Acute Rheumatic Fever and Rheumatic Heart Disease on the Navajo Reservation, 1962-77

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ACUTE RHEUMATIC FEVER (ARF) and rheumatic heart disease (RHD) occur more frequently among lower socioeconomic groups than among other North American populations (*1, 2*). In recent decades, a marked decline in attack rates for ARF has been observed in Europe and the United States (*1*). However, no studies of ARF among American Indians have been reported.

The Navajo Indian Reservation comprises about 25,000 square miles of semi-arid canyon and plateau country in northeastern Arizona, northwestern New Mexico, and southern Utah. The Navajo live under crowded family conditions and in a relatively harsh environment. Upper resipratory infections and pharyngitis considered to be streptococcal in etiology are common among Navajo children (*3*). Because the Navajo have virtually no coronary artery disease, RHD is the major cause of cardiac morbidity and mortality among them.

To ascertain the frequency, demography, and clinical characteristics of ARF and RHD among the Navajo, all records of patients hospitalized because of either condition over a 16-year period were reviewed. Also, the effect of a streptococcal disease control program was observed over a 3-year period.

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Methods

Ascertainment of clinical care. In 1972 the investigators reviewed records for a 10-year period, 1962–71, of all Navajo patients discharged with diagnoses of ARF (ICD–8, 390–391), chorea (392), or chronic RHD 393–398) from all 6 Indian Health Service hospitals on or adjacent to the reservation and 3 private hospitals also serving the Navajo community. For inclusion in the study, the patient had to be Navajo; the diagnosis of ARF had to meet the revised Jones criteria of the American Heart Association (*4*); and if heart disease was present, it had to be diagnosed as RHD—with no evidence of congenital or other nonrheumatic valvular disease—by the patient's physician. Many of these patients were followed by cardiology consultants from the University of Arizona.

Diagnosis, date of illness, age, and sex were recorded for each patient. Each episode of ARF was characterized as initial or recurrent. Clinical characteristics and prophylaxis for ARF and RHD were studied in detail for patients seen at Fort Defiance Hospital, which at that time had more than 40 percent of the total ARF admissions.

In 1978 lists of all discharges for the same diagnoses during the 6 years from July 1, 1972 through June 30, 1977 were obtained for the same 9 hospitals that still provided almost all medical care for Navajos living on

the reservation. The same criteria for including patients were used, and the same kinds of data were abstracted from records as in the earlier study. However, clinical characteristics of ARF and RHD were recorded for all cases rather than just for Fort Defiance admissions.

Because the time for filing hospital discharge diagnoses was changed from calendar year to fiscal year, a 6-month period (January 1, 1972 through June 30, 1972) was omitted from this study.

Population estimates. Three widely different estimates of the total Navajo population have been used in recent years: the 1970 census, the Navajo Area BIA (Bureau of Indian Affairs) Population Register, and data from the U.S. Office of Revenue Sharing. Of these estimates, the BIA has accepted the Revenue Sharing data as the best (*5*), and this estimate was used in determining ARF occurrence rates over the 16-year period. The July 1, 1975, population was specified as 134,340. An annual population growth factor of 2.9 percent, based on available birth and death rates (*5*), was then used to calculate the population for each of the 16 years.

Streptococcal disease control program. In mid-1974, a streptococcal disease control program was initiated in certain areas of the Navajo Reservation under the auspices of the Navajo Tribe through a contract with the Indian Health Service. In this program, school nurses or health aides took throat specimens for culture from children in the first through sixth grades at the beginning of the academic year in both public and Bureau of Indian Affairs schools. Thereafter, throat cultures were done periodically (usually monthly) for asymptomatic children, but they were done immediately for any child who presented to the nurse's room with a sore throat. Swabs were plated on standard sheep blood agar plates and sent to one of two special laboratories funded by the program. If group A beta hemolytic streptococci was identified, the nurse was notified and the child was treated with pencillin or erythromycin, if parental permission was obtained. Records were collected of (*a*) schools and districts served by the program and (*b*) numbers of cultures and positivity rates from January 1, 1975 through December 31, 1977.

Schools adopted the control program at different times, and many participated intermittently. During 1975–77, the program was offered in some of the eight Indian Health Service Units into which the reservation is divided. Within given Service Units, there was also a gradient in percentage of schools covered under the program. For analyses in the present study, the eight Service Units were divided into those with some coverage (Fort Defiance, Tuba City, Kayenta, Crownpoint, and Chinle), in which many or most of the schools participated most of the time, and those with no coverage (Gallup, Shiprock, and Winslow). The occurrence of ARF in covered and uncovered areas was compared for 1972–74 (before the program) and for 1975–77 (during the program).

Results

Acute rheumatic fever. Table 1 presents the average annual attack rates for initial episodes of ARF by 2-

year periods. No significant trend was observed for the period as a whole. A peak rate occurred during the 4 years 1968 through 1971, while earlier and more current rates were lower and similar to each other. During the first 10 years, 16.4 percent of all cases were recurrent ARF; in the later period, the percentage was 24.7. The mean time elapsed before recurrence in the 1962–71 timespan was 24.3 months, with a median of 16 months.

The age-specific attack rates are shown in table 2. Of the 184 initial cases of ARF, 93 were in males. The highest rate occurred in the 10–14 age group, and about 80 percent of all cases occurred in the age group 5–24. There were 18 first attacks per 100,000 in the 5–24 age group from 1962 through 1971 and 13.4 per 100,000 from 1972 through 1977.

A comparison of age distributions in the two time periods revealed that initial attacks tended to occur at an earlier age in the more recent group. For example, children aged 5 to 9 increased in representation from 16.2 percent to 28.4 percent ($\chi^2 = 3.82$, 1 *df*, $P \approx 0.05$), while persons aged 20 to 24 decreased in representation from 16.2 percent to 7.5 percent. Of 80 cases of ARF with date of diagnosis from 1972 through 1977, 21 occurred in January, February, and March; 22 in April, May, and June; 21 in July, August, and September; and 16 in October, November, and December.

The clinical characteristics of ARF cases are shown in table 3. Clinical data were available from 1962 through 1971 only for patients seen at Fort Defiance Hospital; however, from 1972 through 1977 these data were available for all patients. The proportion of patients with chorea declined from 26.2 percent in the first time period to 11.2 percent in the second, but the proportion with carditis and arthritis remained essentially unchanged. The proportion of patients with leukocytosis increased, and the number of throat cultures positive for streptococcus decreased. The striking increase in the percentage of patients with prolonged P-R intervals on electrocardiograms must be interpreted cautiously because it is not known if electrocardiograms were performed on all the earlier patients.

Table 1. Acute rheumatic fever attack rates on Navajo Reservation 1962–77, by 2-year intervals

Years	Initial attacks	Estimated population	Initial attack rate [1]	Recurrent attacks	Recurrence rate [1]	Total attacks [1]
1962–63	20	93,721	10.7			
1964–65	14	99,281	7.0			
1966–67	18	105,170	8.6	23	2.2	13.2
1968–69	28	111,408	12.6			
1970–71	37	118,016	15.7			
1972–73	20	125,017	8.0			
1974–75	24	132,432	9.1	22	2.8	11.2
1976–77	23	140,288	8.2			

[1] Per 100,000.

Table 2. Initial attacks of rheumatic fever among Navajos, by age groups

Age group (years)	All cases		Percent population	Index population [1]	Rate [2]
	Number	Percent			
0– 4	3	1.6	13.5	15,480	1.2
5– 9	38	20.7	14.6	16,741	14.2
10–14	60	32.6	14.0	16,053	23.4
15–19	25	13.6	11.6	13,301	11.8
20–24	24	13.1	9.5	10,893	13.8
25–34	23	12.5	14.5	16,626	8.6
35–44	8	4.2	9.1	10,434	4.8
45 and over	3	1.6	13.2	15,136	1.2
Total	184	100.0	100.0	114,664	...

[1] Population estimate at midpoint of series, with age-group breakdown as used by the Bureau of Indian Affairs Statistics Department.

[2] Per 100,000 per year.

Table 3. Clinical characteristics of acute rheumatic fever among Navajos, 1962–77

Clinical characteristics	1962–71 [1]		1972–77 [2]	
	Number	Percent	Number	Percent
Major criteria				
Carditis	38	52	45	51
Arthritis	37	61	57	64
Chorea	[3] 16	26	[3] 10	11
Nodules				
Erythema marginatum			4	4
Other signs and symptoms				
Arthralgias	19	31	35	39
Fever	15	30	31	51
Positive streptococcal throat culture	14	23	10	11
Increased P-R interval on electrocardiogram	[4] 3	5	[4] 19	21
Leukocytosis	[5] 16	26	[5] 46	52
Elevated erythrocyte sedimentation rate	51	84	72	81
Elevated ASO titer (≧ 300)	38	62	63	71

[1] 61 cases seen at Fort Defiance Hospital; diagnosed according to modified Jones criteria.
[2] Total of 89 cases seen; diagnosed according to modified Jones criteria.
[3] $\chi^2 = 5.68$, $P < 0.05$.
[4] $\chi^2 = 7.81$, $P < 0.01$.
[5] $\chi^2 = 9.67$, $P < 0.01$.

The amount of prophylaxis documented on charts differed in the two time periods. From 1962 through 1971, the charts showed that penicillin or other prophylaxis was ordered for 46 of the 61 ARF patients (75.4 percent) at Fort Defiance Hospital. From 1972 through 1977, prophylaxis was ordered for 82 of the 89 ARF patients (92.1 percent) throughout the Navajo area ($\chi^2 = 8.09$, $P < 0.001$). The charts were also audited for documentation of specific bacterial endocarditis prophylaxis at the time of dental or surgical procedures. Of 27 dental and surgical procedures performed on ARF patients at Fort Defiance Hospital in the first 10 years, 13 (48.1 percent) received prophylaxis, whereas prophylaxis was ordered for each of 65 procedures performed in the second period.

Streptococcal disease control. As part of the control program, 172,700 throat cultures were performed from 1975 through 1977, and 11,931 (6.9 percent) of these cultures were positive for group A beta hemolytic streptococci. Overall, the positivity rate was 8.6 percent in September, declined to 5.7 percent in October, rose again to 8.4 percent from November through January, and then dropped to about 6 percent from February through May. Cultures were not done in the summer months.

Location of residence was ascertained for 80 of the 89 ARF patients seen from 1972 through 1977. The Navajo Service Units were grouped according to some coverage under the control program or no coverage. Table 4 shows the attack rates for ARF for the 2 years before the program (July 1, 1972 through June 30, 1974) and the 3 years covered by the program (July 1, 1974 through June 30, 1977). Covered areas' ARF rate was 39 percent lower in the second period, while the rates in areas with no coverage showed little change. However, covered Service Units started with substantially higher annual attack rates than the uncovered ones during the 2 years before the program.

The control program was aimed primarily at elementary school children. In the covered areas, 21 of 28 cases (75 percent) were in children aged 6 through 16 before the program, while 9 of 18 cases (50 percent, $\chi^2 = 3.01$, $P < 0.10$) were in children of that age

Table 4. Occurrence of acute rheumatic fever before and and during streptococcal disease control program coverage for Navajos, 1972–77

Coverage	Population	Number cases	Rate [1]
	Before program, 7/1/72–6/30/74		
Some	68,997	28	13.5
None	56,060	16	9.5
	During program, 7/1/74–6/30/77		
Some	73,089	18	8.2
None	59,385	18	10.1

[1] Per 100,000 per year.

Table 5. Age distribution of Navajo patients admitted for rheumatic heart disease

Age group (years)	1962–71			1972–77		
	Number	Percent	Cumulative percent	Number	Percent	Cumulative percent
0– 4	2	1.1	1.1	1	0.7	0.7
5– 9	7	3.8	4.9	2	1.4	2.1
10–14	19	10.3	15.2	4	2.7	4.8
15–19	26	14.1	29.3	8	5.4	10.2
20–24	30	16.3	45.6	14	9.5	19.7
25–29	25	13.6	59.2	23	15.7	35.4
30–34	21	11.5	70.7	19	12.9	48.3
35–39	19	10.3	81.0	14	9.5	57.8
40–44	16	8.7	89.7	15	10.2	68.0
45–49	9	4.9	94.6	8	5.5	73.5
50–54	3	1.6	96.2	10	6.8	80.3
55–59	2	1.1	97.3	12	8.2	88.5
60 and over	5	2.7	100.0	17	11.5	100.0
Total	184	100.0	....	147	100.0	....

group during the program. The percentages were 50 and 61 for the noncovered areas in the two periods.

Rheumatic heart disease. The numbers of hospitalizations for a primary or secondary diagnosis of RHD were 187 during 1962–71 and 147 during 1972–77. However, these number are not additive because patients in the second cohort could also be represented in the first. The annualized rates of persons hospitalized for RHD during the two periods were 17.5 and 18.5 per 100,000 population, respectively. Table 5 shows the age distribution of RHD in the two groups. The younger age groups, especially those under 24 years, had far less RHD in 1972–77 than in 1962–71 ($\chi^2 = 20.5$, $P < 0.001$) but the opposite was true for persons aged 50 and over ($\chi^2 = 28.8$, $P < 0.001$). In both groups of patients with RHD, the percentages of males remained about the same, 35.5 (65) and 29.9 (44). The kinds of valvular lesions seen in RHD patients are shown in table 6. The clinical patterns were similar for the two periods, with about 75–80 percent mitral lesions, mostly mitral insufficiency, and more than half of the patients had two or more lesions.

Discussion

Methodological problems in determining the incidence of ARF in large populations include lack of uniform diagnostic criteria, incomplete ascertainment, inadequate data sources, and skewing of data by use of nonrepresentative population samples—for example, hospital patients only (*1*). In the present survey, some of these problems were avoided or were of lesser significance. The Navajo health facilities have a uniform reporting system, and all use the modified Jones criteria. Mild cases may have been missed because only patients hospitalized during sometime in the course of their disease were studied. However, the proportion of patients with ARF who were hospitalized was likely high because long distances, poor transportation, and lack of understanding about the disease among patients make ambulatory followup difficult.

Four general observations about ARF and RHD among Navajo people are suggested by the results of this study: the overall ARF rate was modest but unchanged; ARF seems to have become milder; prophylactic management of ARF has improved; and the streptococcal control program was associated with a decline in ARF in the population served.

Table 6. Valvular lesions in Navajo patients with rheumatic heart disease

Lesions	1962–71 [1]		1972–77 [2]	
	Number	Percent	Number	Percent
Mitral insufficiency	37	54	127	54
Mitral stenosis	15	22	63	27
Aortic insufficiency	9	13	26	11
Aortic stenosis	6	9	16	7
Tricuspid insufficiency	2	3	3	1
Total	69	100	235	100
Lesions per patient	2	...	2	...
Pericarditis	2	...	1	...
Conduction defects	3	...	13	...

[1] 44 patients seen at Fort Defiance Hospital.
[2] Total series of 147 patients with rheumatic heart disease.

Rate. The annualized ARF attack rate for all Navajos from 1962 through 1967 was 12.4 per 100,000, and there was no evidence of a secular trend. Although the Navajo population has a low socioeconomic status and a high incidence of streptococcal infections (*3*), the ARF incidence rate is modest. This modest rate also may have held 30 years ago, because Smith (*6*), in an analysis of Navajo death certificates, found that Navajos had about the expected number of deaths from ARF and less than the expected number from chronic RHD when age corrected and compared with the number for the white population.

Acute rheumatic fever has decreased in frequency and severity during the past three decades, particularly in North America and Europe (*1,2,7,8*). This decline has been most precipitous in the Scandinavian countries; for example, in Sweden the ARF attack rate declined from 38 per 100,000 from 1940 to 1944 to less than 2 per 100,000 from 1960 to 1964 (*9,10*). Despite the overall lowering trend, the high incidence of ARF in the United States is still associated with lower socioeconomic class, crowded living conditions, and inadequate treatment and prevention of streptococcal pharyngitis (*1,2*).

In table 7 the Navajo ARF rates are compared with those found in some other American surveys in the 1960s. The ARF incidence among Navajos was similar to that in Baltimore (*11,12*) and Nashville (*13*) and much lower than that reported for New York City (*14*). Predominantly rural areas, such as in Oklahoma (*15*) and Colorado (*16*), have reported lower rates (4.2 per 100,000 in 1969 and 5.9 per 100,000 in 1973, respectively), but these were based on surveys of physicians who did not use uniform diagnostic criteria.

Table 7. Acute rheumatic fever attack rates among Navajos compared with other selected U.S. population groups

Study group [1]	Rate per 100,000			
	White	Nonwhite [2]	Total	Navajo
Baltimore, 1960–64, ages 5–9 (*11*)	9.6	24.4	15.6	16.4
Nashville, 1963–69, all ages (*13*)	9.0	17.9	10.8	11.6
Manhattan, 1963–65, ages 5–14 (*14*)	23.0	78.0	61.0	18.8
Baltimore, 1968, ages 5–14 (*12*)	10.6	17.8	13.5	18.8

[1] Numbers in parentheses are references.
[2] In the Manhattan study, predominantly Puerto Rican; in the Baltimore studies, predominantly black.

Disease severity. There is some evidence that ARF is becoming a milder disease among the Navajo. Their ARF experience is similar to that of other populations in the proportion of recurrent attacks—19.6 percent compared to 19.7 percent in Manhattan (*14*) and 19 percent in Nashville (*13*)—and in clinical manifestations (*2,16,17*). The change in the percentages of patients with chorea between 1962–71 (26.6 percent) and 1972–77 (11.2 percent) is consistent with a decreasing occurrence of this manifestation noted by others (*16, 18*). Proportionately more RHD patients were in the older age groups in 1972–77 than in 1962–71, perhaps because people with known RHD are becoming older —as the data in table 5 suggest—and relatively few new cases of RHD in young people have been diagnosed recently. Because development of RHD relates to ARF severity (*19–21*), fewer new diagnoses of RHD may indicate that rheumatic fever has become, on the average, less severe.

RHD prophylaxis. More than 90 percent of the ARF patients in the 1972–77 series received streptocccal prophylaxis and were placed in a followup program, as opposed to 75 percent in 1962–71. Almost every patient who had a dental or surgical procedure received endocarditis prophylaxis in 1972–77, compared to only 48 percent in 1962–71. The increased ordering of prophylaxis reflects the institution, in 1972, and maintenance of a Rheumatic Fever Control Registry and the incorporation of stricter followup and control procedures.

Streptococcal surveillance programs have decreased the endemicity of group A beta hemolytic streptococcal throat colonization, as well as the incidence of ARF (*22,23*). In the Navajo surveillance program, cultures were done for all symptomatic children and periodically (for example, monthly) for some asymptomatic children—the relative proportions and positivity rates varied by school and district.

Streptococcal control. The streptocooccal surveillance program may have had a favorable effect. The frequency of ARF cases in areas covered by the program decreased from 13.5 to 8.2 cases per year. The rates in the noncovered areas were 9.5 before the program was started and 10.1 after. Specifically, the apparent decrease in new cases occurred in the target group of the surveillance program, elementary school children. The rates are based on small numbers, and the difference in pre-control rates between the two areas (13.5 versus 9.5, higher in the covered Service Units) is larger than the difference observed during the control program (8.2 versus 10.1, lower in the covered Service Units). It is not likely that special characteristics of the Navajo Service Unit areas and population would

lead to meaningful or consistent discrepancies in true ARF occurrence. Thus, if the pre-control difference is an artifact, the subsequent changes must be viewed with caution. Nevertheless, although the figures are consistent with a modest effect on ARF rates, more years of observation are required.

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SYNOPSIS

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The occurrence of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) among Navajos was ascertained for the period from 1962 through 1977 by means of hospital discharge diagnoses and patients' charts. The annualized attack rate for ARF was 12.4 per 100,000 population, with no clear evidence of an overall secular trend. The proportion of recurrences (19.6 percent) and clinical features were similar to those reported elsewhere, but no seasonal variation in attack rates was noted. Between 1962–71 and 1972–77, the age of RHD patients increased, suggesting few newly diagnosed cases and the aging of known patients.

A streptococcal disease control program was instituted in many Navajo elementary schools before 1975. In the program, throat cultures were performed routinely for some asymptomatic children and for all symptomatic children. During the subsequent 3 years, ARF rates declined from 13.5 to 8.2 per 100,000 in areas covered by the program, while in the noncovered areas the rates showed little change—9.5 to 10.1 per 100,000.