HANDIGODU SYNDROME IN SOUTHERN INDIA

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ABSTRACT

Handigodu syndrome is a rare and peculiar osteoarthritic disorder prevalent in Shimoga and chikmaglur in the Karnataka State, southern India. Since it was first noticed in Handigodu village in 1975. About 20% of people from 150 villages are suffering with this disease without any cure. The tragic incidence of 'Handigodu syndrome' has been attributed to be due to the 'long-term' consumption of pesticide-poisoned crabs and fish, As these persons marry only within their communities but no inbreeding was observed in the three families. Clinical presentation is based on the X-ray analysis, Deformity of flexion hip, lumbar lordosis, and pain in hip joints, difficulty in walking, truncal shortening, short stature, dwarfism, and epiphyseal dysplasia at hips, knees, hands and wrists. Lower femoral metaphysis, shortness of femoral necks and broadening, widening of the lower ends of radius and ulna, dysphatic changes shows in hip joints such as flattening of the femoral heads, cox vara and broad short necks. Surgical hip replacement therapy has indicated by the third and fourth decade of life. Life style modifications such as fresh eating habits and avoiding consumption of pesticide-poisoned crabs and fish. Marriages from same communities or from same families should be stopped.

KEY WORDS: Handigodu syndrome, flexion hip, lumbar lordosis.

INTRODUCTION

Handigodu Syndrome, also known as Handigodu Joint Disease (HJ D) is a rare and painful, peculiar of the osteoarthritic disorder prevalent in several villages of two districts viz, Shimoga and chikmaglur in the Karnataka State, southern India. This disorder name derives from the village name Handigodu, since it was first noticed in Handigodu village in 1975.
About 20% of people from 150 villages are suffering with this disease without any cure.\textsuperscript{[1]} the data obtained during subsequent multidisciplinary study by ICMR (Indian Council of Medical Research) revealed that HJD is a late onset spondylo epi (meta) physeal dysplasia.\textsuperscript{[2]}

The tragic incidence of 'Handigodu syndrome' has been attributed to be due to the 'long-term' consumption of pesticide-poisoned crabs and fish by the local population (National Institute of Nutrition 1977) and possible causes such as exposure to toxins in food or pesticides have been disproved.\textsuperscript{[3]}

**AETIOLOGY**

Communities like Channaggi and Chaluvadi in which HJD is prevalent are small, isolated caste groups who live in a localized geographical area. As these persons marry only within their communities but no inbreeding was observed in the three families. Their ethnological and nutritional status, life style, is different from higher caste people like Brahmins of the same area. Lower socioeconomic groups who are residing in the same area were also not affected. And the villages in which HJD occurred were not contiguous to each other, and likewise not all contiguous villages were affected. The communities were apparently aware of the hereditary nature of the condition and it was customary for unaffected individuals not to marry into villages with affected persons.\textsuperscript{[4]}

**Handigodu disease:** Two persons on the right are affected and two women on the Left are normal.

Clinical studies have shown that about 50% of persons in affected families (234 out of 475 studied) suffer from HJD, males and females are being equally prevalent. These
investigations have also shown three generation vertical transmission, which suggests autosomal dominant inheritance. The dwarfed individuals who had widespread changes may represent the most severe phenotypical expression of the same mutated gene or homozygosity for the mutated gene. Studies of possible environmental pathogens have yielded negative results, further pointing towards a genetic cause.\textsuperscript{[5]}

**CLINICAL PRESENTATION AND DIAGNOSIS**

HJD was classified into three sub types on the basis of X-ray analysis namely Type I (predominantly Arthritic), Type II (predominantly Dysplastic) and Type III (Dwarf). The X-ray pictures showing the marked abnormalities are depicted in the **Figure 1 and Figure 2**

![Figure 1](image_url)

**Figure 1.** X-Ray radiograph of hip joints from Handigodu Disease (HD) (A-E): The radiograph osteoarthritis in hip joints of Type I subgroup are shown in A-C whereas dysplasia of hip joints from Type II and Type III subgroups are presented in D and E respectively

The type I characterized by the late presentation of disease (45-50 years of age), by pain in hip joints, difficulty in walking and on examination these individuals had normal body proportions (without significant truncal shortening) and height, inability to sit cross legged, Deformity of flexion hip, lumbar lordosis. Radiologically these individuals showed characteristic changes of osteoarthritis at the hip joints bilaterally, without any abnormality of spine or involvement of other bones. Other Malformations are some of the feature of this syndrome. The type II was characterized by short stature, particularly truncal shortening and the main complaint was inability to walk for a longer distance. These patients were relatively younger (25-35 years of age). The X-rays of these patients showed dysplastic femoral heads
bilaterally, and varying degrees of platyspondyly. The type III was marked by dwarfism and associated with varying types of skeletal anomalies, in knees and hands, there was marked epiphyseal dysplasia at hips, knees, hands and wrists besides significant platyspondyly.\[^{3,4}\]

![Figure 2. X-Ray radiograph of Lumbar spine from Handigodu Disease (HD) (A-C): Lateral view Lumbar spine showing varying degrees of platyspondyly in Type I (A), Type II (B) and Type III (C) subgroups.](image)

Radiological changes are typically spondyloepiphyseal dysplasia plus modelling deformities of the lower femoral metaphysis, shortness of femoral necks and broadening, widening of the lower ends of radius and ulna. Dysphatic changes shows in hip joints such as flattening of the femoral heads, cox vara, and broad short necks. The changes in spine present in 75% of affected persons, were marked platyspondyly, anterior disc space widening, and posterior verbal humping.

**Phenotype of Handigodu disease**

**Hip joint involvement**

- Osteoarthritis
- Dysplastic changes
- Dysplasia spine only
- Dysplasia hips and spine with or without Involvement of other bones\[^{5}\]

The major groups of Spondyloepiphyseal dysplasia (SED), which were similar to HJD, are SED congenita, SED tarda and pseudoachondrodysplasia. A similar disease is found in South
Africa namely MSeleni Joint Disease, having clinical presentations similar to HJD. The only difference is that a posterior hump in the spine is absent and rheumatoid arthritis like changes in Metacarpophalanges were observed in MSeleni Joint Disease.\(^8\)

**TREATMENT**

In advance degenerative arthritis surgical hip replacement therapy has indicated by the third and fourth decade of life. Although surgery is not always practical, when both hips are invariably involved the best option would be total hip replacement. The techniques of surgery would differ quite clearly depending on whether there is subluxation or protrusio, as it would enable reconstructive hip surgeons to plan the most appropriate way of performing total hip replacement in these patients and showed the improved situation of Handigodu disease sufferers. Life style modifications such as fresh eating habits and avoiding consumption of pesticide-poisoned crabs and fish. Marriages from same communities or from same families should be stopped.\(^9\)

**REFERENCE**
