

RAPIDLY PROGRESSIVE DEMENTIA

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DISCLOSURES

None

LEARNING OBJECTIVES

- Know how to diagnose Rapidly Progressive Dementia.
- Recognize curable causes of Rapidly Progressive Dementia..
- Know how to rule out infectious causes such as Neuro-Syphilis and Neuro-HIV.
- Recognize autoimmune encephalopathies, the main antibodies involved and the appropriate treatment.
- Recognize paraneoplastic encephalopathies and look for the visceral neoplasm involved.
- Know clinical, brain imaging, EEG and CSF features of Creutzfeldt-Jakob Disease.
- Recognize neurodegenerative dementia (such as Alzheimer' disease or Fronto-Temporal Dementia) with Young-Onset and Rapid Progression.
- Know the Genetic Metabolic diseases responsible for dementia

KEY MESSAGE

- RPD is defined as progression to dementia or death within 2 years.
- The age of onset and the mode of progression are key to the diagnosis.
- Brain MRI features can guide the etiological diagnosis.
- Biological, immunological and infectious tests are essential to rule out a curable cause.
- Prion diseases and neurodegenerative diseases are among the most frequent causes.
- Biomarkers in the CSF or by PET scan allow the diagnosis of AD or FTD
- Autoimmune encephalitis require specific antibody testing and appropriate treatment
- In case of paraneoplastic encephalopathy the search for the responsible neoplasm must be active.
- The prognosis of curable forms depends on the prompt use of an appropriate treatment

REFERENCES (I)

- Banks SA, Elia Sechi E, Flanagan EP(2021). Ther Adv Neurol Disord 2021, Vol. 14: 1–16.
- Benabdeljlil M, Boutbibe F, Rahmani M, Benbelaïd F, Bennani M, Aïdi S, El Alaoui Faris M. (2015). Dementia and Alzheimer's disease (AD). Experience of the memory center of rabat. Journal of the Neurological Sciences, 357, e125.
- El Alaoui Faris M (2019). Dementia in Arab World. <https://wfneurology.org>
- Figgie, M.P., Jr.; Appleby, B.S. (2021). Clinical Use of Improved Diagnostic Testing for Detection of Prion Disease. *Viruses*, 13, 789.
- Geschwind MD (2016). Rapidly Progressive Dementia. *Continuum (Minneapolis)* 22(2):510–537.
- Josephs KA, Ahlskog JE, Parisi JE, et al (2009). Rapidly progressive neurodegenerative dementias. *Arch Neurol* 66(2):201-207.
- Geschwind MD, Shu H, Haman A, et al (2008). Rapidly progressive dementia. *Ann Neurol*;64(1): 97-108.

REFERENCES (2)

- Gregory SD (2019). Reversible Dementias (2019). Continuum (Minneap Minn) ;25(1, Dementia):234–253.
- McKeon A.(2016). Autoimmune encephalopathies and dementias. Continuum (Minneap Minn) 2016;22(2 Dementia): 538-558 Papageorgiou SG, Kontaxis T, Bonakis A, et al (2009). Rapidly progressive dementia: causes found in a Greek tertiary referral center in Athens. Alzheimer Dis Assoc Disord 2009; 23(4): 337-346.
- Sala I, Marquie´ M, Sa´ nchez-Saudino´ s MB, et al.(2012). Rapidly progressive dementia: experience in a tertiary care medical center. Alzheimer Dis Assoc Disord ;26 (3): 267-271.
- Schmidt C, HaBk S, Satoh K, et al.(2012) Rapidly progressive Alzheimer’s disease: a multicenter update. J Alzheimers Dis;30(4):751-756.
- Shrestha R, Wuerz T, Appleby D S. Rapidly Progressive Young-Onset Dementias Neuropsychiatric Aspects. Psychiatr Clin North Am. 2015 Jun;38(2):221-32.
- Vitali P, Maccagnano E, Caverzasi E, et al (2011). Diffusion-weighted MRI hyperintensity patterns differentiate CJD from other rapid dementias. Neurology;76(20) : 1711-1719.