Multiple system atrophy (MSA)

- sporadic neurodegenerative disease (one of the synucleinopathies) characterised by varying degrees of
  - cerebellar ataxia
  - autonomic dysfunction
  - parkinsonism
  - corticospinal dysfunction.

Multiple system atrophy

- Typically symptoms begin between 40 and 60 years of age.
- Clinical presentation is variable, but typically presents in one of three patterns (initially described as separate entities) [1-2]:
  - Shy-Drager syndrome is used when autonomic symptoms predominate
  - Striatonigral degeneration shows predominant parkinsonian features
  - Olivopontocerebellar atrophy demonstrates primarily cerebellar dysfunction

Divided in to 2 Forms

- **MSA-C:** predominance of cerebellar symptoms (olivopontocerebellar atrophy)
- **MSA-P:** predominance of parkinsonian signs and symptoms (striatonigral degeneration)
- Some older texts refer to **MSA-A** to denote **Shy-Drager syndrome.**
  - In the latest consensus however autonomic symptoms are considered part of both MSA-C and MSA-P and thus the term MSA-A is no longer used.

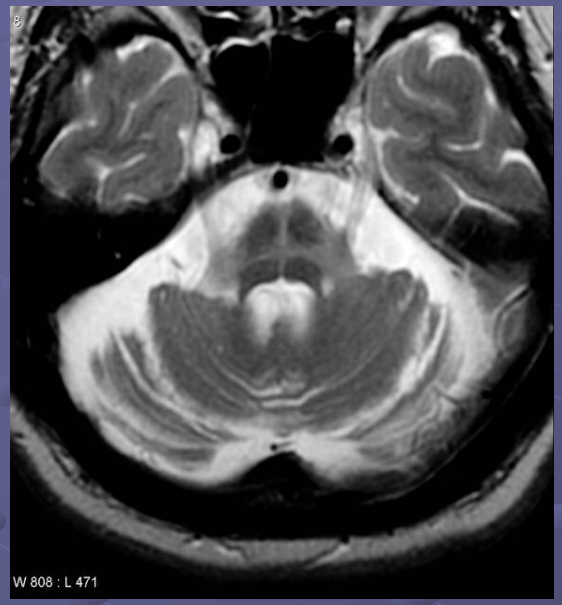
MSA

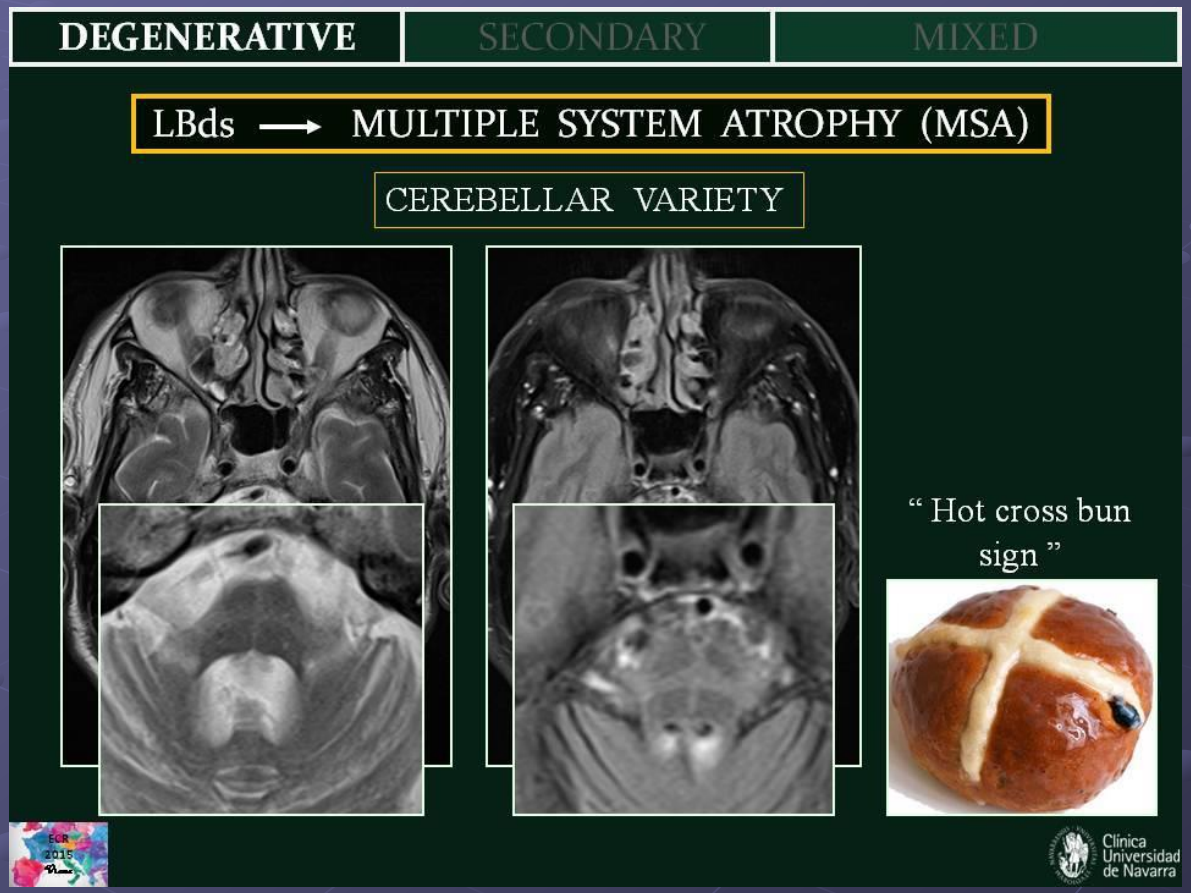
- Like other synucleinopathies, multiple systemic atrophy results from abnormalities of alpha-synuclein metabolism, resulting in intracellular deposition.
- Unlike Parkinson disease and Lewy body dementia (two other synucleinopathies) these intracellular deposits are found not only in **neurons** but also **in oligodendroglia** .

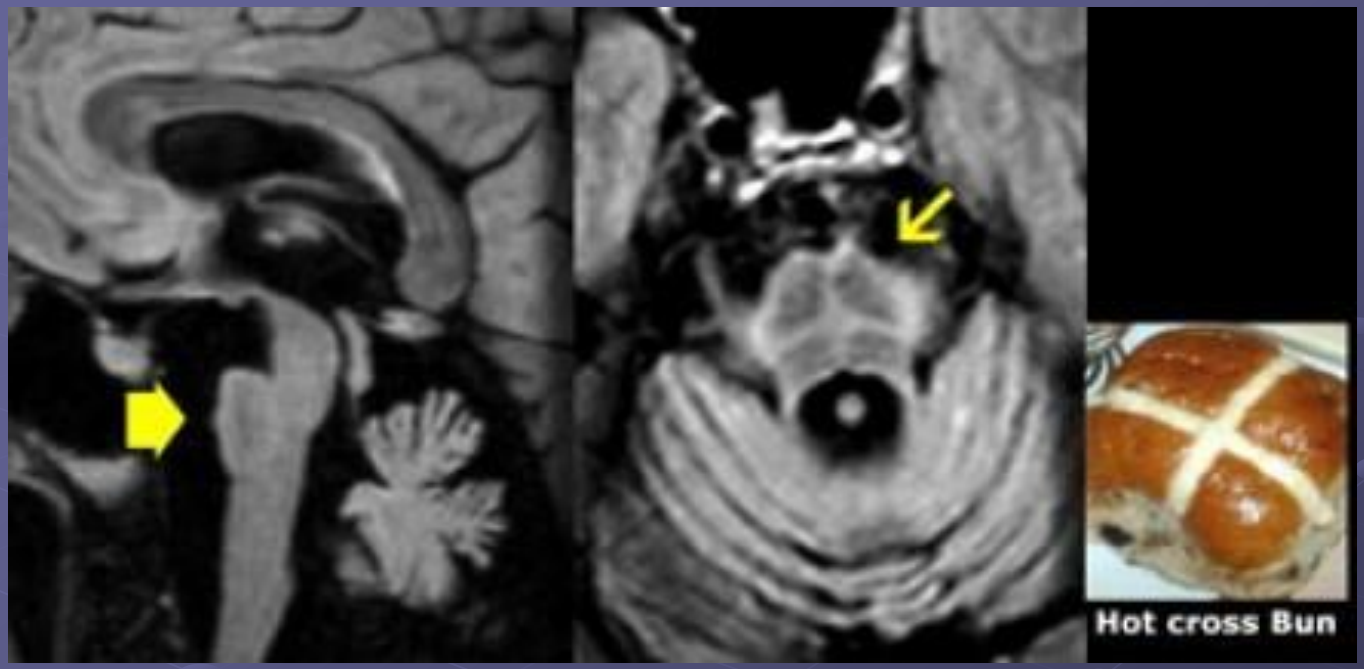
MRI

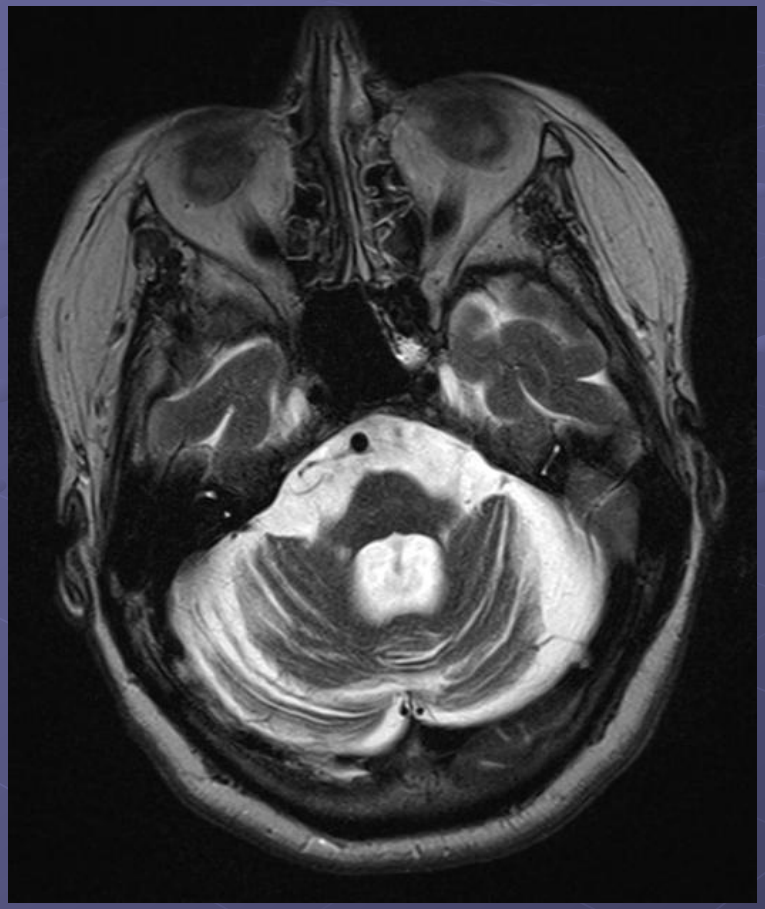
- **T2:** hyperintensities typically present in the pontocerebellar tracts
  - pons: hot cross bun sign (MSA-C)
  - middle cerebellar peduncles
  - cerebellum
- putaminal findings in MSA-P [5]:
  - reduced volume
  - reduced GRE and T2 signal relative to globus pallidus
  - reduced GRE and T2 signal relative to red nucleus
  - abnormally high T2 linear rim surrounding the putamen ("putaminal rim sign"), seen at 1.5T (this is normal at 3T) [7] (see case 3)
- MSA-C
  - disproportionate atrophy of the cerebellum and brainstem (especially olivary nuclei and middle cerebellar peduncle)
- ADC values: higher in the pons, cerebellum, and putamen than in Parkinson disease or controls
- **Generalized Brain Atrophy,**

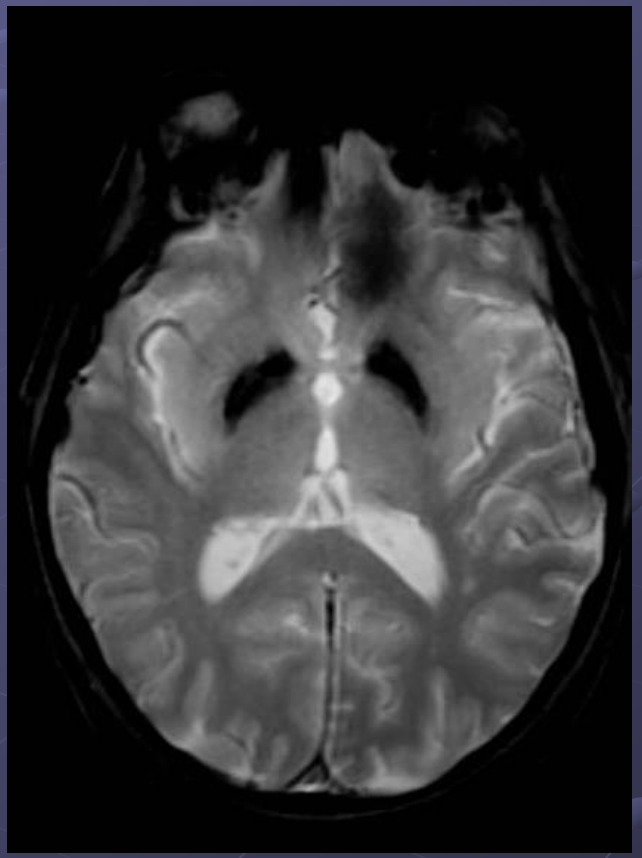
Hot Cross Bun Sign






Hot cross Bun





Putaminal rim sign