Riunione annuale SIN Calabria

La diagnosi clinica e di laboratorio delle diverse malattie responsabili di decadimento cognitivo

Università Magna Graecia di Catanzaro
Aula D2

Catanzaro, 23 novembre 2019

REGIONE CALABRIA
AZIENDA OSPEDALIERA
"Pugliese-Ciaccio"
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Altre etiologie di decadimento cognitivo

Emilio Le Piane

Struttura Operativa Complessa di Neurologia "F. PETITTO"

malattie responsabili di decadimento cognitivo

Neurology: Clinical Practice ▮ September 2012

Diagnosis and treatment of rapidly progressive dementias

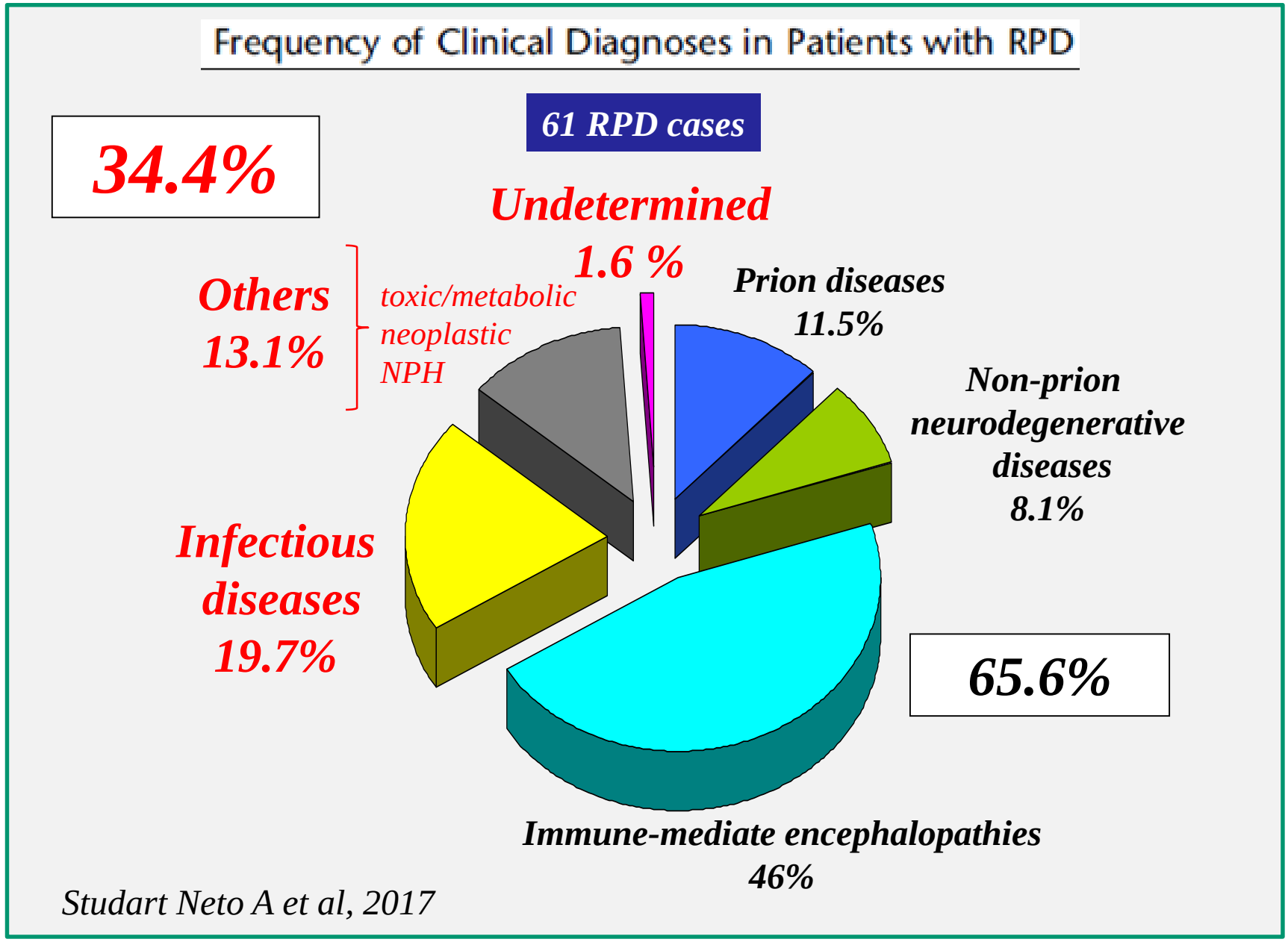
Ross W. Paterson, MRCP*
Leonel T. Takada, MD*
Michael D. Geschwind, MD, PhD

Although no formal definition exists for tia (RPD), generally we use the term from illness onset, but more comm conditions are relatively uncommon,

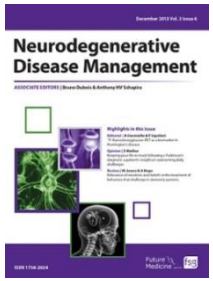
what constitutes a rapidly progressive demen- when dementia occurs in less than 1–2 years only over weeks to months.[1] Because these

- Malattia di Alzheimer
- Demenza fronto-temporale
- Malattia di Parkinson
- demenza a corpi di Levy
- Malattia cerebro-vascolare
- Encefalopatie spongiformi
- Malattie infiammatorie

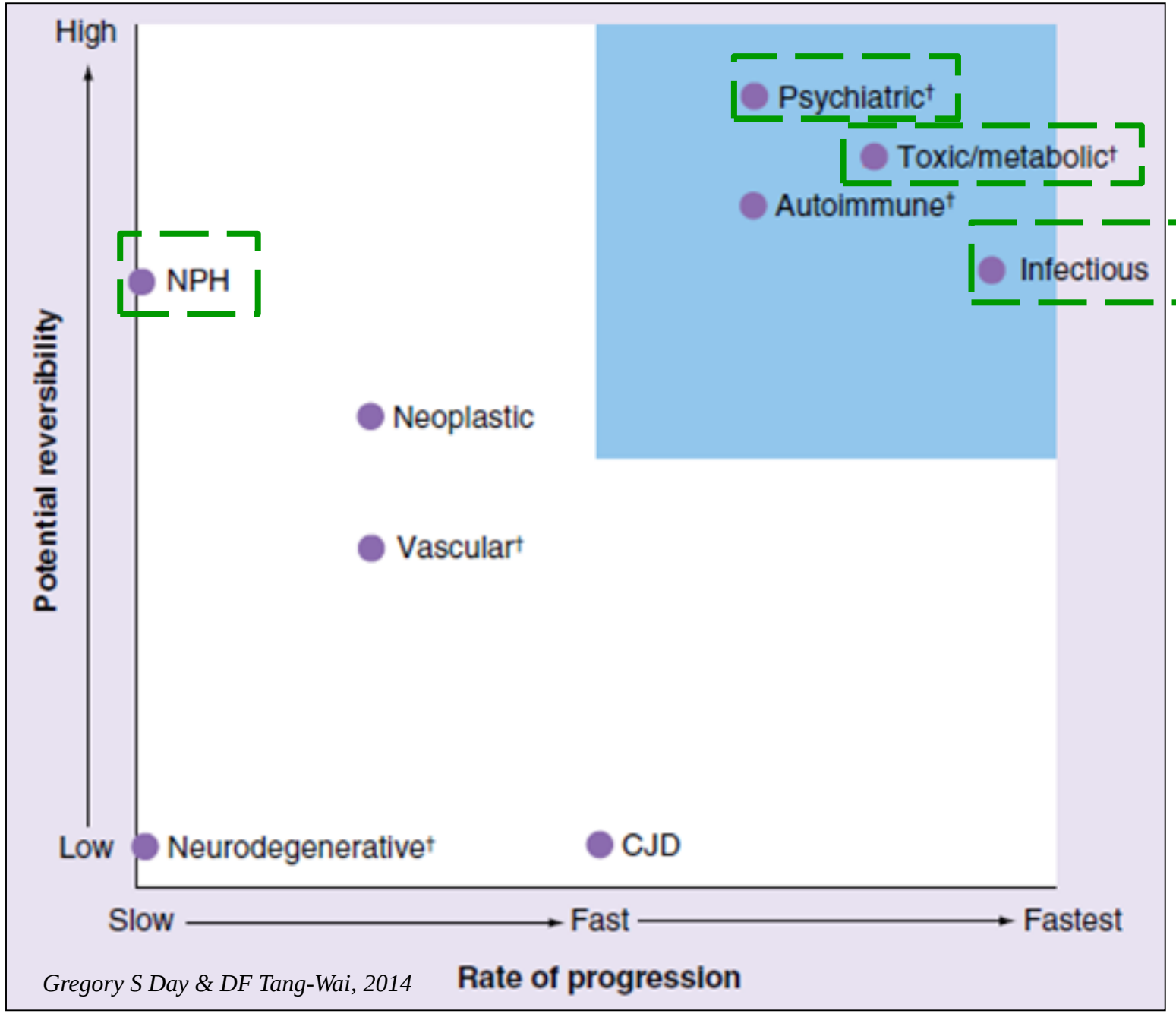
Altre etiologie di decadimento cognitivo



Neurodegen. Dis. Manage. (2014) 4(1), 41–56



Potential reversibility

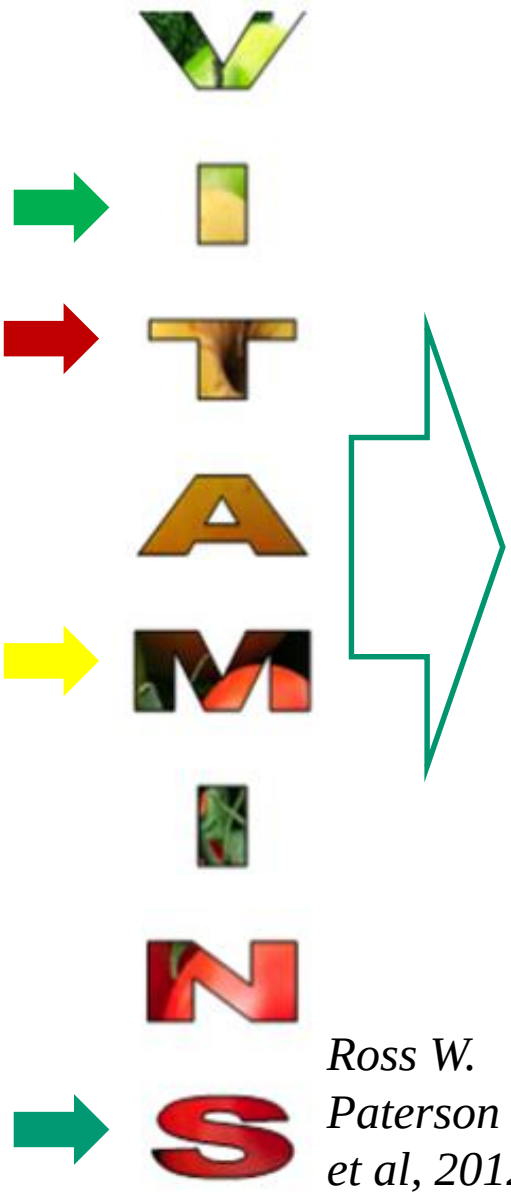


Gregory S Day & DF Tang-Wai, 2014

Etiology of RPDs: use of the mnemonic VITAMINS

Ross W. Paterson et al, 2012

Etiology of RPDs: use of the mnemonic VITAMINS



Ross W. Paterson et al, 2012

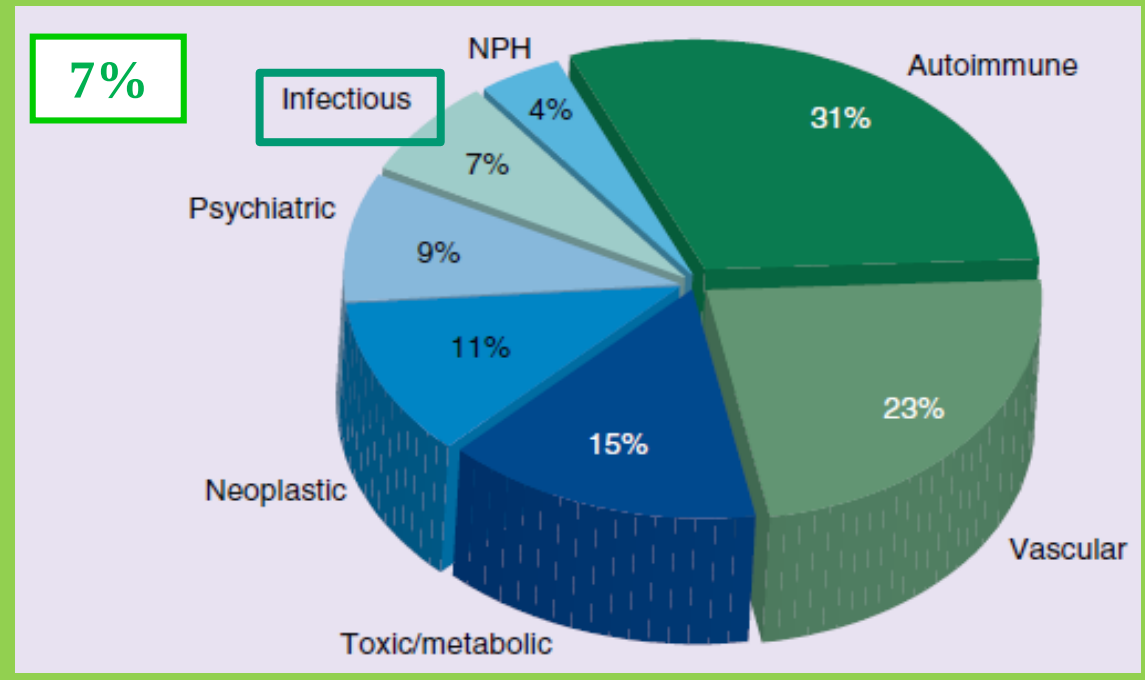
- V Vascular
- I Infectious
- T Toxic-metabolic
- A Autoimmune
- M Metastases/neoplam
- I Iatrogenic/inborn error of metabolism
- N Neurodegenerative
- S Systemic/Seizures

Etiology of RPDs: use of the mnemonic VITAMINS

Ross W. Paterson et al, 2012

V Vascular

I Infectious



Gregory S Day & D.F. Tang-Wai, 2014

Diagnostic challenges in rapidly progressive dementia

EXPERT REVIEW OF NEUROTHERAPEUTICS
https://doi.org/10.1080/14737175.2018.1519397

I. ZERR AND P. HERMANN

Table 1. Differential diagnoses of RPD reported by tertiary referral centers.

	Athens, Greece [61] (n = 68*)	Zhejiang, China [106] (n = 310**)	Sao Paulo, Brazil [14] (n = 61)	Chandigarh, India [15] (n = 187)
Infectious encephalitis (%)	5.9*	21.9	19.7	20.6
Immune-mediated disease (%)	8.8	9.0	45.9	18.2
Creutzfeldt–Jakob disease (%)	13.2	7.1	11.5	7.5
Neurodegenerative diseases (%)	47.0	24.8	8.2	14.4
Alzheimer's disease (%)	17.6	14.5	n.a.	n.a.
Others (%)	29.4	10.3	n.a.	n.a.
Vascular dementia (%)	13.2	**	n.a.	9.6
Toxic + metabolic (%)	*	10.3	n.a.	16.0
Others (%)	11.8	26.9	14.7	13.4

Infectious agents (other than prions) possibly causing RPD

Viral

- HIV
- JC-virus (progressive multifocal leukoencephalopathy)
- Herpes simplex virus 1, 2 (6, 7)
- Varicella zoster virus
- Measles (sclerosing subacute panencephalitis)
- Mumps
- Epstein–Barr virus
- Influenza virus
- Cytomegalovirus
- Japanese encephalitis virus
- West Nile virus
- Rabies
- Parvovirus B19
- Hepatitis C

Bacterial

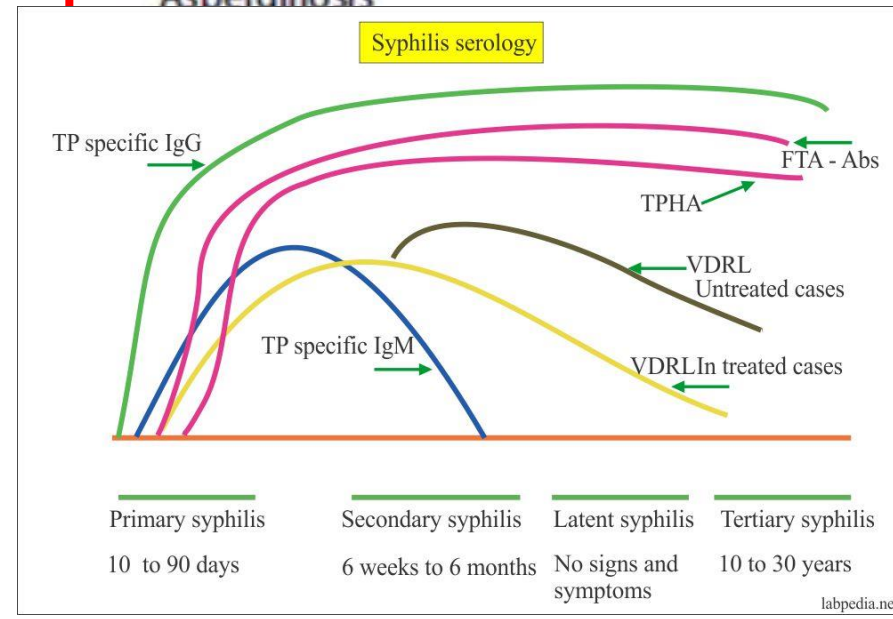
- *Treponema pallidum* (Lues)
- *Tropheryma whipplei* (Whipple's disease)
- *Mycobacterium tuberculosis*
- *Listeria monocytogenes*
- *Borrelia burgdorferi*

Parasitic/Fungal

- *Toxoplasma gondii*
- Balmuthia
- *Cryptococcus neoformans*
- Aspergillosis

Viral encephalitis – clinical symptoms

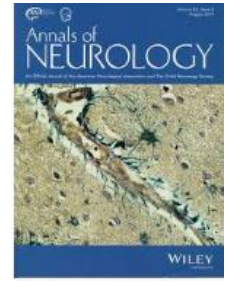
- **Typical presentation**
  - Acute flu-like prodrome
  - High fever, severe headache
  - Altered consciousness (lethargy, drowsiness, confusion, coma)
  - Seizures
- **More subtle presentations**
  - Low grade fever
  - Speech disturbances (dysphasia, aphasia)
  - Behavioural changes
  - Subacute and chronic presentations can be caused by CMV, VZV, HSV (immuno-compromised)



Inga Zerr & Peter Hermann, 2018

CLINICAL UPDATE

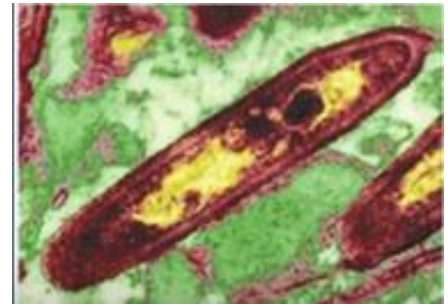
Ann Neurol 2008;64:97–108



Rapidly Progressive Dementia

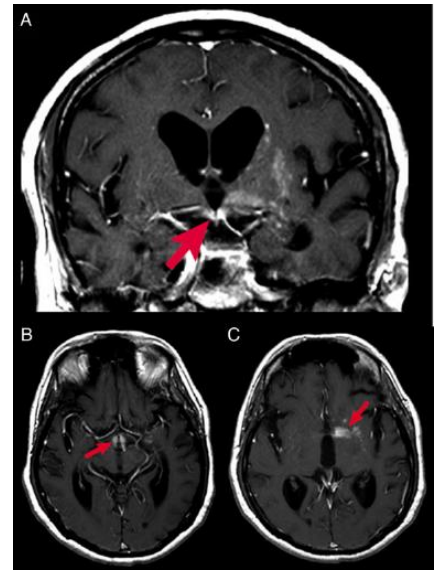
Michael D. Geschwind, MD, PhD,[1] Huidy Shu, MD, PhD,[1] Aissa Haman, MD,[1] James J. Sejvar, MD,[2] and Bruce L. Miller, MD[1]

Whipple's disease is a rare bacterial (*Tropheryma whippelii*) infection that often begins as a malabsorption syndrome, but 5% of cases start as a neurological syndrome with dementia, movement disorder (myorrhythmia and ataxia), or psychiatric signs. The triad of dementia, ophthalmoplegia, and myoclonus occurs in only 10% of cases, but this combination strongly suggests Whipple's disease, whereas oculomasticatory myorrhythmia is pathognomonic. Diagnosis is made by the demonstration of periodic acid-Schiff–positive inclusions, *T. whippelii* on jejunal biopsy, or polymerase chain reaction from jejunal biopsy or CSF.[106]



***Tropheryma* whipplei**

batterio bastoncelliforme



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Teaching NeuroImage: Oculomasticatory myorhythmia

Pathognomonic phenomenology of Whipple disease

Fredy J. Revilla, MD
Rafael de la Cruz, MD
Nancy Khardori, MD, FACP
Alberto J. Espay, MD, MSc

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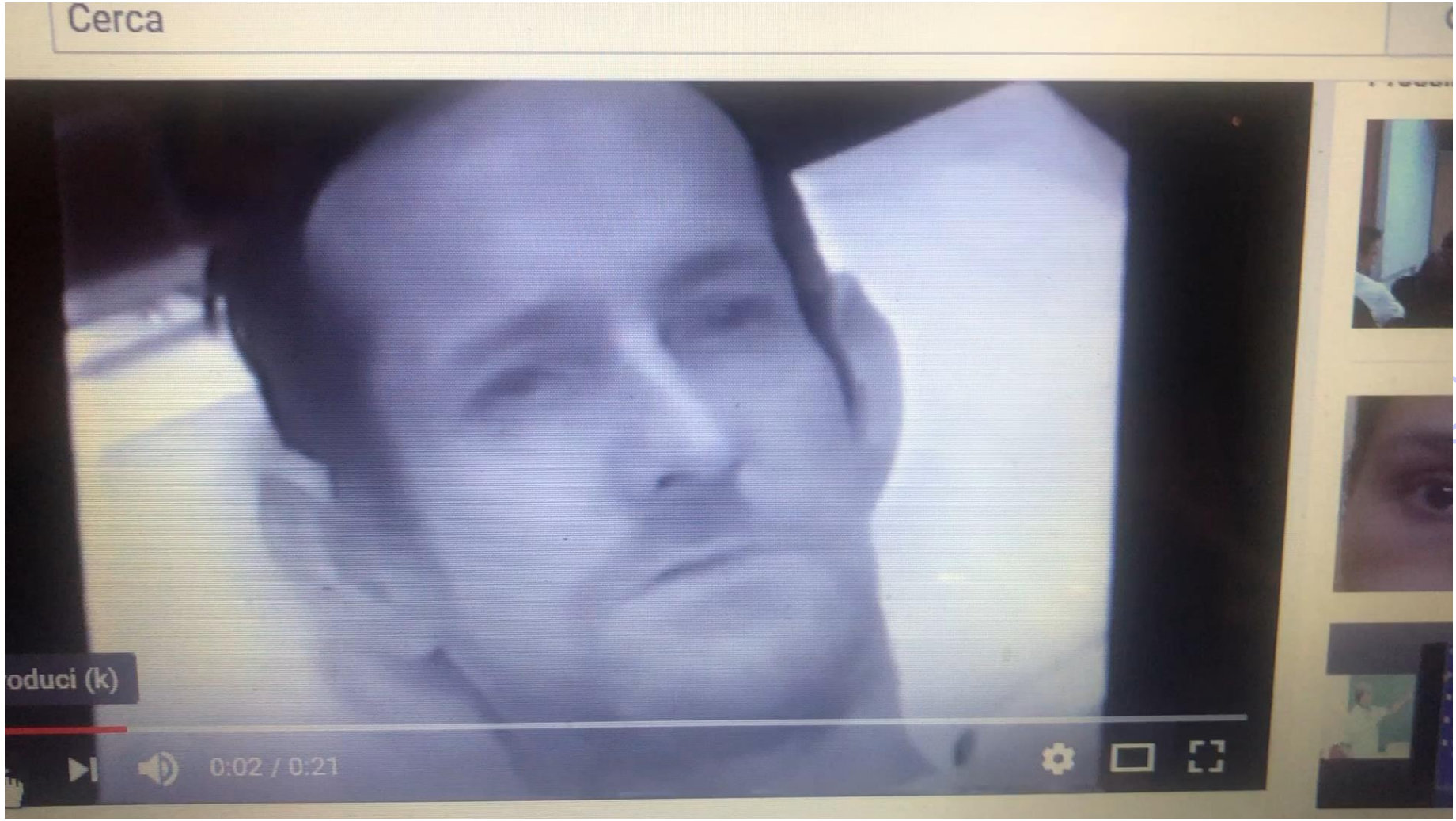
alberto.espay@uc.edu

A 41-year-old patient developed diplopia, imbalance, and weight loss. Examination showed pendular vergence oscillations of the eyes and synchronous contractions of the masticatory but not palatal muscles, i.e., oculomasticatory myorhythmia (OMM; figure). There was complete supranuclear vertical and, to a lesser extent, horizontal gaze palsy. The remainder of the examination was unremarkable. Brain MRI was normal. OMM is pathognomonic of Whipple disease.[1] In its presence, neither jejunal biopsy nor blood or CSF PCR of *Tropheryma whippelii* is necessary for the initiation of trimethoprim-sulfamethoxazole.[2] This patient became symptom free after 6 months of treatment. Video footage of the typical presentation should assist clinicians in recognizing this highly treatable neurologic disorder.

REFERENCES

1. Schwartz MA, Selhorst JB, Ochs AL, et al. Oculomasticatory myorhythmia: a unique movement disorder occurring in Whipple's disease. Ann Neurol 1986;20:677–683.
2. Louis ED, Lynch T, Kaufmann P, Fahn S, Odel J. Diagnostic guidelines in central nervous system Whipple's disease. Ann Neurol 1996;40:561–568.



The patient is a 46 year old man who, over a period of six months, lost the ability to read and complained of excessive somnolence, occasional urinary incontinence, and recent 6-kg weight loss. A disabling, seronegative, non-deforming migratory arthritis had developed three years earlier. Gastrointestinal symptoms and fever had not occurred.

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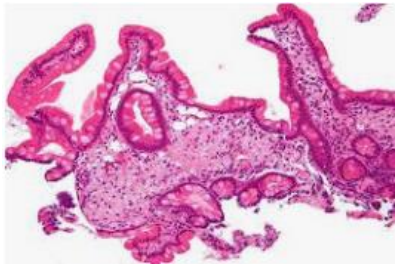
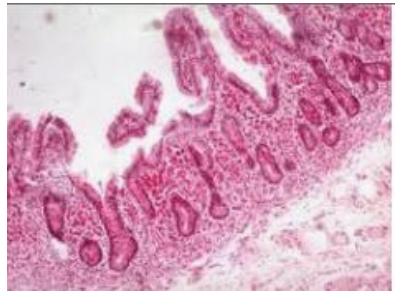
Home > Contents > Infectious Disorders Center > Article

Peter K Panegyres MD PhD et al, 2018

Diagnosi: biopsia duodeno-digiunale
PCR: sequenza 16SRNA





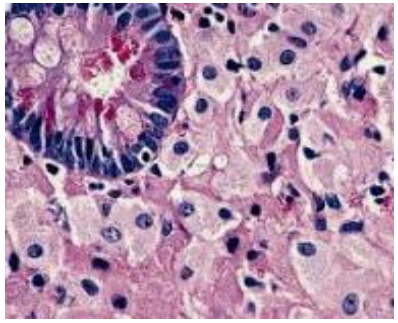


Table 4. Antibiotic Guidelines to Treat Whipple Disease of the Brain

A. (i) Ceftriaxone (2 g IV daily) or meropenem (1 g IV 3 times daily) for 2 weeks

(ii) Trimethoprim-sulfamethoxazole (trimethoprim 160 mg + sulfamethoxazole 800 mg) 2 tablets orally twice daily [indefinite]

(iii) Folinic acid (15 mg orally daily) [indefinite to combat the antifolate effect of (ii)

B. If allergic or lack of response:

(i) Streptomycin (2 g IV daily) + penicillin G (1.2 x 106 units IV daily) for 2 weeks, then

(ii) Doxycycline (100 mg orally twice daily) [indefinite] or

(iii) Hydroxychloroquine (200mg orally twice daily) [indefinite]

C. If allergic or lack of response:

Consider chloramphenicol or cefepime intravenously

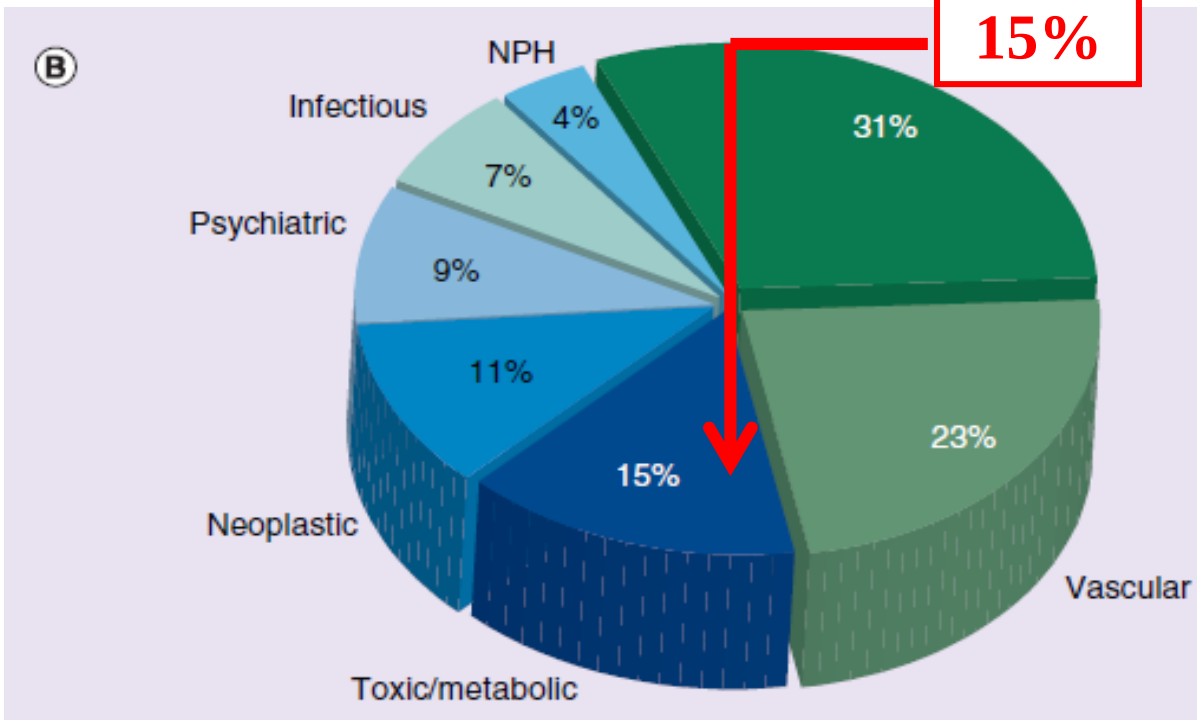
Etiology of RPDs: use of the mnemonic VITAMINS

Ross W. Paterson et al, 2012

V Vascular

I Infectious

T Toxic-metabolic



Gregory S Day & D.F. Tang-Wai, 2014

Etiology of RPDs

Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment
Toxic-metabolic							
Wernicke syndrome	A	Risk factors: alcoholism, malnutrition	Cognitive impairment, eye movement abnormalities, ataxia	T2 hyper in medial thalamus and mammillary bodies[e7]	Nondiagnostic	—	Thiamine

Neurology: Clinical Practice ▮ September 2012

Ross W. Paterson et al, 2012

Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management

GianPietro Sechi, Alessandro Serra

Lancet Neurol 2007; 6: 442–55

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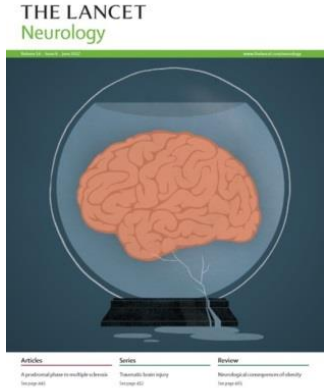


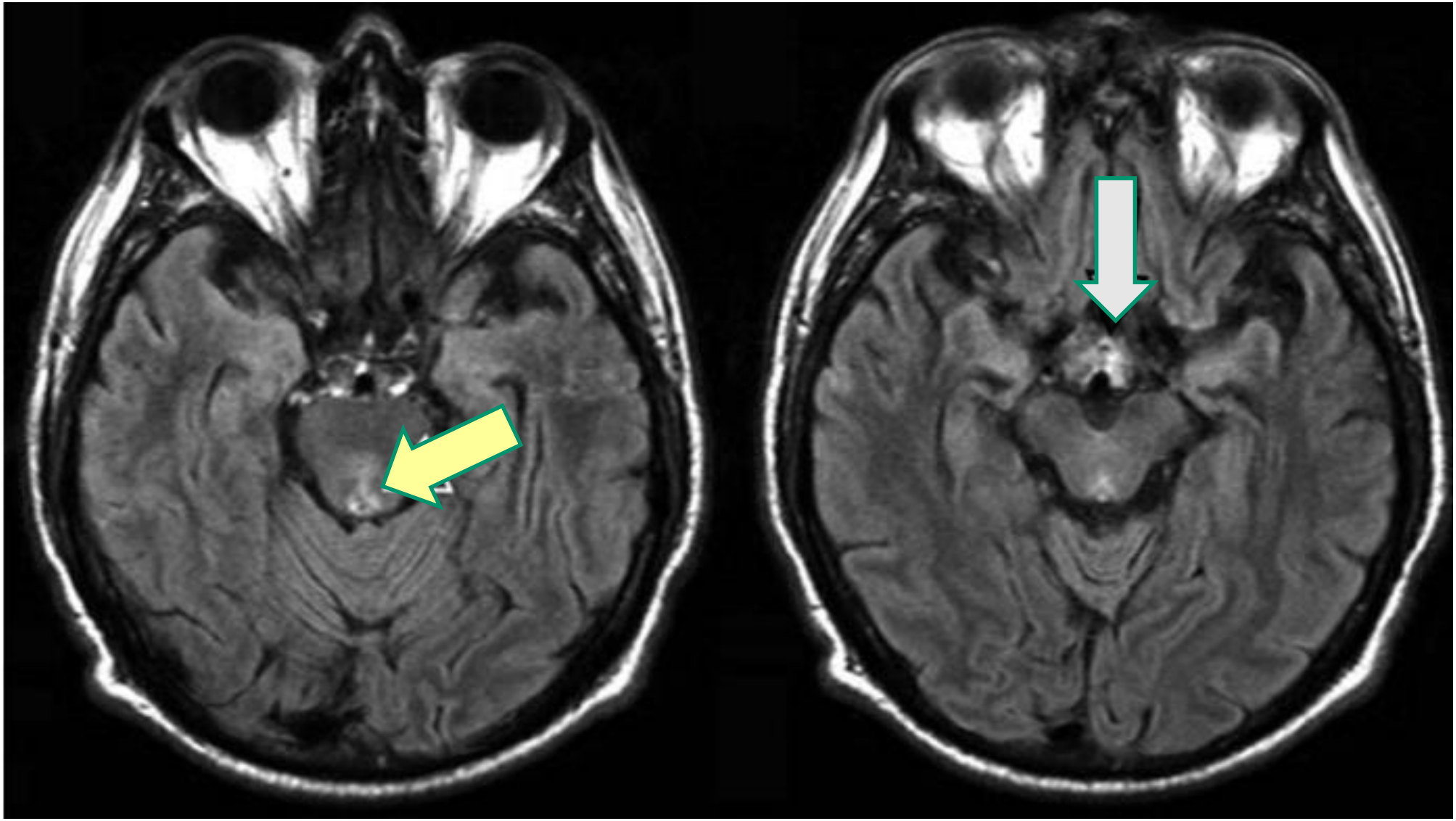
Panel 1: Clinical features of Wernicke's encephalopathy

Common symptoms or signs at presentation
Ocular abnormalities
Mental status changes
Incoordination of gait and trunk ataxia

Uncommon symptoms or signs at presentation
Stupor
Hypotension and tachycardia
Hypothermia
Bilateral visual disturbances and papilloedema
Epileptic seizures
Hearing loss
Hallucinations and behavioural disturbances

Late-stage symptoms
Hyperthermia
Increased muscular tone and spastic paresis
Choreic dyskinesias
Coma

Wernicke's encephalopathy: RMN sequenze assiali FLAIR



sostanza grigia periacqueduttale

corpi mammillari

Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management

GianPietro Sechi, Alessandro Serra

Review

Lancet Neurol 2007; 6: 442–55

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Sechi GP et al, 2007

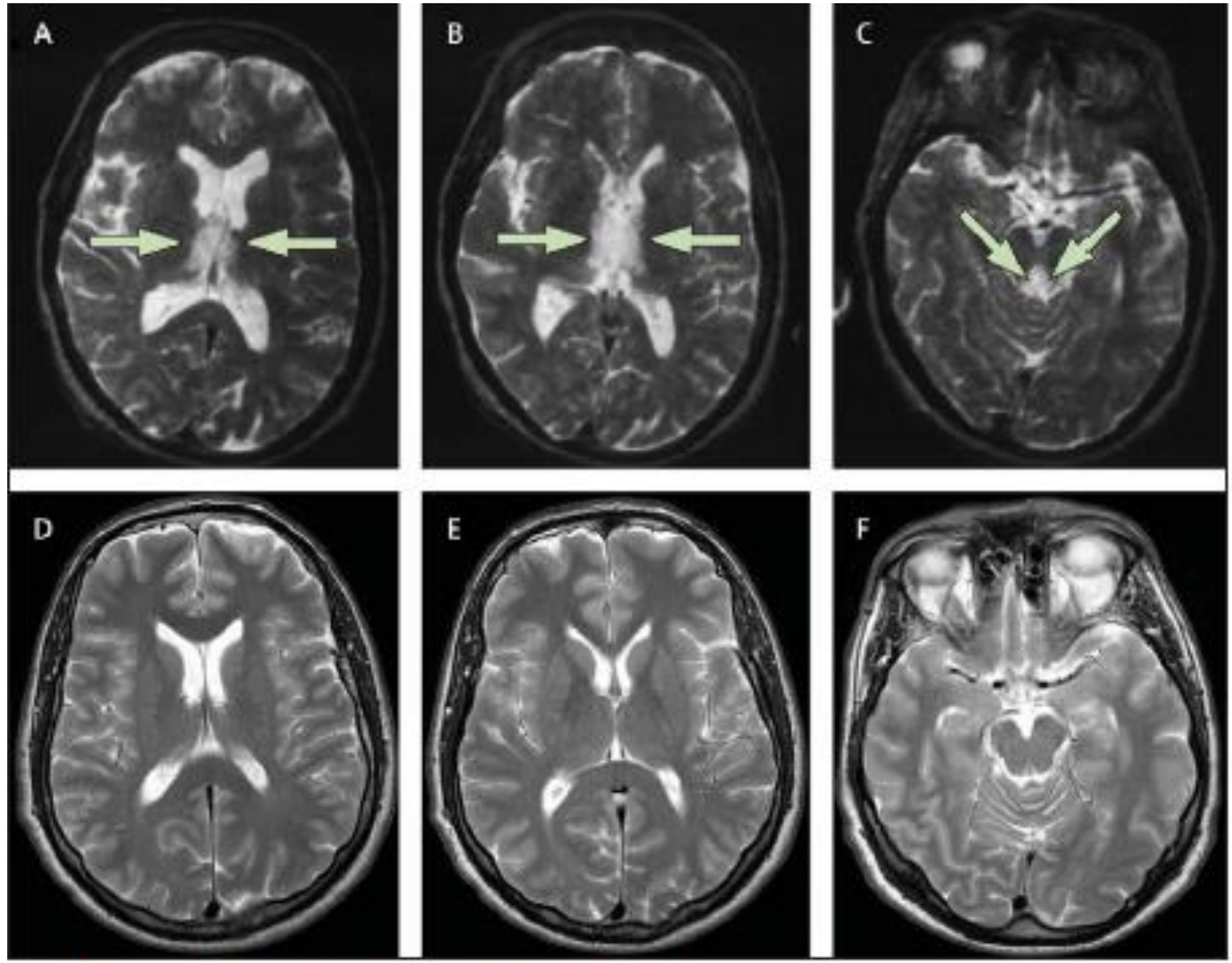
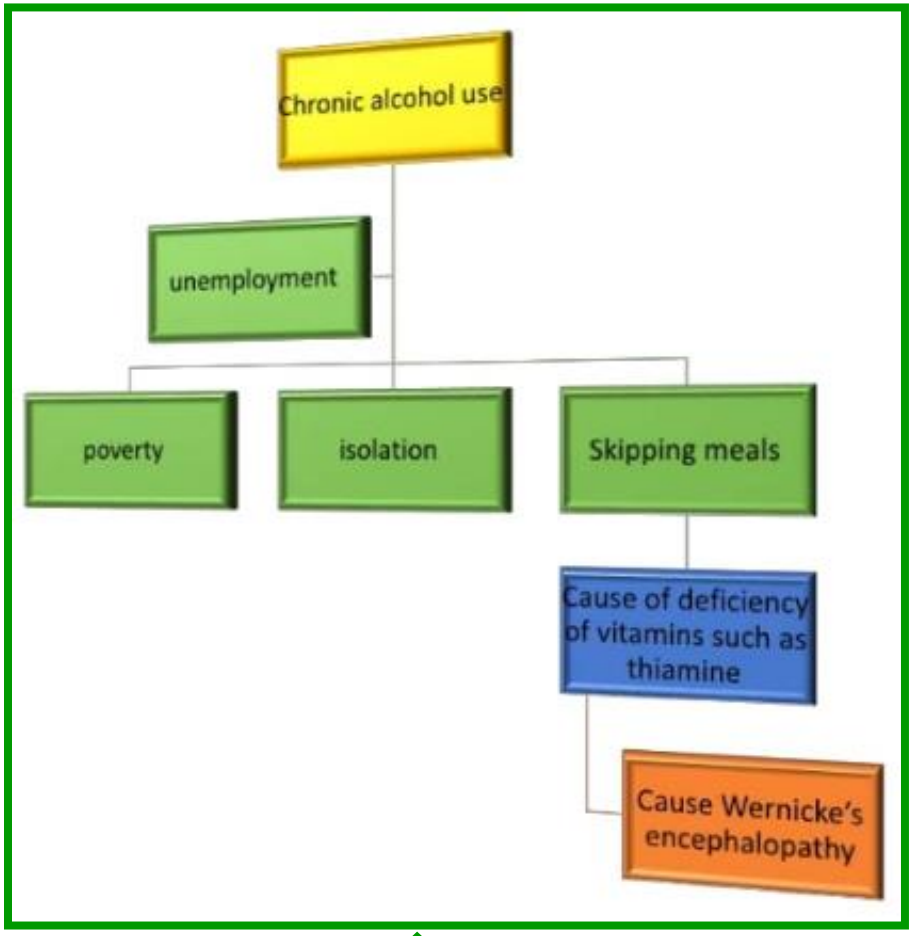
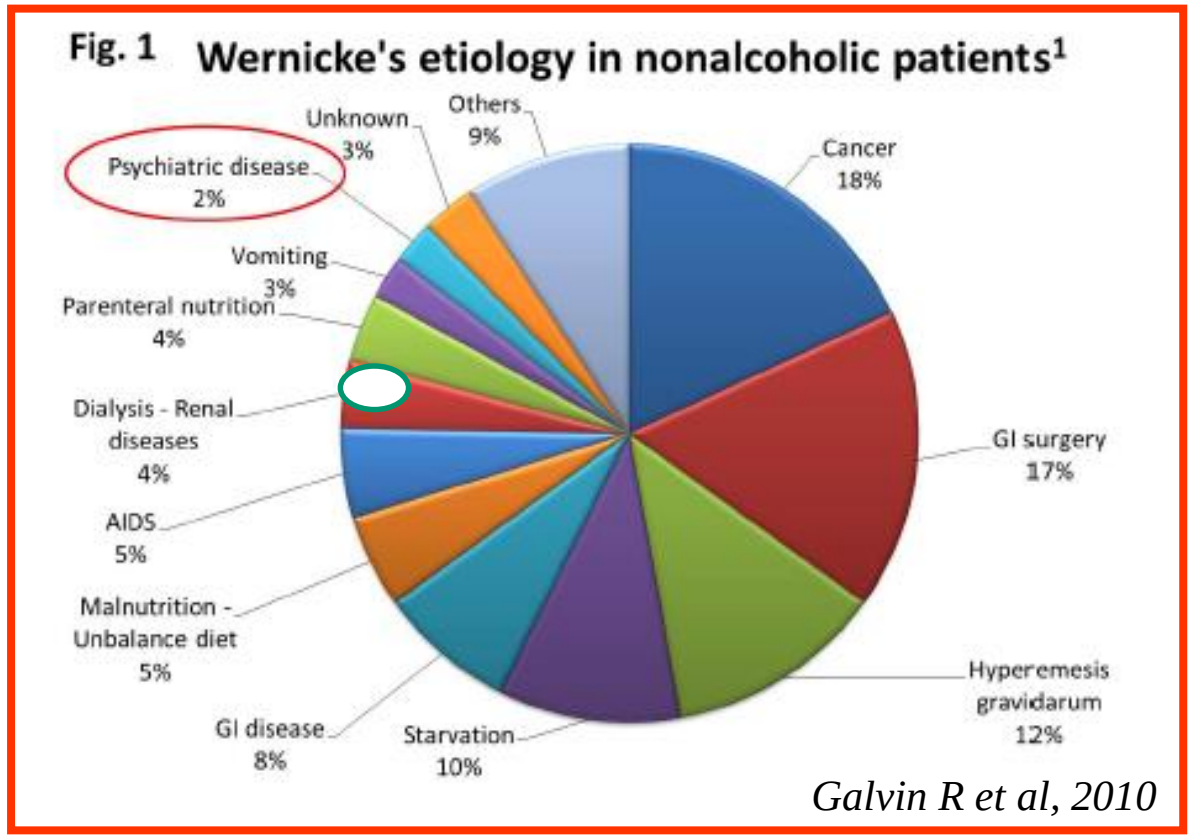


Figure 4: **T2-weighted axial MRI in Wernicke's encephalopathy**
Symmetrical high-intensity lesions in the medial thalami (A, B), as well as in the periaqueductal grey matter of the midbrain (C), are evident in a patient about 2 weeks after onset of neurological symptoms. MRI of a healthy person (D, E, F).



Alcoholic Wernicke's encephalopathy

Non-alcoholic Wernicke's encephalopathy



Galvin R et al, 2010

Therapeutic Apheresis and Dialysis

Wernicke–Korsakoff Syndrome in End-Stage Renal Disease: A Case Report

Ramona Nicotera,[1] Giuseppe Leonardi,[1] Alberto Castagna,[2] Valeria Cernaro,[3] Giuseppe Coppolino[1] and "Pugliese-Ciaccio" Hospital of Catanzaro collaborators*
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*"Pugliese-Ciaccio" Hospital of Catanzaro Collaborators: Salvatore Mileti, Maria Teresa Panzino, Salvatore Chiarella, Giordano Placida, Antonio Panzino, Luciano De Paola, Maria Antonia Panzino, Cinzia Donato, Emilio Le Piane, Antonio Sergi, and Luigi Santaguida

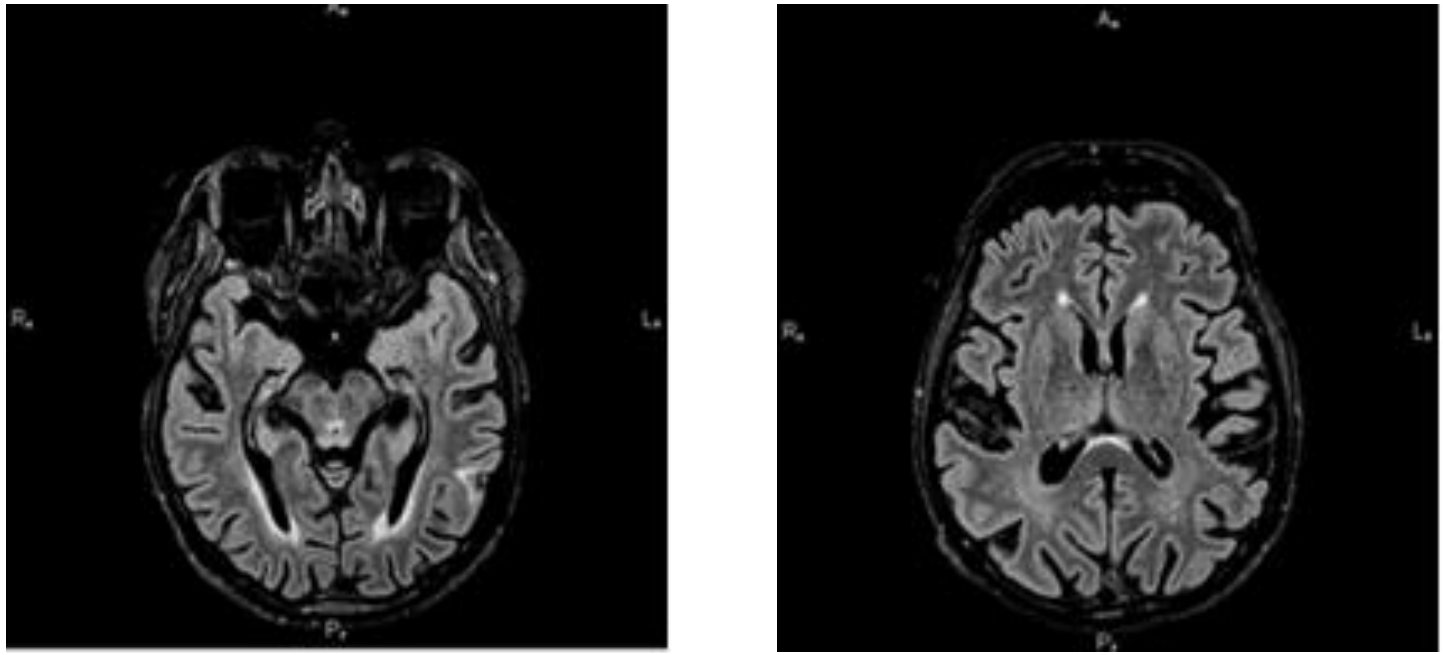

FIG. 1. MRI showing hyperintensities of back-medial thalami and periaqueductal membranes compatible with Wernicke's encephalopathy.

Volume 22, Issue 6
December 2018
Pages 676-679

Nicotera R et al, 2018

Tumori, 93: 207-209, 2007

WERNICKE'S ENCEPHALOPATHY AFTER ALLOGENEIC STEM CELL TRANSPLANTATION

Giuseppe Messina[1], Eugenia Quartarone[1], Giuseppe Console[1], Giuseppe Irrera[1], Massimo Martino[1], Giulia Pucci[1], Emilio Le Piane[2], Damiano Branca[2], Francesco Cafarelli[3], Antonio Gangemi[3], and Pasquale Iacopino[1]

[1]Centro Trapianti Midollo Osseo "A. Neri", [2]Divisione di Neurologia, [3]Unità Operativa di Neuroradiologia, Azienda Ospedaliera "Bianchi-Melacrino-Morelli", Reggio Calabria, Italy

Wernicke's encephalopathy is an acute neuropsychiatric condition due to thiamine deficiency frequently associated with chronic alcohol abuse. We describe 2 cases of patients who experienced acute Wernicke's encephalopathy after allogeneic stem cell transplantation associated with the use of commercial total parental nutrition. Early diagnosis with magnetic resonance imaging and timely treatment with thiamine resulted in rapid resolution of clinical and radiological signs. In conclusion, the prolonged use of commercial total parental nutrition formulas must be supplemented with thiamine in the form of intramuscularly administered multivitamins.

Key words: allogeneic stem cell transplantation, thiamine, total parental nutrition, Wernicke's encephalopathy.

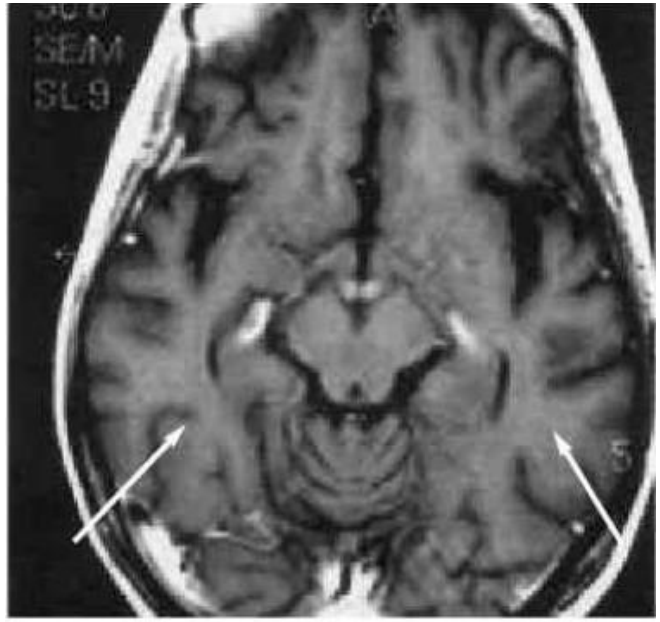

Figure 2 - Hyperintense lesions in mammillary body after gadolinum infusion in case 1.

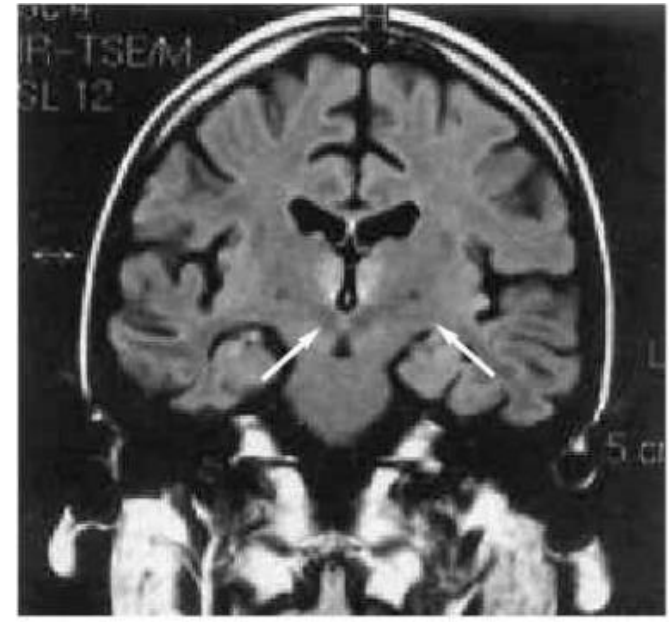

Figure 1 - Evidence of lesions in thalamic region of case 1.

Differences Between Alcoholic and Nonalcoholic Patients With Wernicke Encephalopathy: A Multicenter Observational Study

Antonio J. Chamorro, PhD[a], Beatriz Rosón-Hernández, PhD[b], José-A. Medina-García, MD[c], Roberto Muga-Bustamante, PhD[d], Joaquín Fernández-Solá, PhD[e], M.-Candelaria Martín-González, MD[f], Elena Seco-Hernández, MD[g], Ignacio Novo-Veleiro, PhD[h], Carlos Suárez-Cuervo, PhD[i], Ana M. Mateos-Díaz, MD[a], Rafael Monte-Secades, PhD[j], Begoña Machado-Prieto, MD[k], Rubén Puerta-Louro, PhD[l], Cristina Prada-González, MD[m], Álvaro Fernández-Rial, MD[n], Patricia Sabio-Repiso, MD[o], Rocío Vázquez-Vigo, MD[p], Ana-C. Antolí-Royo, PhD[q], Aina Gomila-Grange, MD[b], Nieves-C. Felipe-Pérez, MD[c], Arantza Sanvisens-Bergé, MS[d], Emilia Antúnez-Jorge, PhD[e], Camino-M. Fernández-Rodríguez, MD[f], Lucía Alvela-Suárez, MD[h], Alba Fidalgo-Navarro, MD[i], Miguel Marcos, PhD[a,*] on behalf of the ⊞ Wernicke-SEMI Group , Alcohol and Alcoholism Group, Spanish Society of Internal Medicine (SEMI)

MAYO CLINIC PROCEEDINGS

June 2017 Volume 92, Issue 6, Pages 899–907

Results

Among the 468 patients, the most common risk factor was alcoholism (n=434 [92.7%]). More than one-third of patients (n=181 [38.7%]) had the classic WE triad of symptoms (ocular signs, cerebellar dysfunction, and confusion). Among 252 patients for whom magnetic resonance imaging data were available, 135 (53.6%) had WE-related lesions and 42 (16.7%) had cerebellar lesions. Of the 468 patients, 25 (5.3%) died during hospitalization. Alcoholic patients presented more frequently than nonalcoholic patients with cerebellar signs (P=.01) but less frequently with ocular signs (P=.02). Alcoholic patients had a significantly higher frequency of hyponatremia (P=.04) and decreased platelet count (P=.005) compared with nonalcoholics. Alcoholic patients were diagnosed earlier than nonalcoholics (median time to diagnosis, 1 vs 4 days; P=.001) and had shorter hospitalizations (13 vs 23 days; P=.002).

Conclusion

Compared with nonalcoholic patients, alcoholic patients with WE are more likely to present with cerebellar signs and less likely to have ocular signs. Diagnosis may be delayed in nonalcoholic patients. Mortality in the present series was lower than described previously.

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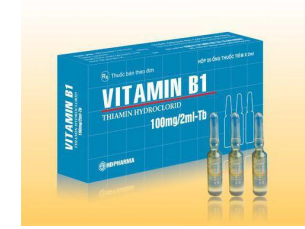
Neeraj Kumar al, 2019



A commonly used thiamine replacement regimen is 100 mg intravenously every 8 hours for 3 to 5 days (Foster et al 2005; Latt and Dore 2014; Flynn et al 2015). A Cochrane review suggests that clinically significant thiamine deficiency be treated with at least 200 mg of parenteral thiamine daily for 2 days (Day et al 2004). Higher doses may be required in Wernicke encephalopathy, particularly when Wernicke encephalopathy occurs in the setting of alcoholism (Thomson and Marshall 2006b). Lower doses may suffice with predominantly nutritional thiamine deficiency. It has been suggested that patients with signs of Wernicke encephalopathy should receive 500 mg thiamine hydrochloride (diluted in 100 mL of normal saline) by infusion over 30 minutes, 3 times a day, for 2 to 3 days (Cook et al 1998; Hope et al 1999; Cook 2000; Thomson 2000; Thomson et al 2002; Thomson and Marshall 2006a; Thomson and Marshall 2006b; Sechi and Serra 2007). Subsequently, the dose may be reduced to 250 mg of thiamine given intravenously or intramuscularly daily for 3 to 5 days. Similar doses may be given prophylactically in conditions of alcohol withdrawal or severe malnutrition. The parenteral form is used when there is doubt about adequate gastrointestinal absorption. Long-term oral maintenance with 50 to 100 mg thiamine is commonly employed. Higher

tiamina
schema posologico

1500/die

↓

250/die

↓

100/die

1. Treatable causes of adult-onset rapid **cognitive impairment**.
Wang S, Zhang J, Liang J, Song H, Ji X.
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Pennisi M, Malaguarnera G, Bartolo GD, Lanza G, Bella R, Chisari EM, Cauli O, Vicari E, Malaguarnera M.
Nutrients. 2019 Oct 20;11(10). pii: E2531. doi: 10.3390/nu11102531.
PMID: 31635199 **Free Article**
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4. Dietary intakes and biomarker patterns of folate, **vitamin** B_6, and **vitamin** B_{12} can be associated with **cognitive impairment** by hypermethylation of redox-related genes NUDT15 and TXNRD1.
An Y, Feng L, Zhang X, Wang Y, Wang Y, Tao L, Qin Z, Xiao R.
Clin Epigenetics. 2019 Oct 11;11(1):139. doi: 10.1186/s13148-019-0741-y.
PMID: 31601260 **Free PMC Article**
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5. Assessment of the Dose-Response Relationship Between Folate Exposure and **Cognitive Impairment**: Synthesizing Data from Documented Studies.

Metabolic

1. Hyponatremia and other electrolyte disturbances
2. Wernicke's encephalopathy (thiamine) and other vitamin deficiency syndromes (e.g. B12, folate, niacin, biotin)
3. Hepatic encephalopathy
4. Uremic encephalopathy
5. Hypothyroidism
6. Hypoparathyroidism
7. Porphyria
8. Adrenal insufficiency
9. Extrapontine myelinolysis

Inga Zerr & Peter Hermann, 2018

ORIGINAL CONTRIBUTION

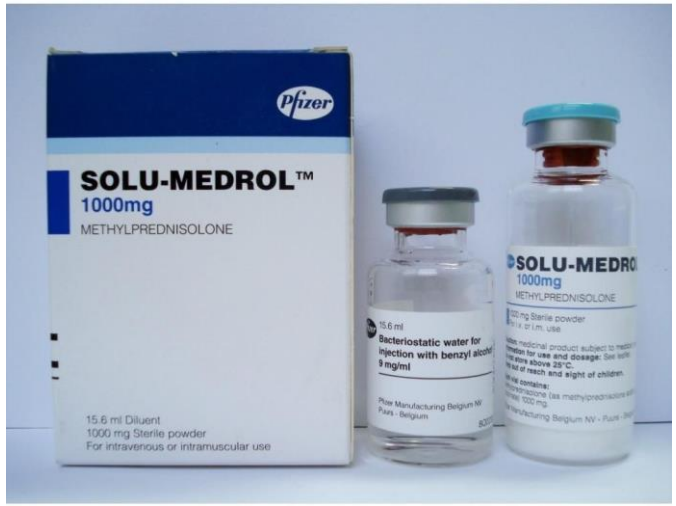
ARCHIVES OF NEUROLOGY

Arch Neurol. 2006;63:197-202

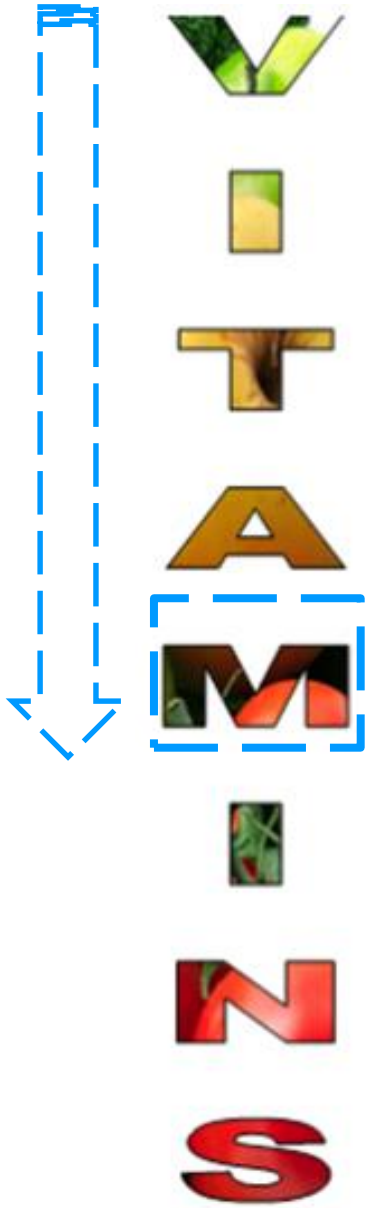
Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Pablo Castillo, MD; Bryan Woodruff, MD; Richard Caselli, MD; Steven Vernino, MD, PhD; Claudia Lucchinetti, MD; Jerry Swanson, MD; John Noseworthy, MD; Allen Aksamit, MD; Jonathan Carter, MD; Joseph Sirven, MD; Gene Hunder, MD; Vahab Fatourechi, MD; Bahram Mokri, MD; Daniel Drubach, MD; Sean Pittock, MD; Vanda Lennon, MD, PhD; Brad Boeve, MD

SREAT

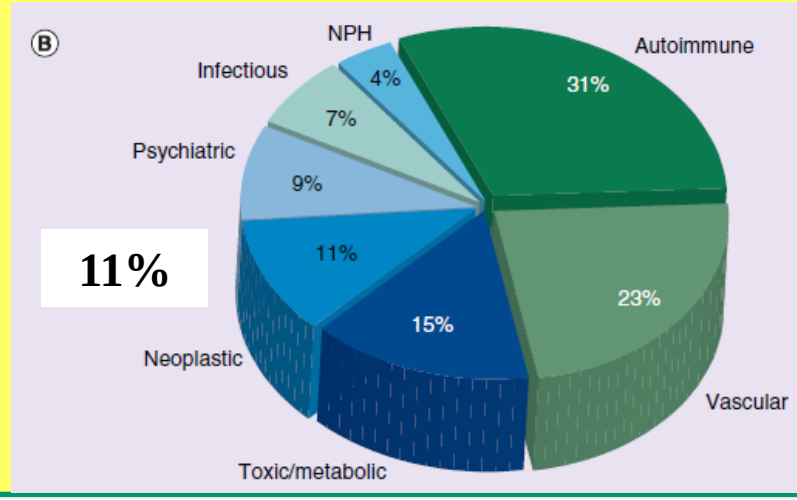


Etiology of RPDs: use of the mnemonic VITAMINS



Ross W. Paterson et al, 2012

V	Vascular
I	Infectious
T	Toxic-metabolic
A	Autoimmune
M	**Metastases/neoplam**



Gregory S Day et al, 2014

Etiology of RPDs

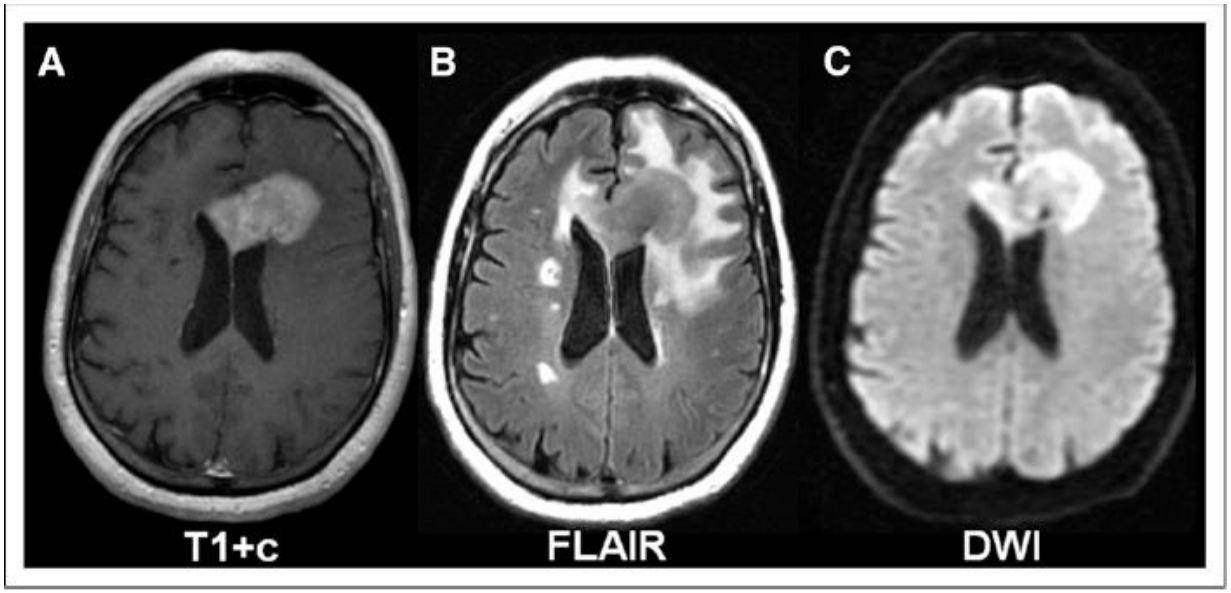
CNS neoplasia

1. CNS lymphoma
2. Solid neoplasia (primary in the CNS or metastatic)
3. Diffuse gliomatosis
4. Leptomeningeal spread of other tumors

Neurology: Clinical Practice ▮ September 2012

Ross W. Paterson et al, 2012

Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment
Metastasis/neoplasia							
Primary CNS lymphoma	S	Most 50–70 years	Neuropsychiatric symptoms, focal neurologic deficits, seizures	Focal hypo or hyper T2 lesions with CE; seldom DWI hyper[a15]	Lymphocytic pleocytosis; flow cytometry for lymphoma cells	High LDH, ESR; biopsy	Specific lymphoma treatment
Gliomatosis cerebri	S	Older adults	AMS, dementia, seizures, headache, focal deficits	T2/FLAIR hyper in 2+ lobes; ± mass effect; ± CE[a16]	—	Brain biopsy	Radiation ± chemotherapy



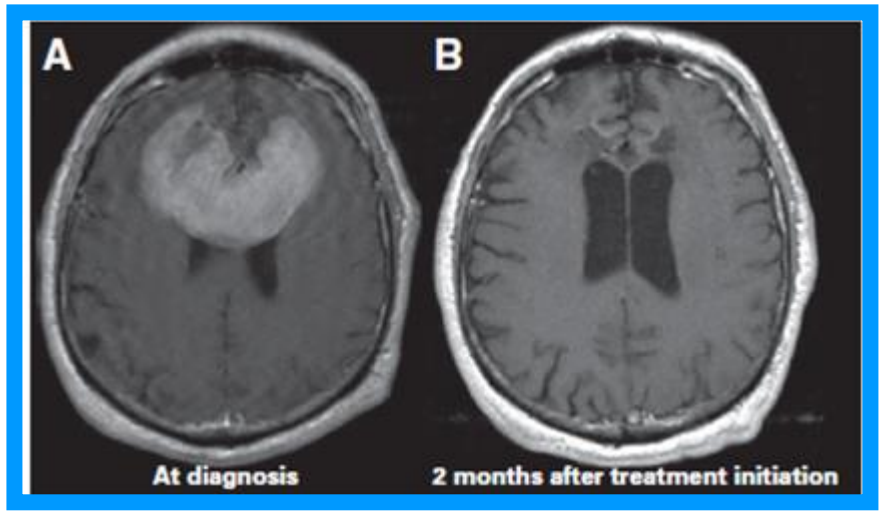


Fig 3. PCNSL is highly chemosensitive. (A) Magnetic resonance imaging (T1+ gadolinium) demonstrates a large, frontal-enhancing brain lesion. (B) Follow-up magnetic resonance imaging demonstrates resolution of the large lesion 2 months

CLINICA NPH
Disturbi della deambulazione

Cause:
- insufficiente input dalla corteccia sensitivo-motoria alla formazione reticolare
- Interessamento del tratto cortico-spinale che decorre in prossimità dei ventricoli laterali dilatati

- Primi sintomi a comparire e primi a risolversi dopo trattamento
- Il disturbo della deambulazione principale è l'"aprassia motoria": la postura eretta è instabile, con tendenza alla retropulsione. La marcia è caratterizzata da passi piccoli e strascicati (marcia magnetica)

Clinica NPH
deficit cognitivo

- Compare generalmente dopo il disturbo motorio
- Esordisce come disturbo di memoria a breve termine
- Disturbi del comportamento (abulia, apatia, disinteresse, isolamento, indifferenza, inerzia, rallentamento ideativo, depressione)
- Disturbi attentivi, difficoltà a pianificare
- Unica demenza potenzialmente reversibile dopo trattamento

Clinica NPH
disturbo urinario

- Esordio clinico con disturbi incompleti come pollachiuria, nicturia, urgenza minzionale che precedono per lungo tempo l'instaurarsi di una incontinenza vera e propria – dd ipertrofia prostatica
- Nelle fasi avanzate incontinenza urinaria e fecale
- Miglioramento significativo dopo derivazione liquorale

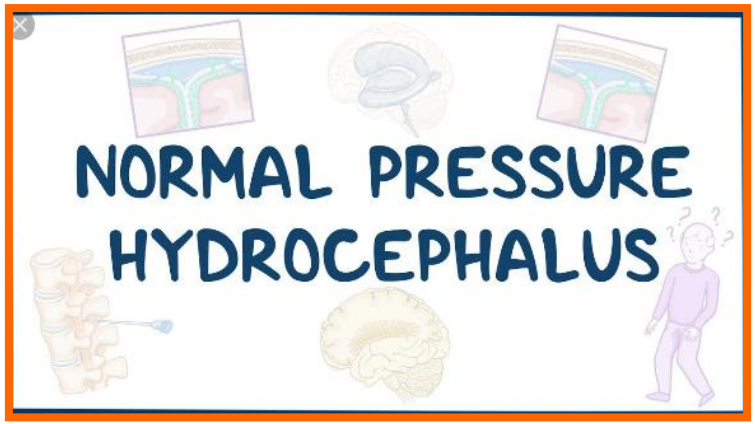


Table 3 Spectrum and frequency of secondary cognitive deterioration in current study

RPCD	Cases, n (%)	Time to cognitive deterioration (days)
Viral encephalitis	68 (41.5)	8.65±9.61
Bacterial encephalitis	2 (1.2)	3.00±1.41
Autoimmune encephalitis	28 (17.2)	29.5±48.02
Neurosyphilis	8 (4.9)	257.63±227.97
ADEM	2 (1.2)	18.50±16.26
Demyelination	4 (11.6)	71.25±72.84
Brain tumor	5 (2.4)	23.60±22.69
Toxic	19 (11.6)	30.53±66.32
Metabolic	13 (7.9)	25.38±40.48
Mitochondrial encephalopathy	6 (3.7)	10.00±10.99
Psychiatric	4 (2.4)	54.25±59.87
NPH	5 (3.0)	85.00±113.77
Total	164 (100)	33.74±81.57

Zhang Yun et al, 2017

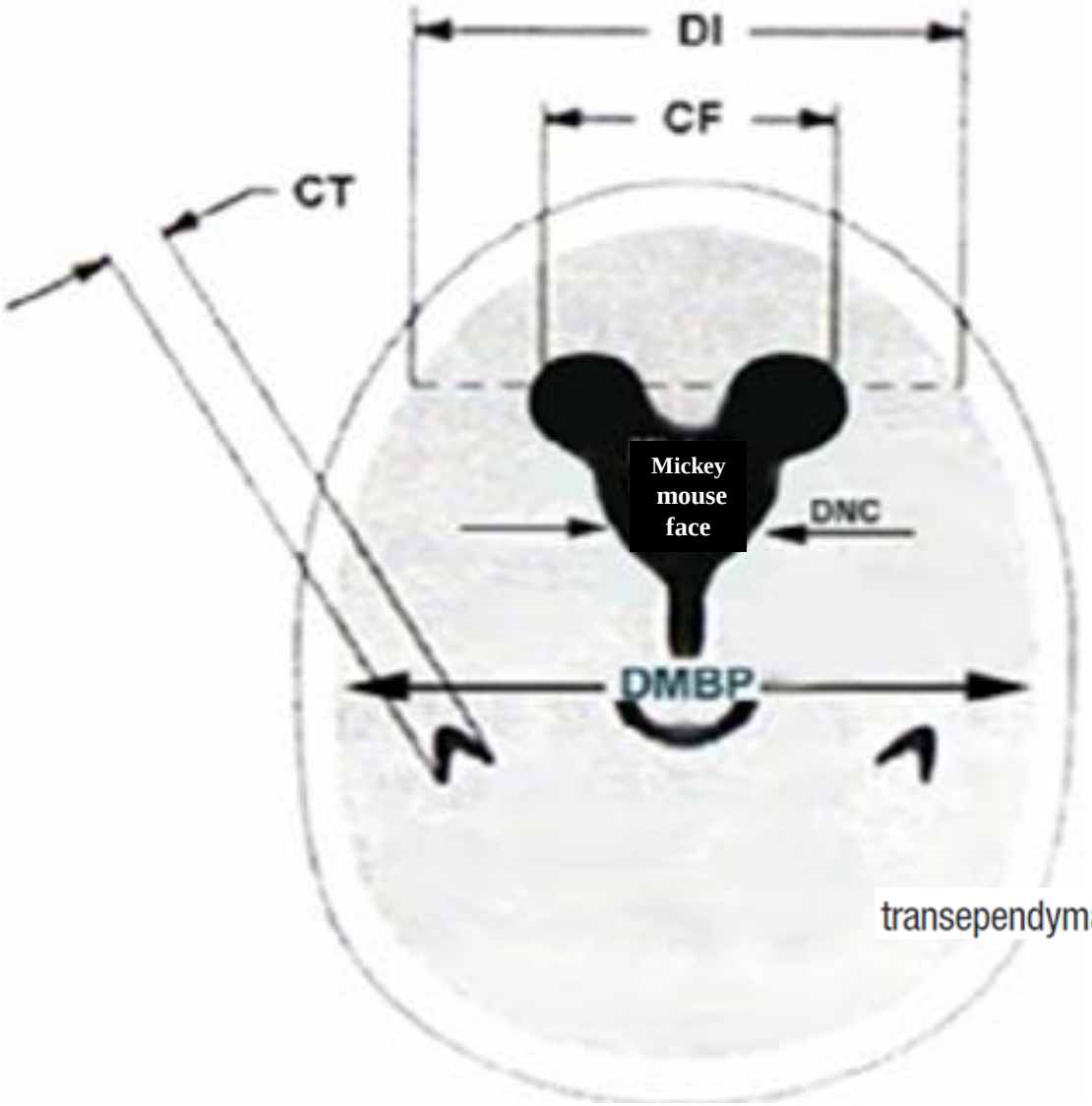
Normal-pressure hydrocephalus

A critical review

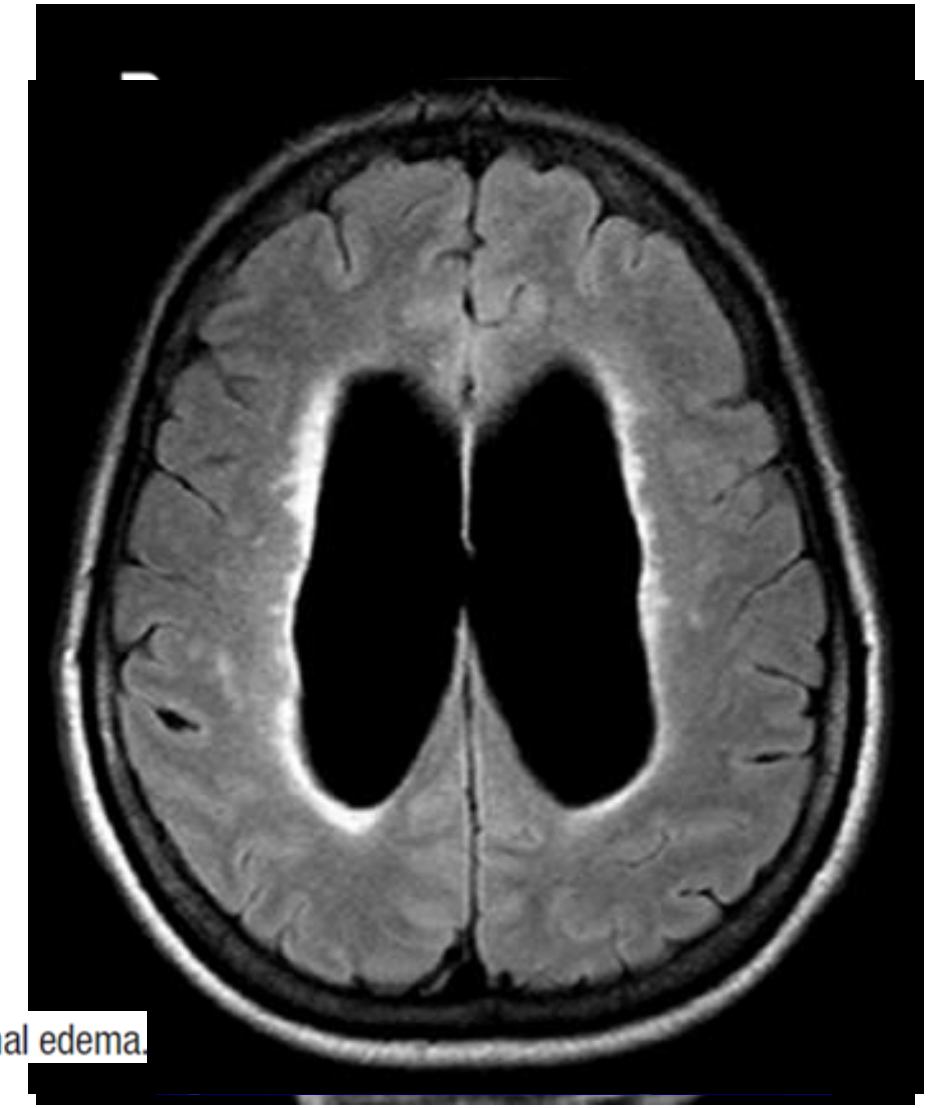
Louise Makarem Oliveira[1], Ricardo Nitrini[2], Gustavo C. Román[3]

Dement Neuropsychol 2019 June;13(2):133-143

Views & Reviews

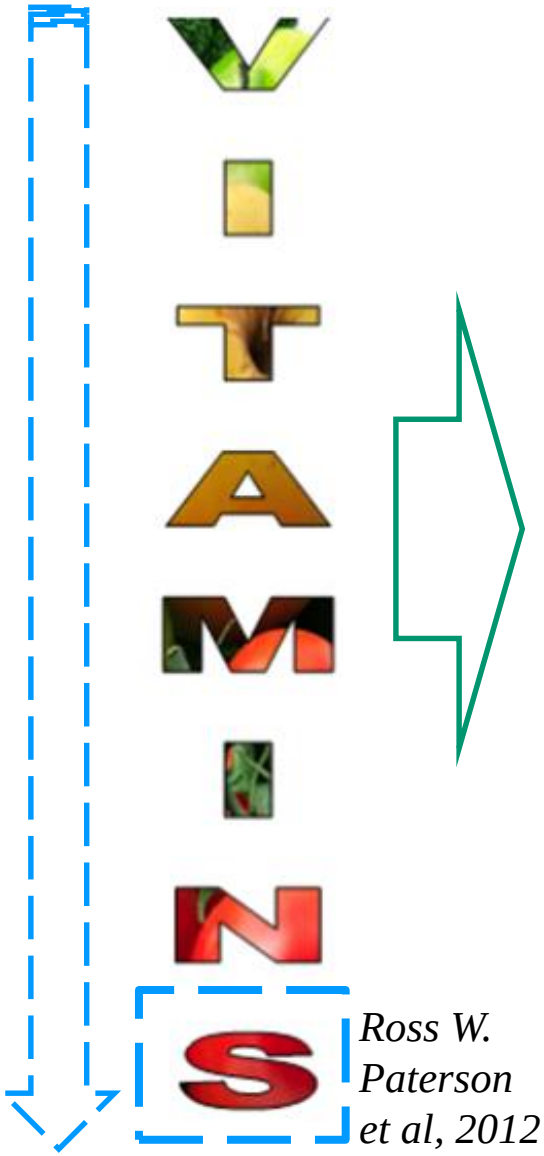


transepend**y**mal edema.



(B) T1-weighted coronal gadolinium-enhanced MRI scan showing reduced callosal angle.

Etiology of RPDs: use of the mnemonic VITAMINS



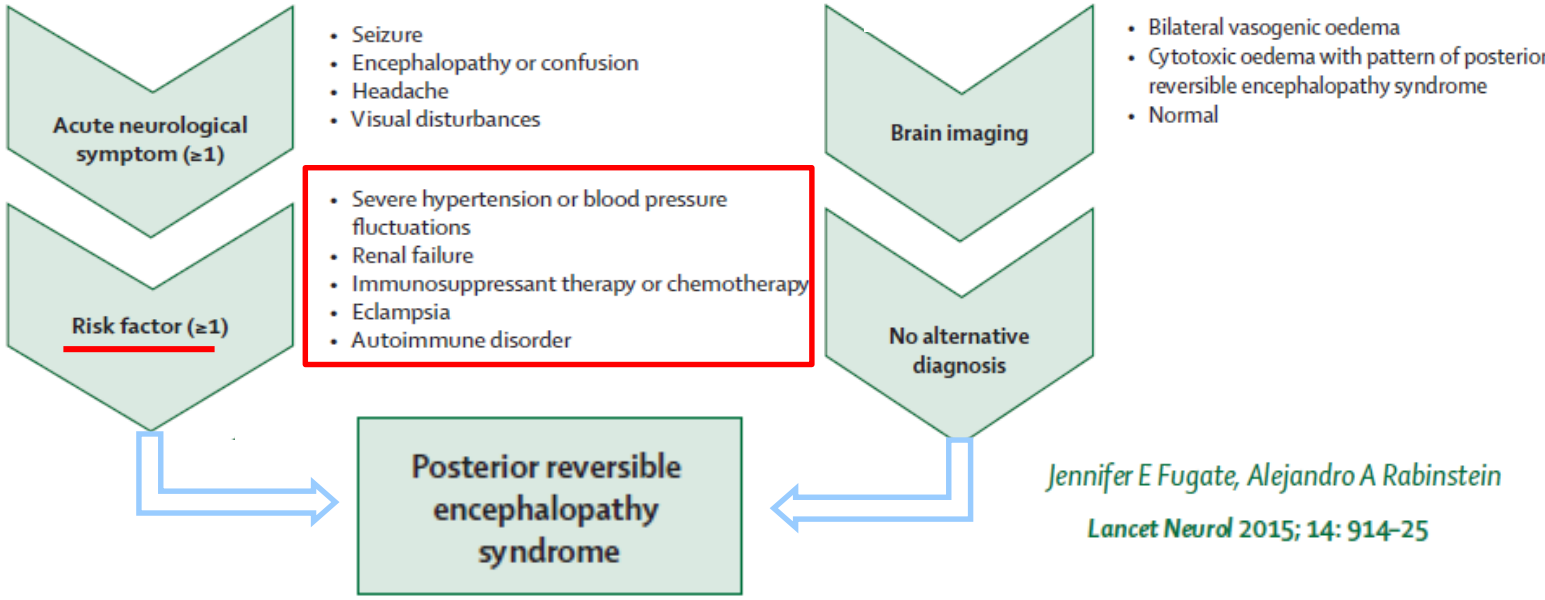
Ross W. Paterson et al, 2012

V	Vascular
I	Infectious
T	Toxic-metabolic
A	Autoimmune
M	Metastases/neoplam
I	Iatrogenic/inborn error of metabolism
N	Neurodegenerative
S	**Systemic/Seizures**

Etiology of RPDs

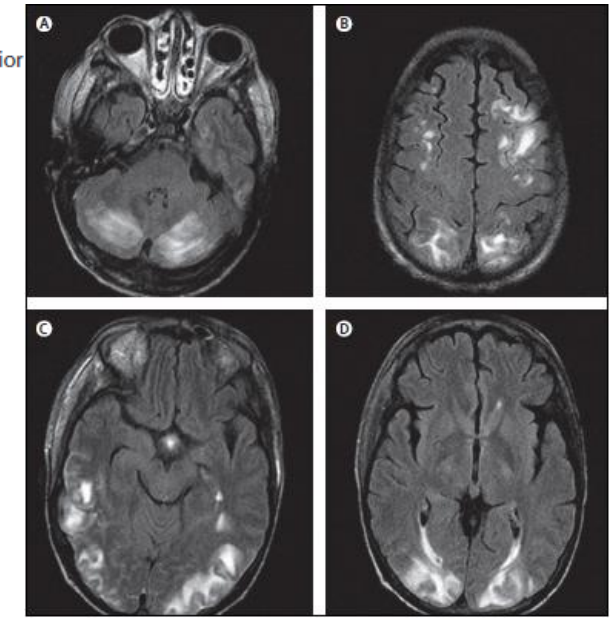
Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment
Neurology: Clinical Practice ▮ September 2012							
Systemic/seizures							
Hypertensive encephalopathy	A	Uncontrolled hypertension, eclampsia, chemotherapy	Headaches, confusion, visual changes, seizures, coma	FLAIR hyper in occipitoparietal WM	Nondiagnostic	—	Treatment of hypertension

Ross W. Paterson et al, 2012



Jennifer E Fugate, Alejandro A Rabinstein

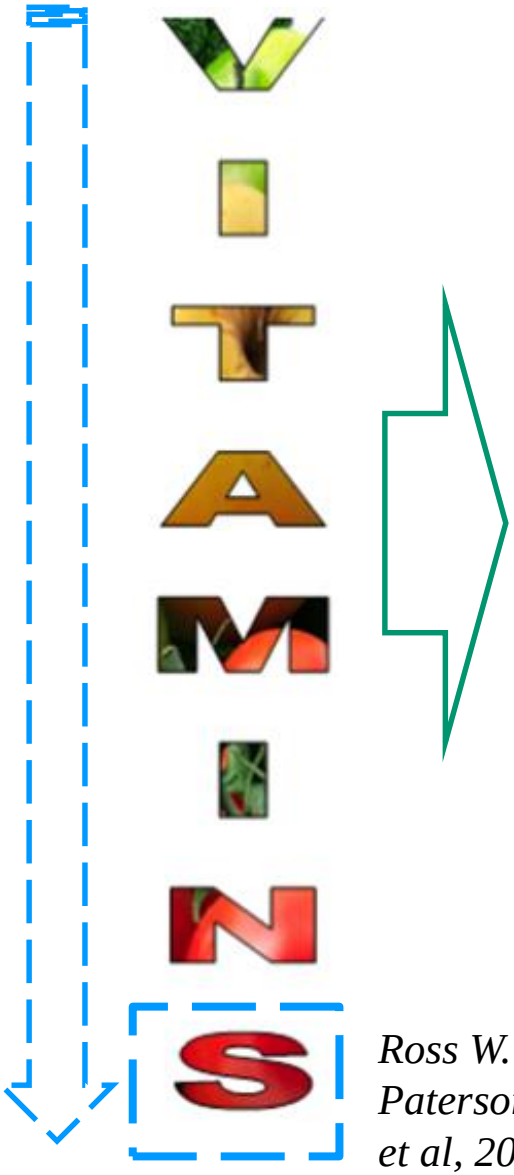
Lancet Neurol 2015; 14: 914–25



RMN sequenze assiali FLAIR

Figure 7: Proposed algorithm for the diagnosis of posterior reversible encephalopathy syndrome

Etiology of RPDs: use of the mnemonic VITAMINS



Ross W. Paterson et al, 2012

V	Vascular
I	Infectious
T	Toxic-metabolic
A	Autoimmune
M	Metastases/neoplam
I	Iatrogenic/inborn error of metabolism
N	Neurodegenerative
S	**Systemic/ Seizures (NCSE)**

Etiology of RPDs

Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment

Neurology: Clinical Practice ▮ September 2012

Ross W. Paterson et al, 2012

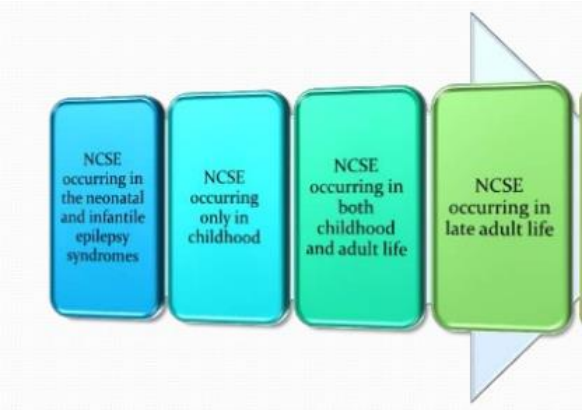
Seizures/NCSE	A	Older adults	Cognitive dysfunction, fluctuations in alertness	DWI hyper in cortical or	Might have mild pleocytosis	EEG	AEDs

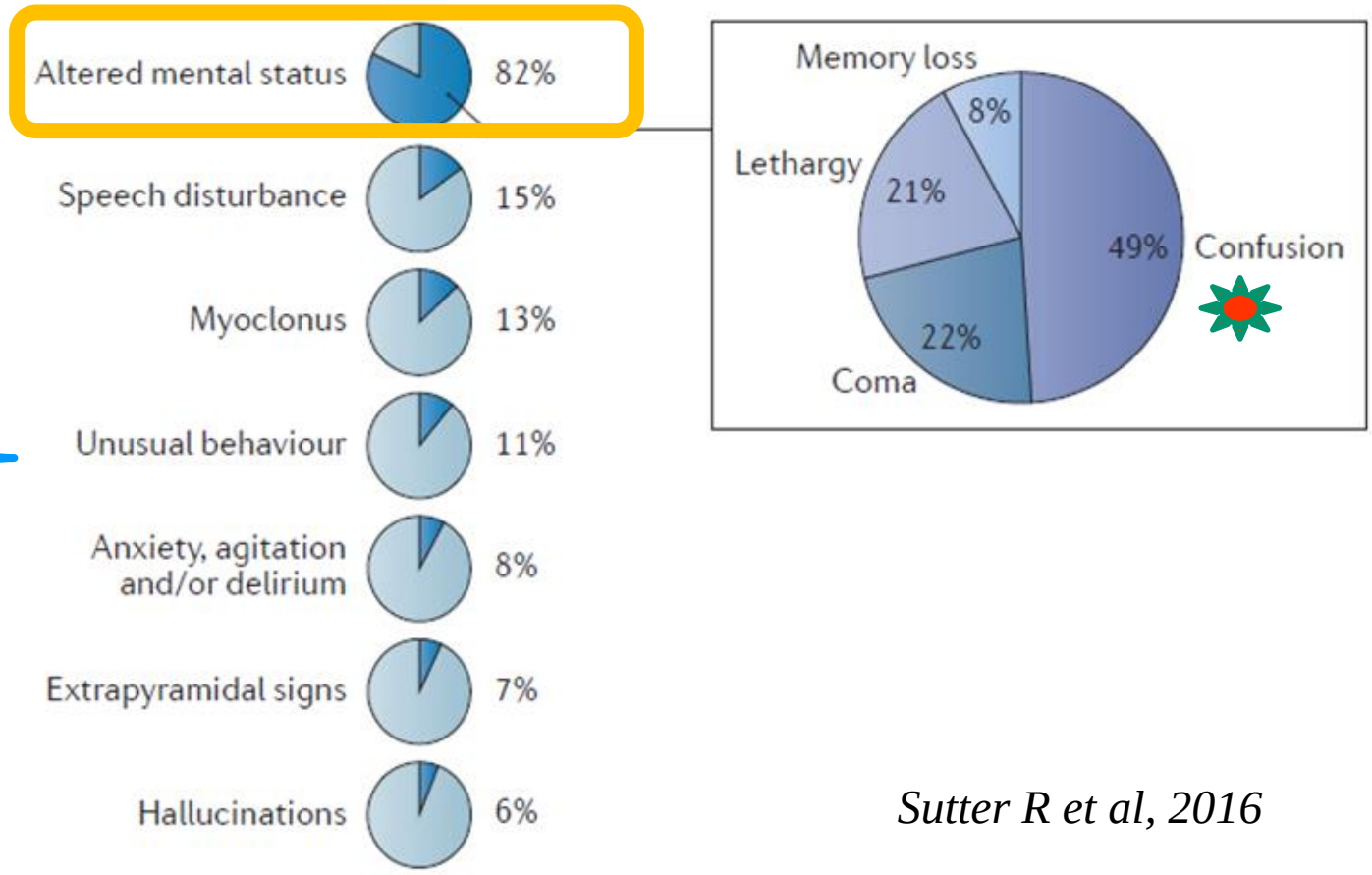
NATURE REVIEWS | NEUROLOGY
VOLUME 12 | MAY 2016 | 281

Nonconvulsive status epilepticus in adults — insights into the invisible

Raoul Sutter[1,2], Saskia Semmlack[1] and Peter W. Kaplan[3]

Major symptoms of NCSE





Sutter R et al, 2016

Etiology of RPDs

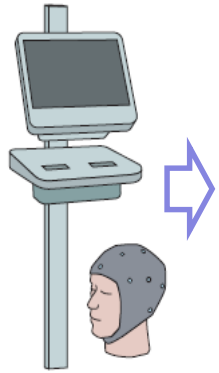
Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment
Seizures/NCSE	A	Older adults	Cognitive dysfunction, fluctuations in alertness	DWI hyper in cortical or	Might have mild pleocytosis	EEG	AEDs

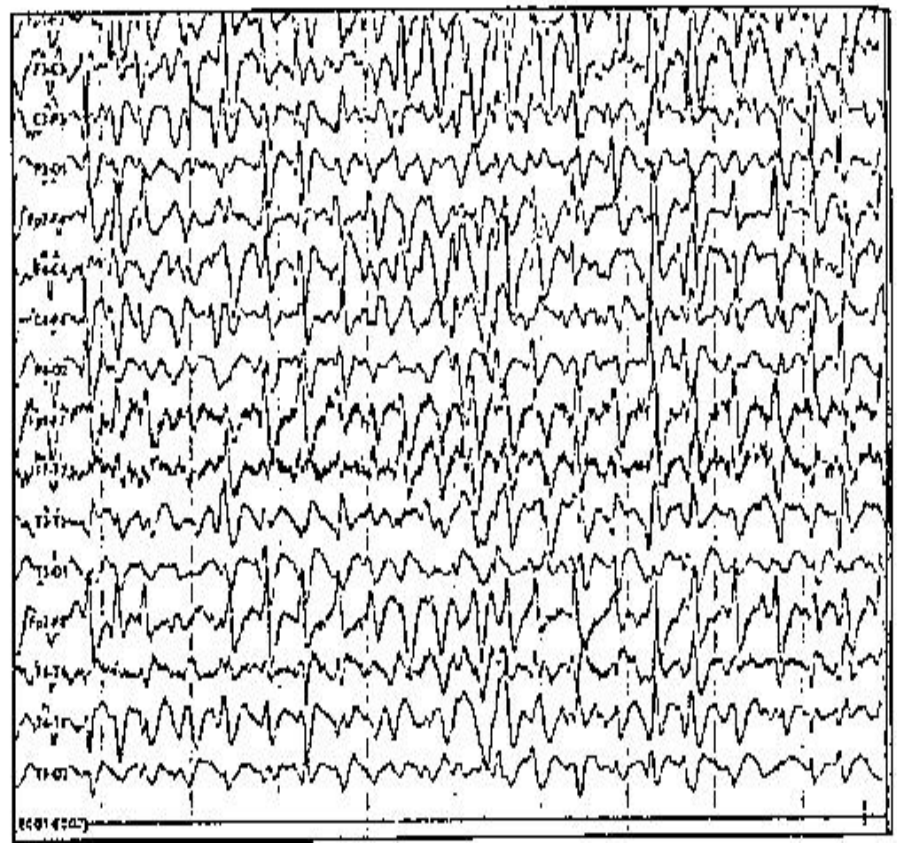
Neurology: Clinical Practice ▮ September 2012

Ross W. Paterson et al, 2012

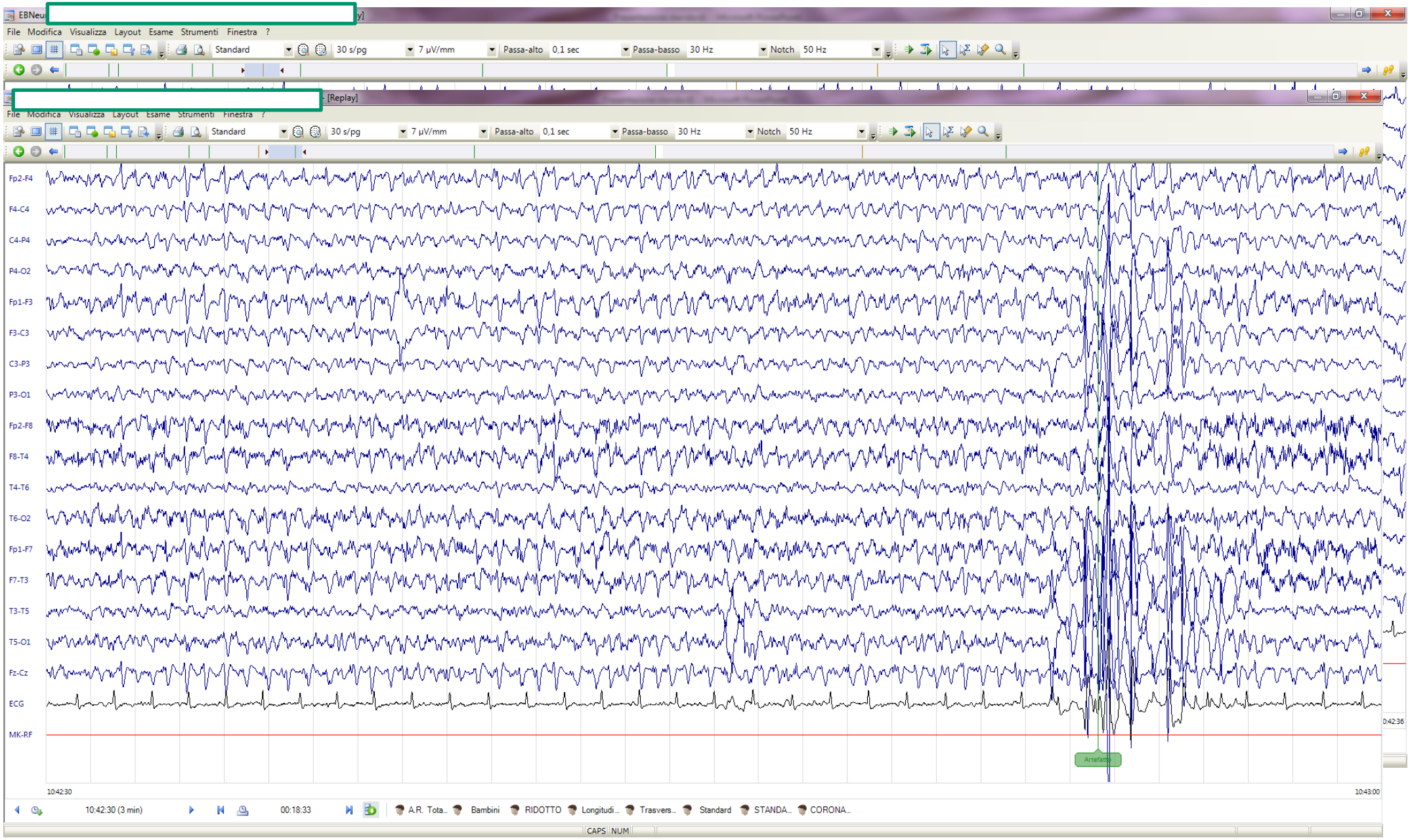
Diagnosis of Status Epilepticus

- Convulsive SE
  - Easy to diagnose clinically
  - Obvious motor signs
- NCSE
  - Difficult to diagnose clinically
  - No obvious motor manifestations
  - Altered consciousness
  - EEG-confirmed (electroclinical) diagnosis

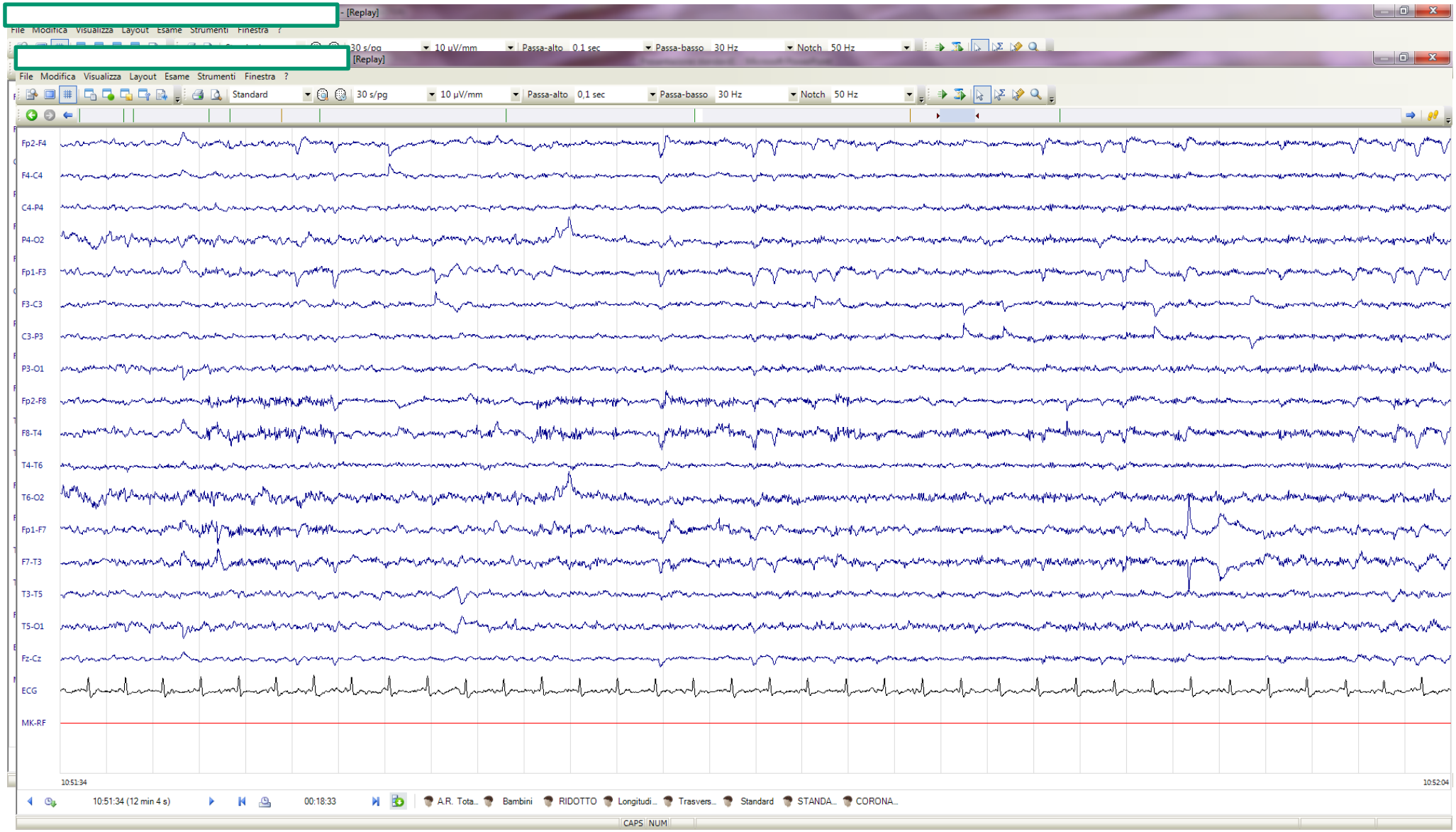




EEG critico: etiologia iponatriemia



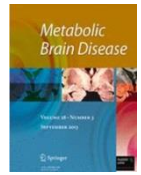
EEG critico: risposta alla BdZ (lorazepam)



De Novo Epileptic Confusional Status in a Patient with Cobalamin D

Metabolic Brain Disease, Vol. 10, No. 3, 1995

Umberto Aguglia
Giovambattista D



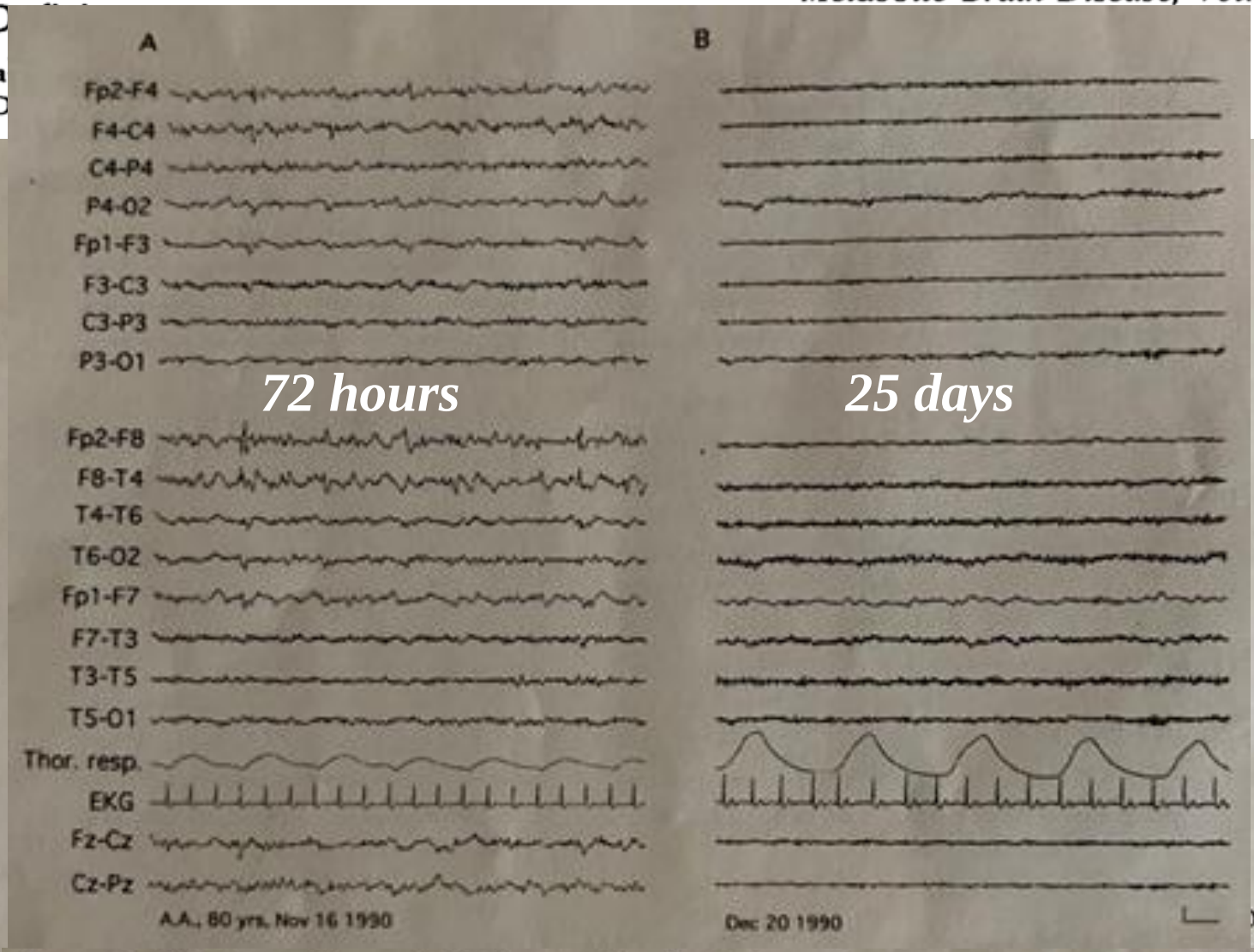


Figure 2: EEG recording. A) 72 hours after beginning of replacement therapy. Normal background activity and sporadic interictal spike-waves over the right frontal region at F8 and F4 phase reversal. B) 25 days after beginning replacement therapy. Normal posterior background and lack of any abnormal activity over the right frontal region. Calibration: 1 sec, 50 μV.

malattie responsabili di decadimento cognitivo

ascular (strokes, clotting of brain veins);

nfectious (HIV, encephalitis, fungal, parasites);

oxic-Metabolic (medicines, vitamin excess/deficiency, toxins);

utoimmune (Antibody-mediated, rheumatological, cancer-related);

etastases/Neoplasm (cancer);

atrogenic (brought forth by your doctor);

eurodegenerative (Alzheimer's, Parkinson's, Primary Progressive Aphasia, Lewy Bo

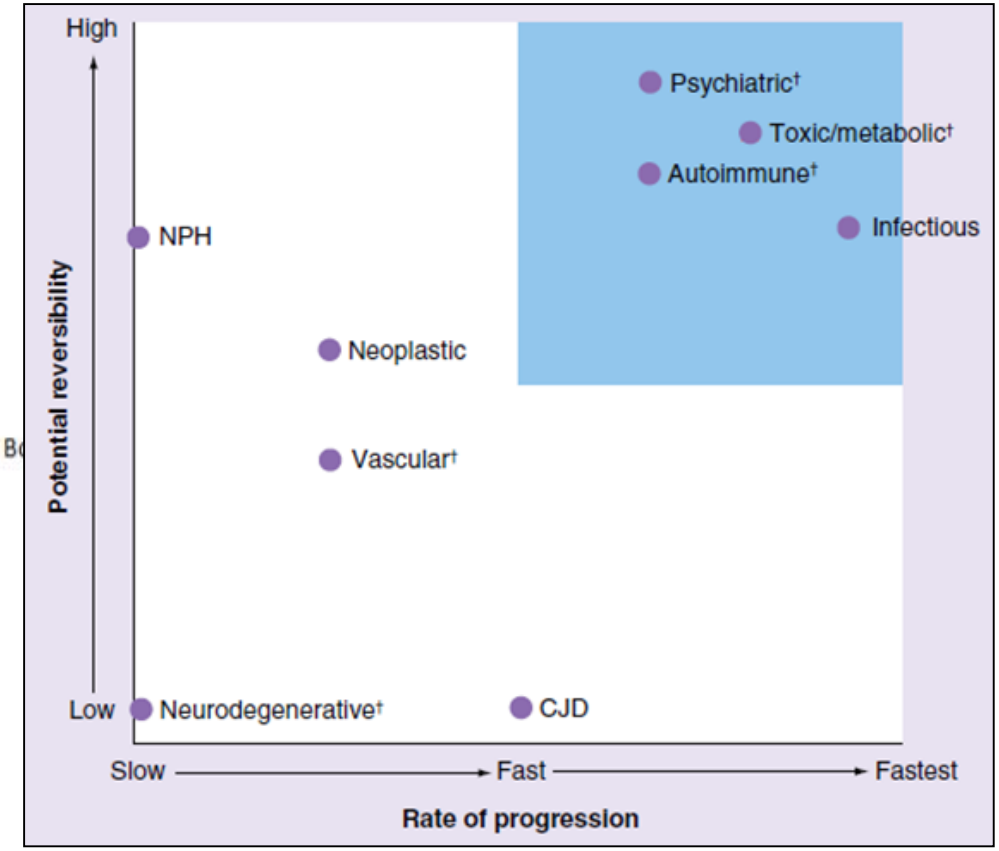
ystemic/Seizures/Structural.

Ross W. Paterson et al, 2012

Neurology® Clinical Practice

Diagnosis and treatment of rapidly progressive dementias

Ross W. Paterson, MRCP*
Leonel T. Takada, MD*
Michael D. Geschwind, MD, PhD



Maria C. 51 anni

Anamnesi familiare:

padre di 93 affetto da FA; madre di 84 anni, 2 sorelle ed 1 fratello in a.b.s.; 1 fratello è stato sottoposto all'età di 42 anni a trapianto di midollo osseo per *«mielodisplasia»;* 1 fratello di 42 anni è affetto da *«valvulopatia cardiogena»* in attesa di intervento cardiochirurgico.

Anamnesi fisiologica:

nulla da segnalate; impiegata comunale

Anamnesi patologica remota:

vitiligine; fibromioma uterino; allergia al nichel ed ad alcuni alimenti (pomodori, fragole, etc).

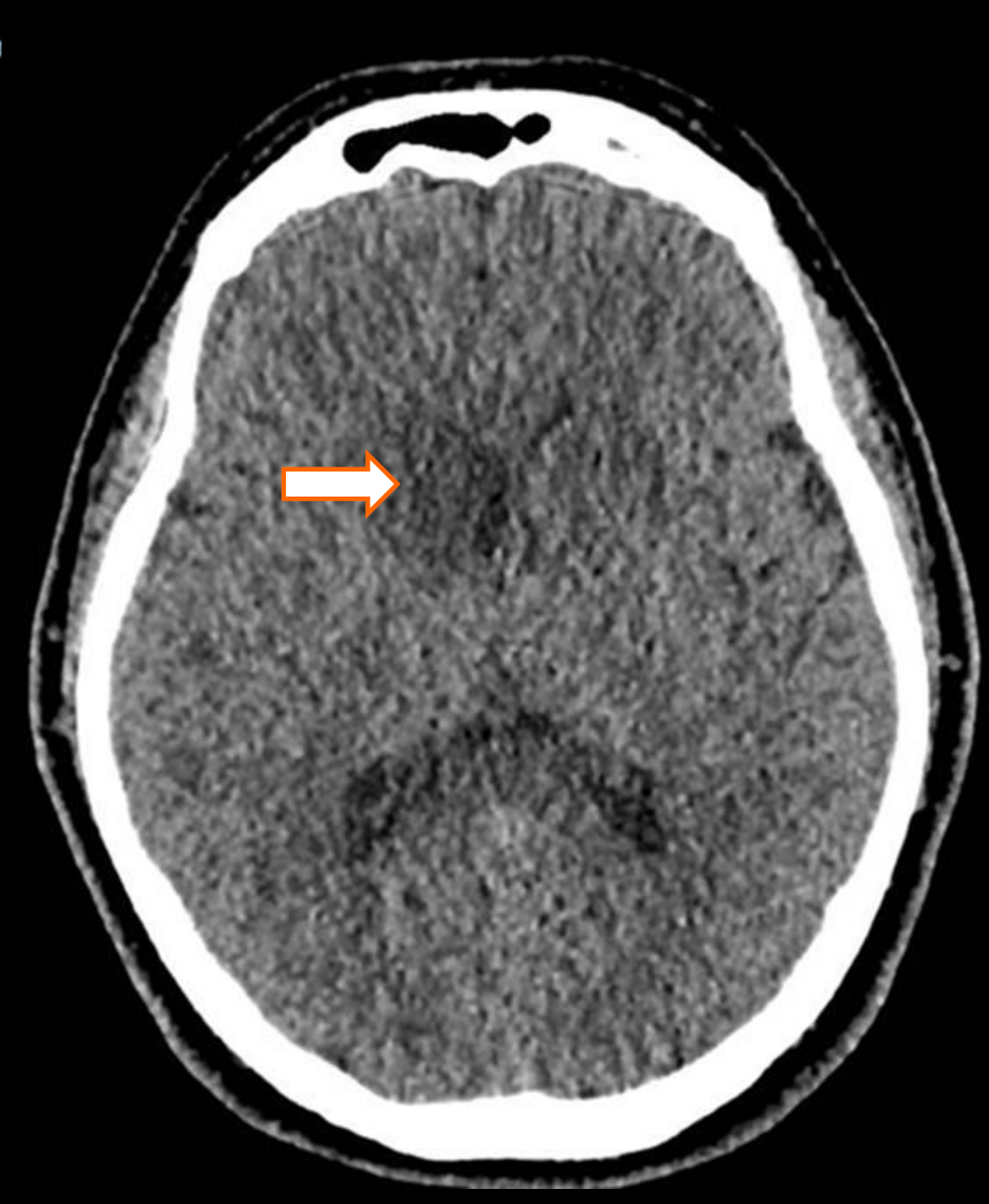
Anamnesi patologica prossima:

da giugno 2019 disturbi mnesici e comportamentali, insonnia, depressione. 24.10.19: TC encefalo (P.S.):
lesioni multiple su base «vascolare»;
dimissione: riferiti disturbi della memoria;
test neuropsicologici ambulatoriali.

28 ottobre 2019 P.S. AO Pugliese

valutazione neurologica:
disorientata/confusa

TC encefalo



TC encefalo urgente (28 ottobre)

Ricovero per 2 giorni in OBI

(consulenza psichiatrica/talofen)

31.10 ricovero: laboratorio

Routine ematochimica: nella norma

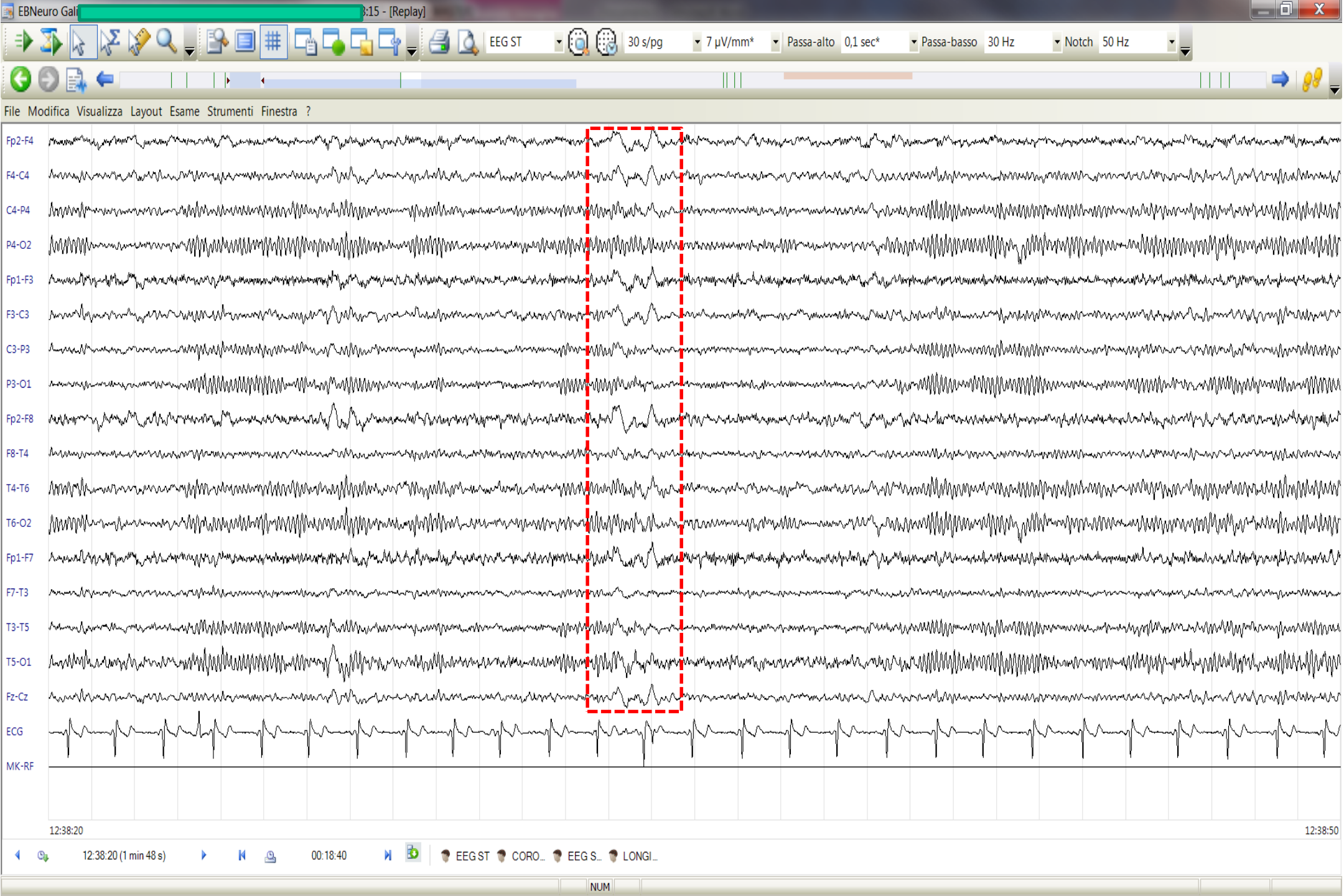
Studio trombofilico: nella norma. Assenza di mutazioni Leiden e protrombina

Screening tiroideo: nella norma

Screening autoimmune: nella norma

Dosaggio vitamina B12, folati: nella norma

Sierologia Lue, HIV: negativi


EBNeuro Gali
:15 - [Replay]
EEG ST
30 s/pg
7 µV/mm*
Passa-alto
0,1 sec*
Passa-basso
30 Hz
Notch
50 Hz
File Modifica Visualizza Layout Esame Strumenti Finestra ?
Fp2-F4
F4-C4
C4-P4
P4-O2
Fp1-F3
F3-C3
C3-P3
P3-O1
Fp2-F8
F8-T4
T4-T6
T6-O2
Fp1-F7
F7-T3
T3-T5
T5-O1
Fz-Cz
ECG
MK-RF
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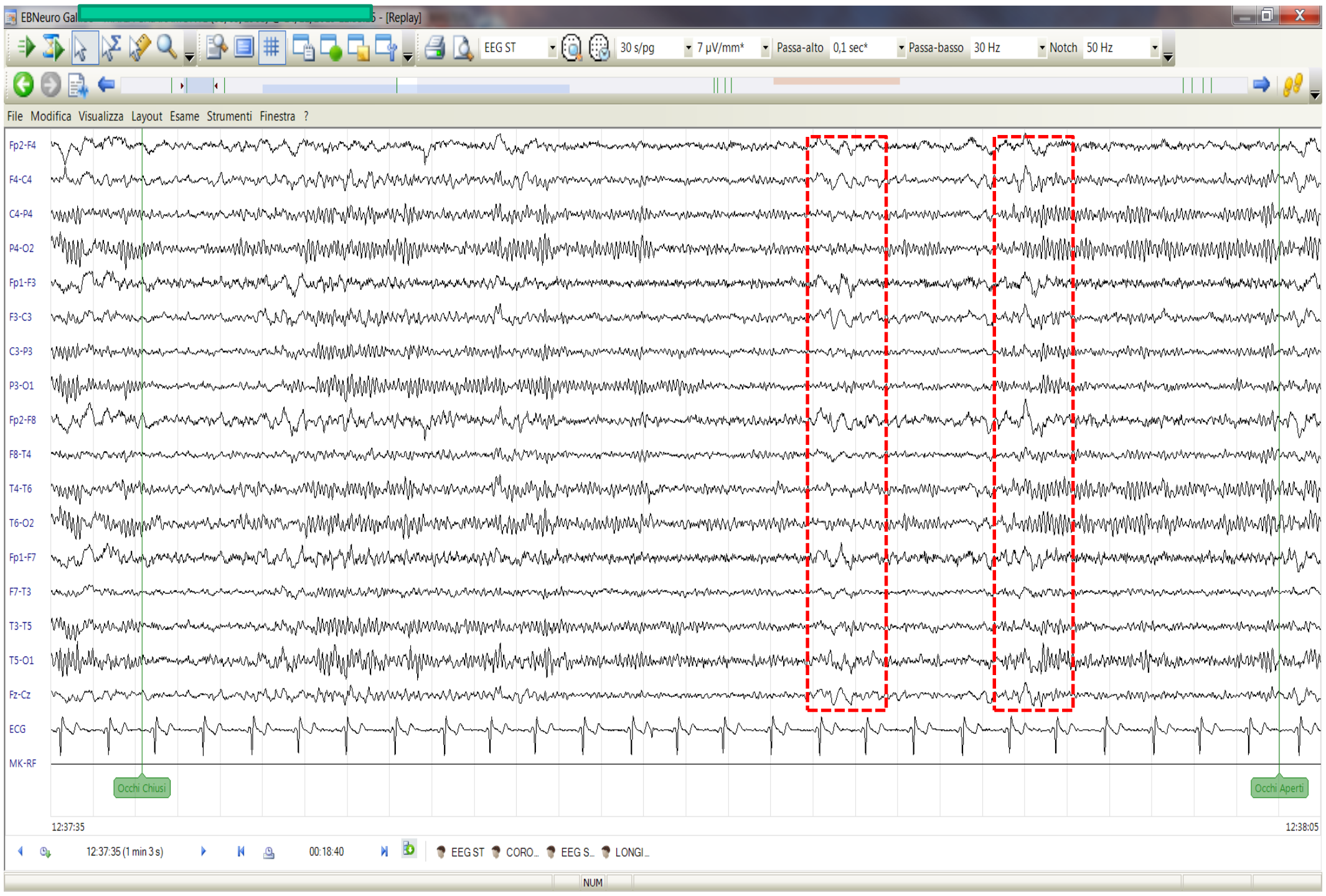
12:38:20 (1 min 48 s)
00:18:40
EEG ST
CORO...
EEG S...
LONGI...
NUM


EBNeuro Gal
- [Replay]
EEG ST
30 s/pg
7 µV/mm*
Passa-alto
0,1 sec*
Passa-basso
30 Hz
Notch
50 Hz
File Modifica Visualizza Layout Esame Strumenti Finestra ?
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F4-C4
C4-P4
P4-O2
Fp1-F3
F3-C3
C3-P3
P3-O1
Fp2-F8
F8-T4
T4-T6
T6-O2
Fp1-F7
F7-T3
T3-T5
T5-O1
Fz-Cz
ECG
MK-RF
Occhi Chiusi
Occhi Aperti
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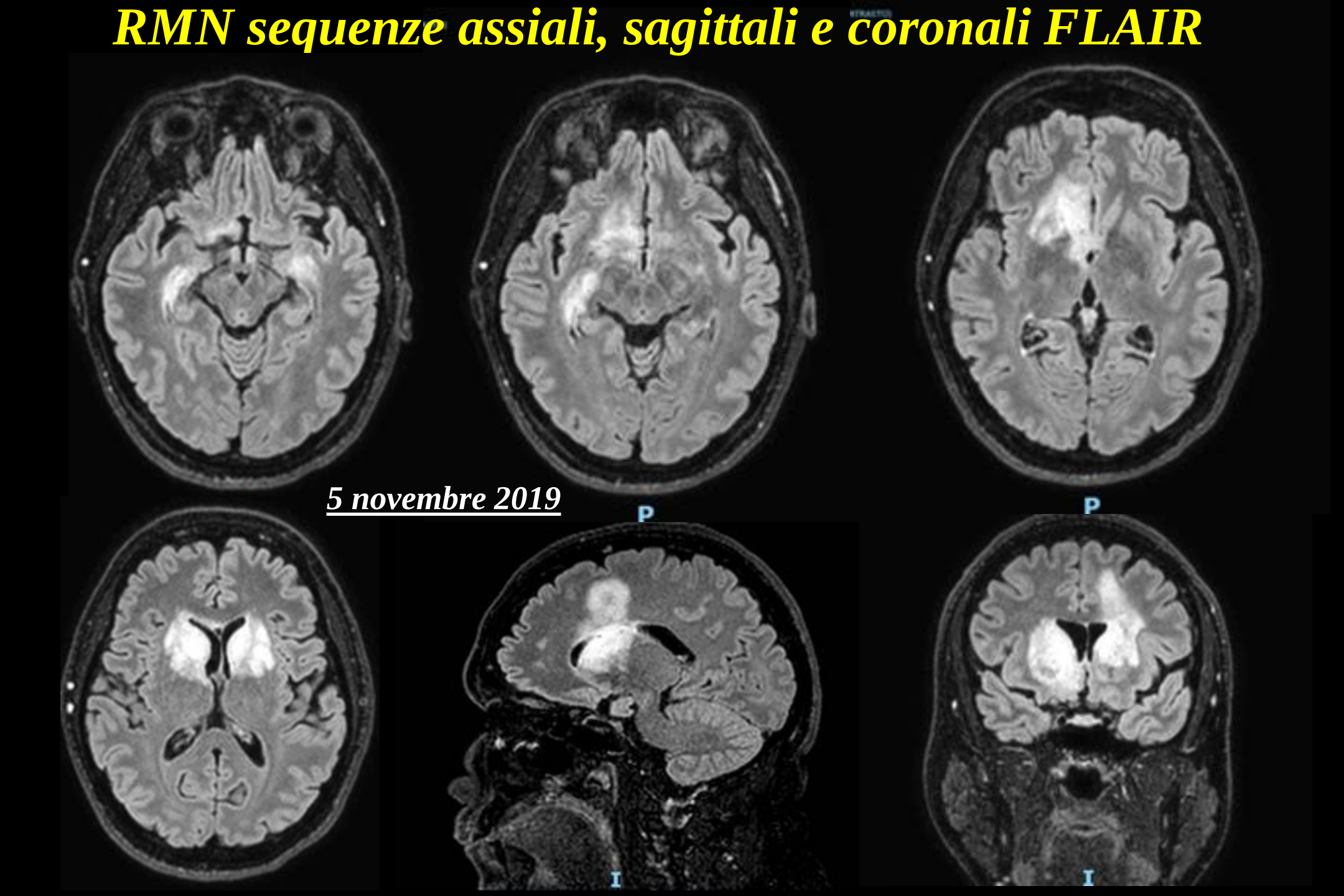
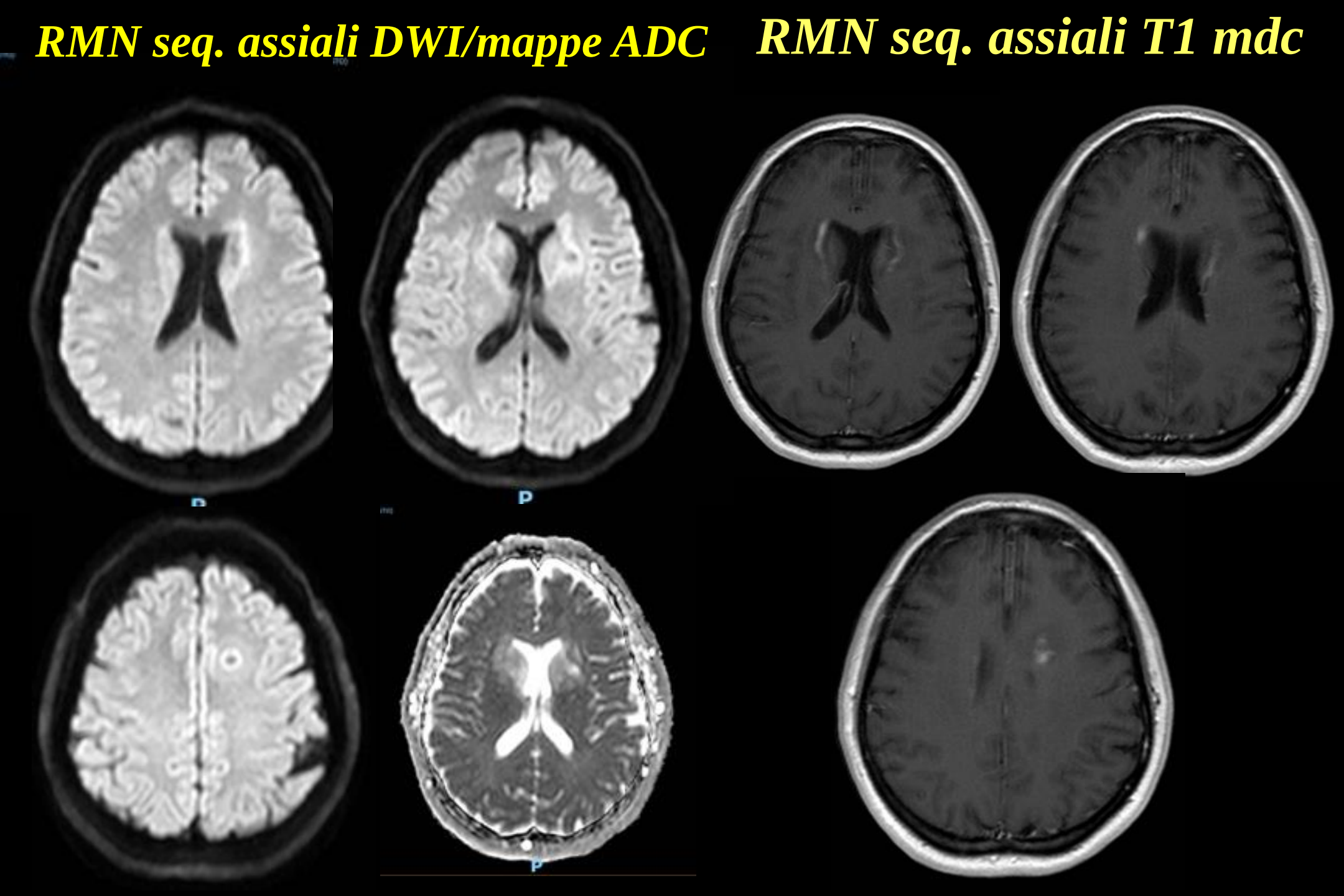
12:38:05
12:37:35 (1 min 3 s)
00:18:40
EEG ST
CORO...
EEG S...
LONGI...
NUM

RMN sequenze assiali, sagittali e coronali FLAIR

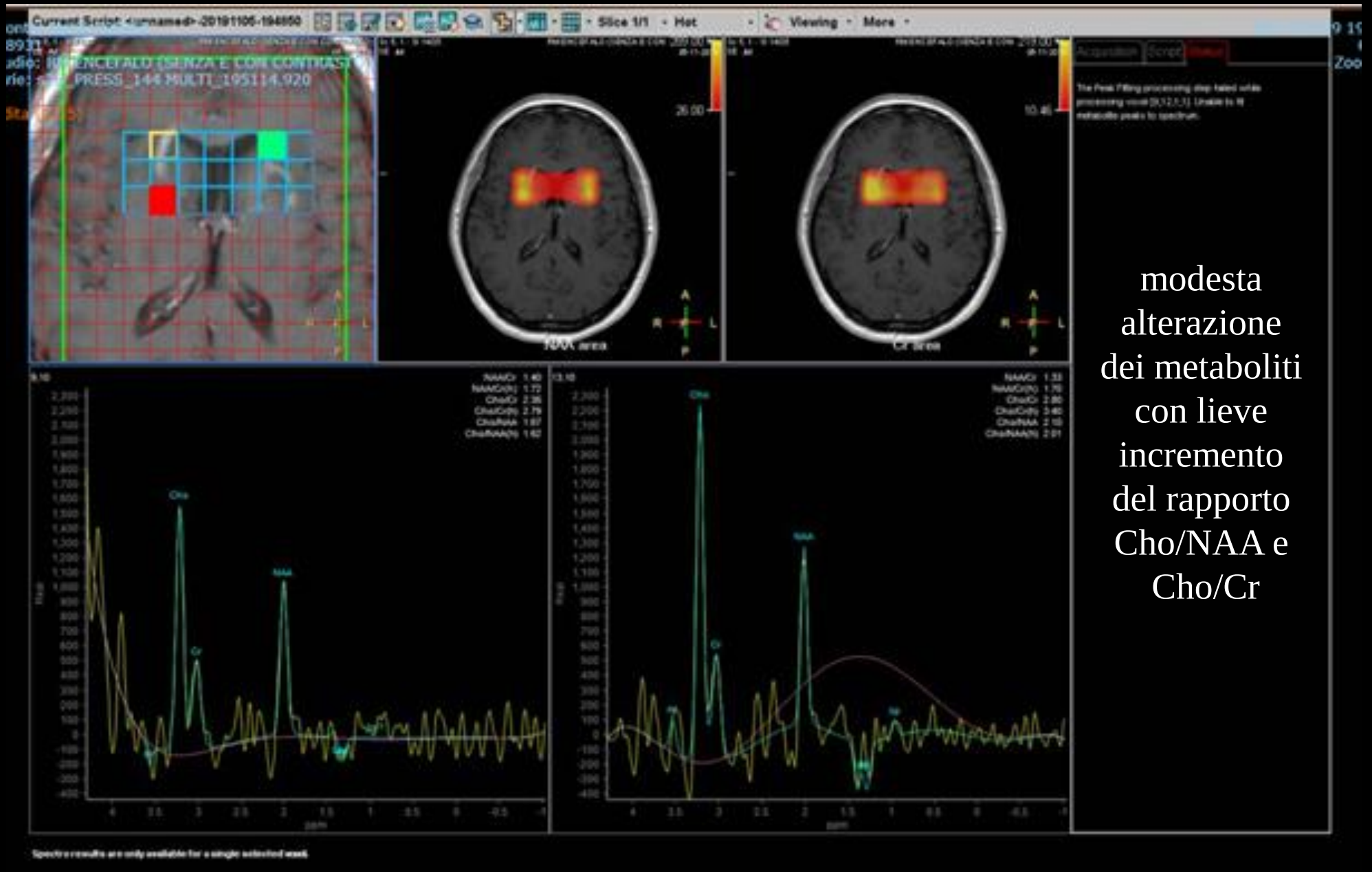


RMN seq. assiali DWI/mappe ADC

RMN seq. assiali T1 mdc



RMN spettroscopica



modesta alterazione dei metaboliti con lieve incremento del rapporto Cho/NAA e Cho/Cr

BIOLOGIA MOLECOLA

Identificazione genomica su
(Met. Nested multiplex PCR Sy

- E. coli K1:	Sensibilità: 1000 CFU/mL
- Haemophilus influenzae:	Sensibilità: 1000 CFU/mL
- Listeria monocytogenes:	Sensibilità: 1000 CFU/ml
- N. meningitidis (A, B, C, D	Sensibilità: 100 CFU/ml
- Streptococcus agalactiae	Sensibilità: 1000 CFU/ml
- Streptococcus pneumonia	Sensibilità: 100 cellule/ml
- Cytomegalovirus (CMV):	Sensibilità: 100 TCID50/ml
- Enterovirus (specie A, B,	Sensibilità: 50 TCID50/ml
- Herpes simplex virus 1 (H	Sensibilità: 1500 copie/ml
- Herpes simplex virus 2 (H	Sensibilità: 1300 copie/ml
- Human herpesvirus 6 (HH	Sensibilità: 10000 copie/ml
- Human parechovirus:	Sensibilità: 500 TCID50/ml
- Varicella zoster virus (VZ	Sensibilità: 1600 copie/ml
- Cryptococcus neoforman	Sensibilità: 100 CFU/ml

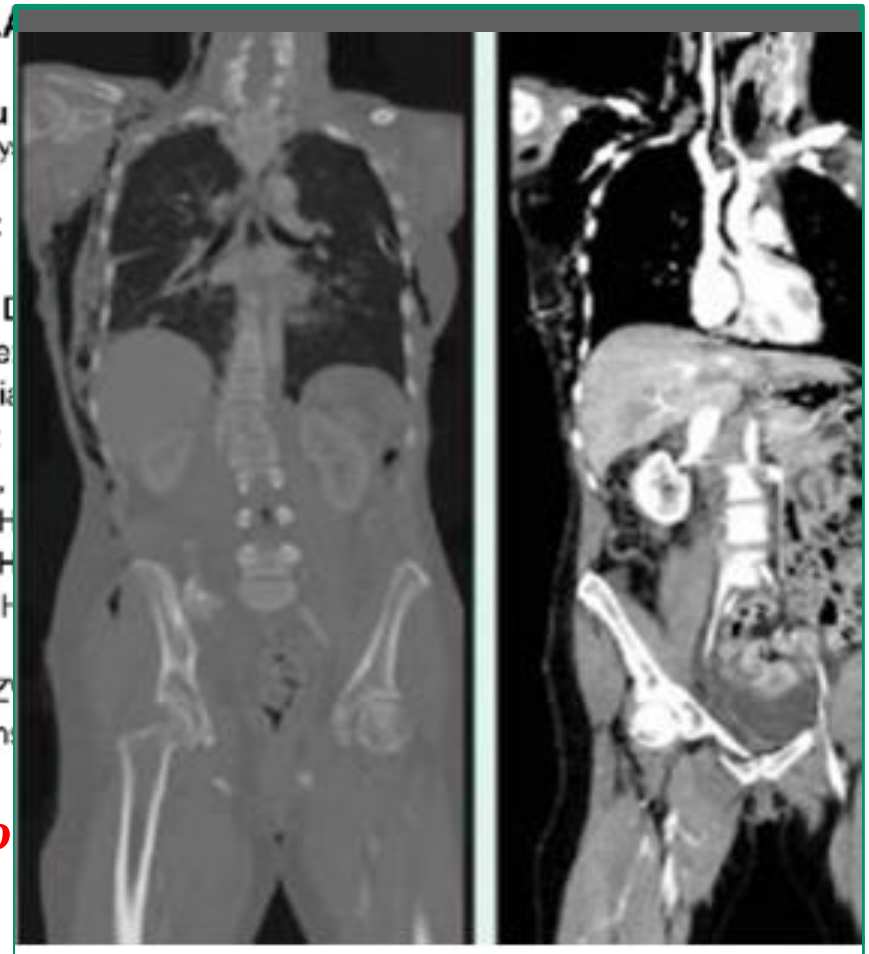
citologico: negativo

MICROBIOLOGIA

ligoclonali: presenti

INQUADRAMENTO DIAGNOS

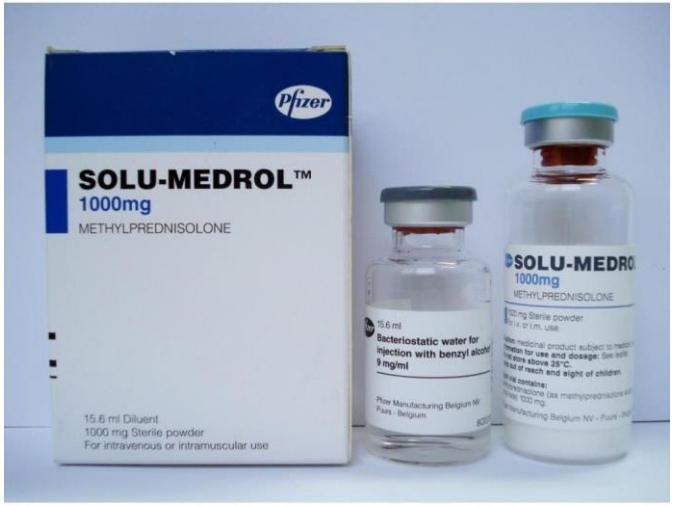
- ES. FISICO: (Met. ottico)	
- GLUCOSIO su LIQUOR: (Met. enzimatico)	(40 - 70)
- ALBUMINA su LIQUOR: (Met. nefelometrico)	(0 - 350)
- IgG su LIQUOR: (Met. nefelometrico)	(0 - 34)
- CONTA CELLULE AL CC (Contaglobuli: citofluorimetria in	POSITIVO > 25 cellule/ul

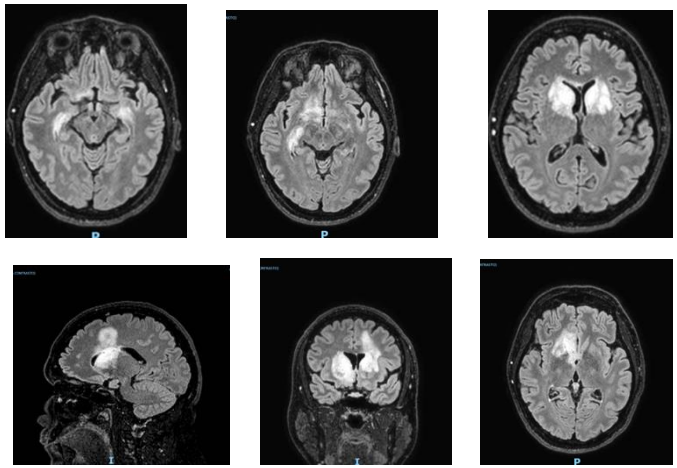


TC total body negativa

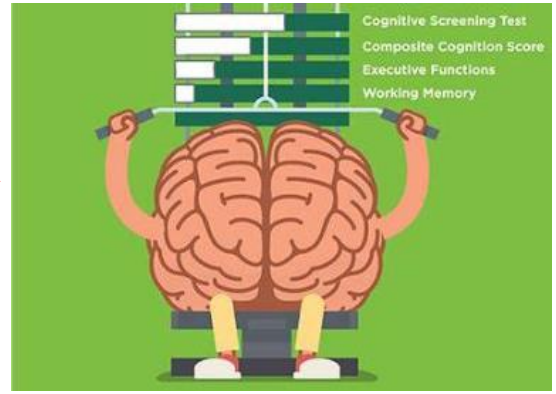
Gestione terapeutica

1 gr/die per 7 giorni

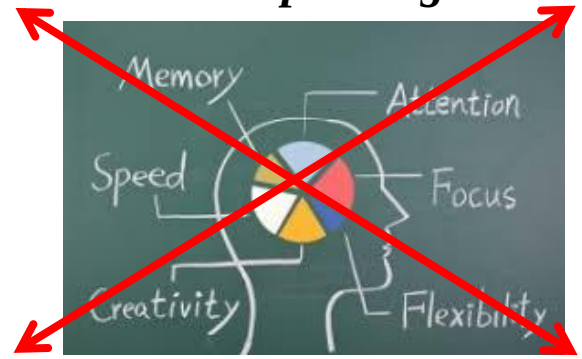




migliora cognitivamente

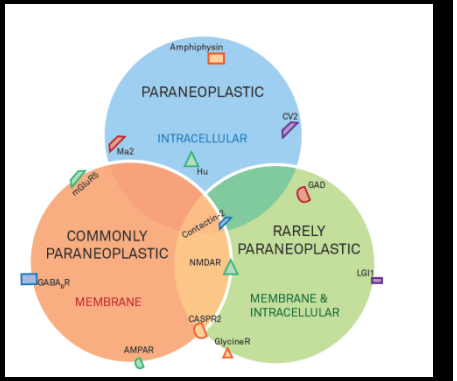


test neuropsicologici



in corso dosaggio……

anticorpi onconeurali/ anti-LGI1, NMDAr, etc) (Reggio C/Milano)



bande oligoclonali: presenti

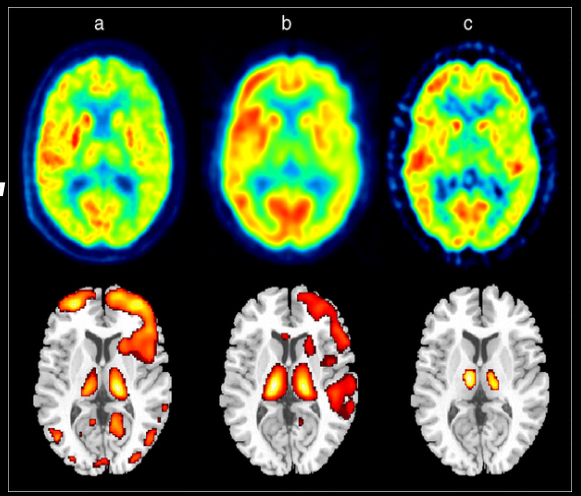
Table 2. CSF abnormalities in different causes of RPD.

Etiology	Cell count	Oligoclonal bands	CSF markers of neuronal damage or other surrogates
Prion diseases	↔	Not present	14-3-3: ↑ – ↑↑↑ Tau: ↑ – ↑↑↑
(rp)AD	↔	Not present	14-3-3: ↔ – ↑ Tau: ↔ – ↑↑
Other neurodegenerative diseases	↔	Not present	14-3-3: ↔ – (↑) Tau: ↔ – (↑)
Cerebrovascular disease	↔ (except for vasculitis)	Not present (except for vasculitis)	14-3-3: ↔ – (↑↑)* Tau: ↔ – (↑)*
Immune-mediated encephalitis	↔ – ↑↑	Present in most cases	14-3-3: ↔ – ↑↑↑ Tau: ↔ – ↑↑
Infectious encephalitis	↑ – ↑↑↑	Present	14-3-3: ↔ – ↑↑↑ Tau: ↔ – ↑↑

Ingar Zer & Peter Herman, 2018

da eseguire al Policlinico……

FDG-PET



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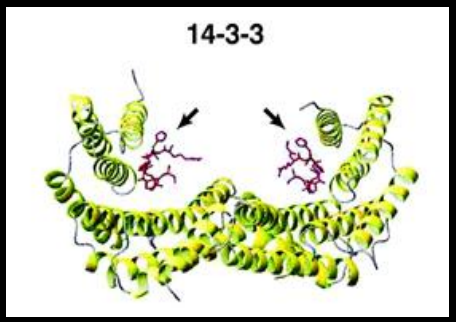


altri suggerimenti diagnostici??

FINE

Diagnostic challenges in rapidly progressive dementia

Table 5. Summary of various other diseases and conditions that may lead to or mimic RPD.

Other diseases and conditions that may lead to or mimic RPD		
CNS neoplasia 1. CNS lymphoma 2. Solid neoplasia (primary in the CNS or metastatic) 3. Diffuse gliomatosis 4. Leptomeningeal spread of other tumors **Metabolic** 1. Hyponatremia and other electrolyte disturbances 2. Wernicke's encephalopathy (thiamine) and other vitamin deficiency syndromes (e.g. B12, folate, niacin, biotin) 3. Hepatic encephalopathy 4. Uremic encephalopathy 5. Hypothyroidism 6. Hypoparathyroidism 7. Porphyria 8. Adrenal insufficiency 9. Extrapontine myelinolysis	**Storage diseases and other genetic causes for RPD** 1. Wilson's disease 2. NBIA 3. Lysosomal storage diseases (selected): mucopolysaccharidoses, sphingolipidoses, and neuronal ceroid lipofuscinoses 4. Huntington's disease 5. CADASIL resp. CARASIL 6. Mitochondrial diseases (selected): MELAS, Leigh disease 7. PME: Lafora disease, ULD 8. Porphyria 9. Methylmalonic academia 10. DRPLA	**Toxic** 1. Alcohol-related dementia 2. Benzodiazepine-related dementia 3. Methyl intoxication 4. Metal intoxications (e.g. lithium, mercury) 5. Side effects of chemotherapeutics or neuroleptic drugs **Psychiatric** 1. Severe depression 2. Schizophrenic disorders 3. Hypochondriac delusions **Others and secondary conditions** 1. Hypoxic brain damage 2. Epilepsy and secondary seizures 3. Systemic inflammation (e.g. systemic immune-mediated diseases, septic encephalopathy) 4. NPH

NBIA: Neurodegeneration with brain iron accumulation; CADASIL: cerebral autosomal dominant arteriopathy with subcortical Infarcts and leukoencephalopathy; MELAS: mitochondrial encephalopathy, lactic acidosis and stroke-like episodes; PME: progressive myoclonic epilepsies; ULD: Unverricht–Lundborg disease; DRPLA: dentatorubral-pallidolysial atrophy; NPH: normal pressure hydrocephalus.

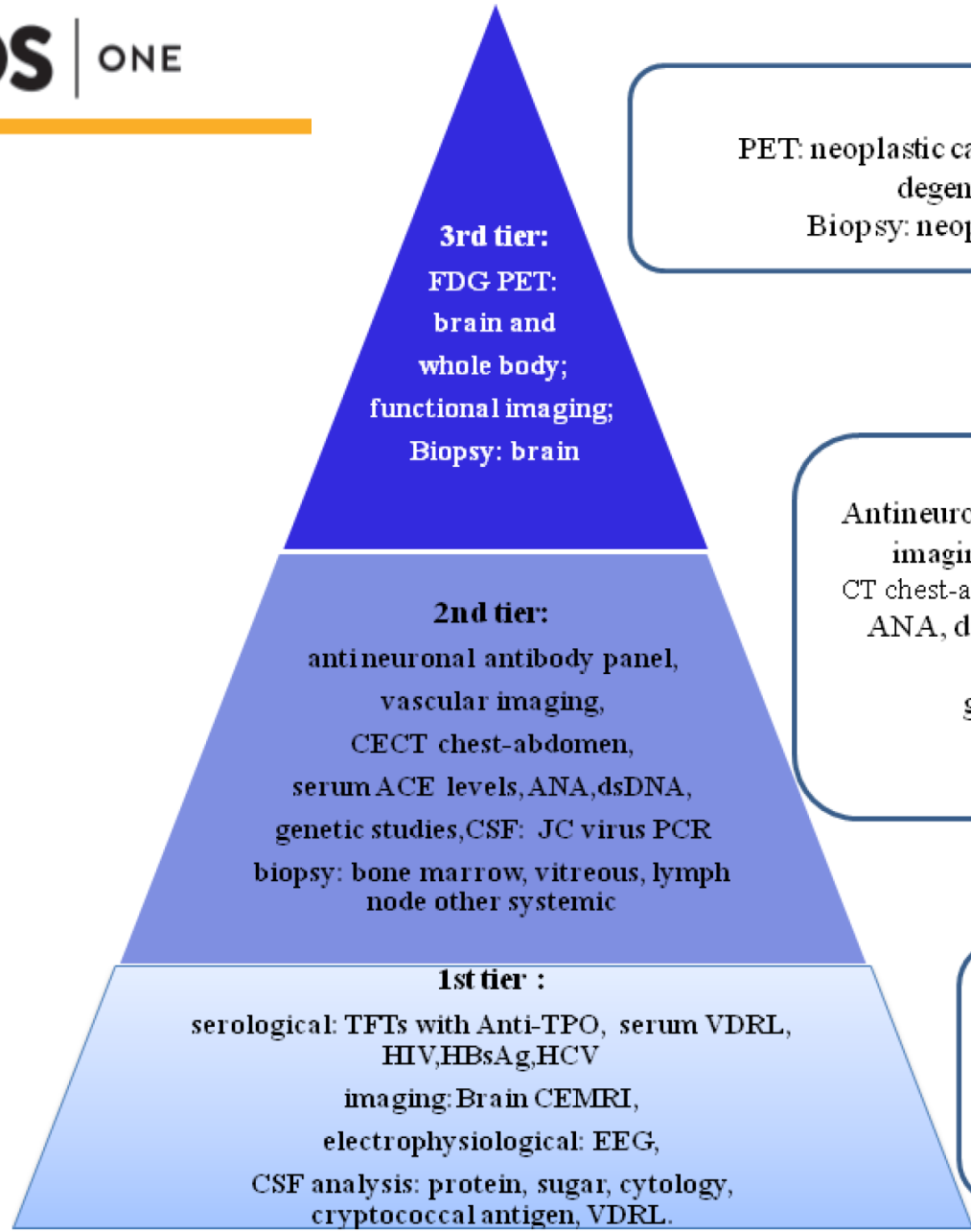
Inga Zerr & Peter Hermann, 2018

PLOS ONE

RESEARCH ARTICLE

Rapidly progressive dementia: An eight year (2008–2016) retrospective study

Patil Anuja[1☯], Vishnu Venugopalan[2‡*], Naheed Darakhshan[1☯], Pandit Awadh[2‡], Vinny Wilson[1‡], Goyal Manoj[1‡], Modi Manish[1‡], Lal Vivek[1‡]



Storage diseases and other genetic causes for RPD

1. Wilson's disease
2. NBIA
3. Lysosomal storage diseases (selected): mucopolysaccharidoses, sphingolipidoses, and neuronal ceroid lipofuscinoses
4. Huntington's disease
5. CADASIL resp. CARASIL
6. Mitochondrial diseases (selected): MELAS, Leigh disease
7. PME: Lafora disease, ULD
8. Porphyria
9. Methylmalonic academia
10. DRPLA

RAPPORTO TRA DIAMETRO DEI CORNI FRONTALI E BIPARIETALE (d1/d2)
Se >0.3 = idrocefalo

Toxic

1. Alcohol-related dementia
2. Benzodiazepine-related dementia
3. Methyl intoxication
4. Metal intoxications (e.g. lithium, mercury)
5. Side effects of chemotherapeutics or neuroleptic drugs

Etiology of RPDs: use of the mnemonic VITAMINS

ascular (strokes, clotting of brain veins);

nfectious (HIV, encephalitis, fungal, parasites);

oxic-Metabolic (medicines, vitamin excess/deficiency, toxins);

utoimmune (Antibody-mediated, rheumatological, cancer-related);

etastases/Neoplasm (cancer);

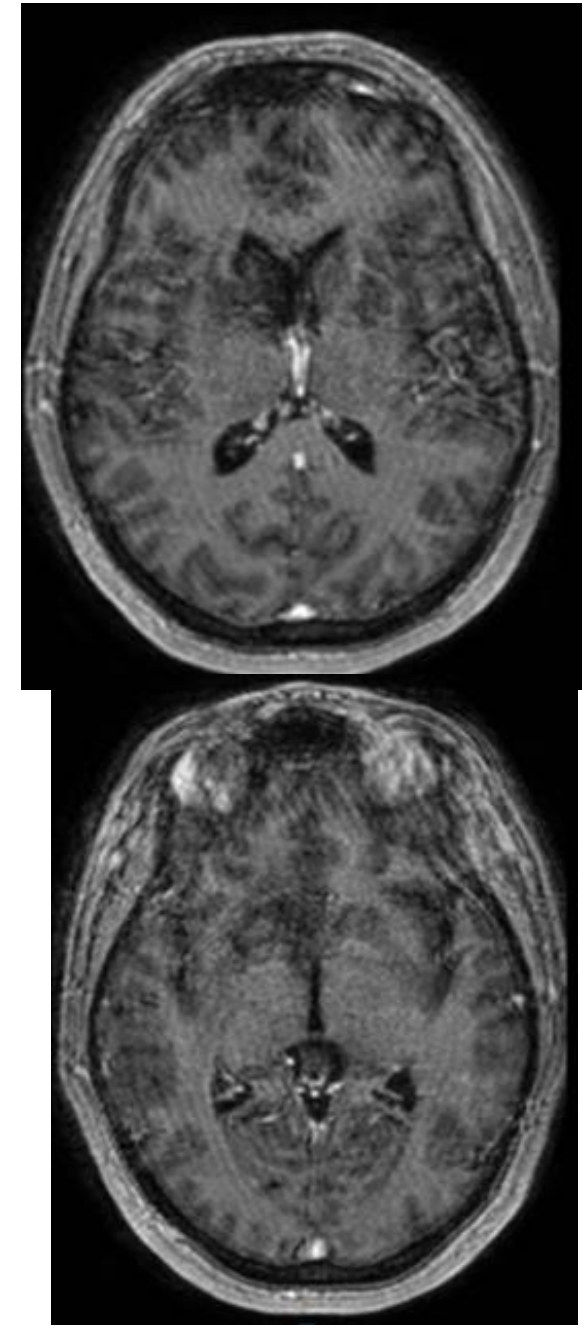
atrogenic (brought forth by your doctor);

eurodegenerative (Alzheimer's, Parkinson's, Primary Progressive Aphasia, Lewy Bodies);

ystemic/Seizures/Structural.

Ross W. Paterson et al, 2012

Neurologia
Emergenza
Urgenza



Anteprima PDF — Pag. 2 di 2 — Qualità anteprima: Alta (lenta) — Adatta immagine — Zoom

Esame	Risultato	U.M.

BIOLOGIA MOLECOLARE

Identificazione genomica su liquor di patogeni (pannello 14 target)
(Met. Nested multiplex PCR System FilmArray)

- E. coli K1:	NON RILEVATO
- Haemophilus influenzae:	NON RILEVATO
- Listeria monocytogenes:	NON RILEVATO
- N. meningitidis (A, B, C, D, Y, W-135)	NON RILEVATO
- Streptococcus agalactiae:	NON RILEVATO
- Streptococcus pneumoniae:	NON RILEVATO
- Cytomegalovirus (CMV):	NON RILEVATO
- Enterovirus (specie A, B, C, D)	NON RILEVATO
- Herpes simplex virus 1 (HSV1):	NON RILEVATO
- Herpes simplex virus 2 (HSV2):	NON RILEVATO
- Human herpesvirus 6 (HHV6):	NON RILEVATO
- Human parechovirus:	NON RILEVATO
- Varicella zoster virus (VZV):	NON RILEVATO
- Cryptococcus neoformans/gattii:	NON RILEVATO

MICROBIOLOGIA

INQUADRAMENTO DIAGNOSTICO LIQUOR

- ES. FISICO: (Met. ottico)	Limpido, incolore	
- GLUCOSIO su LIQUOR: (Met. enzimatico)	73	mg/dl
- ALBUMINA su LIQUOR: (Met. nefelometrico)	343	mg/l
- IgG su LIQUOR: (Met. nefelometrico)	47	mg/l
- CONTA CELLULE AL CONTAGLOBULI: (Contaglobuli: citofluorimetria in fluorescenza)	4	cellule/ul

Liquor: limpido, incolore, trasparente; cellularità, glucosio, albumina, indice di Link: *nella norma*. IgG liquor 7.35 (0.48-5.86). Esame colturale negativo. **Bande oligoclonali: presenti.** Identificazione genomica su liquor di patogeni (pannello 14 target): *negativo*

Anteprima PDF — Pag. 5 di 6 — Qualità anteprima: Alta (lenta) — Adatta immagine — Zoom — Chiudi

MPO (pANCA)
(Immunofluorescenza multiparametrica)

PR3 (cANCA)
(Immunofluorescenza multiparametrica)

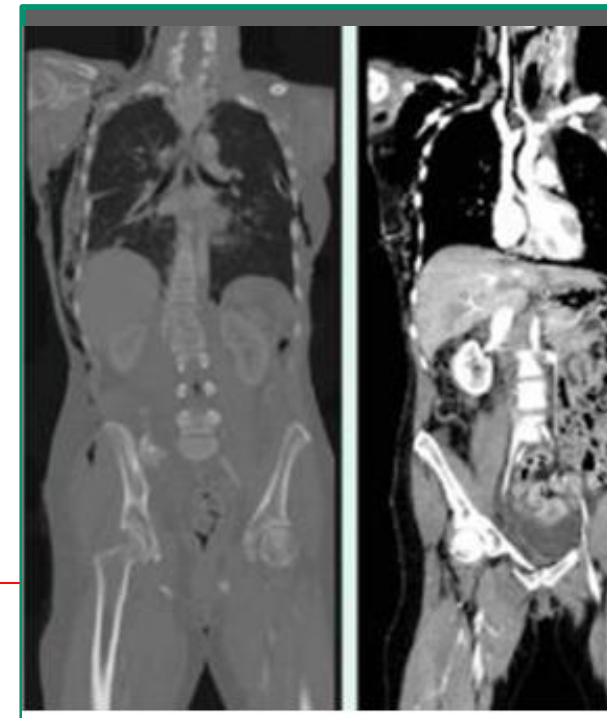
Anti-Fosfolipidi IgG
(Immunofluorescenza multiparametrica)

Anti-Fosfolipidi IgM
(Immunofluorescenza multiparametrica)

Beta2-Glicoproteina IgG
(Immunofluorescenza multiparametrica)

Beta2-Glicoproteina IgM
(Immunofluorescenza multiparametrica)

Cardiolipina IgG
(Immunofluorescenza multiparametrica)



TC total body negativa

MARKERS EPATITE / HIV

Anti HIV 1/2 - Ag p24
(Chemiluminescenza)

DIAGNOSTICA TREPONEMICA

VDRL
(Flocculazione)

TPHA QUANTITATIVO
(Emoagglutinazione)

FTA-Abs IgG
(IFI)

FTA-Abs IgM — NEGATIVO
(IFI)

Screening Sifilide TPA (Ab anti-Treponema p. IgG ed IgM)
(Chemiluminescenza)

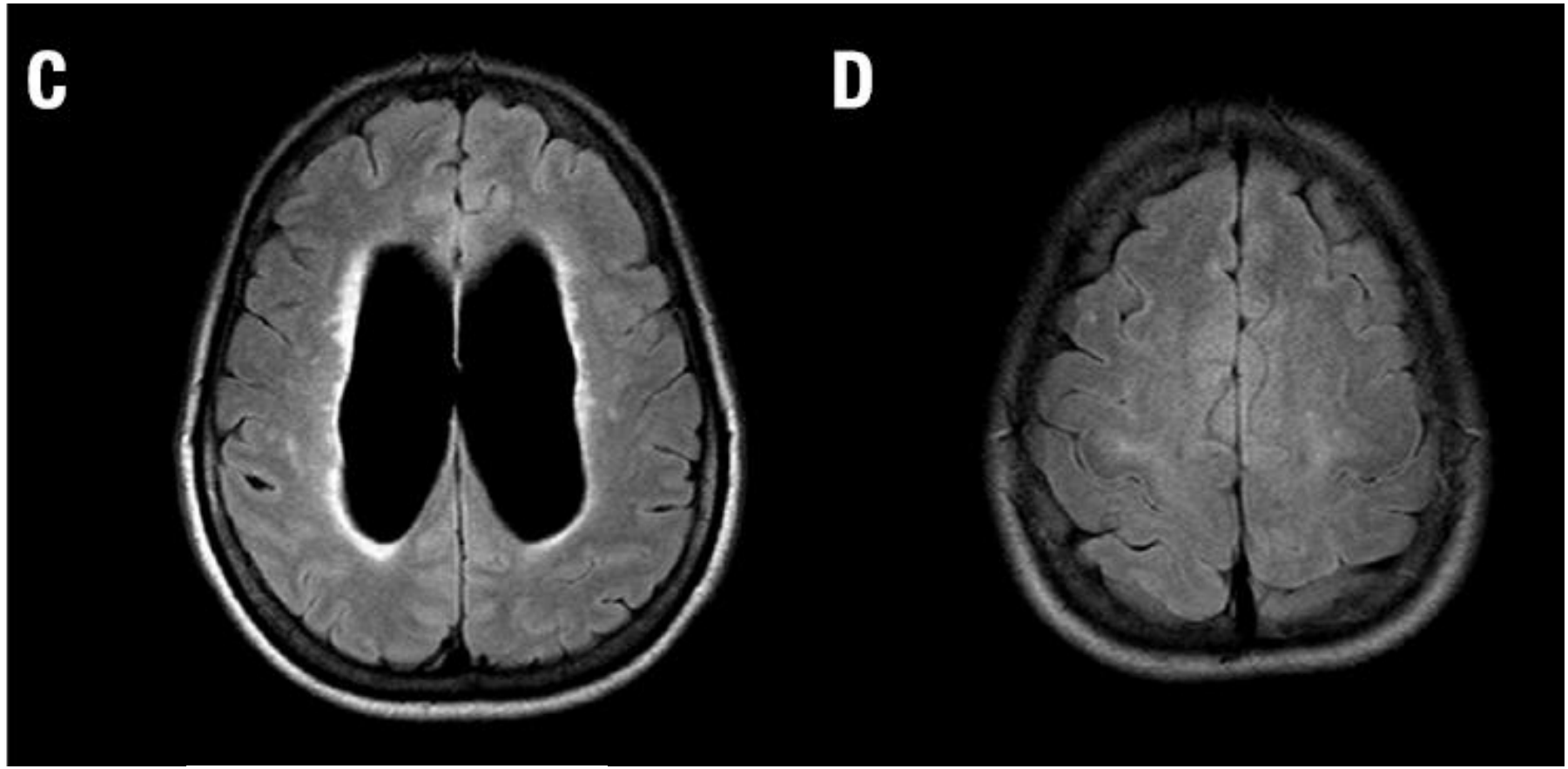
NEGATIVO
0,01 Index Negativo <0.80

Normal-pressure hydrocephalus

A critical review

Louise Makarem Oliveira[1], Ricardo Nitrini[2], Gustavo C. Román[3]

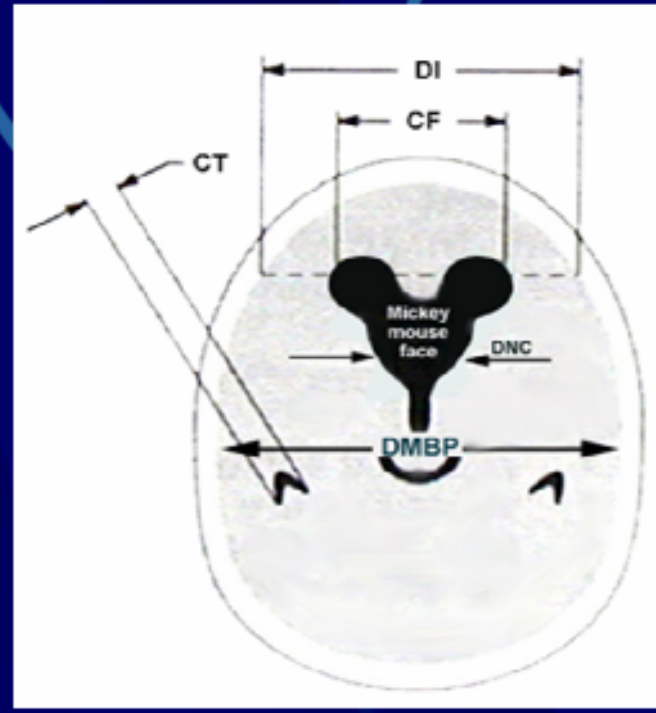
Dement Neuropsychol 2019 June;13(2):133-143



(C) Axial FLAIR MRI scan revealing enlarged lateral ventricles with bright signal in the surrounding white matter, suggestive of transependymal edema. **(D)** Axial FLAIR MRI showing narrowing of the sulci and subarachnoid spaces over the high convexity and midline surface in the frontoparietal regions.

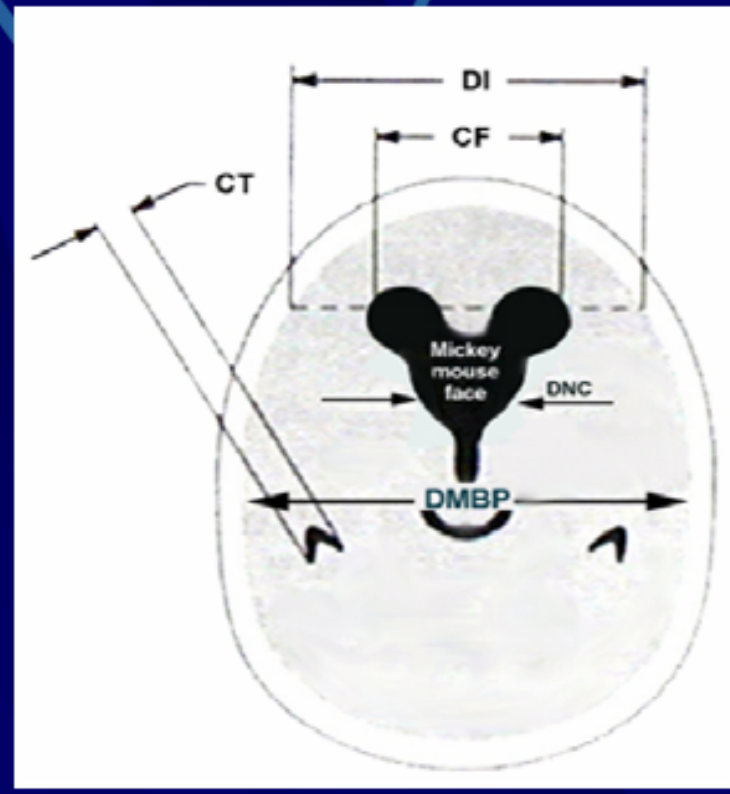
Nell'adulto normale, il rapporto della massima ampiezza dei corni frontali diviso per il diametro trasversale interno del cranio allo stesso livello (indice di Evans) è in genere il 30%.

Nell'idrocefalo i ventricoli dilatati hanno profilo arrotondato e pareti stirate e i corni frontali assumono un aspetto "globoso" fino a costituire, nei tagli assiali, quella che viene definita " Mickey Mouse Face", causata dello scomparsa della fisiologica impronta della testa del nucleo caudato sui ventricoli laterali.



DI = Diametro Interparietale CF = Corno Frontale CT = Corno Temporale
DMBP = Diametro Medio Biparietale DNC = Distanza Nuclei Caudati

Indice di Evans	CF\DMBP > 30%
Mickey mouse face	Ballooning dei corni frontali dei ventricoli laterali
Indice bicaudato	Rapporto tra la distanza tra i profili mediali dei nuclei caudati e la distanza trasversale tra due punti della teca interna misurati allo stesso livello. Patologico se > 0,15
Angolo calloso	Diminuzione dell'angolo compreso tra i due corni frontali. Valore normale 120°
Rapporto CF\DI	Patologico se > 50%
Dimensione CT	Patologica se entrambi i corni sono > 2mm

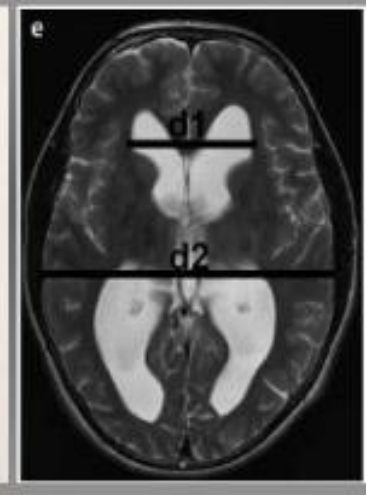


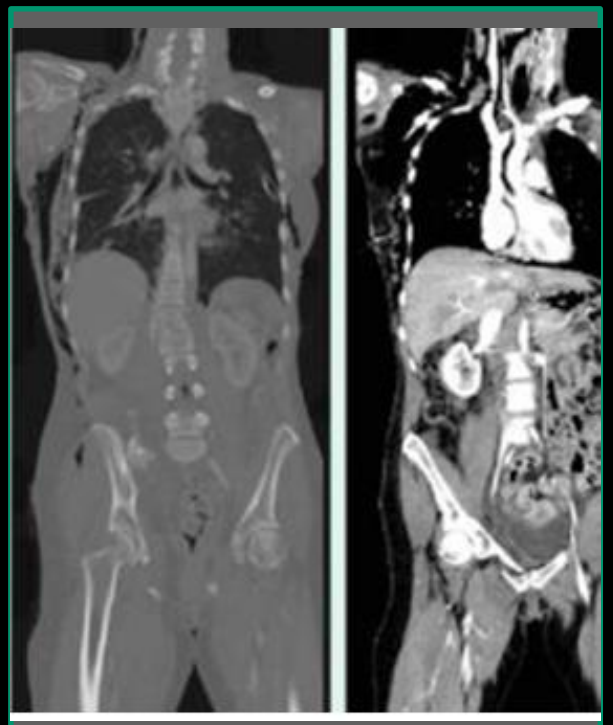
INDICE DI EVANS

RAPPORTO TRA DIAMETRO DEI CORNI FRONTALI E BIPARIETALE (d1/d2)

Se >0.3 = idrocefalo

Evans, W.A., Jr.: An encephalographic ratio for estimating ventricular enlargement and cerebral atrophy. Arch. Neurol. Psychiat. 47, 931-937 (1942)





TC total body negativa

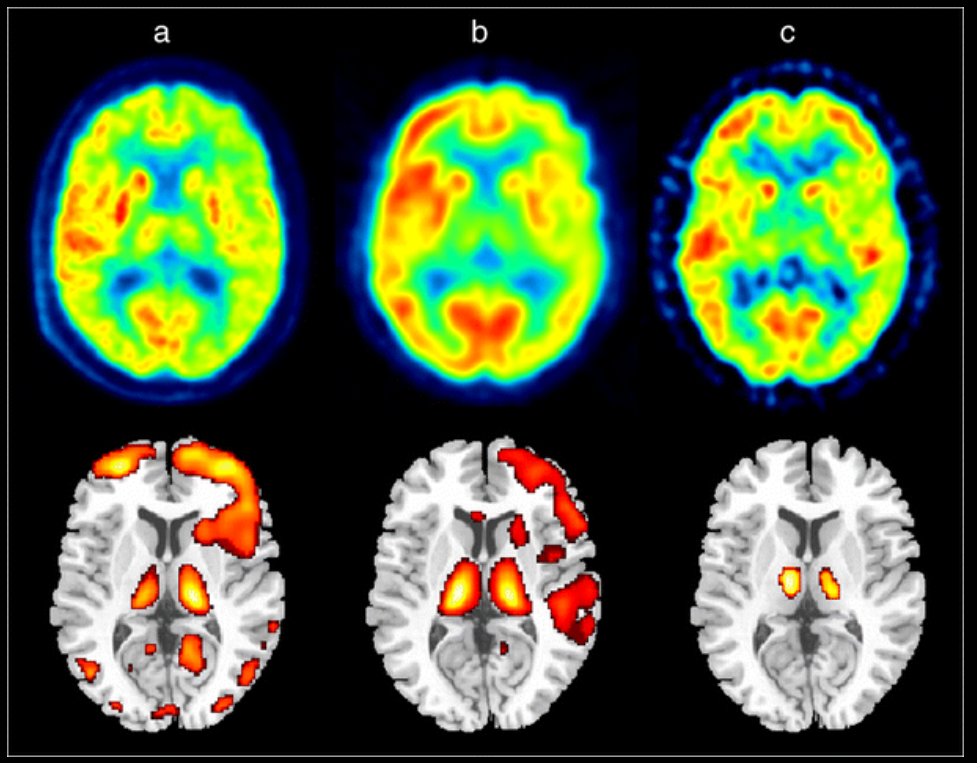
In programma…..

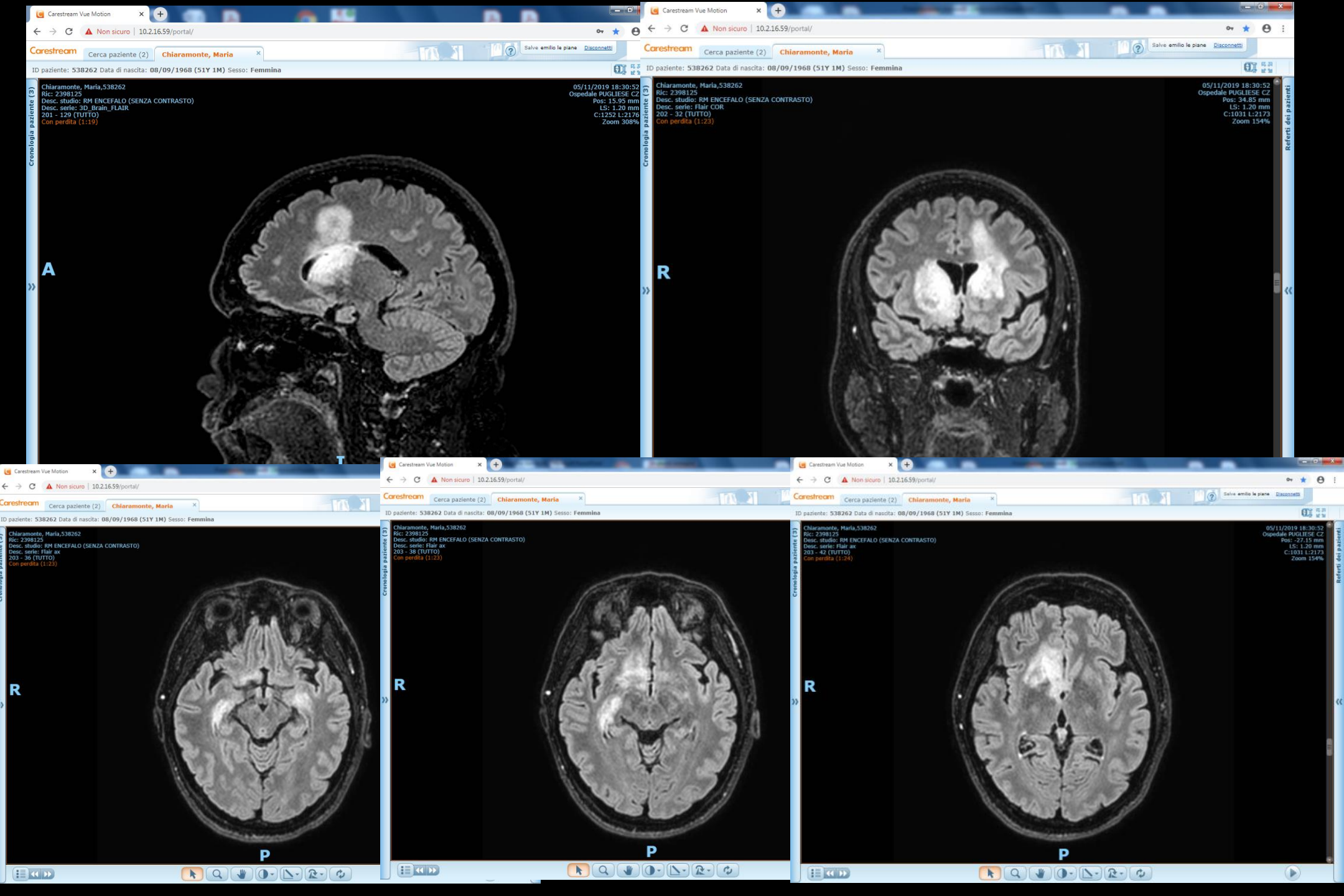
antigeni onconeurali (intracellulari)

Anti-Hu anti-Ma2 CV2-CRMP5 anti-Ri anti-amfifisina

anticorpi anti-antigeni di superficie (presenti anche in forme non paraneo)

Anti VGKC anti VGCC anti GABABR anti AMPAR antiNMDAR




Carestream Vue Motion
Non sicuro | 10.2.16.59/portal/
Carestream
Cerca paziente (2)
Chiaramonte, Maria
ID paziente: 538262 Data di nascita: 08/09/1968 (51Y 1M) Sesso: Femmina
Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: 3D_Brain_FLAIR
201 - 129 (TUTTO)
Con perdita (1:19)
05/11/2019 18:30:52
Ospedale PUGLIESE CZ
Pos: 15.95 mm
LS: 1.20 mm
C:1252 L:2176
Zoom 308%
A
T
Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: Flair COR
202 - 32 (TUTTO)
Con perdita (1:23)
05/11/2019 18:30:52
Ospedale PUGLIESE CZ
Pos: 34.85 mm
LS: 1.20 mm
C:1031 L:2173
Zoom 154%
R
Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: Flair ax
203 - 36 (TUTTO)
Con perdita (1:23)
R
P
Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: Flair ax
203 - 38 (TUTTO)
Con perdita (1:23)
R
P
Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: Flair ax
203 - 42 (TUTTO)
Con perdita (1:24)
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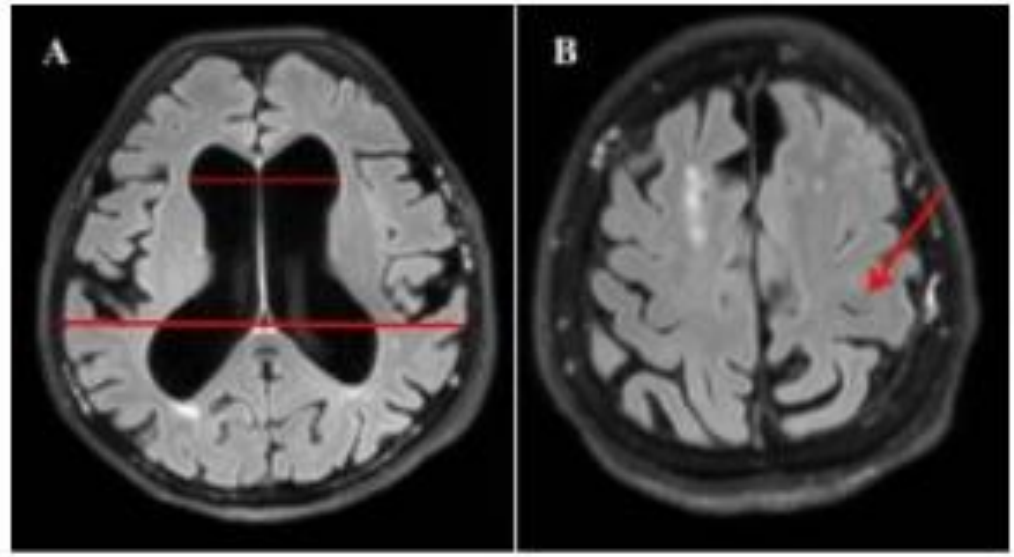
Pos: -27.15 mm
LS: 1.20 mm
C:1031 L:2173
Zoom 154%
R
P



A) Indice di Evans: rapporto tra la massima ampiezza dei corni frontali (prima linea orizzontale rossa) e il massimo diametro interno del cranio allo stesso livello (seconda linea orizzontale rossa); nell'idrocefalo normoteso il rapporto è patologico (> 0,3). B) I solchi corticali al vertice nell'idrocefalo normoteso possono essere assottigliati (freccia rossa).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Cerca paziente (2)
Chiaramonte, Maria
ID paziente: 538262 Data di nascita: 08/09/1968 (51Y 1M) Sesso: Femmina
Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: Diffusione
301 - 190 (TUTTO)
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Ospedale PUGLIESE CZ
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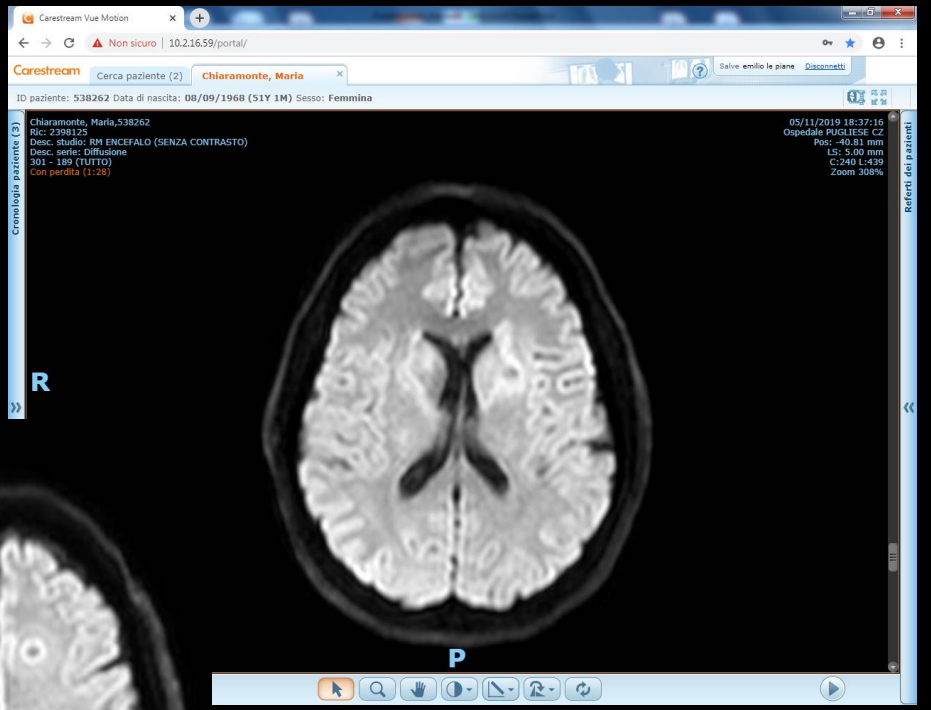
C:237 L:438
Zoom 308%
Cronologia paziente (3)
Referti dei pazienti
R
P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Chiaramonte, Maria,538262
Ric: 2398125
Desc. studio: RM ENCEFALO (SENZA CONTRASTO)
Desc. serie: Diffusione
301 - 189 (TUTTO)
Con perdita (1:28)
05/11/2019 18:37:16
Ospedale PUGLIESE CZ
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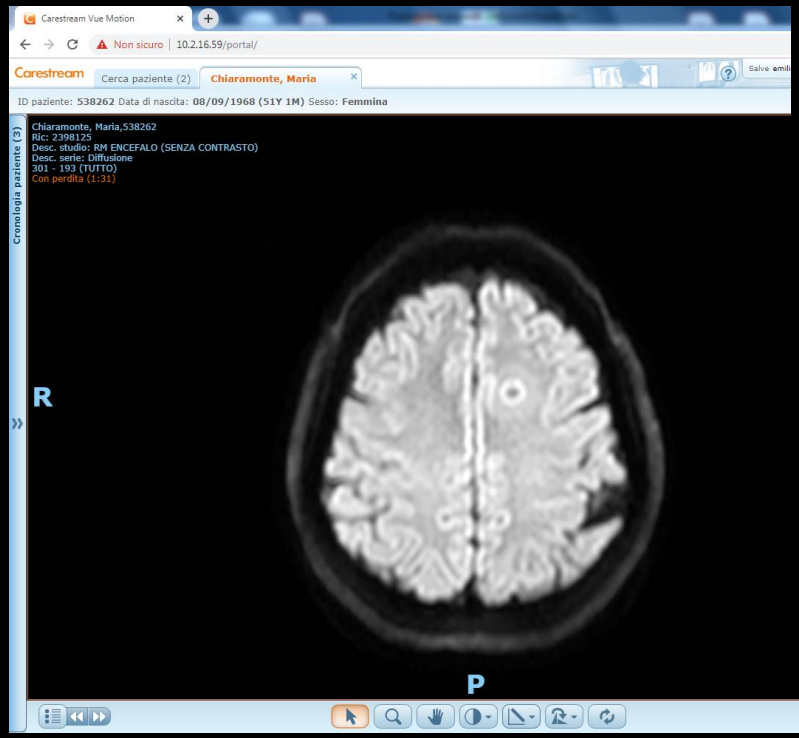
LS: 5.00 mm
C:240 L:439
Zoom 308%
Cronologia paziente (3)
Referti dei pazienti
R
P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Chiaramonte, Maria,538262
Ric: 2398125
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Desc. serie: Diffusione
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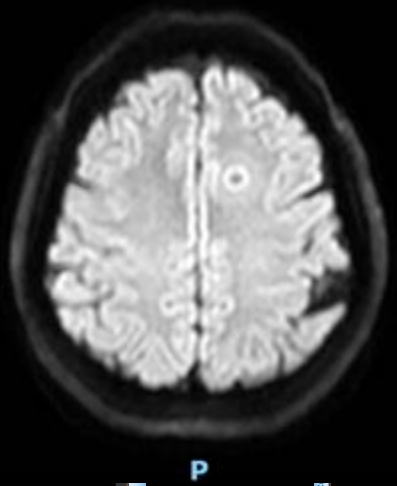
Con perdita (1:31)
Cronologia paziente (3)
R
P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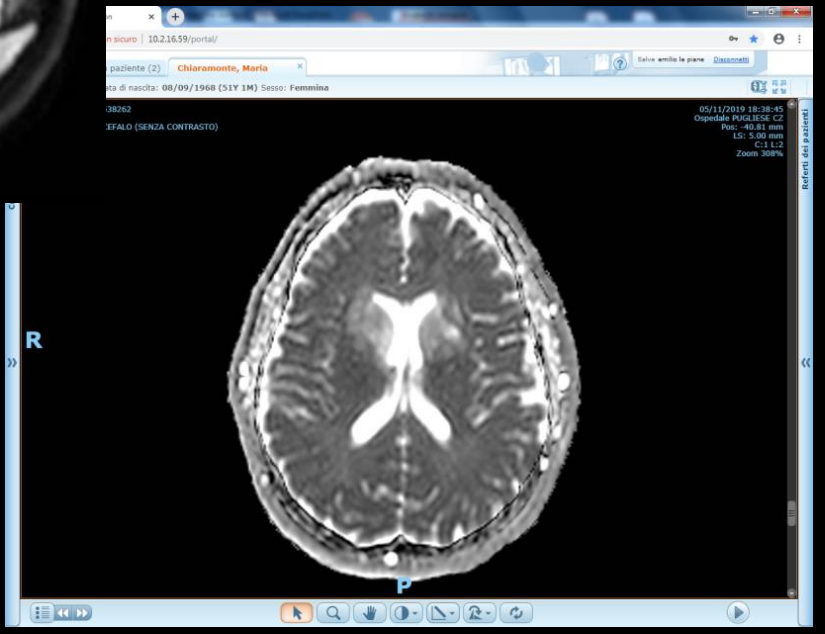
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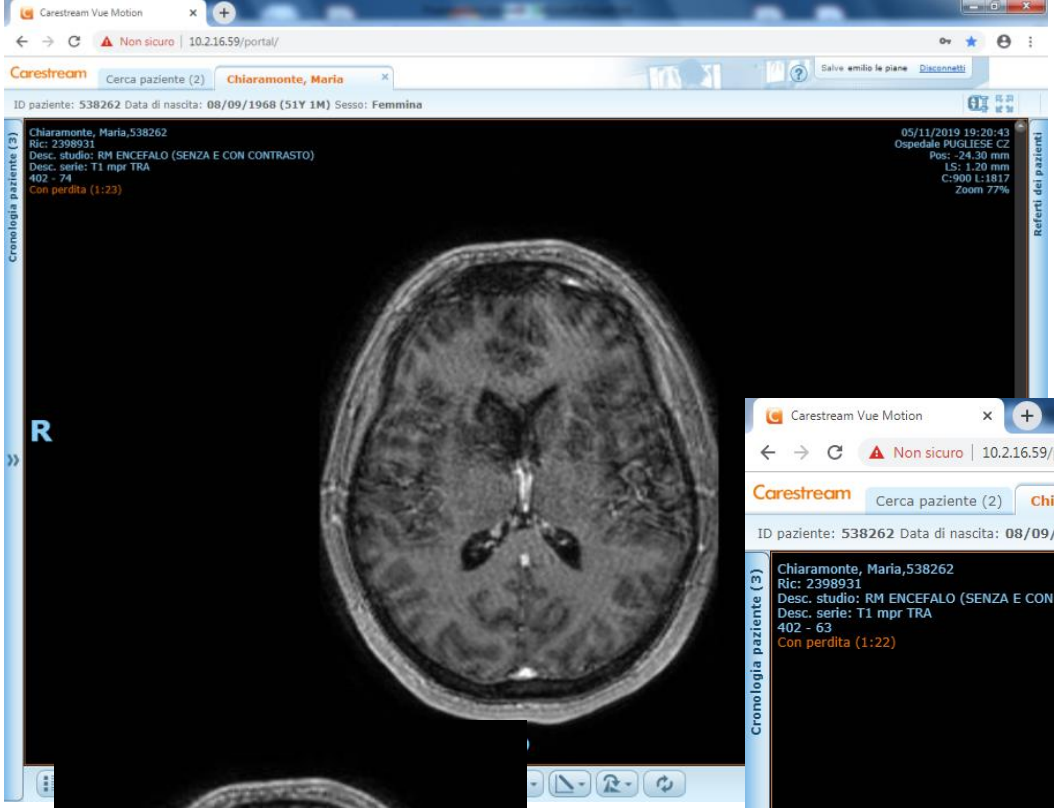
ID paziente: 538262 Data di nascita: 08/09/1968 (51Y 1M) Sesso: Femmina
05/11/2019 18:38:45
Ospedale PUGLIESE CZ
Pos: -40.81 mm
LS: 5.00 mm
C:1 L:2
Zoom 308%
Referti dei pazienti
R
P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Carestream
Cerca paziente (2)
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Salve emilio le piane
Disconnetti
ID paziente: 538262 Data di nascita: 08/09/1968 (51Y 1M) Sesso: Femmina
Chiaramonte, Maria,538262
Ric: 2398931
Desc. studio: RM ENCEFALO (SENZA E CON CONTRASTO)
Desc. serie: T1 mpr TRA
402 - 74
Con perdita (1:23)
05/11/2019 19:20:43
Ospedale PUGLIESE CZ
Pos: -24.30 mm
LS: 1.20 mm
C:900 L:1817
Zoom 77%
Cronologia paziente (3)
Referti dei pazienti
R
P


Carestream Vue Motion
Non sicuro | 10.2.16.59/portal/
Carestream
Cerca paziente (2)
Chiaramonte, Maria
Salve emilio le piane
Disconnetti
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Ric: 2398931
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Metabolic Brain Disease, Vol. 10, No. 3, 1995

Metabolic

De Novo Epilep
Cobalamin Defi

Umberto Aguglia[1,3],
Giovambattista De S

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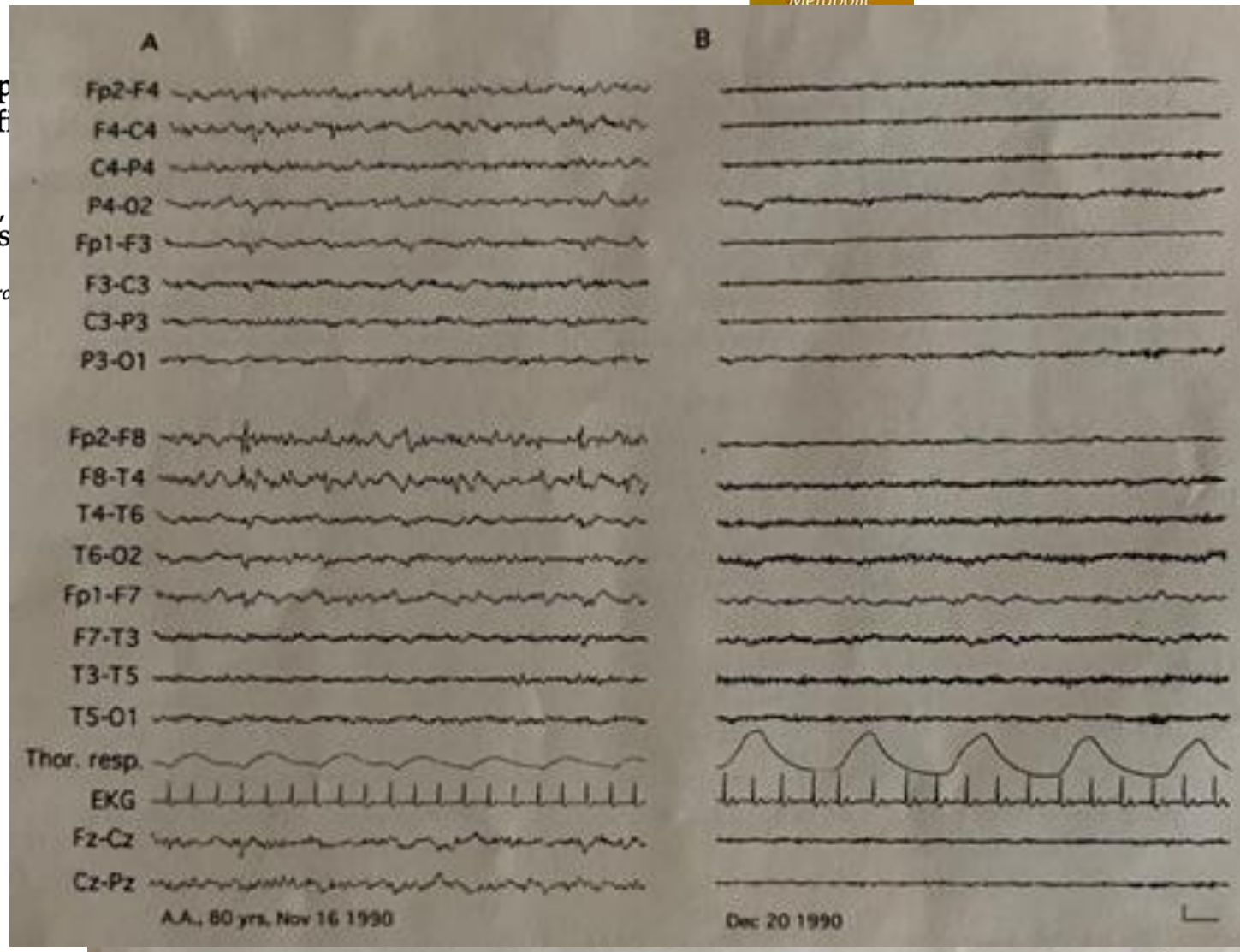


Figure 2: EEG recording. A) 72 hours after beginning of replacement therapy. Normal background activity and sporadic interictal spike-waves over the right frontal region at F8 and F4 phase reversal. B) 25 days after beginning replacement therapy. Normal posterior background and lack of any abnormal activity over the right frontal region. Calibration: 1 sec, 50 μV.

Storage diseases and other genetic causes for RPD

1. Wilson's disease
2. NBIA
3. Lysosomal storage diseases (selected): mucopolysaccharidoses, sphingolipidoses, and neuronal ceroid lipofuscinoses
4. Huntington's disease
5. CADASIL resp. CARASIL
6. Mitochondrial diseases (selected): MELAS, Leigh disease
7. PME: Lafora disease, ULD
8. Porphyria
9. Methylmalonic academia
10. DRPLA

Toxic

1. Alcohol-related dementia
2. Benzodiazepine-related dementia
3. Methyl intoxication
4. Metal intoxications (e.g. lithium, mercury)
5. Side effects of chemotherapeutics or neuroleptic drugs

Psychiatric

1. Severe depression
2. Schizophrenic disorders
3. Hypochondriac delusions

Others and secondary conditions

1. Hypoxic brain damage
2. Epilepsy and secondary seizures
3. Systemic inflammation (e.g. systemic immune-mediated diseases, septic encephalopathy)
4. NPH

Inga Zerr & Peter Hermann, 2018

Figure 1 Suggested diagnostic tests for initial rapidly progressive dementia evaluation

Blood tests	CSF	Imaging	Urine/Other
Basic panel of tests			
- Complete blood count - Basic metabolic panel (+Ca,P,Mg) - Liver function tests (including ammonia) - Renal function tests - Thyroid function tests - Anti-TG and Anti-TP antibodies - Vitamin B12/MMA/homocysteine - Rheumatologic screen (ANA, ESR, CRP, RF, ANCAs, SSA, SSB) - Rapid plasma reagin (RPR) - HIV serology - Paraneoplastic/autoimmune antibodies	- Cell count and differential - Protein - Glucose - IgG index - Oligoclonal bands - VDRL - 14-3-3/NSE/total tau	- Brain MRI (including FLAIR, DWI and ADC sequences), at least one scan with and without contrast	- Urine analysis (and culture if indicated) - EEG

Figure 1 Suggested diagnostic tests for initial rapidly progressive dementia evaluation

Blood tests	CSF	Imaging	Urine/Other
Basic panel of tests			
- Complete blood count - Basic metabolic panel (+Ca,P,Mg) - Liver function tests (including ammonia) - Renal function tests - Thyroid function tests - Anti-TG and Anti-TP antibodies - Vitamin B12/MMA/homocysteine - Rheumatologic screen (ANA, ESR, CRP, RF, ANCAs, SSA, SSB) - Rapid plasma reagin (RPR) - HIV serology - Paraneoplastic/autoimmune antibodies	- Cell count and differential - Protein - Glucose - IgG index - Oligoclonal bands - VDRL - 14-3-3/NSE/total tau	- Brain MRI (including FLAIR, DWI and ADC sequences), at least one scan with and without contrast	- Urine analysis (and culture if indicated) - EEG
Tests to consider in selected cases			
- Lyme disease (in endemic areas) - Cancer screen - Blood smear - Coagulation profile - Hypercoagulability testing - Copper and ceruloplasmin - Additional rheumatologic tests (complement, dsDNA, anti-Sm, anti-RNP, anticardiolipin, anti-SCL 70, Anti-Jo, anti-centromere antibodies)	- Bacterial, fungal, acid-fast bacilli stains and cultures - Cytology - Flow cytometry - Whipple PCR - Cryptococcal antigen - Viral PCRs and cultures	- Cancer screen (CT chest, abdomen, and pelvis with and without contrast; mammogram; body PET scan) - MR angiography or brain angiogram - MR spectroscopy - Carotid ultrasound - Echocardiogram	- Heavy metal screen (24h urine) - Copper (24h urine) - Porphobilinogen (PBG)/delta-aminolevulinic acid (ALA) in urine (24h) - EMG/nerve conduction study - Brain biopsy

Seizure: European Journal of Epilepsy 60 (2018) 94–95

Contents lists available at ScienceDirect

Seizure: European Journal of Epilepsy

journal homepage: www.elsevier.com/locate/seizure

Short communication

Seizures with Migraine-like Attacks after Radiation Therapy (SMART): A new meaning of an old acronym

Edoardo Ferlazzo[a,b,c], Michele Ascoli[a,b], Sara Gasparini[a,b], Vittoria Cianci[b], Damiano Branca[b], Chiara Sueri[b], Umberto Aguglia[a,b,c,*]

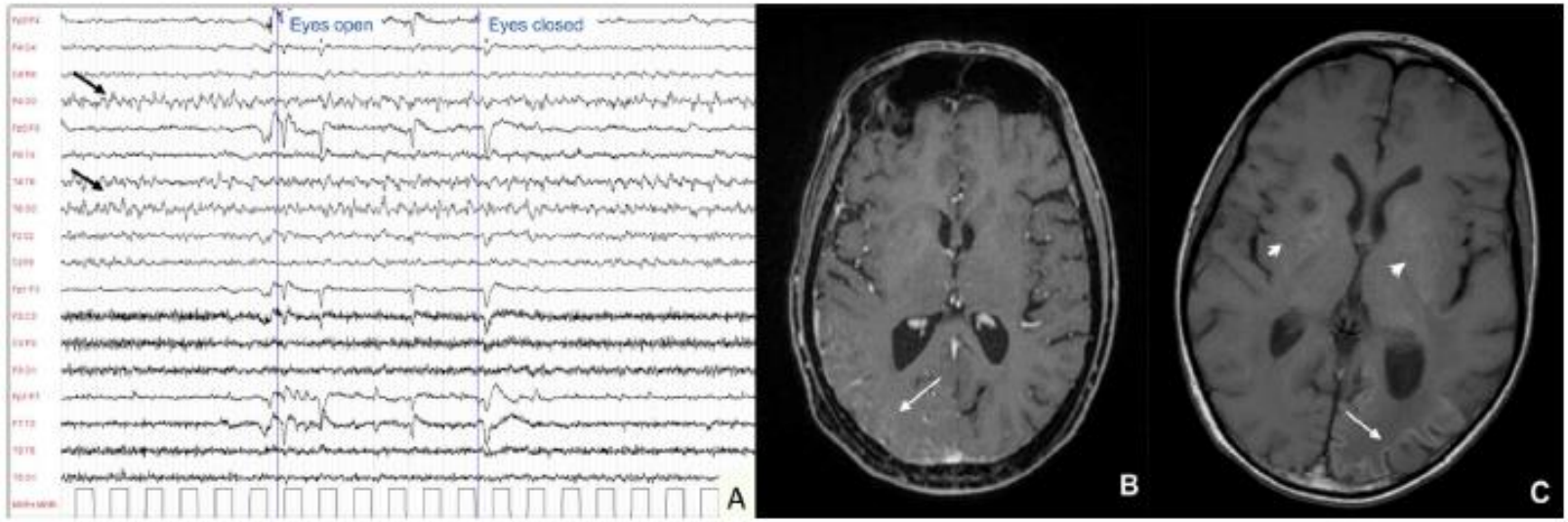
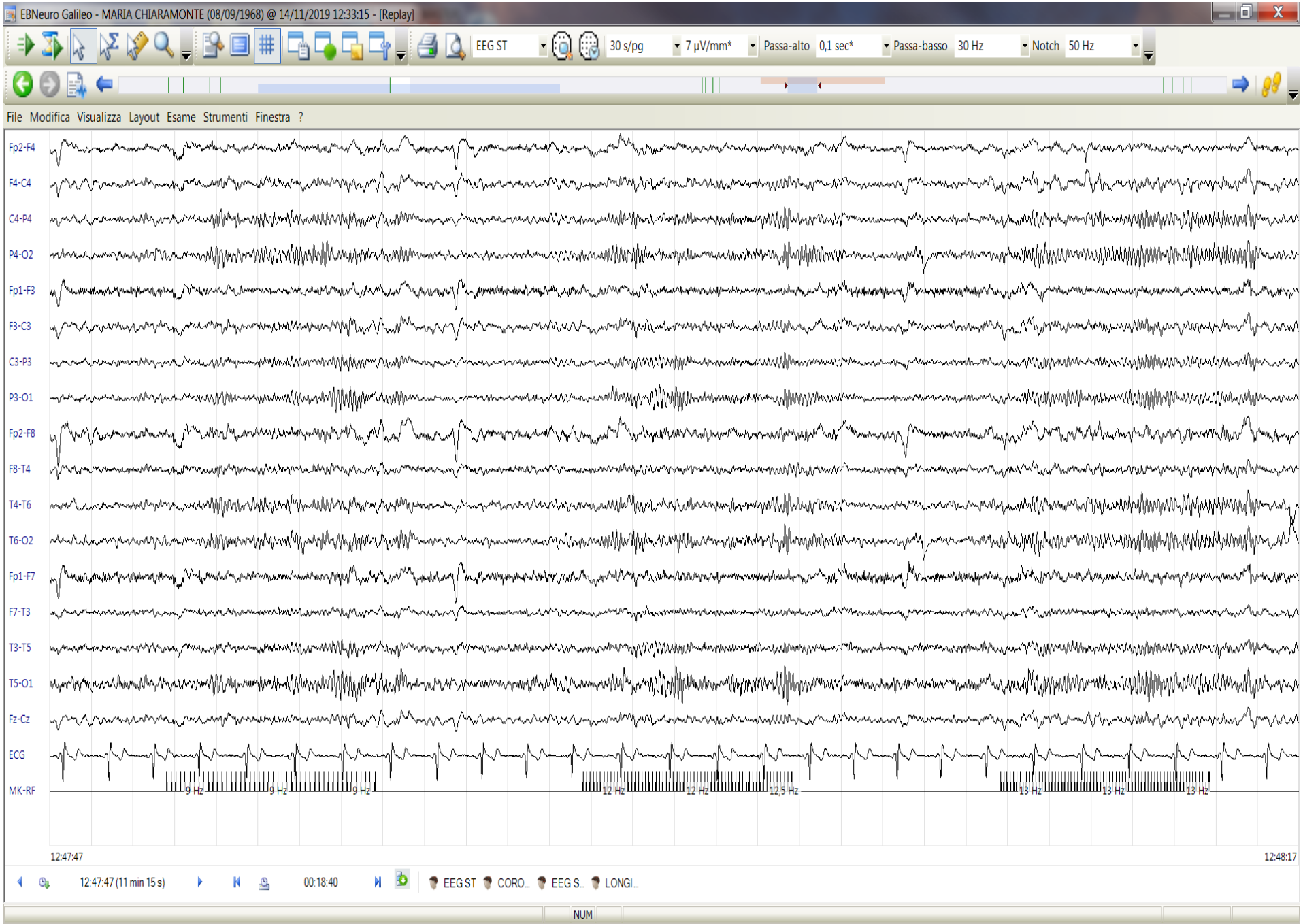


Fig. 1. a: Patient 1. Rhythmic spikes evident over the right occipital region (black arrows). b: patient 1. Axial T1-weighted image with gadolinium showing gyral enhancement over the right parieto-occipital regions (white arrow). c: Patient 2. Axial T1-weighted image. Note linear gray matter hyperintensity on the left occipital lobe (laminar necrosis: white arrow), and post-radiation leukoencephalopathy with well-evident bilateral calcifications of the basal ganglia (arrowheads).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F8-T4
T4-T6
T6-O2
Fp1-F7
F7-T3
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Clinical Interventions in Aging

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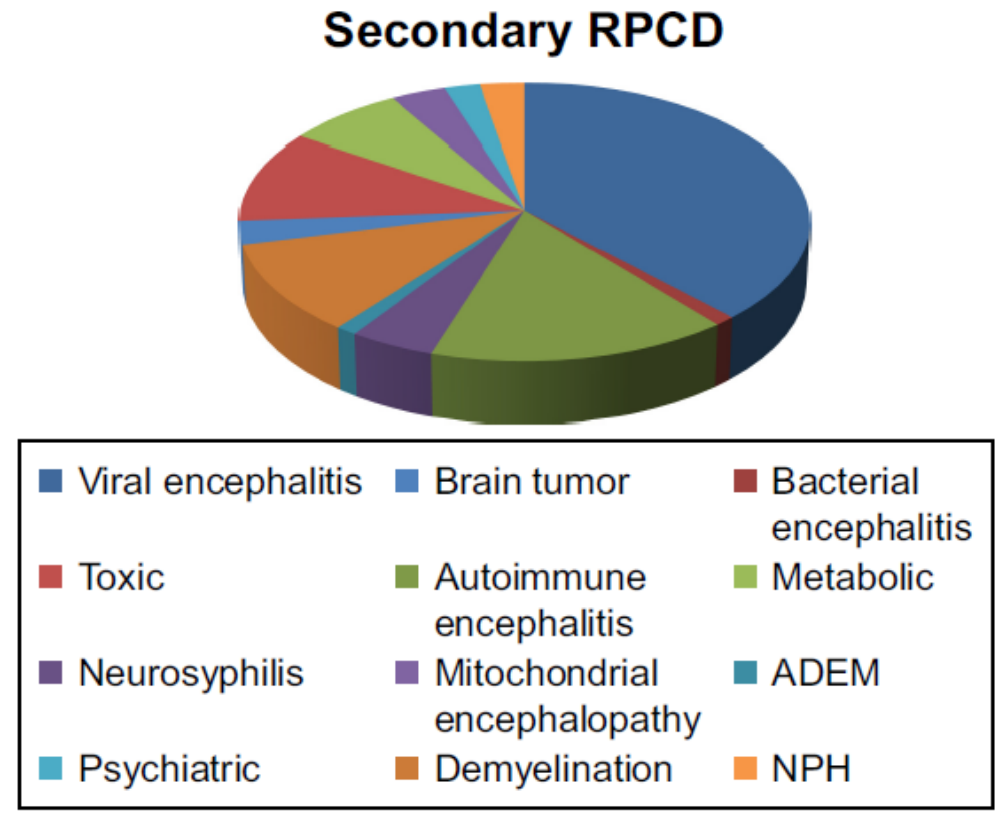
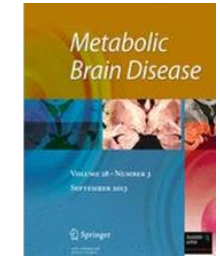
ORIGINAL RESEARCH

Spectrum of noncerebrovascular rapidly progressive cognitive deterioration: a 2-year retrospective study



Zhang et al, 2017

Metabolic Brain Disease, Vol. 10, No. 3, 1995



De Novo Epileptic Confusional Status in a Patient with Cobalamin Deficiency

Umberto Aguglia[1,3], Antonio Gambardella[1], Rosario Luciano Oliveri[1], Giovambattista De Sarro[2], Mario Zappia[1], Aldo Quattrone [1]

Received 15 March 1995; Revised version received 23 June 1995; Accepted 18 July 1995

A 80-year-old man with cobalamin deficiency and no history of epilepsy developed a partial complex epileptic confusional status (ECS) unresponsive to acute i.v. diazepam. Brain CT scan and MRI investigation ruled out a focal cerebral lesion. Therapy with high doses (10,000 µg i.m. daily) of cobalamin alone was started, and the patient fully recovered in the following 72-hour. Control EEGs repeatedly performed days and weeks later showed progressive disappearance of the frontal interictal spiking, while the patient was on monotherapy with cobalamin (5,000 µg i.m. weekly). A month later the patient unfortunately discontinued replacement therapy and 13 weeks later he developed a fatal convulsive epileptic status. To our knowledge the association of ECS and cobalamin-deficiency has not been previously reported.

Box 1. Primary and secondary causes of rapidly progressive dementia, divided by etiology.

Neurodegenerative

- Alzheimer's disease
- Corticobasal degeneration
- Dementia with Lewy bodies
- Familial spastic paraparesis
- Frontotemporal lobar degenera
- Motor neuron disease
- Progressive supranuclear palsy
- Prion disease (i.e., Creutzfeldt–J
- Progressive subcortical gliosis

Autoimmune/inflammatory[†]

- Acute disseminated encephalor
- Antibody-mediated brain disea
- Anti-GAD65 autoimmunity
- Behçet's disease
- Celiac sprue
- Limbic encephalitis
- Multiple sclerosis
- Neuropsychiatric lupus
- Sarcoidosis
- Sjögren's syndrome
- Steroid-responsive encephalop

Vascular[†]

- CADASIL
- Cerebral amyloid angiopathy
- CNS vasculitis[‡]
- MELAS
- Strategic infarction
- Subdural hematoma
- Vascular dementia

Metabolic[†]

- Cerebrotendinous xanthomatosis
- Extrapontine myelinolysis
- Liver failure
- MELAS
- NBIA
- Neuronal ceroid lipofucinosis
- Nutritional deficiency (i.e., vitamin B1, B3 or B
- Porphyria
- Renal failure

Box 1. Primary and secondary causes of rapidly progressive dementia, divided by etiology (cont.).

Metabolic[†] **(cont.)**

- Thyroid/parathyroid dysfunction
- Wilson's disease

Toxic[†]

- Bismuth
- Heavy metal (lead, arsenic and mercury)
- Manganese
- Medication-induced
- PRES
- Radiation-induced leukoencephalopathy

Neoplastic

- Brain metastases
- Lymphomatoid granulomatosis
- Paraneoplastic limbic encephalitis
- Primary CNS neoplasms

Infectious[†,§]

- Bacterial
- Fungal
- Parasites
- Viral
- Other

- Bipolar affective disorder
- Hypoxic-ischemic encephalopathy
- Normal pressure hydrocephalus
- Other psychiatric illnesses
- Schizophrenia

Box 5 Chameleon presentations of non-convulsive status epilepticus (NCSE presentations that might be overlooked)

Disorders of consciousness
- Alert and unresponsive
- Coma

Disturbances of speech/language
- Aphasia
- Reduced verbal fluency to mute
- Echolalia
- Stuttering

Psychiatric/behavioural disturbance
- Psychosis
- Mood disturbance
- Fear/agitation

Cognitive dysfunction
- Amnesia
- Confusion
- Alexia
- Confabulation

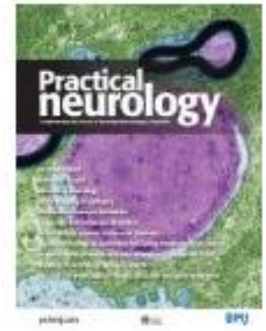
Movement/motor disorders
- Catatonia
- Myoclonus
- Gaze deviation
- Limb paralysis

Perceptual/sensory disturbances
- Hallucinations (olfactory, gustatory, auditory, visual)
- Blindness
- Sensory disturbance and pain (including headache)

Box 7 Mimics of NCSE

Common disorders
- **Postictal state**
- **Toxic/metabolic encephalopathy**
- Non-epileptic attack disorder (psychogenic non-epileptic status)

Less common disorders
- **Autoimmune encephalitis**
- **Cerebral infarction**
- Persistent vegetative state
- **Drug withdrawal**
- Primary psychiatric presentation, for example, psychosis in schizophrenia
- Malingering
- Migraine with aura
- Transient global amnesia
- Neuroleptic malignant syndrome
- Serotonin syndrome
- **Mitochondrial disorder such as mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes**
- **Stroke-like migraine attacks after radiation therapy (SMART syndrome)**



Kinney MO et al, 2018

Panel 2: **Clinical settings related to Wernicke's encephalopathy**

Staple diet of polished rice

Chronic alcohol abuse and malnutrition

Gastrointestinal surgical procedures
Surgical procedures: gastrectomy; gastrojejunostomy; partial or subtotal colectomy; gastric bypass surgery; vertical banded gastroplasty; therapy with an intragastric balloon
Disorders: peptic ulcer; gastric cancer; colon cancer; ulcerative colitis with megacolon; severe obesity

Recurrent vomiting or chronic diarrhoea
Pyloric stenosis; peptic ulcer; drug-induced gastritis; biliary colics; Crohn's disease; intestinal obstruction or perforation; lithium-induced diarrhoea; migraine attacks; anorexia nervosa; pancreatitis; hyperemesis gravidarum

Cancer and chemotherapeutic treatments
Cancer and related conditions: gastric carcinoma; non-Hodgkin's lymphoma; myelomonocitic leukaemia; large B-cell lymphoma; myeloid leukaemia; allogenic bone marrow transplantation
Chemotherapeutics: erbulozole; ifosfamide

Systemic diseases
Renal diseases; AIDS; chronic infectious febrile diseases; thyrotoxicosis

Magnesium depletion
Secondary to chronic diuretic therapy; intestinal tract resection; Crohn's disease

Use of chemical compounds and drugs
Intravenous infusion of high-dose nitroglycerin; tolazamide

Unbalanced nutrition
Absolute deficiency of food/thiamine: dietary restrictions owing to economic reasons or political trade embargoes; psychogenic food refusal; fasting for religious-philosophical reasons; starvation for treatment of obesity; hunger strike; neglect in old age or Alzheimer's disease
Relative deficiency of thiamine: unbalanced total parenteral nutrition; unbalanced intravenous hyperalimentation; re-feeding syndrome; use of dietary commercial formulae; slimming diets; excessive cooking of food; chronic use of food containing thiaminases or antithiamine factors; chronic use of sulphites as food additives (dogs)

Table 1. Findings on history and physical examination supportive of an etiologic diagnosis.

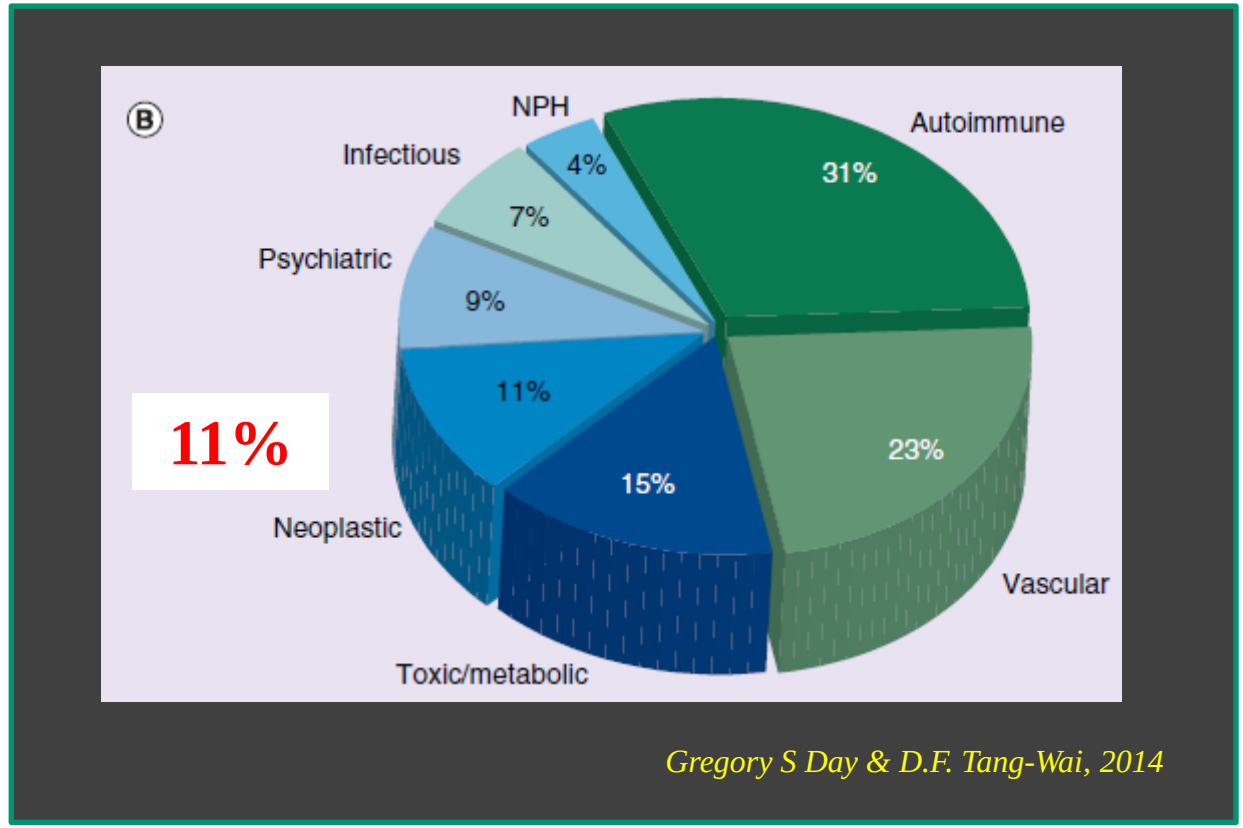
Finding	CJD	Neurodegenerative	Autoimmune/ inflammatory	Vascular	Metabolic	Toxic	Neoplastic/ paraneoplastic	Infectious
History								
Age (years):								
Young (<50)	vCJD		X	X	X	X		X
Old (≥50)	sCJD	X	X	X	X	X	X	X
Onset:								
Acute†	X		X	X	X	X		X
Subacute‡	X	X					X	
Relapsing–remitting		X (DLB)	X	X	X	X		
Symptoms of limbic encephalitis			X				X	
Systemic signs			X				X	X
Neurological examination								
Upper motor neuron signs	X		X	X	X		X	X
Parkinsonism	X	X		X	X	X		X (SSPE)
Myoclonus	X	X			X	X		X
Asterixis	X	X			X			
Peripheral neuropathy					X	X		

†Acute onset occurs over days to weeks.
‡Subacute onset occurs over months to years.
CJD: Creutzfeldt–Jakob disease; DLB: Dementia with Lewy bodies; sCJD: Sporadic Creutzfeldt–Jakob disease; SSPE: Subacute sclerosing panencephalitis; vCJD: Variant Creutzfeldt–Jakob disease; X: Expected findings.

Table 4. EEG patterns of interest in rapidly progressive dementia.

EEG finding	Definition	Potential etiologies
Periodic complexes	Generalized discharges of synchronous high-voltage spikes or sharp waves	CJD [93] SSPE [94] Rarely Alzheimer's disease [93] or DLB [95]
Extreme δ-brush	Rhythmic δ activity (1–3 Hz) with superimposed bursts of 20–30 Hz β-frequency activity riding on each δ-wave	Anti-NMDA receptor encephalitis [15,96]
Triphasic waves	Synchronous, frontally predominant, rhythmic triphasic waves, usually with background slowing	Metabolic disorders (i.e., hepatic encephalopathy) [97], nonconvulsive status epilepticus
PLEDs	High-voltage sharp potentials over one or both lobes, occurring every few seconds	Herpes simplex encephalitis (temporal PLED) [80], other focal lesions
Frontal intermittent rhythmic δ-activity	Rhythmic, discontinuous high-voltage δ-frequency (1–3 Hz) activity that predominates in frontal regions	Processes involving deep midline structures (i.e., hydrocephalus), other focal lesions [63]

CJD: Creutzfeldt–Jakob disease; DLB: Dementia with Lewy bodies; PLED: Periodic lateralized epileptiform discharge; SSPE: Subacute sclerosing panencephalitis.



Gregory S Day & D.F. Tang-Wai, 2014

PLOS ONE

RESEARCH ARTICLE

Rapidly progressive dementia: An eight year (2008–2016) retrospective study

Patil Anuja[1☯], Vishnu Venugopalan[2‡*], Naheed Darakhshan[1☯], Pandit Awadh[2‡], Vinny Wilson[1‡], Goyal Manoj[1‡], Modi Manish[1‡], Lal Vivek[1‡]

PLOS ONE | https://doi.org/10.1371/journal.pone.0189832 January 18, 2018

Table 3. Associated neurological deficits in patients with rapidly progressive dementia.

Sr.no	Category:		No of patients: n (%)
1	Vision loss		17 (9.09)
2	Other cranial nerve deficits		8 (4.2)
3	Pyramidal signs		37 (19.7)
4	Extrapyramidal signs and/or gait		54 (28.8)
5	Cerebellar signs		26 (13.9)
6	Small fiber neuropathy		8 (4.2)
7	Visual hallucinations		9 (4.8)
8	Generalized and/or focal seizures		40 (21.3)
9	Sleep disturbances	REM disorders	9 (4.8)
		Hypersomnolence	5 (2.6)
10	Hyperkinetic movement disorders:	Myoclonus	32 (17.1)
		Opsoclonus	4 (2.1)
		Choreo-athetosis	6 (3.1)
		Dystonia	12 (6.4)
		Dyskinesia	6 (3.1)

Altre etiologie

ascular (strokes, clotting of brain veins);

nfectious (HIV, encephalitis, fungal, parasites);

oxic-Metabolic (medicines, vitamin excess/deficiency, toxins);

utoimmune (Antibody-mediated, rheumatological, cancer-related);

etastases/Neoplasm (cancer);

atrogenic (brought forth by your doctor);

eurodegenerative (Alzheimer's, Parkinson's, Primary Progressive Aphasia, Lewy Bodies);

ystemic/Seizures/Structural.

Open Access Full Text Article

RAPID COMMUNICATION

Practical way of creating differential diagnoses through an expanded *VITAMINSABCDEK* mnemonic

This article was published in the following Dove Press journal:
Advances in Medical Education and Practice
22 April 2016
Number of times this article has been viewed

Zabidi A Zabidi-Hussin

Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia

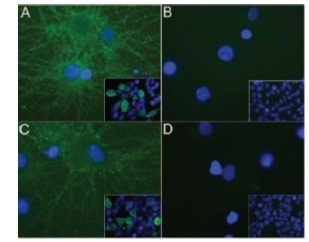
Abstract : Having an organized, structured thinking process is critical in medicine. It is through this thinking process that enables one to go through the method of history-taking, which will eventually lead to making a definitive diagnosis and all other processes that follow. The use of mnemonic has been found to be useful for this purpose. The mnemonic VITAMINSABCDEK, is a convenient and practical way to assist in expanding the differential diagnoses and covers all possible causes of an illness. It is also easy to remember, as the vitamins whose letters are represented in this mnemonic cover the entire range of vitamins known.

Keywords: mnemonic, differential diagnosis

“V” stands for “vascular”, where the disease can be caused by “vessel” (bleed or blocked), or anything related to hematology. “I” is for “infective” or “post infective” causes. “T” is for “trauma” or anything caused by mechanical factors such as obstructions or pressure. “A” is for causes of “autoimmune”-related illnesses or “allergy”. “M” is for “metabolic” causes affecting lipids, proteins, carbohydrates, or micronutrients. “I” is for “idiopathic” or “iatrogenic” causes. “N” is for “neoplasia”. “S” is for diseases caused by “social” reasons, such as child abuse and social deprivation. “A” is for diseases caused by “alcohol”-related issues. “B” is for diseases caused by “behavioral” or psychosomatic disorders. “C” is for diseases caused by “congenital” problems (the entire VITAMINSABCDEK could be applied again in the congenital causes). “D” is for diseases caused by “degenerative” disor-

Neurotherapeutics (2016) 13:147–162
DOI 10.1007/s13311-015-0410-6

REVIEW



Antibody-Mediated Autoimmune Encephalopathies and Immunotherapies

Matteo Gastaldi[1,2] · Anaïs Thouin[3] · Angela Vincent[1]

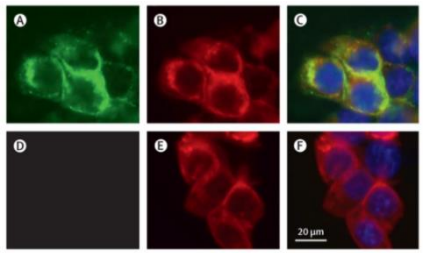
JCN Open Access REVIEW
pISSN 1738-6586 / eISSN 2005-5013 / J Clin Neurol 2016;12(1):1-13 / http://dx.doi.org/10.3988/jcn.2016.12.1.1

The Diagnosis and Treatment of Autoimmune Encephalitis

Position Paper

Eric Lancaster

Department of Neurology, University of Pennsylvania, Philadelphia, PA, USA



A clinical approach to diagnosis of autoimmune encephalitis

Francesc Graus, Maarten J Titulaer, Ramani Balu, Susanne Benseler, Christian G Bien, Tania Cellucci, Irene Cortese, Russell C Dale, Jeffrey M Gelfand, Michael Geschwind, Carol A Glaser, Jerome Honnorat, Romana Höftberger, Takahiro Iizuka, Sarosh R Irani, Eric Lancaster, Frank Leypoldt, Harald Prüss, Alexander Rae-Grant, Markus Reindl, Myrna R Rosenfeld, Kevin Rostásy, Albert Saiz, Arun Venkatesan, Angela Vincent, Klaus-Peter Wandinger, Patrick Waters, Josep Dalmau

Encephalitis is a severe inflammatory disorder of the brain with many possible causes and a complex differential diagnosis. Advances in autoimmune encephalitis research in the past 10 years have led to the identification of new syndromes and biomarkers that have transformed the diagnostic approach to these disorders. However, existing criteria for autoimmune encephalitis are too reliant on antibody testing and response to immunotherapy, which might delay the diagnosis. We reviewed the literature and gathered the experience of a team of experts with the aims of developing a practical, syndrome-based diagnostic approach to autoimmune encephalitis and providing guidelines to navigate through the differential diagnosis. Because autoantibody test results and response to therapy are not available at disease onset, we based the initial diagnostic approach on neurological assessment and conventional tests that are accessible to most clinicians. Through logical differential diagnosis, levels of evidence for autoimmune encephalitis (possible, probable, or definite) are achieved, which can lead to prompt immunotherapy.

Lancet Neurol 2016; 15: 391–404
Published Online
February 19, 2016
http://dx.doi.org/10.1016/S1474-4422(15)00401-9

See Comment page 349

Neuroimmunology Program, Institut d'Investigacions Biomèdiques August Pi i Sunyer, Barcelona, Spain (Prof F Graus MD, Prof M R Rosenfeld MD,

Totino, Michela,531094
Ric: 2352569
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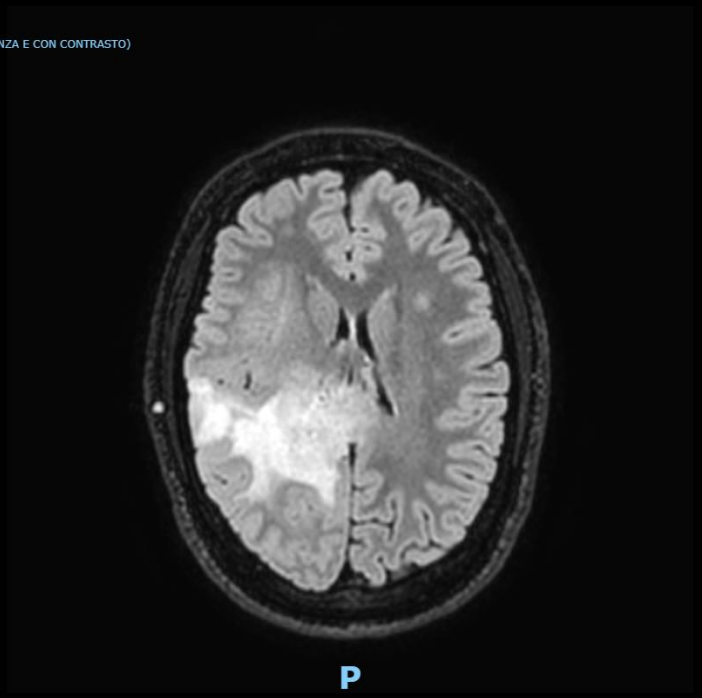

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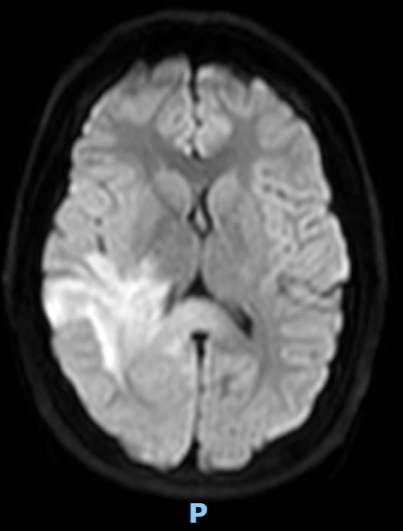

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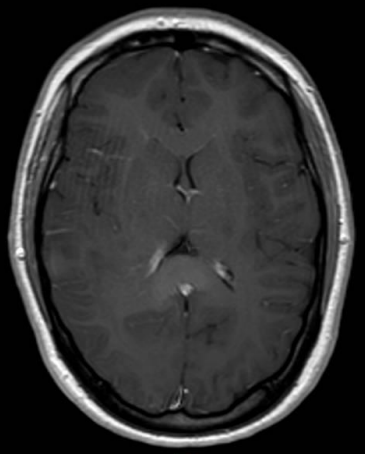

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BMJ Case Rep. 2019 May 14;12(5). pii: e229044. doi: 10.1136/bcr-2018-229044.

Reversible dementia, psychotic symptoms and epilepsy in a patient with vitamin B_{12} deficiency.

Silva B[1,2], Velosa A[1], Barahona-Corrêa JB[1,2,3].

Author information

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Abstract

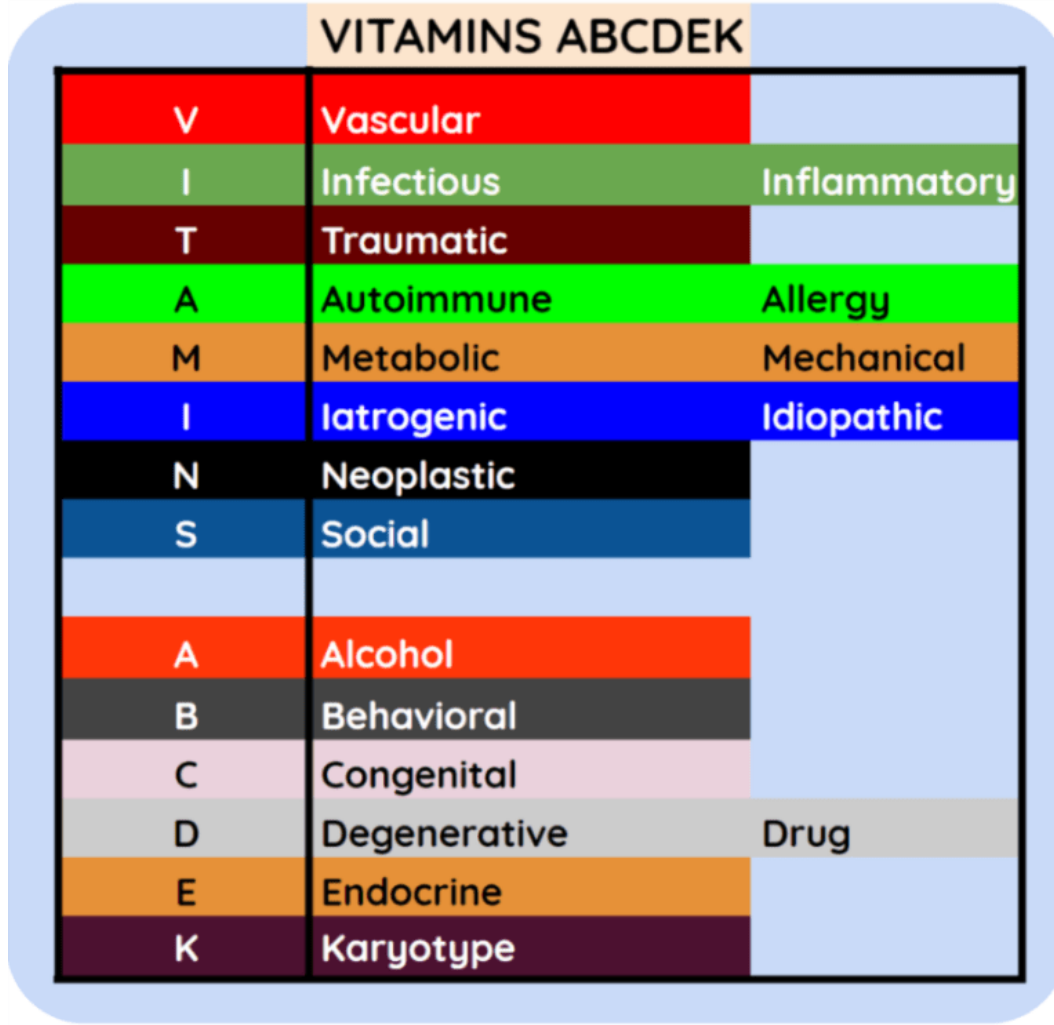
Vitamin B_{12} deficiency is a common condition, typically associated with megaloblastic anaemia, glossitis and neuropsychiatric symptoms. We report the case of a patient presenting with progressive cognitive and functional deterioration, psychosis and seizures, later found to be secondary to pernicious anaemia. Importantly, the diagnosis of pernicious anaemia was only established 5 years after symptom onset and was overlooked even when the patient was under medical care, in part due to the lack of classic neurological and haematological signs associated with the condition. The patient had a remarkable neuropsychiatric recovery after vitamin replacement and psychopharmacological management. We discuss similar presentations of vitamin B_{12} deficiency found in the literature, symptom reversibility and the importance of its early recognition and treatment.

DIFFERENTIAL DIAGNOSIS – VITAMIN CDE

Organizing principle for differential diagnoses based on etiology:

- **V**ascular
- **I**nfectious / Inflammatory
- **T**raumatic / Toxic
- **A**utoimmune
- **M**etabolic
- **I**atrogenic / Idiopathic
- **N**eoplastic

- **C**ongenital
- **D**egenerative
- **E**ndocrine



A clinical approach to diagnosis of autoimmune encephalitis

Francesc Graus, Maarten J Titulaer, Ramani Balu, Susanne Benseler, Christian G Bien, Tania Cellucci, Irene Cortese, Russell C Dale, Jeffrey M Gelfand, Michael Geschwind, Carol A Glaser, Jerome Honnorat, Romana Höftberger, Takahiro Iizuka, Sarosh R Irani, Eric Lancaster, Frank Leypoldt, Harald Prüss, Alexander Rae-Grant, Markus Reindl, Myrna R Rosenfeld, Kevin Rostásy, Albert Saiz, Arun Venkatesan, Angela Vincent, Klaus-Peter Wandinger, Patrick Waters, Josep Dalmau

Lancet Neurol 2016; 15: 391–404

Panel 1: Diagnostic criteria for possible autoimmune encephalitis

Diagnosis can be made when all three of the following criteria have been met:

1 Subacute onset (rapid progression of less than 3 months) of working memory deficits (short-term memory loss), altered mental status*, or psychiatric symptoms
2 At least one of the following:
   - New focal CNS findings
   - Seizures not explained by a previously known seizure disorder
   - CSF pleocytosis (white blood cell count of more than five cells per mm^3)
   - MRI features suggestive of encephalitis†
3 Reasonable exclusion of alternative causes (appendix)

*Altered mental status defined as decreased or altered level of consciousness, lethargy, or personality change. †Brain MRI hyperintense signal on T2-weighted fluid-attenuated inversion recovery sequences highly restricted to one or both medial temporal lobes (limbic encephalitis), or in multifocal areas involving grey matter, white matter, or both compatible with demyelination or inflammation.

Panel 2: Diagnostic criteria for definite autoimmune limbic encephalitis

Diagnosis can be made when all four* of the following criteria have been met:

1 Subacute onset (rapid progression of less than 3 months) of working memory deficits, seizures, or psychiatric symptoms suggesting involvement of the limbic system
2 Bilateral brain abnormalities on T2-weighted fluid-attenuated inversion recovery MRI highly restricted to the medial temporal lobes†
3 At least one of the following:
   - CSF pleocytosis (white blood cell count of more than five cells per mm^3)
   - EEG with epileptic or slow-wave activity involving the temporal lobes
4 Reasonable exclusion of alternative causes (appendix)

*If one of the first three criteria is not met, a diagnosis of definite limbic encephalitis can be made only with the detection of antibodies against cell-surface, synaptic, or onconeural proteins. †^{18}Fluorodeoxyglucose (^{18}F-FDG) PET can be used to fulfil this criterion. Results from studies from the past 5 years suggest that ^{18}F-FDG-PET imaging might be more sensitive than MRI to show an increase in FDG uptake in normal-appearing medial temporal lobes.[44,45]

Therapeutic Apheresis and Dialysis

Letter to the Editor | Full Access

Wernicke–Korsakoff Syndrome in End-Stage Renal Disease: A Case Report

The patient was started on therapy with intravenous (IV) high-dose thiamine immediately: 500 mg thrice a day for 5 days, continuing with 250 mg thrice a day for seven days and modulating the dose based on response. Considering the higher loss of water-soluble vitamins in peritoneal dialysis patients, we decided to position a temporary central venous catheter in the right jugular vein and start HD treatment in order to reach a better metabolic balance.

There was a noticeable recovery in the patient in the next 24 h, with improvement of the state of consciousness; the agitation was attenuated and the speech became more articulated. However, only during the following week did complete recovery occur, with continued dose of thiamine at 100 mg thrice a day.

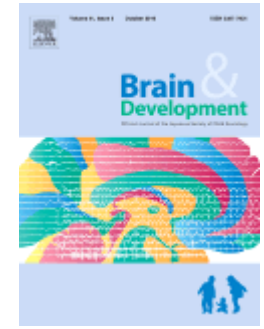
Nicotera R et al, 2018

ELSEVIER

Brain & Development 40 (2018) 42–48

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev



Original article

Hashimoto encephalopathy in pediatric patients: Homogeneity in clinical presentation and heterogeneity in antibody titers

Jiwon Lee [a], Hee Joon Yu [b], Jeehun Lee [a,*]

[a] Department of Pediatrics, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Republic of Korea
[b] Department of Pediatrics, Hanllym University Sacred Heart Hospital, Anyang, Republic of Korea

Received 1 May 2017; received in revised form 19 July 2017; accepted 20 July 2017

Abstract

Objective: Hashimoto encephalopathy is an autoimmune encephalopathy characterized by elevated antithyroid antibodies and a favorable response to corticosteroid. This study delineated the clinical characteristics of pediatric Hashimoto encephalopathy and the significance of low antithyroid antibody titers in diagnosis and treatment.

Subjects and methods: Clinical manifestations, antibody titers, and treatment responses were retrospectively reviewed in six consecutive children diagnosed with Hashimoto encephalopathy between August 2008 and July 2016.

Results: Age at diagnosis was 10–17 years. Presenting symptoms were seizures, altered consciousness, behavioral changes, psychosis, tremor, and dystonia. Thyroid function was normal in five patients, and one had hypothyroidism prior to the encephalopathy. Antithyroid antibody titer was increased at presentation in five patients and one week later in the other. Antibody levels were extremely varied (anti-thyroglobulin, 20.5–2318.0 U/ml; anti-thyroid peroxidase, 12.5–2231.0 U/ml; reference range, <60 U/ml) and <180 U/ml in two patients. Electroencephalogram was abnormal in five patients. Brain magnetic resonance imaging was unremarkable. Four patients responded to high-dose corticosteroid and one improved with additional intravenous immunoglobulin. The remaining patient did not respond to both treatments and normalized after plasmapheresis. Autoantibody titers decreased with treatment response in the acute stage. Two patients with low antibody titers showed similar clinical presentations and responses.

Conclusions: The clinical presentations and treatment responses in Hashimoto encephalopathy were similar, irrespective of antithyroid antibody titer. Because the initial antithyroid antibody titers can be normal or mildly-elevated, follow-up testing of antithyroid antibodies is required in patients who are clinically suspect for Hashimoto encephalopathy.

Table 1
Clinical characteristics and laboratory findings.

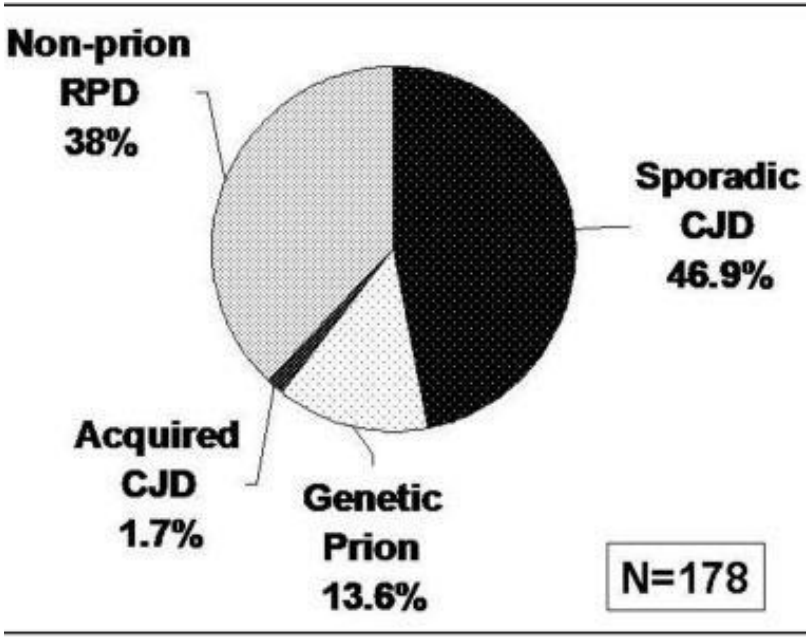
Total patients	$n = 6$
Female: male	5:1
Age at diagnosis, years (range)	14.1 ± 3.2 (10–17.6)
Duration of admission, days (range)	38.0 ± 31.6 (9–86)
Response to treatment, days (range)	6.3 ± 4.5 (1–14)
Relapse, *n* (%)	1 (16.7)
Neurologic symptoms, n (%)	
Seizures	4 (66.7)
Alternation of consciousness	6 (100)
Behavioral changes	5 (83.3)
Psychosis	4 (66.7)
Abnormal movements	2 (33.3)
Transient amnesia	1 (16.7)
Thyroid function, n (%)	
Euthyroid, subclinical hypothyroid	5 (83.3)
Hypothyroid	1 (16.7)
Elevated antithyroid antibodies initially, n (%)	
Anti-thyroglobulin antibody	5 (83.3)
Anti-thyroid peroxidase antibody	2 (33.3)
Anti-TSH receptor antibody	1 (16.7)
Treatment to encephalopathy, n (%)	
Corticosteroid	6 (100)
Immunoglobulin	3 (50)
Plasmapheresis	1 (16.7)

CLINICAL UPDATE

Rapidly Progressive Dementia

Michael D. Geschwind, MD, PhD,[1] Huidy Shu, MD, PhD,[1] Aissa Haman, MD,[1] James J. Sejvar, MD,[2] and Bruce L. Miller, MD[1]

Ann Neurol 2008;64:97–108



ORIGINAL CONTRIBUTION

ARCHIVES OF NEUROLOGY

Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Pablo Castillo, MD; Bryan Woodruff, MD; Richard Caselli, MD; Steven Vernino, MD, PhD; Claudia Lucchinetti, MD; Jerry Swanson, MD; John Noseworthy, MD; Allen Aksamit, MD; Jonathan Carter, MD; Joseph Sirven, MD; Gene Hunder, MD; Vahab Fatourechi, MD; Bahram Mokri, MD; Daniel Drubach, MD; Sean Pittock, MD; Vanda Lennon, MD, PhD; Brad Boeve, MD

Arch Neurol. 2006;63:197-202

Table 2. Initial Clinical Diagnoses in 20 Patients With Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Initial Clinical Diagnosis	No. (%) of Patients
Viral encephalitis	5 (25)
Creutzfeldt-Jakob disease	3 (15)
Stroke or transient ischemic attack	3 (15)
Alzheimer disease	3 (15)
Migraine	2 (10)
Lewy body dementia	1 (5)
Psychosis	1 (5)
Metabolic encephalopathy	1 (5)
Delirium	1 (5)

Table 3. Serologic and Other Laboratory Abnormalities in 20 Patients With Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Serologic or Laboratory Test	No. of Patients*
Thyroperoxidase antibody positive	13/13
Thyroid microsomal antibody positive	7/7
Thyroglobulin antibody positive	6/10
Antinuclear antibody positive	6/20
Extractable nuclear antigen antibody positive	1/20
Rheumatoid factor positive	1/17
Gliadin antibody positive	1/11
Erythrocyte sedimentation rate elevated	5/19
C-reactive protein level elevated	3/9
Liver aminotransferase levels (AST/ALT) elevated	11/20

REVIEW
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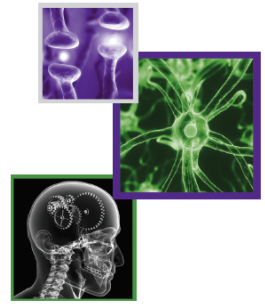
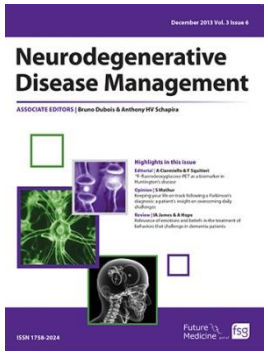
When dementia progresses quickly: a practical approach to the diagnosis and management of rapidly progressive dementia



Gregory S Day*[1] & David F Tang-Wai[1]

Neurodegen. Dis. Manage. (2014) 4(1), 41–56



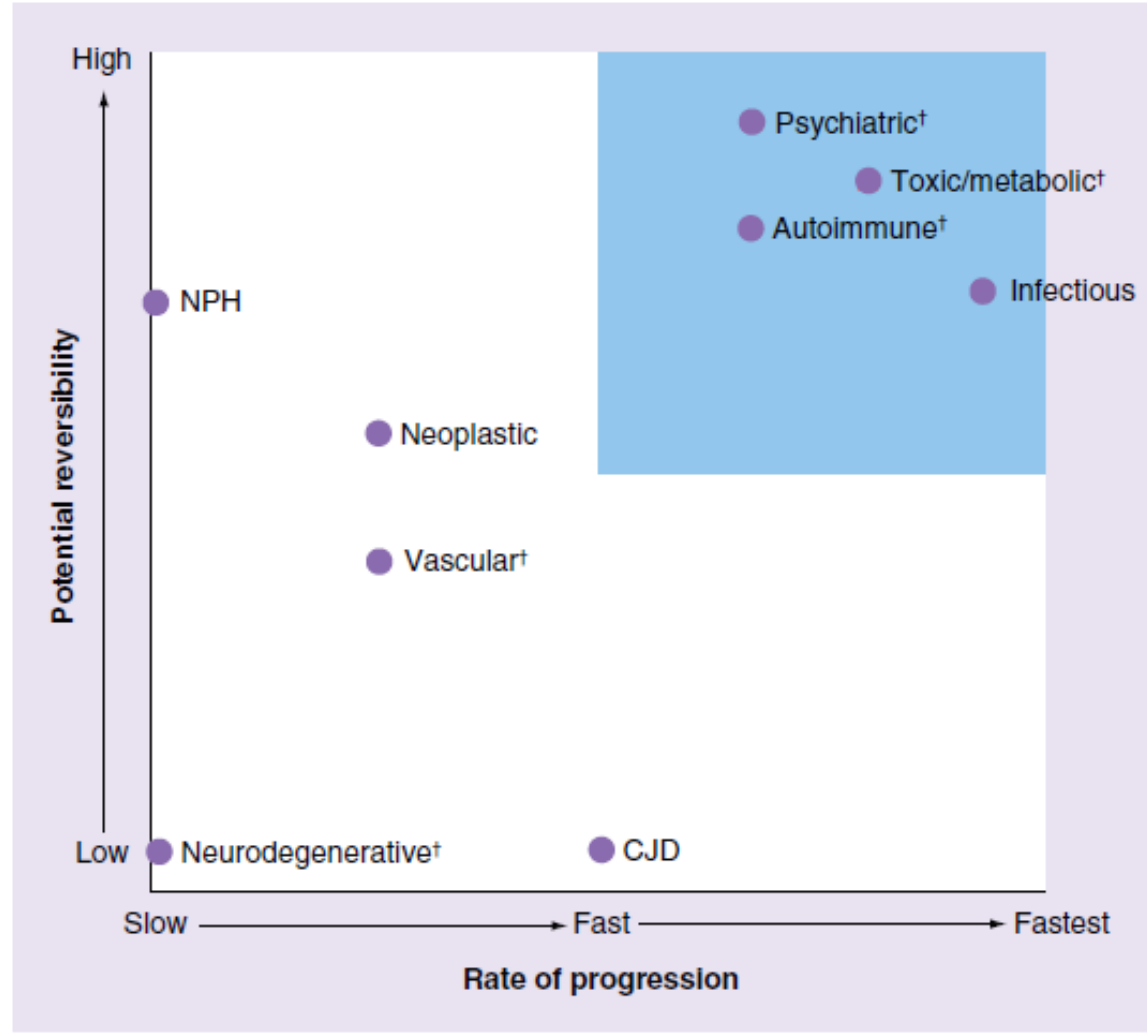
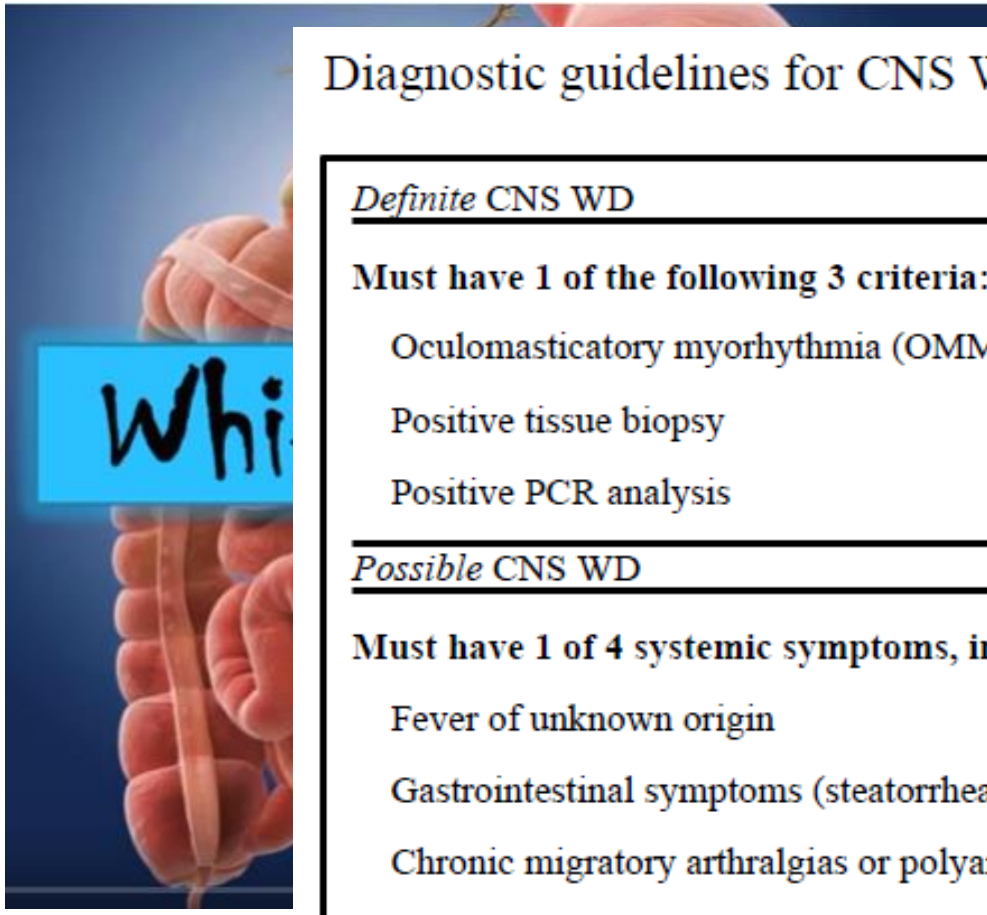


Figure 2. Causes of rapidly progressive dementia stratified by rate of progression and potential reversibility. Disease etiologies in the top right quadrant (shaded) are typically associated with the most rapidly progressive presentations and the greatest potential for response to treatment with appropriate treatments.

Gregory S Day & David F Tang-Wai, 2014



Diagnostic guidelines for CNS Whipple's disease*

Definite CNS WD

Must have 1 of the following 3 criteria:

- Oculomasticatory myorhythmia (OMM) or oculofacial skeletal myorhythmia (OFSM)
- Positive tissue biopsy
- Positive PCR analysis

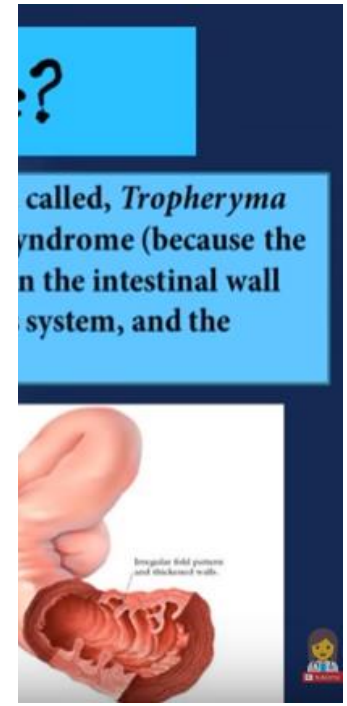
Possible CNS WD

Must have 1 of 4 systemic symptoms, including:

- Fever of unknown origin
- Gastrointestinal symptoms (steatorrhea, chronic diarrhea, abdominal distention, pain)
- Chronic migratory arthralgias or polyarthralgias
- Unexplained lymphadenopathy, night sweats, or malaise

Must also have 1 of 4 unexplained neurological signs, including:

- Supranuclear vertical gaze palsy
- Rhythmic myoclonus
- Dementia with psychiatric symptoms
- Hypothalamic manifestations



Clinical Infectious Diseases

PHOTO QUIZ

Anthony Amoroso, Section Editor

A 63-Year-Old Man With Rapidly Progressive Dementia

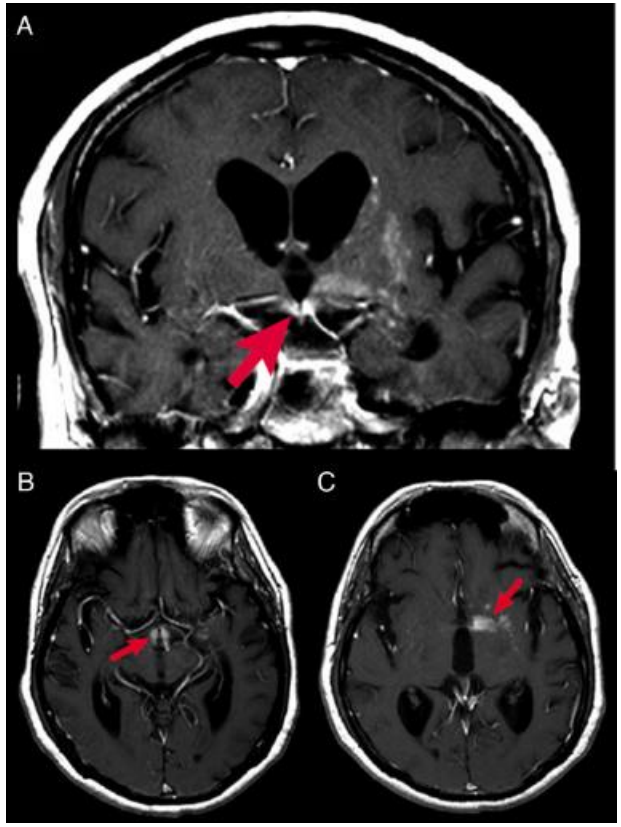


Figure 1. *A*, T1-weighted, coronal magnetic resonance imaging reveals contrast-enhancing lesions bilaterally in the ventral hypothalamus (red arrow). Additional contrast-enhancing lesions extend from the left hypothalamus into the basal forebrain, striatum, and temporal lobe. *B*, Axial views of this contrast-enhancing lesion in the bilateral hypothalamus (red arrow) extending superior-laterally (*C*) into the left basal forebrain and striatum (red arrow).

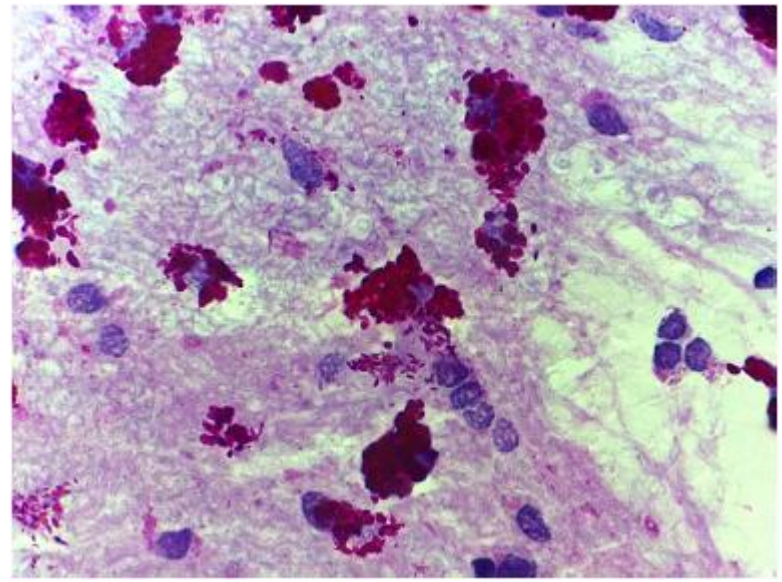


Figure 2. Histological examination of brain biopsy specimen (periodic acid-Schiff stain).

A 63-year-old man presented with 7 months of progressive cognitive decline, personality change, gait instability, and abdominal pain. Recently unemployed, he started to exhibit odd behaviors and became dependent on a wheelchair and personal care assistant. His history included diabetes, alcoholism, and tobacco use. He was originally from the Dominican Republic. There was no family history of dementia or movement disorders. On initial examination, he demonstrated abulia, with minimal speech and movement, and was unable to count backwards

REVIEW
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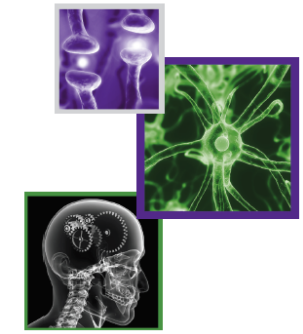
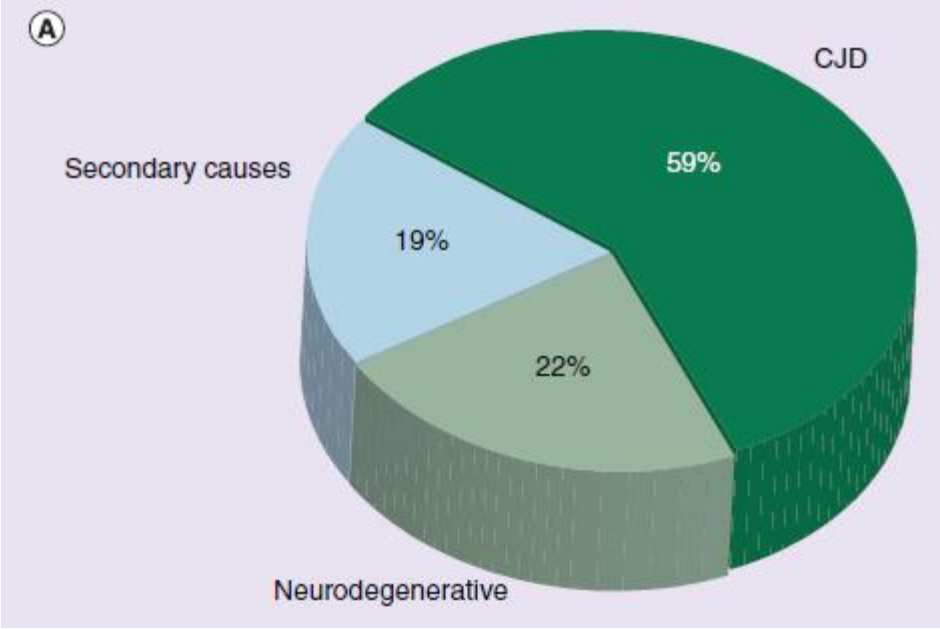
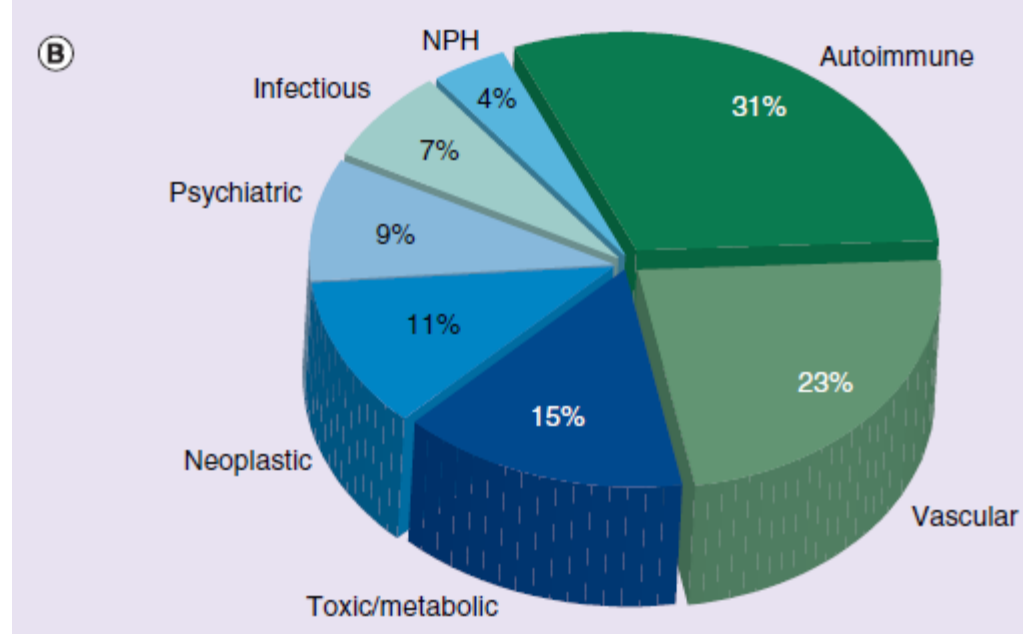
When dementia progresses quickly: a practical approach to the diagnosis and management of rapidly progressive dementia



Gregory S Day*[1] & David F Tang-Wai[1]

Neurodegen. Dis. Manage. (2014) 4(1), 41–56





REVIEW

Learning from history: Lord Brain and Hashimoto's encephalopathy

Grace F Crotty,[1,2] Colin Doherty,[3] Isaac H Solomon,[4] James D Berry,[2] Martin A Samuels[1]

Crotty GF, *et al*. *Pract Neurol* 2019;**19**:316–320. doi:10.1136/practneurol-2018-002107

Key points

- Thyroid dysfunction, both hypothyroidism and hyperthyroidism, can cause neurological disorders.
- Hashimoto's encephalopathy is a poorly defined entity, first described in 1966 by Lord Brain of Eynsham.
- Hashimoto's encephalopathy is a diagnosis of exclusion, requiring exclusion of infections, toxins, metabolic, neoplastic and other neuronal antibody syndromes.
- Giving a corticosteroid trial in unexplained encephalopathy is reasonable while awaiting antibody results or more definitive testing.

Lord Brain[1]

Box 1 Diagnostic criteria for Hashimoto's encephalopathy: diagnosis made when all six criteria present: (modified from Graus *et al*[18])

1. Encephalopathy with seizures, myoclonus, hallucinations or stroke-like episodes.
2. Thyroid disease (subclinical or mild overt).
3. MR scan of brain—normal or with nonspecific abnormalities.
4. Serum thyroid antibodies present (no specific disease—cut-off value).
5. Absence of other neuronal antibodies in serum or CSF.
6. Exclusion of alternative causes.

ORIGINAL CONTRIBUTION

ARCHIVES OF NEUROLOGY

Arch Neurol. 2006;63:197-202

Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Pablo Castillo, MD; Bryan Woodruff, MD; Richard Caselli, MD; Steven Vernino, MD, PhD; Claudia Lucchinetti, MD; Jerry Swanson, MD; John Noseworthy, MD; Allen Aksamit, MD; Jonathan Carter, MD; Joseph Sirven, MD; Gene Hunder, MD; Vahab Fatourechi, MD; Bahram Mokri, MD; Daniel Drubach, MD; Sean Pittock, MD; Vanda Lennon, MD, PhD; Brad Boeve, MD

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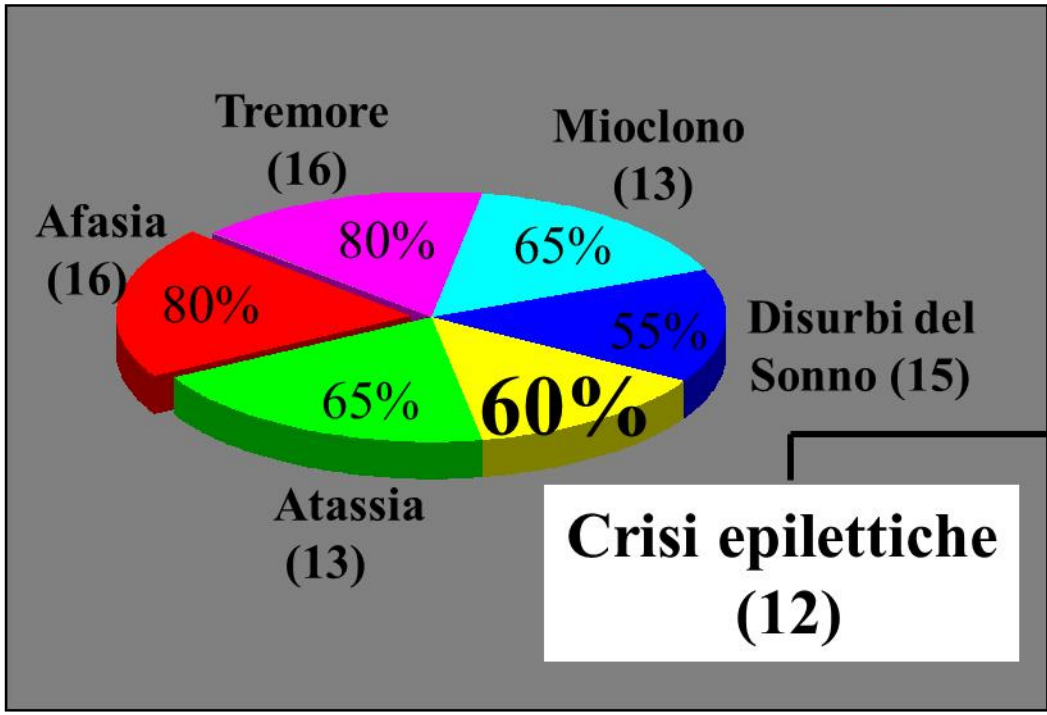


Table 1. Clinical Findings in 20 Patients With Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Abnormal Clinical Finding	No. (%) of Patients
Cognitive impairment and behavioral changes	20 (100)
Fluctuating symptoms	19 (95)
Transient aphasia	16 (80)
Tremor	16 (80)
Myoclonus	13 (65)
Ataxia or gait disorder	13 (65)
Seizures	12 (60)
Generalized	10 (50)
Partial	7 (35)
Both	5 (25)
Sleep disturbance	11 (55)
Hypersomnolence	8 (40)
Insomnia	3 (15)
Headache	10 (50)
Lateralized motor or sensory deficits	5 (25)
Psychosis or paranoia	5 (25)

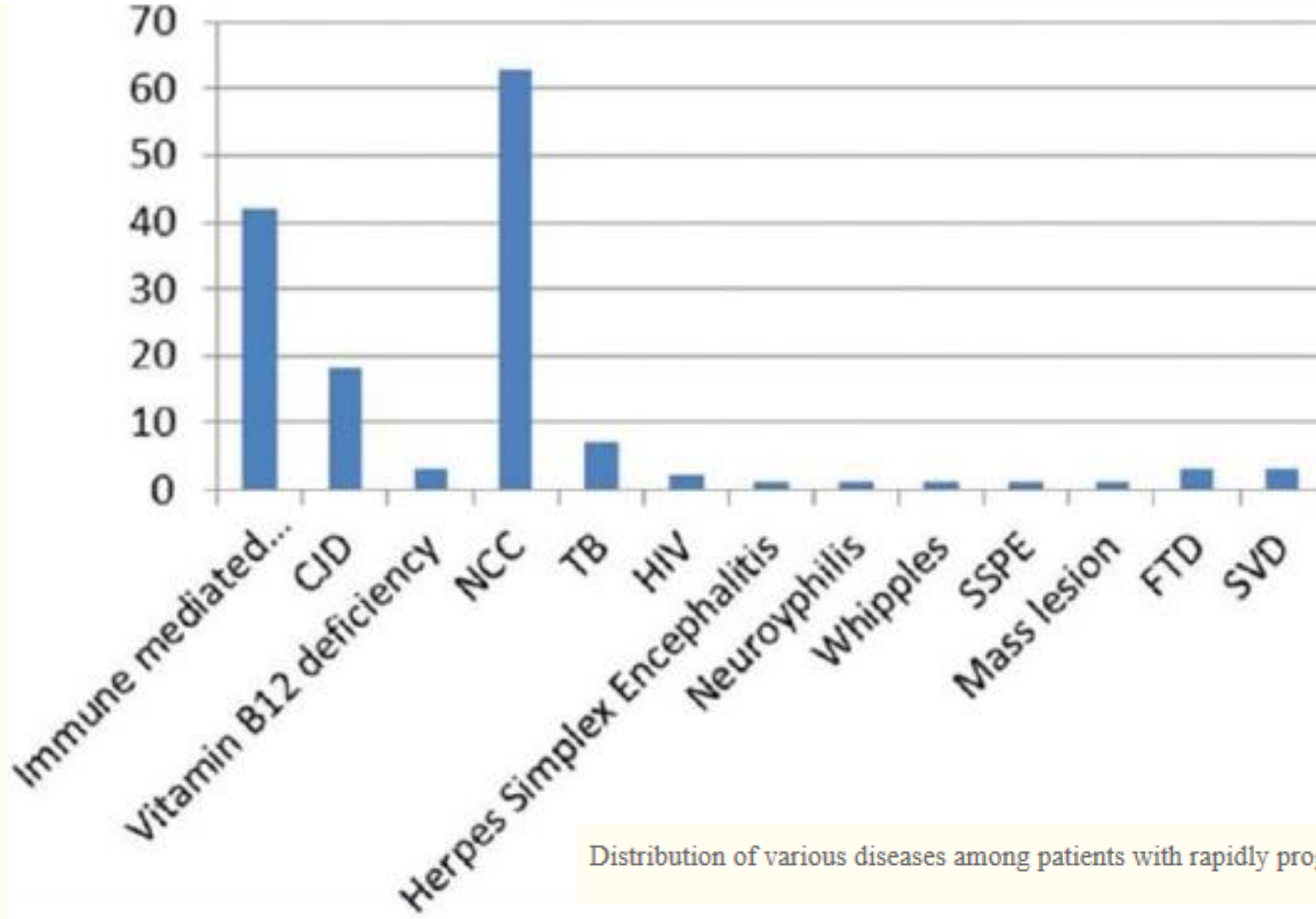
Journal of Neurosciences in Rural Practice

J Neurosci Rural Pract. 2017 Aug; 8(Suppl 1): S66–S71.
doi: 10.4103/jnrp.jnrp_100_17

PMCID: PMC5602264
PMID: 28936074

Syndromes of Rapidly Progressive Cognitive Decline-Our Experience

Sadanandavalli Retnaswami Chandra, Lakshminarayanapuram Gopal Viswanathan, Anupama Ramakanth Pai,[1] Rahul Wahatule, and Suvarna Alladi



Distribution of various diseases among patients with rapidly progressive dementia

VOLUME 35 · NUMBER 21 · JULY 20, 2017

JOURNAL OF CLINICAL ONCOLOGY

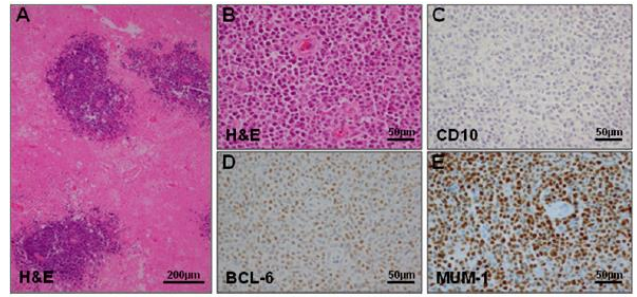
REVIEW ARTICLE

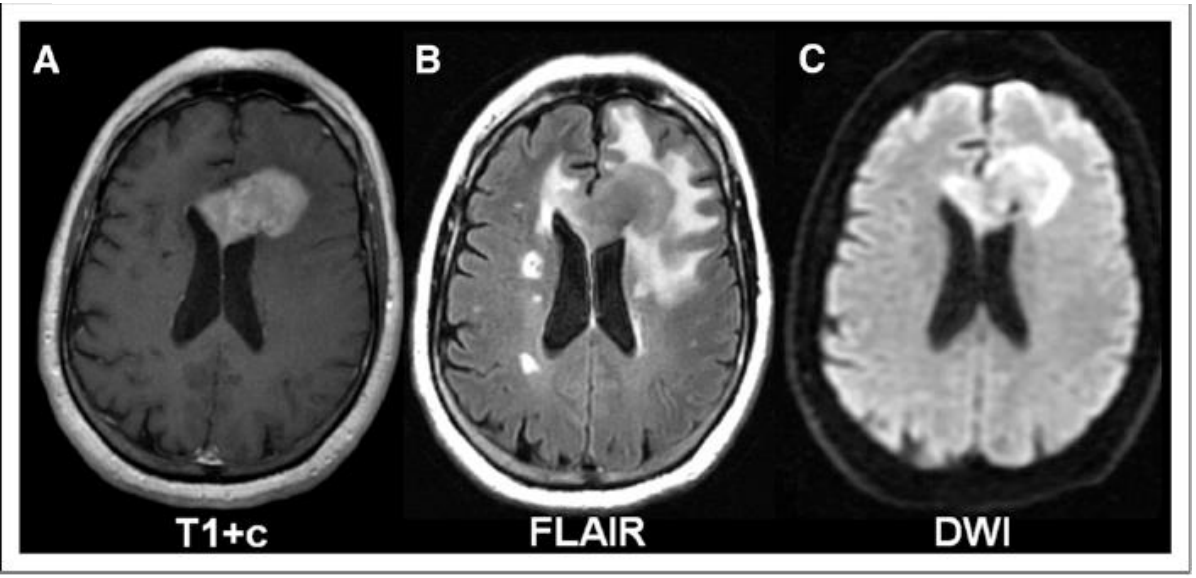
Primary CNS Lymphoma

Christian Grommes and Lisa M. DeAngelis

A B S T R A C T

Primary CNS lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin lymphoma that is typically confined to the brain, eyes, and cerebrospinal fluid without evidence of systemic spread. The prognosis of patients with PCNSL has improved during the last decades with the introduction of high-dose methotrexate. However, despite recent progress, results after treatment are durable in half of patients, and therapy can be associated with late neurotoxicity. PCNSL is an uncommon tumor, and only four randomized trials and one phase III trial have been completed so far, all in the first-line setting. To our knowledge, no randomized trial has been conducted for recurrent/refractory disease, leaving many questions unanswered about optimal first-line and salvage treatments. This review will give an overview of the presentation, evaluation, and treatment of immunocompetent patients with PCNSL.





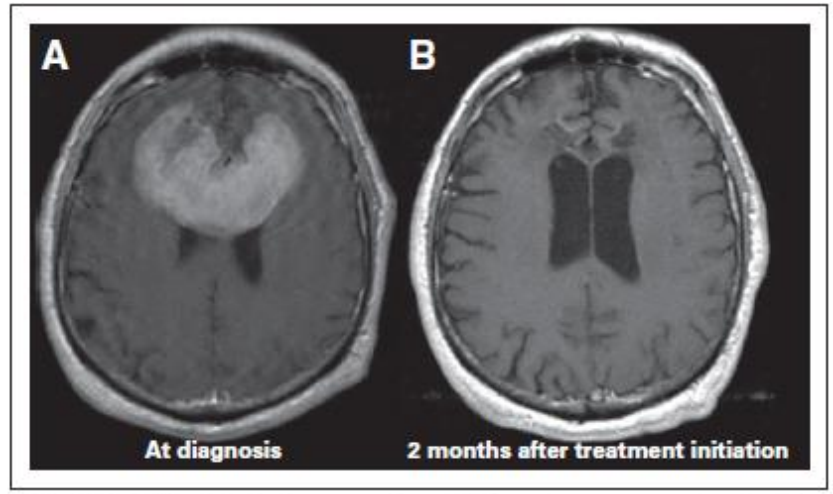
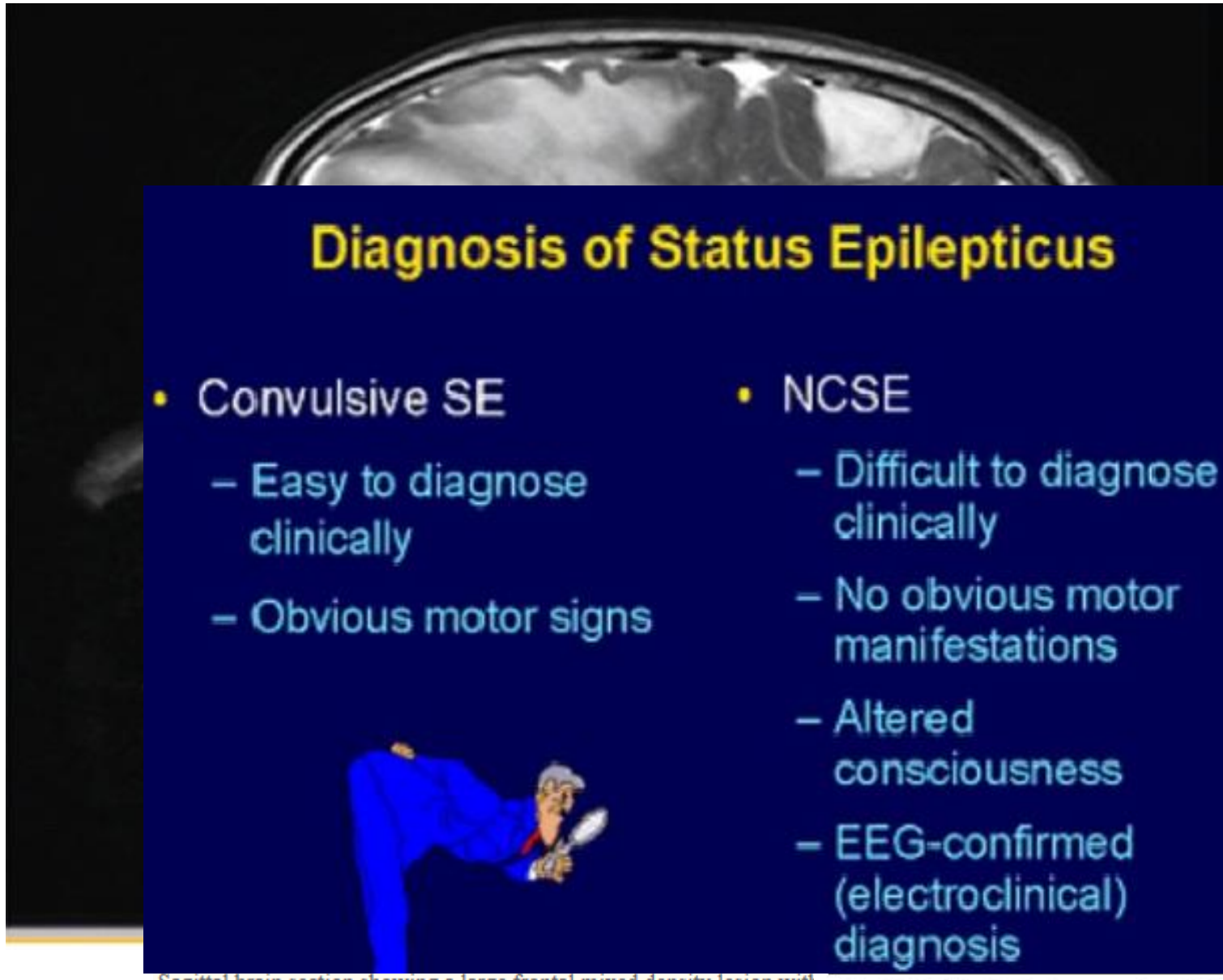


Fig 3. PCNSL is highly chemosensitive. (A) Magnetic resonance imaging (T1+ gadolinium) demonstrates a large, frontal-enhancing brain lesion. (B) Follow-up magnetic resonance imaging demonstrates resolution of the large lesion 2 months after treatment initiation with a high-dose methotrexate-based regimen.



Diagnosis of Status Epilepticus

- Convulsive SE
  - Easy to diagnose clinically
  - Obvious motor signs
- NCSE
  - Difficult to diagnose clinically
  - No obvious motor manifestations
  - Altered consciousness
  - EEG-confirmed (electroclinical) diagnosis

Sagittal brain section showing a large frontal mixed density lesion with
frontoparietal region

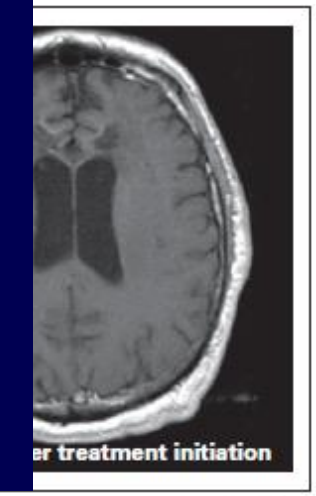
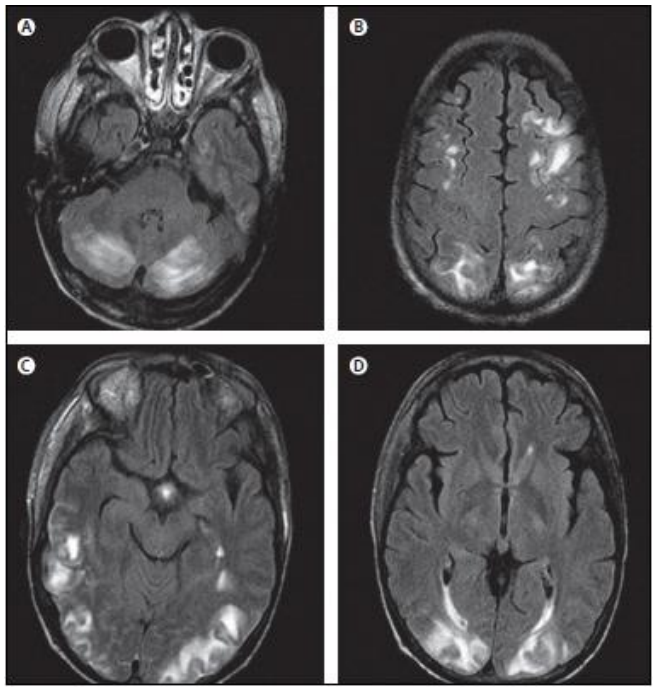


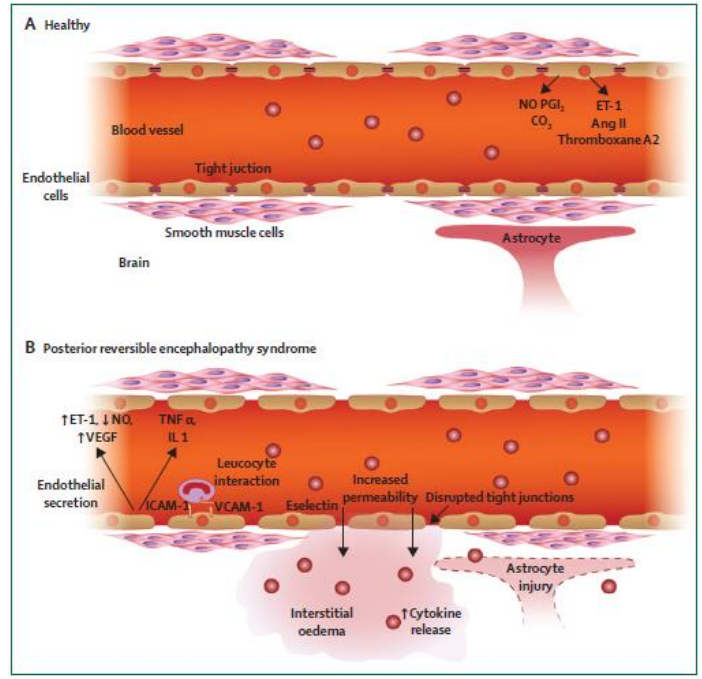
Fig 3. PCNSL is highly chemosensitive. (A) Magnetic resonance imaging (T1+ gadolinium) demonstrates a large, frontal-enhancing brain lesion. (B) Follow-up magnetic resonance imaging demonstrates resolution of the large lesion 2 months after treatment initiation with a high-dose methotrexate-based regimen

Lancet Neurol 2015; 14: 914–25

Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions

Jennifer E Fugate, Alejandro A Rabinstein





Panel 2: **Differential diagnoses of posterior reversible encephalopathy syndrome (PRES)**

Infectious encephalitis
- CSF pleocytosis
- Positive CSF Gram stain or culture
- Positive CSF microbial serology or PCR
- Fever
- Peripheral leucocytosis
- Can be unilateral in brain imaging

Autoimmune or paraneoplastic encephalitis
- History of malignancy or tumour
- Antigen-specific antibody in serum or CSF
- Can be unilateral in brain imaging

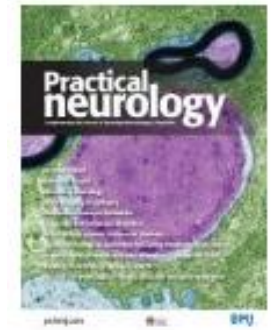
Malignancy or tumour (lymphoma, gliomatosis cerebri, metastatic disease)
- Subacute-to-chronic clinical presentation
- History of malignant tumour
- History of unintentional weight loss
- Abnormal CSF cytology
- Absence of clinical and radiological resolution
- Can be unilateral in brain imaging

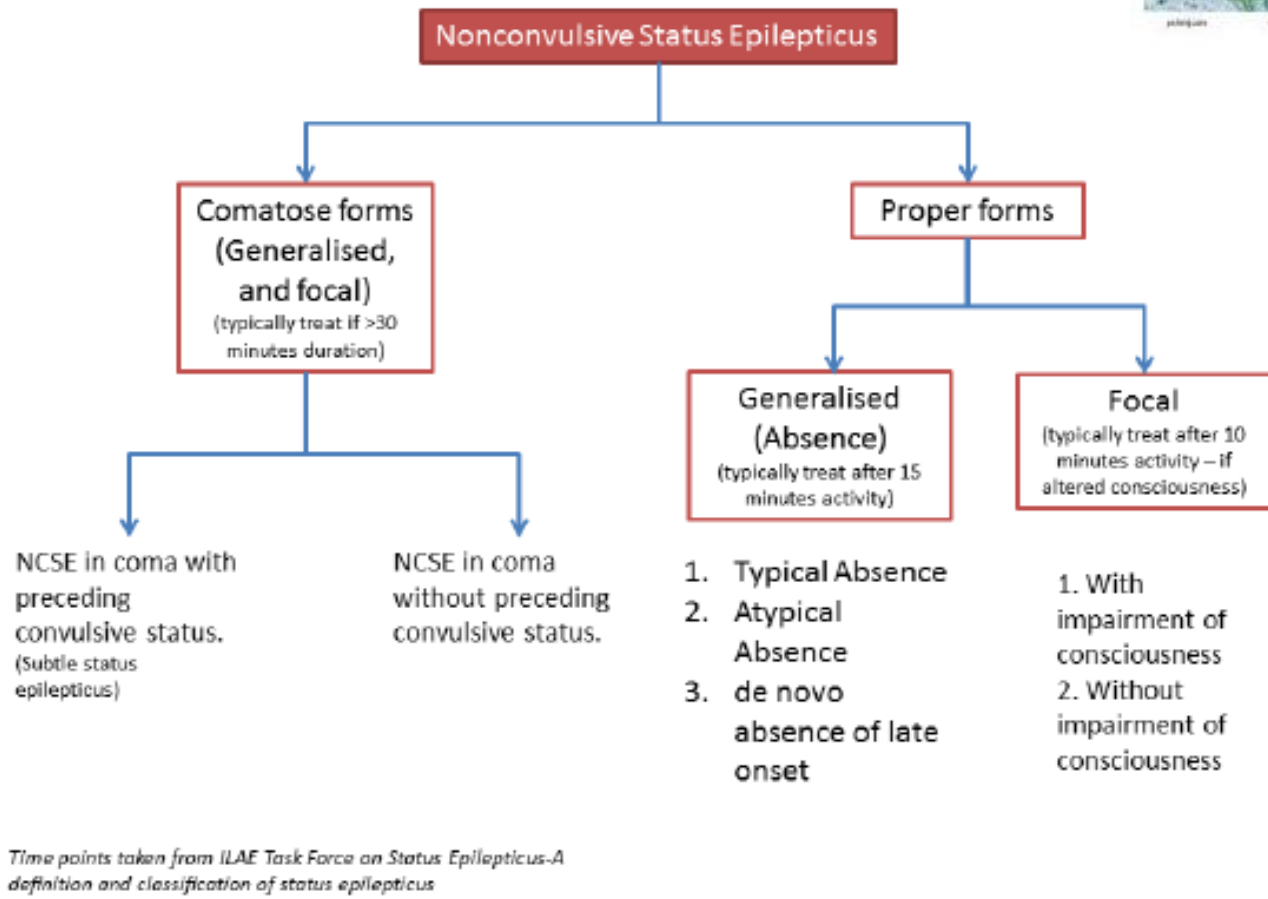
Subcortical leucoaraiosis
- No acute clinical presentation
- Radiographical signal abnormality is particularly confluent and periventricular

CNS vasculitis
- Often subacute clinical presentation
- CSF pleocytosis
- Cytotoxic oedema in non-PRES-like pattern

Non-convulsive status epilepticus: mimics and chameleons

Michael Owen Kinney,[1] John J Craig,[1] P W Kaplan[2]

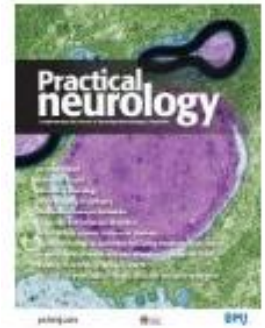




Kinney MO et al, 2018

Figure 1 Classification of non-convulsive status epilepticus (NCSE); Based on the International League Against Epilepsy (ILAE) Task Force classification[1].

Box 3 Working diagnostic criteria for EEG diagnosis of NCSE[18]



Patients without epileptic encephalopathy (either criteria 1 or 2)

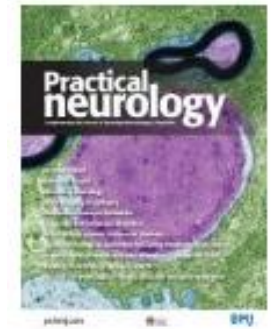
1. Repetitive epileptiform discharges at >2.5 Hz (focal or generalised spikes, polyspike, sharp waves, spike-and-wave or sharp-and-slow wave complexes).
2. Epileptiform discharges ≤2.5 Hz or rhythmic delta/theta activity (>0.5 Hz) including one of the additional criteria: 2a, 2b or 2c.
   a. Clinical improvement after intravenous benzodiazepines, with improved EEG reactivity, and appearance of EEG background. If the EEG improves without clinical improvement, then this is considered possible NCSE.
   b. Focal ictal symptoms during the observed EEG pattern (eg, facial twitching, nystagmus, myoclonus).
   c. Spatiotemporal evolution: incrementing onset (increase in voltage, with increase or decrease in frequency), pattern evolution (increase or decrease in frequency >1 Hz or location), decrementing termination (voltage or frequency) or post-periodic epileptiform discharges with background slowing or attenuation.

Patients with epileptic encephalopathy

1. Frequent or continuous generalised spike–wave discharges that increase in profusion or frequency when compared with baseline EEG with observed change in clinical state.
2. Improved clinical or EEG features with intravenous benzodiazepines. If the EEG improves without clinical improvement, this is again best considered as possible NCSE.

Kinney MO et al, 2018

Box 4 Trial of benzodiazepine or other antiepileptic medications



- Use an intravenous benzodiazepine trial under appropriate ventilator precautions, when the diagnosis
- If the EEG improves without clinical change, this may reflect possible NCSE, but note that benzodiazepines can suppress many interictal EEG patterns.
- If benzodiazepines are contraindicated (hypotension or concern about reducing consciousness), consider an alternative non-sedating antiepileptic drug (levetiracetam, lacosamide, valproate, phenytoin).
- The effects from an intravenous drug can be delayed.

lorazepam, or until there is a secure diagnosis of NCSE with clinical and EEG improvement.

Kinney MO et al, 2018

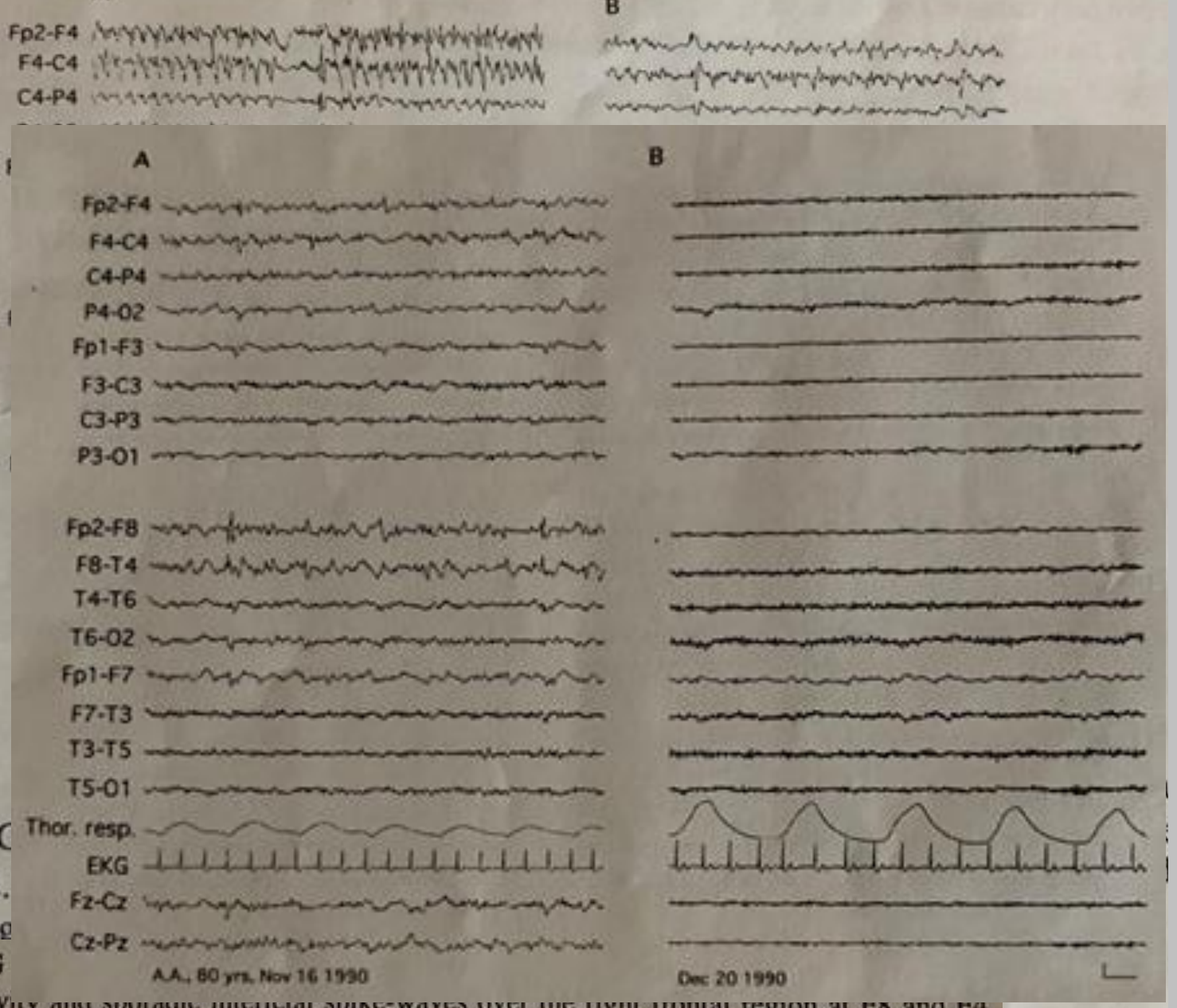


Figure 1: EEG
rhythmic (2-2.
activity lasting
di

and
fast
ictal

Figure 2: EEG
background activity and sporadic interictal spike-waves over the right frontal region at F8 and F4 phase reversal. B) 25 days after beginning replacement therapy. Normal posterior background and lack of any abnormal activity over the right frontal region. Calibration: 1 sec, 50 μV.

Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management

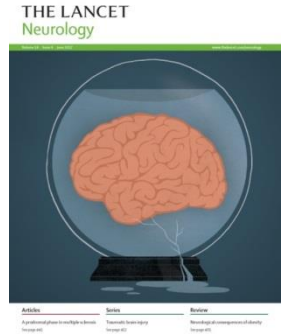
Review

GianPietro Sechi, Alessandro Serra

Lancet Neurol 2007; 6: 442–55

Institute of Clinical Neurology, University of Sassari, Italy
(G Sechi MD, A Serra MD)

Correspondence to:
Prof GianPietro Sechi, Institute of Clinical Neurology, University of Sassari, Viale S. Pietro 10, 07100, Sassari, Italy
gpsechi@uniss.it



Panel 1: Clinical features of Wernicke's encephalopathy

Common symptoms or signs at presentation
Ocular abnormalities
Mental status changes
Incoordination of gait and trunk ataxia

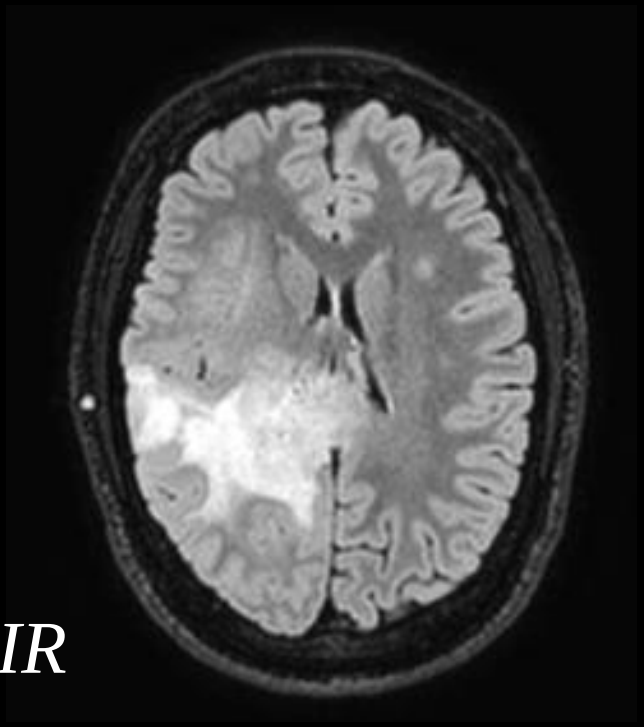
Uncommon symptoms or signs at presentation
Stupor
Hypotension and tachycardia
Hypothermia
Bilateral visual disturbances and papilloedema
Epileptic seizures
Hearing loss
Hallucinations and behavioural disturbances

Late-stage symptoms
Hyperthermia
Increased muscular tone and spastic paresis
Choreic dyskinesias
Coma

Etiology of RPDs

Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment
Toxic-metabolic							
Wernicke syndrome	A	Risk factors: alcoholism, malnutrition	Cognitive impairment, eye movement abnormalities, ataxia	T2 hyper in medial thalamus and mammillary bodies[e7]	Nondiagnostic	—	Thiamine
Extrapontine myelinolysis	A	Rapid correction of electrolyte disturbance (e.g., hyponatremia)	May take few days to develop symptoms; encephalopathy, movement disorders, para/quadriparesis	Hyper T2 lesions (CE) in pons, cerebellum, basal ganglia, thalamus; may take days to appear[e8]	Nondiagnostic	—	Symptomatic
Vitamin B12 deficiency	S	Older adults, pernicious anemia, veganism, fad diets	Cognitive impairment (infrequent, but treatable), sensory ataxia, paresthesias	Nondiagnostic	Nondiagnostic	↓Vitamin B12, ↑MMA, ↑homocysteine	Vitamin B12
Acquired hepatocerebral degeneration	S	Cirrhosis (portosystemic shunting)	Apathy, inattention, parkinsonism, cranial dyskinesia	Pallidal T1 hyper, T2 normal[e9]	Nondiagnostic	—	Treatment of liver disease, but might be irreversible; liver transplant
Acute intermittent porphyria[e10,e11]	A/S	20s-30s; F > M	Abdominal pain, autonomic dysfunction, behavioral changes, altered consciousness	Normal	Nondiagnostic	Elevated PBG/ALA in urine	Carbohydrates, intravenous haem arginate; avoid certain medications and metabolic disturbances

Ross W. Paterson et al, 2012



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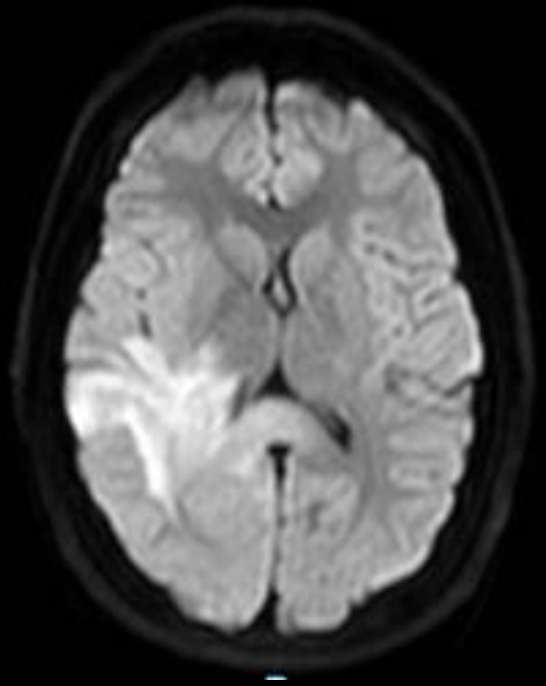
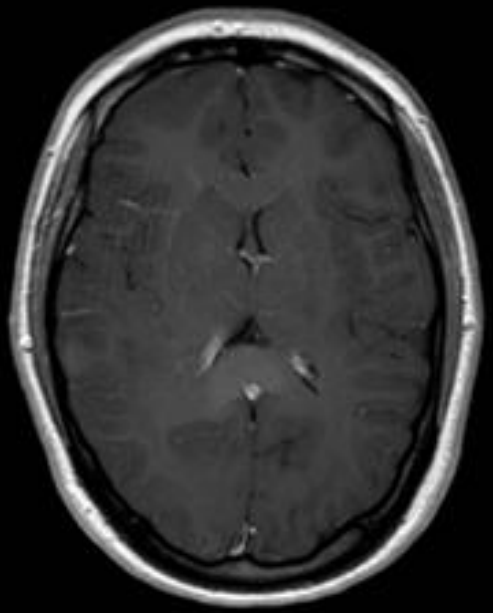
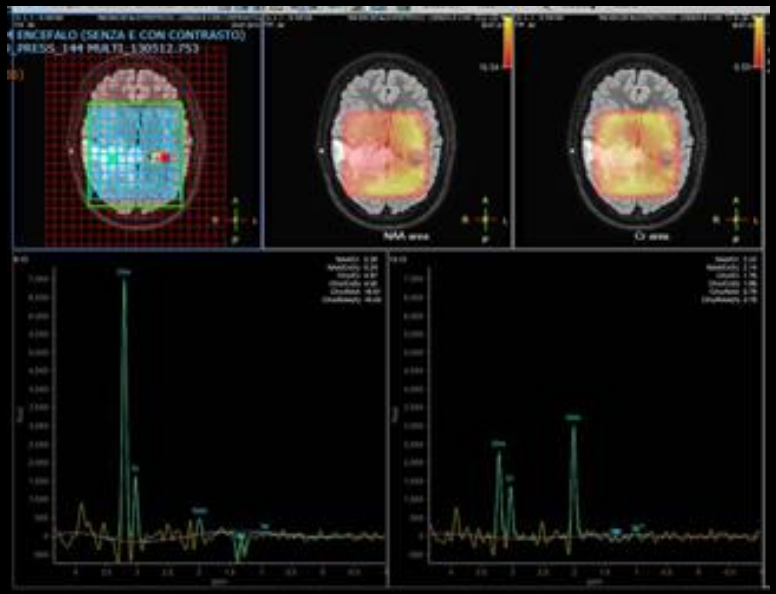


DWI



T1 mdc



REVIEW
EXPERT REVIEW OF NEUROTHERAPEUTICS
https://doi.org/10.1080/14737175.2018.1519397

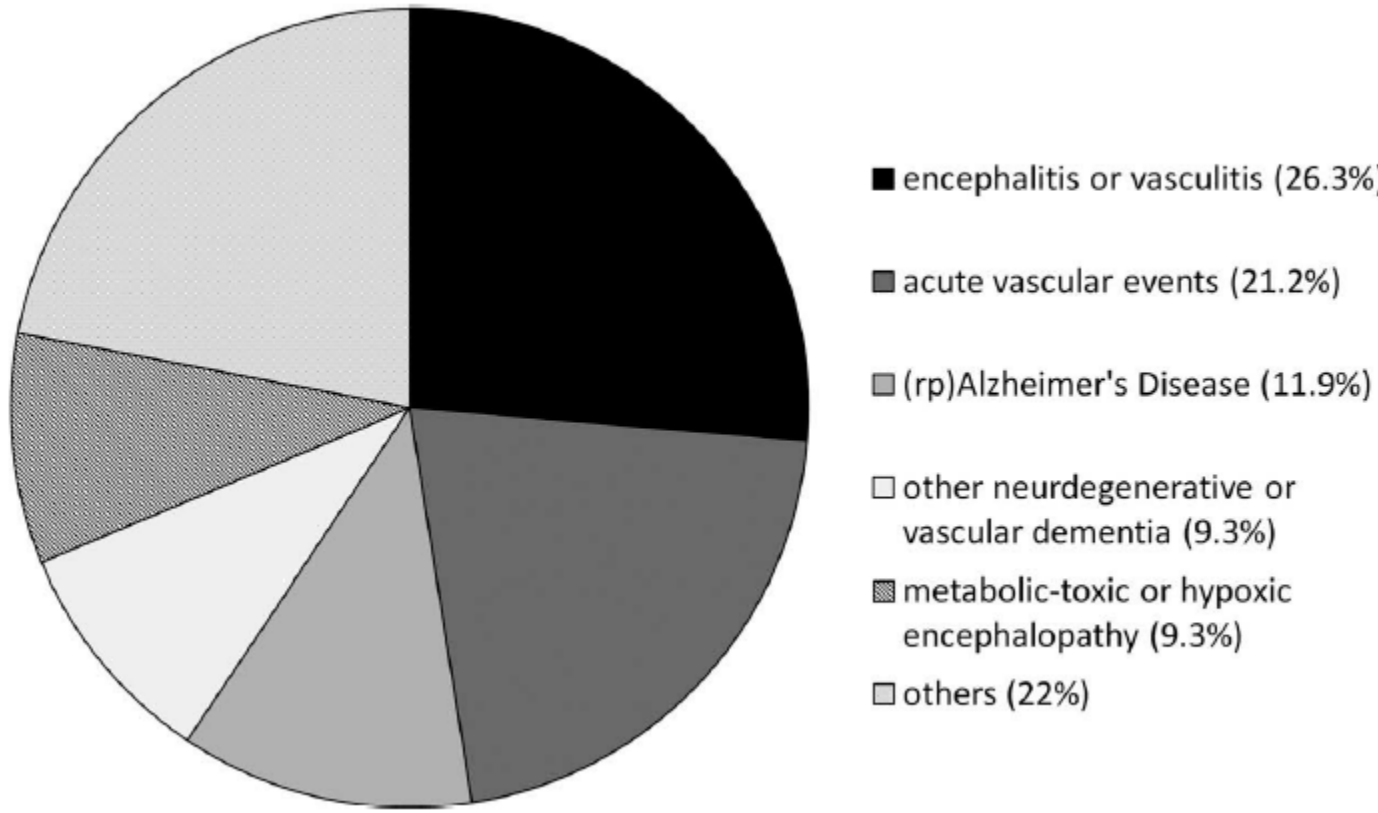
Diagnostic challenges in rapidly progressive dementia

Inga Zerr and Peter Hermann

Clinical Dementia Center and National TSE Reference Center, Department of Neurology, Goettingen University Medical Center, Goettingen, Germany

Table 1. Differential diagnoses of RPD reported by tertiary referral centers.

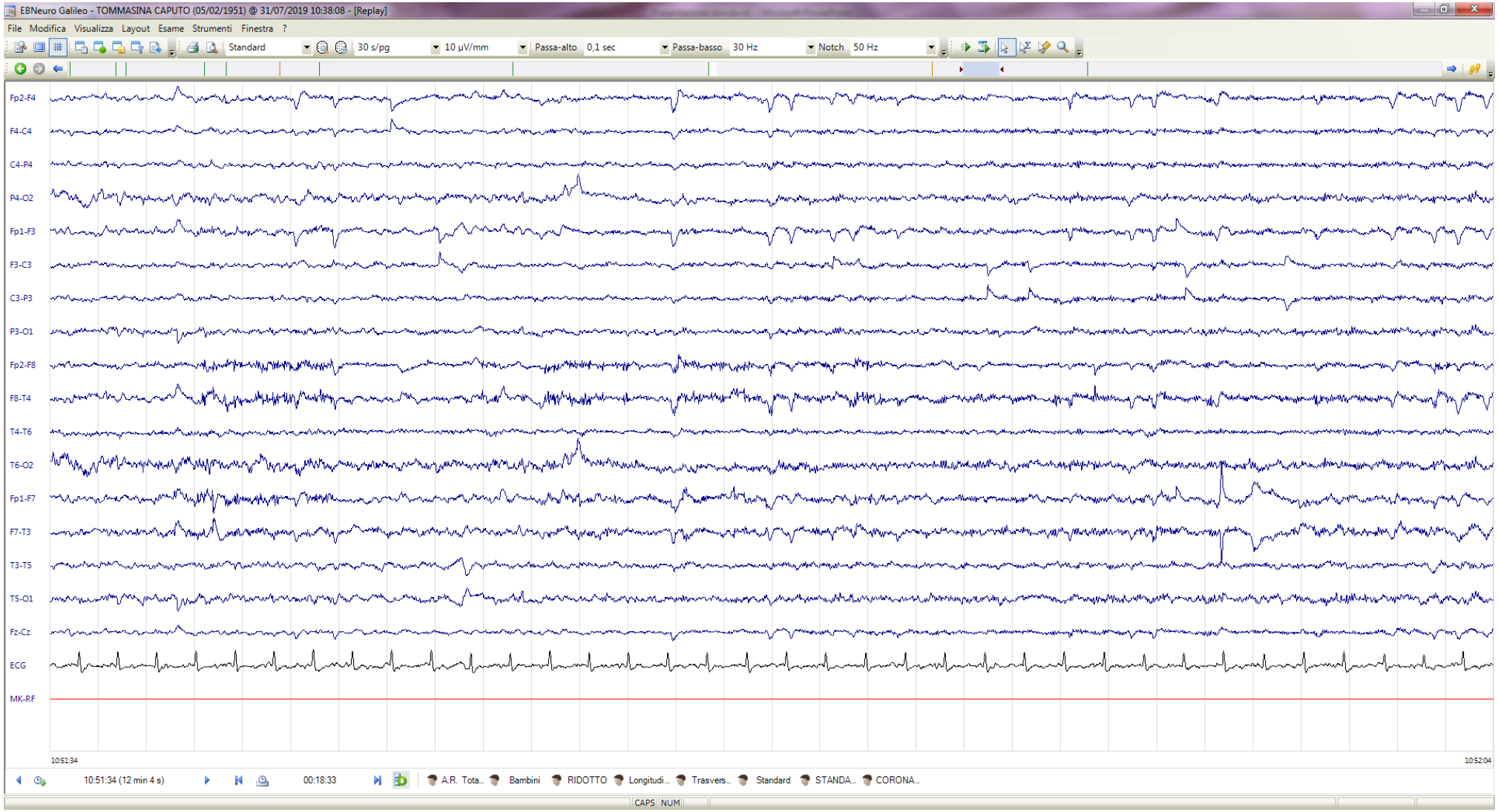
	Athens, Greece [61] (n = 68*)	Zhejiang, China [106] (n = 310**)	Sao Paulo, Brazil [14] (n = 61)	Chandigarh, India [15] (n = 187)
Infectious encephalitis (%)	5.9*	21.9	19.7	20.6
Immune-mediated disease (%)	8.8	9.0	45.9	18.2
Creutzfeldt–Jakob disease (%)	13.2	7.1	11.5	7.5
Neurodegenerative diseases (%)	47.0	24.8	8.2	14.4
Alzheimer's disease (%)	17.6	14.5	n.a.	n.a.
Others (%)	29.4	10.3	n.a.	n.a.
Vascular dementia (%)	13.2	**	n.a.	9.6
Toxic + metabolic (%)	*	10.3	n.a.	16.0
Others (%)	11.8	26.9	14.7	13.4


encephalitis or vasculitis (26.3%)
acute vascular events (21.2%)
(rp)Alzheimer's Disease (11.9%)
other neurdegenerative or vascular dementia (9.3%)
metabolic-toxic or hypoxic encephalopathy (9.3%)
others (22%)

Etiology of RPDs: use of the mnemonic VITAMINS

Ross W. Paterson et al, 2012

- **V** Vascular
- **I** Infectious
- **T** Toxic-metabolic
- **A** Autoimmune
- **M** Metastases/neoplam
- **I** Iatrogenic/inborn error of metabolism
- **N** Neurodegenerative
- **S** Systemic/seizures


EBNeuro Galileo - TOMMASINA CAPUTO (05/02/1951) @ 31/07/2019 10:38:08 - [Replay]
File Modifica Visualizza Layout Esame Strumenti Finestra ?
Standard
30 s/pg
10 µV/mm
Passa-alto 0,1 sec
Passa-basso 30 Hz
Notch 50 Hz
Fp2-F4
F4-C4
C4-P4
P4-O2
Fp1-F3
F3-C3
C3-P3
P3-O1
Fp2-F8
F8-T4
T4-T6
T6-O2
Fp1-F7
F7-T3
T3-T5
T5-O1
Fz-Cz
ECG
MK-RF
10:51:34
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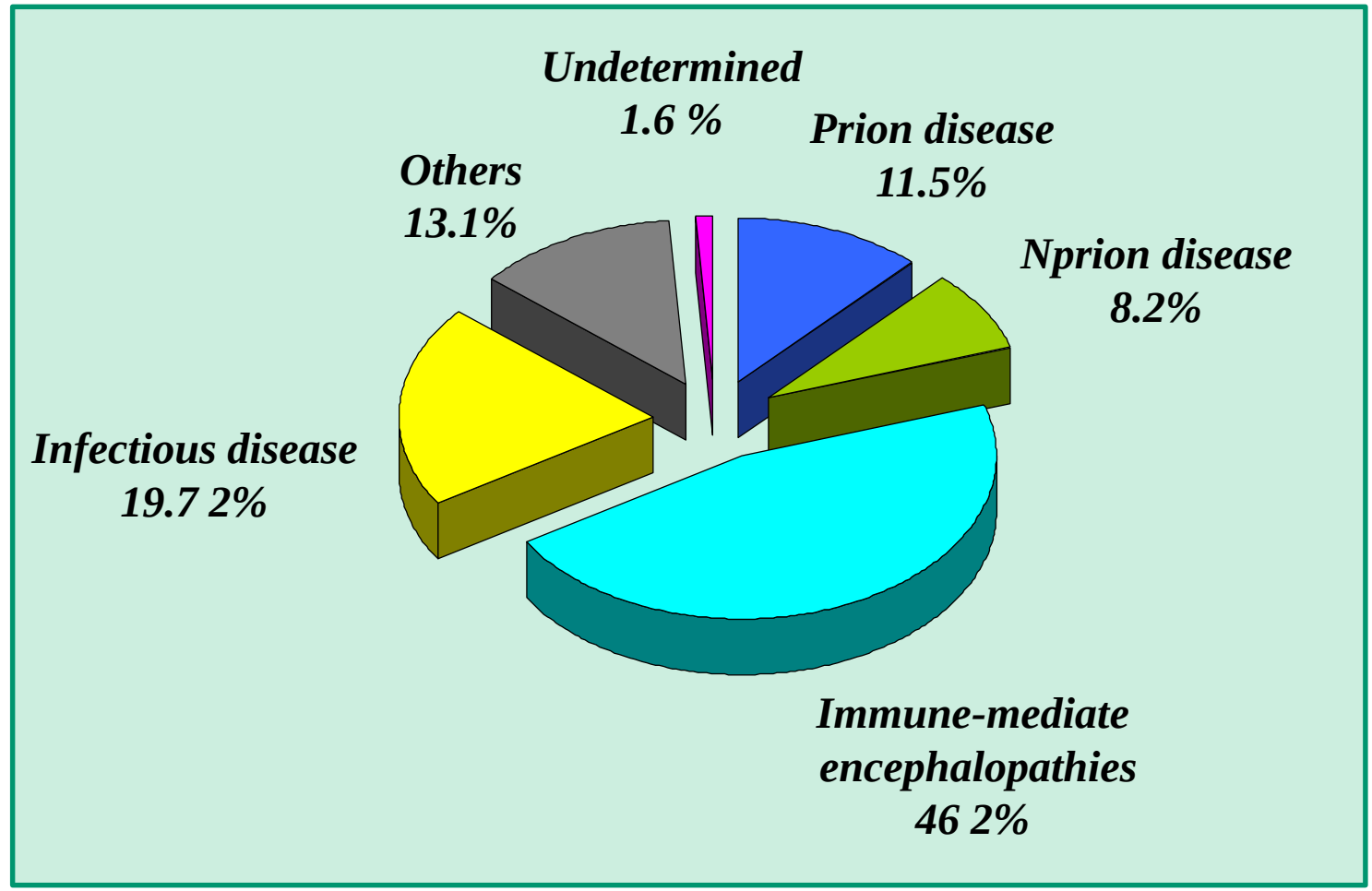

Undetermined
1.6 %
Prion disease
11.5%
Others
13.1%
Nprion disease
8.2%
Infectious disease
19.7 2%
Immune-mediate
encephalopathies
46 2%

Rapidly Progressive Dementia: Prevalence and Causes in a Neurologic Unit of a Tertiary Hospital in Brazil

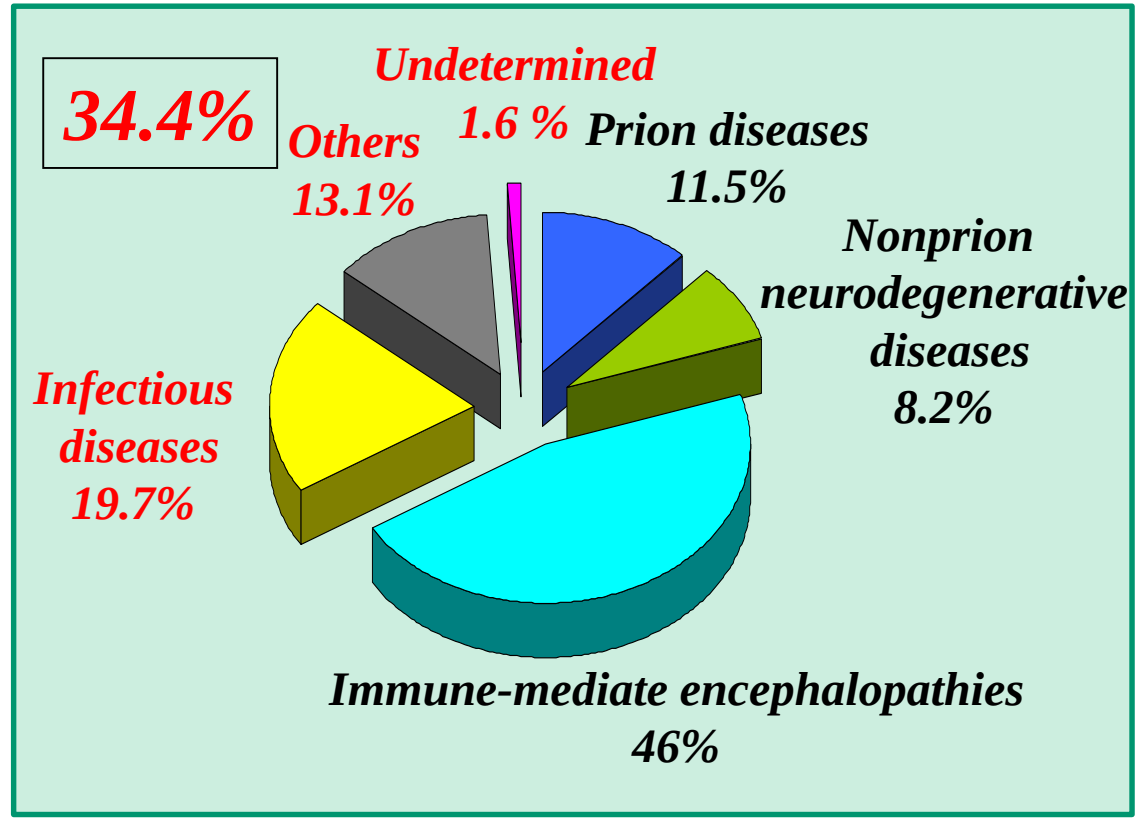
Adalberto Studart Neto, MD, Herval R. Soares Neto, MD, Mateus M. Simabukuro, MD, Davi J.F. Solla, MD, Márcia R.R. Gonçalves, MD, Ida Fortini, MD, Luiz H.M. Castro, MD, and Ricardo Nitrini, MD, PhD

(*Alzheimer Dis Assoc Disord* 2017;31:239–243)

Results: We identified 61 RPD (3.7%) cases among 1648 inpatients. Mean RPD patients' age was 48 years, and median time to progression was 6.4 months. Immune-mediated diseases represented the most commonly observed disease group in this series (45.9% of cases). Creutzfeldt-Jakob disease (11.5%) and nonprion neurodegenerative diseases (8.2%) were less common in this series. Outcome was favorable in 36/61 (59.0%) RPD cases and in 28/31 (89.3%) of immune-mediated cases. Favorable outcome was associated with shorter time from symptom onset to diagnosis and abnormal cerebrospinal fluid findings.

TABLE 2. Frequency of Clinical Diagnoses in Patients with RPD

Clinical Diagnoses	N (%)
Prion diseases	11.5
Probable CJD	4
Definite CJD	3
Nonprion neurodegenerative diseases	8.2
Probable dementia with Lewy bodies	1
Probable frontotemporal dementia	2
Wilson's disease	2
Immune-mediated encephalopathies	29.5
Anti-NMDA	10
Paraneoplastic encephalopathies	2
Unidentified neuronal surface antibodies	1
Unknown etiology	5
Other immune-mediated diseases	16.4
Behçet's disease	1
Probable sarcoidosis	3
CNS vasculitis	3
Hashimoto's encephalopathy	2
Encephalopathy associated with relapsing polychondritis	1
Infectious diseases	19.7
Herpes viral encephalitis	6
Infectious meningoencephalitis (unknown etiology)	3
Cryptococcosis	2
Coxsackie virus (common variable immunodeficiency)	1
Others*	8 (13.1)
Undetermined	1 (1.6)



Frequency of Clinical Diagnoses in Patients with RPD

Studart Neto A et al, 2017

Neurology® Clinical Practice

Diagnosis and treatment of rapidly progressive dementias

Ross W. Paterson, MRCP*
Leonel T. Takada, MD*
Michael D. Geschwind, MD, PhD

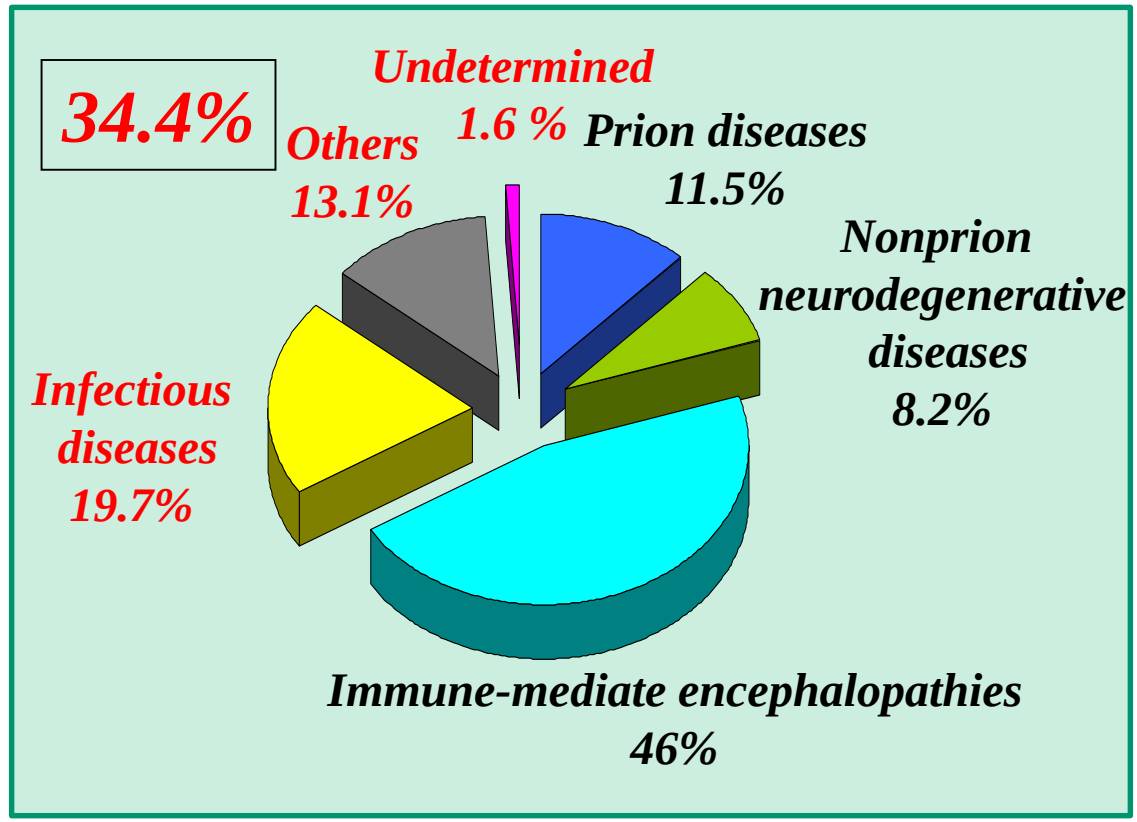
Neurology: Clinical Practice ▮ September 2012

Although no formal definition exists for what constitutes a rapidly progressive dementia (RPD), generally we use the term when dementia occurs in less than 1–2 years from illness onset, but more commonly over weeks to months.[1] Because these conditions are relatively uncommon, the appropriate diagnostic workup and treatments often are unfamiliar to many neurologists. Accurate, thorough, and prompt diagnosis is important as many RPDs are treatable, and even curable. In this article, we present a practical, systematic approach for RPD diagnosis as well as treatment algorithms for the management of immunotherapy-responsive and other dementias.

malattie responsabili di decadimento cognitivo

- Malattia di Alzheimer
- Demenza fronto-temporale
- Malattia di Parkinson
- demenza a corpi di Levy
- Malattia cerebro-vascolare
- Encefalopatie spongiformi
- Malattie infiammatorie

61 RPD cases



Frequency of Clinical Diagnoses in Patients with RPD

Studart Neto A et al, 2017

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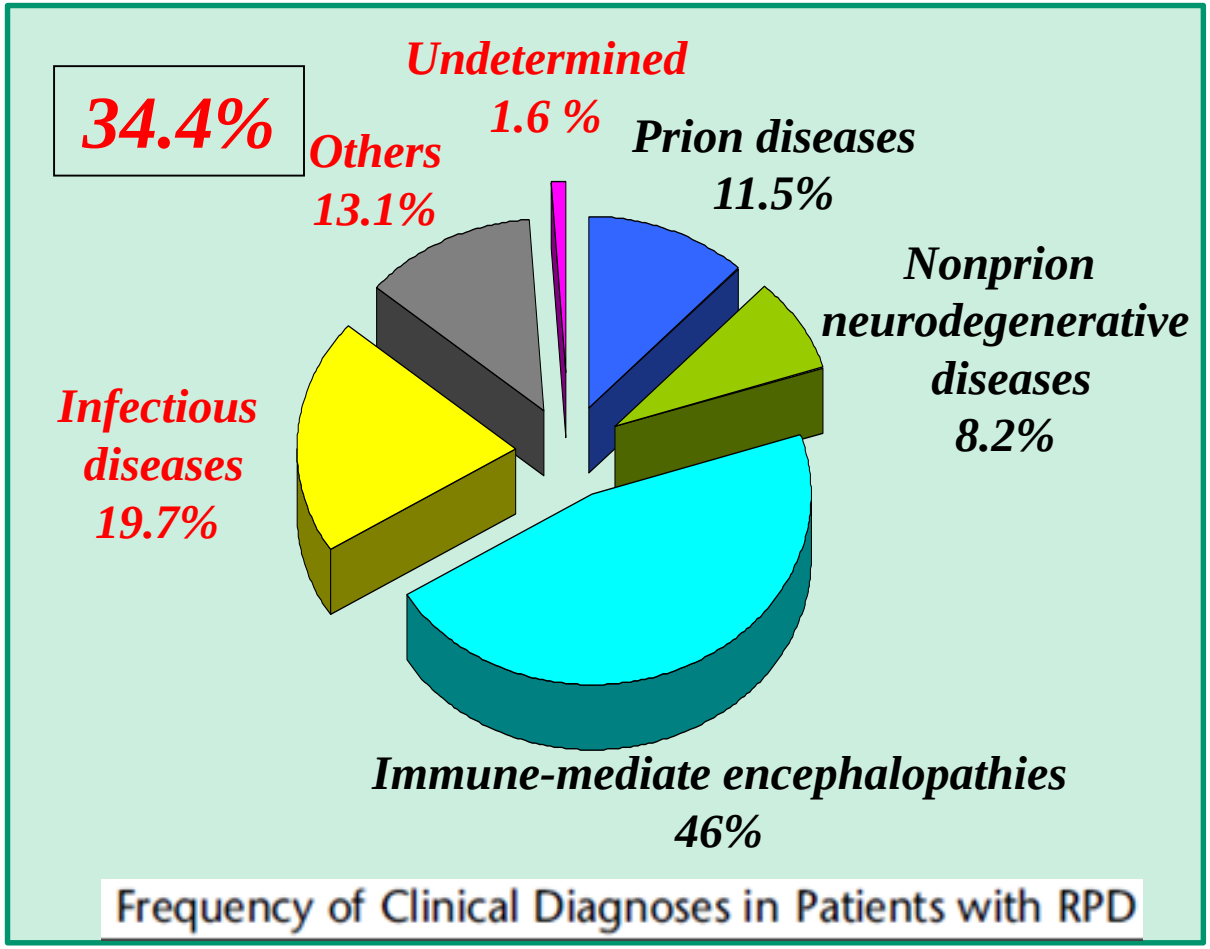

Studart Neto A et al, 2017

Nonconvulsive status epilepticus in adults — insights into the invisible

Raoul Sutter[1,2], Saskia Semmlack[1] and Peter W. Kaplan[3]

NATURE REVIEWS | NEUROLOGY VOLUME 12 | MAY 2016 | 281

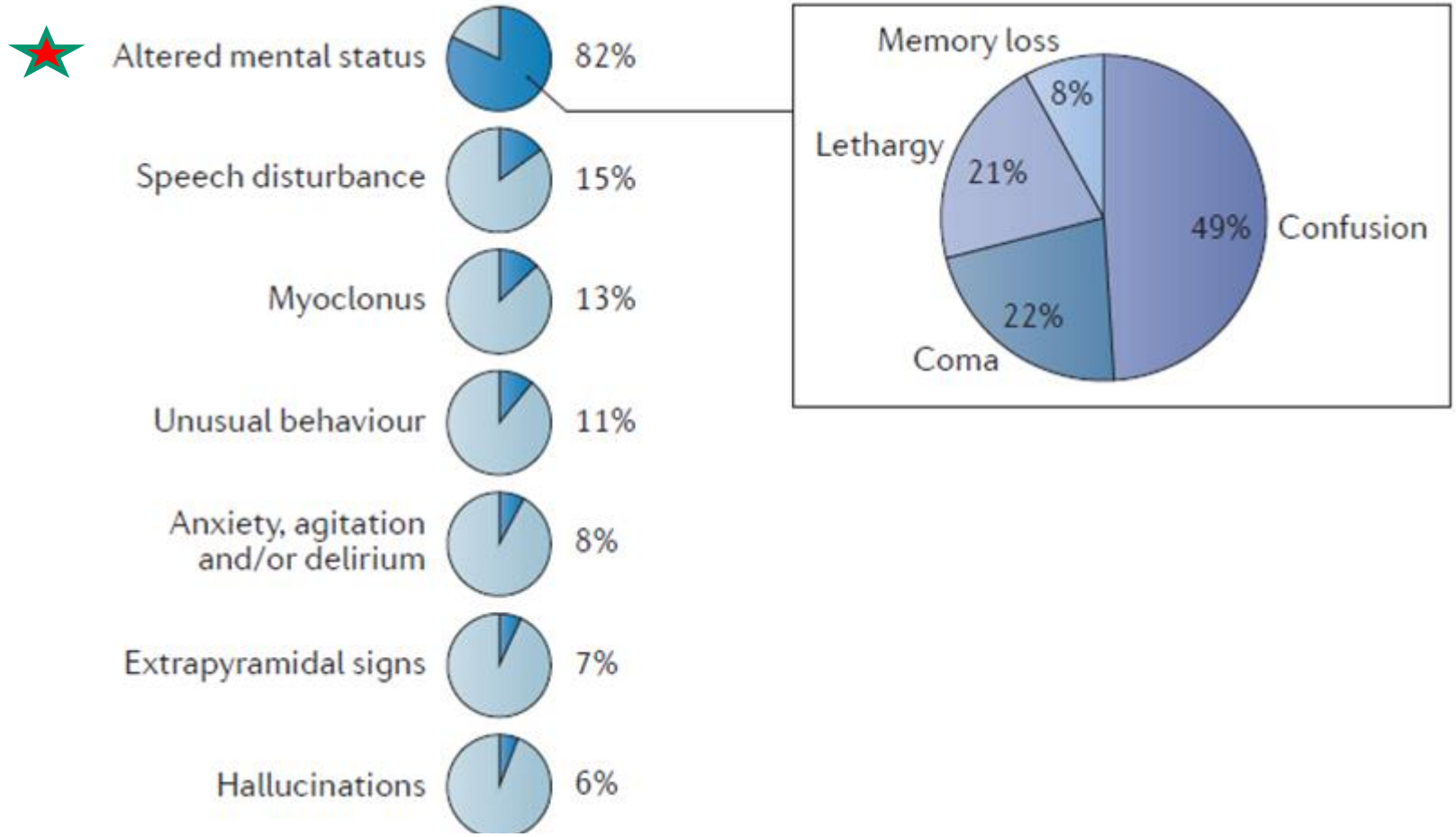

Figure 1 | **Major symptoms of nonconvulsive status epilepticus.**

SHORT REPORT

Non-convulsive status epilepticus: a profile of patients diagnosed within a tertiary referral centre

S Haffey, A McKernan, K Pang

J Neurol Neurosurg Psychiatry 2004;**75**:1043–1044. doi: 10.1136/jnnp.2003.019612

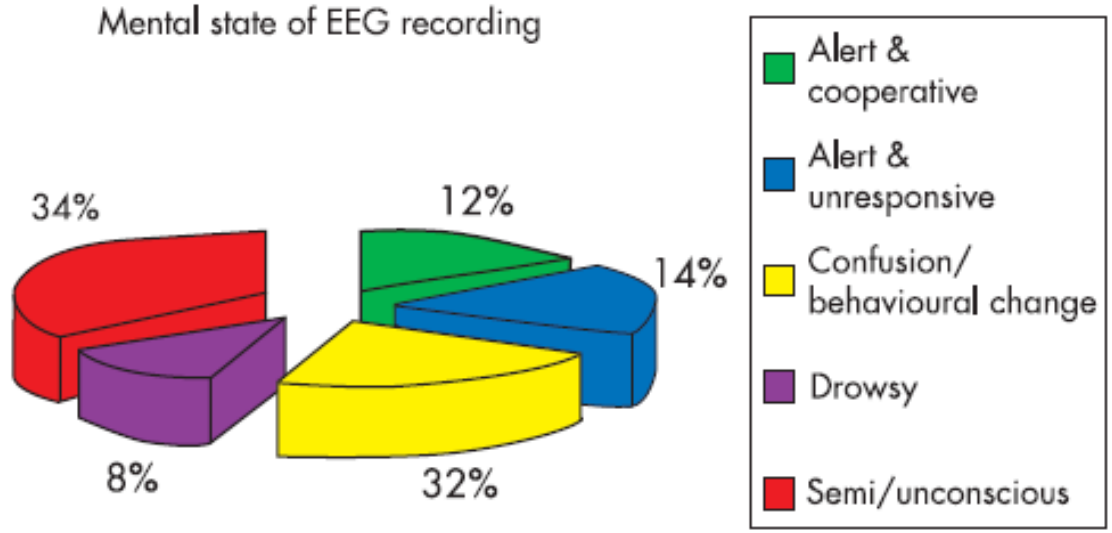

Figure 1 Mental state categories of the 45 patients at the time of electroencephalogram recording.

Viral encephalitis – clinical symptoms

- **Typical presentation**
  - Acute flu-like prodrome
  - High fever, severe headache
  - Altered consciousness (lethargy, drowsiness, confusion, coma)
  - Seizures
  - Focal neurological signs
- **More subtle presentations**
  - Low grade fever
  - Speech disturbances (dysphasia, aphasia)
  - Behavioural changes
  - Subacute and chronic presentations can be
    VZV, HSV (immuno-compromised)

Table 2. Initial Clinical Diagnoses in 20 Patients With Steroid-Responsive Encephalopathy Associated With Autoimmune Thyroiditis

Initial Clinical Diagnosis	No. (%) of Patients
Viral encephalitis	5 (25)
Creutzfeldt-Jakob disease	3 (15)
Stroke or transient ischemic attack	3 (15)
Alzheimer disease	3 (15)
Migraine	2 (10)
Lewy body dementia	1 (5)
Psychosis	1 (5)
Metabolic encephalopathy	1 (5)
Delirium	1 (5)

R.A. , F 68 a

Luglio 2018 → Accede al DEA del P.O. Misericordia di Grosseto per crisi epilettiche subentranti→ S.E. convulsivo

Anamnesi patologica recente:

- da alcune settimane disturbi mnesici e comportamentali e del sonno
- crisi focali motorie (pregresso accesso al DEA di altro Presidio)

IOT
ricovero presso
Terapia Intensiva

Anamnesi patologica remota: negativa per epilessia.

Ipertensione arteriosa, IGT, OSAS.

RICOVERO in NEUROLOGIA

EON: vigile, accessibile al colloquio, disorientata nei parametri spaziali, orientata nel tempo. Deficit di memoria di rievocazione a breve termine. Restante obiettività nei limiti.

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	Athens, Greece [61] (*n* = 68*)	Zhejiang, China [106] (*n* = 310**)	Sao Paulo, Brazil [14] (*n* = 61)	Chandigarh, India [15] (*n* = 187)
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Diagnostic challenges in rapidly progressive dementia

I. ZERR AND P. HERMANN

EXPERT REVIEW OF NEUROTHERAPEUTICS
https://doi.org/10.1080/14737175.2018.1519397

January 18, 2018

PLOS ONE

RESEARCH ARTICLE

Rapidly progressive dementia: An eight year (2008–2016) retrospective study

Patil Anuja[1☯], Vishnu Venugopalan[2‡*], Naheed Darakhshan[1☯], Pandit Awadh[2‡], Vinny Wilson[1‡], Goyal Manoj[1‡], Modi Manish[1‡], Lal Vivek[1‡]

Table 2. Infectious causes presenting with rapidly progressive dementia.

Sr. no	Diagnosis:	No. of patients:	Symptom duration:
1	HSV encephalitis	1	15days
2	Tubercular meningitis	2	5.5months
3	Cryptococcal meningitis	1	4months
4	TBM+ Cryptococcal meningitis	1	12months
5	Neurocysticercosis	3	3.3months
6	SSPE	17	3.94months
7	HIV dementia	1	3months
8	HIV + PMLE	6	4.33months
9	Neurosyphilis	7	9.14months

Etiology of RPDs

Neurology: Clinical Practice ▮ September 2012

Disease	Onset	Demographics[b]	Clinical features	MRI	CSF	Other tests	Treatment
Infectious							
Neurosyphilis	S	Consider risk	Cognitive decline	Nonspecific	CSF VDRL	Serum RPR	Crystalline IV

Infectious agents (other than prions) possibly causing RPD**

Viral
- HIV
- JC-virus (progressive multifocal leukoencephalopathy)
- Herpes simplex virus 1, 2 (6, 7)
- Varicella zoster virus
- Measles (sclerosing subacute panencephalitis)
- Mumps
- Epstein–Barr virus
- Influenza virus
- Cytomegalovirus
- Japanese encephalitis virus
- West Nile virus
- Rabies
- Parvovirus B19
- Hepatitis C

Bacterial
- *Treponema pallidum* (Lues)
- *Tropheryma whipplei* (Whipple's disease)
- *Mycobacterium tuberculosis*
- *Listeria monocytogenes*
- *Borrelia burgdorferi*

Parasitic/Fungal
- *Toxoplasma gondii*
- Balmuthia
- *Cryptococcus neoformans*
- Aspergillosis

Inga Zerr & Peter Hermann, 2018

HIV dementia	A/S	Seroconversion, older HIV-positive adults, low CD4	Psychomotor slowing, executive dysfunction, depression, movement disorders	Cortical atrophy; nonspecific white matter changes	Increased protein, mild pleocytosis	HIV serology; serum and CSF viral loads	CNS penetrating HAART
Herpetic meningoencephalitis	A	Any age	Altered level of consciousness, focal deficits, seizures, behavioral changes; fever	Medial temporal lobe hyper on FLAIR, asymmetric; later hemorrhagic necrosis[e6]	Lymphocytic pleocytosis, ↑RBC, HSV-1 PCR+	EEG: focal abnormalities, PLEDs	IV acyclovir for 14–21 days (start early if suspected)[e6]

Ross W. Paterson et al, 2012

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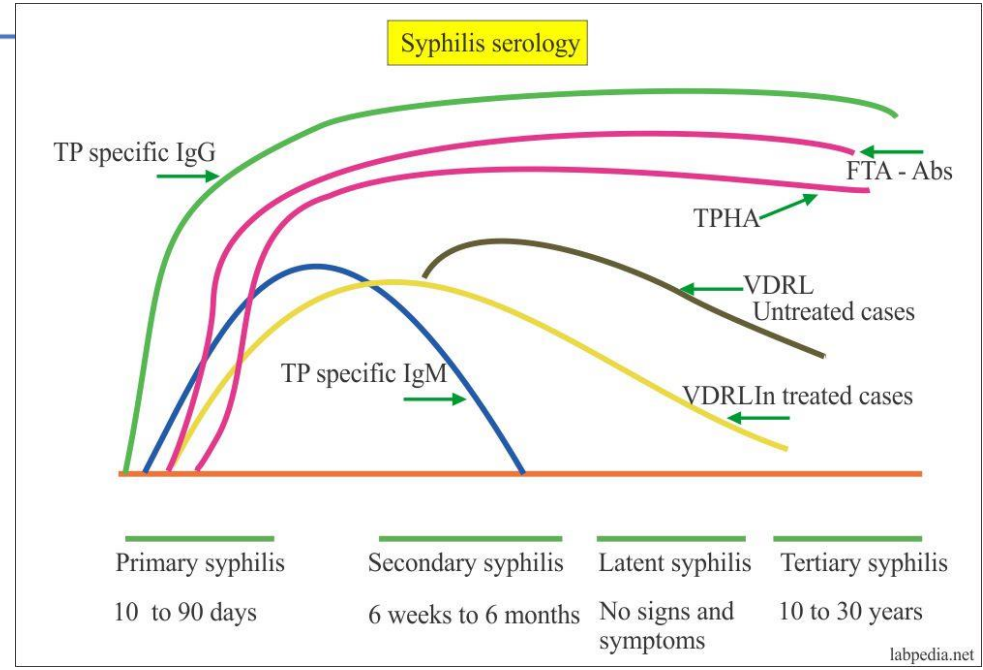

malattie responsabili di decadimento cognitivo

Malattia di Alzheimer

Malattia di Parkinson e demenza a corpi di Levy

Demenza fronto-temporale

Malattia cerebro-vascolare

Encefalopatie spongiformi

Malattie infiammatorie

REVIEW
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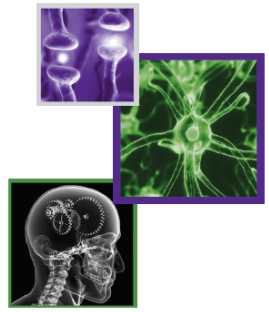
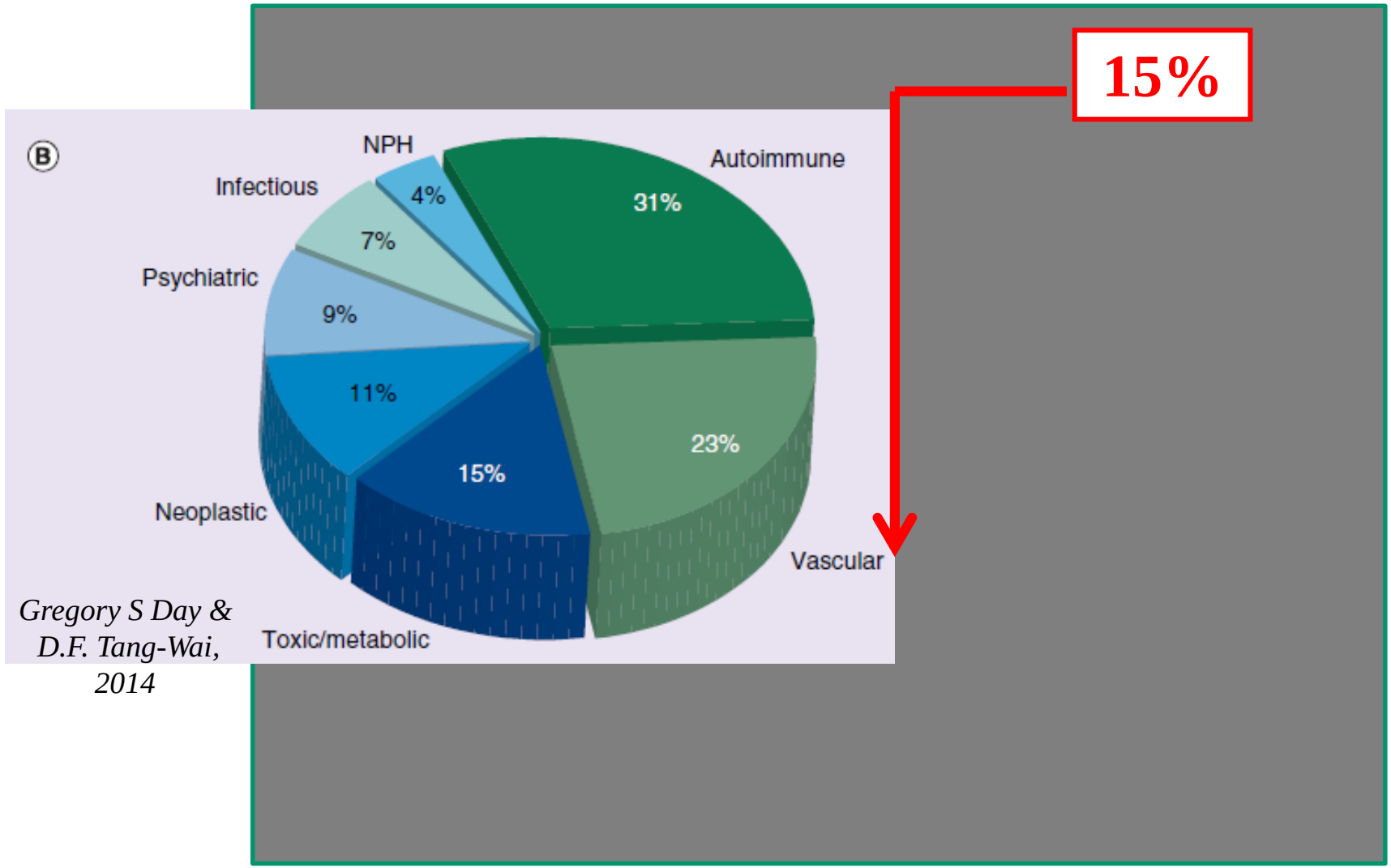
When dementia progresses quickly: a practical approach to the diagnosis and management of rapidly progressive dementia



Gregory S Day*[1] & David F Tang-Wai[1]

Neurodegen. Dis. Manage. (2014) 4(1), 41–56



Gregory S Day & D.F. Tang-Wai, 2014

Case report

Clin Ter 2015; 166 (2):68-71. doi: 10.7417/CT.2015.1818

Posterior Reversible Encephalopathy Syndrome in late postpartum eclampsia

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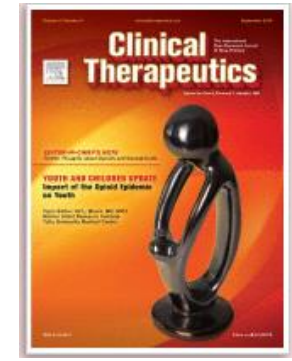
Departments of [1]Anesthesia and Intensive Care, [2]Neurology and Radiology, [3]"Pugliese-Ciaccio" Hospital, Catanzaro, Italy



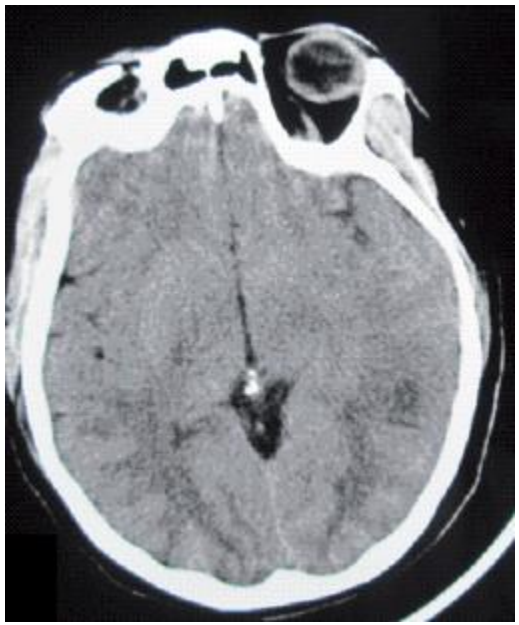


Fig. 1. Computed tomography:

hypodensity area in the left temporo – occipital region.

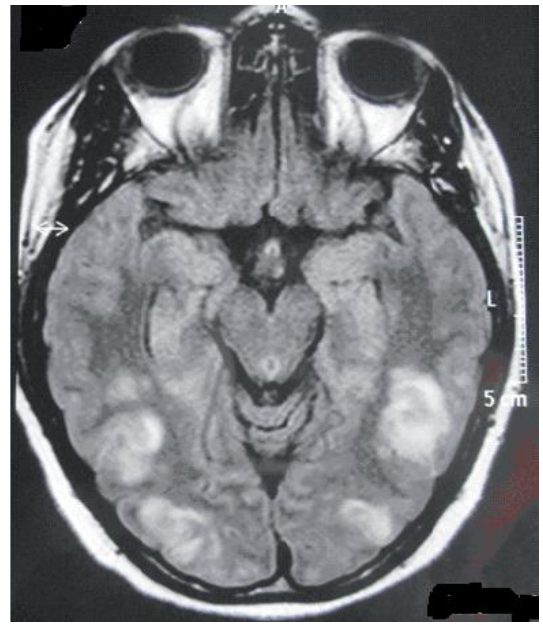


Fig. 2. Encephalic MRI: Presence of multiple patches of increased signal in weighted sequences in T2 mainly in the parietal, temporal and occipital lobes bilaterally, with cortical-subcortical localization, by parenchymal edema.

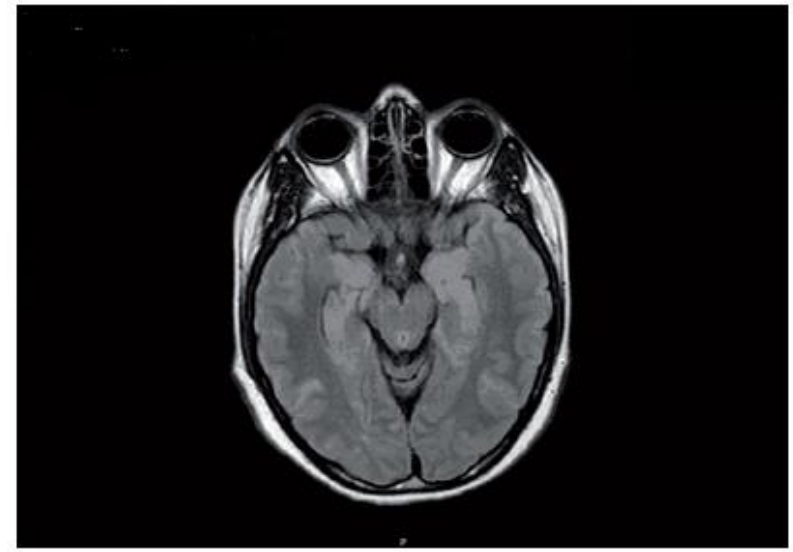


Fig. 3. Encephalic MRI: It highlights the disappearance of all lesions.

malattie responsabili di decadimento cognitivo

Neurology: Clinical Practice ▮ September 2012

Diagnosis and treatment of rapidly progressive dementias

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Leonel T. Takada, MD*
Michael D. Geschwind, MD, PhD

Although no formal definition exists for tia (RPD), generally we use the term from illness onset, but more comm conditions are relatively uncommon, what constitutes a rapidly progressive demen- when dementia occurs in less than 1–2 years only over weeks to months.[1] Because these

Malattia di Alzheimer

Malattia di Parkinson

demenza a corpi di Levy

Demenza fronto-temporale

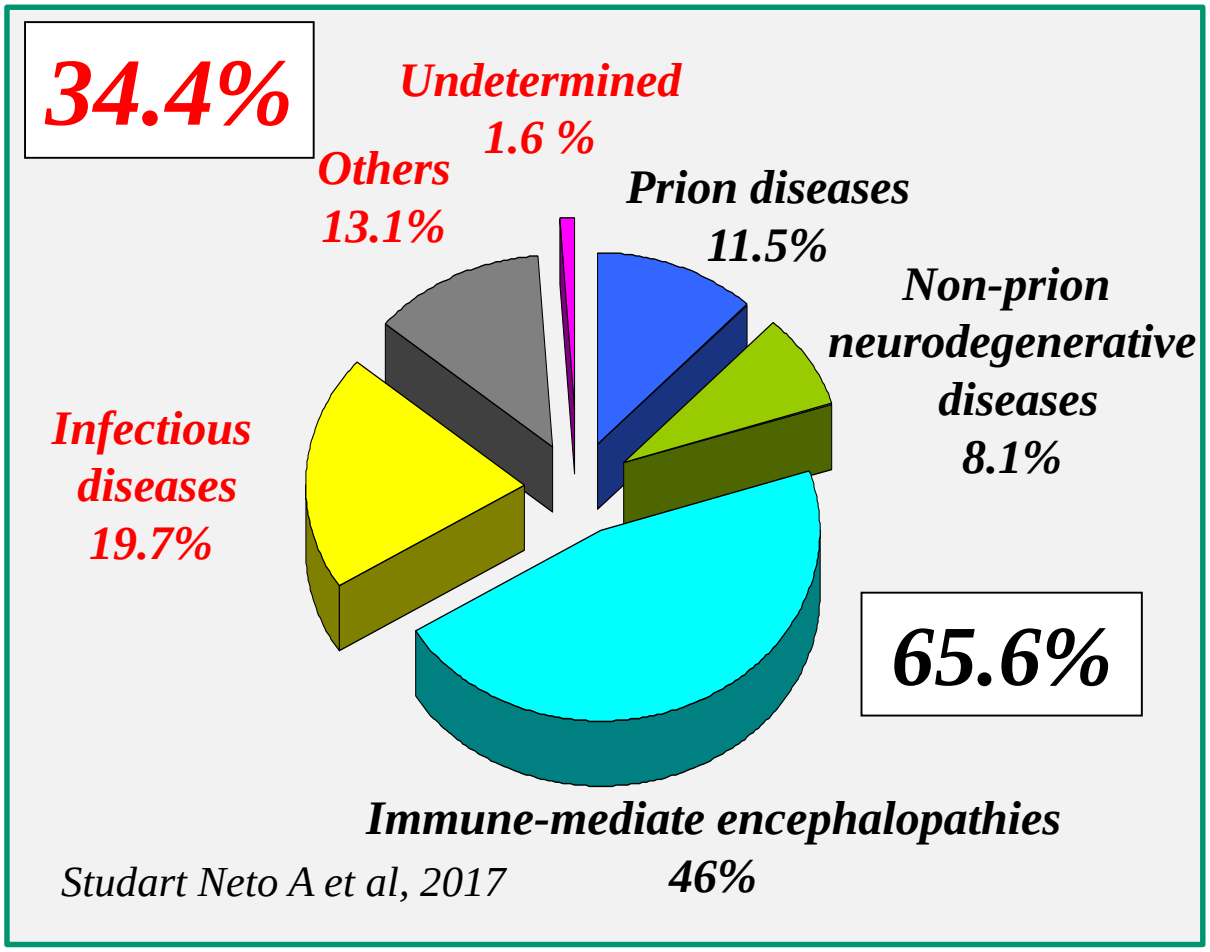
Malattia cerebro-vascolare

Encefalopatie spongiformi

Malattie infiammatorie

Altre etiologie

61 RPD cases



Frequency of Clinical Diagnoses in Patients with RPD

- Altered level of consciousness, ranging from confusion to coma
- Headache
- Seizures
- Visual symptoms (usually, blindness or hemianopia)
- Neuroradiology findings
- Exclusion of other pathology (eg. encephalitis or stroke)
- Acute onset, and reversibility over days or weeks
- Hypertension