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Intrathecal baclofen for intractable spasticity in amyotrophic lateral sclerosis

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Sirs: Amyotrophic lateral sclerosis (ALS) is a motor neuron disorder with poor prognosis because the cause is unknown [2]. Spasticity and painful muscle cramps are common symptoms of the disease. Antispastic drugs can relieve spasticity, but severe side effects restrict their use when the maximum daily dose is exceeded. The efficacy of intrathecal baclofen has been well demonstrated in treating spasticity of cerebral and spinal origin [1, 3, 4]. The use of baclofen intrathecally in ALS has not yet been reported to our knowledge. This prompted us to describe the case of a patient suffering from ALS treated in this manner, thanks to which mobility is still maintained.

Two years previously a now 23-year-old man noticed progressive gait disturbance and weakness of his right foot. At night he experienced painful muscle cramps in the legs. Physical examination revealed generalized fasciculations and weakness of the right hand and foot (MRC grade 4/5). Muscular tone was markedly elevated, with signs of spasticity and hyperreflexia. His mental state was normal, and there were no sensory deficits. Magnetic resonance imaging of brain and spinal cord were normal, as were the findings of CSF. Electromyography confirmed the suspected diagnosis of ALS. Antispastic medication was initiated, with baclofen and memantine. During the next year the patient remained mobile but progression of the disease led to worsening spasticity and requires an increase in baclofen dose. At a daily oral dose of 80 mg baclofen spasticity was no longer ameliorated, but the patient developed side effects, such as weakness, daytime fatigue, and sleepiness.

He was then admitted to our Neurosurgical Clinic, giving informed consent to an intrathecal treatment. Clinically he showed severe paraspasticity and was able to walk only a few steps with help. According to the El Escorial criteria for the diagnosis of ALS [5], the patient was classified as definite ALS with involvement of the lower and upper motor neurons in three regions; he continued to show progression of the disorder. In the first procedure an intrathecal catheter was implanted at the L4-L5 level to assess the efficacy of intrathecal baclofen. The tip of the catheter was placed at the midthoracic level, and it was connected to a portsystem. Baclofen was administered continuously at increasing doses using an external pump device. This made it possible to raise the dose step by step, creating a steady concentration of the medicament within the CSF. Thus a drug overdose with repeated bolus injections can be safely avoided. Moreover, the individually required daily dose administered continuously by the implanted pump can be exactly defined and simulated in this test phase under clinical observance. At a dose of 160 µg the patient showed only minimal clinical signs of spasticity, was able to walk without help and could even climb stairs. In a second procedure the portsystem was removed and replaced by an implantable pump. The pump is percutaneously refilled every 6 weeks ambulatorily, and the follow-up period is now 1 year. Spasticity showed further increase due to progression of the disease, requiring an adjustment of dose. The patient presently receives 480 µg baclofen daily and is still able to walk.

ALS is a degenerative motor neuron disorder characterized by progressive paresis, fasciculations, and spasticity. Since there is no causative therapy available for ALS prognosis is poor and treatment is symptomatic [2]. Spasticity and painful muscle cramps are disabling symptoms markedly reducing the patient's quality of life. There are several antispastic drugs such as baclofen, memantine or benzodiazepines which can effectively relieve spasticity. Due to progression of the disease, however, spasticity can increase and oral medication can no longer ameliorate spasticity when the maximum daily dose is reached and side effects occur. This is generally the fact with baclofen when a daily oral dose of 80 mg is exceeded. Weakness, daytime fatigue and sleepiness are the most common side effects. Since ALS patients need adequate palliative treatment more than anything else [2], the intrathecal administration of baclofen offers the temporary maintenance of mobility. Before the implantation of the pump our patient was able to walk only a few steps with help, despite high doses of oral antispastic drugs, while after the implantation, he was able to walk without help and even climb stairs. The implantation of a drug-delivering pump device is a safe procedure without significant risks. Increased spasticity due to progression of the disease can be dealt with by adjusting the delivered dose. In our patient we raised the dose from 160 to 480 µg baclofen daily within 1 year. Although spasticity continues to increase, he is still able to walk, now with help of a rollator because of progression of the weakness. Maintenance of mobility as long as possible is of paramount importance for ALS patients and their caregivers.

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