

## Chorea Revisited

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### Abstract

#### Objectives

To re-evaluate the clinical signs and etiology of chorea in the modern era of antibiotics and health delivery system.

#### Material and Methods

This prospective descriptive study was conducted in the Department of Medicine, Khyber Teaching Hospital, Peshawar, from January 2001 to April 2008. Twenty eight patients presenting with chorea were included in the study. Patients were thoroughly examined for various clinical signs and their distribution. Relevant investigations were done to confirm etiology of chorea.

#### Results

Rheumatic chorea was the commonest (14 patients, 50%) among 28 patients in total. Most patients were in the age range of 11-20 years (42.9%). Male to female ratio was 1.3:1. Generalized chorea was seen in 20 patients (71.4%), hemi chorea in five patients (17.7%) and focal chorea in three patients (10.7%). Right sided chorea (Pure / predominant, focal / generalized) was more common i.e. 14 (50%). Milkmaid grip was seen in 25 (89.3%), piano movements were present in 24 (85.7%) and tongue signs in 18 (64.3%). Pendular deep tendon reflexes were found in seven patients (25%).

#### Conclusions

Rheumatic chorea is still the commonest cause of chorea and involuntary movements are more common in right sided limbs.

#### Key words

Chorea, Rheumatic Fever.

#### Introduction

Chorea is a hyperkinetic state characterized by spontaneous, abrupt, random and irregular movements<sup>1</sup>. It may be hereditary or non-hereditary (sporadic)<sup>2</sup>. Non-hereditary chorea may be unilateral (usually due to contralateral hemispheric lesions) or bilateral (due to degenerative, metabolic or toxic brain diseases)<sup>3</sup>. The association between rheumatic fever and chorea was recognized in the 19<sup>th</sup> century<sup>4</sup>. Isolation of streptococci and their relationship to chorea and rheumatic fever was confirmed in early 20<sup>th</sup> century<sup>5</sup>. The recognition of post-streptococcal

tics in the 1980s<sup>6</sup> and subsequent recognition of Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections (PANDAS)<sup>7</sup> led to renewed interest in rheumatic chorea. Chorea is believed to be caused by disturbance at the basal ganglia<sup>8</sup>. The most consistent pathologic finding is inflammation<sup>9</sup> and enlargement<sup>10</sup> of corpus striatum in the acute phase of chorea with normalization during remission<sup>11</sup>. Biochemically, hypermetabolism and increased glucose consumption in the basal ganglia<sup>12</sup>, and reduction of n-acetyl aspartate in the striatum<sup>13</sup> (indicative of neuronal dysfunction or loss) or enhanced dopaminergic activity<sup>14</sup> has been demonstrated. Immunologically, anti-neuronal antibodies binding to caudate nucleus (antibody hypothesis)<sup>15</sup> has been proposed as a possible mechanism for chorea.

With the widespread use of antibiotics and prompt treatment of streptococcal infection, the incidence of rheumatic fever has gone down in the west, but has probably been replaced by PANDAS<sup>7</sup>. We wanted to see the possible etiological factors and the frequency of various signs in chorea, in the present era when antibiotics are widely available and people have reasonably good access to medical help.

#### Material and Methods

This prospective, descriptive study was conducted in the Department of Medicine, Khyber Teaching Hospital, Peshawar, from January 2001 to April 2008. All patients presenting with symptoms and signs of chorea were included in the study. During hospitalization detailed history was taken and patients underwent a thorough physical examination. Patients were observed for two minutes for spontaneous movements. Symmetry/asymmetry between right and left side of the body or between upper and lower limbs was recorded. Milkmaid grip and piano movements were also assessed for two minutes to see equality/inequality between right and left side of the body. Blood tests including full blood count, urinalysis, liver function tests, thyroid function test, blood sugar levels, serum electrolytes, autoantibody screen, antistreptolysin O (ASO) titers, serum copper and ceruloplasmin level were requested. Electrocardiography and imaging studies including Magnetic Resonance Imaging (MRI) / Computed Tomography (CT) of brain were also performed to confirm the cause of chorea.

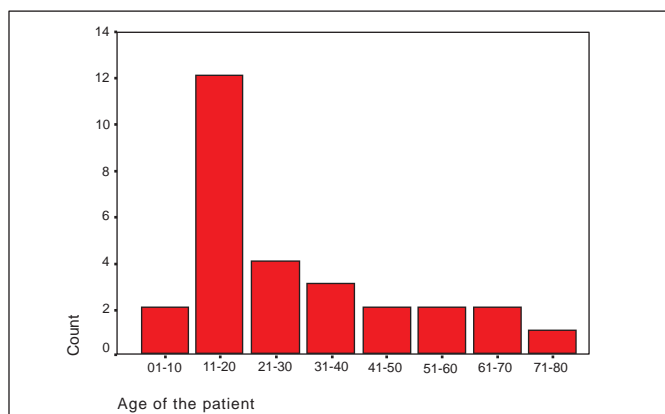
#### Results

A total of 28 patients (16 male [57.1%]) and 12 female [42.9%])

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with minimum age of 6 years and maximum age of 74 years, were included in the study. Distribution of chorea in different age groups is shown in Fig. 1. Most of the patients were in the age range of 11-20 years (42.9%). Male to female ratio was 1.3:1 in general but the reverse was true in subgroup of patients with rheumatic chorea. Rheumatic chorea was seen in 14 patients (50%), with 12 patients (85.7%) below 20 years age. No cause was found in two patients (7.1%). Etiology of chorea is shown in Table 1. Generalized chorea (Table 2) was found in 20 patients (71.4%), hemi chorea in five (17.7%) and focal chorea in three patients (10.7%). Involuntary spontaneous movements characteristic of chorea were seen in all 28 patients; however, 14 patients (50%) had pure / predominant involvement of the right side of the body (focal / generalized) and 11 patients (39.3%) had predominant involvement of the upper limbs (Table 2). Milkmaid grip was present in 25 patients (89.3%) with equal involvement of both upper limbs in 15 (53.6%), and predominant involvement of the right side in six patients (17.9%). Tongue signs were present in 18 patients (64.3%), with 'jack in the box sign' in 12 (42.9%) and 'bag of worms sign' in six (21.4%). Piano movements of the fingers and toes were present in 24 patients (85.7%) with 10 patients (35.7%) having right sided or predominantly right sided involvement. Deep tendon reflexes were pendular in seven patients (25%).

**Figure 1. Age of the patients.**



## Discussion

Chorea is a clinical manifestation of disorder of the corpus striatum but it is not yet clear why some patients present with chorea while others present with tics, dystonia or myoclonus. The predominance of Sydenham's chorea in females, its relapse during pregnancy<sup>16</sup> and predominance of post-streptococcal tics in males<sup>17</sup> suggests a possible influence of sex hormones on the phenotypic expression of movement disorders. Results in our study reconfirm the female predominance in rheumatic chorea (male to female ratio of 1:1.3), the same as reported by Dale et al<sup>18</sup> and Cardoso F et al<sup>19</sup>. When combined with other causes of chorea this ratio was reversed with male predominance (male to female ratio of 1.3:1). We report 50% patients in our study group with rheumatic chorea, 7.1% patients with drug-induced

**Table No 1. Cause of chorea in cases studied (n=28)**

Cause of chorea	n	%
Rheumatic chorea	14	50%
Drugs (Levodopa)	2	7.1%
Brain atrophy / ischemia	2	7.1%
Intracranial tumor	1	3.6%
Adenoleukodystrophy	1	3.6%
Right occipital cyst	1	3.6%
Chorea gravidarum	1	3.6%
Huntington's chorea	1	3.6%
Wilson's disease	1	3.6%
Systemic lupus erythematosus	1	3.6%
Thyrotoxicosis	1	3.6%
Idiopathic	2	7.1%

**Table No 2. Spontaneous movements involving limbs (n=28)**

Generalized chorea	n	%
All 4 limbs equally involved	4	14.3%
All 4 limbs involved but Right more than left limbs (Predominant right hemichorea)	8	28.6%
All 4 limbs involved but Left more than right limbs (Predominant left hemichorea)	4	14.3%
All 4 limbs involved but upper more than lower limbs (Predominant cephalad chorea)	4	14.3%
<b>Hemichorea</b>		
Right limbs involved only (Pure right hemichorea)	4	14.3%
Left limbs involved only (Pure left hemichorea)	1	3.6%
<b>Focal chorea</b>		
Only upper limbs but equally involved	1	3.6%
Only upper limbs but right more than left	1	3.6%
Only right upper limb involved	1	3.6%

chorea, 7.1% with chorea due to brain atrophy/ischemia and 3.6% with Huntington's chorea. Mendes et al<sup>20</sup> reported 51.3% rheumatic chorea, 18.5% Huntington's chorea and 9.2% post-stroke chorea; figures similar to ours. Piccolo et al<sup>21</sup> reported drug-induced chorea in 11.7% patients and Huntington's chorea in 10% patients. No cause was found in 7.14% in our study which is comparable to 5.9% reported by Piccolo et al<sup>21</sup>. et al<sup>22</sup> reports drugs, pregnancy, vascular disease, thyrotoxicosis, and

systemic lupus erythematosus as the major causes of chorea. The same causes were responsible for chorea in our study as well (Table 1). Rheumatic chorea was the most common form of chorea (85.7%) in young patients (<20 years age) while non-rheumatic chorea was seen in older persons in our study. et al<sup>14</sup> reports that Sydenham's chorea is the most common form of childhood chorea, whereas Huntington's disease and drug induced chorea account for the majority of adult onset cases.

Involuntary spontaneous movements characteristic of chorea were seen in all 28 patients, the same as reported by Al-Eissa A<sup>23</sup>. Distribution of chorea as to the body parts involved (Table2) was variable in our study even if the cause of the chorea was the same, which is quite consistent with the earlier published work. Chorea may involve any part of the body<sup>24</sup>, or may predominantly involve one side, or be completely one sided<sup>25</sup>. Choreiform movements involving the right side or predominantly the right sided limbs were seen in 14 patients (50%) and were completely unilateral in six patients (21.5%) in our study. et al<sup>26</sup> reports that chorea is usually bilateral but in up to 30% patients it may be unilateral. Al-Eissa A<sup>23</sup> has reported hemichorea as the most common presentation, often affecting the right side. We report 71.4% patients with generalized chorea while Aron AM et al<sup>27</sup> and Nausieda PA et al<sup>28</sup> has reported >80% patients presenting with generalized chorea.

Milkmaid grip was the commonest sign, followed by piano movements of the fingers and toes, 'jack in the box sign', 'bag of worms sign', and pendular deep tendon reflexes. Chorea paralytica/Mollis, a rare form of chorea<sup>29</sup>, was not seen in our study.

### Conclusion

We conclude that rheumatic fever is still the commonest cause of chorea despite advances in knowledge and availability of antibiotics. We suggest the need for early diagnosis and treatment of streptococcal infection. Organized and continuous public awareness campaigns might help in this regard. The right sided dominance may suggest the dominant role of left sided corpus striatum. However, additional research is needed for further understanding of this condition.

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