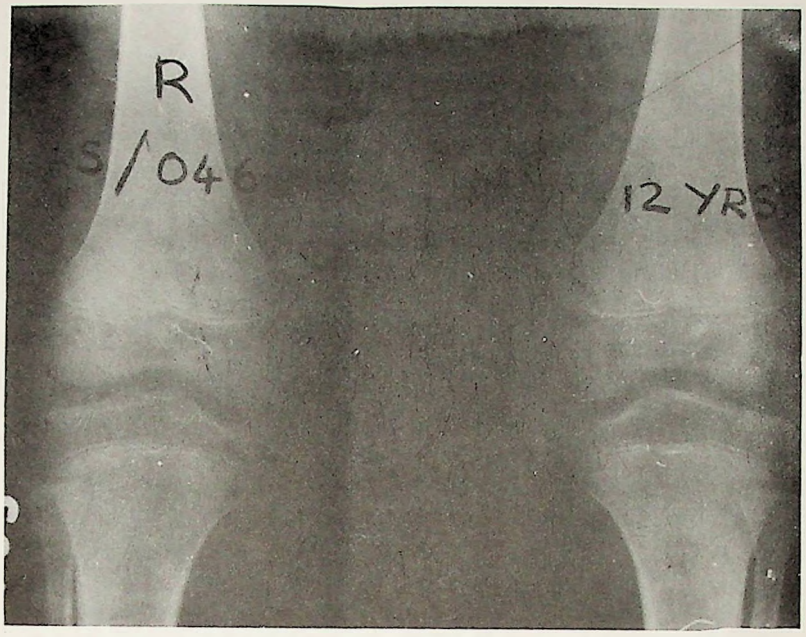

Fig 10. Normal knee joints - AP view

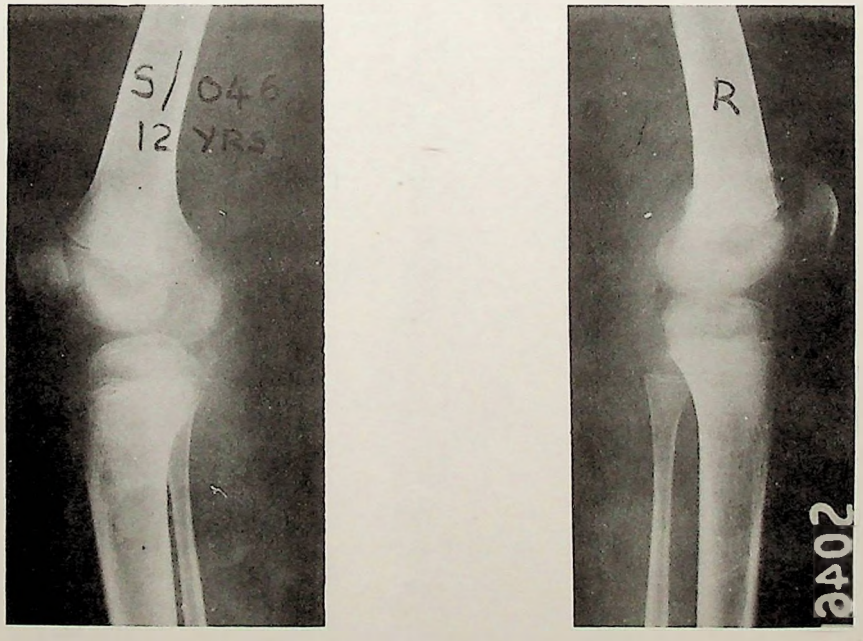

Fig 11. Normal knee joints - Lateral view

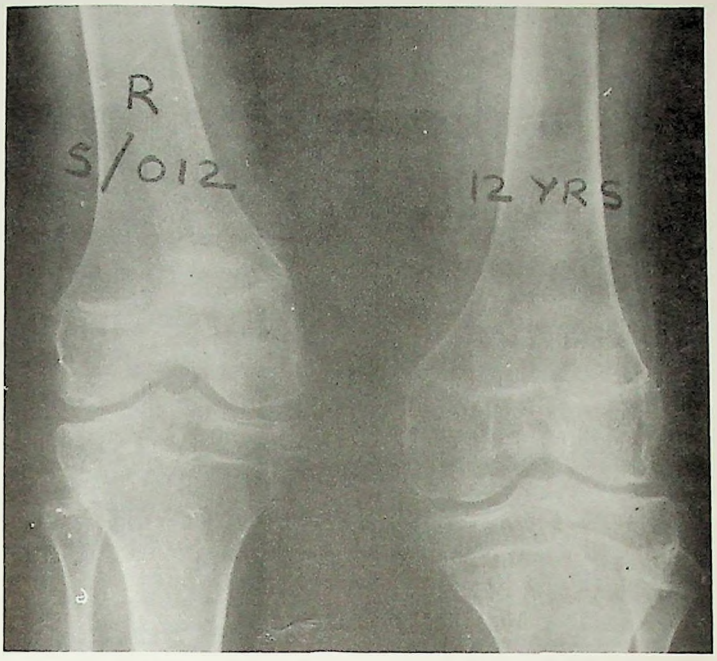

Fig 12. Knee joints - Moderate changes. AP view. Note premature closure of the epiphyseal lines, narrowing of the joint space and cystic areas with marginal sclerosis

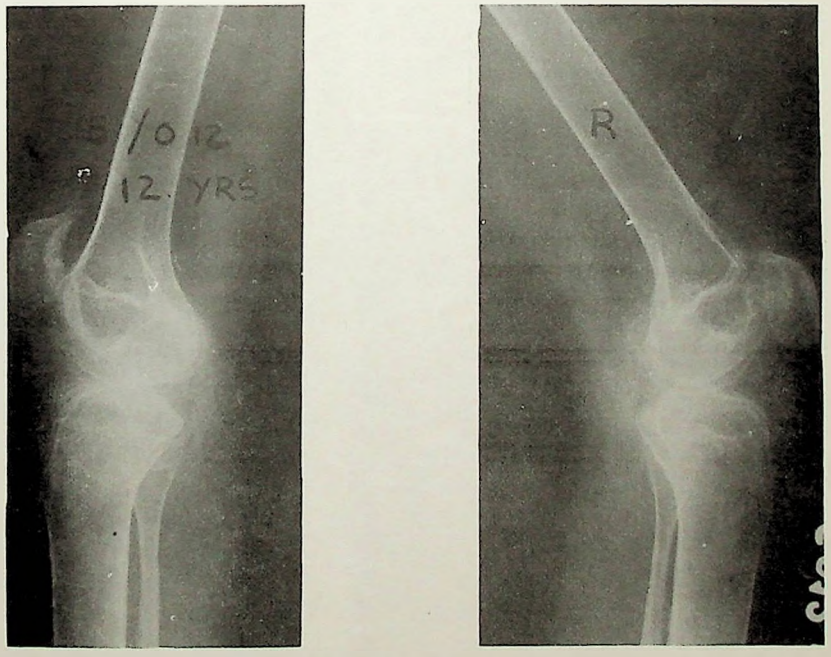

Fig 13. Knee joints - Moderate changes. Lateral view. Note excavated articular surface of the patellae

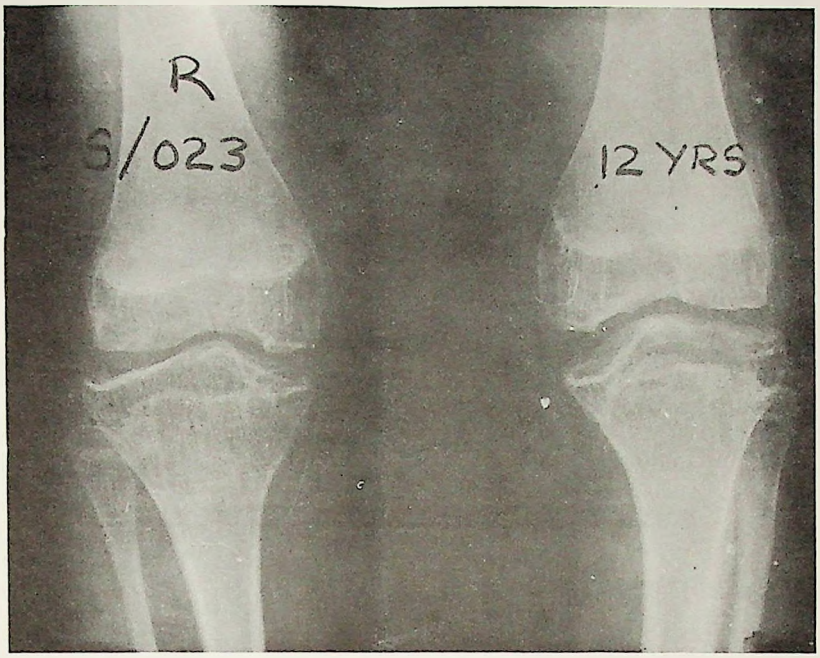

Fig 14. Knee joints - Severe changes. AP view.
As in Fig 4, but in the knee joints

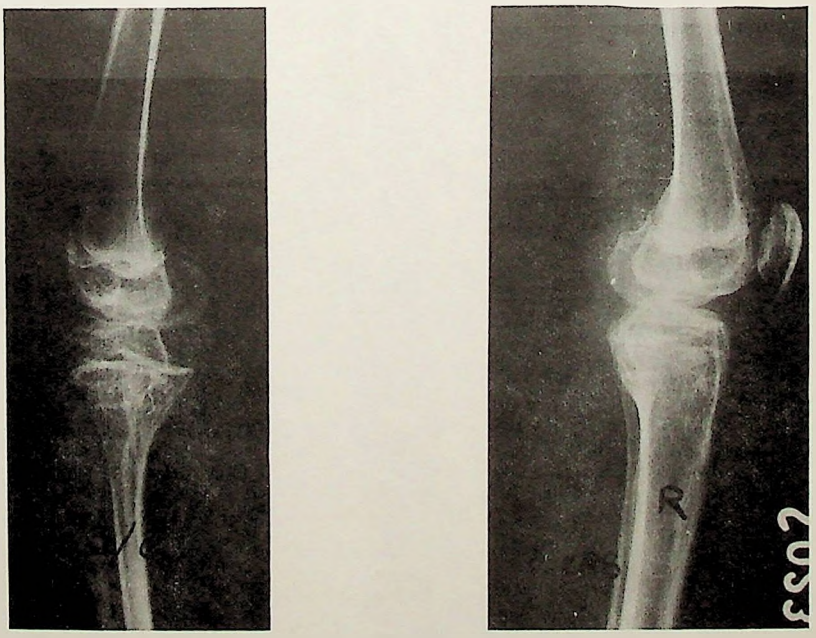

Fig 15. Knee joints - Severe changes. Lateral view

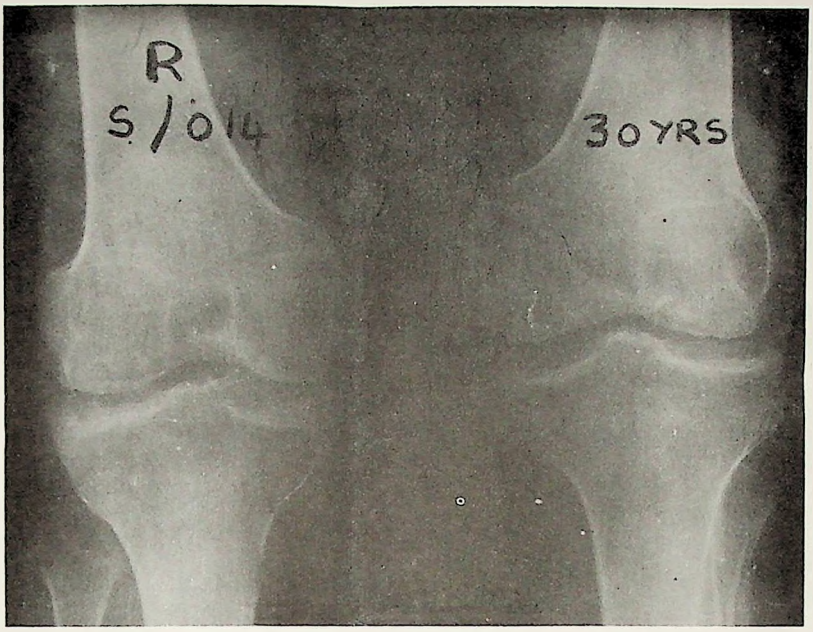

Fig 16. Knee joints - Moderate changes. AP view.
As in Fig 12, but in an adult

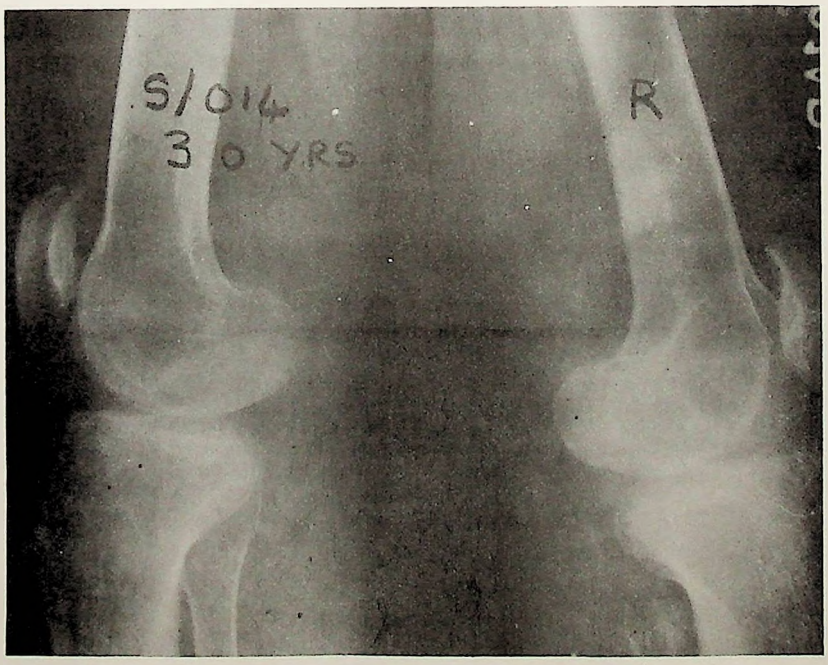

Fig 17. Knee joints - Moderate changes. Lateral view

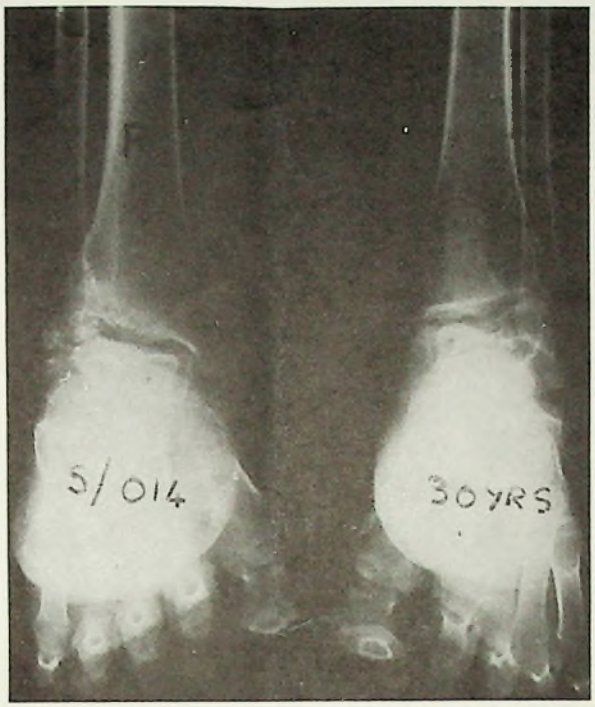

Fig 18. Ankle joints - AP view. Note flattering of the superior surface of the body of the talus

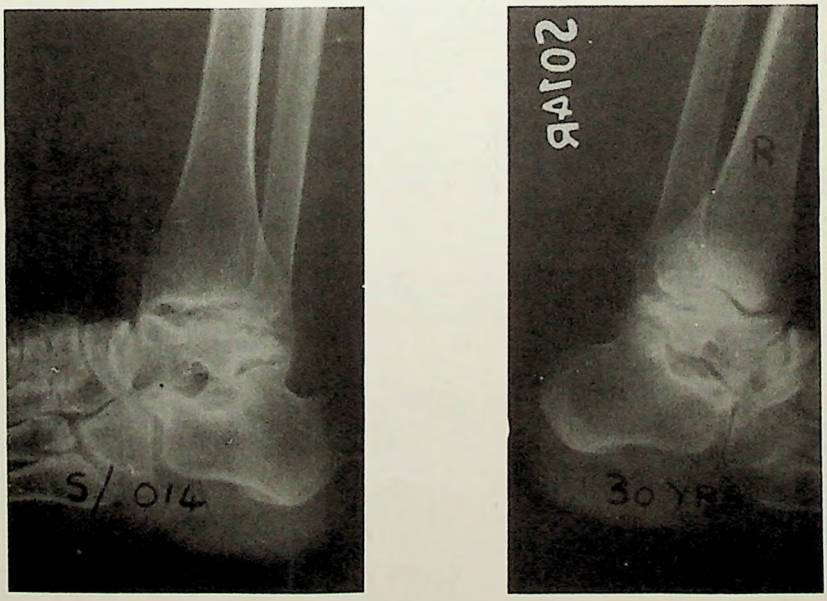

Fig 19. Ankle joints - lateral view. As in Fig 18

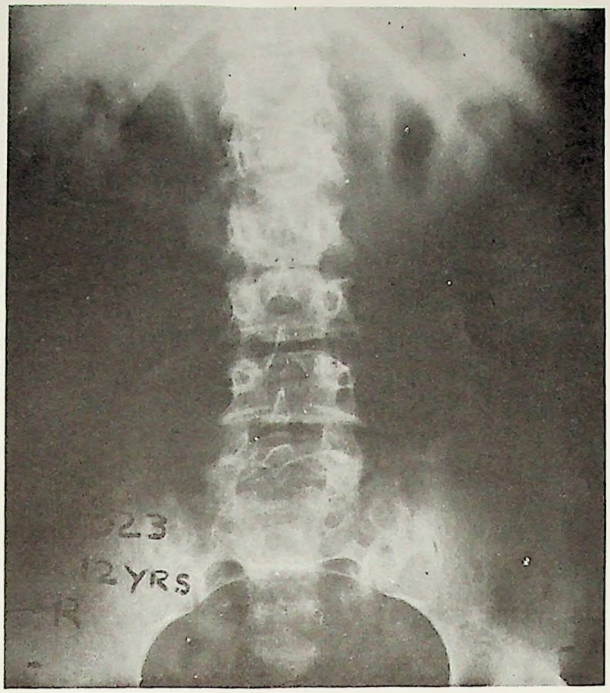

Fig 20. Lumbar Spine - AP view. Note flattering of the vertebral bodies with osteophytosis

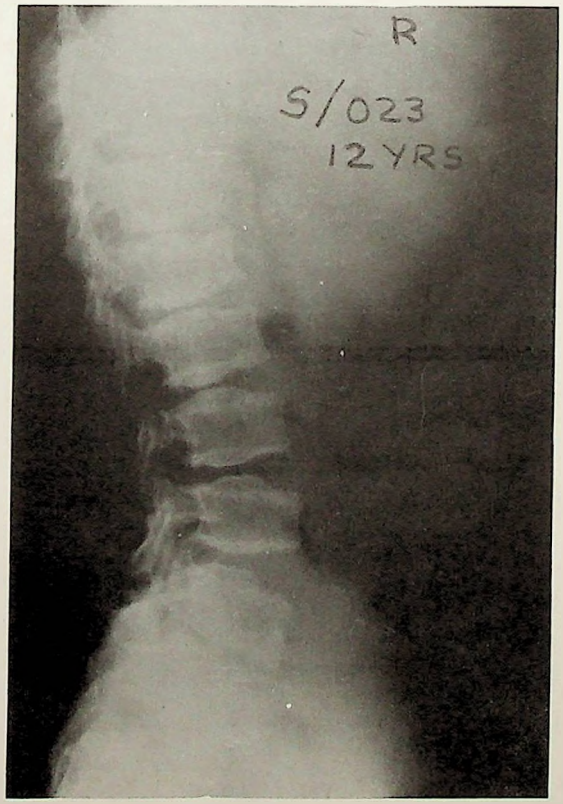

Fig 21. Lumbar Spine - Lateral view. As in Fig 20

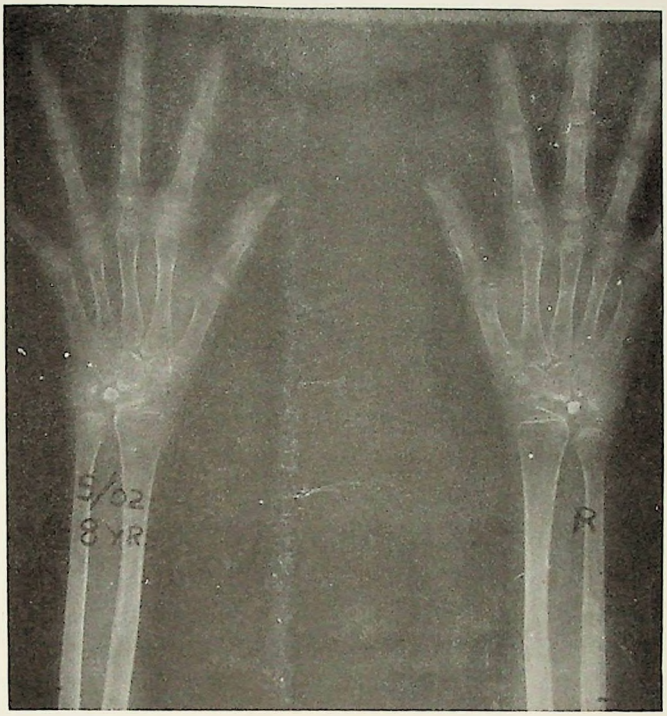

Fig 22. Wrist joints - PA view.
Note increased density of the
lunate bones - osteochondritis.

ICMR Bulletin: Vol.7, No.6, June 1977

HANDIGODU SYNDROME
(Endemic Familial Arthritis)

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In April 1976, preliminary studies were undertaken by the National Institute of Nutrition, Hyderabad. It was noted that 8 years earlier there were no reports of this disease; the disease was seen predominently among harijans and appeared to have a strong familial tendency; it appeared to have some relation to dwarfism and rickets and there was close resemblance to Mseleni joint disease reported from parts of South Africa in 1973.

A detailed epidemiological study was undertaken by a team of the National Institute of Nutrition, Hyderabad in November 1976.

DEMOGRAPHIC STUDIES

The disease was found to be confined to a few villages around the towns of Sagar in Shimoga district and Balehonur in Chikkamagalur district (parts of the Malnad). Sagar taluk has a population of around 0.13 million according to the 1971 census. The social caste structure in this area in the order of population strength, are Deevaru or Idiga, followed by Brahmins of Havyaka sect, Vokkaligas (with a number of sub-division), Gowdas, Lingayats and Harijans. The population figure for scheduled castes and tribes in Sagar taluk was 11,741. Although all the scheduled castes and tribes identified themselves as either Harijans or Adi Karnatakas, there were atleast 4 sub-castes among the scheduled tribes in addition to

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several nomadic tribes. Mixing and co-existence of different sub-caste was rare and marriages almost invariably occurred within the sub-caste and that too, within a radius of 5-10 km. resulting in a very high degree of inbreeding.

A house to house study was carried out in 40 villages of which 34 were affected. In view of the small number of households in these villages, it was possible to obtain detailed information on almost all the affected households.

THE DISEASE

Out break: The disease first appeared 6-8 years ago, with the majority of cases developing 4-6 years ago. Very few cases have been reported in the last two years. It did not appear to be ineffective in nature, nor was it seasonal or follow any environmental catastrophe. It was confined to humans. The onset was gradual, punctuated by pain of moderate degree in the lower limbs, particularly in the lumbo-sacral regions, hips and knee joints. Pain was bilateral and of variable severity, sometimes necessitating immobilisation. A proportion of cases suffered from crippling flexion deformities of the knee and hip joints. Involvement of other joints was rare. The characteristic radiological features were destruction of articular surfaces of the femur and tibia, osteophytic lipping of several bones, rarefaction of the bones around the area of destruction, varying degrees of osteoarthritic changes in the hip joint and in some cases cystic changes in the femoral head.

Subjects of all ages between 6 and 50 years were affected and there appeared to be no sex prediliction. The disease per se was not fatal. It was noted that over 85 percent of the affected subjects belonged to the Harijan community while the rest belonged to Vokkaliga and Deevaru communities. Not a single case was found among the brahim community.

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Familial characters of the disease: In 40 of the 60 affected households, more than one member was affected. A detailed study revealed the existence of a close social relationship not only between the affected households of the same village but also between affected households of different villages. Although subjects belonging to three different generations suffered from the disease in some familieis it was significant that in all of them, the disease made its appearance within a short period of 1-2 years. Equally significant was the observation that in many households, the children showed the manifestation first followed by the parents and on a occassions by the grand parents. The disease was also seen in both the local inhabitants and their partners who originally came from other non-affected neighbouring villages but had subsequently settled down in affected villages.

Relation to achondroplasic dwarfism: A striking observation was that in the affected villages in both Sagar and Chikkamagalur districts, achondroplasic dwarfs were frequently seen. As many as 22 dwarfs of different ages were encountered during the survey, the oldest being over 60 years of age. These dwarfs were seen in villages whose inhabitants were most severely affected with arthritis, which is clinically a distinct syndrome not related to any congenital lesion. All the dwarfs were found to be closely related by birth to members of affected households and some of them were related to other dwarfs in the same village and to the dwarfs in the same village and to the dwarfs in other villages. In a few cases the dwarfs had suffered from arthritis.

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A variety of malformations ie., phocomelia, congenital absence of eye balls, congenitalscoliosis and microcephaly were observed in young children in the affected areas. Congenital icthyosis was seen in four adults of the affected households.

AGRICULTURAL PRACTICES IN AFFECTED VILLAGES

Paddy cultivation and tending of plantations are the main activities of the inhabitants. Crops grown in these villages include arecanut, pepper, cardamom and coffee. The farmers of the areas are progressive and have adopted mechanised cultivation High yielding varieties of paddy have been introduced during the last decade. These and otheragricultural developments have madeit necessary for them to go in for extensive use of fertilizers and pesticides.

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FOOD HABITS

The staple in these areas is rice. It is supplemented with dhal, vegetables, and milk and milk products in the case of economically well off groups. Vokkaliga, Gowda, and Deevaru communities in addition consume animal foods.

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In view of the clustering of the affected villages around the two major rural towns of Sagar and Balehonnur, it was considered important to extend these studies to villages situated at a distance from these towns, beyond the affected villages. Accordingly the study was carried out in 15 neighbouring villages. In all these villages the 4 communities were represented but no case of disease had been reported. It was noted that there was a definite change in the environment of these villages as compared to the affected villages. Food habits were also changed showing increasingly dependency in milets.

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C O N C L U S I O N S

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In spite of the strong familial tendency epidemiological studies suggest that the disease, a recent one, has been precipitated by certain changes in the microecological system. One striking change over the last decade is the use of high yielding varieties of paddy with attendant intensive agricultural practices and the use of large amounts of pesticides. Pond water being contaminated is a very high risk and the immediate and direct effect on the fauna after spraying operations has already been indicated by the wholesale mortality in fishes and crabs.

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DIS 12.2

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KHA-2 DIS 12.3

DIS

REPORT OF INVESTIGATION TEAM OF EXPERTS ON HANDIGODU SYNDROME IN SHIMOGA AND CHIKKAMAGALUR DISTRICTS.

References: 1. EST(1)CR-502/82-83 from the Principal, B.M.C. Bangalore dated 23.11.1982

2. OM.E.M.C./24/82-83 dated 3.11.82 of D.M.E. Bangalore

MEMBERS OF THE TEAM

1. Dr Mohamed Nuruddin (Leader of the team), Professor of Medicine

2. Dr Deshikachar, Professor of Surgery

3. Dr M N Shivaprakash, Professor of Orthopaedics

4. Dr Krishna Murthy, Professor of Pathology

5. Dr Parthasarathy, Professor of Radiology

6. Dr P V Aswath, Assistant Professor of P&S Medicine

Accompanied by: to Handigodu and other villages

1. Dr Gopinath, Assistant District Health & F W Officer, Sagar

2. Dr Lokappa, Medical Officer, Sagar

With para medical staff.

Places visited in Shimoga District

1. Handigodu Village
2. Keladipura
3. Kolisaru
4. Bandagadde

Accompanied by the following at Chikkamagalur

1. Dr Prasanna, District Health & FW Officer, Chickmagalur

Places visited at Chikkamagalur District

1. Kadaba Gadde
2. Hallandur
3. Sangameshwarpet

Patients from the affected areas were seen in the above centres.

At Sagar Taluk 49 cases were examined and at Chikkamagalur division 46 cases were examined.

AGE GROUP : METHODS

Patient age group varied from 4 - 40 years. Most of the affected persons belong to the age group of 5 - 15 years.

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Religion & Caste

All the patients belong to Hindu Religion. Most of the patients belong to Harijans, Vokkaligas and Edigas.

Detailed clinical examination of each case was done.

An attempt was made to study the Pedigree of each family.

Blood samples were drawn for investigation. Living conditions and the surrounding area were studied.

History of origin of the disease, their food habits and type of diet were studied.

Clinical photographs of the patients were taken.

OBSERVATIONS

1. Most of the patients belong to socially economically backward class

2. Highest incidence of the disease is seen in the age group of 5-15 years.

3. Sex predilection is equal

4. No significant nutritional deficiency was noticed. No gross symptoms of deficiency was water and fat soluble vitamins seen. No gross protein deficiency noticed. No calorie malnutrition observed.

5. Upper part of the body was seen to be normally built than to the lower part.

TABLE 1

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** Patient is unable to stand. Tibia and fibula were bowed. Fibula is almost by the side of medial side of tibia.

OBSERVATIONS

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Span length was found to be abnormal. It is observed that in majority of the individuals this abnormality ranged from 1 - 7 cms.

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Villagewise details of the Pedigree studies were tabulated in Table 2.

Sl No	No. of cases	No. of families in				No. of families with history of consanguineousmarriage
		Handigodu	Kaldipura	Sangameswar-pet	Kadaba-gadde	
1	1	1*	2	1	3*	3(1+2)
2.	2	-	1	-	1	-
3.	3	2	2	1	1*	1
4.	4	2*	2*	3*	-	3(1+1+1)
5.	5	1	1	-	-	-
6.	6	1	-	-	-	-
7.	7	-	-	1*	-	1
Total No. of families		7	8	6	5	8
NO. OF CASES		26	23	23	8	

*Family with history of consanguineous marriage found.

In 4 villages where detailed history of 80 cases were collected The cases were distributed in 26 families and history of consanguineous marriage was observed in eight families.

All these families originally belonged to Karkala Area (Gattada Kelage) and migrated to these places about 2 centuries back. (Keladi Shivappa Nayaka's time).

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FOOD HABITS

Rice with sambar or rasam without dhal with available vegetables and roots, like Kesavedantu (Colocasiae), Basale (spinache) Dantu (amaranth).

Rarely they take non-vegetarian diet once in a year. During rainy season crabs and fishes are consumed rarely. Some food is consumed by other people of village.

As it was not the rainy season fish and crab samples were not collected for examination.

General environmental conditions and housing were quite satisfactory. Lighting ventilation of the houses were satisfactory. Water sources were common both for affected and non affected persons in the villages.

In many families many persons were affected. Symptoms of this syndrome were suggestive of a developmental defect as a result of flexion deformity of hip and knee, upper limbs are normal in these people.

Majority of the individuals are able to move about for their routine activity without much of complaint except perhaps that their efficiency was lower to the extent of disability.

From history narrated by local people and from affected area, no definite date could be fixed as to when the disease was first seen. It is likely that this disease appears to be present in the community since a few generations.

Members of this community are in the habit of consuming illicit liquors from their teen age in both the sexes. Blood samples collected showed normal serum calcium levels and in two patients eosinophilia was present. Anaemia was present in a few cases. Anaemia is of normocytic hypochromic type. No radiological investigation could be done.

On the day of our visit to Handigodu village one person who was suffering from the Handigodu syndrome had died because of pulmonary tuberculosis as told by the relatives.

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Following are the names of the persons who were subjected to blood examination.

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3.	Nagaraja	13	M
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5.	Ramachandra	18	M
6.	Bharathi	9	F
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10.	Varadappa	18	M
11.	Rudramma	25	F
12.	Raju	18	M
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15.	Pokuramma	35	F
16.	Thimmamma	45	F
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18.	Guttiyamma	40	F
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CONCLUSIONS

1. The disease appears to be a herido families disorder affecting endochnodral ossification.
2. Subsequent crippling appears to be due to super added traume on a joint which is not mechanically sound.

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3. The crippling is also due to lack of treatment at the crucial time of its development.
4. Crippling is appeared to be a preventable condition.

OBSERVATIONS

In these crippled persons their earning capacity is considerably reduced. Consequently their nutritional status decreases. They become susceptible tomany other diseases due to above factors.

RECOMMENDATIONS

1. The individuals who develop the signs of inflammation of the joints have to be shifted to a hospital under the care of an orthopaedic surgeon.
2. Immobilisation of the joint, traction, analgesics, steroids and antibiotics have to be used whenever felt necessary.
3. Deformed joints should be surgically corrected and o this will increase the earning capacity of the individual enermously.
4. The persons who underwent corrections of the joints should be rehabilitated with occupational therapy.
5. Genetic counselling and marriage counselling should be done to control the disease.
6. Services of social and phylanthropic associations could be taken for the social upliftment of the community and rehabilitation of the cases.
7. Chromosomal and genetic studies have to be undertaken. For the detailed study of cases a selected number of cases from the affected willages from each group of crippled and non crippled persons have to be taken. To pin point the herido familial character of the disease and to adopt practical measures in the management and prevention of this disease.
8. In view of the magnitude and endemicity of the problem and in the interest of long term treatment and in the prevention of disease it is suggested that a creation of a special cell headed by an officer with his associated staff.
9. Such rehabilitation centres should be located in a place where the persons of both the places could be treated similar to Kyasanur Forest Disease team.

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10. The team suggests that a detailed survey has to be conducted in Karkala for detection of cases from where these people have migrated.

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DIS 12.3

REPORT OF INVESTIGATION TEAM OF EXPERTS ON HANDIGODU SYNDROME IN SHIMOGA AND CHIKKAMAGALUR DISTRICTS.

References: 1. MST(1)CR-502/82-83 from the Principal, B.M.C. Bangalore dated 23.11.1982

2. CM.D.M.C./24/82-83 dated 3.11.82 of D.M.C. Bangalore

MEMBERS OF THE TEAM

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5. Dr Parthasarathy, Professor of Radiology

6. Dr P V Aswath, Assistant Professor of P&S Medicine

Accompanied by: to Handigodu and other villages

1. Dr Gopinath, Assistant District Health & F W Officer, Sagar

2. Dr Lokappa, Medical Officer, Sagar

With para medical staff.

Places visited in Shimoga District

1. Handigodu Village
2. Koladipura
3. Kolisara
4. Bandagadde

Accompanied by the following at Chikkamagalur

1. Dr Prasanna, District Health & FW Officer, Chickmagalur

Places visited at Chikkamagalur District

1. Kadaba Gadde
2. Hallandur
3. Sangameshwarpet

Patients from the affected areas were seen in the above centres.

At Sagar Taluk 49 cases were examined and at Chikkamagalur division 46 cases were examined.

AGE GROUP : METHODS

Patient age group varied from 4 - 40 years. Most of the affected persons belong to the age group of 5 - 15 years.

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Religion & Caste

All the patients belong to Hindu Religion. Most of the patients belong to Harijans, Vokkaligas and Edigas.

Detailed clinical examination of each case was done.

An attempt was made to study the Pedigree of each family.

Blood samples were drawn for investigation. Living conditions and the surrounding area were studied.

History of origin of the disease, their food habits and type of diet were studied.

Clinical photographs of the patients were taken.

OBSERVATIONS

1. Most of the patients belong to socially economically backward class

2. Highest incidence of the disease is seen in the age group of 5-15 years.

3. Sex predilictions is equal

4. No significant nutritional deficiency was noticed. No gross symptoms of deficiency was water and fat soluble vitamins seen. No gross protein deficiency noticed. No calorie malnutrition observed.

5. Upper part of the body was seen to be normally built than to the lower part.

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Karnataka Medical Journal, Vol.XLVIII, pp.103-107, April-June, 1983

HANDIGODU SYNDROME - A MYSTERIOUS DISEASE

P V Aswath* and M K Sudarshan**

Introduction

In January 1975, 4 patients from Handigodu village in Karnataka were examined at the local hospital for the complaint of inability to walk. In the next week there were about 30 similar cases from the same village. Initially it was thought to be a neurological disorder. However, soon a team of specialists who examined these cases identified the disease to be an orthopaedic problem and ruled out any primary neurological involvement. The disease was labelled as "Handigodu syndrome" named after the village Handigodu from where the cases were reported first[1]. Following this many investigations have been conducted on this disease. But unfortunately the aetiological factor(s) responsible are yet to be identified. An approximate estimate has been made that more than 600 cases are affected by this disease. Though the disease has been reported about 8 years back, publi[illegible] feel that nothing substantial has been made to contain the problem, as the disease permanently cripples the individuals.[2,3]. These incapacitated individuals who are invariably from a lower class, became a socio-economic burden not only to their families but also to the community.

A profile of the disease has been presented in brief and a few suggestions have been made.

Topography

The disease has been reported from a few villages (only) around the towns of Sagar in Shimoga district and Balehonnur in Chikmagalur district (parts of Malnad area) ofKarnataka State.

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* Assistant Professor of Preventive and Social Medicine, Bangalore Medical College, Bangalore.

** Assistant Professor of Community Medicine, Kempegowda Institute of Medical Sciences, Bangalore 4

The affected region is predominantly an agricultural area and has no industries. These villages are situated along the Western ghats and mostly at an altitude of 2000-3000 feet above sea level. The annual rainfall is about 80-120 inches. The region is thick in forest wealth and certain areas have a classical tropical forest. The temperature ranges from upper nineties in summer to lower seventies (Faranheit) in winter. The relative humidity is quite high.

Demography

The population of Chikmagalur district and Shimoga district is about 0.9 million and 1.6 million respectively[4]. The caste wise dominance in the area is idigas, brahmins, vokkaligas, lingayats and harijans in that order.[5] Because of the stringent social mores, marriages take place within the same sub-caste and that too within a radius of 5-10 kms resulting in high degree of inbreeding. About 76% of the population is rural and 86% 'Hindus'. The literary rate is about 43%. The estimated crude birth rate and the crude death rate are 29.2 and 12.0 respectively (Karnataka State figures). The infant mortality rate is 83.3 and the projected expectation of life at birth by sex in Karnataka is 61.4 years for males and 59.4 years for females.

Environmental sanitation

It is observed that there is a fairly clean surroundings around the affected households in these villages. The houses are mostly kucha with thatched roofing, though a few houses are pucca with tiled roofing. The source of drinking water is well. There is no drainage system and the villagers go to the adjacent forest for their excretory functions.

Agricultural activities

Most of the people in the area are land cultivators and their chief agricultural produces include rice, arecanut, pepper, cardamom and coffee. The farmers are progressive and have

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mechanised cultivation. Following the introduction of high yielding varieties of paddy, since last decade there is a wide scale use of pesticides. The paddy fields and adjacent ponds contain a large variety of fish, crabs and frogs. However, following pesticide spraying of paddy fields, there is large scale death of local fauna. Farm labourers, particularly the poor harijans and vokkaligas though deny eating this dead fauna, it has been found to be not true. And consumption of crabs and fish has increased in recent years because of the tendency of discontinuing the practice of serving food (as a part of their wages) to these land labourers by their landlords.

Dietary practices

Rice forms the staple diet and is consumed 2-3 times a day along with tamarind soup. Though a detailed dietary history has not been elicited, it has been found that green leafy vegetables are consumed once or twice a month. Egg/meat are taken hardly 3-4 times a year and milk and milk products practically never. The tuber ordinary yam (which is known to have a high cyanide content) however, is soaked in water, washed (removes cyanide), cooked and consumed. Another interesting feature is the consumption of illicit country liquor, even by some of the women and children. However, as the wild flora in the area is abutting on the backyard of the houses, there is a suspicion about the type(s) of articles consumed in raw states.

The Disease (fig.1)

The disease per se is not fatal and is confined to humans.

ICMR as early as in 1976 conducted an house to house survey in 40 villages and found 34 villages to be affected. Subsequently many teams have investigated the disease. But it appears that an exact estimate of the number of cases affected is not available. The first investigating team reports that there was no history of similar illness in the previous generations. But a recent investigation team observes that this disease appears to be present in the community since a few generations.[6] It has also made a study of family pre

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pedigree in some cases and after observing a high frequency of consanguineous marriages in these cases, the disease is now suspected to be herido-familial. It is significant that in some households members of the three generations were affected almost at the same time. In many cases children suffered earlier and more severely than the elders of the household[7]. However, it appears that one of the drawbacks of these studies is the non-inclusion of adequate and appropriate controls and absence of sampling methods.

The disease has been found to affect people in the age group of 5-64 years. Infants and preschool children appear to have been spared. However, the disease is reported to have a higher predeliction towards younger age group (5-15 years). Most of the patients are from the socioeconomically backward classes (mostly harijans and a few vokkaligas and idigas) and not a single case has been reported from the brahmin community. No significant nutritional dificiencies are seen. The disease appears to be neither infectious nor contagious.

The onset is insidious, associated with pain and swelling of joints, particularly of the lower limbs. The frequently affected joints are hips and knees. In some cases, due to severity there is crippling flexion deformities and disuse muscular atrophy The characteristic radiological picture is destruction of articular surfaces of femur and tibia, osteophytic lipping of many bones, rarefaction of the bones around the area of destruction, varying degrees of osteoarthritic changes in the hip joint and sometimes with cystic changes in the femoral head. The disease is suspected to be affecting endochondral ossification. The subsequent crippling is thought to be due to superadded trauma on a joint which is not mechanically sound. It is opined that this crippling is preventable as it is due to lack of treatment at the crucial time ofits development.

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In one of the surveys, 22 dwarfs were encountered among 1000 harijans studied in the affected villages. It was found that all the dwarfs were inter-related, in addition, they were free from the disease except for three dwarfs.[7] Osteomalacia and rickets like manifestations have been frequently encountered in this area. A variety of congenital malformations such as phocomelia, congenital absence of eye ball with rudementary palpebral fissure, microcephaly, congenital scoliosis and icthyosis have beenobserved in the affected population.[8]

A team of investigators who recently investigated the disease in April 1983, suspect the disease as an autoimmmune reaction akin to rheumatoid arthritis or the auto-immune reaction being triggered of by the KFD viral infection prevalentin the region.[9]

The disease has been identified tobe similar to Kashinbeck syndrome (Urov's disease) reported among Siberians, which is presumed to be due to consumption of cereals contaminated with the fungus fusarium sporotrichiella. The disease also has a striking resemblance with Mseleni joint disease in South Africa, which is thought to be due to an unidenfified environmental factor.

Investigation

The haemotological investigations done include haemoglobin, ESR, peripheral Eosinophilia, random blood sugar, blood urea, serum electrolytes (Na K Mg), total serum proteins, total serum calcium, serum inorganic prosphorous and alkaline phosphatase. The other investigations done include tests of liver function like serum bilirubbin, vanden bergh reaction and thymol turbidity urine analysis, rose waaler test, C-reactive protein, CSF examination, ECG, EKG and electromyography. The results of these investigations have failed to throw light on the disease.

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C O N C L U S I O N

This crippling disease appears to be primarily an orthopaedic problem mostly affecting the poor lower socio economic ethnic f group, with a tendency for familial segregation. Apparently industrial effluents have no role in the genesis of this disease as there are no industries in this area.

Though the pathological process of the disease appears to have been studied, the aetiological factor(s) responsible for the same are still at large. The disease is thought to be herido familial following an history of high frequency of consanguineous marriages in the affected families. It is also suspected to be nutritional, suspected factors include (a) illicit country liquor, (b) pesticides (concentrated) in crabs and fishes consumed by the affected people, (c) unidentified food article consumed in raw state from the adjacent thick forest.

Lastly, it is to be noted that a striking observation of a number of achondroplasic dwarfs in the affected area has been made. It should also be remembered that this area is known for the occurrence of Kyasanur Forest Disease (KFD).

Suggestions

In the light of the above observations, few suggestions are made for future work on the disease:

a. There is an urgent need for an immediate establishment of a special research cum service cell at a suitable place (preferably at Sagar Town) to carry out further investigations and to provide therapeutic and preventive services. The cell should be staffed by an Epidemiologist, Orthopaedic Surgeon(s), Radiologist, Genetic Specialist, Physiotherapist(s), other ancillary and field staff with adequate clinical and independent transport facilities.

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b. The immediate task is to identify all the cases and mapping of the disease. Those persons in whom the disease is still in early stages, should be provided with physiotherapy and the required treatment facilities at the centre.

c. The persons who are already permanently crippled have to be rehabilitated vocationally to help them to earn their livelihood. It is also desirable that the Government should provide a suitable compensation to these individuals.

d. As one of the investigation teams has observed the disease to be associated with consanguineous marriages, the 'at risk' population in the area must be properly educated in this regard. The people should also be advised against the consumption of crabs and fish from the paddy fields.

e. There is scope for a detailed field epidemiological investigation of this mysterious disease.

Acknowledgement

The authors are thankful to Dr A N Armugam, Professor and Head of the Department of Community Medicine, Kempegowda Institute of Medical Sciences, for his valuable suggestions and help.

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HANDIGODU SYNDROME - A MYSTERIOUS DISEASE

P V Aswath* and M K Sudarshan**

Introduction

In January 1975, 4 patients from Handigodu village in Karnataka were examined at the local hospital for the complaint of inability to walk. In the next week there were about 30 similar cases from the same village. Initially it was thought to be a neurological disorder. However, soon a team of specialists who examined these cases identified the disease to be an orthopaedic problem and ruled out any primary neurological involvement. The disease was labelled as "Handigodu syndrome" named after the village Handigodu from where the cases were reported first[1]. Following this many investigations have been conducted on this disease. But unfortunately the aetiological factor(s) responsible are yet to be identified. An approximate estimate has been made that more than 600 cases are affected by this disease. Though the disease has been reported about 8 years back, public feel that nothing substantial has been made to contain the problem, as the disease permanently cripples the individuals.[2,3]. These incapacitated individuals who are invariably from a lower class, became a socio-economic burden not only to their families but also to the community.

A profile of the disease has been presented in brief and a few suggestions have been made.

Topography

The disease has been reported from a few villages (only) around the towns of Sagar in Shimoga district and Balehonnur in Chikmagalur district (parts of Malnad area) ofKarnataka State.

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The affected region is predominantly an agricultural area and has no industries. These villages are situated along the Western ghats and mostly at an altitude of 2000-3000 feet above sea level. The annual rainfall is about 80-120 inches. The region is thick in forest wealth and certain areas have a classical tropical forest. The temperature ranges from upper nineties in summer to lower seventies (Faranheit) in winter. The relative humidity is quite high.

Demography

The population of Chikmagalur district and Shimoga district is about 0.9 million and 1.6 million respectively[4]. The caste wise dominance in the area is idigas, brahmins, vokkaligas, lingayats and harijans in that order.[5] Because of the stringent social mores, marriages take place within the same sub-caste and that too within a radius of 5-10 kms resulting in high degree of inbreeding. About 76% of the population is rural and 86% 'Hindus'. The literary rate is about 43%. The estimated crude birth rate and the crude death rate are 29.2 and 12.0 respectively (Karnataka State figures). The infant mortality rate is 83.3 and the projected expectation of life at birth by sex in Karnataka is 61.4 years for males and 59.4 years for females.

Environmental sanitation

It is observed that there is a fairly clean surroundings around the affected households in these villages. The houses are mostly kucha with thatched roofing, though a few houses are pucca with tiled roofing. The source of drinking water is well. There is no drainage system and the villagers go to the adjacent forest for their excretory functions.

Agricultural activities

Most of the people in the area are land cultivators and their chief agricultural produces include rice, arecanut, pepper, cardamom and coffee. The farmers are progressive and have

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mechanised cultivation. Following the introduction of high yielding varieties of paddy, since last decade there is a wide scale use of pesticides. The paddy fields and adjacent ponds contain a large variety of fish, crabs and frogs. However, following pesticide spraying of paddy fields, there is large scale death of local fauna. Farm labourers, particularly the poor harijans and vokkaligas though deny eating this dead fauna, it has been found to be not true. And consumption of crabs and fish has increased in recent years because of the tendency of discontinuing the practice of serving food (as a part of their wages) to these land labourers by their landlords.

Dietary practices

Rice forms the staple diet and is consumed 2-3 times a day along with tamarind soup. Though a detailed dietary history has not been elicited, it has been found that green leafy vegetables are consumed once or twice a month. Egg/meat are taken hardly 3-4 times a year and milk and milk products practically never. The tuber ordinary yam (which is known to have a high cyanide content) however, is soaked in water, washed (removes cyanide), cooked and consumed. Another interesting feature is the consumption of illicit country liquor, even by some of the women and children. However, as the wild flora in the area is abutting on the backyard of the houses, there is a suspicion about the type(s) of articles consumed in raw states.

The Disease (fig.1)

The disease per se is not fatal and is confined to humans.

ICMR as early as in 1976 conducted an house to house survey in 40 villages and found 34 villages to be affected. Subsequently many teams have investigated the disease. But it appears that an exact estimate of the number of cases affected is not available. The first investigating team reports that there was no history of similar illness in the previous generations. But a recent investigation team observes that this disease appears to be present in the community since a few generations.[6] It has also made a study of family pre

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pedigree in some cases and after observing a high frequency of consanguineous marriages in these cases, the disease is now suspected to be heredo-familial. It is significant that in some households members of the three generations were affected almost at the same time. In many cases children suffered earlier and more severely than the elders of the household[7]. However, it appears that one of the drawbacks of these studies is the non-inclusion of adequate and appropriate controls and absence of sampling methods.

The disease has been found to affect people in the age group of 5-64 years. Infants and preschool children appear to have been spared. However, the disease is reported to have a higher predeliction towards younger age group (5-15 years). Most of the patients are from the socioeconomically backward classes (mostly harijans and a few vokkaligas and idigas) and not a single case has been reported from the brahmin community. No significant nutritional dificiencies are seen. The disease appears to be neither infectious nor contagious.

The onset is insidious, associated with pain and swelling of joints, particularly of the lower limbs. The frequently affected joints are hips and knees. In some cases, due to severity there is crippling flexion deformities and disuse muscular atrophy The characteristic radiological picture is destruction of articular surfaces of femur and tibia, osteophytic lipping of many bones, rarefaction of the bones around the area of destruction, varying degrees of osteoarthritic changes in the hip joint and sometimes with cystic changes in the femoral head. The disease is suspected to be affecting endochondral ossification. The subsequent crippling is thought to be due to superadded trauma on a joint which is not mechanically sound. It is opined that this crippling is preventable as it is due to lack of treatment at the crucial time ofits development.

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In one of the surveys, 22 dwarfs were encountered among 1000 harijans studied in the affected villages. It was found that all the dwarfs were inter-related, in addition, they were free from the disease except for three dwarfs.[7] Osteomalacia and rickets like manifestations have been frequently encountered in this area. A variety of congenital malformations such as phocomelia, congenital absence of eye ball with rudementary palpebral fissure, microcephaly, congenital scoliosis and icthyosis have beenobserved in the affected population.[8]

A team of investigators who recently investigated the disease in April 1983, suspect the disease as an autoimmmune reaction akin to rheumatoid arthritis or the auto-immune reaction being triggered of by the KFD viral infection prevalentin the region.[9]

The disease has been identified tobe similar to Kashinbeck syndrome (Urov's disease) reported among Siberians, which is presumed to be due to consumption of cereals contaminated with the fungus fusarium sporotrichiella. The disease also has a striking resemblance with Mseleni joint disease in South Africa, which is thought to be due to an unidentified environmental factor.

Investigation

The haemotological investigations done include haemoglobin, ESR, peripheral Eosinophilia, random blood sugar, blood urea, serum electrolytes (Na K Mg), total serum proteins, total serum calcium, serum inorganic prosphorous and alkaline phosphatase. The other investigations done include tests of liver function like serum bilirubbin, vanden bergh reaction and thymol turbidity urine analysis, rose waaler test, C-reactive protein, CSF examination, ECG, EKG and electromyography. The results of these investigations have failed to throw light on the disease.

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C O N C L U S I O N

This crippling disease appears to be primarily an orthopaedic problem mostly affecting the poor lower socio economic ethnic f group, with a tendency for familial segregation. Apparently industrial effluents have no role in the genesis of this disease as there are no industries in this area.

Though the pathological process of the disease appears to have been studied, the aetiological factor(s) responsible for the same are still at large. The disease is thought to be herido familial following an history of high frequency of consanguineous marriages in the affected families. It is also suspected to be nutritional, suspected factors include (a) illicit country liquor, (b) pesticides (concentrated) in crabs and fishes consumed by the affected people, (c) unidentified food article consumed in raw state from the adjacent thick forest.

Lastly, it is to be noted that a striking observation of a number of achondroplasic dwarfs in the affected area has been made. It should also be remembered that this area is known for the occurrence of Kyasanur Forest Disease (KFD).

Suggestions

In the light of the above observations, few suggestions are made for future work on the disease:

a. There is an urgent need for an immediate establishment of a special research cum service cell at a suitable place (preferably at Sagar Town) to carry out further investigations and to provide therapeutic and preventive services. The cell should be staffed by an Epidemiologist, Orthopaedic Surgeon(s), Radiologist, Genetic Specialist, Physiotherapist(s), other ancillary and field staff with adequate clinical and independent transport facilities.

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b. The immediate task is to identify all the cases and mapping of the disease. Those persons in whom the disease is still in early stages, should be provided with physiotherapy and the required treatment facilities at the centre.

c. The persons who are already permanently crippled have to be rehabilitated vocationally to help them to earn their livelihood. It is also desirable that the Government should provide a suitable compensation to these individuals.

d. As one of the investigation teams has observed the disease to be associated with consanguineous marriages, the 'at risk' population in the area must be properly educated in this regard. The people should also be advised against the consumption of crabs and fish from the paddy fields.

e. There is scope for a detailed field epidemiological investigation of this mysterious disease.

Acknowledgement

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DIS 12.5

Disease that seeks out SCs in Handigodu

by N.C. Gundu Rao

HER voice is hoarse. She is clad in coarse clothes. The wrinkles on her frail face probably cannot outnumber the trials in her life.

As 60-year-old Maramma toils with her feeble hands to peel raw areca fruits, she replies to questions with a contempt that serves only to conceal her helplessness and sorrow in life.

Maramma has deformed legs, the effect of the Handigodu a wry smile: "I am not sure of passing it."

In all there are about 500 such crippled people in Handigodu and 36 other villages of Sagar Taluk who are affected by the Handigodu syndrome, a mysterious disease named after the village, located five km off Sagar, in which it occurred first in 1975. Some four cases were reported then. The village itself has got its name due to the sticky character of the soil. An adjoining village where the soil structure is loose, is called

Pain in the limb joints is the initial symptom of the disease. The victims later find it difficult to move their limbs and this leads to bow-legs and further deformities. The disease strikes during all parts of the year and at least four fresh cases are reported annually, the doctors say.

Mr. Chandrashekhar, who has conducted a socio-economic survey of the victims for the ICMR project, says that Cheluvadi and Channangi, two sub-sects among the Scheduled Castes are most susceptible to the disease. While the population of Channangi in 37 affected villages of Sagar Taluk is 2,227, there were 186 cases of the Handigodu syndrome among this sub-sect. The tally for the Cheluvadi which has a population of 354 is 87 cases.

The population and disease ratio among other communities as per the survey is: Vokkaliga: 1118-35, Uppar 236-4 and Idiga: 2731-11. The survey says that the Ramakshatriya and Achari communities are also vulnerable to the disease and claims that the disease is restricted to those who eat non-vegetarian food. The disease is not contagious and it is not noticed among children below six years, according to the survey which covered Handigodu, Bandagadde, Haregoppa, Keladipura, Lingadahalli, Karehonda, Kalasepet, Bommatthi, Beleyur, Balegody and 27 other affected villages. Studies are under way to determine if the disease is hereditary.

A MEDICAL survey which covered 45 villages in Sagar, Sorab, Hosanagar and Siddapur taluks reveals that there are eight cases of the disease among men in the age group of 0 to nine years, 52 cases among men between 10 and 18 years and 97 cases among men above 19 years.

Similarly, there were eight cases among women in the age group of 0 to nine years, 39 cases among women above 19 years. In all, 45 villages reported 364 cases and the doctors this year are still examining 11 doubtful cases, the survey reveals. The doctors have also classified 127 cases of mild deformity, 113 moderate deformity cases and 36 cases of severe deformity. Whereas the deformity on knee joints needs corrective surgeries like *genu valgus* and *genu varus*, the one on hip joints calls for *bilateral adductor tenotomy*.

Handigodu has not wiped out his smile.

Harijan women affected by Handigodu returning from work.

Dr. H.K. Srinivasa Murthy, orthopaedic surgeon of Bangalore, visits Sagar every second Saturday and Sunday of each month to perform operations on the victims. Both the operation and post-operative treatment are free as far as patients are concerned. The doctors and the social worker say that it is not possible for them to motivate over two victims at a time to undergo operations during Dr. Murthy's visit. The villagers who are scared of operations are reluctant to come to hospital though the ICMR authorities provide a vehicle at their door-step.

These people were earlier very much opposed to medical treatment since they firmly believed that this disease was due to the wrath of family deities like Chowdy and Bhoota. They used to throw away medicines given since they thought that wearing amulets was the only panacea to their illness. Now, the social workers and para-medical personnel have, to some extent eradicated these superstitions. Even then, many patients are reluctant to undergo operations since staying in the hospital for one month after the operation and a minimum of six months' physiotherapy will affect their daily earnings. Their reluctance is all the more understandable if the patient happens to be the bread-winner of his family.

Dr. Srinivasa Murthy has conducted 25 operations so far including six repeat cases. Manja and other victims who have undergone operations say that the surgery has helped improve their limb movement but there is no total relief from pain.

DR. K. Venkata Rao, a private medical practitioner, has arranged a donation of Rs. 200 per operation case through the Sai Samiti of Bangalore.

The doctors feel more help should be forthcoming from both the Government and voluntary agencies since the severe deformity among some patients needs more than one surgical operation. This means that the patients and the relatives looking after them have to stay in hospital for months together. Most patients cannot afford to stay so long forgoing their earnings, the doctors say.

Both the Zilla Parishads of Shimoga and Chikmagalur districts where the disease prevails should make joint efforts to secure aid from the Centre, the Indian Red Cross Society and even external agencies to help the victims, feels Mr. Chandrashekhar. His efforts in securing medical treatment for the victims have won accolades from many. He also suggests that the Government should start

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syndrome, a strange disease prevailing in parts of Shimoga District. The malady which has killed her two sons, Ramappa and Nagaraj, has also afflicted Nagaraj's wife Neelavva. It has not spared Nagaraj's children (Maramma's grandchildren), Manja (15) and Rama (10). Maramma's daughter Subbamma (26) who has also fallen a victim to the scourge, has been unable to marry. Maramma's husband Fakeerappa died a natural death long back. Thus her woes are endless.

With her emaciated hands and crippled legs, Maramma cannot sit for more than two hours to peel areca and thus she can earn only Rs. two a day which is hardly enough to keep the pot boiling in a house of so many crippled members. There are also days when she is too tired to come to work. Asked how she will survive those days, she says bluntly: "We have learnt to starve when there is no income."

Nagaraj (18), another crippled person in the village, is arduously managing to get around with the help of a tricycle. H.G. Manjappa (18) who is lucky to have his deformity cured substantially through surgery, is braving the SSLC examination and says with

Measures needed

THE weaving centre at Handigodu must be re-activated at once to ease the desperate plight of the Handigodu victims. Other steps that would contribute to the containment of the disease would be —

- **Shimoga Zilla Parishad must ensure that the weaving centre at Handigodu resumes its work and admits new batches of victims for training.**
- **[redacted] Government and other agencies should raise resources to supply handloom to the trained victims.**
- **Establishment of a residential workshop with an intake of 300 is desirable.**
- **Both Shimoga and Chikmagalur Zilla Parishads should join hands to demand aid from the Centre and other external agencies.**
- **Deployment of a field staff headed by an Assistant Surgeon to be in charge of the Handigodu disease cases exclusively. Construction of a separate ward with a bed [redacted]trength of 10.**

Sulagodu.

The disease afflicting the villagers is being studied by two teams of the Indian Council of Medical Research (ICMR)). The study under the guidance of Prof. S.P.S. Tiotia of the Lalalajpath Roy Memorial Medical College, Meerut and Dr. S.S. Agarwala of the Sanjay Gandhi Memorial Medical College, Lucknow, is aimed at identifying the cause of the disease and recommending remedies. The three-year research project ends after the ICMR teams submit their final report.

The disease is prevalent in Anandapura, Kasaba, Talaguppa and Avinahalli hoblies and Bharangi and Karur. The other two hoblies of the taluk are free from the disease. Handigodu syndrome cases are also reported from two villages of Sorab Taluk, three villages of Hosanagar Taluk and one village of Siddapur Taluk of Uttara Kannada District, according to field staff of the ICMR Project. The staff this year have noticed the occurrence of the disease in Devangi and Untoor villages of Tirthahalli Taluk. In all 21 cases were reported from these two villages till February this year. The doctors who confirm the prevalence of the disease in parts of Chikmagalur District, however, deny its occurrence in Indi Taluk of Bijapur District. The ICMR personnel who visited Indi Taluk recently, did not find any positive proof of the disease, the doctors in Sagar say.

THE State Government has sanctioned a monthly pension of Rs. 50 to 211 victims so far under its welfare programme for the handicapped. The pension applications of some more victims are being processed.

The authorities have also set up a weavers' training centre in the village as part of their rehabilitation programme. Both Dr. H. Srinivas, who is in charge of the ICMR project and social worker H.M. Chandrashekhar feel that training the victims in weaving is ideal since it involves a lot of exercise for the limbs. This can be a good substitute for physiotherapy which the victims require regularly.

But the centre which has trained two batches offering a monthly stipend of Rs. 200 per individual is yet to admit the third batch. The trained victims have not got any handloom either to become self-reliant. The absence of a meaningful rehabilitation programme has forced the victims to rely on the job of peeling raw areca nuts. Each victim thus engaged will get 60 paise for peeling the areca in a particular local measure, *Gidna*. Hence their daily earnings are too meagre to buy enough food to combat malnutrition which some doctors believe is one of the causes for the disease.