Clinical Profile and Management in Children with Rheumatic Heart Disease in a Tertiary Cardiac Care Center of Nepal

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Abstract

Backgrounds and Aims: Rheumatic heart disease is the commonest acquired heart disease in children of developing world presenting earlier and with a more severe valvular lesion. This study was conducted to assess the clinical and demographic profile, pattern of valvular involvement and management modalities among children with rheumatic heart disease.

Methods: This was a hospital-based observational study, performed at Shahid Gangalal National Heart Center where all children less than 15 years of age with confirmed echocardiographic diagnosis of rheumatic heart disease from June 2015 to May 2016 were evaluated and relevant data were recorded.

Results: Two hundred and eleven children were diagnosed with rheumatic heart disease. The mean age was 11.9 ± 2.2 years with (100) 47.4% male and (111) 52.6% female. The median duration of symptoms among new cases was 1 year (interquartile range of 0.5 to 2 years). The most common symptom was dyspnea present in (187) 88.6% children followed by palpitation (34.5%). Mitral valve was most commonly affected in (141) 96.5% while isolated aortic valve involvement was seen in (8) 3.5%. Isolated mitral regurgitation was the most common lesion present in 118 (55.9%) children, while 50 (23.7%) children had combination of mitral and aortic regurgitation. Mitral valve was significantly more commonly affected in females and aortic valve commonly affected in males (p= 0.003).

Conclusions: Rheumatic heart disease although declining in developed countries, remains to be the commonest acquired heart disease in developing and underdeveloped countries. Efforts are needed to improve primary and secondary preventive measures.

Key Words: Acquired heart disease, children, Rheumatic heart disease.
Results:
A total of 211 children with RHD attended the pediatric cardiology clinic and emergency department of SGNHC during the study period. Among them 111 children (52.6%) were female. The mean age of children with RHD was 11.9 ± 2.2 years with the youngest child being 5.9 years and the oldest being 15 years. The mean age of male children was 12.1 ± 2.0 years while females were slightly younger with mean age of 11.8 ± 2.3 years.

Figure 1 demonstrated the distribution of cases from different districts of Nepal. Children from almost all parts of Nepal have been affected with RHD. However, we did not have any cases from few districts of hilly and mountains of western and far western region of Nepal because of difficulty in access either to the capital or because of lack of awareness regarding the disease in those areas. The maximum number of children was from Rukum followed by Surkhet and Sindhuli.

Among all cases of RHD, 103 children (48.8%) presented to the hospital for the first time and 108 (51.2%) children were known cases who come for follow up. The median duration of symptoms among new cases was 1 year (ranging from 1 week to 9 years). Similarly, only 35 children (34%) who presented to the hospital for the first time received medication from outside while 68 children were referred to this tertiary center without any medication. The most common symptom in children with RHD was dyspnea. Table 1 shows the frequency of various presenting symptoms. Past history suggestive of Rheumatic fever (joint pain and swelling associated with fever) was present only in 47 (22.3%) children. There were a total of 19 children with infective endocarditis during the study period, among whom 16 children (84%) was those with RHD.

Mitrval valve was the most common valve affected with RHD present in 96.3% of children while isolated aortic valve involvement was seen in eight (3.5%) children. Table 2 shows the gender distribution of the various valves involved in RHD. Mitrval valve involvement was significantly more common in female children whereas aortic valve involvement was significantly more in males (p = 0.003). Similarly, Table3depicts the various combinations of valvular involvement in the study population.

### Table 1. Frequencies of various symptoms in a child with RHD

<table>
<thead>
<tr>
<th>Presenting symptoms</th>
<th>Number of children</th>
<th>Percentage</th>
<th>Total (n=935)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyspnea</td>
<td>187</td>
<td>88.6</td>
<td>385</td>
</tr>
<tr>
<td>Palpitation</td>
<td>73</td>
<td>34.5</td>
<td>251</td>
</tr>
<tr>
<td>Chronic cough</td>
<td>54</td>
<td>25.5</td>
<td>232</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>38</td>
<td>18</td>
<td>67</td>
</tr>
<tr>
<td>Fever</td>
<td>24</td>
<td>11.3</td>
<td></td>
</tr>
<tr>
<td>Arthritis</td>
<td>7</td>
<td>3.3</td>
<td></td>
</tr>
<tr>
<td>Chorea</td>
<td>3</td>
<td>1.4</td>
<td></td>
</tr>
</tbody>
</table>

### Table 2. Gender distribution of different types of valvular lesion in children with RHD

<table>
<thead>
<tr>
<th>Valvular lesion</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mitral</td>
<td>59</td>
<td>82</td>
<td>141</td>
<td>0.003</td>
</tr>
<tr>
<td>Aortic</td>
<td>8</td>
<td>0</td>
<td>8</td>
<td>0.003</td>
</tr>
<tr>
<td>Combined</td>
<td>33</td>
<td>29</td>
<td>62</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>Valvular lesion</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mitral</td>
<td>85</td>
<td>105</td>
<td>190</td>
<td>0.02</td>
<td></td>
</tr>
<tr>
<td>Aortic</td>
<td>13</td>
<td>18</td>
<td>31</td>
<td>0.5</td>
<td></td>
</tr>
<tr>
<td>Combined</td>
<td>39</td>
<td>29</td>
<td>68</td>
<td>0.04</td>
<td></td>
</tr>
<tr>
<td>Mitral</td>
<td>5</td>
<td>5</td>
<td>10</td>
<td>0.86</td>
<td></td>
</tr>
</tbody>
</table>

MV: mitral valve; AV: aortic valve; MR: mitral regurgitation; MS: mitral stenosis; AR: aortic regurgitation; AS: aortic stenosis
Percent with 470,000 reported new cases and 233,000 deaths annually.1 As the commonest acquired heart disease in children of poor nations, rheumatic fever and RHD, though rare in developed nations, is receiving the attention they require.7 The global burden of RHD affects the poorest people of the world, they often fail to receive the attention they require.7 The global burden of RHD varies according to various regions with unreliable estimates due to missing data in many countries and the use of different methods for diagnosis.2

In this study, 147 (69.6%) children were managed in the outpatient department with oral medications, 24 (11.4%) in the inpatient department with oral medications, 24 (11.4%) were referred to the surgical department for valve repair/replacement and/or heart failure and 40 (19%) were referred to the surgical department for valve repair/replacement.

DISCUSSION

Rheumatic fever and RHD, though rare in developed nations, is the commonest acquired heart disease in children of poor nations with 470,000 reported new cases and 233,000 deaths annually.2 As RHD affects the poorest people of the world, they often fail to receive the attention they require.7 The global burden of RHD varies according to various regions with unreliable estimates due to missing data in many countries and the use of different methods for diagnosis.2

Few studies reported, more so only in and around Kathmandu valley to estimate the incidence and prevalence of RHD in Nepalese children. Although the prevalence was initially estimated to be 1 to 1.3/1000 in school children, presence of subclinical carditis in normal children results in much higher prevalence of RHD in the population.4,8,9 Shrestha et al in their recent population based study in school going children of Eastern Nepal have reported an alarming prevalence of 10.2/1000 school children.10

The mean age of children with RHD was 11.9 ± 2.2 years. This is consistent with the result from Nigeria where the mean age of children with RHD was recorded to be 10.8 ± 3.2 years.10 Females were slightly more affected in this study with male to female ratio of 0.9:1 which is similar to studies done in different parts of the world.11 Further we have observed that girls with RHD present in 88% of children followed by palpitation. This finding is in contrast to that observed by Zhang et al from Uganda where palpitation was the most common observed symptom followed by fatigue and dyspnea.14 This could be because their study was done in adult population and children are less likely to notice or verbalize more subjective symptoms like palpitation or fatigue. The symptoms a child presents in the clinic are usually those that are observed by the mother/caretaker. Mitral valve was the most common valve affected with RHD present in 96.5% of children. Isolated aortic valve involvement was seen in eight (3.5%) children. Similarly, mitral valve involvement was significantly more common in female children whereas aortic valvulitis was observed to be more in males. This was similar to the study from eastern Nepal where mitral stenosis was more common in females and aortic regurgitation in males.15

Infective endocarditis was observed in 16% of children with RHD which is the cause for 84% of cases with infective endocarditis. The association of this deadly and potentially fatal complication with RHD is almost present only in Asian countries.7 Studies from India and Turkey have shown RHD to be the most common cause for infective endocarditis representing 33% to 66% of cases6,17,18 whereas in Singapore RHD was responsible for only 4% of endocarditis cases.19

RHD is the second commonest cause of open heart surgery in Nepal.20 However, valvular surgery in pediatric population has its own difficulties regarding the procedure, the size of the valve and immediate postoperative complications.21 Further there is an additional burden of lifelong oral anticoagulants required after valvular replacement therapy along with the complication of valve thrombosis and systemic embolic disasters.22 However, 40 children of less than 15 years of age had to undergo valvular repair/replacement surgery as their symptoms despite adequate and aggressive medical therapy. This is probably because these children presented late in the course of the illness with severely deformed valves and significant dilatation of the heart chambers. This observational study has certain limitation. Being a single center study done it does not reflect the prevalence of the whole country. Further being a tertiary level referral center it may represent only advanced and severe cases that were referred from the periphery. Similarly, we could not analyze the status of pulmonary hypertension and specific chamber dilatation. Finally follow up analysis was not done to look for the outcome after medical/surgical management.

Conclusion

RHD although in declining phase in developed countries, remains to be the commonest acquired heart disease in poorly developed nations. Its increased incidence is complicated by late presentation to the hospital increasing the severity of valvular involvement of this potentially preventable condition. Proper education and awareness along with effective primary and secondary preventive strategies remain the cornerstone for the control of this common childhood condition.

References: