

50 Years of Progressive Supranuclear Palsy

Palliative Care in Progressive Supranuclear Palsy
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Patients with chronic neurologic disorders suffer from the burden of disease progression without the hope for a cure. Therefore, symptom management and palliative care approaches should be included early on. Palliative care aims at improving a patient's quality of life and meaning in life by alleviating suffering due to physical, psychosocial and spiritual illness. Since no curative and only limited life-prolonging treatment options are available for patients with Progressive Supranuclear Palsy (PSP), a palliative care approach can help to create a treatment plan that considers all aspects of the disease. A palliative approach to PSP does not mean to limit treatment and focus on pain. Instead, the whole "unit of care", consisting of the patient and his relatives and caregivers, should be perceived with all their needs.

It has become clear in recent years that the "total symptom" concept, the multiprofessional approach, early palliative care integration and academic models, all are very relevant to many patients suffering from movement disorders. However, still only a few patients find their way to palliative care units or into hospices. Patients with PSP are not traditionally managed by palliative care teams. However, as there are no disease-modifying agents available, and the wide spectrum of symptoms have a significant impact on the quality of life of the patients and families, palliative intervention has a lot to offer in the management of these conditions.

Disease trajectory in PSP is frequently divided into the supportive phase, the phase of transition and the terminal phase. In contrast to the normal life expectancy of patients suffering from PD, the median survival of PSP is estimated at 6 – 9 years.

Distressing motor symptoms include bradykinesia, muscle rigidity, dystonia and instability of gait. Physiotherapy is the mainstay of non-pharmacologic therapy to improve balance and self-

confidence. Painful dyskinesias and dystonias are distressing for patients and carers and require aggressive management. Increased sweating, delayed gastric emptying, constipation, sialorrhea and urinary urge incontinence are part of the dysautonomic spectrum of PSP.

Progressive dysphagia is due to rigidity and hypokinesia as well as the gradual involvement of the dorsal motor nucleus of the vagus nerve in the disease process. So far, there is no evidence to suggest a survival or quality of life benefit by feeding tube placement in advanced PSP. This remains an individual decision following discussion with patient and caregivers.

Despite PSP is rapidly progressive, there is now evidence that quality of life can be preserved with high quality palliative care. However, in the first years of the disease almost one third of patients has suicidal ideation and might seek assisted suicide.

As the patient becomes more and more disabled and persistent neuropsychiatric problems develop, the priority of care shifts to preservation of quality of life through effective symptom control.

In their last days of life, patients with PSP often suffer from pneumonia. Bradykinesia increases dramatically, and often even spasticity is seen. Communication might not been possible due to severe generalized dystonia (including larynx and pharynx muscles as well as mouth opening dystonia). Eyes are often wide open, but apparently there is an inability to control eye movements. Seizures as well as myoclonic jerks occur frequently. It is not rare for patients to exhibit profound cachexia, even if tube feeding has been placed earlier on. However, patients are often able to understand and want to participate in decisions. Therefore, decisions regarding life limiting procedures should be discussed not only with the relatives but also with the patient. In our experience, most patients with PSP refuse life-prolonging therapies.