CURRENT OPINION

Rheumatic heart disease control: the time for a paradigm shift

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ABSTRACT

Rheumatic heart disease (RHD) is a completely preventable, life-threatening complication of group A streptococcal pharyngitis and the commonest cause of acquired heart disease in children and young adults in low- and middle-income countries. Conventional control measures are faced with many obstacles including the difficulty of early diagnosis of bacterial pharyngitis and acute rheumatic fever (ARF) leading to late presentation with established RHD which is not curable. Recent evidence confirmed the role of echocardiography screening of asymptomatic children in the early detection of ‘latent’ RHD. Benzathine penicillin prophylaxis was shown to be effective in halting the progression of latent RHD. There is enough evidence to warrant the implementation of control strategies that use lower thresholds for the diagnosis of group A streptococcal infection and ARF and we believe that it is high time to introduce an echocardiography screen-to-treat policy in endemic areas.

KEYWORDS
Rheumatic heart disease; Control; Echocardiography; Screening; Latent.

Rheumatic heart disease (RHD) is a completely preventable life-threatening sequel of a relatively simple infection with Group A beta-haemolytic streptococcus (GAS) that results in acute rheumatic fever (ARF). If not treated early, ARF can result in carditis leading to permanent, incurable destruction of heart valves which rapidly progresses to heart failure and death. The estimated prevalence of RHD is 40 million people with the maximum burden in South Asia and sub-Saharan Africa [1]. RHD has almost disappeared from developed countries since the 1940s except for pockets in marginalised communities. Therefore, the disease was neglected for many decades leading to flaring in low-income countries. Global control measures were initiated in the 1990s by the World Health Organisation (WHO) but soon they ceased and were almost non-existing till mid-year 2000 [2]. In the year 2007, echocardiographic (echo) screening of large cohorts of asymptomatic people revealed a huge burden of subclinical disease [3]. Simultaneously, control initiatives were introduced by many organisations including the Pan African Society of Cardiology (PASCAR) [4], and the World Heart Federation (WHF) [5]. In 2018, WHO issued a resolution for RHD which

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Received: 17 May 2019 | Accepted: 23 October 2019

How to cite this article:
signifies and is expected to be the start of a global campaign against RHD [6].

THE ERA OF ECHO-DIAGNOSED CARDITIS

Since 2004, the WHO recognised that echo can diagnose RHD in patients without clinically detected signs [6]. With the rapid improvement of echo technology, smaller (portable) echo machines became available which resulted in echo screening of asymptomatic people revealing a striking discrepancy between clinical and echo-diagnosed RHD [2]. The echo prevalence was found to be several folds higher than that discovered by auscultation. Subsequently, many studies from several highly endemic countries revealed similar findings unmasking a huge burden of subclinical disease [7–10]. These findings led to the re-classification of RHD into the following categories:

1. **Subclinical** (echo diagnosed/ silent/latent) RHD
2. **Clinical asymptomatic** RHD with heart murmurs (early RHD)
3. **Clinical symptomatic** (severe/late) RHD presenting with heart failure or other complications (such as stroke and endocarditis)

In 2012, the WHF published guidelines for the diagnosis of subclinical carditis that included two-dimensional echo, colour and spectral Doppler criteria from two echo views [11]. However, these criteria need either a standard or portable echo machine with spectral Doppler which limited their use in remote areas. Subsequent studies tested a focused protocol modified from the WHF criteria using handheld echo machines and a single echo view. The protocol utilises two-dimensional and colour Doppler echo, omitting spectral Doppler which allowed more accessibility in remote areas. The protocol was found to have a reliability of 98.7% for definite and 94.7% for borderline disease [12]. Using this focused protocol, we established RHD control sites based on echo screen-to-treat policy in the nine states in Sudan. The programs were coupled with health worker training and public education in highly endemic areas [13,14].

Further studies documented that subclinical RHD can progress to the clinical stage. However, the role of antibiotic prophylaxis in halting the progression of the disease was not confirmed [15]. In 2022, a randomised controlled study from Uganda proved that Benzathine penicillin G (BPG) can stop the progression of subclinical RHD paving the way for establishing an echo-based control program [16].

RHD control: the conventional policies and their limitations

There had been many models for RHD control including the PASCAR and the WHF [5,17]. Most of the objectives of these models depend on primary prevention (targeting early diagnosis and treatment of GAS), secondary prevention (targeting early diagnosis and treatment of ARF) and tertiary prevention which aims to prevent complications of established RHD. However, many limitations exist to these parameters as shown in Figure 1.

Time for a paradigm shift

Due to the described limitations in the conventional model of RHD control, and the new evidence indicating the benefit of BPG in halting the progression of echo-diagnosed RHD, a need for a paradigm shift in RHD control is evident. The followings are the main features of the suggested model (Figure 2) and the tools needed to implement it (Figure 3) [18].

Setting simple algorithm for diagnosis and treatment of GAS pharyngitis

In the absence of a reliable and affordable test for GAS pharyngitis, it is widely accepted to use protocols for the diagnosis of bacterial pharyngitis. The stricter the rules, the more likely for them to miss positive cases of GAS; therefore, it is desirable to use the most sensitive criteria in RHD endemic areas [19]. Stratification of patients according to their susceptibility to ARF/RHD is useful, this has been applied to New Zealand
Figure 1. Limitations of conventional RHD control policies. ARF, Acute rheumatic fever; BPG, Benzathine penicillin G; GAS, Group A streptococcal infection; RHD, Rheumatic heart disease.

Figure 2. Suggested policy for RHD control. ARF, Acute rheumatic fever; GAS, Group A streptococcal infection; JCC, Joint, cardiac, chorea.

Figure 3. Tools needed to implement RHD control. ASOT, Antistreptolysin O titre; ESR, Erythrocyte sedimentation rate, INR; International normalisation ratio; JCC, Joint, cardiac, chorea.
GAS protocols. In high-risk categories, empiric treatment of sore throat is advised [20].

The standard treatment for GAS pharyngitis is one injection of BPG or a 10-day course of oral penicillin [21]. However, BPG is preferred due to its better bactericidal effect as well as guaranteed compliance. Therefore, the minimum number of clinical symptoms and signs together with the use of a single injection of BPG is recommended in order to improve the primary prevention of ARF/RHD.

Lowering the threshold for diagnosis and treatment of ARF

In primary and secondary care settings, it might be cumbersome for health workers to refer to Jones Criteria due to their complexity and multiple items. The main objectives of the 2015 update of the Jones Criteria were to include echo as a major criterion and to lower the threshold for diagnosis of ARF in endemic areas by including mono arthralgia and introducing a new category of ‘probable ARF’ which indicates the use of a combination of symptoms suggestive of ARF without fulfilling the whole criteria [22]. The most common symptoms of ARF are joint and cardiac symptoms followed by chorea (JCC). Therefore, health workers can be trained to treat any of these symptoms as probable ARF, start BPG and refer to the next level of care. On the other hand, many endemic diseases can mimic ARF which leads to overlapping of diagnoses, however, the clinical picture and investigations will reveal the correct diagnosis at the referral hospital where an echo is expected to be done by specialists.

Echo screen-to-treat policy in endemic areas

This indicates active surveillance for RHD in endemic areas by screening asymptomatic school children using portable/handheld echo machines. The operators can be trained physicians or other health professionals who can record the images for later review by cardiologists. Positive cases need to be started immediately on BPG and referred to the next level of care for re-evaluation.

Expected limitations and suggested solutions

BPG and oral penicillin

BPG is the cornerstone drug for the treatment of GAS and secondary prevention of ARF with proven efficacy for both conditions. The drug has inherent problems which face the patients as well as health workers (Table 1). However, there is currently no ideal alternative to BPG, moreover, GAS is still sensitive to it with no reported resistance. WHO and other health organisations are urged to improve the supply of BPG and oral penicillin in RHD-endemic countries. Protocols for safe administration need to be made available and training of health workers should be intensified in endemic areas.

Over-diagnosis of ARF

The new protocol entails the diagnosis of any joint symptom as ARF, and this can lead to overdiagnosis as there are many other causes of arthritis.

<table>
<thead>
<tr>
<th>Problem</th>
<th>Suggested solution</th>
</tr>
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<tbody>
<tr>
<td>The drug is heavy</td>
<td>-Dissolve in the recommended volume of diluent</td>
</tr>
<tr>
<td>The drug may obstruct the needle</td>
<td>-Use a large bore needle</td>
</tr>
<tr>
<td></td>
<td>- Avoid cold diluent</td>
</tr>
<tr>
<td>The drug is painful</td>
<td>Use local anaesthetic as a diluent</td>
</tr>
<tr>
<td>Patients fear allergy</td>
<td>Allergy is rare, ask patients to report previous reactions</td>
</tr>
<tr>
<td>Health workers fear reactions</td>
<td>Use a protocol for health worker training</td>
</tr>
<tr>
<td>The drug can lead to sudden collapse</td>
<td>-Avoid using in uncontrolled heart failure</td>
</tr>
<tr>
<td></td>
<td>-Patients need to have oral fluids to avoid dehydration before the injection</td>
</tr>
<tr>
<td>Drug supply shortage</td>
<td>Global efforts to improve availability</td>
</tr>
</tbody>
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Table 1. Problems of BPG and suggested solutions.
of acute arthritis in children. In an endemic area, malaria and typhoid fever as well as viral infections can lead to joint symptoms that can mimic ARF. Health workers tend to diagnose these ‘mimickers’ more often than ARF as has been observed during daily practice. It is safer to start BPG prophylaxis and send the patient for an echo while investigating the cause of joint symptoms. A specified protocol is needed to direct health workers to the right diagnosis and treatment.

Over-diagnosis of RHD by echo

Training of health workers needs to be guided by protocols set by expert cardiologists. The health workers performing echo could be physicians or non-physicians; task shifting utilising nurses and other health professionals has been tested in some countries with acceptable results [23,24]. After the screening echo, patients need to be referred for evaluation by a trained cardiologist to re-evaluate the diagnosis and treatment which will decrease the potential for overdiagnosis.

CONCLUSION

Echo has reshaped the spectrum of RHD unmasking a huge burden of subclinical carditis. Evidence has shown that BPG stopped the progression of subclinical RHD. The conventional approach to controlling RHD has many limitations. A new approach utilising fewer criteria for ARF and active echo surveillance is suggested.

CONFLICT OF INTERESTS

The authors declare that they have no competing interests.

FUNDING

None.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

REFERENCES


