

Prevalence of Sydenham's chorea in patients with Acute Rheumatic fever in Nepal.

Regmi PR¹, Shrestha A², Khanal HH¹, Nepal BP¹, Chapagain P¹

¹Cardiology Unit, Bir Hospital, NAMS

²Neurology Unit, Bir Hospital, NAMS

Abstract

Background

Sydenham's Chorea (SC) is a major manifestation of acute rheumatic fever (ARF) and is the only evidence of ARF in approximately 20% of cases. There are no published reports on prevalence of SC in patients with ARF in Nepal.

Objectives

This retrospective study was done to find out the prevalence of SC in patients with ARF in Nepal.

Methods

Four years data of 672 patients with ARF from the RF/RHD registry of National RF/RHD prevention & control programme launched by Ministry of Health and Population & Nepal Heart Foundation in 32 hospitals throughout Nepal were analyzed.

Results

Prevalence of SC as a major manifestation of ARF in Nepalese patients was found to be 3.87%. Females were mostly affected with a female-to-male ratio of 3:1. Patients of age group 10-15 years were affected the most. SC was the third most common manifestation of ARF after Arthritis and Carditis.

Keywords Acute Rheumatic Fever, RF/RHD prevention and control programme, Sydenham's chorea,

Introduction:

Sydenham's chorea (SC) or chorea minor (historically referred to as Saint Vitas' Dances)¹ is a disease characterized by rapid, uncoordinated jerking movements affecting primarily the face, and hands. Sydenham's chorea results from childhood infection with group A Beta hemolytic Streptococci² and is reported to occur in 5 - 36 % of patients with Acute Rheumatic Fever (ARF).³ The disease is usually latent, occurring up to 6 months after the acute infection, but might occasionally be the presenting symptom of rheumatic fever. SC is one of the major manifestations of ARF described in Jones criteria for diagnosis of ARF. SC is more common in females than in males and most patients are children below 18 years of age. Adult onset of SC is comparatively rare and most of the adult cases are associated with exacerbation of chorea following childhood SC. It is named after British Physician Thomas Sydenham (1624 – 1689).⁴ There are no published studies on prevalence of SC in patients with ARF in Nepal.

There are several published studies on prevalence of RF / RHD in Nepal. Prevalence of RF / RHD has been reported to be 1.0 to 1.2 per 1000 school aged children in Nepal.⁵⁻⁷ None of these studies have data on manifestations of ARF. There are very few published studies on manifestations of ARF in international medical literature. We have tried to collect data on RF / RHD with SC as manifestation of ARF.

Ministry of health and population (MoHP) is running National RF/RHD prevention & control programme throughout the country with technical support from Nepal Heart Foundation (NHF). Registry of RF/RHD is maintained in 32 participating hospitals throughout the country from east to far west. The registry includes details of the RF/RHD patients receiving secondary prophylaxis. A central Registry of RF / RHD patients is also maintained at the NHF central office located in Kathmandu.⁸ The purpose of this study was to find out the prevalence of SC in Patients with Acute RF in Nepal.

Methods:

Four years data (June 2007 – Oct 2011) from the registers of National RF / RHD Prevention and control programme from 32 hospitals of Nepal were collected and analyzed in this retrospective study. There were 6028 RF / RHD patients registered for secondary prevention. Out of them 672 had ARF. Only this group of patient was enrolled in our study. These patients were suffering from ARF and were receiving 3 weekly Benzathine Penicillin injections or oral antibiotic for secondary prevention of RF. Details of these patients were noted in a previously designed format. Details of age, sex and manifestations were collected and analyzed for the prevalence of SC.

Results

There were 672 patients with ARF in this study. Among them 374 (55.64%) were females and 298(44.34%) were males. 168(25%) were under 10 years of age, 402(59.8%) from 10-16 years and 102(15.2%) were above 16 years. Out of 672 a total of 26(3.87%) patients had chorea. Among them 20(76.92%) were females and 6(23.08%) were males. 7(26.9%) were below 10 years, 19(73.1%) 10 to 16 years and none were above 16 years age. (**Table 1**). In 16(2.31%) patients chorea was associated with Carditis, and in 6(0.89%) chorea was associated with Arthritis. Only 4(0.67%) patients had chorea as the only manifestation of ARF. The prevalence of SC was found to be 3.87% among patients with ARF.(**Table 2**)

Table 1 Age and sex of ARF patients with Sydenham's chorea

Age group		Sex	
<10yrs	7(26.9%)	Female	20(76.92%)
10-16 yrs	19(73.1%)	Male	6(23.08%)
>16yrs	0		
Total	26	Total	26

Table 2 Chorea as a major manifestations of ARF

Major manifestations	No. of patients
Chorea + carditis	16(2.31%)
Chorea + arthritis	6(0.89%)
Chorea alone	4(0.67%)
Total	26(3.87%)

Discussion

A major manifestation of AFR, SC is the result of an autoimmune response that occurs following infection by group A β -hemolytic streptococci that destroys cells in the corpus striatum of the basal ganglia.⁹⁻¹¹ The prevalence of ARF and Sydenham's chorea has declined progressively in developed countries over the last decades.^{12,13}

According to some published reports chorea is a major manifestation of ARF and is the only evidence of RF in approximately 20% of cases. In some outbreaks, chorea had been present in more than 30% of patients with ARF. The female- to -male ratio is approximately 2:1 and most patients present between 5-15 years of age. Studies have demonstrated a high frequency of a positive family history in the patients with SC and ARF. Arun et al found that 3.5% of patients and 2% of siblings of children with SC had been affected.¹⁴ SC usually develops in those aged 5-15 yrs. The patient may have no history of ARF and a preceding streptococcal infection cannot always be documented. Infections can be subclinical and often precede the development of neurologic symptoms by age 1-6 months. At least 25% of patients with SC fail to have serologic

evidence of prior infection. Chorea alone is sufficient for diagnosis of ARF provided other causes of the condition have been excluded.¹⁵

Our study has reported the prevalence of SC among patients with ARF to be 3.87% which is much lower than 5-38% reported by WHO.³ The reason behind the variation might be related to differences in susceptibility to chorea in the host population or to differences in case finding methods. It is unknown whether particular strains of group A streptococci vary in their propensity to elicit chorea. Why the prevalence of SC among ARF patients in Nepal is lower than in other countries is a subject for further studies. In our study female-to-male ratio was 3:1 which is slightly different than 2:1 reported in international literature.³ Patients of age group 10-16 yrs were mostly affected. This is similar to the reports of other published studies.

Conclusion

Prevalence of SC as the major manifestation in patients with ARF in Nepal is low and is reported to be 3.87% in our study. Females of age group 10-15 yrs are mostly affected with female-to male ratio 3:1. SC is the third common manifestation of ARF after arthritis and carditis. Chorea as the only manifestation of ARF is not uncommon but is mostly associated with carditis and arthritis in Nepalese population.

Acknowledgement

I acknowledge Mr. Ishwor Rayamajhi, a technical staff of Nepal Heart Foundation, central office who travelled throughout Nepal from east to west visiting all the 32 hospitals where RF/RHD registers are maintained and have collected data for this study.

References

1. National Institute of Neurological Disorders and Stroke (NINDS). NINDS Sydenham's chorea Information page. Accessed April 26, 2008.
2. Sydenham's chorea: Symptoms/ Findings from We MOVE. Org Accessed April 26,2008.
3. Bisno A. Non cardiac manifestations of rheumatic fever. In: Narula et al., eds. Rheumatic fever. Washington DC, American registry of pathology, 1994. p. 245-256.
4. Walker K, Lawrenson J, Wilmshurst JM. Sydenham's chorea – clinical and Therapeutic updates 320 years down the line. *SAMJ* 2006;96(9):906-12.
5. Shrestha UK, Bhattarai TN, Pandey MR. Prevalence of rheumatic fever and rheumatic heart disease in school children in a rural community of the hill region of Nepal. *Indian Heart J* 1991 Jan- Feb;43(1):39-41.
6. KC MB, Sharma D, Shrestha MP, et al. Prevalence of rheumatic and congenital heart disease in school children of Kathmandu valley in Nepal. *Indian Heart J* 2003;55:615-8.
7. Regmi PR, Pandey MR. Prevalence of RF and RHD in school children of Kathmandu city. *Indian Heart J* 1997 Sept-Oct;49(5):518-20.
8. Regmi PR, Upadhaya AB. RF and RHD prevention and control program in Nepal. *Nepalese Heart J* 2009;6(1):88-93.
9. Swedo SE, Leonard HL, Shapiro MB. Sydenham's chorea: Physical and Psychological symptoms of st vitus Dance. *Pediatrics*1993;91(4):706-13.
10. Sydenham's chorea symptoms. Accessed September 24,2009
11. Faustino PC, Terreri MT, da Rocha AJ et al. Clinical, laboratory, Psychiatric and Magnetic resonance findings in patients with Sydenham's chorea. *Neuroradiology* 2003;45(7):456-62.
12. Nausieda PA, Grossman BJ, Koller WC, et al. Sydenham's Chorea: An update. *Neurology* 1980;30(3):331-4.
13. Eshel E, Cahat E, Azizi E, et al. Chorea as a manifestation of rheumatic fever- a 30 year survey. *Eur J Pediatr* 1993;158(8):645-6.
14. Aron AM, Freeman JM, Carter S. The Natural History of Sydenham's chorea. Review of the Literature and long – term evaluation with emphasis on cardiac squeal. *Am J Med* 1965;38:83-95.
15. Special writing group of the AHA. Guidelines for the diagnosis of rheumatic fever. Jones criteria, 1992 updates. *JAMA* 1992;268(15):2069-73.