THE CERVICAL SPINE IN ATHETOID CEREBRAL PALSY

A RADIOLOGICAL STUDY OF 180 PATIENTS

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We have reviewed the cervical spine radiographs of 180 patients with athetoid cerebral palsy and compared them with those of 417 control subjects.

Disc degeneration occurred earlier and progressed more rapidly in the patients, with advanced disc degeneration in 51%, eight times the frequency in normal subjects. At the C3/4 and C4/5 levels, there was listhetic instability in 17% and 27% of the patients, respectively, again six and eight times more frequently than in the control subjects. Angular instability was seen, particularly at the C3/4, C4/5 and C5/6 levels.

We found a significantly higher incidence of narrowing of the cervical canal in the patients, notably at the C4 and C5 levels, where the average was 14.4 mm in the patients and 16.4 mm in normal subjects.

The combination of disc degeneration and listhetic instability with a narrow canal predisposes these patients to relatively rapid progression to a devastating neurological deficit.

Received 18 October 1995; Accepted after revision 11 January 1996

Patients with the athetoid type of cerebral palsy (athetoid CP) are considered to have a stable neurological condition, but many of them develop new functional deficits caused by cervical myelopathy and become progressively more disabled. These problems are crucial for the care of athetoid CP patients, but the disorders of the cervical spine are not widely recognised. Some patients may progress to a completely dependent state because of the lack of timely treatment.

Several papers discuss degenerative changes in the cervical spine (Anderson et al 1962; Ebara et al 1989, 1990) and surgery at this level in athetoid CP (Nishihara et al 1984; Fuji et al 1987), but no previous reports address the cause of the high incidence of cervical myelopathy in these patients.

We have made a radiological survey of the cervical spine in patients with athetoid CP and compared it with normal subjects to establish the characteristics and the factors leading to myelopathy.

PATIENTS AND METHODS

We invited 180 patients with a diagnosis of athetoid CP under treatment at five hospitals to participate in our study. We excluded all those who already had had cervical spine surgery. The mean age of the patients was 34.3 years (16 to 60); there were 102 men and 78 women (Table I).

We considered them in two groups: an operative group who had surgical treatment for cervical myelopathy during our study and a non-operative group who had neither cervical myelopathy nor any operation for a cervical spine problem. The operative group included 24 men and 19 women (53 patients) and the non-operative group included 78 men and 23 women (127 patients) (Table I).

Table I. The age distribution of the 180 patients with athetoid CP and the 417 control subjects

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Athetoid CP Operative</th>
<th>Non-operative</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 24</td>
<td>3</td>
<td>32</td>
<td>93</td>
</tr>
<tr>
<td>25 to 34</td>
<td>16</td>
<td>50</td>
<td>103</td>
</tr>
<tr>
<td>35 to 44</td>
<td>13</td>
<td>32</td>
<td>107</td>
</tr>
<tr>
<td>45 to 54</td>
<td>8</td>
<td>13</td>
<td>76</td>
</tr>
<tr>
<td>≥ 55</td>
<td>3</td>
<td>10</td>
<td>38</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td>137</td>
<td>417</td>
</tr>
</tbody>
</table>

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women; their mean age was 37.9 years (19 to 60). The non-operative group consisted of 78 men and 59 women; their mean age was 33.2 years (16 to 60).

Our control group was 417 patients who attended for conditions other than spinal problems. All gave fully informed written consent for carefully screened radiography of the cervical spine. Their mean age was 36.4 years (15 to 64) (Table I). There were 230 men and 187 women. The age and gender distribution of the patients and the control group was not significantly different.

We studied lateral radiographs in the neutral position and in maximum flexion and extension to detect disc degeneration, listhetic instability, angular instability and flattening of the vertebral body. We measured the sagittal diameter of the spinal canal. The CP patients were given 5 to 10 mg of diazepam orally about one to two hours before examination, and they were immobilised with a fixation device to the trunk which helped to obtain an exact lateral view of the cervical spine.

All data were evaluated and statistical analysis used the chi-squared or Student’s t-tests as appropriate. A p value of less than 0.05 was considered statistically significant.

Disc degeneration was assessed according to Lawrence (1969): grade 1 is slight anterior wear of the vertebral lip; grade 2 shows anterior osteophytes; grade 3 has both anterior osteophytes and narrowing of the disc space; and grade 4 is as grade 3 with sclerosis of the vertebral plates. The presence of a single osteophyte of less than 2 mm was recorded as a grade-1 change.

Listhetic instability was diagnosed when flexion or extension caused movement of greater than 3 mm, either anteriorly or posteriorly, between two adjacent vertebrae. At the atlantoaxial joint listhetic instability was recorded when the atlantodental interval (ADI) was over 3 mm (Hinck and Hopkins 1960).

Angular instability was recorded when excessive rotation in the sagittal plane was seen between two adjacent vertebrae during extension-flexion. Dvorak et al (1988), using Penning’s superimposing method (Penning 1978), reported that the maximum range (mean plus 2 sD) of sagittal rotation in healthy adults is 20° at C1/2, 15° at C2/3, 23° at C3/4, 26° at C4/5, 28° at C5/6, and 26° at C6/7.

The vertebral body ratio was recorded as the vertical diameter divided by the anteroposterior diameter of the vertebral body (Mochida, Hirabayashi and Satomi 1983). We calculated the mean value of the ratio minus 2 sD as the maximal flattening of normal vertebrae in the control group which showed no features of degenerative change or instability.

We defined the sagittal diameter of the spinal canal as the distance between the posterior border of the vertebral body and the closest point on the cortical line at the junction of the laminae and the spinous process which is in the midline of the spinal canal (Wolf, Khilnani and Malis 1956).

RESULTS

Incidence of disc degeneration. The incidence and grade of disc degeneration increased progressively with age in both men and women (Fig. 1). In male patients cervical disc degeneration of grade 1 or more was found in 67% of patients aged between 15 and 24 years, in 97% in those between 25 and 34 years and in all over this age. Disc degeneration of grades 3 or 4 was present in half of all the patients. Gender made no difference to prevalence. Disc degeneration of grades 3 or 4 was found in 65% of operated and 46% of non-operated cases, a statistically significant difference (Table II).

As expected, the control group showed a gradual increase in degeneration with age (Fig. 2) and again no significant gender difference. Disc degeneration of grades 3 to 4 was seen in only 6%.
Levels of cervical disc degeneration. In the patients, degeneration most often affected levels C3/4, C4/5, C5/6 and C6/7. In patients aged 15 to 34 years, the C5/6 disc was most frequently affected (Fig. 3). It seemed that degeneration started at the C5/6 disc level and progressed to other levels with increasing severity, eventually even affecting the C2/3 and C7/T1 levels (Fig. 4). In the control group, disc degeneration, usually of grades 1 or 2, was rare at C2/3, C3/4 or C7/T1 (see Fig. 3b).

The sites of onset of disc degeneration were similar, usually at the C4/5, C5/6 and C6/7 disc levels, in both the control group and in the athetoid CP patients (Fig. 5a). In five of the athetoid CP patients, however, disc degeneration started at C3/4 (Fig. 5b), a pattern shown by none of the
Instability. Listhetic instability of the atlantoaxial joint was not seen in control subjects, but was evident in 18 patients (Fig. 6), 11 in the operative group and 7 in the non-operative group. Three of these patients showed an os odontoideum and the incidence of C1/2 instability increased significantly with age (p = 0.0401).

At other levels, listhesis was found mainly at C3/4 and C4/5, with increasing frequency at C2/3 over 35 years of age. It was rare at the C6/7 and C7/T1 disc levels for all age groups (Table III). At the C3/4 level the incidence was six times that seen in the control subjects and at the C4/5 level eight times greater. Again, there was a statistically significant difference between operative and non-operative groups (Table II).

An increased incidence of angular instability was found at the C3/4, C4/5 and C5/6 disc levels in the patients (Table IV), and was very rare in the control group.

**Flattening.** At all levels of the cervical spine in all age groups, the vertebral body ratio in the athetoid CP patients and the control group showed a statistically significant increase in flattening (p < 0.0001; Table V), with no significant difference between the operative and non-operative groups.

**Sagittal diameter.** The cervical spinal canal in the athetoid CP patients at all levels was significantly narrower than that in the control group (Table VI). It was narrowest at C4 and C5, and narrower in the operative group.

**DISCUSSION**

Our findings show that patients with athetoid CP develop disc degeneration in the neck at a younger age and that it progresses rapidly and involves more disc levels than in normal subjects. Angular and listhetic instabilities also appeared at a younger age (Fig. 7) and were much more frequent. In patients with athetoid CP, an already narrow spinal canal is further compromised by osteophyte forma-
Table III. Incidence (percentage) of listhetic instability according to age and level in the athetoid CP patients and in the control group

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>C1/2</th>
<th>C2/3</th>
<th>C3/4</th>
<th>C4/5</th>
<th>C5/6</th>
<th>C6/7</th>
<th>C7/T1</th>
</tr>
</thead>
<tbody>
<tr>
<td>15 to 24 (n = 35/93)*</td>
<td>3/0*</td>
<td>3/0</td>
<td>9/0</td>
<td>34/0</td>
<td>6/0</td>
<td>0/0</td>
<td>0/0</td>
</tr>
<tr>
<td>25 to 34 (n = 66/103)</td>
<td>8/0</td>
<td>0/0</td>
<td>17/2</td>
<td>23/2</td>
<td>0/0</td>
<td>0/0</td>
<td>0/0</td>
</tr>
<tr>
<td>35 to 44 (n = 45/107)</td>
<td>9/0</td>
<td>7/0</td>
<td>18/2</td>
<td>24/6</td>
<td>4/2</td>
<td>0/0</td>
<td>0/0</td>
</tr>
<tr>
<td>45 to 54 (n = 21/76)</td>
<td>14/0</td>
<td>19/0</td>
<td>24/7</td>
<td>33/4</td>
<td>100/0</td>
<td>0/0</td>
<td>0/0</td>
</tr>
<tr>
<td>55 to 64 (n = 13/38)</td>
<td>38/0</td>
<td>15/0</td>
<td>23/8</td>
<td>31/8</td>
<td>15/0</td>
<td>18/0</td>
<td>0/0</td>
</tr>
</tbody>
</table>

* athetoid CP/control

Table IV. Incidence (percentage) of angular instability according to age and level in the athetoid CP patients and in the control group

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>C1/2</th>
<th>C2/3</th>
<th>C3/4</th>
<th>C4/5</th>
<th>C5/6</th>
<th>C6/7</th>
</tr>
</thead>
<tbody>
<tr>
<td>15 to 24 (n = 35/93)*</td>
<td>9/4*</td>
<td>17/1</td>
<td>11/0</td>
<td>31/1</td>
<td>12/3</td>
<td>11/0</td>
</tr>
<tr>
<td>25 to 34 (n = 66/103)</td>
<td>6/2</td>
<td>9/2</td>
<td>18/0</td>
<td>24/4</td>
<td>9/3</td>
<td>0/0</td>
</tr>
<tr>
<td>35 to 44 (n = 45/107)</td>
<td>13/4</td>
<td>0/1</td>
<td>11/1</td>
<td>20/1</td>
<td>17/0</td>
<td>0/0</td>
</tr>
<tr>
<td>45 to 54 (n = 21/76)</td>
<td>10/1</td>
<td>0/0</td>
<td>0/3</td>
<td>0/4</td>
<td>0/0</td>
<td>0/0</td>
</tr>
<tr>
<td>55 to 64 (n = 13/38)</td>
<td>0/0</td>
<td>0/0</td>
<td>0/0</td>
<td>0/5</td>
<td>0/0</td>
<td>0/0</td>
</tr>
</tbody>
</table>

* athetoid CP/control

Table V. Mean vertebral body ratio according to age and level in the athetoid CP patients and in the control group at all levels and in all age groups (p < 0.0001)

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>C3 CP</th>
<th>C4 CP</th>
<th>C5 CP</th>
<th>C6 CP</th>
<th>C7 CP</th>
</tr>
</thead>
<tbody>
<tr>
<td>15 to 24</td>
<td>0.651</td>
<td>0.807</td>
<td>0.679</td>
<td>0.805</td>
<td>0.669</td>
</tr>
<tr>
<td>25 to 34</td>
<td>0.647</td>
<td>0.796</td>
<td>0.659</td>
<td>0.772</td>
<td>0.649</td>
</tr>
<tr>
<td>35 to 44</td>
<td>0.618</td>
<td>0.752</td>
<td>0.602</td>
<td>0.750</td>
<td>0.579</td>
</tr>
<tr>
<td>45 to 54</td>
<td>0.563</td>
<td>0.724</td>
<td>0.565</td>
<td>0.709</td>
<td>0.589</td>
</tr>
<tr>
<td>55 to 64</td>
<td>0.522</td>
<td>0.714</td>
<td>0.526</td>
<td>0.719</td>
<td>0.469</td>
</tr>
</tbody>
</table>

Table VI. Mean sagittal diameter (mm; ± SD) of the spinal canal at all levels in the athetoid CP patients and in the control group

| C1 Control | 21.5 ± 2.6 | 19.0 ± 2.0 | 17.0 ± 2.1 | 14.9 ± 1.7 | 14.4 ± 1.7 | 16.4 ± 1.7 | 16.6 ± 1.7 | 16.6 ± 1.4 |
| Athetoid CP | 19.7 ± 2.6 | 17.1 ± 2.1 | 14.9 ± 1.7 | 14.4 ± 1.7 | 14.4 ± 1.6 | 14.8 ± 1.5 | 15.2 ± 1.5 | 15.0 ± 1.2 |
| Operative | 19.7 ± 2.9 | 16.3 ± 1.9 | 14.3 ± 1.7 | 13.7 ± 1.6 | 13.7 ± 1.3 | 14.4 ± 1.5 | 15.0 ± 1.5 | 15.3 ± 1.6 |
| Non-operative | 19.6 ± 2.3 | 17.4 ± 2.1 | 15.1 ± 1.7 | 14.6 ± 1.7 | 14.7 ± 1.6 | 15.0 ± 1.5 | 15.3 ± 1.6 |

* p < 0.0001
† p < 0.01
‡ p < 0.001
A 57-year-old woman with athetoid CP. The atlanto-dental interval is 7 mm and there is anterior listhesis of 4 mm at C3/4 and at C4/5.

Spinal degeneration is regarded as part of the normal ageing process (Lawrence 1969; Sasaki 1980; Kelsey 1982; Hayashi et al 1987), and the abnormal neck motion in athetoid CP patients accelerates this. Excessive neck motion may cause a ‘fatigue failure’ of the cervical discs. The resulting degeneration and instability may lead to rapid neurological deterioration.

Early surgical intervention can prevent irreversible neurological injury in these patients. We have used anterior decompression with fusion and laminoplasty with posterior fusion, usually from C3 to C7, in over 50 patients (Harada et al 1993,1994). There have been few problems in the early postoperative period after these stabilising procedures.

REFERENCES


