## ABSTRACT

**Objective:** To investigate the efficacy of carbamazepine in children with chorea.

**Patients and Method:** Six children (three boys, three girls, mean age 11.5 years) with the diagnosis of chorea were treated with carbamazepine (5-20 mg/kg/day). Response to the treatment was assessed based on improvement of hand-writing and Archimedes spiral test.

**Results:** Clinical improvement was observed in all children within the first two weeks. Chorea totally disappeared in four patients. In the other two patients chorea did not disappear totally, however markedly improvement was observed. No side effects were seen.

**Conclusion:** Carbamazepine is an efficient and safe drug in the treatment of chorea in children.

**Keywords:** Chorea, Carbamazepine, Childhood

---

## METHODS

Six patients (three boys, three girls) between the ages of 8-15 (mean 11.5 years) were diagnosed as having chorea and treated by CBZ between September 2000 and June 2005. The etiologies of chorea were as follows: Sydenham chorea (n: 4), dyskinetic cerebral palsy (n:1), basal ganglia infarction due to antiphospholipid syndrome (n:1).

The diagnosis of “Acute Rhematic Fever” was made on the basis of modified Jones criteria and all patients with Sydenham chorea had carditis. All of the children presented hemichorea except for the patient with dyskinetic syndrome. CBZ was used in two patients as the first line therapy, in four children.
who were unresponsive to haloperidole as a second choice drug. CBZ was started in 5mg/kg/day and the daily dose was increased until the involuntary movements seemed to decrease or disappear. Response to treatment was assessed based on improvement of hand-writing and Archimed spiral test as being mild or moderate improvement and complete remission.

RESULTS
Table I shows clinical findings and the patient responses to treatment. Clinical improvement was observed within the first two weeks of treatment in all patients and no side effects were seen. Chorea totally disappeared in four patients with Sydenham chorea, and the treatment was sustained between 7 – 14 months (mean 11.5).

Table I: Findings of patients treated with carbamazepine

<table>
<thead>
<tr>
<th>Patient No</th>
<th>Age/ Gender</th>
<th>Etiology of chorea</th>
<th>Neurological findings</th>
<th>CBZ dose max. mg/kg/d</th>
<th>Duration of treatment (month)</th>
<th>Recurrence</th>
<th>Following period (month)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>14/M</td>
<td>Stroke Antiphospholipid antibody syndrome</td>
<td>Right hemiparesis, right hemichorea</td>
<td>20</td>
<td>48</td>
<td>-</td>
<td>48</td>
</tr>
<tr>
<td>2</td>
<td>15/F</td>
<td>Sydenham chorea</td>
<td>Left hemichorea</td>
<td>10</td>
<td>14</td>
<td>-</td>
<td>36</td>
</tr>
<tr>
<td>3</td>
<td>8/M</td>
<td>Sydenham chorea</td>
<td>Left hemichorea</td>
<td>20</td>
<td>7</td>
<td>-</td>
<td>9</td>
</tr>
<tr>
<td>4</td>
<td>11/M</td>
<td>Sydenham chorea</td>
<td>Left hemichorea</td>
<td>20</td>
<td>7</td>
<td>-</td>
<td>9</td>
</tr>
<tr>
<td>5</td>
<td>14/F</td>
<td>Kernicterus, dyskinetic cerebral palsy</td>
<td>Dyscooordination, generalized choreathetosis, motor retardation</td>
<td>10</td>
<td>48</td>
<td>-</td>
<td>48</td>
</tr>
<tr>
<td>6</td>
<td>15/F</td>
<td>Sydenham chorea</td>
<td>Right hemichorea</td>
<td>7</td>
<td>12</td>
<td>-</td>
<td>17</td>
</tr>
</tbody>
</table>

Table II: Literature overview: Results of patients diagnosed as having chorea and treated with carbamazepine.

<table>
<thead>
<tr>
<th>Authors</th>
<th>n</th>
<th>Age (yr)/ Sex</th>
<th>Etiology</th>
<th>CBZ dose (daily)</th>
<th>Duration of treatment</th>
<th>Side effect</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ring et al (1988)</td>
<td>5</td>
<td>7-16/8F,3M</td>
<td>2 Sydenham C</td>
<td>4-10 mg/kg</td>
<td>3-6 months</td>
<td>Rash</td>
<td>-</td>
</tr>
<tr>
<td>Hare et al (2000)</td>
<td>10</td>
<td>7-16/8F,3M</td>
<td>9 Sydenham's C</td>
<td>5-10 mg/kg</td>
<td>1-15 months</td>
<td>-</td>
<td>2 cases</td>
</tr>
<tr>
<td>Green et al (2002)</td>
<td>17</td>
<td>5-14/19F,3M</td>
<td>14 Sydenham's C</td>
<td>3-10 mg/kg</td>
<td>1-15 months</td>
<td>-</td>
<td>3 cases</td>
</tr>
<tr>
<td>Perez et al (2002)</td>
<td>6</td>
<td>7-15/10F,3M</td>
<td>14 Sydenham's C</td>
<td>15-20 mg/kg</td>
<td>1-14 months</td>
<td>-</td>
<td>1 case</td>
</tr>
</tbody>
</table>

Abbreviations:
F = Female  M = Male
n = number of cases
Sydenham's C = Sydenham's Chorea
APS = Antiphospholipid antibody syndrome
SLE = Systemic lupus erythematosus
CP = Cerebral palsy
As soon as the symptoms were under control, the dose of CBZ was tapered slowly. In the other two patients, chorea did not disappear totally but markedly improved and the patients’ daily life quality concerning their capacity of using required motor function abilities (fine motor function) was better.

**DISCUSSION**

Neuroleptics (haloperidol, risperidone, fluphenazine) and antiepileptics (phenobarbital, clonazepam, valproate, carbamazepine) have often been used in the treatment of chorea. There are few studies regarding the clinical effects of these drugs, minimum effective doses, and therapeutic blood levels in childhood2-5. Table II shows the results of studies in the literature, demonstrating the effect of CBZ.

Harel et al. reported that clinical improvement occurred in children who were treated with low dose of CBZ (4-10 mg/kg/days) and blood levels of CBZ were under the required level for the treatment of epilepsy2. CBZ and valproate have been found equally effective and safe drugs in the treatment of Sydenham chorea3,5. The effective mechanism of carbamazepine to the basal ganglia is not obvious, but it is postulated that it could be through the blockage of dopaminergic postsynaptic receptors and through the stimulation of cholinergic pathways4.

Most of the reported children treated with CBZ were diagnosed as Sydenham chorea, and the duration of treatment in these patients was reported as lasting 1-15 months. Similarly, none of our four patients with Sydenham chorea were treated longer than 14 months. However, our two patients with permanent chorea are still under treatment for 48 months. Knowledge in the literature about the duration of treatment in children with different etiology is still insufficient.

Based on the results of previous studies and our own results, we conclude that carbamazepine (4-20mg /kg /day) is an efficient and safe drug in the treatment of chorea. Because of the wide range of effective dose, CBZ should be started clinically in minimum effective doses and the dose may be increased until a sufficient clinical response is achieved.

**REFERENCES**