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Research Letter

Rheumatic heart disease on an Eastern Arizona Reservation, 2007-2022

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ABSTRACT

Contemporary studies of rheumatic heart disease (RHD) within American Indian communities are lacking, despite recent work indicating high rates of group A streptococcus, the precursor to RHD. Utilizing retrospective chart review of the Indian Health Service, we sought to characterize the burden of acute rheumatic fever and RHD within an American Indian tribe in Eastern Arizona. Our study found that, in line with other high-income countries, RHD in the US continues to disproportionately impact native peoples, with rates 10 times that of the general population.

1. Background

Rheumatic heart disease (RHD) results from recurrent untreated group A streptococcal (GAS) infection and acute rheumatic fever (ARF), with subsequent valvular damage. RHD burden is inequitable, impacting mainly those in low- and middle-income countries (LMIC) and indigenous populations within high-income countries (HIC) [1]. Historical studies examining RHD in American Indian populations found rates seven times that of the general population [2]. Recent work within an American Indian population found rates of invasive GAS, the precursor to RHD, 30 times higher than their non-native counterparts [3]. Despite this knowledge, contemporary studies of ARF and RHD within American Indian communities are lacking. We sought to characterize the burden of ARF and RHD within one American Indian tribe in Eastern Arizona.

2. Methods

We conducted a retrospective chart review of the electronic health record (EHR) of the Indian Health Service (IHS) facility, including American Indians 5 years of age or older between January 1, 2007 to December 31, 2022. Patients were excluded if they had congenital heart disease. An inclusive taxonomy for ARF/RHD was built of ICD-9/10 codes to pull patient charts. Extracted variables included demographic data and ARF/RHD related management. Disease severity was based on echocardiogram findings as either mild (mild mitral and/or aortic regurgitation), moderate (mild to moderate mitral and/or aortic regurgitation and/or mild LV dysfunction), or severe (moderate to severe mitral and/or aortic regurgitation, and/or more than mild LV dysfunction, and/or the presence of mitral stenosis). In cases where no echocardiogram data was available, severity was based on clinical notes

referencing disease severity. For individuals diagnosed prior to 2007, paper charts were requested and referenced when available dating back as far as 1960s for the earliest diagnosed patient. Data were managed using REDCap hosted at Cincinnati Children's Hospital. Descriptive statistics were used to enumerate the distribution of all variables analyzed. All procedures were performed in compliance with relevant laws and institutional guidelines and were approved by the Tribal Council (resolution 10-2020-237), Cincinnati Children's Institutional Review Board (IRB) (2020-0913), and the Indian Health Service IRB (PXR 21.09).

3. Results

Twenty-five patients had a diagnosis of ARF or RHD. Of these, 11 (44 %) were male and median age was 61 years (IQR 45.9–76.4) at the time of this study. For all patients, the median age at diagnosis was 37 years (IQR 23.3–62.7) with 21 (84 %) presenting with RHD and 4 (16 %) with ARF. Of those presenting with ARF, all eventually developed RHD. The median time from initial concern prompting work up to final diagnosis was 13.5 days (IQR 2.0–65.3), with six patients diagnosed more than a year after initial presentation. Based on echocardiograms obtained at time of diagnosis ($n = 19$, 6 missing data), 68 % ($n = 13/19$) presented with severe RHD. Six patients (25 %, $n = 24$, 1 missing data) required hospitalization at time of diagnosis, with 2 (8 %, $n = 24$, 1 missing data) requiring cardiac intervention at time of diagnosis. In total, nine patients (38 %, $n = 24$, 1 missing data) required either cardiac surgery and/or catheter-based interventions. Of these, six patients (25 %, $n = 24$) required valve replacement, with two of these (33 %, $n = 6$) undergoing multiple procedures (one patient underwent valve replacement twice, and one patient had catheter-based valvuloplasty with subsequent valve

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Table 1
Patient demographics and RHD diagnosis and management.

Age (at chart extraction), years ($n = 25$) (median, IQR)	61.1, 45.9–76.4
Age at diagnosis, years ($n = 22$) (median, IQR)	36.9, 23.3–62.7
Native American race ($n = 25$)	25 (100 %)
Non-Hispanic Ethnicity ($n = 25$)	25 (100 %)
Health insurance ($n = 25$)	
Indian Health Services Medicaid	10 (40 %)
Other medicaid	3 (12 %)
Medicare	6 (24 %)
Other medicaid + medicare	1 (4 %)
Indian Health Services Medicaid + Medicare	5 (20 %)
Private health insurance	0 (0 %)
Tribal affiliation ($n = 25$)	
Apache	21 (84 %)
Navajo	2 (8 %)
Hopi	1 (4 %)
Choctaw	1 (4 %)
Primary language ($n = 25$)	
English	12 (48 %)
Apache	12 (48 %)
Navajo	1 (4 %)
Presenting clinical concern ($n = 18$)	
Concern for ARF	4 (22 %)
Arrhythmia	4 (22 %)
Patient reported history of ARF	2 (11 %)
Murmur	2 (11 %)
Other	6 (34 %)
Disease severity at time of diagnosis ($n = 19$)	
Mild	3 (16 %)
Moderate	3 (16 %)
Severe	13 (68 %)
Total days from initial concern to definitive diagnosis ($n = 16$) (median, IQR)	13.5, 2.0–65.3
Required hospitalization at diagnosis ($n = 24$)	6 (25 %)
Required intervention at diagnosis ($n = 24$)	2 (9 %)
Catheterization	1 (4.5 %)
Surgery	1 (4.5 %)
Patient deaths during study period ($n = 25$)	4 (16 %)
Death from RHD complication	1 (4 %)
Prescribed prophylaxis ($n = 24$)	10 (42 %)

replacement). One patient (4.2 %, $n = 24$) underwent surgical valve commissurotomy. Ten (42 %, $n = 24$, 1 missing) patients were prescribed secondary antibiotic prophylaxis. There were four patient deaths during the study period, one confirmed cardiac, leaving 21 people living with RHD in the service unit user population ($n = 18,500$ in 2022). This translates to an RHD prevalence at the end of 2022 of 0.11 % (95 % CI, 0.1055 %, 0.1145 %). See Table 1 for further details.

4. Discussion

In line with other HIC [4], this contemporary study demonstrates that RHD in the US continues to disproportionately impact native peoples, with rates 10 times that of the general population (estimated overall RHD prevalence in the US of 0.018 %) [1]. Similar to presentation in LMICs [5], most individuals in our study were diagnosed late in the disease process and less than half were receiving secondary antibiotic prophylaxis. These data highlight the urgent need to better understand the epidemiology of RHD across our indigenous populations and for implementation strategies to improve diagnosis and adherence to guideline-directed care.

The study has some limitations. This was a retrospective study utilizing an EHR first implemented in 2006, thus data preceding this date was limited. Moreover, some patients may have been misclassified at time of presentation and not coded as RHD, therefore leading to under-reporting and lower calculated prevalence estimates. Additionally, details surrounding secondary antibiotic prophylaxis, or the lack thereof,

were inadequate to determine the reason for low secondary antibiotic prophylaxis prescription. We were unable to account for other potential contributing causes of reduced left ventricular function in determining disease severity. Finally, our findings are limited to one population and therefore may not be generalizable to other American Indian territories within the US.

RHD continues to exist as a disease of disparity both in the US and around the world. The prevalence of RHD has declined dramatically in the whole of the US, but high-risk populations, such as Native American populations, continue to be affected disproportionately. Additional research is needed to understand the scale of this burden across different populations and regions in the United States. Additional work examining community and provider awareness of RHD in American Indian populations could also offer valuable insight into potential avenues to improve guideline-based care and outcomes.

5. Conclusion

The burden of RHD within the US continues to be shouldered disproportionately by American Indian populations. Opportunities exist to improve RHD detection and prevention. Future research to better characterize RHD disease burden and community awareness within American Indian populations is needed.

Disclaimer

The opinions expressed in this paper are those of the author and do not necessarily reflect the views of the IHS.

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Ethical statement

All procedures were performed in compliance with relevant laws and institutional guidelines and were approved by the Tribal Council (resolution 10-2020-237), Cincinnati Children's Institutional Review Board (IRB) (2020-0913), and the Indian Health Service IRB (PXR 21.09).

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Jacob Hoekzema: Writing – original draft, Investigation, Formal analysis, Data curation. **LeCario Benashley:** Writing – review & editing, Investigation, Data curation. **Ryan Close:** Writing – review & editing, Methodology, Conceptualization. **Andrea Beaton:** Writing – review & editing, Methodology, Conceptualization. **Sarah de Loizaga:** Writing – review & editing, Methodology, Formal analysis, Conceptualization.

Declaration of competing interest

The authors have none.

References

- [1] D.A. Watkins, C.O. Johnson, S.M. Colquhoun, G. Karthikeyan, A. Beaton, G. Bukhman, M.H. Forouzanfar, C.T. Longenecker, B.M. Mayosi, G.A. Mensah, B. R. Nascimento, A.L.P. Ribeiro, C.A. Sable, A.C. Steer, M. Naghavi, A.H. Mokdad, C.J. L. Murray, T. Vos, J.R. Carapetis, G.A. Roth, Global, regional, and National Burden of rheumatic heart disease, 1990–2015, *N. Engl. J. Med.* 377 (2017) 713–722.
- [2] W.L. Schaffer, J.M. Galloway, M.J. Roman, V. Palmieri, J.E. Liu, E.T. Lee, L.G. Best, R.R. Fabsitz, B.V. Howard, R.B. Devereux, Prevalence and correlates of rheumatic heart disease in American Indians (the strong heart study), *Am. J. Cardiol.* 91 (2003) 1379–1382.
- [3] C. Sutcliffe, R.M. Close, A.M. Davidson, A. Reid, D. Quay, K. Nicolet, L.B. Brown, L. Grant, R. Weatherholtz, J. McAuley, L. Hammit, 453. High burden of invasive and severe group A Streptococcus disease among native Americans on the White Mountain apache tribal lands, *Open Forum Infect. Dis.* 6 (2019) S223.

- [4] A.C. Steer, J.R. Carapetis, T.M. Nolan, F. Shann, Systematic review of rheumatic heart disease prevalence in children in developing countries: the role of environmental factors, *J. Paediatr. Child Health* 38 (2002) 229–234.
- [5] L. Zühlke, G. Karthikeyan, M.E. Engel, S. Rangarajan, P. Mackie, B. Cupido-Katya Mauff, S. Islam, R. Daniels, V. Francis, S. Ogendo, B. Gitura, C. Mondo, E. Okello, P. Lwabi, M.M. Al-Kebisi, C. Hugo-Hamman, S.S. Sheta, A. Haileamlak, W. Daniel, D. Y. Goshu, S.G. Abdissa, A.G. Desta, B.A. Shasho, D.M. Begna, A. ElSayed, A. S. Ibrahim, J. Musuku, F. Bode-Thomas, C.C. Yilgwan, G.A. Amusa, O. Ige, B. Okeahialam, C. Sutton, R. Misra, A. Abul Fadl, N. Kennedy, A. Damasceno, M. U. Sani, O.S. Ogah, T.O. Elhassan, A.O. Mocumbi, A.M. Adeoye, P. Mntla, D. Ojji, J. Mucumbitsi, K. Teo, S. Yusuf, B.M. Mayosi, Clinical outcomes in 3343 children and adults with rheumatic heart disease from 14 low- and middle-income countries: two-year follow-up of the global rheumatic heart disease registry (the REMEDY study), *Circulation* 134 (2016) 1456–1466.

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