AXIAL MYOCLONUS OF PROPRIOSPINAL ORIGIN

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SUMMARY

Three patients are described with nonrhythmic repetitive axial myoclonic jerks causing symmetric flexion of the neck, trunk, hips and knees. No electrophysiological evidence of a cortical or brainstem reticular origin for the myoclonus was found. In the first patient the axial jerks only occurred spontaneously. The latencies of recruitment of spinal segments during a jerk indicated that the discharge arose in the mid thoracic cord and then slowly spread at about 5 ms\(^{-1}\) up and down the cord to involve rostral and caudal segments. No structural lesion was identified in this patient. In the second patient spontaneous and reflex axial jerks developed following the excision of a cervical haemangioblastoma. In the stimulus-induced jerks the relative latencies of muscles innervated by rostral and caudal spinal segments suggested that the myoclonus originated between the upper cervical and midthoracic cord. In the final patient, EMG activity during spontaneous and stimulus-induced jerks commenced in the rectus abdominis, and was followed by later activity in muscles innervated by rostral spinal segments, suggesting that the myoclonus originated in the midthoracic cord. No structural lesion was identified in this patient. Electrophysiological evidence is used to argue a spinal origin for these axial jerks in all 3 cases. There are striking features common both to this form of human myoclonus and to long propriospinal pathways identified in animals. The new concept of propriospinal myoclonus is suggested.

INTRODUCTION

Myoclonus may arise from generators in many sites within the nervous system. Cortical myoclonus is usually focal or multifocal and is due to discharges in the sensorimotor cerebral cortex which are propagated via corticomotoneuron pathways (Shibasaki et al., 1978; Hallett et al., 1979; Obeso et al., 1985). Brainstem reticular myoclonus causes generalized body jerks via slower conducting reticulospinal pathways (Hallett et al., 1977; Shibasaki et al., 1988). Brainstem generators may also activate cortical discharges (Thompson et al., 1989), possibly via the thalamus (Milhorat, 1967), to produce focal or multifocal jerks. In addition to these cerebral mechanisms generating myoclonus, focal or segmental myoclonus may also be caused by local spinal generators (Halliday, 1967), or lesions of the spinal roots (Sotaniemi, 1985), cervical or lumbosacral plexus (Banks et al., 1985), or peripheral nerves (Said and Bathien, 1977).

Spinal myoclonus, as described so far, typically has been confined to muscles innervated by a few adjacent spinal segments (hence the description as spinal segmental myoclonus), has been rhythmic and has often persisted in sleep (Halliday, 1967). Local spinal pathology, be it infectious (Ellis, 1920; Hopkins and Michael, 1974), inflammatory (Campbell and Garland, 1956), neoplastic (Garcin et al., 1968), traumatic (Lhermitte, 1919) or vascular (Davis et al., 1981), is believed to cause loss of local inhibitory spinal
interneuronal function, thereby liberating the spontaneous repetitive discharge of local segmental anterior horn cell pools to cause focal myoclonic jerks (Howell et al., 1979).

The spinal cord, however, possesses not only local segmental organization, but also powerful and complicated long propriospinal pathways linking activity in many segments. Abnormal activity in these propriospinal systems could, conceivably, generate a more complicated and more extensive form of spinal myoclonus. Here we present evidence from 3 patients for the existence of such propriospinal myoclonus in man. It is characterized by repetitive, usually flexor and nonrhythmic jerks of the neck, trunk, both hips and knees.

METHODS

Neurophysiological studies included routine electroencephalography (EEG), polymyography, back-averaging of the EEG activity preceding spontaneous jerks and somatosensory evoked potentials (SEPs). Electromyographic (EMG) recordings were made using bipolar silver/silver chloride electrodes placed 2 cm apart longitudinally over the muscle bellies. Those muscles most regularly involved and with well defined bursts of EMG activity in each jerk were selected to trigger the collection of EEG data for back-averaging. The same muscles were used to trigger the collection of EMG data from other muscles to define their order of recruitment during jerks. The latter was calculated from single trials rather than averaged data.

SEPs were recorded following stimulation of the tibial nerve at the ankle with a constant voltage rectangular pulse of 0.3 ms duration at intensities just above motor threshold. SEPs were the average of 128–256 stimuli and their amplitude was measured from the first major cortical positive peak following the stimulus (conventionally known as the P1, here termed P1) to the second major negative peak (the N2, here termed N2). Controls (14 healthy subjects, aged 18–40, mean 27 yrs) had a mean P1 latency of 40.4 ms (range 35.8–44.1 ms) and a mean P1N2 amplitude of 3.3 μV (range 0.5–11 μV) with stimulation of the tibial nerve.

The EEG was recorded from silver/silver chloride electrodes fixed to the scalp with collodion and referred to linked ear-lobe electrodes. The signals were band pass filtered at 2.5 kHz with a time constant of 1.0 s, and data were collected by a PDP 12 computer. The EEG recording sites were standard positions of the 10–20 system with additional electrodes used between these sites for the localization of focal cerebral activity, as necessary. In the EEG records shown, positivity is downward.

Magnetic stimulation of the cortex or spinal roots (Ugawa et al., 1989) was performed with a 9 cm diameter stimulator coil using a Novametrix Magstim 200. Spinal segmental lengths were calculated using data derived from postmortem examinations (Donaldson and Davis, 1903).

CASE HISTORIES

Case 1

A 47-yr-old man spontaneously developed involuntary generalized jerks of his body with flexion at the knees, hips, trunk and neck. The frequency and severity of the jerks increased when lying flat, but the jerks were evident on sitting or standing, and to a lesser extent when walking. They did not interfere with gait and did not make him fall. The jerks also occurred during sleep. They were not preceded by a build-up of ‘inner tension’ and could not be suppressed by an effort of will. The strength of the arms and legs was normal, and there was no sphincter or sensory disturbance. In the past the patient had been in good health with the exception of a 6 yr history of low lumbar back pain and right sciatica cured by a L5/S1 discectomy 3 yrs before the development of the truncal jerks.

Over the ensuing 2 yrs the jerks occurred every few minutes despite trials of baclofen 30 mg daily, sodium valproate 800 mg daily and milacemide 2400 mg daily. The combination of clonazepam 1 mg daily and temazepam 60 mg noxe was of modest benefit.

General examination early in his illness was normal. However, when seated or lying there were sudden simultaneous flexion movements of the neck, trunk, hips and knees. These could not be provoked by cutaneous stimuli or tendon taps. The jerks were most violent on lying flat when they would momentarily
bring the knees onto the chest. They were less frequent and less marked on sitting and standing, and did
not interrupt walking. The cranial nerves, muscle tone, tendon reflexes, abdominal reflexes and strength
were normal, but initially the plantar responses were extensor. Sensory examination, including over the
trunk and buttocks, was normal. These findings remained unchanged over the ensuing 4 yrs, with the
exception of the plantar responses which became flexor. Investigations including cerebrospinal fluid
examination, EEG and computerized tomography (CT) of the brain were normal. Myelography showed
a very small C5/6 disc protrusion without cord compression.

Case 2

A 40-yr-old man first noticed paraesthesiae in the fingers of the right hand. Over the next 3 yrs the
paraesthesiae spread to involve the right leg and right side of the face and head. He had a history of ankylosing
spondylitis and gout, but no relevant family history.

When first seen, 2 yrs after the onset of symptoms, abnormalities on examination were limited to brisk
upper limb tendon reflexes and reduced pin prick and temperature sensation in the right hand and leg.
Myelography showed an extensive cervical syrinx, extending from the craniovertebral junction to T4.
Vertebral angiography revealed an intradural mass at the level of C2. At subsequent operation a
haemangioblastoma was completely removed.

After the operation he developed brief involuntary flexion jerks involving the trunk and, if severe, the
neck, legs and right arm. These were more frequent on standing. The jerks were not preceded by any
build up of ‘inner tension’ and he could not control them. They partially improved when taking carbamazepine
800 mg daily. Six months later he developed extension jerks of the right knee, particularly when turning
the head to the left. Over the next 18 months he deteriorated still further with flexion jerks occurring every
few minutes and with progression of the right sided paraesthesiae.

Examination at this time revealed no evidence of retinal angioblastoma on fundoscopy and normal cranial
nerves. The right brachioradialis and both knee tendon jerks were brisk. The right plantar response was
extensor and there was hyperreflexia over the perineum and right side of the face. Pin prick sensation was
diminished below both knees. Magnetic resonance imaging (MRI) showed backward displacement and
expansion of the posterior aspect of the upper cervical cord but no syrinx (see fig. 5). Vertebral angiography showed no evidence of tumour recurrence and subsequent surgical exploration found and excised adhesions
between the posterior cervical cord and dura mater. After the operation there was marked reduction in
the frequency of both the generalized flexion jerks and the extension jerks of the right knee. The paraesthesiae
were unchanged.

The examination findings remained largely unchanged 1 yr after the surgical division of the upper cervical
adhesions. The spontaneous truncal flexion jerks occurred 5—15 times a day, but now, for the first time,
taps to both right and left biceps and patellar tendons elicited brisk, but small, axial flexion jerks.

Case 3

A 32-yr-old man spontaneously developed flexion jerks of his trunk which occurred only when lying
down and persisted during sleep. At times they were accompanied by noises due to the forced expiration
produced by vigorous abdominal contraction. There was no limb weakness, sensory, sexual or sphincter
disturbance. He was otherwise healthy with no personal or family history of significance.

General and neurological examination was entirely normal except that when lying down the patient
developed irregular shock-like contractions of axial muscles producing flexion of the trunk, neck and hips.
The jerks could be elicited by light touch over the anterior abdominal wall and by tapping the left rectus
abdominis with a tendon hammer. His condition remained unchanged over the ensuing 5 yrs. Medical
treatment was declined.

RESULTS

Case 1

Truncal jerks occurred irregularly every 20 to 60 s during polymyography in the supine position. They involved the sternocleidomastoid, rectus abdominis, quadriceps, hamstrings and to a lesser and more variable extent the gastrocnemius, tibialis anterior,
biceps and deltoid muscles. EMG bursts were small in erector spinae and pectoralis major. The duration of EMG bursts was variable ranging from 100 to 1400 ms in rectus abdominis. Homologous muscles were activated synchronously bilaterally and cocontraction was evident in agonist and antagonist muscle pairs (figs 1, 2).

![EMG Record](image)

**Fig. 1.** EMG record of a single spontaneous jerk in Case 1. Muscles are sternocleidomastoid, deltoid, biceps, rostral aspect of rectus abdominis, caudal aspect of rectus abdominis and quadriceps. Contraction of sternocleidomastoid, rectus abdominis and quadriceps occurred in all jerks, but activity in deltoid and biceps (evident here) was variable. Vertical calibration line = 200 μV and 100 μV for bottom and top 3 channels, respectively.

There was considerable variability in the relative latencies of individual muscles in the jerks (Table). Despite this, the jerks tended to show a characteristic electromyographic pattern (fig. 1). Within each jerk, the first EMG activity to be recorded was in the superior portion of rectus abdominis. EMG activity in the inferior portion of rectus abdominis...
**AXIAL MYOCLONUS**

FIG. 2. EMG record of a single spontaneous jerk in Case 1 showing cocontraction of agonist and antagonist muscles on both sides. Muscles are left (L) and right (R) quadriceps, tibialis anterior (TA) and gastrocnemius.

<table>
<thead>
<tr>
<th>Muscle</th>
<th>n</th>
<th>Mean (ms)</th>
<th>SD</th>
<th>Efferent delay (ms)</th>
<th>Spinal delay (ms)</th>
<th>Spinal root</th>
<th>Distance (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>L sternomastoid</td>
<td>51</td>
<td>35.2</td>
<td>17.1</td>
<td>5.2</td>
<td>39.9</td>
<td>C1, 2</td>
<td>21.2</td>
</tr>
<tr>
<td>L deltoid</td>
<td>19</td>
<td>37.2</td>
<td>4.8</td>
<td>6.5</td>
<td>40.6</td>
<td>C5, 6</td>
<td>16.9</td>
</tr>
<tr>
<td>L biceps</td>
<td>28</td>
<td>30.6</td>
<td>18.5</td>
<td>6.8</td>
<td>33.7</td>
<td>C5, 6</td>
<td>15.6</td>
</tr>
<tr>
<td>L superior rect.</td>
<td>49</td>
<td>2.9</td>
<td>9.1</td>
<td>10.3</td>
<td>2.5</td>
<td>T7</td>
<td>0.0</td>
</tr>
<tr>
<td>L middle rect.</td>
<td>34</td>
<td>8.9</td>
<td>18.7</td>
<td>9.9</td>
<td>8.9</td>
<td>T10</td>
<td>7.2</td>
</tr>
<tr>
<td>L inferior rect.</td>
<td>47</td>
<td>8.5</td>
<td>7.5</td>
<td>9.4</td>
<td>9.0</td>
<td>T12</td>
<td>11.4</td>
</tr>
<tr>
<td>R quadriceps</td>
<td>83</td>
<td>20.6</td>
<td>10.0</td>
<td>9.0</td>
<td>21.5</td>
<td>L3, 4</td>
<td>16.7</td>
</tr>
<tr>
<td>L quadriceps</td>
<td>83</td>
<td>21.9</td>
<td>10.0</td>
<td>9.0</td>
<td>22.8</td>
<td>L3, 4</td>
<td>16.7</td>
</tr>
<tr>
<td>L tibialis ant.</td>
<td>33</td>
<td>34.6</td>
<td>8.0</td>
<td>12.0</td>
<td>32.5</td>
<td>L4, 5</td>
<td>17.2</td>
</tr>
<tr>
<td>L hamstrings</td>
<td>20</td>
<td>50.5</td>
<td>16.1</td>
<td>9.9</td>
<td>50.5</td>
<td>S1</td>
<td>18.6</td>
</tr>
<tr>
<td>L gastrocnemius</td>
<td>34</td>
<td>46.9</td>
<td>9.3</td>
<td>12.0</td>
<td>44.8</td>
<td>S1, 2</td>
<td>19.4</td>
</tr>
</tbody>
</table>

* n = number of jerks. Mean = mean latency relative to the superior aspect of the right (R) rectus abdominis (Rect.); the SD is given to illustrate the jitter in the relative latencies. Efferent = peripheral efferent delay calculated from magnetic stimulation of spinal roots. Spinal delay = estimated extra time for the recruitment of spinal segments relative to T7 (the spinal segment supplying the superior portion of right rectus abdominis, the muscle with the earliest recorded activity in a jerk). Spinal delay for each muscle was calculated as: (latency relative to superior aspect of right rectus abdominis) minus (efferent conduction time to each muscle, less 9.9 ms which was the efferent conduction time to superior aspect of right rectus abdominis). Spinal root = principal segmental innervation of each muscle. Distance = distance of the principal spinal segments supplying each muscle from T7, calculated from postmortem data (Donaldson and Davis, 1903).
followed about 8.5 ms later ($P < 0.001$). EMG activity was then recorded in muscles both cranial and caudal to rectus abdominis. The latencies of these muscles (relative to the superior portion of rectus abdominis) increased with their distance from the superior portion of rectus abdominis both up and down the body.

It was possible to estimate the conduction velocity of the spinal efferent pathways involved in this form of myoclonus from the relative latencies of the individual muscles (Table). Peripheral efferent times were calculated for each muscle using magnetic stimulation of nerve roots, the coil being placed over the appropriate spinal segment. These peripheral efferent times, less 9.9 ms (the peripheral efferent time for the superior portion of right rectus abdominis), were then subtracted from the delay in onset of EMG activity in each muscle relative to the superior portion of rectus abdominis, to give the order of recruitment (spinal delay) of the spinal segments innervating individual muscles involved in the jerks. When the spinal delay for each muscle was considered as a function of the distance of its innervating segment along the cord from T7 (the segment innervating the superior portion of rectus abdominis) a highly significant positive correlation was found ($r = 0.882, P = 0.0001$). The slope of the regression line was consistent with a conduction velocity of about $5 \text{ ms}^{-1}$ in spinal efferent pathways leading up and down the cord from T7. This calculation of the conduction velocity was only approximate as it was based on average segmental lengths in postmortem specimens (Donaldson and Davis, 1903) and included delays at both root and synaptic levels. The latter will include the time necessary for temporal facilitation, which is likely to increase with increasing distance from the spinal myoclonic generator, leading to a further underestimation of the true conduction velocity in the spinal efferent pathways involved in the myoclonus. (A similar calculation was made using an estimate of the spinal delay derived from the minimum relative latencies of each muscle. There was a significant correlation between this minimum spinal delay and the distance of segmental innervations from the midthoracic spine ($r = 0.710, P = 0.01$). The slope of the regression line was consistent with a conduction velocity of about $6.5 \text{ ms}^{-1}$ in the spinal efferent pathways involved in the myoclonus.)

The effect of the jerks on tonic voluntary EMG was investigated, the patient being instructed to keep the right leg relaxed and the left leg extended in the air (producing a moderately steady EMG activity 10–20% of maximum). In the left tibialis anterior, tonic voluntary EMG activity was reduced for 150 (SEM ±24) ms starting 35 (±9.6) ms before the EMG burst in the right tibialis anterior, and in the left quadriceps tonic EMG activity was reduced for 72 (±7) ms starting at the same time as the EMG burst in the right quadriceps (fig. 3).

There was no evidence that the jerks were stimulus sensitive. Cortical SEPs from stimulation of the right and left posterior tibial nerves at the ankle were of normal latency ($P$, at 41 and 42 ms, respectively), and amplitude ($P_{\text{N}}$, 2.8 and 1.3 $\mu$V, respectively). Cutaneous muscular reflexes to a train of stimuli delivered to the right hallux habituated normally. Back-averaging the EEG activity in the 125 ms before each jerk did not reveal any short latency time-locked cortical activity. Magnetic stimulation of the cortex produced EMG responses of normal latency in biceps (10 ms), rectus abdominis (20 ms) and quadriceps (22 ms).

Finally, the possibility that the jerks were voluntary was investigated. Back-averaging
the EEG activity in the 2 s before each spontaneous jerk did not reveal any premovement potential, although a normal premovement potential was recorded before voluntary trunk flexion (fig. 4). When normal subjects were asked to mimic the truncal flexion jerks they exhibited a very different picture of muscle activation to those in Case 1. The muscle activity in agonists and antagonists tended to alternate as seen in normal ballistic limb movements. In addition, unlike Case 1, the superior and inferior aspects of the abdominal recti contracted synchronously. Finally in mimicked jerks, EMG activity was not usually recorded first in the rectus abdominis, and no significant correlation was found between the spinal delay of various muscles and the distance of the respective innervating spinal segments from T7.

**Conclusion.** The jerks were involuntary. No evidence for a supraspinal origin for the myoclonus was found. The recruitment of muscles within the jerks was determined by the distance of their segmental supply from T7, suggesting a spinal myoclonic generator at about this level.

**Case 2**

The first opportunity to examine this patient electrophysiologically occurred the day before his surgical reexploration. He was treated preoperatively with dexamethasone 4 mg daily which had greatly reduced jerk frequency so that polymyography (fig. 6A) only recorded 4 jerks. These occurred irregularly over a period of 1 h and involved rectus abdominis, erector spinae and usually quadriceps and tibialis anterior synchronously.
Fig. 4. Back-averaging of EEG activity over 2 s preceding voluntary truncal and hip flexion movements mimicking the spontaneous myoclonic jerks (left, average of 88 recordings) and spontaneous jerks (right, average of 100 recordings) in Case 1. A Bereitschaftspotential is evident preceding the voluntary movements, but not the spontaneous jerks. The postmovement potentials are probably caused by movement artefact. The sweep time is 2.5 s.

Fig. 5. MRI of the cervical cord in Case 2 (sagittal spin echo image with gadolinium enhancement), showing backward displacement and expansion of the posterior aspect of the upper cervical cord.
on both sides. Bursts of muscle activity lasted 40–200 ms, with cocontraction of the abdominal recti and erector spinae. There was no stimulus sensitivity at this initial examination. Cortical SEPs from stimulation of the tibial nerves at both the right and left ankles were delayed (P1 at 46 and 49 ms on right and left tibial nerve stimulation, respectively), but of normal amplitude (P1N2 of 0.6 and 0.5 μV, respectively). Back-averaging was precluded by the low frequency of the jerks. Magnetic stimulation of the cortex elicited responses of normal latency in the right tibialis anterior (31 ms) and right quadriceps (28 ms). No responses could be obtained on the left.

The patient was reexamined electrophysiologically 1 yr later. At this time taps to both biceps and quadriceps tendons elicited brief axial flexion jerks. During these, EMG activity was recorded in sternocleidomastoid, rectus abdominis, quadriceps and often in pectoralis major. There was considerable jitter in the relative latencies of muscles involved in the jerks. Despite this jitter, EMG activity was recorded first in rectus abdominis (137.9±5.8 ms, n = 18), and significantly later in sternocleidomastoid (148.8±3.7 ms, n = 16, P = 0.014) and quadriceps (149.8±4.3 ms, n = 13, P = 0.022) following taps to the left patellar tendon (fig. 6B). The duration of EMG activity during these reflex jerks was brief (50–200 ms).

Conclusion. The patient developed spontaneous axial myoclonic jerks, very similar to those seen in Case 1, following the excision of a cervical haemangioblastoma. The myoclonus worsened with subsequent distortion of the cervical cord by dural adhesions seen on MRI and confirmed during surgical exploration. The spontaneous jerks greatly improved following division of the adhesions. In the reflex jerks that developed after the second operation, the EMG activity was recorded first in the abdominal recti, and later in more rostral and caudal muscles. We conclude, therefore, that the jerks were spinal in origin.
Case 3

During polymyography, jerks occurred irregularly every 20—50 s. There was bilateral and relatively synchronous activation of thoracic and lumbar paraspinal muscles, rectus abdominis, pectoralis major, deltoid and, less often, quadriceps and hamstrings. The duration of EMG bursts was variable, ranging from 100 to 400 ms in rectus abdominis.

In both spontaneous and reflex myoclonus there was considerable jitter in the relative latencies of muscles involved in the jerks. Thus, in spontaneous jerks, EMG activity in the lumbar paraspinal muscles occurred from 12 to 44 ms (mean 26.2 ms) after that in the abdominal recti. Despite this jitter, EMG activity was always recorded first in the abdominal recti (fig. 7). The EMG activity in rostral muscles occurred later (the

![Diagram of EMG activity](https://example.com/diagram)

**Fig. 7.** EMG of a single spontaneous jerk in Case 3. The first muscles to contract are the abdominal recti.

mean (± SEM) latencies of pectoralis major and deltoid relative to rectus abdominis in 22 jerks were 19.8 ± 2.6 ms and 30.2 ± 1.8 ms, respectively. Too few spontaneous jerks involving the lower limb muscles were recorded to comment on the relative latencies of these muscles.

Single taps with a tendon hammer to the anterior abdominal wall elicited several bursts of activity in the abdominal recti. Taps to the left abdomen were unique in eliciting an early burst of EMG activity in the left rectus abdominis at 33.0 ± 1.9 ms, n = 9
AXIAL MYOCLONUS

(fig. 8). This was separate from the earliest burst of activity seen at about 19 ms in some trials (not evident in fig. 8), which represented the tendon reflex in this muscle.

In addition, taps to either the right or left abdomen elicited a series of decreasing bursts of activity in both abdominal recti at 106.0 ± 1.8, 206.7 ± 2.4, 287.8 ± 4.2 and 360.0 ± 6.0 ms. The duration of the first EMG burst in this series was 70.8 ± 2.7 ms (n = 15).

Taps with a tendon hammer to either the right or left patellar tendon elicited single bursts of activity in the abdominal recti (at 113.0 ± 5.3 ms, n = 12 for taps to the right patella), and significantly later in the right pectoralis major (at 127.6 ± 5.7 ms, n = 8 for taps to the right patella, P < 0.001). The difference between the latency of right pectoralis major and rectus abdominis in spontaneous jerks did not differ significantly from that in jerks elicited by taps to either patellar tendon. The duration of the burst of activity in rectus abdominis to taps to the right patellar tendon was 57.2 ± 3.8 ms (n = 12). Reflex jerks did not involve the lower limbs.

The cortical SEP from stimulation of the right tibial nerve was of normal amplitude (P1N2 = 8 μV) and latency (P1 at 41 ms).

Conclusion. A spinal myoclonic generator seems highly probable in Case 3. In both spontaneous and reflex jerks, EMG activity was recorded first in the abdominal recti, and later in more rostral muscles. Although the most marked reflex responses were, as in Case 2, of long latency, comparable (Roby-Brami and Bussel, 1987) or longer.
(Shahani and Young, 1971) spinal reflex latencies have been reported in patients with cord transections. In addition, taps to the left abdomen elicited a response in the left rectus abdominis at 33 ms. Such a short latency response (only 14 ms longer than the probable monosynaptic tendon reflex in the abdominal recti) is incompatible with a cortical reflex loop. Taps to the abdomen may elicit an abdominal skin reflex (Kugelberg and Hagbarth, 1958), but the latter usually has a longer (Duensing, 1940) and much more variable (Kugelberg and Hagbarth, 1958) latency than the reflex elicited in Case 3. In addition, the abdominal skin reflex is readily elicited by electrical stimulation of the skin (Kugelberg and Hagbarth, 1958). The early response in the left rectus abdominis was absent on electrical stimulation of the abdomen.

DISCUSSION

Clinical and electrophysiological features

Three cases with an unusual, distinctive and clinically similar movement disorder are presented. Each exhibited jerks resulting in shock-like flexion at the trunk, hips and often knees and neck. The jerks developed in middle age. In Case 2 jerks developed following excision of a cervical haemangioblastoma and subsequent arachnoiditis, and improved following the surgical division of dural adhesions. In the other 2 cases no definite cause was identified. In these patients no other neurological abnormalities developed except for the bilateral extensor plantar responses present in Case 1 on first presentation alone. The course in these 2 patients was relatively benign, with the myoclonus remaining unchanged in nature for up to 5 yrs. All the patients were active and independent, and only 1 was unable to work (Case 1).

The jerks were very similar in all 3 patients. They consisted of irregular symmetric, relatively synchronous bursts of muscle activity occurring up to twice per second, with a duration varying between 40 ms and 2 s in the abdominal recti, lumbar and thoracic paraspinous muscles, quadriceps, hamstrings and often in the sternocleidomastoids, biceps, deltoids, glutei, tibialis anterior and gastrocnemius. The contraction of flexor muscles, particularly the abdominal recti, dominated. As a result a typical jerk caused flexion of the trunk and often also of the neck, hips and knees. Jerks were most marked on lying down in 2 patients (Cases 1, 3) and in this position the jerks produced a violent movement bringing the flexed knees towards the chest. The jerks often occurred in sleep. In 2 of the 3 patients (Cases 2, 3), the jerks were stimulus sensitive.

Differential diagnosis

The jerks were myoclonic in appearance. The jerks were involuntary and were not preceded by a premovement potential in Case 1. In addition, during the jerks there was cocontraction of agonist and antagonist muscles (e.g., the abdominal recti and paraspinal muscles) in Cases 1, 2 and 3. This was quite unlike the alternating contraction seen in these muscles when normal subjects voluntarily mimicked such movements. Accordingly, we concluded that this myoclonus was real and not psychogenic.

At no time did any of the 3 patients exhibit any sustained abnormal postures that might have suggested that the jerks were part of a dystonic syndrome (Obeso et al., 1983).
Nor did they have any evidence of dystonia elsewhere. Axial torsion dystonia can produce flexor or extensor spasms of the trunk, but these typically appear on action, especially on walking, and decrease or disappear when lying flat or relaxed. The converse was the case in 2 of the 3 patients described here.

On clinical grounds the jerks were also unlikely to represent tics. They did not vary or migrate to involve other parts of the body, they were not preceded by any of the subjective feelings associated with tics, and they were not suppressible (Jankovic, 1987). There were no vocalizations or behavioural disturbances. The absence of any family history and the onset in middle-life would also be unusual. Stimulus sensitivity is not a feature of dystonia or tic disorders.

For these reasons we conclude that these 3 patients had a peculiar type of axial myoclonus. The question then is where did it come from? There was nothing to suggest a cortical (Shibasaki et al., 1978; Hallett et al., 1979; Obeso et al., 1985), subcortical (Thompson et al., 1989) or reticular origin (Hallett et al., 1977) for the myoclonic jerks. EEG and cortical SEP amplitudes were normal. Back-averaging of the EEG activity preceding the jerks did not reveal any cortical correlates. None of the patients exhibited any specific clinical signs of cerebral cortical or brainstem dysfunction. The duration of the EMG bursts responsible for the jerks was often much longer than that seen in cortical (Hallett et al., 1979; Obeso et al., 1985), subcortical (Thompson et al., 1989) or brainstem reticular (Hallett et al., 1977) myoclonus. Thus there was no evidence for a supraspinal origin for this myoclonus.

A spinal origin for the myoclonus

The features of all 3 cases are, however, consistent with a spinal origin of the jerks. In Case 1 the order of recruitment of muscles involved in the jerks was determined by the distance of their segmental supplies from T7, suggesting a myoclonic generator at about this level. In Case 2 jerks followed excision of a cervical haemangioblastoma and worsened with subsequent distortion of the cervical cord by dural adhesions seen on MRI and confirmed during surgical exploration. Jerks diminished following the division of the adhesions. The difference in latency between quadriceps and the middle portion of rectus abdominis in the stimulus-sensitive jerks (11.9 ms) was similar to that seen in the spontaneous jerks in Case 1 (13.0 ms). In contrast, the difference in latency between sternocleidomastoid and rectus abdominis (10.9 ms) was shorter than in Case 1 (44.1 ms), suggesting a more rostrally placed spinal myoclonic generator in Case 2. In Case 3 EMG activity was recorded first in the abdominal recti in both spontaneous and reflex jerks. The differences in latency between rectus abdominis and more rostral muscles were similar to those recorded in Case 1, and suggest a spinal origin for the myoclonus near the midthoracic cord. In addition there was a response in the left abdominal rectus to abdominal taps at a very short latency. The latter was incompatible with a transcortical reflex loop.

Hitherto, myoclonus of spinal origin has been described as focal (Halliday, 1967; Jankovic and Pardo, 1986), consisting of synchronous rhythmic jerks with a fairly constant frequency ranging from 20 to 180 per min, and confined to a group of muscles supplied by either one segment or several contiguous segments of the spinal cord (Halliday, 1967). Bursts of EMG activity may be relatively long (Hopkins and Michael, 1974; Davis et al., 1981). Various spinal pathologies may give rise to segmental myoclonus (Lhermitte,
The myoclonus described here was quite different from simple spinal segmental myoclonus. The axial flexion jerks were nonrhythmic and involved the recruitment of muscles innervated by spinal segments from the upper cervical cord to the lower lumbosacral cord. We suggest that this form of myoclonus arose in propriospinal systems.

**Propriospinal myoclonus**

The relatively stereotyped pattern of muscle involvement seen in the 3 patients suggests that the generalized axial jerks were subserved in each case by a similar pathway. In Case 1 the estimated conduction velocity within the spinal cord was slow (5.0–6.5 m·s⁻¹). The differences in relative latencies between rectus abdominis and quadriceps (11.8 ms) in the stimulus-induced jerks in Case 2, and between rectus abdominis and deltoid (30.2 ms) in the spontaneous jerks in Case 3, were long and also suggest a slow spinal conduction velocity. In contrast, the conduction velocity in spinal efferent pathways is rapid in those forms of myoclonus with a supraspinal origin (Hallett et al., 1977, 1979; Obeso et al., 1985; Thompson et al., 1989).

The slow spinal conduction velocity and the large jitter in latencies suggests a slowly conducting polysynaptic pathway. It is our hypothesis that these jerks arose in one or more adjacent spinal segments and then spread up and down the cord to other segments via propriospinal pathways.

There has been relatively little work on the propriospinal system in man. However, Giok (1958), studying the pattern of fibre degeneration in cases of ventrolateral cordotomy, concluded that a descending spinospinal fibre system exists in the ventrolateral funiculus, and there is some electrophysiological evidence to suggest the existence of both short (Malmgren and Pierrot-Deseilligny, 1988a, b) and long (Kearney and Chan, 1979; Meinck and Piesiur-Strehlow, 1981) propriospinal pathways in man. Meinck and Piesiur-Strehlow (1981), investigating a long-loop reflex from the rostral trunk and arms to lumbosacral motoneurons in healthy subjects, found a decrease in latency when more caudal segments were stimulated, and proposed that these reflexes were mediated by directly descending propriospinal pathways in man. In their study the difference in mean latency of the facilitation of the H reflex in quadriceps and gastrocnemius following ulnar nerve stimulation was 10 ms. Allowing for the shorter peripheral efferent time for quadriceps gives a 5 ms delay for central conduction from the L3/4 to the S1/2 segmental level, a distance of approximately 2.7 cm (Donaldson and Davis, 1903). The derived estimate of spinal conduction velocity in these propriospinal pathways in healthy subjects is slow (5.4 m·s⁻¹) and compares favourably with the calculated conduction velocity in Case 1. The presence of inhibition at, or just preceding the onset of a jerk, as seen in the tonic EMG activity in Case 1, is also a feature shared by long propriospinal reflexes in man (Meinck and Piesiur-Strehlow, 1981).

Long propriospinal systems in the cat have been studied in more detail, and again bear close similarities to the generalized axial jerks. Long propriospinal pathways in the cat may originate in the cervical (Lloyd, 1942; Baldissera et al., 1972; Jankowska et al., 1974; Vasilenko, 1975) or midthoracic (Baldissera et al., 1972) segments and project to lumbar segments, or they may originate in lumbar segments and project rostrally (Gernandt and Megirian, 1961; Barilari and Kuypers, 1969; Miller et al., 1973). They
are slowly conducting and may be polysynaptic (Lloyd, 1942; Vasilenko, 1975). In
the 2 patients in whom jerks developed spontaneously, jerks were strikingly dependent
on posture. In the cat, long cervical propriospinal neurons receive a very large vestibular
input (Alstermark et al., 1987a) as well as input from neck and limb afferents (Alstermark
et al., 1987b).

In all 3 cases the jerks involved axial and proximal limb muscles bilaterally, with
cocoonaction in antagonist and agonist muscle pairs. Similarly, the long propriospinal
systems defined in the cat recruit mainly trunk (Baldissera et al., 1972) and proximal
limb (Vasilenko, 1975) motoneurones bilaterally (Barilari and Kuypers, 1969). Long
propriospinal systems in the cat (Lloyd, 1942; Vasilenko, 1975) and man (Meinck and
Piesier-Streelow, 1981) also share a lack of reciprocal action on antagonists. The
functional importance of long spinospinal pathways principally innervating proximal
muscle groups, without reciprocal organization, seems obscure until it is remembered
that, in the cat at least, the spinal cord itself can generate forelimb locomotor movements
coordinated with those of the hindlimbs (Miller and Van der Meche, 1976). Long
propriospinal pathways are particularly suited to a role in interlimb coordination as the
pattern of response generated by propriospinal projections varies systematically with
limb position (Grillner and Rossignol, 1978) in static situations, and with the phase
of the step cycle during locomotion (Forssberg et al., 1977; Miller et al., 1977).

Finally, an experimental model of propriospinally mediated myoclonus exists in the
cat, where spinospinal pathways are thought to mediate the development of generalized
rhythmic myoclonus following the inoculation of Newcastle disease virus into the lumbar,
midthoracic or cervical cord, since the myoclonus in segments distal to the site of
inoculation is abolished by thoracic spinal cord transection (Luttrell et al., 1959). The
myoclonus in this animal model persists after high cervical cord transection (Luttrell
et al., 1959).

Conclusion

Here we present 3 cases of axial jerking resulting in flexion at the neck, trunk, hips
and knees, and suggest that the common clinical features of this new type of myoclonus
are determined by the involvement of long propriospinal pathways in man.

It has always seemed likely that, because of the complex organization of its
propriospinal systems, the spinal cord may give rise not only to simple focal segmental
myoclonus, but also to a more generalized and complex form of myoclonus. Bussel
et al. (1988) recently reported a patient with a cervical cord transection who developed
rhythmic generalized extensor jerks, with relatively long EMG bursts, below the level
of transection. Our 3 cases exhibited clinically similar generalized flexion jerks, again
with relatively long EMG bursts in spontaneous jerks, and there was strong
electrophysiological and pathological evidence for a spinal origin for the myoclonus
in these patients. In addition to the 3 cases described here, and studied neuro-
physiologically, we have seen 3 other patients with clinically similar unexplained truncal
flexor jerks, who may have a similar condition. Bressman and Fahn (1986) have also
described 2 other patients (Cases 11, 12) whose clinical features are remarkably similar.

There is therefore an argument for expanding the present concept of spinal myoclonus
to include two types: first, simple spinal segmental myoclonus, with focal repetitive
rhythmic jerks confined to one or more adjacent spinal segments; and secondly,
propriospinal myoclonus, with predominantly axial and often arhythmic flexor or extensor (Bussel et al., 1988) jerks involving many spinal segments linked by long propriospinal pathways.

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AXIAL MYOCLONUS 213


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