

Rheumatic Fever In Micronesia

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Abstract

Rheumatic fever is an important health problem in Micronesia. A retrospective review of hospital discharges for the years 1986 through 1988 was conducted in Yap, Chuuk, Pohnpei, and Kosrae states of the Federated States of Micronesia for all patients coded as acute rheumatic fever (ICD 390-392.9) and rheumatic heart disease (ICD 393-398.9). Suspected cases were identified from a variety of sources and the diagnosis was confirmed utilizing the Modified Jones criteria and WHO modifications. The annual incidence of acute rheumatic fever was found to be 10.4/100,000 over that three year period. Forty-two cases of rheumatic heart disease were identified. A registry of cases would allow more accurate evaluation of the extent of acute rheumatic fever and rheumatic heart disease and facilitate better secondary prophylaxis. The goal of primary prophylaxis is difficult but attainable through education of the health workers, utilization of lab. testing, and community awareness.

Introduction

Acute rheumatic fever (ARF) is the most serious nonsuppurative complication of upper respiratory tract infection by group A beta-hemolytic *Streptococcus pyogenes*. Although the incidence of ARF has generally decreased in developed countries, the disease is still a major Public health problem in developing countries¹⁻³ and in some ethnic groups, such as Samoans, living in developed countries.^{4,6} It is estimated that the annual

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incidence of ARF in the developing world and the Western Pacific is 100 per 100,000 children^{3,7} and that 30 million children worldwide in less developed countries (LDCs) have rheumatic heart disease.¹ This disease is considered to be the most preventable of all cardiovascular disorders but is the leading cause of cardiac related death in the first four decades of life.⁸ The economic and social costs of ARF and rheumatic heart disease (RHD) impose considerable burdens on the health resources of the LDCs⁹ even though this is largely preventable.^{2,10} Although ARF is thought to be a significant Public health problem in Micronesia, studies have not been conducted to determine the incidence of ARF or RHD.

Study Population

The Federated States of Micronesia (FSM) are comprised of four separate states with a total 1988 estimated population of 101,884 scattered over 45 inhabited islands of the Caroline archipelago. Their land area is only 271 square miles lying within 2,000,000 square miles of the Pacific Ocean. The climate is tropical with variable rainfall, generally greater on the higher islands than on the atolls. The people are predominately Micronesian with about 1,000 Polynesians. There are four base

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hospitals, one in each state capital, that offer diagnostic facilities and are staffed by doctors. A number of outlying dispensaries provide care in villages and the smaller islands. Medical referrals within the states are hampered by distance and lack of airstrips on most islands.

Over the period 1981-1982 there were only three cases of ARF reported from the former Trust Territories of the Pacific Islands,^{11,12} including the FSM and the Republic of Belau but excluded the Marianas Islands, giving an average annual incidence of 0.98/100,000. Over that same period the incidence of ARF among the United States was 0.24/100,000.^{11,12}

In contrast, the incidence of ARF in Samoan children 4-18 years of age living in Hawaii was 206/100,000⁴ over a 4 year period (1980-1984), and that of New Zealand Maoris ages 5-29 was 68.6/100,000 over the period

1973-1986.⁵ From the data above, it appears that Micronesians were either unlikely to contract ARF or there was gross under-reporting of the disease throughout the U.S. Trust Territories. This study was undertaken

in Kosrae the referral list, penicillin prophylaxis records, and hospital laboratory reports were available for review.

Throughout the FSM outpatient and inpatient hospital records are kept in the same binder and this allowed for easy reference from one to the other. All patients with RHD over the period of the study had their charts reviewed for a record of an outpatient encounter consistent with ARF. In addition, the charts of all patients who had a positive streptococcal culture or serological test were reviewed as well as those of patients listed on the Public Health penicillin prophylaxis or the cardiology case logs. Only one inpatient with ARF during 1986-1988 was identified through these steps.

A majority of patients with ARF during 1988 were found on review of Public Health records. Data from dis-

pensary diagnosed and treated patients were not considered in this study as most dispensaries do not keep permanent records of patient encounters.

Diagnosis of ARF was based on the Modified Jones criteria¹³ and the WHO modifications.^{3,14} The Jones criteria require two major manifestations (carditis, polyarthritides, chorea, erythema marginatum, subcutaneous nodules) or one major and two minor manifestations (fever, arthralgia, history of previous rheumatic fever or RHD, EKG, and elevated erythrocyte sedimentation rate, C-reactive protein, or leukocytosis) and evidence of preceding streptococcal infection (recent scarlet fever, increased streptococcal antibodies in serum, or positive throat culture).

Laboratory tests to confirm previous streptococcal infections (ASO titer, etc.) were not always available in Yap, Pohnpei, and Kosrae hospitals and were never available at Chuuk Hospital over the study period. Therefore the diagnosis of ARF was made without this laboratory confirmation as recommended in the 1966 report of the WHO Expert Committee on Prevention of Rheumatic Fever.¹⁴ This report recommended that the requirement of evidence of prior streptococcal infection be omitted in developing countries without sufficient laboratory resources.

In addition, a 1988 WHO technical report recommended that late onset carditis and chorea be exempt from evidence of prior infection as the anti-streptococcal antibody levels may have fallen to normal levels by the time the patient presents with symptoms. This report advised that in patients with previously established RHD who have not taken suppressive drugs for at least two months one major criterion, or fever, arthralgia, elevated acute-phase reactants plus evidence of a preceding streptococcal infection be considered presumptive of

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to determine retrospectively the incidence of ARF and the number of hospital discharges with the diagnosis of RHD in the FSM.

Material and Methods

Medical records of all patients hospitalized in one of the four central hospitals with ARF or RHD during the years 1986 through 1988 were reviewed. A computer-

Table 1. Estimated Midyear Population for the FS of Micronesia (1986-1988)

Year	Chuuk	Kosrae	Pohnpei	Yap	FSM (Total)
1986	48,862	6,598	30,149	9,868	95,447
1987	50,621	6,789	31,084	10,135	98,629
1988	52,443	6,986	32,047	10,408	101,884

ized list of all hospital discharges with the ICD code 390-392.9 (ARF) and 393-398.9 (RHD) for the period 1986-1987 was provided by the Department of Health of the FSM. The 1988 data had not yet been entered onto the central computer and was not available. In Yap State the hospital computer data base was examined along with Public Health's registry of benzathine penicillin recipients, laboratory books for positive throat cultures and ASO titers of hospitalized patients, death certificates, and a pediatric cardiac patient book of the local pediatrician. At Chuuk Hospital pediatric ward log books, the hospital computer, and the cardiac patient book were reviewed. Chuuk Hospital did not carry out streptococcal serology or culture over that period. In Pohnpei the hospital computer, ward nurses, public health penicillin prophylaxis records, ward doctors, death certificates, and laboratory records were utilized as sources. While

ARF. The report further stated that the requirement of evidence of a prior group A streptococcal infection was essential and can be obtained in most developing countries. Unfortunately, this is not the case with the FSM.

The hospital discharge rate for all patients with the diagnosis of RHD was obtained by reviewing charts of patients with ICD codes 393-398.9 entered as the primary or contributing diagnosis. Cardiac referral lists were reviewed and all patients who were sent off-island for evaluation or treatment and who received a confirmed diagnosis of ARF or RHD were included. Estimated midyear population for the years 1986-1988 was based on the information provided by the FSM Office of Statistics and Planning with an annual population growth rate of 3.37%. (see Table 1)

Table 2. Number of Hospitalised Cases of Acute Rheumatic Fever in the FS of Micronesia (1986-1988)

Country	Year	Number of Cases	Incidence - No. per 100,000 Deaths
Chuuk	1986	7	14.3
	1987	1	2.0
	1988	1	1.9
	Mean	3.0	5.9
Kosrae	1986	1	15.1
	1987	1	14.7
	1988	1	71.6
	Mean	2.3	34.4
Pohnpei	1986	7	23.2
	1987	2	6.4
	1988	7	21.8
	Mean	5.0	17.1
Yap	1986	1	10.1
	1987	0	0
	1988	1	9.6
	Mean	0.67	6.6
Totals	1986	16	16.7
	1987	4	4.1
	1988	11	10.8
	Mean	10.3	10.5

Table 3. Age Distribution of Children with Acute Rheumatic Fever

Age Group (Years)	Number of Cases	Frequency (%)
< 4	2	6
4 - 5	0	0
6 - 11	15	48
12 - 15	10	32
16 - 18	1	3
18 - 19	2	6
> 20	3	3

Results

A total of 31 cases of ARF occurred in the FSM over the period of 1986-1988 using this study's criteria. This gave an incidence rate of 10.4/100,000 per year for the FSM. Table 2 shows the number of cases and the incidence of ARF by state, and the number of deaths from ARF.

The annual incidence rate in the 5-19 year age group was 25/100,000 in all four states over the study period. In the 5-29 year old cohort the incidence was 18/100,000 and 14.6/100,000 in the 5-39 years of age group 14.6/100,000 (Table 3). Forty-two percent were males and 58% were females. There was no seasonal variation.

Only 2 of the 31 cases were recurrent attacks of ARF—a recurrence rate of 6%; the remaining 29 were primary cases. Twenty of the 31 cases (64.5%) had an elevated serological test for streptococcal infection,¹⁷ a positive throat culture¹ or a typical scarlet fever rash.² In the other 11 patients no confirmatory laboratory tests were carried out.

No single data base provided an accurate list of ARF cases. Of all the ARF cases listed on the FSM national computer of hospital discharges for the years 1986-1987, only 14/37 met the study's criteria for the diagnosis of ARF (Yap 2/2, Truk 7/12, Pohnpei 3/8, Kosrae 2/15). The other 17 cases (11 from 1988 and 6 from 1986-1987) were located through public health records. Of 63 charts reviewed with a ICD coding of 390-393.9 (ARF), only 26 fulfilled the Modified Jones or the WHO criteria as described above. Four patients with confirmed ARF had been miscoded as RHD and another as rheumatoid arthritis. This accounts for the total of 31 cases of ARF hospitalized over the period 1986-1988.

The number of hospital discharges with an established diagnosis and the number of off-island cardiac referrals RHD are shown in Table 4. There were 48 hospital

discharges with a primary diagnosis RHD over the three year period of 1986-1988. A total of 42 patients were hospitalized. One child admitted with a diagnosis of ARF during the study period was later admitted to the hospital with RHD, and is included in both categories. Twenty-two patients were referred to the United States for cardiac evaluation because of RHD and ten underwent corrective surgery. There were 2 deaths at a referral hospital, and one patient died before referral. The annual hospital discharge rate for RHD was 16.2/100,000 in the FSM. Kosrae state had nearly twice this rate (29.5/100,000) in comparison to the other states.

Discussion

The incidence of ARF in the FSM for the years 1986-1988 (10.4/100,000) appears to be lower than that of many of its Pacific neighbors although it is considerably higher than that of the United States in 1986 (0.12/100,000).¹⁵ Direct comparisons between the rates of this Micronesian population and other Pacific island groups are difficult to interpret because different time periods, data sources and age groups were reported in these studies. Chun et al⁴ found an incidence of 206/100,000 cases of ARF among Samoans living in Hawaii (1976-1980) in the 4-18 age group (though he adds the caveat that this is probably high due to underestimation of actual Samoan population). In contrast, Neutze⁷ reported an annual incidence of 7.6/100,000 from Tonga during the period 1981-1985 in the 5-24 year age group. Among the 5-39 year age group, Fiji had an incidence of 11.3/100,000 (1982-1984), Western Samoa was 16.1/100,000 (1982-1985), while French Polynesia reported 72.2/100,000 (1980-1984).⁷ (see Table 5)

The incidence of ARF we found in Micronesia is slightly higher than that of Tonga and Fiji and very similar to that of Western Samoa, but considerably lower than that of French Polynesia, Samoans living in Hawaii, or New Zealand Maoris. Why the incidence of ARF in Micronesia is lower than that reported in other groups of Pacific islanders may be explained by several factors; the most important of these are likely to be the problems of detection, inaccessibility of outer-islanders to health care, and diagnostic inaccuracy.

Approximately 50% of the people of the FSM do not live on the main island or central atoll.²¹ Their only access to the hospital is via the irregular field trip vessel that calls once every one to six months. A patient who develops ARF on one of these outer islands would be unlikely to be referred to the central hospital and therefore would not have been included in this study. If one can assume that ARF is as common in these isolated communities as in the main centers, then one half of all patients with ARF may have escaped detection. A similar percentage of patients with RHD may also be undetected and does not receive penicillin prophylaxis. This underserved population could

Table 4. Hospital Discharges and Referrals for Cardiac Evaluation for Rheumatic Heart Disease (1986-1988)

	Year	Number of Discharges	Discharge Rate /100,000	Number of Overseas Referrals
Chuuk	1986	6	12.3	2
	1987	8	15.8	4
	1988	9	17.6	2
	<i>Mean</i>	7.7	15.2	2.7
Kosrae	1986	1	15.2	6
	1987	2	29.5	1
	1988	3	43.0	2
	<i>Mean</i>	2	29.5	3
Pohnpei	1986	3	10.0	0
	1987	9	29.0	1
	1988	2	6.2	0
	<i>Mean</i>	4.7	15.1	.3
Yap	1986	2	20.2	2
	1987	0	0	0
	1988	3	28.8	2
	<i>Mean</i>	1.7	16.8	1.3
Totals	1986	12	12.6	12
	1987	19	19.3	4
	1988	17	16.7	6
	<i>Mean</i>	16	16.2	7.3

be screened to determine the prevalence of RHD. The training of health assistants manning the outlying dispensaries in the diagnosis, management, and reporting of ARF, and the development of effective community control programs can increase the number of identified cases.

There is a strong possibility that the lower incidence of ARF in the FSM may be due to under-reporting or to misdiagnosis. The health reporting systems of the more cosmopolitan islands may be more thorough in collecting cases of ARF; physicians are more numerous, and they are equipped with modern diagnostic equipment. This is most certainly the case in New Zealand, Hawaii, and the French-controlled islands of the Pacific. A number of patients who had received a discharge diagnosis of ARF did not meet the defined diagnostic criteria. Over one-half of physician-diagnosed cases did not fulfill diagnostic criteria. This is similar to the findings of

others.^{17,18} Hospital notes were often terse with few positive or negative findings noted.

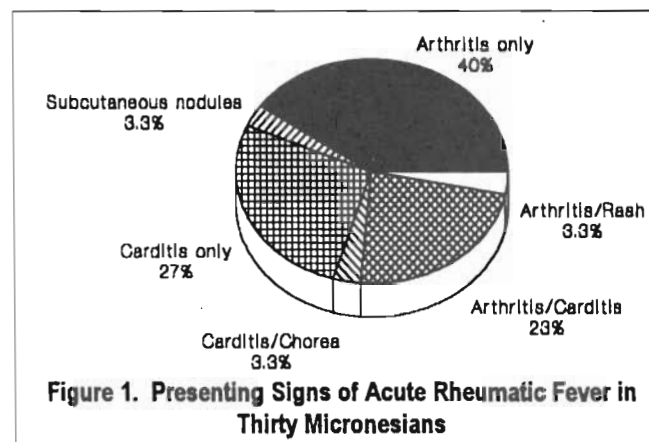
A single patient (3.5%) was identified as having erythema marginatum which may be more difficult to see on the skin of a Micronesian. It is reported to occur in 0.5-10% of patients with ARF.^{17,20} Sydenham's chorea was described in one female patient (range 3-20%^{16,17} and subcutaneous nodules were found in another, which is similar to the usual prevalence of less than 3%.²⁰ Carditis (50%) and arthritis (73%) were the most common major criterion reported. Most cases of ARF in Micronesia were diagnosed on the basis of carditis or arthritis or both (23%) along with a fever (100%), an elevated sedimentation rate, and an ASO titer done in the United States. The type of symptoms in the Micronesian patients (Figure 1) is similar to that reported from Hawaii⁴ where arthritis was present in 73%, carditis in 34%, chorea in 10%, and subcutaneous nodules and erythema marginatum in 2% of children with ARF.

Micronesians may be genetically less susceptible to attacks of ARF in comparison to their Polynesian neighbors.

Region	Period	Age Range (Years)	Incidence /100,000
New Zealand Maori	1973-1986	5 - 29	68.6
Samoans in Hawaii	1976-1980	4 - 18	206
Western Samoa	1982-1985	5 - 39	16.1
French Polynesia	1980-1984	5 - 39	72.2
Tonga	1981-1985	5 - 24	7.6
Fiji	1982-1984	5 - 39	11.9
Fed. States of Micronesia	1986-1988	-	10.4

The hypothesis of rheumatic susceptibility is currently under investigation and inherited host factors do appear to play a part in who contracts ARF.¹⁶ Polynesians in Tonga, Western Samoa, Hawaii, and French Polynesia are genetically similar but there has been a large variation in their risk of ARF. This suggests that other factors are at least partially responsible for the observed differences such as the completeness of the reporting system, changes in diagnostic criteria over the years,¹⁷ or differences in the socioeconomic status of the various groups.^{4,5}

It is possible that the prevalence of rheumatogenic strains of group A streptococci or of streptococcal pharyngitis infections may be lower in Micronesia than in the other Western Pacific areas. No studies of the serological types of streptococci have been carried out in the FSM, and this remains a field for research. Throat cultures were rarely done during this study period, and bacteriology facilities were available in only 3 of the 4 central hospitals.



The 48 overseas referrals for RHD contributed to the high cost of off-island care. The cost of referral in 1986 to the FSM government was more than \$11,000²² per RHD case, while secondary prevention with benzathine penicillin would have cost approximately \$100 per person per year.^{7,23} There is ample evidence that secondary prophylaxis is effective and that national RHD registries are a practical method to improve compliance, monitor the epidemiology of the disease, and educate the public. A national registry of cases of ARF and RHD would enable the Department of Health to determine the true extent of the problem and permit the improvement of secondary prophylaxis.^{5,10}

An extensive and coordinated primary health care program adequately supplied with simple diagnostic tests for streptococci and appropriate drugs, staffed with aware and motivated health workers, and supported by the community can begin to approach the task of primary prevention. An efficient primary and secondary prevention program would more than pay for itself. If one assumes that the prevalence of ARF in the FSM is the same as that of Western Samoa (2.3/1000), then there are approximately 234 patients in the FSM that require penicillin prophylaxis at a total cost of \$23,000 per year. This is equal to the cost of two off-island medical referrals.

Conclusions

Rheumatic fever is common in Micronesia with an incidence of 10.4/100,000 over the period 1986-1989. We believe that the problem is much greater because of the population's poor access to appropriate medical care

and that the incidence of ARF is probably several times larger than reported here. End-stage RHD resulted in 22 referrals off-island for tertiary cardiac care over the same three year period. The true prevalence of RHD and

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“The Jones criteria require two major manifestations . . . or one major and two minor manifestations . . . and evidence of preceding streptococcal infection.”

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incidence of ARF remains unknown. This study has provided the minimum rates of ARF and RHD in the FSM.

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