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Spina bifida occulta. Foot deformities, enuresis and vertebral cleft: clinical picture and neurophysiological assessment

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Aim. **The aim of the study was to investigate the relationship between the clinical evidence of foot deformities in spina bifida occulta and the associated neurophysio-logical damage.**

Methods. **The authors studied 47 patients with foot deformities (37 flat foot, 10 pes cavus) and vertebral cleft, variably associated with enuresis, midline cutaneous lesions, and further orthopaedic deformities. An electrophysiological evaluation was performed in an attempt to investigate the peripheral nervous system in greater detail, including conventional motor and sensory nerve conduction, F-wave recording and electromyogram (EMG) testing.**

Results. **The peroneal nerve F wave latency was longer in patients with pes cavus than in those with flat foot (P<0.04). Conversely, the posterior tibial nerve F-wave latency was longer in patients with flat foot than in those with pes cavus (P<0.02). Needle EMG showed large amplitude motor unit potentials during voluntary recruitment in all patients, suggesting a neurogenic origin of these EMG changes. Neurophysiological study makes it possible to distinguish between myogenic and lower motor neuron involvement. The existence of some degree of spinal cord dysraphism may be pathophysiologically associated with foot deformities.**

Conclusion. **Children with foot deformities and clinical evidence of occult spinal dysraphism should have a neuro-physiological assessment in order to obtain an early diagnosis and avoid ineffective foot surgery.**

KEY WORDS: Spina bifida occulta - Spinal dysraphism - Enuresis - Flatfoot - Foot deformities - Electromyography.

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Spinal dysraphism may include different conditions, such as bone deformities, spinal cord anomalies and midline skin lesions. The particular combination of these disorders affects the severity of the subsequent neurological and orthopaedic damage.[1, 2]

Spina bifida occulta is a variable clinical syndrome with such manifestations as vertebral cleft, foot deformities, midline skin lesion, spondylolysis, genitourinary dysfunction, intraspinal lipoma, syringomyelia and tethered cord syndrome.[3]

Enuresis may be the only clinical evidence.[4] Sometime foot deformities and scoliosis are signs of a muscular imbalance.[5, 6]

A number of spinal cord malformation (*e.g.* tethered cord) may remain subclinical in childhood and appear only later in life, when the clinical picture is more evident and possibly progressive. Electrophysiological techniques have been used to study both clinical and pathophysiological aspects of these conditions. Somatosensory evoked potentials (SEPs) have been used in selected cases [7] and in experimental investigations.[8] Conventional conduction studies (motor and sensory) and electromyogram (EMG) are routinely performed to assess peripheral nerve function. The F wave is a late muscle response resulting from

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antidromic activation of motoneurons following distal stimulation of their axons. F-wave recording may be clinically useful in neuro-orthopedic patients for assessing both peripheral nerve and lumbosacral radicular structures.[9, 10]

The authors identified a group of patients referred to the orthopedic unit for foot deformities, in which the authors observed associated clinical abnormalities (scoliosis, low back pain, midline cutaneous lesions of the back, lower limb length discrepancy) possibly related to spina bifida occulta.

The frequent association between spina bifida occulta and enuresis prompted us to ask these patients about their history — if any — of enuresis and to perform lumbosacral X-rays. The hypothesis was that the presence of enuresis foot deformities and vertebral cleft, could be in most cases associated with spinal dysraphism.[11]

The aim of this study was to clarify possible occult spinal dysraphism in patients with foot deformities and vertebral cleft, with the aid of an extensive electrophysiological investigation including EMG, nerve conduction studies and F-wave recording.

Materials and methods

A group of 47 patients with foot deformities and vertebral cleft were included in the study. The study sample consisted of 23 males and 24 females, with an average age of 14.3 years (range: 8 to 21 years); 32 out of 47 (68%) had flat foot (19 bilateral and 13 unilateral), and 15 out of 47 (32%) *pes cavus* (6 bilateral and 9 unilateral).

All 47 patients had a bony cleft, involving L5 in 11 (23.5%) and S1 in 36 (76.5%) and investigated by standard x-rays. Fifteen patients had undergone magnetic resonance imaging (MRI) elsewhere, revealing no abnormalities of the lower cord (level of the conus medullaris corresponding to the L1 vertebral body).

All patients and their parents were questioned about enuresis, or involuntary voiding of the bladder after the age of five. Thirty-seven out of 47 patients (78.7%) had experienced enuresis, which resolved spontaneously at the mean age of 12.7 (range 10.3 to 20).

Twenty-seven of the 47 patients (57.4%) had deformity of the spine. Twenty-five of them had scoliosis. Scoliosis was seen at the lumbar or dorsolumbar level in all cases, with the apex at D11. The average degree of curvature was 24° (range: 18°-35°). A brace was used in 8 patients. No major double curvatures were found.

Two out of 27 (4.2%) patients had spondylolysis with spondylolisthesis.

Eighteen out of 47 patients (38.3%) complained of low back pain.

Twelve patients (25.5%) had a skin lesions along the midline (6 hairy patches, 4 atrophic scars, 2 lumbar cutaneous capillary haemangiomas).

Eight out of 47 patients (17%) had lower limb length discrepancy less than 3 centimeters.

None of these patients had an occult intraspinal lesion such as an epidermoid or dermoid tumour, lipoma, diastematomyelia, dural band or tethered spinal cord.

In each patient an electrophysiological investigation was conducted in order to assess lower limb nerves and corresponding lumbo-sacral roots (L4-S1 mostly). Motor nerve conduction velocity was calculated by individually stimulating the peroneal and tibial posterior nerve between distal (ankle) and proximal (popliteal fossa) sites; the latency and the amplitude of the compound motor action potential (c-AMP) from extensor digitorum brevis and flexor hallucis brevis respectively were analyzed. The late response F wave was recorded from the same muscles by stimulating the peroneal nerve and tibial posterior nerve at the ankle: the mean minimum latency and the rate frequency (*i.e.* the percentage of F waves) were calculated on a raster of at least 16 consecutive traces. Sensory action potentials (Saps) was recorded by antidromic stimulation of the superficial peroneal nerve and sural nerve: distal latency and amplitude were measured. Electrophysiological tecnique was performed according to Kimura.[12, 13]

Results

The conduction velocities of both the peroneal and tibial nerves were normal in all patients, as were the amplitude and latency of the antidromic sensory potentials (superficial peroneal and sural nerves).

No statistically significant difference in F-wave latency was found between patients and controls ($P>0.05$).

It was observed that the F-wave latency was differently affected in subgroups of patients with *pes cavus* and flat foot. The mean F-wave latency in response to peroneal nerve stimulation in the patients with flat foot was 45.7 ms (SD=2.6), while in the

patients with *pes cavus* it was 47.3 ms (SD=3.8). The statistical analysis showed a slightly significant difference (P=0.04). On the other hand, the mean F-wave latency in response to tibial nerve stimulation was longer in patients with flat foot (49.0 ms; SD=3.1) than in patients with *pes cavus* (47.9 ms; SD=2.5). The statistical analysis showed a significant difference (P=0.02). No statistical correlation on studying the relationships between distal latency of motor action potentials and F-wave latency was observed: this should exclude a distal effect on the F wave and may suggest a more proximal site of involvement (Table I).

Needle EMG during voluntary recruitment showed motor unit potentials of large amplitude in all patients. The motor unit potentials was selected with a short rise time (500 µs or less) for analysis purposes in order to guarantee their proximity to the recording electrode. During voluntary recruitment, the mean amplitude of the EMG signal varies from 1 to 3 mV. Motor unit potentials larger than 4 mV in the recruitment condition of non-maximal effort ere considered "large motor unit potentials".

The absence of spontaneous EMG activity ruled out signs of acute denervation.

TABLE I.—*Mean F-wave latency and standard deviation in patients and control group, and in patients with flat foot and pes cavus.*

F-wave latency (ms)	Normal	Patients	Flat foot	Pes cavus
Peroneal nerve	46.4 (3.0)	45.7 (3.3)	45.7 (2.6)	47.3 (3.8)
	P=0.37		P=0.04	
Posterior tibial nerve	48.5 (2.7)	47.4 (2.9)	49 (3.1)	47.9 (2.5)
	P=0.33		P=0.02	

Discussion

While there are can be no doubt as to the etiopathogenesis of the ensuing bone deformity in the case of spina bifida aperta, closed spinal lesions may remain undetected or be found only incidentally, and are likely to be unrelated to orthopaedic deformities and normally classified as idiopathic.

EMG examination showed motor unit potentials of increased amplitude and a reduced interference pattern: these findings generally suggest signs of disorder of the lower motor neurons or peripheral nerves. Contrasting changes in amplitude and duration of the motor unit potentials may help to distinguish between myopathies and peripheral nerve disease. Electromyography and histochemical findings from muscle biopsies have an overall concordance of 90% or greater.[14] The findings of the present study are not strictly characteristic and cannot be used to differentiate between the topography of the lesions since they are present at the same time in different territories depending on the lower spinal roots. Nevertheless, they can be considered important as a diagnostic marker of neurogenic damage.

The motor conduction velocities of the peroneal and tibial nerves were normal in all patients. Abnormalities of the F-waves in spite of normal conduction velocity in the peripheral nerve, are suggestive of proximal nerve involvement: peroneal nerve F wave is mostly related to conduction along L5 root motor fibers while tibial posterior nerve F wave is mostly related to conduction along S1 root motor fibers. F-wave analysis proved useful for evaluating proximal conduction. An interesting correlation was revealed by the analysis of F-wave latency and foot deformities. Patients with *pes cavus* had a longer latency of the peroneal nerve F wave than patients affected by flat foot. Conversely, patients affected by flat foot had a longer tibial nerve F-wave latency than patients affected by *pes cavus*.

Since there was no correlation whatsoever between the distal latency of motor potentials and the F wave of the corresponding nerve, the authors postulated that the F wave abnormalities might be predominantly due to damage to the proximal part of the nerve (root, spinal cord).

Foot deformities may be early evidence of disease in patients with hereditary sensory motor neuropathy (HSMN). In Charcot-Marie-Tooth (CMT) type 2 (axonal type), EMG shows increased-amplitude motor unit potentials in leg and foot muscles. In mildly diseased nerves, a slow motor conduction may be present in distal segments alongside normal conduction in the proximal segment.[15] Moreover, the sensory action potentials were completely normal in both latency and amplitude. This result is particularly important, since it provides additional evidence of a preganglionic origin of the EMG signs.

Conclusions

On the basis of these findings, the authors suggest that the neurological damage probably occurs in the

proximal part of the nerve (root, spinal cord). Fifteen patients had previously undergone spinal MRI elsewhere: in agreement with other studies, no structural defects were detected in these patients because the lesion is probably beyond the resolution power of MRI, as claimed by other investigators in the case of isolated bladder neuropathy. MRI is a useful tool in evaluating these patients: in fact, it ruled out any abnormality of lower spinal cord signal intensity and, that's more, showed that conus medullaris is lying in the right position without thickness of phylum terminale.[16-18] Although the question is still controversial, myelo-computed tomography (CT) scans or ultrasound could also be used in order to exclude minimal spinal disease.[2, 3, 13, 18, 19]

The diagnosis of spina bifida occulta is easier when skin lesions are present along the midline on the back associated with foot deformities.[6]

The authors suggest enquiring about enuresis and performing lumbosacral x-rays when foot deformities (flat foot and *pes cavus*) are classified as idiopathic, especially if bilateral, and associated with other clinical evidence possibly pointing to occult spinal dysraphism. The high incidence of enuresis observed in this group of patients (78.8%) suggests that enuresis could be considered the result of delayed lower spinal cord development affecting bladder innervation.

It should be added that neurophysiology can be considered as a useful diagnostic aid and should be performed before foot surgery in patients with occult spinal dysraphism or in patients with recurrence of foot deformity after surgery.

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