Dancing Eyes Syndrome – Brainstem Acoustic Evoked Potential Approach

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Abstract
Three patients with dancing eyes syndrome of childhood are reported focussing on brainstem acoustic evoked potentials (BAEPs) recorded at different phases of the disease. In the first child in the acute phase BAEPs revealed pontine disturbance, which was less severe in a following milder attack. In the second child slight BAEP abnormalities were shown in the period of remission. In the case of the third child minimal pontine abnormalities were registered by BAEPs.

These electrophysiological findings, in accordance with the clinical features, suggest tegmento-pontine or pontocerebellar localisation of pathology in the dancing eyes syndrome.

Introduction
In 1962, Kinsbourne (12) reported cases of six children with opsoclonus, myoclonus, ataxia, dysynergia and irritable behaviour which produced a consistent and distinctive clinical picture unlike that seen in any other disorder. The term “dancing eyes and feet syndrome” was used first by Ford in 1966 (9). Several synonyms of this syndrome have been used later such as “infantile polymyoclonia” (7), “opsoclonus-myoclonus syndrome” (20), “syndrome of rapid irregular movements of eyes and limbs in childhood” (15).

In 1985, Talon (20) reviewed the literature of 110 cases. The syndrome occurs in association with viral infection, or with neuroblastoma in childhood. In adulthood it exists much more rarely in association with carcinoma or viral infection (6, 8, 13).

Postmortem studies have failed to localize the critical anatomic substrate(s) of the dancing eyes syndrome (DES) definitively. Several cases of childhood DES have been associated with no demonstrable pathology in the brainstem or cerebellum (1), however, in 1978 Cogan (4) found in other cases diffuse perivascular mononuclear-cell infiltrates in the thalamus, hypothalamus, basal ganglia, mesencephalon, pons. In 1970 Moe and Nelhaus (14) showed in some neuroblastoma-associated cases the cerebellum to be normal; Ziter et al (23) showed in other cases mild loss of Purkinje’s cells, peridental gliosis and also slight loss of myelin. In 1984, Graus et al (10) reported minimal perivascular infiltrates in the brainstem and in the meninges overlying the cerebellum. Ross and Zemann (16) found a decrease in succinate dehydrogenase activity in the dentate nucleus with perivascular infiltration in the pons, mesencephalon, hypothalamus in adults with DES associated with neoplasm.

The actual pathophysiological mechanisms causing DES with, or without an associated neoplasm is unclear, but most of the authors suppose an autoimmune process (2, 11, 17, 21).

In order to have an electrophysiological approach of the localisation in DES, we have studied the brainstem acoustic evoked potentials (BAEPs) in three girls with “idiopathic” DES in different phases to seek electrophysiological evidence of localisation of the disease process.

Method
BAEPs were recorded by the Madsen system. The auditory stimulus consisted of monoaural rarefaction clicks of 0.1 msec duration at the intensity of 0, 20, 40, 60, 80 dB above the mean threshold, as determined for people with normal hearing. Clicks were delivered at a rate of 12.8 per second through shielded headphones. The left and right ears were independently stimulated, and vertex-earlobe recordings were obtained. Signals were filtered at 150 and 3600 Hz. Analysis time was 10 msec. Two hundred samples were averaged. BAEPs were plotted on an X-Y recorder. BAEPs at 60 dB HL and 80 dB HL were analysed. Wave latencies from wave I to wave V interpeak latencies for I-II, I-III, III-V, I-V, and I/V amplitude ratio were measured. BAEPs were judged to be abnormal if one of these values was beyond 2.5 SD of the control mean. If an interwave separation was at or near the upper limit of normal, and the inter-ear interwave latency difference was abnormal, that segment was deemed to be abnormal (3).

Patients
Clinical symptoms of all the three patients were similar. The first patient was 15 months old, the second 21 months, and the third was four years old at the onset of the first symptoms,
in the case of the first child occurring five days after measles vaccination. In the second child the symptoms occurred with fever. The patients had opsonoclonus, ataxia, hypotonia, severe dyssynergia and asynchronous myoclonus in the muscles of the face, trunk and extremities. None of the children was able to stand or walk. They were excited and anxious. Neuroblastoma was excluded. CT scans of the brain were normal.

Cerebrospinal fluid

Patient 1: protein: 120 mg%, cell count: 12/3 (mononuclear cells), IgG/alb. ratio: 0.62, oligoclonal gammopathy.

Patient 2: protein: 33 mg%, cell count: 10/3 (mononuclear cells), IgG/alb. ratio: 0.98, oligoclonal gammopathy.

Patient 3: protein: 30 mg%, cell count: 320/3 (mononuclear cells), IgG increased, oligoclonal gammopathy.

Therapie

Each of the three patients was treated by corticosteroids (cortison, dexamethason) or by ACTH.

Outcome

Patient 1 has been closely followed for the past two and a half years, and had five relapses related to the withdrawal or reduction of corticosteroid dosage.

Patient 2 has been followed for two and a half years, and had four relapses related to upper respiratory or enteral viral infections.

Patient 3 has been followed over the past 14 years, had seven relapses during a five years' period of time, and when she reached 10 years of age, the condition resolved completely with mild residual fine motor dyscoordination and reduced IQ.

Results

BAEP values are shown on the Table 1a and b.

Patient 1

The first recording was performed at the time of the fourth relapse in the acute phase. The second recording was timed at the fifth relapse, when clinical symptoms were less severe (Fig. 1).

At the first time at 80 dB stimulus intensity on the left side I–V and III–V interpeak latencies were prolonged, the interpeak latency difference was prolonged. At 60 dB stimulus intensity on the left side waves III, V were delayed and I–III, I–V interpeak latencies were prolonged. At the second testing at 60 dB stimulus intensity on both sides wave III was delayed, and on the left side I–III, interpeak latency was prolonged, the inter-ear interpeak latency difference for I–V was abnormal with interpeak latency on the left side near the upper limit of normal.

Patient 2

BAEP recording was carried out after the patient’s fourth relapse in remission, when she had minimal dyssynergia, and sometimes brief opsonoclonic jerks (Fig. 2).

At 80 dB stimulus intensity wave IV on the left side, wave V on both sides was delayed, and interpeak latency of I–V was prolonged on both sides. At 60 dB stimulus intensity on the left side waves II, III, IV, V were at the upper limit of normal.
Dancing Eyes Syndrome

Table I

<table>
<thead>
<tr>
<th></th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>I-II</th>
<th>I-III</th>
<th>III-V</th>
<th>I-V</th>
<th>I/V</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control mean</td>
<td>1.69</td>
<td>2.79</td>
<td>3.71</td>
<td>4.83</td>
<td>5.42</td>
<td>1.03</td>
<td>2.03</td>
<td>1.61</td>
<td>3.65</td>
<td>74%</td>
</tr>
<tr>
<td>± SD</td>
<td>0.12</td>
<td>0.18</td>
<td>0.15</td>
<td>0.32</td>
<td>0.15</td>
<td>0.20</td>
<td>0.14</td>
<td>0.21</td>
<td>0.19</td>
<td>38%</td>
</tr>
<tr>
<td>II LD mean</td>
<td>0.15</td>
<td>0.11</td>
<td>0.09</td>
<td>0.10</td>
<td>0.14</td>
<td>0.07</td>
<td>0.09</td>
<td>0.06</td>
<td>0.09</td>
<td></td>
</tr>
<tr>
<td>± SD</td>
<td>0.14</td>
<td>0.07</td>
<td>0.06</td>
<td>0.09</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>R</td>
<td>1.85</td>
<td>2.4</td>
<td>3.9</td>
<td>5.8</td>
<td>0.55</td>
<td>2.05</td>
<td>1.9</td>
<td>3.95</td>
<td>14%</td>
<td></td>
</tr>
<tr>
<td>a. L</td>
<td>1.75</td>
<td>2.8</td>
<td>3.8</td>
<td>5.3</td>
<td>6.2*</td>
<td>2.05</td>
<td>2.4*</td>
<td>4.45*</td>
<td>62%</td>
<td></td>
</tr>
<tr>
<td>II LD</td>
<td>0.5*</td>
<td>0</td>
<td>0.5*</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

F.N.
- R
  - 1.7  2.5  4.0
  - 1.75 2.65 3.95
  - 1.7  2.8  3.9
  - 1.75 2.85 4.05
  - 1.7  2.75 3.9
  - 1.75 2.75 4.05

B.V.
- L
  - 1.75 2.85 4.95
  - 1.75 2.85 4.05
  - 1.7  2.8  4.0
  - 1.75 2.85 4.05
  - 1.7  2.8  4.0
  - 1.75 2.85 4.05

B.Zs.
- L
  - 1.85 2.75 3.70
  - 1.85 2.75 3.70
  - 1.85 2.75 3.70
  - 1.85 2.75 3.70

Note: Denote abnormal value beyond 2.5 SD of the control mean.

II LD: Interaural interwave latency difference

Table I b

<table>
<thead>
<tr>
<th></th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>I-II</th>
<th>I-III</th>
<th>III-V</th>
<th>I-V</th>
<th>I/V</th>
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<tbody>
<tr>
<td>Control mean</td>
<td>1.93</td>
<td>2.87</td>
<td>3.93</td>
<td>5.05</td>
<td>5.76</td>
<td>0.96</td>
<td>2.03</td>
<td>1.80</td>
<td>3.81</td>
<td>29%</td>
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<tr>
<td>± SD</td>
<td>0.15</td>
<td>0.18</td>
<td>0.13</td>
<td>0.33</td>
<td>0.26</td>
<td>0.20</td>
<td>0.14</td>
<td>0.20</td>
<td>0.22</td>
<td>15%</td>
</tr>
<tr>
<td>II LD mean</td>
<td>0.19</td>
<td>0.09</td>
<td>0.15</td>
<td>0.13</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>± SD</td>
<td>0.12</td>
<td>0.06</td>
<td>0.11</td>
<td>0.12</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>1.9</td>
<td>2.8</td>
<td>4.0</td>
<td>5.5</td>
<td>0.9</td>
<td>2.1</td>
<td>1.5</td>
<td>3.6</td>
<td>50%</td>
<td></td>
</tr>
<tr>
<td>a. L</td>
<td>1.75</td>
<td>2.85</td>
<td>4.9*</td>
<td>8.7*</td>
<td>1.1</td>
<td>3.15*</td>
<td>1.8</td>
<td>4.95*</td>
<td>15%</td>
<td></td>
</tr>
<tr>
<td>II LD</td>
<td>0.2</td>
<td>1.05*</td>
<td>0.3</td>
<td>1.35*</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

F.N.
- R
  - 2.05 2.8 4.5* 5.4
  - 1.9  2.95 4.7* 5.5
  - 2.05 3.2 4.25
  - 2.05 3.2 4.25
  - 2.15 3.2 4.1
  - 2.15 3.2 4.1

B.V.
- L
  - 2.1  3.3 4.3* 5.9*
  - 2.1  3.3 4.3* 5.9
  - 2.15 3.2 4.1
  - 2.15 3.2 4.1
  - 2.15 3.2 4.1
  - 2.15 3.2 4.1

B.Zs.
- L
  - 2.1  3.55* 4.15 5.2
  - 2.1  3.55* 4.15 5.2
  - 2.1  3.55* 4.15 5.2
  - 2.1  3.55* 4.15 5.2

Note: Denote abnormal value beyond 2.5 SD of the control mean.

II LD: Interaural interwave latency difference

Patient 3

Each of three patients investigated had BAEP abnormalities. The most severe abnormalities were found in the acute phase of the disease. The abnormalities were less marked in remission, and were minimal-borderline years after the recovery. BAEP abnormalities of this type can be found in patients with disorders affecting the pons, damaging the auditory pathways from the cochlear nuclei through the lateral lemniscus (18, 19).

In 1975, Dichtgans and Jung (5) suggested that opsoclonus is caused by a lesion of the cerebello-pontine integrators. In 1979, Zee and Robinson (22) explained it hypothetically by the instability of saccadic pulse-generators situated in the paramedian pontine formatio reticularis. In 1985, Leopold

Discussion

The localisation of the site of pathology in DES is not clear. Postmortem studies of patients with this syndrome have not given an uniform picture.
(13) stipulated that a local lesion in the brachium conjunctivum and/or in the cerebellar nuclei was the cause of the DES.

Our BAEP findings are in accordance with the clinical features and suggest that the lesions in our patients were located in the oral tegmental area of the pons affecting the lateral Lemniscus, brachium conjunctivum, probably together with some parts of the nearby lateral tegmental area of the pons and ponto-cerebellar connections.

Résumé
Trois maladies avec “dancing eyes syndrome” de l’enfance sont présentées centralisées aux réponses auditives du tronc cérébral, qui (PATC) ont été enregistrées aux phases différentes de la maladie. Chez le premier enfant à la phase aigue PATC signalèrent la dysfonction de la protubérance, qui devait moins grave au cours d’une attaque plus modérée suivante. Chez le deuxième enfant des anomalies douces de PATC ont été observées dans la rémission. Chez le troisième enfant une dysfonction minima de la protubérance était enregistrée à l’aide de PATC.

Ces résultats électrophysiologiques en accord avec les symptômes cliniques suggèrent une localisation protubérancielle (à la calotte) ou ponto-cérébelleuse de la pathologie dans le “dancing eyes syndrome”.

References