SUMMARY

AND

CONCLUSION

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Sydenham's chorea is a disease of childhood or early adolescence the age of primary affection was between 3.5-13 years. *(Aron et al 1965)* with average age of onset at 10.8 years *(Ben Hamida 1979).* According to Aron et al 1965, chorea is a disease of childhood characterized by the occurrance of rapid irregular aimless involuntary movements of extrimities, face and trunk. Most cases of chorea are preceded by streptococcal infection or rheumatic fever. However, the interval between the bacterial infection and the onset of neurologic symptoms is usually so long that serologic evidence for streptococcal infection was absent in 27% of children who had a chorea as the only clinical manifestation. *(Taranta and stollerman, 1985 and Manchanda and Ravi paul, 1968).* The appearance of chorea is conditioned by the presence of genes controlling the immune response gene (Ir.gene) which are beleived to be linked to the HLA chromosomal region *(Menkes, 1980).*

The aim of our study is to ascertain the freguency of HLA antigens among unrelated patients with rheumatic chorea to uncover the unknown insights in the pathogenesis and throw beems of light on the genetic control of such disease.

For this purpose we have chosen (30) patients, (21) females and (9) males ranged in age from (5-16) years. These patients subjected to :

* Full history taking.
* Thorough clinical examination.
* ESR, c-reactive protien and ASO.
* ECG and ECHO.
* HLA. typing according to NIH microcytolymphocytotoxi city technique (Bodmer - 1978):

(9) Alleles at A locus (A_1, A_2, A_3, A_9, A_{10}, A_{11}, Awlg, A_{28}, and A_{29})., (15) alleles at B- locus (B_5, B_8, B_{12}, B_{13}, B_{14}, B_{15}, B_{16}, B_{17}, B_{18}, B_{21}, B_{22}, B_{27}, B_{37} and B_{40}) and (6) alleles at DR-lous (DR_1, DR_2, DR_3, DR_4, DR_5 and DR_7) were tested.

We classified the patients into two groups :

1- Group I: (Isolated rheumatic chorea).
This group comprised (15) patients: (11) females and (4) males with their ages ranging from (5-16) years.

2- Group II: (chorea with carditis)
This group comprised (15) patients: (10) females and (5) males with their ages ranging from (6-13) years.

Our study cleared up the following results :

Among the patients with rheumatic chorea (total), the frequency of only B_5 and DR_2 were significantly high compared to controls. For B_5, antigen frequency was 56.6% while the relative risk was 6.11 and the etiologic fraction was 0.473, for DR_2, the antigen frequency was 53.3% while the relative risk was 4.39 and the etiologic fraction was 0.411. As there is no linkage disequilibrium between B_5 and DR_2, both of them have an important role in the pathogenesis of rheumatic chorea. Also, our study revealed that there was significant correlation between the occurrance of rheumatic chorea and the presence of B_5 and DR_2 reflecting the role of each allele in the development of chorea.

On studying the frequency of B_5 and DR_2 in patients with chorea associated with carditis and those with isolated chorea, we found that there was significant differance of the two alleles among the two groups.(Fisher exacts were 0.00001 and 0.00005) respectively.

Although the antigen frequency of HLA - B_5 is significantly high in the total patients (G_1 + G_2), it was found that the antigen frequency of B_5 is significantly high in the G_2 group of patients only (not G_1).

This suggests that the higher frequency of B_5 antigen in the total patients is related to the associated carditis with high acute phase reactants. This means that if any patient presenting

with Chorea and HLA - B_5 positive, Chorea is rheumatic in origin and the patient may have carditis or has a higher risk of development of carditis later.

Also, Although the antigen frequency of HLA - DR_2 is significantly high in the total patients (G_1 + G_2), it was found that the antigen frequency of DR_2 is significantly high in the G1 group of patients only (not G_2). This suggests that the higher frequency of DR_2 in the total patients is truly related to the isolated chorea and the G_1 group of patients are not rheumatic and will not develop carditis; also the pathogenesis of the isolated chorea is definitely different from that of chorea of rheumatic origin where the pathogenesis may be due to mild touch of encephalitis, autoimmune ...., allergic ... etc.

So far, follow up of patients with haplotype B_5 and / or DR_2 who developed chorea is recommended for probability of occurrance of carditis specially in cases of chorea with HLA-B_5 positive.