Parkinson's syndrome/PGS/diagnosis

Parkinson's syndrome of other degenerative etiology

It is usually distinguished from PN by a more rapid progression, a more severe prognosis, the presence of non-motor symptoms early in development, and little (usually early) or no response to dopaminergic therapy. It is typically manifested by symmetrical HRS (hypokinetic rigid syndrome) minimal tremor and some of the other symptoms: postural instability, dementia, apraxia, cerebellar syndrome, dystonia, dysphonia, oculomotor disorder, and vegetative dysfunction.

The diagnosis is primarily clinical and can only be definitively determined post-mortem.

Progressive supranuclear palsy (PSP)

A sporadic disease with a prevalence of around 5/100,000 and onset after the age of 40, typically between the 50s and 70s. a year. HRS is accompanied by: oculomotor disorder (supranuclear paresis of vertical gaze - more down, hypometric saccades, convergence disorders, eyelid retraction or blepharospasm), early postural instability with falls, severe dysarthria/dysphonia and cognitive deficit with progression to dementia of the frontal type and severe psychomotor retardation. Accentuated rigidity in the area of the axial and neck muscles twists the torso and neck into hyperextension. Rapid progression quickly leads to insufficiency. 2 clinical types are distinguished: more benign **PSP-parkinsonism** with partially preserved reactivity to dopaminergic therapy and slower progression and classic **PSP- Steele-Richardson-Olszewski**. An MRI is a finding of atrophy of the dorsum of the mesencephalon. There is no causal or effective symptomatic therapy. The response to the dopaminergic stimulus is often poor or gradually disappearing, however, it is advisable to test the effect of levodopa up to high doses. Amantadine and tricyclic antidepressants may also have an effect.

Multisystem atrophy (MSA)

Sporadic disease with a prevalence of 2–5/100 000 and onset after age 30. It leads to degenerative changes in the striatum, stem, cerebellum and autonomic ganglia. Symptoms from the degeneration of the above-mentioned structures can be expressed differently in individual patients. This gave rise to the classic clinical subtypes: striatonigral degeneration (dominated by parkinsonian syndrome), olivopontocerebellar atrophy (dominated by cerebellar syndrome) and Shy-Drager syndrome (autonomic dysfunction dominated). Today, MSA is divided into the **parkinsonian subtype – MSA-P** (80 %) and the **cerebellar subtype – MSA-C** (20 %), which are always accompanied by an autonomic disorder. HRS is often accompanied by orofacial, cervical (antecolis) or limb dystonia, dysphagia, typical dysarthria with a high-pitched voice, sometimes inspiratory stridor, sleep apnea syndrome, and REM sleep behaviour disorder. Postural disorders and autonomic dysfunction (erectile dysfunction, incontinence and orthostatic hypotension) appear earlier and more pronounced. MSA-C may initially be indistinguishable from the cerebellar syndrome of other etiology. Progression is usually rapid with early disability. Of the auxiliary examinations, the following are important: orthostatic test, MRI of the brain (hypointense putamen, trunk/cerebellar atrophy, hot cross-bun sign in the trunk) and EMG urethral or anal sphincter (finding denervation potentials). There is no causal or effective symptomatic therapy, we try to treat the most bothersome symptoms:

Tab.4: Symptomatic therapy of MSA

Akinesia, rigidity, postural instability	▪ High dose of levodopa − up to 2000 mg (cave worsening of hypotension) ▪ Amantadine − series of ten infusions or 3x1 tablet. ▪ Gait rehabilitation
Orthostatic hypotension	See complications of PN
Inspiratory stridor, SAS	Continuous positive airway pressure (CPAP) therapy
Urogenital problems	▪ Detrusor hyperfunction → oxybutynin, trospium chloride ▪ Retention → intermittent autocatheterization ▪ Nocturnal polyuria → desmopressin spray 10 µg NN ▪ Erectile dysfunction → sildenafil
Dystonia	Application of botulinum toxin
Dysarthria, dysphagia	▪ Speech therapy ▪ PEG in severe disability

Corticobasal degeneration (CBGD)

Sporadic, rare, smoothly progressive disease with onset usually after age 70. It is characterized by asymmetric HRS accompanied by other subcortical symptoms (dystonia, myoclonus) and at least one isolated cortical symptom (apraxia, discriminative hearing disorder, neglect syndrome, dysphasia, alien-limb syndrome). There is a cognitive deficit that can progress to dementia. On an MRI, unilateral or asymmetric cortical atrophy is usually found in accordance with clinical impairment. Iodobenzamide SPECT and FDG PET will show asymmetric hypometabolism in the cortex and decreased density of dopamine receptors in the striatum. Therapy is insufficient - a small percentage of patients respond to high doses of levodopa. Painful dystonia of the hand is indicated for the application of botulinum toxin.

Lewy body disease (DLBD)

The sporadic disease manifests as a combination of Parkinson's syndrome and cognitive impairment with significant fluctuation during the day, when the patient can be severely affected by dysexecutive syndrome and in a few hours completely fine. Typical are visual hallucinations, complex systematized delusional productions and sensitivity to neuroleptics, which are contraindicated (they cause a sharp deterioration of HRS with the risk of developing a neuroleptic malignant syndrome). HRS syndrome responds more to dopaminergic therapy than other causes of PS, and psychotic symptoms and cognitive deficits respond well to cognitive therapy. In **Alzheimer's dementia with extrapyramidal features,** there are no such significant fluctuations in mental status, and psychotic symptoms and HRS are less pronounced.