

ERN-RND/EAN/MDS-ES Guidelines on the symptomatic treatment of Atypical Parkinsonian Syndromes



DG/WG
Atypical
Parkinsonism

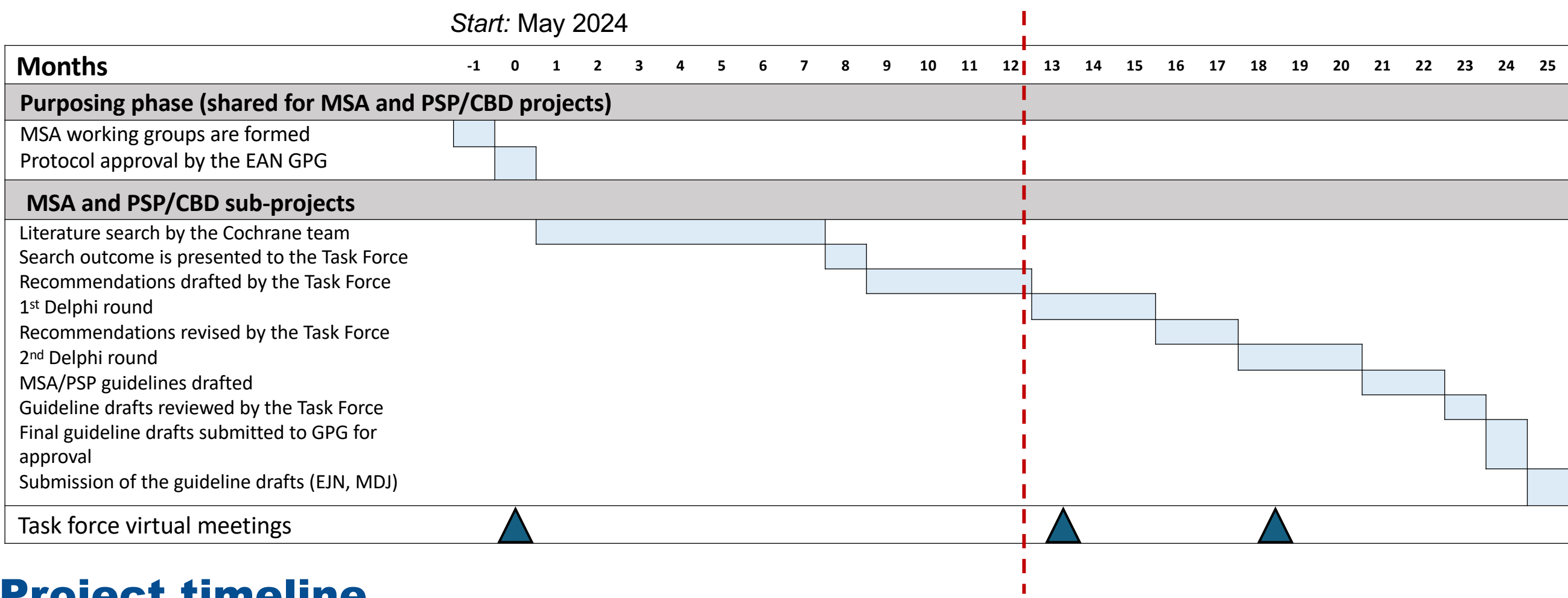
Introduction

Multiple system atrophy (MSA), progressive supranuclear palsy (PSP) and corticobasal degeneration (CBD) are rare, rapidly progressive atypical Parkinsonian Syndromes (APS), characterized by motor, autonomic, sleep, cognitive and behavioral features in various combinations. A lack in systematic evaluated guidelines for APS leads to delayed diagnosis of disease-specific symptoms, inadequate treatment thereof and ultimately higher costs (Winter, Stamelou et al. 2011).

Objective

After the recent international efforts resulting in updated criteria for a more accurate and timely diagnosis of both PSP and MSA (Höglinger, Respondek et al. 2017, Wenning, Stankovic et cal. 2022), the aim is to develop evidence- and consensus-based management guidelines for patients with APS, which meet the highest standards of clinical guidelines.

Results



Project timeline

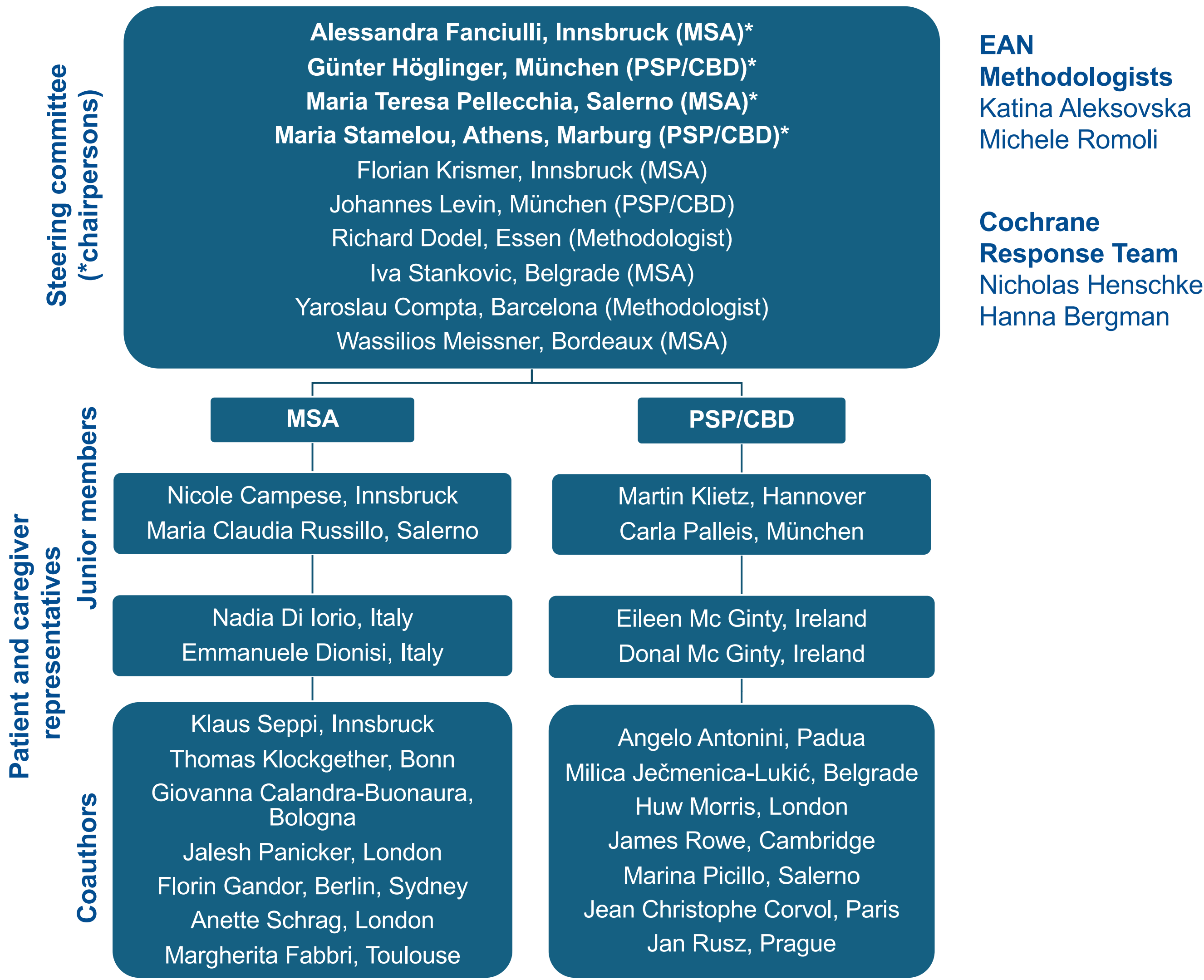
MSA	
PICO No.	Topic
Motor features	
PICO 1	Parkinsonism
PICO 2	Dystonia/spasticity
PICO 3	Cerebellar features
PICO 4	Recurrent falls
Autonomic features	
PICO 5	Bladder disturbances
PICO 6	Urinary tract infections
PICO 7	Sexual dysfunction
PICO 8	Orthostatic hypotension
PICO 9	Supine/nocturnal hypertension
PICO 10	Hypo/anhydrosis
PICO 11	Gastrointestinal dysfunction
Sleep disorders	
PICO 12	REM-sleep behaviour disorder
PICO 13	Stridor
PICO 14	Sleep disorders of breathing
PICO 15	Insomnia
PICO 16	Restless legs syndrome
Drooling	
PICO 17	Drooling
Dysphagia	
PICO 18	Dysphagia
Cognitive and behavioural symptoms	
PICO 19	Cognitive impairment
PICO 20	Depression/anxiety
PICO 21	Hallucinations/psychosis
PICO 22	Social isolation
Palliative care/pain	
PICO 23	Pain
PICO 24	Quality of life

PICOs for MSA and PSP/CBD

PSP/CBD	
PICO No.	Topic
Motor features PSP/CBD	
PICO 1	Parkinsonism (PSP)
PICO 2	Dystonia/spasticity (PSP)
PICO 3	Oculomotor (PSP)
PICO 4	Parkinsonism advanced (PSP)
PICO 5	Parkinsonism (CBD)
PICO 6	Dystonia/spasticity (CBD)
PICO 7	Myoclonus (CBD)
PICO 8	Parkinsonism advanced (CBD)
PICO 9	Complications of falls (PSP/CBD)
Non-Motor features PSP/CBD	
PICO 10	Bladder disturbances
PICO 11	Urinary tract infections
PICO 12	Gastrointestinal dysfunction
PICO 13	Insomnia
PICO 14	Cognitive impairment
PICO 15	Depression/anxiety
PICO 16	Hallucinations/psychosis
PICO 17	Drooling
PICO 18	Dysphagia
PICO 19	Dysarthria
PICO 20	Quality of life

Work Plan

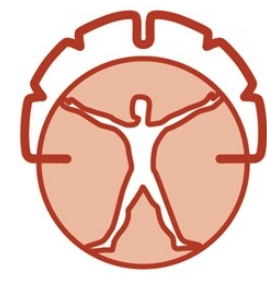
- **Target population:** Individuals with MSA, PSP/CBD.
- **PICOs:** developed by a DGN guideline initiative.
- **Methodology:** Systematic review of literature and evidence synthesis performed in cooperation with Cochrane Response Team.
- **Interventions included:** Pharmacological and non-pharmacological measures to relieve motor and non-motor features., and advanced care planning, including palliation and model of cares.
- **Interventions not included:** Disease-modifying interventions.
- **Main outcomes to be considered:** Symptomatic relief/functional improvement after treatment of a given motor or non-motor feature; for advanced care planning: improvement of quality of life.
- **Target audience:** Neurologists, palliative care experts, healthcare professionals.
- **Setting:** Primary to tertiary care.



Flow chart: The collaborative team

Outlook

- Complete PICO recommendations
- Draft MSA/PSP guidelines
- Submit guidelines for open access publication



European Reference Network
for rare or low prevalence complex diseases

Network
Neurological Diseases
(ERN-RND)



International Parkinson and Movement Disorder Society
European Section



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Winter Y, Stamelou M, ... , Spottke AE. Cost-of-illness in multiple system atrophy and progressive supranuclear palsy. J Neurol. 2011 Oct;258(10):1827-34.
Höglinger GU, Respondek G, Stamelou M, ... , Boxer AL, Golbe LI, Litvan I. Clinical diagnosis of progressive supranuclear palsy: The movement disorder society criteria. Mov Disord. 2017 Jun;32(6):853-864.
Wenning GK, ... , Kaufmann H. The Movement Disorder Society Criteria for the Diagnosis of Multiple System Atrophy. Mov Disord. 2022 Jun;37(6):1131-1148.