

# An endemic neurological disorder in tribal Australian aborigines

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**SUMMARY** Thirteen, and possibly sixteen cases of neurological disorder have been identified in a population of approximately 1100 tribal aborigines living in Groote Eylandt and the adjacent mainland. There were two relatively distinct clinical pictures: one coming on in childhood involved the motor system, the patients often having remarkably lax ligaments; and the other, generally of later onset, comprising cerebellar, upper motor neurone and sometimes supranuclear ophthalmoplegic features. There was some evidence that the two syndromes are varieties of a single condition. No causal factors were identified but there were indications that the disorder might be genetically determined. Attention is drawn to the similarities between this disorder and other ethnic-geographic isolates, particularly the ALS-Parkinsonism-dementia complex of Guam.

This paper reports an endemic neurological complex occurring in the Australian Aborigines living on Groote Eylandt and the adjacent Arnhem Land area of Northern Australia (fig 1). A frequent feature of the condition is involvement of the upper and lower motor neurones. This is present in rather more than half of the patients. In some it produces a teetering gait, leading to their sobriquet "bird people" amongst the Aboriginal tribesmen, who find some resemblance to wading birds such as the heron (fig 2). In other patients cerebellar and ophthalmoplegic features are more prominent and the occasional patient shows evidence of dementia.

This neurological complex may be classed as an ethnic-geographic isolate, a term suggested by Gajdusek<sup>1</sup> to describe those diseases, usually rare, occurring with a high incidence in remote areas. Six ethnic-geographic neurological disease isolates have been identified in the Western Pacific zone all sharing some clinical features though with important differences. These are the endemic neurological disorders occurring in Guam and in Saipan, both islands in Micronesia, those in the Kii Peninsula of Honshu, Japan, those in the southern coastal lowlands of West Irian, Kuru in the Fore people of the Highlands

of New Guinea and now, the Arnhem Land disorder reported in this paper.

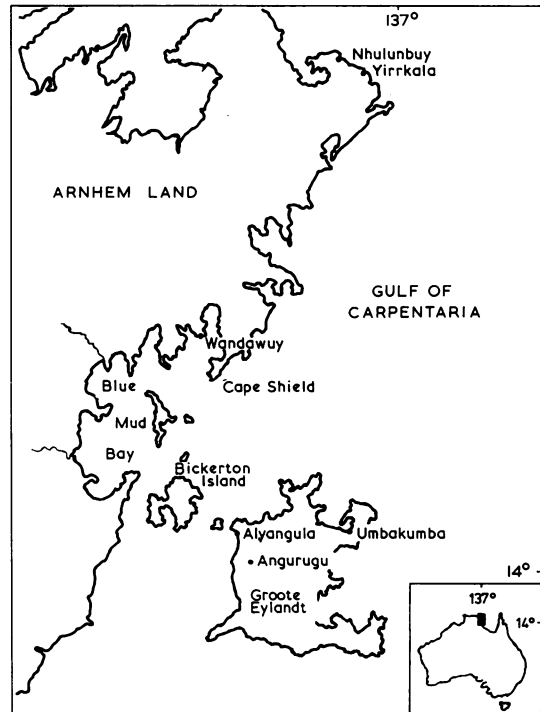


Fig 1 Area of study, Groote Eylandt/Arnhem Land Region of Northern Australia.

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Accepted 21 February 1980

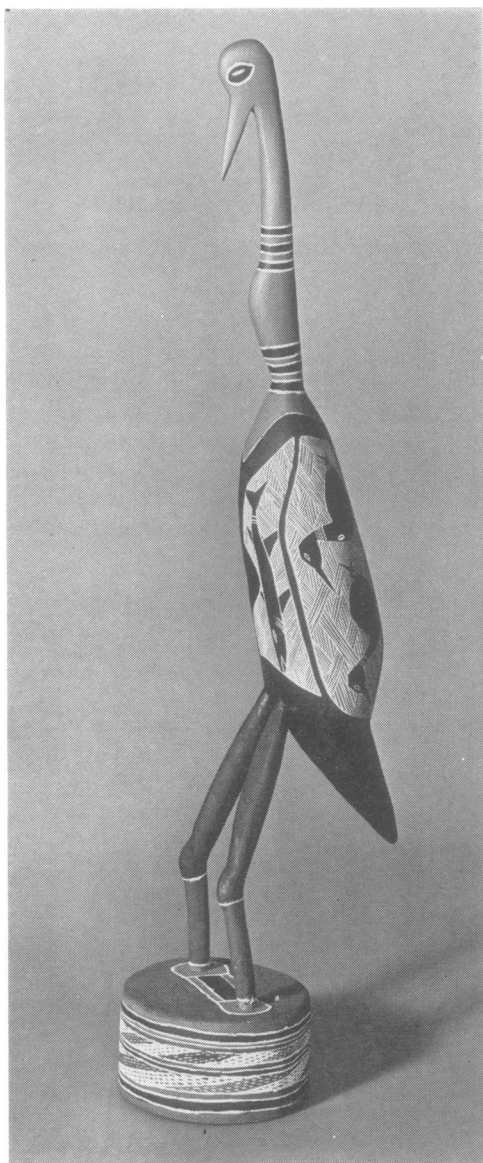


Fig 2 *Aboriginal wood carving of the heron.*

The communities in which all six of these neurological ethnic-geographic isolates occur each have a human ecology completely different from that of Western society. In addition to the social peculiarities arising from their isolation they have recently each been massively disrupted by changes imposed from the outside world. The human ecology and ethnology of the Arnhem Land complex are reviewed in detail in a separate publication by Cawte.<sup>2</sup>

## Patients

### *The Arnhem Land Neurological Cohort*

The locality map shows the three small communities in which the affected individuals live or lived—Angurugu and Umbakumba on the west and east sides of Groote Eylandt respectively, and Wandawuy near the shores of Blue Mud Bay on the Australian mainland. The population of Groote Eylandt is approximately one thousand and that of Wandawuy one hundred. Details of the identified cases are given below; the principal features are summarised in table 1. No names or initials are given, in order to preserve the privacy of the patients concerned. To save undue repetition it can be stated that in no case was there any sensory impairment, nor were fasciculations noted.

*Case 1* Female, b 1968. This girl is the youngest of seven siblings. She had a normal infancy but her gait was noted to be abnormal when two years old. She now walks in a stilted and slightly broadbased fashion on her toes with knees bent and her left leg internally rotated. She has an obvious pes planus. Examination showed slight wasting of the intrinsic hand muscles but she retained good power. The arms were hypotonic with lax ligaments and the deep reflexes reduced. There was marked wasting below the knees with weakness of hip flexion and dorsiflexion of the ankles. Spasticity was evident at the knees with increased knee jerks but her ankles were hypotonic with hyporeflexia. Both plantar responses were considered extensor in 1977 but although there was a right Babinski reflex on a recent visit, no response could be obtained on the left side. Although her mother denies any deterioration in walking there has been a progression in the distal wasting since 1977.

*Case 2* Female, b 1961. Some slowness of movements was noted by her mother soon after birth. At the age of three years she could stand but not walk, and by four years she was able to walk but only feebly. Her gait and neurological signs were similar to those of her sister (Case 1) though rather more marked. The upper limbs showed flaccidity and there was moderately severe wasting of the intrinsic hand muscles. Her legs were generally weak with obvious spasticity. The superficial abdominal reflexes were retained. The deep reflexes in the upper limbs were difficult to elicit except for the finger jerks which were brisk. The knee jerks were exaggerated, the ankle jerks absent and both plantar responses were extensor.

*Case 3* Male, b 1938. The brother of Cases 4 and 5 and a half brother of Cases 10 and 15. In his records at the Mission Health Centre it was noted

Table 1 Summary of cases

Case	Year of birth	Sex	Duration (yr)	Residence	Severity	Clinical features	Joint and ligament abnormalities
1	1968	F	9	Angurugu	moderate	UMN: LMN	present
2	1961	F	17	Angurugu	moderate	UMN: LMN	present
3	1938	M	28	Angurugu	moderate	LMN	present
4	1940	M	7	Angurugu	moderate	Oc. Cer. UMN	—
5	1943	M	6	Angurugu	mild	Oc. Cer. UMN	—
6	1945	F	15	Angurugu	moderate	UMN: LMN	—
7	1958	M	8	Angurugu	moderate	Cer; UMN; Dem	—
8	1960	M	18	Umbakumba	mild	UMN: LMN	present
9	1962	F	16	Umbakumba	mild	UMN: LMN	present
10	1939	F	18	Angurugu	severe	LMN; Bulbar; d.1931	—
11	1937	M	23	Wandawuy	severe	Oc; UMN; ? LMN: Cer.	—
12	1957	M	2	Wandawuy	mild	?Cer (no details)	—
13	1960	M	3	Wandawuy	mild	?Cer (no details)	—
Dubious cases							
14	1936	F	?	Angurugu	slight	?UMN	—
15	1956	F	?	Angurugu	—	Nil	present
16	1967	F	?	Angurugu	slight	LMN	present

UMN=Upper motor neurone;  
Cer =Cerebellar;

LMN=Lower motor neurone;  
Dem =Dementia

Oc=Ophthalmological;

in 1951 that he had a kyphosis, said to be congenital, and an abnormal gait described as "floppy as if his ankle joints were lax". Wasting of the "palm of his right hand" was noted in 1967. A chest x-ray in 1972 showed giant air cysts in the lungs. Although the patient denied any difficulty in walking he walked slowly and was unable to run. There was a mild to moderate degree of distal wasting in the upper and lower limbs with marked hypotonia and laxity of ligaments together with pes planus. There was also slight wasting of the quadriceps muscles. When first seen in 1977 the knee and ankle jerks could be elicited on the right side but not on the left; all were absent on the recent visit. The plantar reflexes were flexor.

**Case 4** Male, b 1940. The brother of Cases 3 and 5 and an important personage in the mission settlement. His main complaints were of unsteadiness of gait, poor balance and weakness and stiffness of his legs. These symptoms began in 1972. He felt that his trouble was quite different from that of his brother (case 5) for whereas he felt "stiff", his brother was "slack". On examination the muscles of his right shoulder girdle and right arm were less well developed than those on the left but he said this had been present since early childhood and he attributed it to a crocodile bite. It caused him no symptoms. He showed a well sustained bilateral nystagmus with only limited superior conjugate eye movements (this latter feature was not noted in 1977), a defect of convergence and a mild posterior internuclear ophthalmoplegia. There was a typical cerebellar syndrome of moderate degree. Tone and the deep reflexes were increased in the lower limbs. The right plantar response was extensor and the left dubiously so. This patient has since been seen by Dr RH Rischbieth in Adelaide who confirmed the clinical observations and noted that his EEG was normal. A

CT scan showed some cortical atrophy. The lateral and third ventricles were normal in size but the fourth showed slight enlargement. There was definite evidence of cerebellar atrophy with shrinkage of the cerebellar folia.

**Case 5** Male, b 1943. The brother of Cases 3 and 4. He first noticed unsteadiness on walking six years ago, and now walks with a slightly broadbased ataxic gait. He had a bilateral horizontal nystagmus with a posterior internuclear ophthalmoplegia and some limitation of superior conjugate ocular movements. There was no muscle weakness or wasting, but there was a mild degree of spasticity in the lower limbs with increased deep reflexes, an extensor plantar response on the right and an equivocal one on the left. Signs of a bilateral symmetrical cerebellar syndrome were present.

**Case 6** Female, b 1945. In 1961 this patient developed a polyarthritis. In 1964 she was admitted to Darwin Hospital and a diagnosis of rheumatoid arthritis was made. She was noted to have weakness and wasting of the muscles of the left arm and forearm. In 1967 some wasting of the hands was noted with contractures of the left fourth finger and right fifth finger. On examination the contractures were confirmed but only mild arthritic changes were evident. There was wasting and weakness of all upper limb muscles, particularly involving the deltoids and the muscles of the upper arms. The left biceps and triceps and brachioradialis reflexes were markedly increased. The left finger jerk was brisk and the jaw jerk was particularly active. There was no abnormality in the lower limbs and both plantar reflexes were flexor.

**Case 7** Male, b 1958. A nephew of Cases 3, 4 and 5 and the only son of Case 10. At the age of 13 years

he was noted to fall easily and complained of weakness of his legs. Children called him "Slacky" because of his loose-limbed slouching gait. In the last few years he has deteriorated. His speech has become slurred, his movements slow and he drags his feet. On examination his gait was a little unsteady. He showed a bilateral horizontal nystagmus but no other ophthalmological abnormalities. There was mild weakness of the intrinsic hand muscles without wasting. In the lower limbs there was a moderate degree of spasticity with increased deep reflexes and bilateral extensor plantar responses. Bilateral cerebellar signs were present and he showed a moderate degree of dementia.

*Case 8* Male, b 1960. His mother said he had always been small in comparison with most of his brothers and sisters. Standing and walking were delayed and when achieved he moved laboriously with his legs slightly flexed at the knees and plantar flexed at the ankles. Eventually he was able to run. When seen in 1977 his upper limbs were normal but on the recent visit there was weakness and wasting of the intrinsic muscles of his hands and of the flexors and extensors of the wrists. In the lower limbs there was wasting of the lower legs, particularly of the tibialis anterior muscles with hypotonia. The deep reflexes were sluggish but the plantar responses were extensor. His joint ligaments were remarkably lax, particularly those of his wrists and ankles.

*Case 9* Female, b 1962. The sister of Case 8 and like her brother was slow to crawl, stand and walk. She too is undersized for her age and has pes planus. On examination her gait was more severely affected than that of her brother. She walked poorly with a "tip toeing" manner, her knees flexed and her ankles plantar flexed. There was wasting and weakness of the forearms. There was some wasting of the thenar eminences but the hands were not affected as severely as the forearms. There was some clawing of the fingers. Distal wasting and weakness were also marked in the lower limbs. The biceps and triceps jerks were just obtainable but the brachioradialis reflexes were absent. The knee and ankle jerks were normal. The plantar responses were flexor. There was very marked hypotonicity with laxity of the joint ligaments.

*Case 10* Female, b 1939. The details of this patient who died in 1970 at the age of 31 years, were obtained from her case records; she was not seen by the authors. She was the mother of Case 7. In 1952 at the age of 13 years she complained of pain in the chest and difficulty in breathing. A "paralysis of the right arm" developed about the same time. She remained small for her age but developed no other unusual features until she was 19 years when it was noted that during the previous few months her gait had become peculiar and that she tended to drop things. When 22 years old she again experienced episodes of difficulty in breathing and swallowing.

By 1964 when aged 25 years, she had deteriorated, having become emaciated and in need of constant nursing care. She was noted to have wasting of her limbs and a dysarthria. In 1965 she developed pressure sores and cystitis. Finally she was unable to speak but could comprehend questions and could respond by weakly pressing her hand. Food and fluids sometimes regurgitated through her nose when she attempted to swallow. Before her death in 1970 she became incontinent of urine and had multiple bed sores.

*Case 11* Male, b 1937. When aged 19 years (in 1956) the patient complained of "contraction" of the finger joints of both hands and wrists. They felt stiff, without feeling, although on examination there was no sensory loss. He said it was more comfortable to hold his hands in a contracted position. He was recorded as having an abnormal gait, throwing out his right leg with each step. In 1969 his gait was again noted to be abnormal and it was described as broadbased. The tone and power in his legs were considered to be normal but his knee jerks were brisk. His ankle jerks were not elicited but the plantar responses were extensor. It was considered that he also showed features of early Parkinson's disease. He was thought to be mentally dull and was described as lethargic and slow to follow instructions. By 1971 he could walk only a few steps and would stagger, falling to either side. In 1975 he had a suprapubic prostatectomy. He was at times aggressive in hospital, attacking the staff. Later that year he was admitted to Gove Hospital where a diagnosis of Parkinson's disease and dementia was made. In 1977 he fractured his right forearm while riding on the back of a truck. It was plated and bone grafted from his left iliac crest, but because he kept removing the plaster it failed to unite and a pseudoarthrosis formed. On examination no real evidence of cognitive defect or memory loss was found. He was co-operative and alert, though from his history a personality change was considered probable. There were gross neurological deficits. He could not stand unaided. He had to be helped to his feet, losing his balance and falling backwards when he tried unaided. Supported on either side he could walk slowly with a reeling gait, lifting his knees and circumducting his legs. There was an intermittent tremor of his head and lips and at times a pill rolling tremor of his hands. There were severe defects of convergence and superior conjugate movement of the eyes, with an anterior internuclear ophthalmoplegia and an ataxic nystagmus. There was slight thenar wasting with some hypotonia evident during movements of the finger and wrist joints. Power and co-ordination appeared to be normal though hard to test in the right upper limb due to the effects of the fracture. The deep reflexes were present and brisk. The abdominal reflexes were present. His legs were thin, presumably due to lack of use, but there was no gross weakness. Both limbs were spastic. There was clonus at the knees but only occasional clonus at the

ankles. The knee jerks were increased and the ankle jerks were normal. Both plantar reflexes were extensor. Co-ordination was grossly impaired, especially on the left side.

*Case 12* Male, b 1957. The son of Case 11 but not seen because he was out in the bush. His father said that he suffered a similar complaint to his. He had grown up normally around Caledon Bay but two years ago he began to have difficulty in walking and occasionally fell down.

*Case 13* Male, b 1960. The son of Case 11 but not seen. Mission health records showed that he was well until he was 16 years old when his gait began to deteriorate. A recent note indicated that he was unable to hop or jump from foot to foot. All reflexes were brisk, especially those in the lower limbs with clonus of both ankles. The plantar reflexes were not mentioned. Sensation was recorded as intact. It was reported that two other offspring of Case 11 and a paternal aunt are affected but these persons have not been examined and no records are available.

#### *Possible Cases*

*Case 14* Female, b 1936. The mother of Cases 1 and 2. She denied any disability but although she was able to sit up from a supine position she required some assistance to stand from a squatting position and had obvious difficulty in attempting to jump off the ground. There was no muscle wasting but there was slight weakness of hip flexion and ankle dorsiflexion. The knee jerks were increased as were the adductor reflexes but the plantar reflexes were flexor. It was concluded that she probably had a slight upper motor neurone disturbance.

*Case 15* Female, b 1956. There was a history of delayed walking. She is markedly undersized and could well pass for a child of 10 years. She is one of eight siblings, the others all being normal. She has marked pes planus. On examination her forearms and legs were very thin with guttering between the radius and ulna on each side. Marked hypotonia was present in the upper and lower limbs with marked laxity of joint ligaments. Her thinness and lack of strength could easily be interpreted as wasting and weakness but this remains uncertain. The deep reflexes were normal and the plantar reflexes were flexor.

*Case 16* Female, b 1967. This girl is the third of six siblings, the others being normal. She was slow to stand and walk but eventually was able to walk and run competently. She has marked pes planus. On examination her forearms and legs were very thin with marked hypotonicity and lax ligaments. There was a definite wasting of the left thenar eminence but as in Case 15 whom she resembles closely, it is debatable whether the other muscle groups were wasted or just thin. The biceps and brachioradialis reflexes were just obtained but the

triceps jerks and the deep reflexes in the lower limbs were normal. The plantar reflexes were flexor.

#### **Laboratory investigations**

The data presented in this paper were gathered in the course of two field visits separated by a little over one year. In the majority of cases little more than history taking and clinical examination were possible. However, four of the patients (Cases 2, 3, 8 and 9) were admitted to the Prince Henry Hospital in Sydney and a number of investigations were carried out.

In nerve conduction studies the sural and median nerves were examined in each patient and the posterior tibial nerve additionally in Case 3. Except for mild abnormalities of the right median nerve in Case 2 and the right sural nerve in Case 3, all conduction values were within normal limits. Electromyograms were obtained in Cases 4 and 8. Neurogenic weakness was evident in the former whilst in Case 8, although the action potentials were of normal form, the interference pattern was slightly reduced (table 2).

Small-bowel biopsies were carried out on Cases 2 and 3 who were considered co-operative enough and able to tolerate the necessary intubation procedure, in order to investigate the possibility of a malabsorption syndrome. Microscopy in Case 2 revealed a partial villous atrophy with an increase in round cells and neutrophils in the lamina propria; examination of the surface epithelium also revealed an increased number of round cells. In Case 3 there was a mildly inflamed duodenal epithelium with irregular or partial villous atrophy. In neither case was the atrophic defect serious enough to suggest that a malabsorption problem might be part of the aetiology. Case 3 showed abnormal red cells on blood examination consistent with foliate deficiency. All other routine investigations were within normal limits. Case 4 was investigated at a later date in Adelaide by Dr Rischbeith.

#### **Discussion**

The questions of greatest interest are firstly whether the neurological syndromes identified in the Arnhem Land cases can be grouped as a single entity or whether they comprise two or more distinct conditions; and secondly to what extent genetic and environmental factors are involved.

The nosological question is probably unanswer-

Table 2 Nerve conduction studies

<b>Case 2</b>		<b>Case 8</b>	
Rt sural nerve	SAP 24 $\mu$ V: 48 m/s	Rt sural nerve	SAP 19 $\mu$ V: 52 m/s
Rt median nerve	SAP 32 $\mu$ V: 45 m/s	Rt median nerve	SAP 34 $\mu$ V: 54 m/s
	MNAP 24 $\mu$ V: 63 m/s		MNAP 65 $\mu$ V: 60 m/s
	Terminal latency 5.0 ms		MCV 62 m/s
			Terminal latency 2.9 ms
<b>Case 3</b>		<b>Case 9</b>	
Rt sural nerve	SAP 6.5 $\mu$ V: 41 m/s	Rt sural nerve	SAP 19 $\mu$ V: 44 m/s
Rt post tibial nerve	MCV 42 m/s	Rt median nerve	SAP 42 $\mu$ V: 58 m/s
	Terminal latency 6.2 ms		MNAP 84 $\mu$ V: 63 ms
R median nerve	SAP 16 $\mu$ V: 53 m/s		MCV 54 m/s
	MNAP 41 $\mu$ V: 55 m/s		Terminal latency 3.0 ms
	MCV 58 m/s		
	Terminal latency 4.5 ms		
SAP —Sensory action potentials		Minimum normal values for subjects up to 60 years of age in this laboratory:	
MNAP—Mixed nerve action potentials		sural nerve SAP 7 $\mu$ V: 40 m/s	
MCV —Motor conduction velocity		median nerve SAP 10 $\mu$ V: 42 m/s	
		MNAP 54 m/s	
		Terminal latency 5.0 ms	

*EMG Studies*

<b>Case 3</b>		
Rt abductor pollicis brevis	AP mildly enlarged and polyphasic	
Rt extensor digitorum communis	IP mild reduction	
Rt tibialis anterior	AP mild—moderately enlarged and polyphasic	
Rt abductor hallucis	AP marginally abnormal	
<b>Case 8</b>		
Rt tibialis anterior	AP normal form	
	IP slightly reduced	

AP—Action potentials  
IP—Interference pattern

able at present. On clinical grounds there could be at least two syndromes, one taking an amyotrophic form with either or both lower and upper motor neurone phenomena and the second characterised by cerebellar, upper motor neurone and supranuclear ophthalmoplegic signs. The later age of onset of this latter group might be considered to support a nosological distinction. On the other hand, the fact that of three affected brothers (Cases 3, 4 and 5) one showed the amyotrophic and the other two the cerebellar form, suggests a single polymorphous condition.

That groups of disparate syndromes may have a single aetiology is well recognised. The wide range of neurological disorders due to syphilis and the numerous conditions associated with vitamin B12 deficiency are examples.

Perhaps more relevant—geographically at any rate—are the cases of amyotrophic lateral sclerosis (ALS) and parkinsonism-dementia (PD) on Guam. When ALS was identified on Guam there seemed to be no immediate diagnostic problem for the clinical features were identical to those of a world wide disease. It was only when its remarkable frequency was appreciated that its importance became apparent. Later,

subtle neuropathological differences were demonstrated.

The relationship of ALS to PD on Guam is still a matter for argument but Kurland and his colleagues<sup>3</sup> are convinced that they are facets of the same disorder. Of those with the PD complex 20 per cent developed ALS sooner or later and 10 of the 321 patients with ALS had features of dementia or extrapyramidal disease. Particularly in the PD cases considerable variation may occur in the picture and some cases show extrapyramidal features only. Such groupings at least suggest the possibility that the Aboriginal cases comprise a single disease entity.

Another area of uncertainty is created by the Aborigines with no definite neurological signs who share with some of the amyotrophic cases the striking abnormality of lax ligaments and hypotonia. Support for the possibility that these may suffer a *forme fruste* of the disorder is provided by case 8 who, when first examined, showed only lax ligaments but on the recent visit was found to have definite muscle wasting and extensor plantar responses. In this respect too there are parallels with the Guam cases in which, as in ordinary motor neurone disease,

there are often dermal changes characterised by an increase in mucopolysaccharides, elastosis and disorganisation of colloid structure. Skin changes are present in about one half and dyschondroplasia is common.

A small series of patients with MND, some with spastic paraplegia, has been noted in the island of Saipan which, like Guam, is in the Marianas. There were six male patients in a population of 2000. No new cases have appeared since 1960. The symptoms progress slowly if at all.<sup>4</sup> Another high incidence area of MND has been located in the Kii peninsula, a remote region of Honshu, in Japan.<sup>5</sup> Of 18 cases, 14 showed a picture of primary lateral sclerosis. Gajdusek<sup>6</sup> described a number of cases similar to those seen on Guam in the Anya and Jakai people of the coastal lowlands of West Irian across the Arafura Sea from the coast of Arnhem Land. All cases occurred in an area populated by some 5000 individuals; no cases were detected in the surrounding area although many of the inhabitants were Anya people. Twenty-four cases of ALS, four cases of parkinsonism without ALS and one case of "myoclonic dementia" were identified. Some of the 24 ALS patients also had extrapyramidal features and two in addition showed dementia.

At first sight Kuru would seem to have little relevance to the Arnhem Land cases. Gajdusek and Gibbs<sup>7</sup> however, have suggested that the Kuru virus may have originated from a single patient with Creutzfeldt-Jakob's disease (CJD) who established appropriate contact with the Fore people in the Highlands of New Guinea and that the CJD virus in turn was developed from the scrapie virus. CJD is another condition with a variety of manifestations including amyotrophic and cerebellar forms although the majority of cases show upper motor neurone features.

### **Genealogy**

In the companion anthropological paper<sup>2</sup> the traditional marriage and kinship patterns of this ethnic group are presented in detail but some features of possible relevance to the disorder described here may be reviewed briefly. This tribal group has firm kinship rules binding upon people who were too "close" to marry. In general, the rules decreed that if a man's father and a woman's mother belonged to the same local group and were of the same generation level, they were too close to marry. It was believed that the offspring of couples who married in defiance

of these rules would be deformed or sickly. In general therefore, women did not marry men of their own generation; an older man would commonly have several younger wives who eventually transferred to other husbands, usually when he died or was not vigorous enough to keep them, or gave them up in return for services received. In this way, any women might well expect to change husbands several times in her lifetime.

In recent times tribal marriage rules have been precipitously disrupted and many women have acquired young husbands and abandoned polygyny. Some marriages have become too "close". To the Aborigines the white kinship rule seemed to be that any man and woman could marry as long as their parents were not siblings. By following white example, marriage within the same generation or age group became permissible, and to the younger members, both male and female, more desirable. The Aborigines were not to realise that the white marriage rule is safe because whites operate within a much larger gene pool. They argue, though without solid evidence, that their present neurological disorders have appeared only since their traditional kinship safeguards were abandoned and consequently they are inclined to blame the white marriage rules, including the practice of monogamy, for their occurrence. There are some signs of resuming the traditional polygyny/gerontocracy which was the former basis of society but this is not likely to become widespread for westernisation is too far advanced.

If, as seems likely, a genetic factor is important, the history of the local groups in this region suggests the likelihood that Bickerton Island, the "bridge" from Groote Eylandt to the mainland for this canoeing people, may hold the key to the genetic understanding of this disorder. Former Bickerton inhabitants now reside largely on Groote Eylandt which contains the majority of cases of the neurological disorder. If this disorder is in fact transmitted as a Mendelian dominant, we are led to consider the hypothesis that a gene mutation occurred on Bickerton Island or the adjacent mainland and was "exported" by women marrying into the nearby population on Groote Eylandt or by the mass population movement to Groote Eylandt that occurred when missions were set up there.

### **Environmental factors**

Our initial reconnoitre of this condition in Arnhem Land revealed a number of features of

the ecosystem that were potentially toxic to the human nervous system. It seemed possible that one or more of these factors, acting as a sufficient cause, or acting upon genetically susceptible individuals, might be incriminated.

The fairly exhaustive enquiry that was conducted into potential noxae in the ecosystem is described in detail by Cawte.<sup>2</sup> Groote Eylandt has large manganese deposits, mined since 1962 by open-cut methods, and a considerable portion of the population lives in close proximity to a dusty dump of manganese ore. Cycad nuts are one of the staple foods of traditional life, and at one of the early mission settlements, cassava became a staple food. Even up to the present time clay cakes are consumed for medicinal and other reasons. Each of these factors has in other settings produced toxic effects upon the human nervous system, although none closely resembles the syndromes here described. No evidence was found suggesting that any of these potentially toxic agents had a role in the neurological disorders described.

Although protein, iron and folic acid deficiencies all occur in the population group there was no indication that any of these was of significance in regard to the neurological disturbances. Alcohol, which in any case is not easily acquired in this particular community, appeared to be excluded by the age range of the patients as well as the nature of the disorders.

### Conclusions

The occurrence of a high incidence of neurological disease in a relatively small and isolated community with a rather stable gene pool but changing cultural patterns, may offer an opportunity for advancing the understanding of the aetiology and pathogenesis of degenerative nervous system disorders, as in the case of the ALS-parkinsonism-dementia complex of Guam. The differences in the Arnhem Land neurological disorders may or may not reflect different aetiologies. It is possible that different populations have different susceptibilities, probably genetically determined, to a variety of environmental hazards.

The data in relation to the neurological disorders which we have seen on Groote Eylandt and the adjacent part of the mainland suggest that genetic determinants are likely to be important, although environmental factors are also likely to be involved. We do not have evidence to incriminate any particular mode of inheritance, although there are suggestions of a dominant

autosomal gene. Nor can one make a firm decision about the number of disease entities suffered by our patients, but the most reasonable choice would seem to lie between two separate entities and a single polymorphous condition.

The Aboriginal people of Groote Eylandt were generally welcoming and many individuals assisted us in the study of this health problem. The Angurugu Council and the Church Missionary Society each gave us every possible help and courtesy. The various officials of the East Arnhem Land Branch of the NT Health Department, especially Dr Stanley Linco, who initially invited us to make the study, gave us full practical support. We are equally indebted to the nursing sisters of Groote Eylandt. Some of the patients described in this paper had been seen previously by Professor J McLeod and Dr J Hargrave. We are grateful to both and to Dr R Rischbieth of Adelaide for allowing us to make use of their clinical observations.

The study was supported by a grant from the National Health and Medical Research Council.

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