Bone and Joint Disorders on Tristan da Cunha

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SUMMARY

In an investigation of bone and joint disorders among the islanders of Tristan da Cunha, a total of 111 individuals aged 35 years and over were examined. These represented 87% of the population in this age group. Serological studies of rheumatoid factor and radiographic examination of the hands were carried out respectively in 74% and 70% of this adult population. The prevalence of rheumatoid arthritis and osteo-arthrosis did not differ to any great extent from that which would be expected to occur in a similar community in Europe. Classical gout was observed in one middle-aged male. Clinodactyly of the fifth finger was transmitted as an autosomal dominant trait in 12 members of the Glass family. Dupuytren's contracture, traumatic lesions and arthralgic complaints were frequently encountered.

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There is considerable geographical variation in the prevalence of the arthritides and other bone and joint disorders. These discrepancies may be due to the ethnic variations in inherited factors, or to the presence or absence of various environmental influences. Island communities are particularly appropriate for studies of situations of this type, as isolation tends to concentrate the gene pool and ensures that the environment remains relatively constant.

For this reason, a survey of bone and joint disorders has recently been carried out on the island of Tristan da Cunha, or Tristan, as it is commonly known. The clinical, radiological and serological observations which were made are presented in this article and discussed in the light of the particular circumstances present on the island.

THE ISLANDERS AND THEIR ENVIRONMENT

The island of Tristan da Cunha is situated in the South Atlantic ocean, on latitude 37° south, longitude 12° east. The nearest permanent mainland habitation is in South Africa, 2 800 km to the east. A fishing company, based in Cape Town, deep-freezes crayfish in a factory on Tristan. Company vessels call intermittently and a Government supply ship makes a twice-yearly visit to the island.

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Tristan da Cunha is a volcanic cone, 24 km in diameter, which arises abruptly out of the ocean. The 294 inhabitants all live in a village, Edinburgh Settlement, which is situated on a small area of flat land, close to a permanent spring. The houses are of stone, thatched with flax and lined with planking obtained from wrecked ships. The design is similar to that of the houses of crofters in the Western Highlands of Scotland. In recent years, all homes have been supplied with running water, electric light and a drainage system.

The island was discovered by a Portuguese admiral, Tristoa da Cunha, in 1506. However, permanent settlement was delayed until 1816, when a British Military Unit was landed. The purpose of this garrison was to prevent the use of Tristan as a base in any French attempt at the rescue of Napoleon Bonaparte, who had been exiled on St Helena. When the garrison was withdrawn, Corporal William Glass and his family stayed on Tristan. Other castaways and wanderers arrived in the years that followed, and womenfolk from St Helena were brought as wives for these lonely bachelors. At the present time, 7 families are represented on the island. These individuals are the inbred descendants of the early settlers.

During the previous century sailing vessels frequently called for fresh water and supplies. The islanders' health is said to have been good at that time, although 'rheumatic pains of the knee' had led one middle-aged male to seek the advice of a visiting naval surgeon (Greany, 1885).[1] After the opening of the Suez Canal, and the decline of sail, Tristan's isolation increased, and many months passed between visits from the outside world.[2] The community survived, but in 1961 a volcanic eruption necessitated their evacuation to England. Here they were extensively investigated,[3,4] although no particular attention was given to serological studies of the rheumatoid factor. Disenchanted with the outside world, they returned to Tristan in 1963, after the volcano had become quiescent.

Until recent years, the islanders' economy was based upon fishing and the cultivation of potatoes, supplemented by barter with passing ships. In the old days, the islanders' diet consisted almost entirely of fish and potatoes, together with meat and fat derived from sea-birds. However, a wide variety of foodstuffs is now available in the government store, and the usual diet is probably very similar to that of an average fishing community in Western Europe.

SURVEY METHODS

Travel, Accommodation and Equipment

P. B. was conveyed to the island in the *Gillian Gaggins*, a 2 000-ton vessel belonging to the Tristan Development Corporation. The return journey was made

in the Company's vessel *Tristania*. Permission for the survey to be carried out was granted by the Foreign Office, London, and the Governors of Tristan and St Helena. Hospitality was provided in the home of the island's Medical Officer.

The survey was conducted in the old Tristan hospital which had been built during World War II, and which has now been replaced with a modern building. Power and running water were available, as were basic radiological facilities and laboratory equipment, including a centrifuge. X-ray film, disposable syringes and needles, specimen bottles and other minor items were taken to the island, the surplus being left behind at the conclusion of the investigation.

Investigation Procedure

Upon arrival, courtesy visits were paid to the Governor, the Headman, and to Mary Swain, the island midwife. With their support, the project was commenced. The survey was based upon an up-to-date census which had been provided by the Medical Research Council, London. Due to limitations imposed by the lack of time, the investigation was confined to individuals aged 35 years and over.

In each instance, a case history was taken and the subject underwent a brief clinical examination, with particular emphasis on the musculoskeletal system. A 20-ml blood specimen was collected, and an anteroposterior radiograph of the hands was taken by Nancy Swain, an islander who had previously received some informal tuition in radiography. Respondents were seen at an approximate rate of 20/ day. The blood specimens were centrifuged during the evening and the supernatant serum stored in a deep freeze.

While the clinical work was proceeding, Pamela Swain called upon respondents in their houses and arranged for them to visit the hospital. Eventually, as the project was drawing to a close, the investigator paid home visits to the unwell and the unwilling. The project was terminated on the day before embarkation, and a traditional dance, attended by the majority of the islanders, was held to celebrate a satisfactory conclusion.

As the *Tristania* had extensive refrigeration facilities, transportation of the frozen sera presented no problems. During the final 2-hour flight from Cape Town to Johannesburg, the sera were packed in dry ice in a polystyrene container. Serum from each patient was used for an investigation of genetic polymorphisms in the population.[5] Further specimens were despatched by air to the laboratory of Dr H. A. Valkenburg, Medical Faculty of Rotterdam, for study of rheumatoid factors.

Rheumatoid factor was determined by the latex fixation test[6] before and after inactivation of the serum at 56°C for half an hour (LFT and LFT_{56}). This procedure destroys a thermal labile inhibitor which is active in unheated sera.[7] A human erythrocyte agglutination test (HEAT), which is a modification of the Waaler-Rose test, was also employed.[8] For the LFT a borderline titre of 1 in 640 is considered to discriminate between a positive and a negative test. For the HEAT this borderline titre is 1 in 32. A positive test in the inactivated serum (LFT_{56}) is sometimes masked by the inhibitor, and a negative result thus obtained. For clinical purposes, this latter result is used.

RESULTS

A total of 111 individuals aged 35 years and over were examined. These represented 87% of the island population in this age group. Details of the population structure and of the patients examined and investigated are shown in Table I.

Arthritides

Inflammatory polyarthritis: Rheumatoid arthritis as defined by the American Rheumatism Association criteria[9] was not encountered during the clinical examinations. However, 2 individuals had abnormalities in the radiographs of their hands which were consistent with rheumatoid arthritis, while 3 had minor changes of doubtful significance. None of them had any psoriatic skin lesions.

The only individuals in this group who had any close degree of kinship were two cousins. From the serological point of view, one woman over 75 years of age had a positive LFT, while 6 other women had a positive LFT_{56}. These represented 2,4% and 14,3% respectively, of the adult female population over the age of 35 years. None of the women were positive for the HEAT.

For the 52 men aged 35 years and over, these percentages were 7,8%, 13,5% and 1,8% respectively.

TABLE I. TRISTAN DA CUNHA POPULATION STRUCTURE AND INVESTIGATION GROUPS

Age group	Total population		Clinical examination		Serological examination		Radiological examination	
	Males	Females	Males	Females	Males	Females	Males	Females
35 - 44	20	15	20	13	18	11	14	12
45 - 54	23	18	21	14	16	13	17	13
55 - 64	11	9	9	8	8	7	7	7
65 - 74	13	9	12	9	9	8	9	7
75+	2	7	1	4	1	3	1	1
Totals	69	58	63	48	52	42	48	40
Completion rates			91%	83%	76%	72%	70%	70%

Gout: One male aged 68 years had classical gout, 3 attacks of acute inflammation in the left first metatarsophalangeal joint having occurred during the past year. These episodes, one of which was observed during the actual investigation, responded rapidly to Colchicine therapy. There was no clinical evidence to suggest underlying renal disease or blood dyscrasia in this individual. Unfortunately, blood could not be obtained for serum uric acid determination.

Degenerative Osteo-arthropathy

Heberden's arthropathy: Heberden's nodes associated with swelling and deformity in the small joints of the fingers were found in 9 females and one 69-year-old male. The presence of osteo-arthrosis in the finger joints was confirmed radiologically in 6 of the 7 of these individuals of whom X-ray films were taken. Clinical evidence of osteo-arthrosis was present in the large joints of the arms or legs in 7 of these patients. The youngest individual in this group was aged 44 years, and the oldest aged 82. The male respondent was the brother of one of the affected females. None of the others in this group were first-degree relatives.

Generalised osteo-arthrosis in the hands: Generalised osteo-arthrosis, diagnosed on a basis of involvement of 3 or more joints, was present in the hands of 3 males and 3 females aged 54 years and over. All had changes in the terminal interphalangeal joints and other small joints of the hands, in the absence of Heberden's nodes. These changes were confirmed radiologically in all 5 of these individuals in whom X-ray films were obtained. Two of these patients were siblings, while the others were unrelated.

Osteo-arthrosis of the large joints (excluding the spine, not associated with Heberden's arthropathy, clinical generalised osteo-arthrosis of the hands or secondary to trauma or deformity). Five males and 3 females fell into this category. The knees were bilaterally affected in 4 of them, while a single knee was affected in 3 individuals. One 75-year-old female had clinical evidence of osteo-arthrosis in the shoulders, wrists and knees. X-ray films of the hands had been obtained in 7 of these patients. In 4 of them, changes indicative of generalised osteo-arthrosis were present in the absence of clinical abnormalities. Three other males, aged 61, 61 and 71 years respectively, had clinical osteo-arthrosis of one hip joint. All had either clinical or radiological evidence of osteo-arthrosis in the hands.

Osteo-arthrosis of the knees, associated with genu varum: Three males and 3 females had clinical evidence of osteo-arthrosis of the knees, associated with genu varum. Three of these individuals also had clinical or radiological evidence of osteo-arthrosis in the hands. There was no close kinship among any of these individuals.

Congenital Deformities

Clinodactyly: Clinodactyly, or incurving of the fifth finger, an isolated symptomless abnormality, was observed in 8 females and 4 males. All these individuals were members of the Glass family, or else descendants of Granny Jane Laverello, the granddaughter of the community's founder, William Glass.

Dupuytren's contracture: This abnormality, of mild to severe degree, was present in 9 males, while 6 other males had gross thickening of the skin of the palms of the hands. It was possible that this latter finding represented an early stage of Dupuytren's contracture, but since flexion deformities of the fingers had not developed, precise diagnosis was not possible. There was no particular preponderance of family names among these individuals.

Miscellaneous Findings

Traumatic lesions: Two middle-aged males had unilateral unreduced dislocations of the elbow. Three patients had wrist deformity which was the sequel of an old Colles fracture, while a 45-year-old female had evidence of a long-standing tear in the gastrocnemius muscle.

Two unrelated males, aged 60 and 73 years, had hard, painless bony swellings of several years' duration situated on the dorsum of their wrists. In each instance, the swelling was the size of a pigeon's egg. In the absence of radiological evidence, a tentative diagnosis of unreduced dislocation of the carpus was made.

Arthralgic complaints: Lumbar backache was a very common complaint, and 14 males and 4 females had experienced this problem for at least one year. Of these individuals, 6 males and 2 females had a history and clinical stigmata which were compatible with a diagnosis of prolapsed intervertebral disc. Three males and 3 females gave a very definite history of persistent muscular pains in the absence of signs and symptoms relating to the joints. These episodes of 'fibrositis' were usually associated with activity in the potato patches or in the fishing boats.

DISCUSSION

Due to their isolation, mid-ocean islands tend to have small, stable populations which are often inbred. The environment to which these islanders are exposed is usually limited in scope and consistent in quality. On this basis, oceanic islands are appropriate situations for studies of disease processes which depend upon the interaction of environmental and genetic factors.[10,11]

Tristan da Cunha fulfilled these criteria of isolation, inbreeding and stability. Furthermore, the compactness of the community, the availability of hospital accommodation and the absence of any language barrier, all facilitated the investigation. As the islanders had been investigated during their stay in Britain, a considerable body of background information had already been collected. For all these reasons, Tristan da Cunha was a suitable locality for the survey. However, although a substantial proportion of the adults in the community were investigated, the fact that the total population was relatively small reduced the value of the results with regard to comparability with other surveys.

The prevalence of rheumatoid arthritis in different parts of the world, based upon the American Rheumatism Association criteria, varies between 2% and 10%. In view of the comparatively small number of individuals in the Tristan survey, it is impossible to ascertain the exact prevalence, but the apparent infrequency of clinical rheumatoid arthritis might have been due to chance. The 2% prevalence of erosive changes which were observed in the hand radiographs is similar to that found in Great Britain. The serological findings in terms of the tests for rheumatoid factor were unremarkable, and the prevalence and age-sex distribution of the positive tests on Tristan were similar to those of other White populations. On a basis of these findings it is probable that the true prevalence of rheumatoid arthritis on Tristan da Cunha is in no way distorted by the particular genetic and environmental circumstances which are present on the island.

The occurrence of Heberden's nodes in association with generalised osteo-arthrosis in 10 individuals represents a prevalence of 9% of the adult population. If the 6 other individuals with generalised osteo-arthrosis of the hands in the absence of Heberden's nodes, are included, the prevalence is even higher (15%). It has been suggested that Heberden's arthropathy might be due to an autosomal dominant gene which is only expressed in females. None of the affected females were first-degree relatives, and there was no obvious genetic background to this arthropathy on Tristan. As the condition does not usually become clinically evident until middle-age, it could not be investigated in their offspring.

It has also been postulated that Heberden's arthropathy is only encountered clinically in males who are homozygous for the gene.[12] The observation that only one male on the island was affected certainly supports the contention that there is indeed a marked discrepancy in prevalence between the sexes. From the genetic point of view, it is noteworthy that one of the other affected females was this individual's sister. However, if radiological osteo-arthrosis in the absence of Heberden's nodes is also considered to represent Heberden's arthropathy, then 8 other males fall into this category, and the marked sex difference is no longer apparent.

The incidence and anatomical distribution of osteo-arthrosis of the large joints were unremarkable. However, 2 of the 3 elderly males with unilateral osteo-arthrosis of the hip joint were moderately disabled, and they would have been likely candidates for surgery, if such facilities had been available.

Six middle-aged individuals had a moderate degree of genu varum. This did not cause any great disability, although all of them had evidence of osteo-arthrosis in the knee joints. The deformity could have been secondary to this osteo-arthrosis but it was also possibly the result of childhood rickets, since the islanders' diet had been relatively poor in the past.

Lumbar backache and prolapsed intervertebral disc were the most common and troublesome musculoskeletal problems which were encountered. The islanders' daily lives involve intermittent bouts of strenuous work in the potato patches and fishing boats, and it seems likely that the back problems were the result of strains and injuries which were sustained during these activities. The 'fibrositis' which was encountered may have had a similar aetiology.

The middle-aged male with gout was of considerable interest. There was little doubt about the diagnosis, and no clinical evidence of any underlying renal or haematological disorder. Gout is thought to have a polygenic hereditary background and probably requires appropriate environmental conditions for the appearance of clinical stigmata.[13,14] In view of the family structure of the population, it is likely that any abnormal genes would be widely distributed throughout the community. The gouty individual was in no way different from his fellows in terms of his diet and daily life, and there seems to be no reason why he should have been exposed to any unique environmental factors. It is difficult, therefore, to explain the development of gout in this single patient.

Clinodactyly of the fifth finger was present in approximately 11% of the survey population. This deformity is inherited as an autosomal dominant trait,[15] and it is evident that the gene was present in the Glass family. Granny Jane, who had transmitted the condition to several of her descendants, was of the opinion that her forebears also had been affected. Because she was the granddaughter of William Glass, the founder of the community, the gene could have reached Tristan at the time of the first colonisation.

Dupuytren's contracture is usually considered to be an autosomal dominant trait, of variable penetrance, which is expressed particularly in males.[16] Alternative theories implicate manual work and liver disease as aetiological factors. Although Dupuytren's contracture (14%) and palmar thickening (10%) were found in 24% of the adult males on Tristan, there was no firm evidence of any genetic background and an environmental agent might have been responsible. The constant trauma sustained by the hands during digging in the potato patches and handling small boats seems to be the most likely explanation.

The wrist deformities and unreduced elbow dislocations were the legacy of traumatic events which occur frequently on Tristan. Both elbow injuries had been sustained during the yoking of an ox which had been used some years previously for drawing heavy loads. This health hazard was removed from the community when the ox featured as the main participant in a feast which was held to celebrate its replacement by a tractor!

Although musculoskeletal problems of one sort or another were very common on Tristan, the islanders had a particularly stoical attitude to their disabilities, and accepted them as a part of everyday life. Indeed, an inquiry after the health of even the most incapacitated individual, using the Tristan vernacular, 'How you h'is?' would invariably elicit the cheerful reply, 'I's wery well.'

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