

REVIEW

Rheumatic Heart Disease in Pregnancy: New Strategies for an Old Disease?

Geraldine Vaughan¹, Angela Dawson², Michael Peek³, Karen Sliwa⁴, Jonathan Carapetis⁵, Vicki Wade⁶ and Elizabeth Sullivan⁷

¹ Central Queensland University, College of Science and Sustainability, Sydney campus, AU

² University of Technology Sydney, Faculty of Health, Sydney, AU

³ The Australian National University, College of Health and Medicine, Canberra, AU

⁴ University of Cape Town, Cape Town, ZA

⁵ Telethon Kids Institute, University of Western Australia, and Perth Children's Hospital, Nedlands WA, AU

⁶ Menzies School of Health Research, NT, AU

⁷ The University of Newcastle, Faculty of Health and Medicine, Newcastle, NSW, AU

Corresponding author: Geraldine Vaughan (g.a.vaughan@cqu.edu.au)

RHD in pregnancy (RHD-P) is associated with an increased burden of maternal and perinatal morbidity and mortality. A sequellae of rheumatic fever resulting in heart valve damage if untreated, RHD is twice as common in women. In providing an historical overview, this commentary provides context for prevention and treatment in the 21st century.

Four underlying themes inform much of the literature on RHD-P: its association with inequities; often-complex care requirements; demands for integrated care models, and a life-course approach. While there have been some gains particularly in awareness, strengthened policies and funding strategies are required to sustain improvements in the RHD landscape and consequently improve outcomes.

As the principal heart disease seen in pregnant women in endemic regions, it is unlikely that the Sustainable Development Goal 3 target of reduced global maternal mortality ratio can be met by 2030 if RHD is not better addressed for women and girls.

Keywords: Rheumatic heart disease; Pregnancy; First Nation peoples; Inequity

A History of Broken Hearts: Rheumatic Heart Disease in Pregnancy

In 2021, a young pregnant Aboriginal woman with rheumatic heart disease (RHD) was told '*...didn't know we had that (RHD) anymore.*' Yet this preventable disease continues to devastate Aboriginal and Torres Strait Islander communities. Despite being a high-income country overall, First Nation Australians experience some of the highest documented rates of RHD in pregnancy in the world, particularly remote communities in Northern Australia which face significant disadvantage and lack of services. However, overall, low-resource regions sustain the global burden of RHD: 80% of people with RHD live in low-and middle-income countries (LMICs) [1].

A sequellae of acute rheumatic fever (ARF) that results in lasting heart valve damage if untreated [2], RHD is twice as common in women, most likely associated with several factors including increased risk of autoimmune disease, accelerated progression of mild RHD during pregnancy and increased exposure to StrepA infection. What is the epidemiology of this ancient disease and how has it been addressed in pregnancy through the millennia? In providing an historical overview, this commentary provides context for the prevention and management in the 21st century. It considers how lessons learnt in the 19th–20th century are of relevance today.

While health systems across the centuries and across the world may differ enormously according to developments in knowledge, technologies, economies, and social structures, four underlying themes inform much of the literature of RHD in pregnancy (RHD-P): the association of RHD with inequities and poverty; recognition of often-complex care requirements during pregnancy; demands for integrated care models to optimise maternal and perinatal outcomes, and a life-course approach to RHD for women and girls.

Risk Through the Millennia for Pregnant Women with RHD

Nearly 2,500 years ago, Hippocrates discussed the classic symptoms of rheumatic mitral stenosis:

'Harpalida's sister, in the fourth or fifth month of her pregnancy, had watery swellings in her legs, swellings in the hollows of her eyes, and her whole body puffed up... Sometimes she was so near to suffocation that she was obliged to sit up in her bed without being able to lie down; and if she tried to sleep it was in a sitting position. Yet there was not much fever. For a long time the foetus did not move, as if it were dead....' Epidemics VII VI (Littre, 1839–1861; Clifton, 1734) in [3].

In the 1800s, several writings noted poor outcomes of pregnancy complicated by cardiac disease, which was overwhelmingly caused by RHD in the overcrowded urbanised centres of the United Kingdom (UK), Europe and North America. The death of a young woman in labour due to (rheumatic) mitral stenosis was a key driver in Scottish cardiologist James Mackenzie's work to improve the clinical management of heart disease [4].

I discovered that [a young woman who had a miscarriage] had a presystolic murmur... I had a vague notion that mitral stenosis was a serious embarrassment to the heart in pregnancy, but of the source of danger and its nature I had no knowledge. The woman ...[later] returned ... seven months pregnant, and with a considerable oedema of the legs. ... As the labour proceeded little progress was made; the suffering was very great, and there was marked distress in breathing... after many hours of suffering she died undelivered [5].

Around the same time, obstetrician Angus Macdonald wrote his defining textbook on cardiac disease in pregnancy. There was little on congenital heart disease: in the nineteenth century, women rarely survived to adulthood. Much of his focus was on mitral stenosis of rheumatic origin:

We have thus nine cases out of the fourteen [pregnancies], or 64.4%, fatal, which indicates a tendency to death in the combination of mitral stenosis with pregnancy which is surely sufficiently grave... [6].

However, despite such outcomes, clinicians such as McKenzie, Macdonald and others did not proscribe pregnancy altogether [7], but counselled according to lesion and clinical status, developing a risk stratification in clinical care, concluding that...

These good results encourage us to believe that judicious and skilful management of similar cases, both throughout the pregnancy and at delivery, might greatly lessen the risks associated with lesions [of mitral stenosis] [6].

Macdonald also mused on the ethical responsibilities of obstetricians to advise women on pregnancy choices (with marriage seen as a proxy), which he argued were largely unconsidered:

I know of no English writer who has striven to put obstetricians in a position to answer with intelligence the question that is every now and again asked of us by a patient who knows she has heart disease, — Should I marry? or do I run great risk in marrying? ... [a] great amount of ignorance exists [6].

In the late 1940s, the first known longitudinal study of maternal outcomes used the recently developed New York Heart Association (NYHA) functional classification of cardiac disease based on clinical severity and prognosis to mark the progression of cardiac disease in 169 pregnant women [8–13].

This article drew on others' research to conclude that RHD of itself was not an indication for caesarean section, counselling that this should be avoided unless indicated by obstetric reasons or severe cardiac compromise [10, 12–14].

'The Cramping Effects of Too-Rigid Specialism': Care Across Disciplines

Integrated care across specialisations was a critical aspect of developing strategies to address the risk for women with RHD-P. There had been calls for collaborative cardiac care for pregnant women from the late nineteenth century. Macdonald admonished his British obstetric colleagues to devote more attention to cardiac disease in pregnancy ... *'commensurate with its importance'* in order to better *'...differentiate the effects of*

the special cardiac lesions, and to define in any way their individual bearings upon either pregnancy or parturition. 'Obstetricians' *'rigid specialism'* he declared, compromised effective management of women under their care, and led to much of the *'disfavour and affected hauteur with which obstetrical medicine has frequently been treated by pure physicians and surgeons'* [6].

Good nursing care was considered *'...likely to be better rewarded ... in warding off the exciting causes of pulmonary disturbances in connection with pregnancy complicated with various cardiac lesions'* [6], but disciplinary references outside cardiac and obstetric specialisations were largely absent.

However, the next half-century did not bring much evidence of better awareness nor improved collaborative care. Some forty years later, McKenzie observed the knowledge of cardiac disease among obstetric physicians [5]. He spoke of the fear of consequences for women and the need to provide informed and educated advice:

These disastrous [cardiac] happenings surround a natural process with dread and mystery, for the dread is aggravated by the fact that the source of danger is not clearly realised. As a result all sorts of signs are looked upon with suspicion—signs innocent as well as signs grave. Needless to say, this obscurity does a great deal of harm. Many women are subjected to unnecessary alarms and restrictions when pregnant ; others have to suppress the natural desire of motherhood... [5].

The collaborative models that Macdonald and McKenzie called for were being slowly developed. A Scottish joint cardiac-obstetric clinic established in 1928 saw cardiac deaths as a percentage of cardiac cases (94% with underlying rheumatic pathology) drop from 6.3 to 0.9% by 1947 [10].

Hamilton described similar trends in North American centres. When a cardiac clinic commenced at Boston Lying-In Hospital in 1921, the maternal mortality rate was 20% (93% due to RHD), which dropped to under 5% in the ensuing three years [15]. Review and treatment (rather than the RHD condition per se) was among the largest prognostic factor in this study, with regular antenatal review during pregnancy stressed as an essential component of care [15].

A 1936 Canadian study called for the... *'establishment of a combined prenatal and cardiac clinic which has permitted personal consultations between cardiologist and obstetrician and a continuity of observation of all heart cases'* [16].

From the 1930s, similar studies in London and Dublin found that, with adequate collaborative cardiac-obstetric care the need for medically advised terminations of pregnancy was reduced significantly and outcomes improved [17, 18]. McIlroy (the first UK female obstetrician and medical professor), in recognising the overwhelming burden of RHD in cardiac pathology, detailed the mutual benefits of obstetric-cardiac collaboration, including earlier diagnosis of cases [17]. A review of women with RHD-P at two UK sites over 28 years (1942–1969) concluded that *'routine medical examinations at about 10, 15, and 20 years of age with appropriate management would render pregnancy virtually safe for every patient with rheumatic heart disease'* [19].

However, such outcomes were challenged by a lack of interdisciplinary collaborative approach to care and gaps in health system support which persists to the current day:

Another difficulty arises from the different way in which the cardiologist and the obstetrician regard the problem. The cardiologist considers the pregnancy as complicating the pre-existing heart disease, but the obstetrician considers that his [sic] patient's pregnancy is complicated by her cardiac lesion. Another cause for ignorance is that the obstetrician rarely studies his patients once the puerperium has been successfully passed, and if further pregnancies do not ensue he may never see the patient again. On the other hand, the cardiologist rarely follows the same patient through several pregnancies unless he is particularly interested in the subject, and his opportunities for observing closely the heart in normal pregnancy are few [13].

The Shifting Global Burden of RHD

The epidemiology of RHD generally mirrors the evolution of nation economies and associated changing health profiles. By the 1960s, the overall prevalence of RHD was waning in high-income countries (HIC) [20–24]. Studies of maternal morbidity and mortality trends in HIC began noting the increasing proportion of women with congenital heart disease compared to a decrease in RHD [25–27] during pregnancy.

Unsurprisingly, this decline was not shared by low-income countries: in the 21st century, the global burden of disease has shifted to (in particular) regions such as Oceania, South Asia, central sub-Saharan Africa [28–30], the Caribbean and Latin America [31], with a correspondingly high burden in pregnancy [32–39].

Reflecting the association of RHD with inequities, the reduced burden of disease has varied *within* HICs [40–45], with an increasing (and persistent) trend among First Nations [2, 40, 46–48], as well as migrants from low-resource countries [2, 49, 50].

The early 2000s saw a shift in policy and political will related to RHD, no doubt galvanised by increasing advocacy and locally-driven initiatives, and supported by increasing evidence of the global burden of RHD [1, 50–58]. Several LMICs have formed national programs dedicated to RHD prevention and control [59–62], with a 2013 World Heart Federation (WHF) goal of a 25% reduction in premature deaths from RF/RHD among individuals aged <25 years by the year 2025 [63]. An important development has been the increased focus on (and working with) people living with RHD and frontline health workers delivering essential RHD services [64–70].

The need to situate RHD strategies within the context of maternal health and access to services was of critical note in the 2018 World Health Organization's first global policy on ARF/RHD [71]. Improved access to reproductive health services for women with RHD and other non-communicable diseases (NCD) is one of the seven key priority actions called for in a roadmap to eliminate RF and eradicate RHD in Africa [52, 72].

Why So Complicated? Barriers to Optimal Care for Women with RHD

While challenges to optimal care in the RHD and pregnancy landscape can be broadly categorised –access to multidisciplinary health services, continuity of care, geographical distances, health workforce resourcing and education (particularly in primary care), sustained support for community-driven initiatives [56, 68–70, 73], fragmented health information silos [74] – they form a complex matrix of contributing factors that can impact directly and on each other. Underpinning each of these are the policies, governance, political will and practical decisions that shape the promotion – or hindering – of culturally safe and effective models of care.

The additional risks in pregnancy of a compromised heart through (possibly undiagnosed) RHD further escalates where anticoagulation and interventional treatments are required, and highlights the necessity for a life-course approach to care including preconception (addressing needs for young people transitioning to adulthood), pregnancy (including early and regular antenatal reviews), the postpartum and interpregnancy periods.

Yet, the compelling need to redress the neglect of RHD and RHD-P in public health and funding arenas persists. Even ignoring the critical social justice issues of RF/RHD, its associated fiscal burden demands significantly better support [75–77]: it costs USD5 to treat a sore throat, compared to the lifetime burden associated with valvular surgery upwards of USD29,000 [56, 62, 78, 79]. The maternal and perinatal costs of the burden of RHD are exponentially higher [80], particularly when the indirect costs of economic impact are taken into account – family, community, work – are taken into account.

Work with Us: Integration and Collaboration

While specific approaches to best-practice care for women with RHD in pregnancy will vary according to country, culture and resources; general principles emphasise the continuity of care, underpinned by collaborative partnerships across sectors [34, 45, 48, 64, 70, 81, 82].

Of course, many principles of care – preconception woman-centred care, a multidisciplinary approach to service development and care guidelines – are equally applicable to women with all-cardiac disease [82, 83]. However, as a sentinel disease of inequity, RHD exemplifies the 'causes of the causes' that underpin compromised health [84], are particularly relevant in pregnancy.

Improved maternal and perinatal outcomes afforded by multidisciplinary models, particularly joint obstetric–cardiac care for women with complex disease are evident across the spectrum of low-resource [81, 85], and high-income settings [45, 82, 86].

Conclusion

This historical perspective illustrates some of the themes and issues central to care pathways for women with RHD-P: diagnosis, pregnancy planning, risk assessment, often-complex care needs, continuity of care and access to woman-centred integrated health services. These physical and structural imperatives are against a backdrop of often-resource challenged environments that test optimal care and outcomes for mother and baby.

The 19th century strategies that called for collaborative models and cross-sectoral partnerships are as relevant today. While there have been some gains particularly in awareness of the impact of RHD in pregnancy, we need the societal and political will, sound policies and funding strategies required to support sustained and meaningful improvements in the RHD landscape and consequently improved care in pregnancy.

As the principal heart disease seen in pregnant women in endemic regions, it is unlikely that the Sustainable Development Goal 3 target of reducing the global maternal mortality ratio can be met by 2030 if RHD is not better addressed for women and girls.

Competing Interests

The authors have no competing interests to declare.

References

1. **Shawar YR, Shiffman J.** Generating Global Priority for Addressing Rheumatic Heart Disease: A Qualitative Policy Analysis. *J Am Heart Assoc.* 2020; 9(8): e014800. DOI: <https://doi.org/10.1161/JAHA.119.014800>
2. **Wyber R, Noonan K, Halkon C,** et al. Ending rheumatic heart disease in Australia: The evidence for a new approach. *Medical Journal of Australia.* 2020; 213(S10): S3–S31. DOI: <https://doi.org/10.5694/mja2.50853>
3. **Katz AM, Phyllis B.** Diseases of the heart in the works of Hippocrates. *British Heart Journal.* 1962; 24(3): 257–64. DOI: <https://doi.org/10.1136/hrt.24.3.257>
4. **Trail TA.** Valvular Heart Disease and Pregnancy. *Cardiology Clinics.* 2012; 30(3): 369–81. DOI: <https://doi.org/10.1016/j.ccl.2012.04.004>
5. **McKenzie J.** *Heart disease and pregnancy.* London: Oxford Medical Publications; 1921.
6. **Macdonald A.** *The bearings of chronic disease of the heart upon pregnancy, parturition and childbed.* London, UK: J&A Churchill; 1878.
7. **Mackness G.** Heart disease in pregnancy and labour. *The Transactions of the Edinburgh Obstetrical Society, Session 1889–1890.* XV. Edinburgh: Oliver and Boyd; 1890.
8. **The Criteria Committee of the New York Heart Association.** 1994 Revisions to Classification of Functional Capacity and Objective Assessment of Patients With Diseases of the Heart: NYHA Functional class. In Dolgin M (Ed.), *Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Great Vessels*, 253–6. 9th ed. Boston, Mass: Little, Brown & Co; 1994.
9. **Jensen J.** *The Heart in Pregnancy.* London: H. Kimpton; 1938. DOI: <https://doi.org/10.1097/00000441-193811000-00035>
10. **Haig D, Gilchrist AR.** Heart disease complicated by pregnancy. *Trans Edin Obst Soc;* 1949.
11. **Bunim J, Appel S.** A principle for determining prognosis of pregnancy in rheumatic heart disease. *JAMA-J AM MED ASSOC.* 1950; 142. DOI: <https://doi.org/10.1001/jama.1950.02910200018005>
12. **Sutherland AM, Mair JT, Bruce DF.** The management of pregnancy associated with cardiac disease. *Glasgow Medical Journal.* 1955; 36(12): 411. DOI: <https://doi.org/10.1136/oem.12.1.36>
13. **Oram S.** Pregnancy and rheumatic heart disease. *Postgraduate Medical Journal.* 1956; 32(364): 76–84. DOI: <https://doi.org/10.1136/pgmj.32.364.76>
14. **Hamilton B, Thomson K.** *The heart in pregnancy and the childbearing age.* Boston: Little Brown and Co; 1941.
15. **Hamilton BE.** Report from the cardiac clinic of the Boston Lying-in Hospital for the first twenty-five years. *American Heart Journal.* 1947; 33(5): 663–8. DOI: [https://doi.org/10.1016/0002-8703\(47\)90082-3](https://doi.org/10.1016/0002-8703(47)90082-3)
16. **Henderson DN.** Pregnancy complicated by rheumatic heart disease. *Canadian Medical Association Journal.* 1936; 35(4): 394–8.
17. **McIlroy DL, Rendel O.** The Problem of the Damaged Heart In Obstetrical Practice. *BJOG: An International Journal of Obstetrics & Gynaecology.* 1931; 38(1): 7. DOI: <https://doi.org/10.1111/j.1471-0528.1931.tb13951.x>
18. **O'Driscoll MK, Coyle CF, Drury MI.** Rheumatic heart disease complicating pregnancy. The remote prospects. *British Medical Journal.* 1962; 2(5307): 767–8. DOI: <https://doi.org/10.1136/bmj.2.5307.767>
19. **Szekely P, Turner R, Snaith L.** Pregnancy and the changing pattern of rheumatic heart disease. *British Heart Journal.* 1973; 35(12): 1293–303. DOI: <https://doi.org/10.1136/hrt.35.12.1293>
20. **Gordis L.** The virtual disappearance of rheumatic fever in the United States: Lessons in the rise and fall of disease. *Circulation.* 1985; 72(6): 1155–62. DOI: <https://doi.org/10.1161/01.CIR.72.6.1155>
21. **Quinn RW.** Comprehensive review of morbidity and mortality trends for rheumatic fever, streptococcal disease, and scarlet fever: The decline of rheumatic fever. *Reviews of Infectious Diseases.* 1989; 11(6): 928–53. DOI: <https://doi.org/10.1093/clinids/11.6.928>

22. **Abajobir AA, Abbafati C, Abbas KM**, et al. Global, regional, and national age-sex specific mortality for 264 causes of death, 1980–2016: A systematic analysis for the Global Burden of Disease Study 2016. *The Lancet*. 2017; 390(10100): 1151–210. DOI: [https://doi.org/10.1016/S0140-6736\(17\)32152-9](https://doi.org/10.1016/S0140-6736(17)32152-9)
23. **Steer AC**. Historical aspects of rheumatic fever. *Journal of Paediatrics and Child Health*. 2015; 51(1): 21–7. DOI: <https://doi.org/10.1111/jpc.12808>
24. **Stanhope JM**. New Zealand trends in rheumatic fever: 1885–1971. *N Z Med J*. 1975; 82(551): 297–9.
25. **McFaul PB, Dornan JC, Lamki H, Boyle D**. Pregnancy complicated by maternal heart disease. A review of 519 women. *BJOG: An International Journal of Obstetrics & Gynaecology*. 1988; 95(9): 861–7. DOI: <https://doi.org/10.1111/j.1471-0528.1988.tb06570.x>
26. **Sachs BP, Brown DA, Driscoll SG**, et al. Hemorrhage, infection, toxemia, and cardiac disease, 1954–85: Causes for their declining role in maternal mortality. *American Journal of Public Health*. 1988; 78(6): 671–5. DOI: <https://doi.org/10.2105/AJPH.78.6.671>
27. **Hibbard LT**. Maternal mortality due to cardiac disease. *Clinical Obstetrics and Gynecology*. 1975; 18(3): 27–36. DOI: <https://doi.org/10.1097/00003081-197509000-00005>
28. **Negus RM**. Rheumatic fever in Western Fiji: The female preponderance. *Medical Journal of Australia*. 1971; 2(5): 251–4. DOI: <https://doi.org/10.5694/j.1326-5377.1971.tb50534.x>
29. **Watkins D, Johnson C, Colquhoun S**, et al. Global, regional, and national burden of rheumatic heart disease, 1990–2015. *New Engl J Med*. 2017; 377(8): 713–22. DOI: <https://doi.org/10.1056/NEJMoa1603693>
30. **Zühlke L, Karthikeyan G, Engel ME**, et al. 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*. 2016; 134(19): 1456–66. DOI: <https://doi.org/10.1161/CIRCULATIONAHA.116.024769>
31. **Ordunez P, Martinez R, Soliz P, Giraldo G, Mujica OJ, Nordet P**. Rheumatic heart disease burden, trends, and inequalities in the Americas, 1990–2017: A population-based study. *Lancet Glob Health*. 2019; 7(10): e1388–e97. DOI: [https://doi.org/10.1016/S2214-109X\(19\)30360-2](https://doi.org/10.1016/S2214-109X(19)30360-2)
32. **Diao M, Kane A, Ndiaye M**, et al. Pregnancy in women with heart disease in sub-Saharan Africa. *Arch Cardiovasc Dis*. 2011; 104(6–7): 370–4. DOI: <https://doi.org/10.1016/j.acvd.2011.04.001>
33. **Sliwa K**. Dealing with late maternal death due to cardiovascular disease. *SA Heart*. 2015; 12(3).
34. **Sliwa K, Johnson M, Zilla P, Roos-Hesselink J**. Management of valvular disease in pregnancy: A global perspective. *European Heart Journal*. 2015; 36(18): 1078–89. DOI: <https://doi.org/10.1093/eurheartj/ehv050>
35. **Soma-Pillay P, Seabe J, Sliwa K**. The importance of cardiovascular pathology contributing to maternal death: Confidential Enquiry into Maternal Deaths in South Africa, 2011–2013. *S Afr Med J*. 2016; 27(2): 60–5. DOI: <https://doi.org/10.5830/CVJA-2016-008>
36. **Mocumbi A, Sliwa K, Soma-Pillay P**. Medical disease as a cause of maternal mortality: The pre-imminence of cardiovascular pathology. *Cardiovasc J Afr*. 2016; 27(2): 84–8. DOI: <https://doi.org/10.5830/CVJA-2016-018>
37. **van Hagen I, Thorne S, Taha N**, et al. Pregnancy outcomes in women with rheumatic mitral valve disease. *Circulation*. 2018; 137(8). DOI: <https://doi.org/10.1161/CIRCULATIONAHA.117.032561>
38. **Watkins D, Sebitleane M, Engel M, Mayosi B**. The burden of antenatal heart disease in South Africa: A systematic review. *BMC Cardiovasc Disor*. 2012; 12(1). DOI: <https://doi.org/10.1186/1471-2261-12-23>
39. **Lumsden R, Barasa F, Park LP**, et al. High Burden of Cardiac Disease in Pregnancy at a National Referral Hospital in Western Kenya. *Global Heart*. 2020; 15(1): 10. DOI: <https://doi.org/10.5334/gh.404>
40. **Gordon J, Kirlew M, Schreiber Y**, et al. Acute rheumatic fever in First Nations communities in north-western Ontario: Social determinants of health 'bite the heart.' *Canadian Family Physician Medecin De Famille Canadien*. 2015; 61(10): 881–6.
41. **Bergsma L, Delgado S, Kizer E**. Navajo County Community Health Status Assessment. Navajo Public Health Services District and the University of Arizona Center for Rural Health; 2012.
42. **Golnick C, Asay E, Provost E**, et al. Innovative primary care delivery in rural Alaska: A review of patient encounters seen by community health aides. *International Journal of Circumpolar Health*. 2012; 71(1): 18543. DOI: <https://doi.org/10.3402/ijch.v71i1.18543>
43. **AIHW**. *Impact and causes of illness and death in Aboriginal and Torres Strait Islander people 2011*. Canberra, Australia: Australian Institute of Health and Welfare; 2016 September 2016. Contract No.: BOD 7.
44. **Wilson N**. Rheumatic heart disease in Indigenous populations—New Zealand experience. *Heart, Lung and Circulation*. 2010; 19: 282–8. DOI: <https://doi.org/10.1016/j.hlc.2010.02.021>

45. **Knight M.** Acquired heart disease in pregnancy – need for a greater focus. *BJOG-INT J OBSTET GY.* 2020; 127(1): 57. DOI: <https://doi.org/10.1111/1471-0528.15973>
46. **Slocumb JC, Kunitz SJ.** Factors affecting maternal mortality and morbidity among American Indians. *Public Health Reports.* 1977; 92(4): 349.
47. **Bocking N, Matsumoto C-L, Loewen K,** et al. High Incidence of invasive Group A streptococcal infections in remote Indigenous communities in Northwestern Ontario, Canada. *Open Forum Infectious Diseases.* 2016; 4(1): ofw243-ofw. DOI: <https://doi.org/10.1093/ofid/ofw243>
48. **Sullivan E, Vaughan G, Li Z,** et al. The high prevalence and impact of rheumatic heart disease in pregnancy in First Nations populations in a high-income setting: A prospective cohort study. *BJOG-INT J OBSTET GY.* 2019; 127(1): 47–56. DOI: <https://doi.org/10.1111/1471-0528.15938>
49. **Knight M, Nair M, Tuffnell D,** et al. Saving Lives, Improving Mothers' Care – Surveillance of maternal deaths in the UK 2012–14 and lessons learned to inform maternity care from the UK and Ireland Confidential Enquiries into Maternal Deaths and Morbidity 2009–14. NPEU, UOxford, National Perinatal Epidemiology Unit U; 2016.
50. **RHDAustralia (ARF/RHD writing group).** The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3rd edition). 2020.
51. **Bhaumik S.** Doctors call for countries to step up the fight against rheumatic heart disease. *BMJ.* 2013; 346. DOI: <https://doi.org/10.1136/bmj.f3504>
52. **Watkins D, Zühlke L, Engel M,** et al. Seven key actions to eradicate rheumatic heart disease in Africa: The Addis Ababa communique. *Cardiovascular journal of Africa.* 2016; 27(3): 184–7. DOI: <https://doi.org/10.5830/CVJA-2015-090>
53. **Maurice J.** Rheumatic heart disease back in the limelight. *The Lancet.* 2013; 382(9898): 1085–6. DOI: [https://doi.org/10.1016/S0140-6736\(13\)61972-8](https://doi.org/10.1016/S0140-6736(13)61972-8)
54. **AMA.** *AMA Report Card on Indigenous Health: A call to action to prevent new cases of rheumatic heart disease in Indigenous Australia by 2031.* Canberra: Australian Medical Association (AMA); 2016.
55. **Mayosi B, Robertson K, Volmink J,** et al. The Drakensberg declaration on the control of rheumatic fever and rheumatic heart disease in Africa. *S Afr Med J.* 2006; 96(3 Pt 2): 246.
56. **Zilla P, Bolman RM, Yacoub MH,** et al. The Cape Town Declaration on Access to Cardiac Surgery in the Developing World. *Cardiovasc J Afr.* 2018; 29(4): 256–9. DOI: <https://doi.org/10.5830/CVJA-2018-046>
57. **Muhamed B, Mutithu D, Aremu O, Zühlke L, Sliwa K.** Rheumatic fever and rheumatic heart disease: Facts and research progress in Africa. *Int J Cardiol.* 2019; 295: 48–55. DOI: <https://doi.org/10.1016/j.ijcard.2019.07.079>
58. **Okello E, Beaton A.** Targeted investment needed to end rheumatic heart disease in Africa. *The Lancet Global Health.* 2021; 9(7): e887–e8. DOI: [https://doi.org/10.1016/S2214-109X\(21\)00215-1](https://doi.org/10.1016/S2214-109X(21)00215-1)
59. **Abrams J, Watkins DA, Abdullahi LH, Zühlke LJ, Engel ME.** Integrating the Prevention and Control of Rheumatic Heart Disease into Country Health Systems: A Systematic Review and Meta-Analysis. *Global heart.* 2020; 15(1): 62. DOI: <https://doi.org/10.5334/gh.874>
60. **Zühlke L, Watkins D, Perkins S,** et al. A Comprehensive Needs Assessment Tool for Planning RHD Control Programs in Limited Resource Settings. *Global Heart;* 2017. DOI: <https://doi.org/10.1016/j.ghheart.2016.10.028>
61. **Wyber R, Johnson T, Perkins S,** et al. *Tools for Implementing RHD Control Programmes (TIPS) Handbook,* 2nd edition. Geneva, Switzerland: RHD Action; 2018.
62. **Coates MM, Sliwa K, Watkins DA,** et al. An investment case for the prevention and management of rheumatic heart disease in the African Union 2021–30: A modelling study. *The Lancet Global Health.* 2021; 9(7): e957–e66. DOI: [https://doi.org/10.1016/S2214-109X\(21\)00199-6](https://doi.org/10.1016/S2214-109X(21)00199-6)
63. **Remenyi B, Carapetis J, Wyber R, Taubert K, Mayosi BM.** Position statement of the World Heart Federation on the prevention and control of rheumatic heart disease. *Nat Rev Cardiol.* 2013; 10(5): 284–92. DOI: <https://doi.org/10.1038/nrcardio.2013.34>
64. **Chang A, Nabbaale J, Nalubwama H,** et al. Motivations of women in Uganda living with rheumatic heart disease: A mixed methods study of experiences in stigma, childbearing, anti-coagulation, and contraception. *PLOSOne.* 2018; 13(3). DOI: <https://doi.org/10.1371/journal.pone.0194030>
65. **Read C, Mitchell Alison G, de Dassel Jessica L,** et al. Qualitative evaluation of a complex intervention to improve rheumatic heart disease secondary prophylaxis. *JAMA-J AM MED ASSOC.* 2018; 7(14): e009376. DOI: <https://doi.org/10.1161/JAHA.118.009376>

66. **Saxena A.** Task shifting rheumatic heart disease screening to non-experts. *The Lancet Global Health.* 2016; 4(6): e349–e50. DOI: [https://doi.org/10.1016/S2214-109X\(16\)30077-8](https://doi.org/10.1016/S2214-109X(16)30077-8)
67. **RHDAustralia.** RHDAustralia Champions4change program 2019. Retrieved from: <https://www.rhdaustralia.org.au/news/champions4change-sharing-caring-and-inspiring>.
68. **Zühlke L, Perkins S, Cembri S.** Rheumatic heart disease patient event: Cape Town hosts 4th Annual Listen to My Heart Rheumatic Heart Disease for patients at the South African Heart Association meeting in 2017. *European Heart Journal.* 2018; 39(19): 1669–71. DOI: <https://doi.org/10.1093/eurheartj/ehy199>
69. **Wyber RNK, Halkon C, Enkel S,** et al. The RHD Endgame Strategy: The blueprint to eliminate rheumatic heart disease in Australia by 2031. Perth: The END RHD Centre of Research Excellence, Telethon Kids Institute; 2020. DOI: <https://doi.org/10.1093/eurpub/ckaa165.059>
70. **Longenecker C, Kalra A, Okello E,** et al. A Human-Centered Approach to CV Care: Infrastructure Development in Uganda. *Global Heart;* 2018. DOI: <https://doi.org/10.1016/j.ghheart.2018.02.002>
71. **World Health Organization (WHO).** Rheumatic fever and rheumatic heart disease: Director-General report. *Seventy-first WHA A71/25.* Geneva: WHO Executive Board; 2018.
72. **Mocumbi AO, Jamal KK, Mbakwem A, Shung-King M, Sliwa K.** The Pan-African Society of Cardiology position paper on reproductive healthcare for women with rheumatic heart disease. *Cardiovasc J Afr.* 2018; 29(6): 394–403. DOI: <https://doi.org/10.5830/CVJA-2018-044>
73. **Belton S, Kruske S, Jackson Pulver L,** et al. Rheumatic heart disease in pregnancy: How can health services adapt to the needs of Indigenous women? A qualitative study. *Aust NZ J Obstet Gynaecol.* 2018; 58(4): 425–31. DOI: <https://doi.org/10.1111/ajo.12744>
74. **Vaughan G, Dawson A, Peek M, Carapetis J, Sullivan E.** Standardizing clinical care measures of rheumatic heart disease in pregnancy: A qualitative synthesis. *BIRTH-ISS PERINAT C.* 2019; 46(4): 560–73. DOI: <https://doi.org/10.1111/birt.12435>
75. **Watkins D, Zühlke L, Engel M, Mayosi B.** Rheumatic fever: Neglected again. *Science.* 2009; 324(5923): 37. DOI: <https://doi.org/10.1126/science.324.5923.37b>
76. **World Heart Federation (WHF).** Rheumatic heart disease: neglected disease of poverty Geneva, Switzerland: World Heart Federation; 2010. Retrieved from <http://www.world-heart-federation.org/what-we-do/rheumatic-heart-disease/>.
77. **Wyber RCJ, Katzenellenbogen J.** The Cost of Inaction on Rheumatic Heart Disease: The predicted human and financial costs of rheumatic heart disease for Aboriginal and Torres Strait Islander people 2016–2031. Perth: The END RHD CRE, Telethon Kids Institute; 2018.
78. **Watkins D, Daskalakis A.** The economic impact of rheumatic heart disease in developing countries. *The Lancet Global Health.* 2015; 3, Supplement 1: S37. DOI: [https://doi.org/10.1016/S2214-109X\(15\)70156-7](https://doi.org/10.1016/S2214-109X(15)70156-7)
79. **Coffey S, Roberts-Thomson R, Brown A,** et al. Global epidemiology of valvular heart disease. *Nature Reviews Cardiology.* 2021; 1–12. DOI: <https://doi.org/10.1038/s41569-021-00570-z>
80. **Schoon M.** Severe morbidity and mortality associated with cardiac disease during pregnancy in the Free State Public Health Service. PhD, University of the Orange Free State; 2001.
81. **Sliwa K, Libhaber E, Elliott C,** et al. Spectrum of cardiac disease in maternity in a low-resource cohort in South Africa. *Heart.* 2014; 100(24). DOI: <https://doi.org/10.1136/heartjnl-2014-306199>
82. **Parsonage WA, Zentner D, Lust K, Kane SC, Sullivan EA.** Heart Disease and Pregnancy: The Need for a Twenty-First Century Approach to Care.... *Heart, Lung and Circulation.* 2021; 30(1): 45–51. DOI: <https://doi.org/10.1016/j.hlc.2020.06.021>
83. **Dawson AJ, Krastev Y, Parsonage WA, Peek M, Lust K, Sullivan EA.** Experiences of women with cardiac disease in pregnancy: A systematic review and metanalysis. *BMJ Open.* 2018; 8(9). DOI: <https://doi.org/10.1136/bmjopen-2018-022755>
84. **Marmot M.** Inclusion health: addressing the causes of the causes. *The Lancet.* 2017; 391(10117): 186–8. DOI: [https://doi.org/10.1016/S0140-6736\(17\)32848-9](https://doi.org/10.1016/S0140-6736(17)32848-9)
85. **Sliwa K, Azibani F, Baard J,** et al. Reducing late maternal death due to cardiovascular disease – A pragmatic pilot study. *Int J Cardiol;* 2018. DOI: <https://doi.org/10.1016/j.ijcard.2018.07.140>
86. **Regitz-Zagrosek V, Roos-Hesselink J, Bauersachs J,** et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy. *Eur Heart J.* 2018; 39(34). DOI: <https://doi.org/10.1093/eurheartj/ehy478>

How to cite this article: Vaughan G, Dawson A, Peek M, Sliwa K, Carapetis J, Wade V, Sullivan E. Rheumatic Heart Disease in Pregnancy: New Strategies for an Old Disease? *Global Heart*. 2021; 16(1): 84. DOI: <https://doi.org/10.5334/gh.1079>

Submitted: 30 September 2021

Accepted: 30 September 2021

Published: 20 December 2021

Copyright: © 2021 The Author(s). This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC-BY 4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited. See <http://creativecommons.org/licenses/by/4.0/>.



Global Heart is a peer-reviewed open access journal published by Ubiquity Press.

OPEN ACCESS The Open Access logo, featuring the words 'OPEN ACCESS' followed by a circular icon containing a stylized padlock with an open keyhole.