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Evidence-based guidelines

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2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease

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Abstract

Rheumatic heart disease (RHD) is an important and preventable cause of morbidity and mortality among children and young adults in low-income and middle-income countries, as well as among certain at-risk populations living in high-income countries. The 2012 World Heart Federation echocardiographic criteria provided a standardized approach for the identification of RHD and facilitated an improvement in early case detection. The 2012 criteria were used to define disease burden in numerous epidemiological studies, but researchers and clinicians have since highlighted limitations that have prompted a revision. In this updated version of the guidelines, we incorporate evidence from a scoping review, an expert panel and end-user feedback and present an approach for active case finding for RHD, including the use of screening and confirmatory criteria. These guidelines also introduce a new stage-based classification for RHD to identify the risk of disease progression. They describe the latest evidence and recommendations on population-based echocardiographic active case finding and risk stratification. Secondary antibiotic prophylaxis, echocardiography equipment and task sharing for RHD active case finding are also discussed. These World Heart Federation 2023 guidelines provide a concise and updated resource for clinical and research applications in RHD-endemic regions.

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Introduction

Rheumatic heart disease (RHD) remains the leading acquired cardiovascular disease among children and young adults in low-income and middle-income countries and in some at-risk populations living in high-income countries[1,2]. RHD results from cumulative valvular damage that is largely preventable by secondary antibiotic prophylaxis (SAP)[3,4]. Given that clinically detectable RHD might present after years of subclinical disease, the detection of early RHD by echocardiography and the subsequent initiation of SAP, alongside other established strategies, may reduce the global burden of RHD[5]. Echocardiography is vastly superior to auscultation for detecting RHD[6]. In 2012, the World Heart Federation (WHF) published the first evidence-based guidelines for the echocardiographic diagnosis of RHD[7] (referred to in this article as the WHF 2012 guidelines). These guidelines included the minimum criteria required to diagnose RHD, harmonized the diagnosis and classification approach for RHD across global regions and were especially useful for epidemiological studies. Although the WHF 2012 guidelines were research-oriented, they have since become a frequently used tool in clinical settings for diagnosing RHD. However, research and real-world experience with the WHF 2012 guidelines have led to the identification of some limitations in the criteria. This includes the heterogeneous outcomes of echocardiography-detected RHD and the practical challenges in applying the criteria in the busy settings in which RHD active case finding occurs[8,9].

Over the past decade, the RHD research community has made important strides in understanding the natural history of RHD[8] and risk factors for its progression[10]. Concurrently, the management strategies for the early stages of RHD were assessed, including the efficacy of SAP[4]. Furthermore, there has been an increasing appreciation of hand-held echocardiography and task sharing for active case finding in resource-limited settings[11–13]. The evidence and experience accrued over the past decade have thus prompted this revision. The WHF 2023 guidelines incorporate evidence gathered from a scoping review, an expert panel and end-user feedback obtained through a global survey on the utility of the WHF 2012 guidelines[7]. In these updated guidelines, we provide the revised minimum echocardiographic criteria for the definitive diagnosis of RHD; a new classification of RHD that is dependent on the risk of disease progression; a two-step echocardiography algorithm for RHD active case finding, including screening criteria for detecting suspected cases and confirmatory criteria for the definitive diagnosis of RHD; management recommendations for early-stage RHD; and guidance for task sharing and the use of hand-held echocardiography in RHD active case finding.

These WHF 2023 guidelines, endorsed by globally representative societies (Box 1), are a pragmatic, contemporary and relevant reference tool that can be utilized for clinical and research applications in RHD-endemic regions across the world. They provide an evidence-based guide for the standardization of disease definitions, diagnostic criteria, risk assessment strategies and management approaches for patients with varying severities of RHD.

Box 1

Endorsements

Organizations that endorse these World Heart Federation 2023 guidelines:

- American Heart Association
- Australian Heart Foundation
- Interamerican Society of Cardiology
- Pan-African Society of Cardiology
- South African Heart Association
- Writing Group of the 2020 Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease

Revising the guidelines

Changes to the WHF 2012 guidelines

In the revised WHF 2023 guidelines, we have introduced a new set of echocardiographic criteria — the 'screening criteria' — to facilitate task sharing in active case-finding programmes. We have also formulated a stage-based classification system for RHD that reflects the continuous spectrum of the disease and risk of progression, as opposed to the discreet classification system used in the WHF 2012 guidelines[7]. These developments are summarized in Boxes 2 and 3.

Aim

These WHF 2023 guidelines aim to define the minimum echocardiographic criteria for diagnosing RHD according to the best-available evidence. These guidelines do not describe all rheumatic valvular lesions and the spectrum of diseases, nor do they intend to provide comprehensive management guidance on the different presentations of valvular heart disease (these topics are discussed in other clinical practice guidelines[14–16]). The focus of this document on the echocardiographic diagnosis of RHD complements, but does not overlap with, the 2023 American Society of Echocardiography report that focuses on providing recommendations for advanced RHD[14]. The current guidelines are designed to provide diagnostic guidance for health-care practitioners and researchers conducting active case-finding programmes, for opportunistic case detection in clinical settings, and for health-care practitioners who detect possible RHD cases as part of routine care.

Methods

An international panel of experts in the screening and echocardiographic imaging of RHD were involved in revising the WHF 2012 guidelines to develop this updated, evidence-based version[7]. A scoping review (not yet published) will summarize the available evidence on the performance of the WHF 2012 criteria, including the specificity (in low-prevalence populations), the inter-rater reliability and various modifications of the criteria for diagnostic simplicity. The panel of experts also conducted an end-user survey to assess the applicability and usability of the WHF 2012 guidelines. The evidence gathered from the scoping review, alongside end-user feedback from the survey, generated three main conclusions: first, the important role of task sharing and the need for a simplified set of criteria that health-care practitioners with limited training using hand-held devices could adopt; second, that the WHF 2012 criteria did not take into account the continuum of early rheumatic valvular changes that might regress, progress and overlap with normal valve morphology; and third, that certain aspects of the WHF 2012 criteria were subjective and difficult to replicate, especially in the clinical setting. Of note, the WHF 2012 criteria were primarily written for use by research teams performing epidemiological studies. However these criteria have now also been widely incorporated into clinical use in RHD-endemic and non-endemic settings. This includes the development of task-sharing and active

Box 2

New features of the World Heart Federation 2023 guidelines

- Introduction of two sets of echocardiographic criteria for RHD:
  - Screening criteria are designed principally for non-experts to apply in appropriate settings for detecting suspected cases of RHD
  - Confirmatory criteria are designed for experts to use to confirm a diagnosis of RHD
- Classification of RHD into stages A, B, C and D on the basis of the risk of progression to more advanced valvular heart disease; the terms 'borderline', 'definite' and 'latent' RHD are no longer recommended
- Weight-based measurements for valvular regurgitation jet length
- Recommendations for the management for early stages of RHD

RHD, rheumatic heart disease.

case-finding programmes for the echocardiographic detection of RHD. Consequently, an updated version of the guidelines was necessary to improve the utility across these various applications.

Grading of evidence

The classification of recommendations and levels of evidence are expressed in accordance with the 2016 ACC/AHA grading system[17].

Summary of recommendations

Figure 1 summarizes the steps from screening to confirmatory echocardiography and subsequent care plans for children and young adults diagnosed with RHD through active case-finding programmes. In addition, Table 1 summarizes the recommendations for RHD care pathways by disease stage, and Table 2 provides an overview of recommendations for at-risk populations, equipment use, task sharing for active case-finding and technology for RHD screening to improve RHD care in endemic regions.

Echocardiographic criteria and staging of RHD

Two sets of criteria are presented that can be integrated into the stage-based diagnostic strategy, including a new set of 'screening criteria' as well as 'confirmatory criteria'. The latter are largely similar to the original WHF 2012 criteria, but with minor modifications. The WHF 2023 guidelines also present a staging process for RHD diagnosis on the basis of the risk of disease progression.

The two-step screening criteria and confirmatory criteria have been developed predominantly to assist in coordinated population-based screening programmes that might or might not involve task sharing. In terms of task sharing, non-experts can use the screening criteria to conduct screening echocardiograms, but the same steps can also be performed by experts to save time during screening or in clinical settings, or when the appropriate equipment for applying the confirmatory set of criteria is not available. Confirmatory echocardiograms can be performed in individuals who have a positive screening result, or as a single step in individuals without a history of acute rheumatic fever (ARF) or RHD by experts who have ready access to the necessary echocardiography equipment.

Screening criteria for the echocardiographic detection of RHD

The screening criteria are intended to rapidly detect possible disease in settings in which RHD is highly prevalent and in which equipment and health-care providers are limited (Box 4). They are only applicable to individuals aged 20 years or less. They can also be applied in high-volume screening programmes that use task sharing and hand-held devices. The criteria have been selected on the basis of the best-known evidence to provide greater sensitivity, as a screening test, at the expense of specificity. The two-step screening strategy should be adopted whenever possible (Fig. 1a).

Following up a screening echocardiogram with a confirmatory echocardiogram might not be possible in certain high-risk populations from resource-limited regions. In some circumstances, a diagnosis on the basis of the screening criteria in combination with clinical features

Box 3

Diagnostic category terms in the World Heart Federation 2012 guidelines that are no longer recommended

Latent RHD

Valvular changes consistent with rheumatic heart disease (RHD) that are detected by echocardiography in an individual with no known history of acute rheumatic fever or symptoms of RHD. This term incorporates a vast range of diseases, from echocardiographic features meeting minimal diagnostic criteria through to clinically significant disease. This term is no longer recommended and is superseded by the stage categories in these revised guidelines.

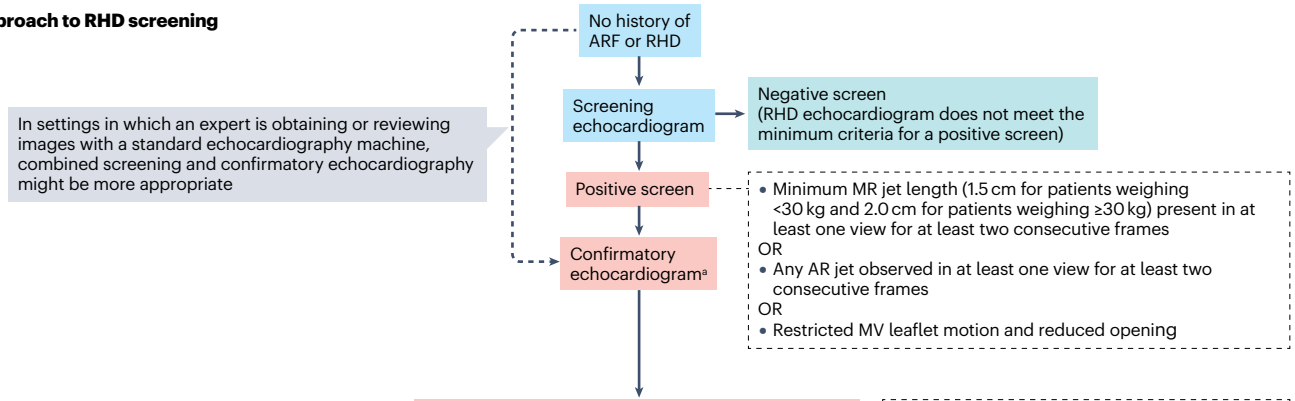
Borderline RHD

This term designates patients with abnormal valve appearance on echocardiography, with some but not all features consistent with RHD. In the 2012 World Heart Federation (WHF) guidelines, this category only applied to individuals aged ≤20 years, given that some valvular features might take time to develop. This category has been recognized to represent a spectrum from 'normal valve' to early-stage RHD. This term is no longer recommended for disease classification and is incorporated within stage A (minimal echocardiographic criteria for RHD) in these revised guidelines.

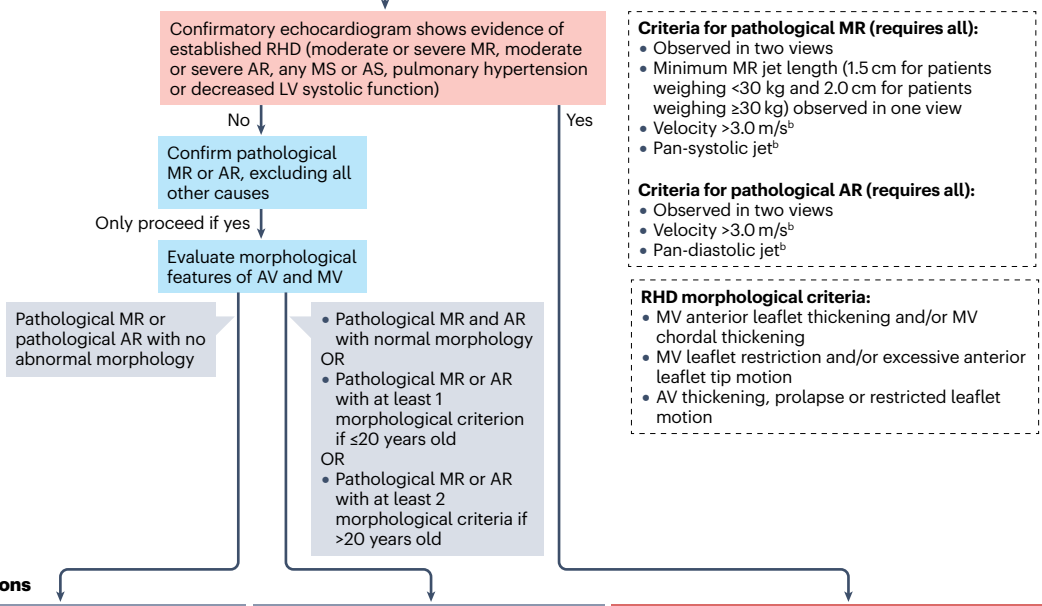
Definite RHD

The term was used to refer to the abnormal valvular appearance with regurgitation or stenosis of aortic or mitral valves on echocardiography meeting the WHF 2012 criteria for RHD. This term is no longer recommended and is incorporated within stage B (mild RHD), stage C (advanced RHD) and stage D (advanced RHD with established complications) in these revised guidelines.

a Approach to RHD screening



b Approach to RHD classification



c Staging and care recommendations

	Stage A[c]	Stage B	Stages C and D
Definition	Minimum echocardiographic criteria for RHD achieved; patient might be at risk of disease progression	Mild RHD; patient is at risk of developing clinical symptoms of valvular heart disease	Advanced RHD; patient is at high risk of developing clinical complications (stage C) or already has clinical complications (stage D) and is needing medical or surgical intervention (in addition to SAP)
Risk of progression	Low risk on the basis of risk score	Moderate or high risk of RHD progression on the basis of risk score	Patients with stage C disease are at high risk of progression to stage D and/or death
Initial care	Enrolment in an RHD registry; SAP initiation is reasonable (class 2A recommendation)[d]; re-review using echocardiogram data in 1–2 years (class 1 recommendation)	Enrolment in RHD registry; SAP is recommended (class 1 recommendation)	Enrolment in RHD registry; SAP (class 1A recommendation) per local guidelines, and specialized care per local, regional or international guidelines
Prophylaxis duration	Cessation of SAP could be considered if echocardiographic findings are normal after 1–2 years (class 2B recommendation)	Per local guidelines, or WHO guidelines if local guidelines are unavailable	Per local guidelines, or WHO guidelines if local guidelines are unavailable

Fig. 1 | The approach to active case finding, classification and care implications for rheumatic heart disease in endemic regions. a, Screening for rheumatic heart disease (RHD). [a]Although confirmatory echocardiography is recommended before the initiation of secondary antibiotic prophylaxis (SAP), in settings in which confirmatory echocardiography cannot be performed immediately, SAP can be initiated in patients while waiting for a confirmatory study. The final decision should be made by or in consultation with a clinician with expertise in RHD diagnosis. **b**, Classification of RHD. [b]These criteria can be achieved in two separate views or from two separate continuous-wave Doppler measures. **c**, Staging and care recommendations. [c]Stage A only applies to individuals aged 20 years or less. [d]Evidence from a randomized, controlled trial showed that SAP reduces the risk of RHD progression in children aged 5–17 years with stage A or stage B RHD[4]. However, SAP is also associated with adverse effects, including allergy and anaphylaxis, and numerous health-system factors should be considered before the roll-out of large-scale SAP programmes. Consequently, there is a need to consider family and clinician decisions, as well as health-system factors before recommending SAP in individuals aged ≤ 20 years with stage A RHD who are at low risk of RHD progression. However, all children with stage A disease must have access to follow-up echocardiography and longitudinal clinical evaluation to monitor for disease progression. AR, aortic regurgitation; ARF, acute rheumatic fever; AS, aortic stenosis; AV, aortic valve; LV, left ventricular; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve.

Table 1 | Recommendations for care pathways for rheumatic heart disease by disease stage

Stage	Initial care	SAP duration	Class of recommendation	Level of evidence
Positive screening criteria	Refer for confirmatory echocardiogram. In special circumstances[a], enrolling in the RHD registry and/or initiating SAP might be reasonable while awaiting a confirmatory echocardiogram	Until a confirmatory echocardiogram has been obtained	2A	C–EO
A	Enrolment in the RHD registry. SAP can be commenced. Re-review with a comprehensive echocardiogram in 1–2 years is recommended	Cessation of SAP could be considered if echocardiographic findings are normal after 1–2 years	2B	B–R
B	Enrolment in the RHD registry. SAP is recommended	According to local guidelines or WHO guidelines for the management of mild RHD, if local guidelines do not exist	1	A
C	Enrolment in the RHD registry. SAP should be administered according to local recommendations and specialized care implemented according to local, regional or international recommendations	According to local guidelines or WHO guidelines for the management of moderate-to-severe RHD, if local guidelines do not exist	1	B–NR
D	Enrolment in the RHD registry. SAP should be administered according to local recommendations and specialized care implemented according to local, regional or international recommendations	According to local guidelines or WHO guidelines for the management of moderate-to-severe RHD, if local guidelines do not exist	1	B–NR

EO, expert opinion; NR, non-randomized; R, randomized; RHD, rheumatic heart disease; SAP, secondary antibiotic prophylaxis. [a]Applicable for patients with a high suspicion of RHD on the basis of the level of risk and echocardiography findings in settings with limited access to confirmatory echocardiography, with the final decision to be made by or in consultation with a clinician with expertise in RHD diagnosis.

and a high pre-test probability of disease might be reasonable, with the final decision to be made by or in consultation with a clinician with expertise in diagnosing RHD.

Rationale and evidence. The screening criteria proposed were derived from an analysis of numerous studies that have evaluated abbreviated (modified) versions of the WHF 2012 criteria[18–22]. Although the modified criteria and the echocardiography protocols have varied between studies, the sensitivity of modified criteria compared with the WHF 2012 criteria across these studies has been satisfactory[12,18,23,24]. Most of these studies have utilized valvular regurgitation alone, excluding morphological features, to screen for RHD, which is reflected in the screening criteria presented in this guideline.

In the current guidelines, the definitions of mitral and aortic regurgitation have been simplified by excluding the need for spectral Doppler imaging. This simplification will enable the application of the screening criteria using hand-held devices that do not have spectral Doppler capacity and allows ease of application in settings with large volume, active case-finding programmes or task sharing.

The detection of mitral regurgitation has demonstrated substantial-to-almost-perfect inter-reviewer reliability for the diagnosis of RHD[8,25,26] and is the most common lesion detected when screening for RHD. Therefore, mitral regurgitation forms the basis of the abbreviated criteria. Several reports have analysed various combinations of mitral and aortic jet lengths from expert-performed and task-sharing echocardiographic screening studies to determine the optimal sensitivity and specificity[20,27,28]. Reporting minimal mitral regurgitation jet lengths as a positive screen has been shown to have a high sensitivity for detecting RHD[18,21,22,27,28]. However, the high rates of false positives that this protocol would produce in clinical practice (requiring confirmatory echocardiography) make this strategy impractical[12]. Available data support a mitral valve jet length of 1.5–2.0 cm as having the best balance between sensitivity (73–81%) and specificity (75–100%) for the detection of RHD[18,20,22]. A study that assessed the inter-rater reliability and individual reviewer performance of the 2012 WHF guidelines demonstrated the highest β-coefficient using a mitral regurgitation jet length of ≥2.0 cm[26]. The two jet lengths for different weight cut-offs presented in these guidelines were proposed by our panel of experts to ensure a high sensitivity of the criteria in the younger paediatric population. Studies that have assessed the WHF 2012 guidelines in populations with a low prevalence of RHD have demonstrated low rates (0.5–0.8%) of pathological mitral regurgitation, presumably representing the upper limit of normal physiological mitral regurgitation[29–31]. The requirement for the regurgitant jet to be observed in more than one consecutive frame can provide a surrogate for spectral Doppler imaging. A jet that is observed in more than one consecutive frame is less likely to be a closing volume than one that is observed in only one frame and is more consistent with pan-systolic regurgitation[21].

As defined by the WHF 2012 criteria, pathological aortic regurgitation has been reported in two studies to have almost perfect inter-reviewer reliability for the detection of RHD (κ = 0.86 and 0.95)[26,32]. The sensitivity and specificity of reducing aortic regurgitation from a minimum jet length of 10 mm to any detected regurgitation have been described in numerous studies and remain adequate as screening criteria[10,18,20,21]. In addition, in a 2019 study, aortic regurgitation remained one of the highly significant variables in a point-based score (β-coefficient = 4.8) used to predict unfavourable outcomes[10]. Ensuring that the regurgitation jet is observed in more than one frame again excludes closing volumes and physiological regurgitation[21].

Confirmatory criteria for the echocardiographic detection of RHD

The criteria for the detection of RHD using pathological valvular regurgitation and mitral stenosis are indicated in Box 5 and those for RHD-defining morphological features are in Box 6.

Rationale and evidence. The confirmatory criteria, which involve the use of 2D imaging, colour Doppler and continuous-wave Doppler

Table 2 | Recommendations for at-risk populations, equipment use and task sharing for rheumatic heart disease screening

Recommendations	Class of recommendation	Level of evidence
Screening for RHD in high-risk settings		
Consider RHD echocardiographic active case finding in children and young adults aged 5–20 years living in endemic regions[2,4,6,34,52]	2A	B
Consider RHD echocardiographic active case finding in first-degree relatives of index cases[53–55]	2A	B
Consider RHD echocardiographic active case finding in pregnant women[56] and young adults (aged 21–39 years) living in endemic regions	2B	B
Confirmatory criteria should be applied for active case finding in individuals aged >20 years, given that there is limited evidence to support the use of the screening criteria in this population[7]	2A	B
Equipment use		
Hand-held echocardiogram devices are a reasonable alternative option for echocardiographic RHD screening[18–22]	2A	B–NR
Task sharing for active case finding in endemic regions		
A structured training programme for teaching non-experts to perform echocardiography screening to facilitate task sharing is recommended[28,68,75]	1	B
Abbreviated diagnostic criteria and echocardiography protocols can be applied for task sharing[18,28,65,68]	2A	B
Expert supervision and review of non-expert images should be provided before a diagnosis is made[28]	2A	B/C
Use of technology to improve care		
Telemedicine can improve education of RHD and related topics for health-care providers, school staff, screening teams and the general population	1	C
Telemedicine and dedicated cloud-based computing systems are helpful for remote upload, storage and interpretation of images and tests[69]	1	C
Tele-echocardiography can aid in task sharing for active echocardiographic case finding in RHD-endemic regions[70]	2B	C

NR, non-randomized; RHD, rheumatic heart disease.

echocardiography, aim to discriminate between physiological and pathological changes in the valves with reasonable accuracy and inter-rater reliability[10]. Differentiating between individual morphological features can be challenging, which is especially true when trying to distinguish between two features of mitral apparatus thickening (anterior leaflet and chordal thickening) and the two features of abnormal mitral valve motion (restrictive leaflet motion and excessive motion of the anterior leaflet tip). In each case, the paired abnormalities probably reflect a similar pathological finding[10]. Furthermore, the abnormalities within the two categories of morphological changes are continuous and often coexist. Accordingly, these features have a lower inter-rater reliability than pathological valvular regurgitation[33,34]. Analyses in multiple populations have led to the development of a risk prediction score that performs well in distinguishing between a low risk of disease progression and a moderate or severe risk of disease progression and includes the criterion of one morphological feature in addition to pathological regurgitation of the associated valve in individuals aged ≤20 years[10]. Additional data from a randomized trial and several natural history studies have shown an increased risk of developing RHD in patients with pathological mitral or aortic regurgitation and at least one morphological feature[4,35]. Given that mild valve thickening is common in adults, two morphological features are still required to diagnose those aged >20 years.

These WHF 2023 guidelines simplify the morphological features needed for the diagnosis and have created two morphological categories for the mitral valve: the thickening of the valvular apparatus (defined by the presence of either or both valvular and chordal thickening) and valve mobility (defined by the presence of either or both restricted and excessive leaflet motion). The morphological features of the aortic valve have not been combined because individual features are more specific for RHD. However, only one morphological feature is needed together with the presence of pathological aortic regurgitation for a diagnosis of mild RHD.

Staging of RHD based on the risk of disease progression

The natural history of early RHD is heterogeneous, and studies have demonstrated that the echocardiographic features of RHD might regress, remain unchanged or progress over time[4,8]. The GOAL study[4] showed that treating early-stage RHD with SAP reduced disease progression at 2 years of follow-up. However, the study also highlighted the heterogeneous nature of early disease, with 50% of borderline and 30% of mild cases regressing spontaneously (without the need for SAP). In high-risk populations, some individuals with mild valvular changes do indeed have RHD (and are at risk of disease progression), whereas the mild changes in other individuals might be classified as being normal. Of note, individuals might oscillate between these two classifications. The stage-based criteria outlined in these guidelines acknowledge the spectrum of early echocardiographic rheumatic changes by incorporating the assessment of the risk of progression and replacing the previously described borderline and definite categories[7] (Fig. 1 and Box 3).

A prediction score reported in 2019 using the parameters in the WHF 2012 echocardiographic criteria (anterior mitral valve thickening, excessive mitral leaflet motion, regurgitant mitral jet >2.0 cm, irregular aortic valve thickening and any aortic regurgitation) showed good discrimination and calibration for a diagnosis of RHD (*c*-statistic of

0.998 and 0.994, respectively) and good discrimination for predicting disease progression (*c*-statistic of 0.811)[10]. The score has been externally validated in international cohorts and has been recalibrated for refinement. The scoring system assigned point values to different variables that were proportional to their regression coefficients and stratified the sum of these values into low (0–6), intermediate (7–9) and high (≥10) risk categories of early RHD progression[10]. By risk-stratifying individuals with RHD at diagnosis, the score has potential value for deciding which patients will benefit most from close clinical monitoring and SAP[36,37]. In keeping with the features of this score and the corresponding level of risk of disease progression, as well as to reflect early RHD as a continuum instead of using distinct diagnostic categories, we present a staging system (stages A–D) for RHD as detected by echocardiography. The criteria to meet these stages and the risk of disease progression are shown in Box 7.

Stage A. Stage A indicates the presence of early valvular changes that meet the minimum diagnostic criteria for RHD in a high-prevalence population. Stage A is only applicable to individuals aged 20 years or less. On the basis of the risk score, individuals with stage A disease might be at risk of developing valvular heart disease, but are considered to have a low risk of disease progression. This stage A category corresponds to the disease stage previously known as 'borderline RHD'[7], but better acknowledges the continuum of echocardiographic features from normal variant to mild RHD. Of note, the presence of morphological features alone, without valvular regurgitation, does not qualify individuals for stage A disease, given that the presence of morphological features alone is observed only in a small number of individuals and is considered unlikely to be associated with the development of RHD.

Stage B. Stage B indicates mild RHD with the presence of both pathological regurgitation and morphological features. The presence of both aortic and mitral pathological regurgitation, but without morphological features, is also classified as stage B, given that the concurrence of pathological mitral and aortic regurgitation is probably indicative of true pathology. Patients with stage B disease are considered to have a moderate or high risk of disease progression on the basis of the risk score[10] and correspond to the previous category named 'definite RHD' (Box 3).

Stage C. Stage C indicates established valvular disease detected by echocardiography, and patients with stage C disease are at risk of developing complications and might require medical treatment or surgical intervention. Mitral stenosis of any severity is included in this category.

Stage D. Stage D indicates established valvular disease detected by echocardiography (the same as stage C), but with overt clinical complications (such as heart failure, arrhythmia, stroke or the need for cardiac surgery).

Limitations of diagnostic criteria

Screening criteria. The screening criteria have been developed for ease and speed of application and are highly sensitive, but have a lower specificity than the confirmatory criteria. This compromise is acceptable for a screening test, given the need for a simplified set of criteria that can easily be applied in settings with limited resources and poor access to specialized care and advanced imaging modalities.

Confirmatory criteria. The limitations of the current confirmatory criteria are essentially unchanged from those described by the WHF 2012 guidelines[7]. However, the challenge of applying morphological features as a criterion has been addressed in this revision. The addition of the body weight-based measurements also addresses concerns about the under-reporting of pathological mitral regurgitation in younger children. The confirmatory criteria are detailed and are best applied by experts with advanced training on RHD diagnosis and with access to full-capacity echocardiogram machines. Although these criteria are appropriate for use in well-resourced settings, their application in certain resource-poor, RHD-endemic regions might be limited.

Furthermore, the comprehensive criteria do not include quantitative severity grading using measurements such as vena contracta, the proximal isovelocity surface area and the effective regurgitant orifice area. These measurements can be challenging to obtain in rheumatic valve lesions with eccentric regurgitant jets[14]. Any quantitative or qualitative evaluation of severity should remain in line with international guidelines on the echocardiographic assessment of valvular heart disease[15,16].

Exclusion of aortic stenosis and right-sided valve lesions from the diagnostic criteria. Neither the screening criteria nor the comprehensive criteria include the presence of aortic stenosis or right-sided cardiac

Box 4

Screening criteria in individuals aged ≤20 years

Screening criteria for the echocardiographic detection of rheumatic heart disease in individuals aged ≤20 years.

MR (requires all the following)

- In individuals weighing <30 kg: MR jet length ≥1.5 cm; in individuals weighing ≥30 kg: MR jet length ≥2.0 cm[a]
- MR jet is observed in at least one view
- MR jet is observed in at least two consecutive frames

AR (requires all the following)

- Any AR
- Observed in at least one view
- Observed in at least two consecutive frames

Mitral stenosis

- Restricted leaflet motion with reduced valve opening

Positive screen

- Presence of any of the defined MR, AR or MS

Negative screen

- Absence of any of the defined MR, AR or MS

AR, aortic regurgitation; MR, mitral regurgitation; MS, mitral stenosis. [a]Cut-off value is derived from expert consensus. If weight is not available, an age cut-off of <10 years or ≥10 years can be applied.

Box 5

Confirmatory criteria for pathological valve dysfunction

Pathological (at least mild) MR (all criteria must be met)

- Observed in at least two views
- Observed in at least one view, MR jet length measures ≥1.5 cm (in individuals weighing <30 kg) or ≥2.0 cm (in individuals weighing ≥30 kg)[a,b]
- Velocity ≥3.0 m/s for one complete envelope[c,d]
- Pan-systolic jet in at least one envelope[d,e]

Pathological (at least mild) AR (all criteria must be met)

- Observed in at least two views
- Observed in at least one view, AR jet length ≥1.0 cm[b]
- Velocity ≥3.0 m/s in early diastole[d]
- Pan-diastolic jet in at least one envelope[d]

Mitral stenosis (all criteria must be met)[f]

- Restricted leaflet motion with reduced valve opening
- Mean peak gradient ≥4.0 mmHg

Other common causes of mild valvular regurgitation, including mitral valve prolapse and bicuspid aortic valve, should be excluded before diagnosing rheumatic heart disease. AR, aortic regurgitation; MR, mitral regurgitation. [a]Cut-off value is derived from expert consensus. If weight is not available, an age cut-off of ≤10 years or >10 years can be applied. [b]A regurgitant jet length should be measured from the vena contracta to the last pixel of regurgitant colour (blue or red). [c]Body of the pan-systolic envelope should be ≥3.0 m/s. [d]Separate colour wave Doppler traces can be used in different views to document the pan-systolic or pan-diastolic envelope and jet velocity. [e]Given the difficulty in aligning spectral Doppler imaging through eccentric regurgitant jets, it is reasonable to use an appearance of a jet being pan-systolic on the basis of qualitative assessment, such as a colour jet observed throughout systole. [f]Two-dimensional evidence of the reduced mitral valve orifice area is adequate when spectral Doppler imaging is unavailable.

valve lesions. As stated in the WHF 2012 guidelines[7], rheumatic aortic valve stenosis is rare in isolation, especially in the absence of mitral valve involvement[38–40]. Similarly, isolated involvement of right-sided cardiac valves — most notably tricuspid regurgitation — in the absence of mitral valve disease that leads to right ventricular dysfunction is an equally unusual finding, especially during screening[41]. Consequently, we have maintained the decision not to include these valve lesions in the minimum diagnostic criteria outlined in these guidelines.

Age-based and weight-based cut-offs for mitral regurgitation jet length. There are limited data on the use of mitral regurgitation jet length as a diagnostic criterion for RHD because it is not routinely included in paediatric echocardiography guidelines. However, in studies comparing left atrial size with body surface area and reporting on nomograms that have compared patient age and weight with body surface area, there is a demonstrable inflexion point where body surface area = 1 m^2, which corresponds closely to an age of 9.5 years and weight of 27–30 kg[42,43]. These data have resulted in a consensus decision to use weight < 30 kg or age < 10 years as thresholds for a smaller mitral regurgitation length cut-off. Ideally, mitral regurgitation jet length should correlate with patient size or left atrial size as a continuous variable, but this criterion is not practical for guidelines in RHD-endemic settings. Therefore, these guidelines recommend the use of 1.5 cm as the cut-off value for patients weighing <30 kg or aged <10 years, for whom shorter mitral regurgitation jet lengths might indicate true rheumatic involvement, especially when smaller atrial sizes are anticipated. Our expert opinion is that the lower cut-off value will prevent the underdiagnosis of RHD in patients weighing < 30 kg or aged <10 years who would otherwise meet the criteria for pathological mitral regurgitation. We believe that the underdiagnosis of younger patients is a more pressing concern than overdiagnosis in RHD-endemic regions.

RHD care implications

The care pathways in these guidelines have been differentiated according to the disease stage, using best-available evidence. Data from a randomized, controlled trial demonstrated that SAP reduced the risk of RHD progression in children aged 5–17 years with stage A or stage B disease[4], with the number needed to treat to prevent one case of RHD progression being 13. However, the adverse effects associated with SAP, including stigmatization, reduced quality of life, allergic reactions and anaphylaxis[44,45], in addition to numerous health-system factors, should be considered before the roll-out of large-scale treatment programmes[4]. Consequently, there is a need to account for family and clinician decisions and health-system factors before recommending SAP in individuals aged ≤20 years with stage A RHD who are at low risk of RHD progression. However, all children with stage A disease must have access to follow-up echocardiography and longitudinal clinical evaluation to monitor for disease progression. We recommend that SAP be continued until a follow-up echocardiogram has been obtained. SAP is recommended for all individuals aged ≤20 years with stage B disease, who have a moderate-to-high risk of disease progression. Finally, SAP is recommended for all individuals with stage C or stage D disease and the treatment regimen should follow relevant local RHD management guidelines. In certain circumstances, such as for individuals without access to a comprehensive echocardiogram report, SAP can be commenced after the criteria for a positive screen have been fulfilled. However, the decision on whether to start SAP should be made with advice from a clinician with RHD experience and all efforts should be made to receive a formal diagnosis with a confirmatory criteria echocardiogram as soon as feasibly possible. These recommendations are summarized in Table 1.

Active case finding

Active case finding refers to active surveillance or screening for disease in at-risk populations within and outside health-care facilities[46,47]. Active case finding for RHD encompasses both coordinated population-based screening programmes and opportunistic case detection in clinical settings. Active case finding for RHD uses echocardiography to identify valvular changes that are consistent with RHD in individuals without a history of ARF and/or RHD. Active case finding can identify a spectrum of valvular changes that are associated with a variable risk of disease progression. Over the past decade, the RHD clinical and research communities have recognized the importance of active case finding to detect mild disease in patients who might benefit from the commencement of SAP and close clinical follow-up[4], as well as patients with moderate or severe disease who require advanced medical and surgical interventions.

RHD meets much of the criteria necessary for conducting population-based screening[33,48,49], albeit with some limitations (Table 3). Population-based echocardiography screening for RHD is resource-demanding. All prospective screening programmes should consider the economic and ethical considerations relevant to that specific jurisdiction, including the effect of the programme on other strategies aimed at addressing the burden of RHD[50]. The current guidelines specifically provide evidence-based guidance on diagnosing RHD by echocardiography that is relevant to various active case-finding strategies. However, the ultimate decision to conduct population-based screening should take into account all other relevant factors and be made by local government and health organizations. Further guidance on population-based screening is outlined subsequently.

Population-based screening for RHD

Candidates. Echocardiography-based screening should be considered only in high-risk populations, as defined by a prevalence of more than 2 cases per 1,000 persons for RHD (all ages) or an incidence of more than 30 cases per 100,000 persons aged 5–14 years annually for ARF[3,51]. Children with no history of ARF (but a presumed subclinical episode) who have developed RHD have the most to gain from the prevention of disease progression. Therefore, most screening programmes for RHD in endemic regions have been undertaken in children and young adults aged 5–20 years[2,6,34,52]. Targeted screening of individuals at an increased risk of RHD, such as siblings (aged 5–20 years) and parents of index cases with ARF or RHD, might also be appropriate[53–55]. The screening of young adults could still be justified for local disease burden management, especially in socially vulnerable and high-prevalence settings. Given the substantial risk of undiagnosed RHD to pregnant women and the fetus, echocardiography screening is also reasonable in pregnant women at high risk of RHD, including those living in endemic regions[56]. These recommendations are summarized in Table 2.

Ethical considerations. Evidence shows that echocardiographic screening combined with SAP for early-stage RHD is more cost-effective than the use of primary prophylaxis for ARF[57]. However, the decision to invest in echocardiographic RHD active case-finding programmes at the expense of other competing health problems with robust evidence-based interventions remains an ethical dilemma, particularly in resource-limited settings[58]. Population-based screening could lead to increased utilization of the already overstretched infrastructure and requires providers to care for more individuals, as well as affecting the quality of life of patients and their families[59]. Although a negative screening result seems to have no adverse effect, a positive screen that might actually be a misdiagnosis can result in stigmatization, unnecessary anxiety, inappropriate SAP use, changes in physical activity levels and a reduction in the quality of life of the individual and their family[60,61]. With these potential harms in mind, implementing population-based screening programmes necessitates the education of health-care professionals about the potential risks and benefits of screening. All programmes should ensure sustainability and the availability of recommended treatment, including SAP and clinical follow-up, for all high-risk individuals, regardless of socio-economic status. This requirement might be even more important in low-resource settings, in which clinical care is prioritized on the basis of disease risk. Furthermore, individuals should be enrolled in a local RHD registry, if available, and the cost of screening should be balanced against potential expenditure on medical care and its effect on other public health strategies[57,62,63].

Echocardiography in acute carditis

Diagnosis of carditis in an index ARF case. A detailed description of echocardiographic findings in ARF is outside the scope of these guidelines and has been reviewed previously[51]. Carditis in the setting of ARF has been described as pancarditis involving the endocardium, myocardium and pericardium. Only endocarditis and, more specifically, valve

Box 6

Confirmatory morphological features of rheumatic heart disease

Morphological characteristics of the mitral valve

- Valve apparatus thickening (defined by the presence of either or both):
  - Anterior leaflet thickening[a,b]
  - Chordal thickening[c]
- Valve mobility abnormalities (defined by the presence of either or both):
  - Restricted anterior or posterior leaflet motion in diastole[d]
  - Excessive anterior leaflet tip motion during systole[e]

Morphological features of the aortic valve

- Cusp thickening[f]
- Cusp prolapse
- Restricted cusp motion
- Coaptation defect in diastole

[a]Anterior mitral valve leaflet (AMVL) thickness should be measured during diastole at the full excursion. Measurement should be taken at the thickest portion of the leaflet, including focal thickening, beading and nodularity. Ideally, the measurement should be performed on a frame with maximal separation of chordae from the leaflet tissue. [b]Abnormal thickening of the AMVL is age-specific and defined as follows: ≥3.0 mm for individuals aged ≤20 years, ≥4.0 mm for individuals aged 21–40 years and ≥5.0 mm for individuals aged >40 years. [c]Chordal thickening might range from individual chordae tendineae to multiple chordae tendineae fusion and calcification. The structures of papillary muscles, chordae tendineae and margins of leaflets might not be clearly distinguishable. Assessment is subjective; however, in most cases, chordal thickening is attributable to fusion of two or more chordae and is seen as an echo-bright structure near to the leaflet tip insertion. Chordal thickening is typically associated with other morphological features. [d]Restricted leaflet motion of either the AMVL or the posterior mitral valve leaflet is usually the result of chordal shortening or fusion, commissural fusion or leaflet thickening. [e]Excessive leaflet tip motion results from elongation of the primary chords and is defined as displacement of the tip or edge of an involved leaflet towards the left atrium, resulting in abnormal coaptation and regurgitation. Excessive leaflet tip motion does not need to meet the standard echocardiographic definition of mitral valve prolapse disease, given that they refer to different disease processes[7]. This characteristic applies only to those aged <35 years. [f]In the parasternal short-axis view, the right and non-coronary aortic cusp closure line often presents as echogenic (thickened) in healthy individuals, which should be considered normal. Image optimization for valve thickness and morphology, including harmonic imaging, should be individualized on the basis of echocardiography devices and those performing echocardiograms.

Box 7

Staging of rheumatic heart disease detected by echocardiography[a,b]

Stage A: minimal echocardiographic criteria for RHD
- Applies only to individuals aged ≤20 years old
- Clinical risk: might be at risk of valvular heart disease progression
- Echocardiographic features: the presence of mild[c] MR or AR without morphological features

Stage B: mild RHD
- Can apply to any age
- Clinical risk: at moderate or high risk of progression and at risk of developing symptoms of valvular heart disease
- Echocardiographic features: evidence of mild[c] valvular regurgitation plus at least one morphological feature in individuals aged ≤20 years and at least two morphological features in individuals aged >20 years[d]; or mild regurgitation in both mitral and aortic valves

Stage C: advanced RHD at risk of clinical complications
- Can apply to any age
- Clinical risk: at high risk of developing clinical complications that require medical or surgical intervention
- Echocardiographic features: moderate or severe MR, moderate or severe AR, any MS or AS[e], pulmonary hypertension and decreased LV systolic function

Stage D: advanced RHD with clinical complications
- Can apply to any age
- Clinical risk: established clinical complications include cardiac surgery, heart failure, arrhythmia, stroke and infective endocarditis
- Echocardiographic features: moderate or severe MR, moderate or severe AR, any MS or AS[e], pulmonary hypertension and decreased LV systolic function

AR, aortic regurgitation; AS, aortic stenosis; LV, left ventricular; MR, mitral regurgitation; MS, mitral stenosis, RHD, rheumatic heart disease. [a]To be applied in high-risk settings[3,51] and requires other causes of valvular heart disease to have been excluded. [b]After the application of the confirmatory echocardiographic criteria, diagnostic categories might include 'normal' and 'other', which encompasses other diseases such as congenital heart disease, cardiomyopathies and pericardial effusion. [c]Fulfilling the confirmatory criteria for pathological regurgitation (Box 4). [d]This cut-off value is derived from expert consensus. [e]Aortic stenosis is defined in accordance with international guidelines on valvular heart disease. A diagnosis of rheumatic aortic stenosis requires the exclusion of other causes, including bicuspid aortic valve and degenerative calcific AS.

dysfunction (valvulitis) are considered major criteria for the diagnosis of carditis[51]. To meet the diagnostic definitions for a major criterion of carditis, pathological mitral and/or aortic regurgitation must be present (as defined in Box 4). Valvular regurgitation in ARF is attributable to various mechanisms, including valvulitis, mitral annular dilatation, leaflet prolapse and chordal elongation or rupture, all of which can be detected on echocardiography[64]. The morphological changes associated with RHD often develop at a later stage of disease or might never transpire and thus are not required for the diagnosis of acute carditis in the setting of ARF.

Diagnosis of acute carditis in the setting of RHD. The differentiation between acute valvulitis and established RHD can pose a clinical dilemma. No evidence exists for the use of echocardiography to differentiate between acute-on-chronic valvulitis and chronic RHD. Echocardiography cannot accurately measure when the rheumatic changes occurred, but a comparison with previous echocardiograms to assess the severity and progression of the valve lesions might be useful in determining whether there is acute valve inflammation in the setting of an ARF recurrence.

Echocardiography equipment

Hand-held echocardiogram devices. The development of hand-held echocardiography devices and improvements in their image quality and functionality have resulted in hand-held devices being a credible option for echocardiographic diagnosis of RHD[65]. All available hand-held devices allow for standard 2D and colour Doppler imaging. The probes of the hand-held devices weigh approximately 130–400 g, making them light enough for comfortable use in clinical practice and active case-finding settings. The phased array (cardiac) probe 'footprint' is small enough to obtain images from most acoustic windows. The hand-held machines can have high frame rates: approximately 40–50 frames/s for 2D imaging and up to 30 frames/s for colour Doppler imaging, depending on the image optimization capacity. The depth (up to 30 cm) and field of view of hand-held devices are adequate for appropriate image acquisition and analysis, and some devices now feature pulsed-wave and continuous-wave Doppler. In addition, certain devices now provide built-in training software, such as artificial intelligence (AI)-guided image acquisition or options to provide real-time teleguidance[66,67].

However, hand-held devices have numerous limitations, including minimal manual image optimization capacity (with most devices now allowing harmonic imaging to be manually turned off), low colour scale and the absence of spectral Doppler imaging. Of note, all these factors associated with hand-held devices can impede the application of the WHF 2023 criteria. Furthermore, the functionality of the hand-held devices varies depending on the specific device characteristics, including battery life, recharge time, memory capabilities, cloud-based file-sharing capacity and the standardization of file types and temperature.

Hand-held echocardiography devices are substantially cheaper than portable ultrasonography machines and currently cost between US $2,000 and US $10,000. Many hand-held devices are sold as stand-alone probes that use widely available, non-brand-specific tablets, allowing operators to use larger screens, which improves disease detection. Finally, hand-held devices are now capable of secure image

upload and storage. Some brands will host their specific study storage options, with limited and proprietary file formats, whereas most devices can upload Digital Imaging and Communications in Medicine (DICOM) formats to non-brand-specific image storage servers.

Portable echocardiogram machines with complete functionality. Portable echocardiography machines are still the mainstay in many RHD active case-finding programmes. Unfortunately, these machines are expensive and thus not accessible in resource-limited settings. In addition to producing high-quality 2D, colour Doppler and continuous-wave Doppler imaging, these portable devices have many extra functions (including global longitudinal strain, automated ejection fraction, stress and 3D imaging), which might not be necessary for RHD detection alone. The recommendations regarding equipment for RHD screening are summarized in Table 2.

Task sharing in RHD-endemic regions

RHD remains prevalent in many low-income and middle-income regions, where there are relatively few cardiologists and cardiac sonographers per capita compared with high-income countries, as well as a substantial disparity in health-care resources between rural and urban regions. An alternative workforce is crucial to the success of population-based active case-finding programmes for RHD in these areas. Task sharing involves strategically redistributing skills or tasks among teams and personnel. Task sharing for RHD active case-finding using echocardiography has been shown to be successful[27,28] and might be a realistic and cost-effective alternative strategy for active case finding. Studies have shown that non-experts can acquire and interpret basic echocardiographic images after brief standardized training, even using limited single-view protocols, and achieve a moderate-to-high sensitivity and specificity[18,28,65,68]. A structured, coordinated and preferably regionally accredited training programme for task sharing is recommended. Furthermore, continuous monitoring and evaluation are vital to the task-sharing process. Discussions with health authorities and medical councils might also be required, given that the regulations in some locations might limit the task sharing of ultrasonography responsibilities to non-physicians. The recommendations for task sharing for active case finding in RHD-endemic regions are summarized in Table 2.

New and emerging technology

RHD active case-finding programmes can benefit from advances in medical technology. Echocardiographic images have been successfully transmitted to experts at an off-site location for analysis and interpretation using cloud-based image storage and viewing platforms (tele-echocardiography)[69]. Task sharing in screening programmes has also been improved by the miniaturization of ultrasonography devices, the increase in image quality and battery life, and the incorporation of dedicated cloud applications and advances features, such as Doppler and probe guidance applications and live videoconferencing[70]. In addition, a clinical trial has tested the utility of smartphone-connected, wireless devices, including pocket echocardiography, in a mobile health clinic to improve outcomes for patients with RHD and other structural heart diseases[71]. Compared with standard care, initial mobile health assessment resulted in a shorter time to diagnosis, increased the probability of referral for advanced surgical care and reduced the risk of adverse outcomes (hospitalization or death) at follow-up[71]. These findings show the potential of integrating technological devices for point-of-care management, especially in resource-limited settings[71].

Table 3 | Suitability of population screening for rheumatic heart disease using echocardiography

Criteria	Criteria met? (level of evidence)	Remaining knowledge gaps
The condition should be an important public health problem	Yes (level B–NR)[1]	The published data are probably an under-estimation of the true global burden of disease, given the scarcity or absolute lack of data from some low-income regions
There should be a recognizable latent or early symptomatic stage	Yes (level B–NR)[76,77]	None
The natural history of the condition, including development from latent to declared disease, should be adequately understood	We have gained a better understanding of the natural history of latent RHD over the past decade (level B–NR)[8,35,78–82]	The longest cohort study had a follow-up of 5 years[82]. Longer follow-up studies are required to elucidate the long-term evolution of the disease
There should be an accepted treatment for patients with recognized disease	The GOAL trial[4] provided evidence that SAP prevents the progression of latent RHD to more advanced stages of the disease over a period of 2 years (level B–R)	The duration of SAP administration remains undefined. In particular, data to support the cessation of SAP for children with latent RHD who show echocardiographic normalization in follow-up studies are lacking
There should be a suitable test or examination that has a high level of accuracy	Yes, but with limitations. Echocardiography is non-invasive and is readily available, but the 2012 WHF echocardiographic diagnostic criteria were not easy to apply at scale (owing to technical and personnel reasons) for population-level screening[32]. This revision of the guidelines has aimed to address these challenges (level B–NR)	Need to test the performance of these revised criteria as they are applied in clinical and research practice
The test should be acceptable to the population	Yes (level B–NR)[83,84]	Further studies required of the potential harms of screening[85]
There should be an agreed policy on whom to treat as patients	Yes, regional and national guidelines are available (level B–NR)	There has not been an update on the international treatment guidelines for RHD since the publication of the GOAL trial[4]. The current guidelines provide guidance with varying levels of evidence, but recommendations need to be refined

NR, non-randomized; R, randomized; RHD, rheumatic heart disease; SAP, secondary antibiotic prophylaxis.

Preliminary data have been published on the use of AI technology to improve automated RHD diagnosis in clinical practice. A Brazilian study showed that a deep learning approach had moderate accuracy (72.8%, 95% CI 69.3–76.3%) for flagging latent RHD in screening images acquired using hand-held devices, with better accuracy for definite RHD[72,73]. AI algorithms have also successfully identified and quantified mitral regurgitation in children, a key feature for RHD diagnosis in screening studies[74]. A novel AI approach for computer-assisted auscultation showed low sensitivity (4.0%) but reasonable specificity (93.7%) for RHD detection compared with echocardiography[22]. Finally, AI-based software has been newly incorporated into echocardiogram devices to guide optimal probe positioning and to facilitate imaging acquisition[66], and AI systems are currently being trained specifically to diagnose RHD. Therefore, although data are still scarce, AI technology holds promise for future development and incorporation into RHD diagnostic flowcharts. A list of recommendations for the use of new technology to detect RHD is presented in Table 2.

Future directions

Validation studies are essential to determine the utility of the new WHF 2023 criteria in communities. Although the abbreviated criteria aim to simplify the echocardiographic screening of RHD, an adequate balance of sensitivity and specificity must be maintained. Therefore, a series of studies are essential to assess whether these criteria are sensitive enough to detect most RHD and, at the same time, maintain adequate specificity. In addition, research is needed to validate and strengthen the age-specific and size-specific recommendations and further characterize the use of rheumatic regurgitant jet length as a criterion, to improve our capacity to distinguish pathology from physiology. Furthermore, continued efforts are required to ensure that the diagnostic criteria can best identify those individuals who will benefit the most from SAP.

In addition, the performance of the newer portable echocardiography devices should also be evaluated, as well as the new applications for imaging guidance, off-site review of images and automated flagging of abnormalities. AI technology for the diagnosis of RHD should continue to be developed.

Finally, cost–effectiveness and implementation research are required to determine the sustainability of various models of RHD active case finding. Guidance on the threshold for implementing active case finding is needed, as well as guidance on the ideal age for screening children. Furthermore, active case-finding programmes might have a role in evaluating vaccine efficacy and safety.

Conclusions

The WHF 2023 guidelines for the echocardiographic diagnosis of RHD provides a contemporary resource for researchers and health-care practitioners in RHD-endemic regions around the world. These guidelines provide an updated, evidence-based set of criteria for diagnosing RHD, which empowers clinicians to make management decisions on the basis of the risk of an individual of disease progression. These diagnostic criteria support the growing demand for task sharing, particularly in resource-limited settings, and complement local, national and international RHD treatment recommendations. In revising the guidelines, we have also identified areas for future research, including continued evaluation of the applicability of this new set of criteria in research and clinical settings to ensure that it remains relevant to end-users.

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Competing interests

The authors declare no competing interests.

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