BENIGN PAROXYSMAL TORTICOLLIS OF INFANCY

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Benign paroxysmal torticollis is a self-limiting condition occurring during infancy. It resolves by the age of two to three years. Periodic episodes of torticollis may randomly alternate from side to side and be associated with other symptoms. The aetiology is unknown and no treatment is effective.

It is relatively uncommon, and has not been previously reported in the orthopaedic literature, although initial referral may well be to an orthopaedic surgeon. We report four cases, and review the literature.

Benign paroxysmal torticollis (BPT) is a self-limiting condition which presents in infancy and generally resolves spontaneously by two to three years of age. There are periodic episodes of torticollis, usually alternating from side to side, which may be associated with vomiting, ataxia, pallor or behavioural changes such as drowsiness or irritability (Hanukoglu, Somekh and Fried 1984). The aetiology remains unknown and no treatment is effective. It was first described by Snyder (1969) who reported 12 cases. Since then, less than 40 cases have been described mainly by paediatricians and neurologists including reports by Gourley 1971; Chutorian 1974; Guerrero et al 1988 and Ishida et al 1990: we could find no previous report in the orthopaedic literature. We present the clinical findings in four patients seen by orthopaedic surgeons at the Royal Children’s Hospital (RCH) between 1977 and 1989, and review the literature in order to bring the condition to the attention of orthopaedic surgeons.

PATIENTS AND METHODS

Between 1977 and 1989 the four patients were all carefully examined by the senior paediatric orthopaedic surgeon (MBM) and a paediatric neurologist. Audiological and ophthalmological examinations were performed and other investigations such as cervical spine radiographs, cerebral CT scans, and EEGs were also obtained when they were indicated. The diagnosis of BPT was made only when the clinical evaluation showed no other

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age at onset (mth)</th>
<th>Frequency (weeks)</th>
<th>Duration</th>
<th>Associated symptoms</th>
<th>At review</th>
<th>Free from episodes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>3</td>
<td>2 to 4</td>
<td>5 to 7 days</td>
<td>Ataxia, irritability</td>
<td>2</td>
<td>6 months</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>2</td>
<td>2</td>
<td>3 to 4 days</td>
<td>None</td>
<td>3</td>
<td>9 months</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>7</td>
<td>1 to 2</td>
<td>6 to 12 hours</td>
<td>Vomiting, ataxia, drowsiness</td>
<td>13</td>
<td>10 years</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>7</td>
<td>2 to 4</td>
<td>6 to 8 hours</td>
<td>Vomiting, ataxia, pallor</td>
<td>10</td>
<td>6.5 years</td>
</tr>
</tbody>
</table>

RESULTS

The findings in the four infants are summarised in Table I. All had normal neurological, audiological and ophthalmological tests. Two had EEGs; both were normal. One

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had a full metabolic investigation which was normal. One case is reported in detail to illustrate a typical clinical presentation.

**Case 1.** The patient was an 18-month-old girl born after an uncomplicated pregnancy by vaginal delivery. From three months of age her mother noticed that she would occasionally wake in the morning with her head tilted to one side. The torticollis did not seem to distress the baby, but was sometimes associated with slight ataxia and irritability. Massage and gentle manipulation by the parents failed to alter the torticollis. There were no apparent precipitating causes, the child was otherwise healthy and had no medication other than occasional paracetamol.

The episodes of torticollis occurred every two to four weeks and typically lasted about seven days; the head was tilted to right or left throughout each episode in an apparently random fashion (Figs 1 and 2).

The family history was unremarkable and physical examination, including detailed neurological examination, was entirely normal. There was no palpable mass in the sternocleidomastoid. Audiological and ophthalmological examinations were also completely normal.

The diagnosis of BPT of infancy was made and the patient was kept under review. The frequency of the episodes steadily decreased over six months, and there has been none for the past three months.

**DISCUSSION**

In most reported cases the torticollis was noted upon awakening in the morning (Hanukoglu et al 1984). The head is tilted to one side with the face slightly rotated in the opposite direction. Attacks last from hours to days and resolve completely, with no residual abnormality in the interim periods. Associated symptoms are common; three of our four patients had occasional vomiting and slight ataxia. The frequency of the episodes tends to decrease with age; they have usually ceased completely by the age of two or three years (Hanukoglu et al 1984).

We found no family history of similar episodes, although this has been reported (Lipson and Robertson 1978; Sanner and Bergström 1979; Deonna and Martin 1981; Roulet and Deonna 1988).

The aetiology of BPT remains obscure. In his original description, Snyder (1969) suggested a possible abnormality in the peripheral vestibular (labyrinthine) apparatus similar to that which is associated with the syndrome of benign paroxysmal vertigo in childhood (BPV). Dunn and Snyder (1976) proposed that BPT was merely a precursor of BPV based upon the fact that four of his original patients developed the latter condition as they became older. We found no evidence of an abnormality in the vestibular apparatus, although we did not perform caloric testing. None of our patients has, as yet, developed BPV.

Sanner and Bergström (1979) and Deonna and Martin (1981) considered that the abnormality was not in the peripheral vestibular apparatus but in the cerebellum or vestibulocerebellar connections; they thought that it might be vascular in origin. This theory was supported by Eeg-Olofsson et al (1982) who concluded that both BPT and BPV could possibly be precursors of recurrent migraine.

None of our patients had a family history of migraine and the two who have been under review for over ten
years have not developed migraine. As regards other aetiologies, we found no evidence of drug-induced dystonia (Casteels van Daele 1970, 1979, 1982) or ophthalmological abnormality (Rabb 1970) in any patient.

BPT of infancy is a benign disorder which may be initially referred to the orthopaedic surgeon (two of our four patients). Lack of familiarity with the condition may lead to a delay in diagnosis or unnecessary investigations. Where there is a typical clinical presentation, we recommend thorough orthopaedic and neurological examination with routine audiological and ophthalmological investigation. No treatment is indicated, but patients should be kept under review until the episodes have completely resolved.

We wish to thank L. K. Shield, FRACP, for his generous permission to include two of his patients in our study.

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REFERENCES