

A Conceptual Framework for Comprehensive Rheumatic Heart Disease Control Programs

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ABSTRACT

The World Health Organization, World Heart Federation, and other organizations recommend comprehensive control programs for rheumatic fever (RF) and rheumatic heart disease (RHD). However, advice on components of control programs are simple lists, with little guidance on program structure or priorities. In particular, there are limited recommendations on “stepwise” implementation and few guidelines on which program components should take temporal priority. An evidence-based framework for describing, prioritizing, and implementing comprehensive RF/RHD control programs is needed. A literature review of existing RF/RHD control program recommendations generated a list of program components. Descriptions and analysis of RF/RHD control programs informed temporal prioritizing of component parts. Relevant programmatic research from other vertical disease control programs was reviewed for generalizable implementation experiences. Twenty-five individual components of comprehensive RF/RHD control programs were identified. These fell into “baseline” program requirements (including burden of disease data, treatment guidelines, and human resources) and requirements for providing primary, secondary, and tertiary interventions. Primordial prevention and research priorities were overarching themes. These components were developed into a conceptual framework schema. Existing literature contains valuable lessons on the design and implementation of comprehensive RF/RHD control programs. Fashioning these guidelines and programmatic experiences into a conceptual framework schema benefits clinicians, policy makers, and RHD advocates.

By the most conservative estimates, more than 15.6 million people globally suffer from rheumatic heart disease (RHD) and 233,000 die of the disease each year around the world [1]. RHD is precipitated by group A streptococcal infection, progressing to the post-infectious syndrome of rheumatic fever (RF) in some people. Approximately 60% of these cases of RF induce the persistent valvular damage of RHD [2]. RF/RHD are endemic in most low- and middle-income countries and in some socioeconomically deprived groups in high-resource settings [1]. RF/RHD disproportionately affect young people and constitute a significant economic burden for low-income families, contribute to maternal mortality, and further strain already overstretched health systems [3–5].

RF/RHD have been successfully controlled in a number of settings through the implementation of register-based control programs [6–9]. This register-based approach to RF/RHD control has been recommended by the World Health Organization (WHO) and World Heart Federation (WHF) for many years [10–12]. Most control programs are “comprehensive” in that they attempt to intervene at multiple points along the protracted etiological pathway of RF/RHD [13,14]. (Although poorly defined, the concept of “comprehensive RF/RHD control programs” functionally reflects a spectrum of approaches with variable weighting of primary and secondary approaches. [14–16].)

Guidelines for the activities and components of comprehensive RF/RHD control programs have historically been simple lists [11,12]. These lists have tended to have few details on program structure or priorities. A stepwise approach to implementation is recommended, though little guidance about initial steps or program structure has been published to date [12,17]. This has spurred concerns that complex interventions—including surgery and active case finding with echocardiography—could occur in isolation, without ensuring program capacity to deliver other essential services, or at the expense of cost effectiveness [14,18–21]. Need for a “preliminary protocol with basic components” for stepwise implementation has already been identified [9].

A new wave of RF/RHD control programs is likely in the wake of pending global burden of disease data, renewed international advocacy, and the expansion of early diagnosis through echocardiographic screening [16,22,23]. Yet, among the greatest challenges in RF/RHD disease control is putting proven approaches into practice [16,23]. An evidence-based framework for describing, designing, implementing, and evaluating comprehensive RF/RHD control programs would maximize the benefit of new and reinvigorated efforts in disease control. A conceptual framework may also provide a common foundation for international collaboration and implementation research.

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This paper collates published recommendations about RF/RHD control programs. Core components are identified and arranged in an attempt to reflect relative complexity and priority. The conceptual framework is intended as a starting point for programmatic discussions by clinicians, managers, policy makers, and government and funding agencies. This first iteration will be open to revision as new evidence and programmatic experience emerges.

METHODS

Benchmark World Health Organization and World Heart Federation guidelines on RF/RHD control programs were reviewed to inform search strategy. Current recommendations have largely appeared in institutional documents, limiting utility of a systematic peer-reviewed approach. Countries with existing RF/RHD guidelines—drawn from World Heart Federation's RHDNet—were reviewed to identify recommendations applicable to other settings. Bibliographic review and a snowball approach were used to identify other program reviews and recommendations.

Listed recommendations for RF/RHD control programs were coded to identify common themes. Program reports that did not make specific recommendations for other initiatives

were excluded from formal thematic analysis. Content from these papers was used qualitatively to inform prioritization and to identify potentially under-represented areas. A number of contemporary areas were specifically reviewed: tertiary interventions; active case finding via echocardiography; and primordial prevention.

Themes identified from the literature review were condensed into a list of components of comprehensive RF/RHD control programs. The list was arranged into domains representing the classic primordial, primary, secondary, and tertiary approach to RF/RHD control (Fig. 1). Within each domain, components were arranged in order of potential priority or temporal sequence based on narrative descriptions of program implementation.

RESULTS

Four institutions have published recommendations for comprehensive control programs [10,12,24–26]. Five more papers identified specific program recommendations applicable to other initiatives [17,27–30]. One book chapter was identified and excluded from formal analysis, because the depth and specificity of the recommendations exceeded the requirements for this review [31]. Thus,

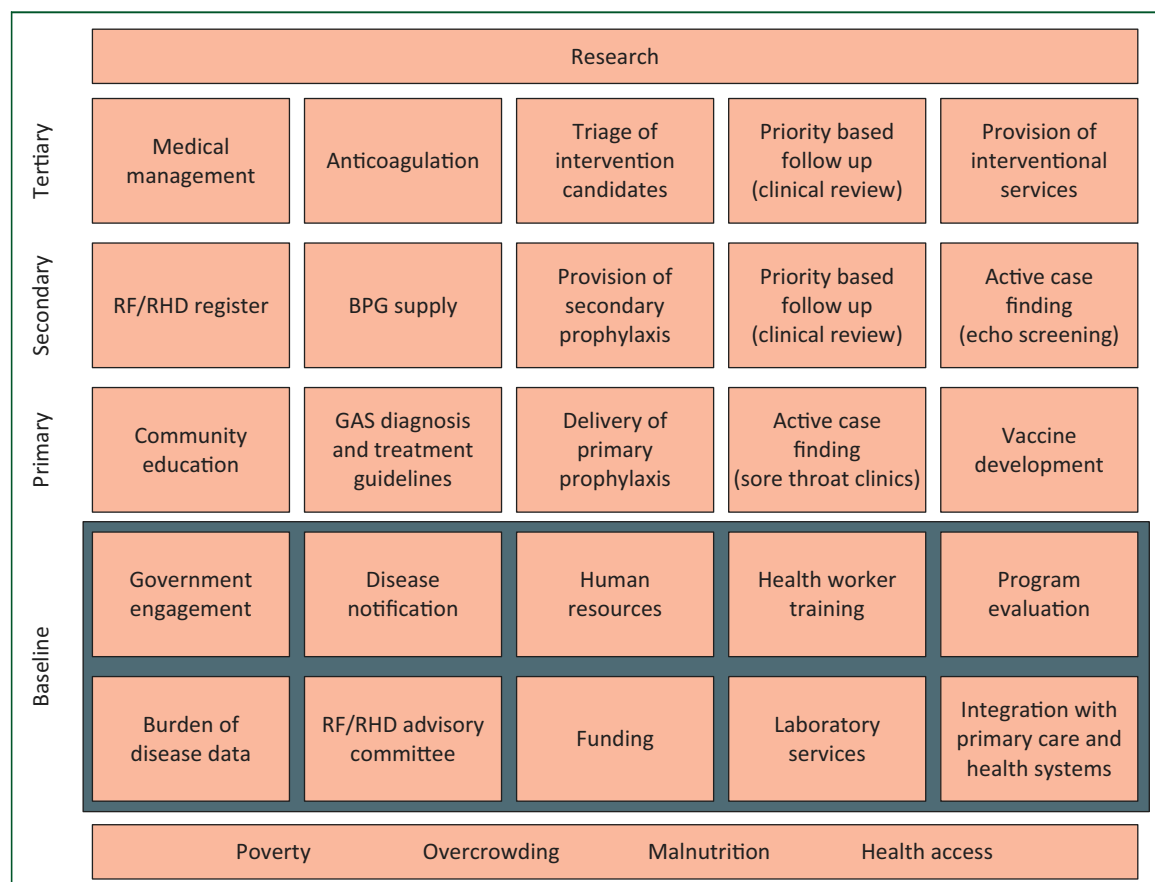


FIGURE 1. A conceptual framework for comprehensive, register-based, rheumatic heart disease control programs. BPG, benzathine penicillin G; GAS, group A streptococcal; RF, rheumatic fever; RHD, rheumatic heart disease.

9 core sources were used to generate a list of comprehensive program components. A much larger number of papers described or reviewed existing programs, but they did not make explicit recommendations directly relevant to other programs [6,9,21,32–45]. These papers were used to generate original recommendations on program components and to inform priority setting.

Twenty-five individual components of comprehensive RF/RHD control programs were identified (Fig. 1). Domains included baseline components and requirements for providing primary, secondary, and tertiary interventions. Primordial prevention and research were identified as overarching themes.

DISCUSSION

Research, advocacy, and funding are generating renewed momentum for the global control of RF/RHD [16,23]. These developments may encourage countries to develop new RF/RHD control programs, revive previous efforts, or evaluate existing programs. A systemic approach to disease control programs would maximize clinical benefit for individuals, reduce inefficiency, and provide opportunities for global knowledge sharing.

The conceptual framework is structured to provide a visual overview of priorities, moving from left to right across the page (Fig. 1). For example, a new RF/RHD program should address burden of disease data, government engagement, community education, register development, and medical management of established cases as initial goals. Programs will necessarily be adaptable to local settings and may elect to have slightly different areas of long-term focus. However, a singular focus on advanced components (e.g., active case finding of sore throats, echocardiographic screening, or interventional services) without attention to interim steps is to be discouraged.

Inclusions in the framework

Many components of RF/RHD control programs are the subject of widespread agreement, including community education, development of a register, and training health workers. However, some areas have been less well explored, necessitating a brief rationale for inclusion:

Primary prevention and active case finding for group A streptococcal pharyngitis Treatment of symptomatic group A streptococcal pharyngitis infections presenting to clinicians is broadly accepted and encouraged [41,46,47]. There is less clarity around active case findings via sore throat clinics in schools, which are widely adopted in some settings and eschewed in others [15,42,48]. Detailed exploration of this issue is outside the scope of this paper. Sore throat clinics are included in the framework as an advanced component of potential relevance in some settings.

Active case finding with echocardiographic screening

The clinical benefits of active case findings via echocardiographic screening is yet to be fully explored [21]. However, the theoretical benefit is dependent upon a functional register-based program, complete with access to benzathine penicillin G and case follow-up [21]. The conceptual framework helps to illustrate this dependency to funders and policy makers, ensuring that screening programs are coupled with prerequisite components of service delivery.

Tertiary components The framework includes a more detailed approach to tertiary intervention—medical management, anticoagulation, surgical or endovascular approaches—than existing program recommendations. The historic disconnect between comprehensive programs and tertiary interventions may reflect lack of access to open and endovascular procedures. However, access to interventional surgical services is increasing in many endemic regions and should align closely with existing RHD programs [49]. Strong links between register-based programs and tertiary services are likely to potentiate triage of candidates and help support post-intervention follow-up. Interventional options should not be delivered in isolation by local providers, charitable groups, or overseas governments. Isolated efforts risk diverting funds from more cost-effective preventative approaches or without ensuring adequate mechanisms for follow-up [21,49].

Vaccine development Few RF/RHD control programs in highly endemic settings will have the capacity for vaccine development activities. However, individual programs can support vaccine development through advocacy to governments and identifying endemic populations appropriate for clinical trials. This programmatic support is critical for eventual vaccination development and delivery. Driving the momentum for an RF/RHD vaccine is a shared responsibility of control programs internationally.

Integration with primary care and health systems

Many existing program recommendations include references to “integration,” primary care, or “existing health care delivery systems” [10,27,29]. However, achieving meaningful integration remains a challenge in most settings [16]. Although “integration” is not a discrete block of activities, it should be given explicit and conscious attention during program planning [50]. Awaiting opportunities for integration to emerge and pervade RF/RHD programs has been historically insufficient. Allocating time, funding, and resources to create space for integration activities is critical.

Primordial prevention is an overarching, but often underarticulated, theme of RF/RHD efforts. Population-level prevention activities have fallen largely outside the scope of traditional comprehensive control programs. An array of complex socioeconomic factors appear to influence manifestation of RF/RHD, including overcrowding, nutrition, and health access [14,42]. Many others, particularly

socioeconomic status and inequality, may also be contributory factors. Although these issues may be difficult for control programs to address directly, program implementers bring a unique perspective to the need for primordial intervention [51]. Policy advocacy is an important role for RF/RHD control programs, particularly given the relative contribution of socioeconomic conditions to disease burden [13]. Broadly strengthening the advocacy capacity of the RF/RHD community is critical for addressing persistent neglect of the disease among decision makers [52].

Research is a second overarching theme for RF/RHD programs. A broad array of disease control questions remain unanswered, including natural history of subclinical carditis, the role of echo screening, program optimization, and the development of novel therapies [16]. Many of these questions can only be answered in highly endemic communities. Coupling research activities with a comprehensive service delivery program is more appropriate and ethically defensible than stand-alone research projects are [53]. Engagement in the research community also fosters collaboration for clinicians and academics in remote and low-resource settings. Maintaining a rigorous evidence base for disease control efforts is a shared responsibility for all register-based programs.

Refining the conceptual framework

Identifying best practices for RF/RHD control programs require comparison and analysis of implementation experiences. A large volume of this information already exists in peer-reviewed literature. However, only some investigators make explicit recommendations for improving existing programs and very few make suggestions applicable to new programs. These formal publications represent only a fraction of program reports provided to governments and external donors. Collating and analyzing this experiential evidence base will be critical for advancing implementation science [16,23].

The proposed conceptual framework provides a starting point for knowledge sharing with a common set of terminology. Calls for standardization of terms have been longstanding [54]; some progress has been achieved in simple disease categories and echocardiographic terms but not in program implementation to date [11,55]. Capitalizing on shared terms and structure, collaborative efforts to build a more robust framework should be encouraged. New components may be added; for example, a protocol for managing anaphylaxis may be prioritized before delivery of secondary prophylaxis is commenced. Pooling international service delivery experience will continue to inform ongoing development of the framework. More substantial program implementation recommendations should be possible. Although “anticoagulation” is identified as a conceptual framework component, there is no detail on possible approaches, relative costs, compliance, or clinical protocols. Exploring the substance of components

within the framework requires significant development and international collaboration.

A number of areas were consciously excluded from this initial version of the framework. Prescriptive lists of staff and equipment have been avoided at this stage (e.g., echo machines, sonographers, information pamphlets, disease notification forms), as these will depend on local conditions. Development of a practical “program implementation handbook” may be a natural extension of the framework in the future.

Application of the conceptual framework

The conceptual framework is applicable to a number of stakeholders in global RF/RHD control. For most, the framework adds value through visual comprehensibility. RF/RHD is a complex disease with unwieldy acronyms, a protracted course, and multiple opportunities for intervention. Few of the constituent groups needed for disease control will be technical experts. Providing a concise snapshot of control program activities enhances communication to all. Some stakeholders may identify specific benefits:

Governments and policy makers The framework may simplify options for decision makers by offering guidance on stepwise program intervention. In resource-limited settings, staged rollout may be more politically and economically feasible than de novo development of a complete program. Governments should be encouraged to take any action toward RF/RHD control, ideally first-tier interventions, with a plan to provide increasingly advanced services.

Donors and funders One of the challenges in securing much-needed resources for RF/RHD is the breadth and complexity of comprehensive programs. Without specialist knowledge of the disease, donors may struggle to understand how their contribution adds to program delivery and prevention efforts. The conceptual framework provides a common ground for funders and implementers to take stock of a proposed program and agree on areas to be strengthened. This approach may help to avoid sequestration of funding for tertiary services, without addressing basic needs such as benzathine penicillin G supply.

Program reporting and evaluation template With the expansion of comprehensive RF/RHD programs comes increasing reporting and evaluation requirements. Without a structured framework, critical program issues may be overlooked. Encouraging programs to report on activities in each of the program components would ensure that core components were addressed and improve comparability between programs. With improvements in the model, it may be possible to develop an objective evaluation template specifying gold-standard practice in each domain and providing customized feedback to programs.

Program handbook The framework provides a broad overview of activities and priorities in comprehensive control programs. It provides no detail about how these can best be achieved. Collating detailed implementation experience—components of register-based programs, technical specifications for echo screening programs—are best addressed in an implementation handbook that could be structured around the headings of the conceptual framework.

SUMMARY

Decades of global experience in the design and implementation of comprehensive RF/RHD control programs already exist. The conceptual framework distills some of this experience, and existing recommendations, into a concise visual overview. Design, development, implementation, and evaluation of programs benefit from a strategic approach. This standardized approach facilitates international collaboration, fostering efforts to achieve excellence in program delivery. Evolution and development of the model is inevitable and encouraged; this first iteration is designed to provide a foundation for policy makers, governments, donors, researchers, and program implementers. Ultimately, a focus on comprehensive control programs benefits the millions of people living with RHD.

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