Journal of Paediatrics and Child Health

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ORIGINAL ARTICLE OPEN ACCESS

Acute Rheumatic Fever and Rheumatic Heart Disease in Children Aged Less Than 5 Years in the Northern Territory Between 2010 and 2020

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Received: 16 January 2025 | **Revised:** 6 May 2025 | **Accepted:** 3 September 2025

Funding: The authors received no specific funding for this work.

Keywords: Aboriginal and Torres Strait Islander people | acute rheumatic fever | paediatrics | rheumatic heart disease

ABSTRACT

Aim: Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) are preventable diseases affecting socioeconomically disadvantaged populations globally, including Australian children. This study aims to describe the clinical presentation and outcomes of ARF and RHD in children aged less than 5 years, to improve recognition and management.

Method: A retrospective audit was undertaken of children aged less than 5 years with ARF and RHD in the Northern Territory (NT) of Australia between 2010 and 2020. Patients were identified from the NT RHD register. Descriptive data analyses were performed to summarise patient demographics, clinical presentation, diagnosis, disease severity, management, and outcomes.

Results: During the study period, 82 children (51% female) aged less than 5 years presented with definite, probable, or possible ARF (78/82), and/or RHD (20/82). Aboriginal and Torres Strait Islander children were disproportionately affected, with average annual incidences (2010–2020) of ARF and RHD of 127 (95% confidence interval [CI]: 99–155) and 33 (95% CI: 18–47) per 100,000 population, respectively. ARF recurrence was common (23%). At diagnosis, RHD was moderate to severe in 55% of cases, and 20% of children with RHD required one or more cardiac surgeries.

Conclusion: Children aged less than 5 years living in remote NT are at risk of ARF and RHD. Aboriginal and Torres Strait Islander children are disproportionately affected. Presentations with sore joints, chorea, new cardiac murmurs or other symptoms suggestive of ARF should be fully investigated. Echocardiographic screening for RHD should be considered for children living in remote NT from the age of 3 years.

1 | Background

Acute rheumatic fever (ARF) results from an immunological response following group A streptococcus (GAS) infection. Carditis associated with ARF can result in rheumatic heart disease (RHD), which most commonly affects the mitral and aortic valves. ARF and RHD are preventable diseases associated with social disadvantage, household crowding, and poverty [1]. Timely treatment of GAS infections, commencement of secondary antibiotic prophylaxis for at-risk individuals, and improved access to appropriate healthcare and adequate living conditions, all reduce the risk of ARF and development of RHD [2].

ARF diagnosis relies on clinician awareness and expertise, and application of revised Jones criteria [3]. Missed ARF diagnoses are associated with increased risk of repeated ARF episodes

https://doi.org/10.1111/jpc.70194

Summary

- What is already known on this topic?
  - Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) are preventable diseases of socioeconomic disadvantage with lifelong health, social, cultural, and financial burdens.
  - Aboriginal and Torres Strait Islander children living in the remote Northern Territory (NT) of Australia are disproportionately affected by ARF and RHD.
  - There are limited data describing the epidemiology, clinical features, and outcomes of ARF and RHD in children aged less than 5 years.
- What this paper adds?
  - Aboriginal and Torres Strait Islander children less than 5 years, living in the NT have an average annual incidence of first known ARF and RHD of 127 per 100,000 population (95% CI: 99–155) and 33 per 100,000 population (95% CI: 18–47), respectively.
  - Similar to older children, the most common manifestations of ARF in this cohort are joint symptoms. Presentations with sore joints, chorea, new cardiac murmurs or other symptoms suggestive of ARF in this population should be thoroughly investigated with blood tests, electrocardiogram and echocardiogram.
  - At initial RHD diagnosis, 55% of children had moderate to severe disease and 20% with RHD underwent one or more cardiac surgeries. Echocardiographic screening for RHD should be considered for children living in remote NT from the age of 3 years.

and progression of RHD, which could be prevented with early recognition of ARF and commencement of secondary antibiotic prophylaxis [4]. In the Northern Territory (NT) following a first episode of ARF, 20% will have a recurrence within 10 years and more than half of patients will develop RHD of which a third will be severe RHD [4, 5]. This can lead to significant morbidity and mortality including complications of heart failure, arrhythmia, stroke, and death [4, 5].

ARF and RHD are most common in low and middle-income countries; however, some of the highest rates in the world are seen in Aboriginal and Torres Strait Islander children living in regional and remote central and northern Australia [6]. In Australia, 91% of ARF and 79% of RHD cases occur in the Aboriginal and Torres Strait Islander population [7].

ARF most commonly affects children aged 5 to 14 years. Although RHD is most prevalent in adults over 35 years, 28% of Australians living with RHD are less than 25 years of age [7]. ARF and RHD do occur in children aged less than 5 years, but the clinical features and epidemiology of ARF and RHD in this age group have not been well described. Limited published data suggest that compared with older children, those diagnosed prior to the age of 5 years are more likely to present with arthritis and moderate to severe carditis and are less likely to present with chorea [8].

Improved understanding of the epidemiology and clinical features of ARF and RHD in children aged less than 5 years is important for informing clinician education and developing appropriate public health strategies for this age group. The NT RHD Control Program and RHD Register was established in 1997, which has allowed for more accurate reporting of ARF and RHD in the NT [4]. In this study, we aim to describe the epidemiology, clinical presentation, disease severity, management, and outcomes of children aged less than 5 years age who reside in the NT of Australia, diagnosed with the first episode of ARF and/or RHD during 2010–2020.

2 | Methodology

A retrospective audit was performed, including children aged less than 5 years who were diagnosed with the first presentation of ARF and/or RHD in the NT between January 2010 and December 2020. Patients with ARF and RHD were identified from the NT RHD Register. Demographic and clinical data from 2010 to 2020 were extracted from the register database, NT Department of Health Hospital Medical Records and the NT Cardiac Database using hospital record numbers. Patients were excluded if diagnosed outside the study dates (January 2010—December 2020), aged ≥ 5 years at the time of diagnosis, or if their residential address was outside the NT at the time of diagnosis. ARF recurrences were noted for each patient; however, they were not included in defining the cohort data.

ARF was classified as either definite, probable, or possible ARF based on the Rheumatic Heart Disease Australia Guidelines for the Prevention, Diagnosis and Management of Acute Rheumatic Fever and Rheumatic Heart Disease (3rd Edition, 2020) [4], which aligns with the 2015 American Heart Association Jones criteria [2–4]. RHD was diagnosed according to the 2012 World Heart Federation guidelines [9]. Streptococcal titres, acute phase reactant markers including creactive protein (CRP) and erythrocyte sedimentation rate (ESR), and P-R intervals were compared against reference ranges in the RHD Australia Guideline [4] for children aged 3 years and older. The reference range for P-R interval for children aged less than 3 years was taken from Park et al. [10].

Descriptive analyses were performed on the data using Microsoft Excel (Microsoft Corporation, Redmond, WA, USA), and proportions are described as a percentage of the cohort of children with ARF or RHD. Incidence rates were calculated using the Australian Bureau of Statistics (ABS) 2016 census population data for the denominator less than 5 years old [11], with case numbers averaged over 11 years from 2010 to 2020 and presented with 95% confidence intervals [12].

Ethics approval was granted by the Human Research Ethics Committee of the NT Department of Health and Menzies School of Health Research (HREC: 2021–4153) and the Central Australian Human Research Ethics Committee (CAHREC: 22–4378).

3 | Results

There were 82 children aged less than 5 years diagnosed with their first episode of ARF and/or RHD and notified to the RHD

register between 2010 and 2020 in the NT. ARF was diagnosed in 78 children, including 36/78 (46%) definite episodes and 42/78 (54%) uncertain episodes (9 probable and 33 possible). RHD was diagnosed in 20/82 children (24%), of which 16 were diagnosed alongside a diagnosis of ARF and four following investigations of a new cardiac murmur without a concurrent or prior ARF diagnosis. Among Aboriginal and Torres Strait Islander children living in the NT aged less than 5 years, the average annual incidence (2010–2020) of the first episode of definite, probable, or possible ARF was 127/100000 population (95% CI 99–155), and of newly diagnosed RHD was 33/100000 population (95% CI 18–47).

Most children diagnosed with ARF and/or RHD were Aboriginal and/or Torres Strait Islander (81/82, 99%) and lived in remote communities (70/82, 85%). Sex distribution was equal among those with ARF (39/78, 50% female), while RHD showed a female predominance (14/20, 70% female). The median age at first diagnosis was 4.4 years for definite ARF (range 2.7–4.9) and 4.5 years for RHD (range 2.8–4.9; Table 1, Figure 1). The median follow-up was 3.3 years (range 0.3–9.3) from initial ARF diagnosis, and 4.6 years (range 0.7–8.3) from initial RHD diagnosis. This reflects that most ARF diagnoses occurred in the final 4 years of the study, while RHD diagnoses were more evenly distributed across the study period.

Of the 78 children diagnosed with ARF, joint involvement was the most common manifestation (69/78, 88%; Table 2, Figure 2). Arthritis was the most frequently fulfilled diagnostic criterion (43/78, 55%), with 29 (29/78, 37%) children presenting with aseptic monoarthritis and 14 (14/78, 18%) with polyarthritis. Arthralgia was reported in 31 children (31/78, 40%), 22 (22/78, 28%) presenting with polyarthralgia and 9 (9/78, 12%) with monoarthralgia. Both monoarthritis and polyarthralgia were recorded in 5 children (5/78, 6%), but appropriately only counted once in the diagnostic criteria. Carditis was confirmed on echocardiography in 16 (16/78, 21%) children. Four (4/78, 5%) children had Sydenham chorea, all of whom had concurrent RHD at initial diagnosis. Subcutaneous nodules and erythema marginatum were not identified in any patients. Elevated inflammatory markers were present in 71 children, with 52 (52/78, 67%) meeting ARF diagnostic criteria as defined by the national guidelines. Among 66 children with an ESR recorded, 49 (49/66, 74%) met criteria, and a CRP value meeting diagnostic thresholds was documented in 34 children (34/78, 44%). The median CRP was 29 mg/L, and median ESR was 47 mm/h. Fever at presentation was noted in 51 (51/78, 65%) children. Electrocardiograms (ECGs) were recorded for 68 children with ARF (68/78, 87%), with 10 (10/78, 13%) unavailable due to incomplete workup or absent documentation. Of those performed, 16 ECGs (16/68, 24%) were abnormal, of which 15 (15/16, 94%) had first-degree heart block and one (1/16, 6%) had a junctional rhythm. Family history was unavailable in 49 cases (49/78, 63%). Among the 29 with available data, 25 (25/29, 86%) had a family history of ARF and/or RHD, and four (4/29, 14%) had no significant family history.

Two-thirds of the cohort had a documented preceding infection (54/82, 66%), of which 26 (26/54, 48%) were skin infections, 18 (18/54, 33%) were a sore throat or upper respiratory tract infection, five (5/54, 9%) were ear infections, four (4/54, 7%) had both skin infection and sore throat, and one patient had skin and ear infections. GAS was isolated from clinical samples from eight patients, two from throat swabs and six from skin swabs.

Most children (67/82, 81%) were hospitalised for additional investigations, acute management, and education, with a median length of stay of 4 days. An echocardiogram was performed at ARF diagnosis in 96% (75/78) children, with the remaining three children having a normal echocardiogram at a later time point (range 4–60 months following initial diagnosis; median 10 months). Steroids were administered to five children (5/82, 6%), four for carditis, and one for chorea.

Of the 20 children with RHD, 9 (45%) had mild RHD at initial diagnosis, eight (40%) had moderate RHD and three (15%) had severe RHD. Isolated mitral valve involvement was present in 11 cases (55%), isolated aortic in four (20%), and combined mitral and aortic in five (25%). Left ventricular dilatation was noted in

TABLE 1 | Demographics of children less than 5 years diagnosed with a first known episode of ARF and/or RHD in the Northern Territory, 2010–2020.

Demographics	Definite ARF ($N=36$)	Probable or possible ARF ($N=42$)	RHD ($N=20$)	Total cohort ($N=82$)
Sex				
Female	21 (58%)	18 (43%)	14 (70%)	42 (51%)
Male	15 (42%)	24 (57%)	6 (30%)	40 (49%)
Geographic residence				
Remote	32 (89%)	35 (83%)	18 (90%)	70 (85%)
Urban	4 (11%)	7 (17%)	2 (10%)	12 (15%)
Age at first diagnosis (years)				
Median (range)	4.4 (2.7–4.9)	4.1 (2.2–4.9)	4.5 (2.8–4.9)[a]	4.3 (2.2–4.9)[b]

Abbreviations: ARF, acute rheumatic fever; RHD, rheumatic heart disease.
[a]Two additional male children were diagnosed with RHD during follow-up at age 5.2 and 8.9 years, one and 5 years after their initial ARF diagnosis respectively.
[b]Age (years) at first diagnosis of ARF and/or RHD.

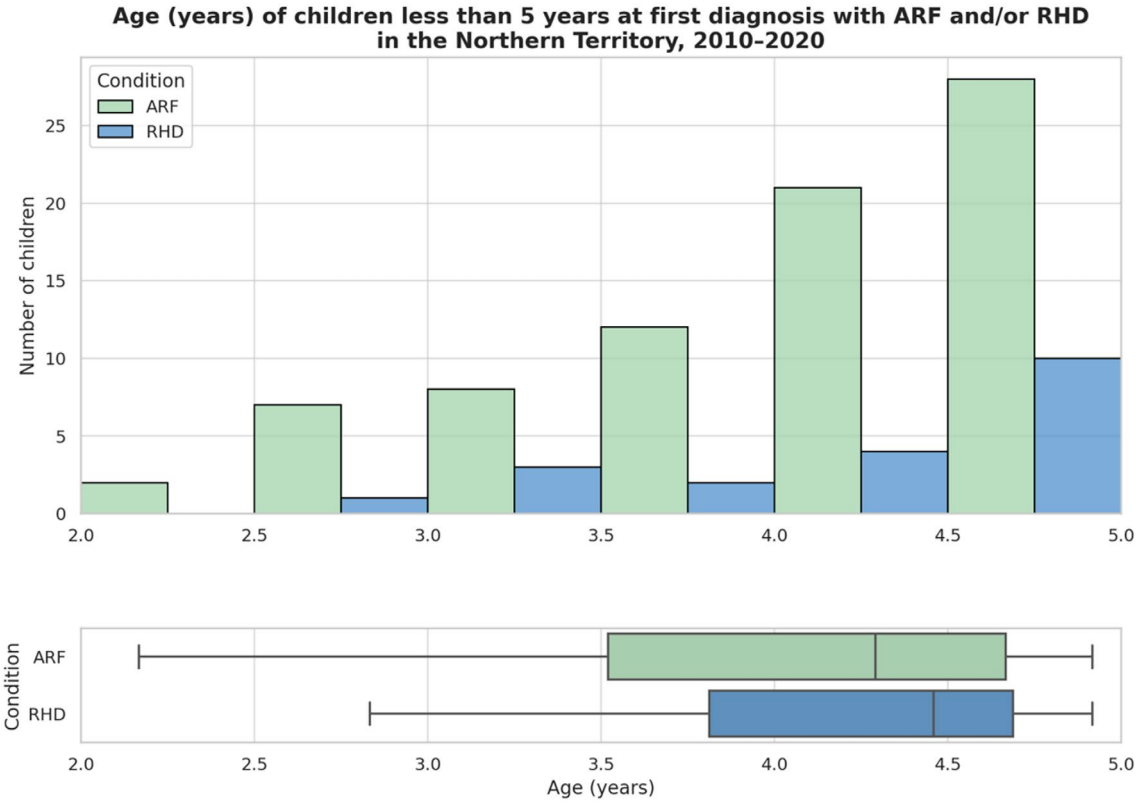

FIGURE 1 | Age (years) of children less than 5 years at first diagnosis with Acute Rheumatic Fever (ARF) and/or Rheumatic Heart Disease (RHD) in the Northern Territory, 2010–2020. Histogram shows the number of children diagnosed with ARF ($n = 78$) and RHD ($n = 20$) by age at diagnosis, displayed in 0.5-year intervals. Box plots depict the median (line), interquartile range (box), and range (whiskers) of age at first diagnosis for each group. ARF: Acute Rheumatic Fever; RHD: Rheumatic Heart Disease. Footnote: Two additional patients were diagnosed with RHD during follow-up at ages 5.2 and 8.9 years, one and 5 years after their initial ARF diagnosis respectively.

three children (15%), and one had pulmonary hypertension (5%). At study completion, 11 (55%) children showed improvement—six (30%) with resolved RHD and five (25%) with reduced severity. Six (30%) remained unchanged, and three (15%) had disease progression. Overall, five children (5/20, 25%) had severe RHD at some point during the study period.

Of the five children with severe RHD, four (80%) underwent cardiac surgery and one (20%) improved without surgery. The median time from RHD diagnosis to surgery was 4.3 years (range 0.1–5.9 years), with a median age at surgery of 8.4 years (range 5.0–10.5 years). Two patients underwent aortic valve repair, and one underwent mitral valve replacement. One child initially required both aortic and mitral valve repairs, followed by further cardiac surgery for a bioprosthetic aortic valve replacement and mechanical mitral valve replacement. Cardiac anti-failure medication was prescribed for four children with severe RHD (4/20, 20%). Two additional children were diagnosed with mild RHD during follow-up, at ages 5.2 and 8.9 years, occurring one and 5 years after their initial ARF diagnosis, respectively. The younger child progressed to severe RHD, requiring an initial and subsequent revision aortic valve repair and was planned for a mechanical aortic valve and aortic root repair.

Penicillin prophylaxis was commenced in 80 of 82 children (98%), with most receiving intramuscular benzathine benzylpenicillin every 3 to 4 weeks. One child initially prescribed prolonged oral prophylaxis was transitioned to intramuscular penicillin following ARF recurrence. Secondary antibiotic prophylaxis was not commenced for one child with possible ARF and another with borderline RHD. ARF recurrence was reported in 23% of children (19/82) between 2010 and 2020 (median follow-up 3.3 years). ARF recurrences occurred in 11 children initially diagnosed with ARF only (three definite, eight possible), six with concurrent RHD and ARF (all definite) and two initially diagnosed with RHD alone, without a recognised prior episode of ARF.

4 | Discussion

ARF and RHD are relatively commonly diagnosed in children aged between 2 and 5 years in the NT, with Aboriginal and Torres Strait Islander children disproportionately affected. The ARF incidence for Aboriginal and Torres Strait Islander children less than 5 years in the NT is more than 70 times higher than the national age-equivalent rate (127/100000 versus 1.7/100000 population) and is similar to the national 5–14 year Aboriginal and Torres Strait Islander population rate (130.9/100000) [7]. High rates of ARF and RHD in this population are a result of repeated exposure to GAS infections from a young age, which is influenced by socioeconomic disadvantage and household crowding [1, 2, 4, 5]. Prevention through improved living conditions, a GAS vaccine, and early recognition and treatment of GAS infections is paramount to reducing the burden of disease in this at-risk population [2, 4].

TABLE 2 | Major and minor modified Jones acute rheumatic fever (ARF) diagnostic criteria of children less than 5 years diagnosed with first known episode of ARF in the Northern Territory, 2010–2020.

Modified jones ARF diagnostic criteria	Definite ARF (*N* = 36)	Probable ARF (*N* = 9)	Possible ARF (*N* = 33)	Total ARF (*N* = 78)
Major criteria				
Arthritis	24 (67%)	3 (33%)	16 (48%)	43 (55%)
Aseptic Monoarthritis	13 (36%)	1 (11%)	15 (45%)	29 (37%)
Polyarthritis	11 (31%)	2 (22%)	1 (3%)	14 (18%)
Carditis	14 (39%)	1 (11%)	1 (3%)	16 (21%)
Sydenham chorea	4 (11%)	0 (0%)	0 (0%)	4 (5%)
Erythema marginatum	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Subcutaneous nodules	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Minor criteria				
Arthralgia	9 (25%)	5 (56%)	17 (51%)	31 (40%)
Monoarthralgia	1 (3%)	0 (0%)	8 (24%)	9 (12%)
Polyarthralgia	8 (22%)	5 (56%)	9 (27%)	22 (28%)
Fever	29 (81%)	6 (67%)	16 (48%)	51 (65%)
Raised acute phase reactants[a]	34 (96%)	6 (67%)	12 (36%)	52 (67%)
CRP ≥ 30 mg/L	25 (69%)	1 (11%)	8 (24%)	34 (44%)
ESR ≥ 30 mm/h	32 (89%)	6 (67%)	11 (33%)	49 (63%)
Abnormal ECG	12 (33%)	1 (11%)	3 (9%)	16 (21%)
First-degree heart block	11 (31%)	1 (11%)	3 (9%)	15 (19%)
Junctional rhythm	1 (3%)	0 (0%)	0 (0%)	1 (1%)

Note: Percentages, including for subcategories, are calculated using the total number of children within each ARF diagnostic group (Definite, Probable, Possible, Total) as the denominator.
Abbreviations: ARF, acute rheumatic fever; CRP, C-reactive protein; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate.
[a]Raised acute phase reactants defined as CRP ≥ 30 mg/L and/or ESR ≥ 30 mm/h.

ARF diagnosis is based on the modified Jones criteria, which includes a combination of clinical, echocardiographic, ECG, and laboratory data. Underdiagnosis of ARF leads to a risk of ARF recurrence and progression to RHD. While improved diagnostic tests are being developed, ARF diagnosis remains challenging [13, 14]. It is critical for clinicians and families to be aware that ARF and/or RHD can occur in younger children and to have a low threshold for investigation. Improved access to echocardiography, including abbreviated scans by trained healthcare workers, may help improve recognition, diagnosis, and early treatment [15].

Clinical findings in this cohort were similar to those observed in older children with ARF and/or RHD [4, 8, 16, 17]. Joint symptoms are the most common presenting symptom of ARF, with monoarthritis predominating over polyarthritis [17]. RHD is moderate to severe in 55% of children at diagnosis and 20% of RHD required at least one cardiac surgery, highlighting the significant lifelong consequences of undetected or untreated GAS infections and ARF. Given the absence of cardiac symptoms in many cases, and the relatively high rates of RHD, inclusion of younger children in existing high-risk population community-wide echocardiography screening for RHD should be considered. In support, ARF incidence in this cohort surpasses the World Heart Federation recommended threshold of 30/100,000 population for 5–14-year-olds, advised when implementing echocardiography-based screening [18]. Most cases of RHD were diagnosed from the age of 4 years, although some cases were as young as two or 3 years.

An ARF and/or RHD diagnosis in a young child has significant lifelong impacts for the individual, their families, and the healthcare system. Secondary penicillin prophylaxis aims to reduce the risk of recurrence of further ARF and development of RHD; however, it can be traumatic, costly, and is long-term [4, 19]. All children less than 5 years with probable or definite ARF are recommended to receive four-weekly IM benzathine penicillin prophylaxis until the age of 21 years, while those with moderate or severe RHD may require prophylaxis until the age of 35 to 40 years, or even for life [4]. In our cohort, ARF recurrence was common (19/82, 23%) and is possibly underestimated due to the short follow-up period from initial diagnosis (median 3.3 years; range 0.2–9.3). While prophylaxis is burdensome and a substantial proportion of children experienced severe RHD

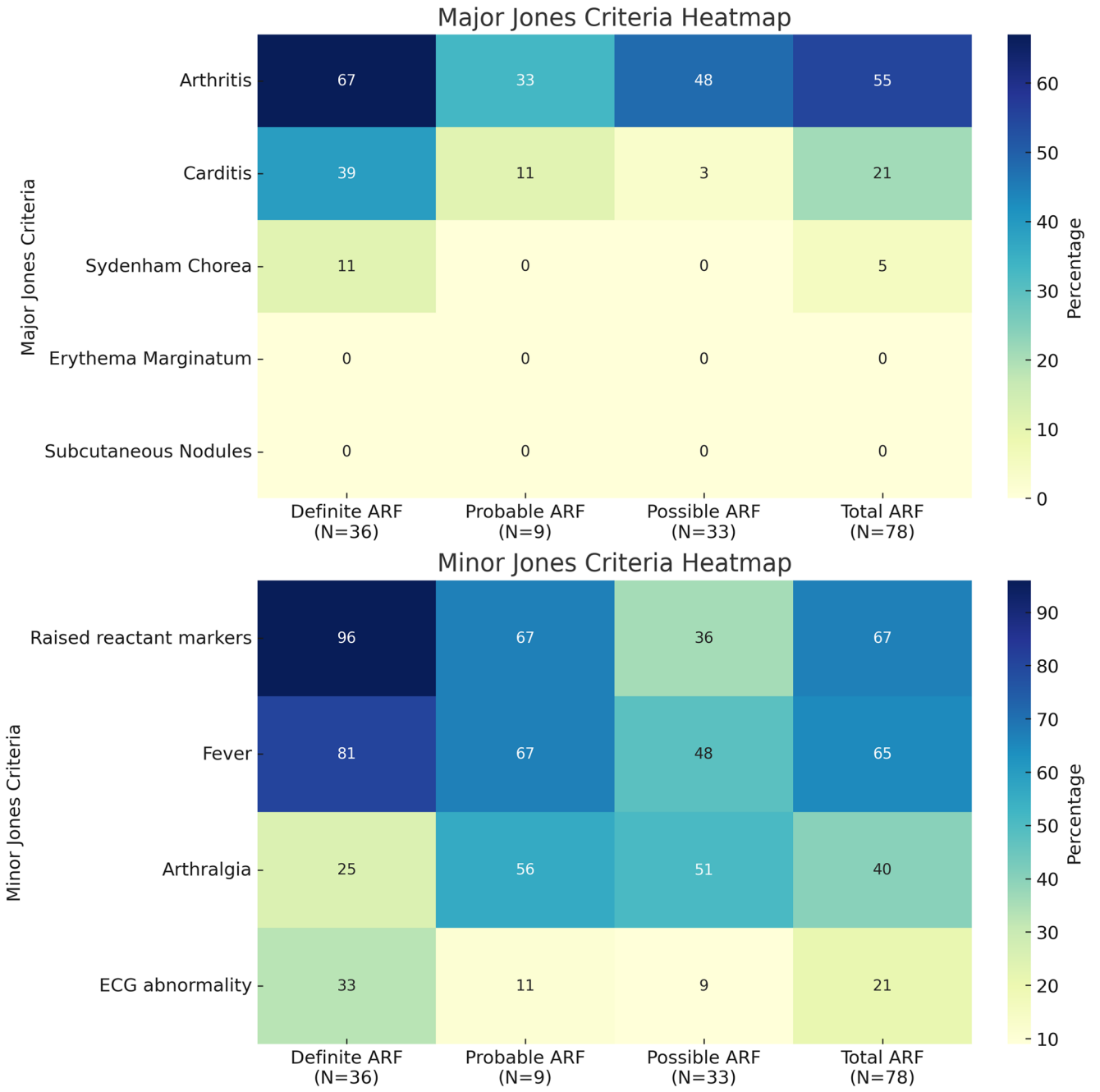

FIGURE 2 | Heatmap of major and minor modified Jones diagnostic criteria met during the first known episode of acute rheumatic fever (ARF) among children less than 5 years of age living in the Northern Territory, 2010–2020. Percentages are calculated using the total number of children within each diagnostic group (Definite ARF, Probable ARF, Possible ARF, Total ARF) as the denominator. ARF, acute rheumatic fever; ECG, electrocardiogram.

requiring long-term management, the overall prognosis was favourable, with 60 children who did not develop RHD and six whose RHD had resolved by the end of the study period.

The retrospective nature of this study is an important limitation, resulting in significant data gaps. Missing data may have arisen from incomplete workup, poor documentation, and/or data from primary care systems not accessible through the methodology used [20]. The study's strength, however, is the use of multiple data sources. There may be a bias towards recognition and diagnosis of more severe symptoms, and rates of ARF and mild RHD may be underestimated due to under-recognition of ARF and RHD in this age group.

ARF and RHD are preventable diseases of socioeconomic disadvantage with lifelong health, social, cultural, and financial burdens. Young children aged less than 5 years living in remote NT are at risk of ARF and RHD. Presentations with sore joints, chorea, new cardiac murmurs, or other symptoms suggestive of ARF should be fully investigated, and in settings where

echocardiographic screening for RHD is being implemented, consideration should be given to extending screening to children living in remote NT from the age of 3 years.

Ethics Statement

Ethics approval was granted by the Human Research Ethics Committee of the NT Department of Health and Menzies School of Health Research (HREC: 2021–4153) and the Central Australian Human Research Ethics Committee (CAHREC: 22–4378).

Conflicts of Interest

The authors declare no conflicts of interest.

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