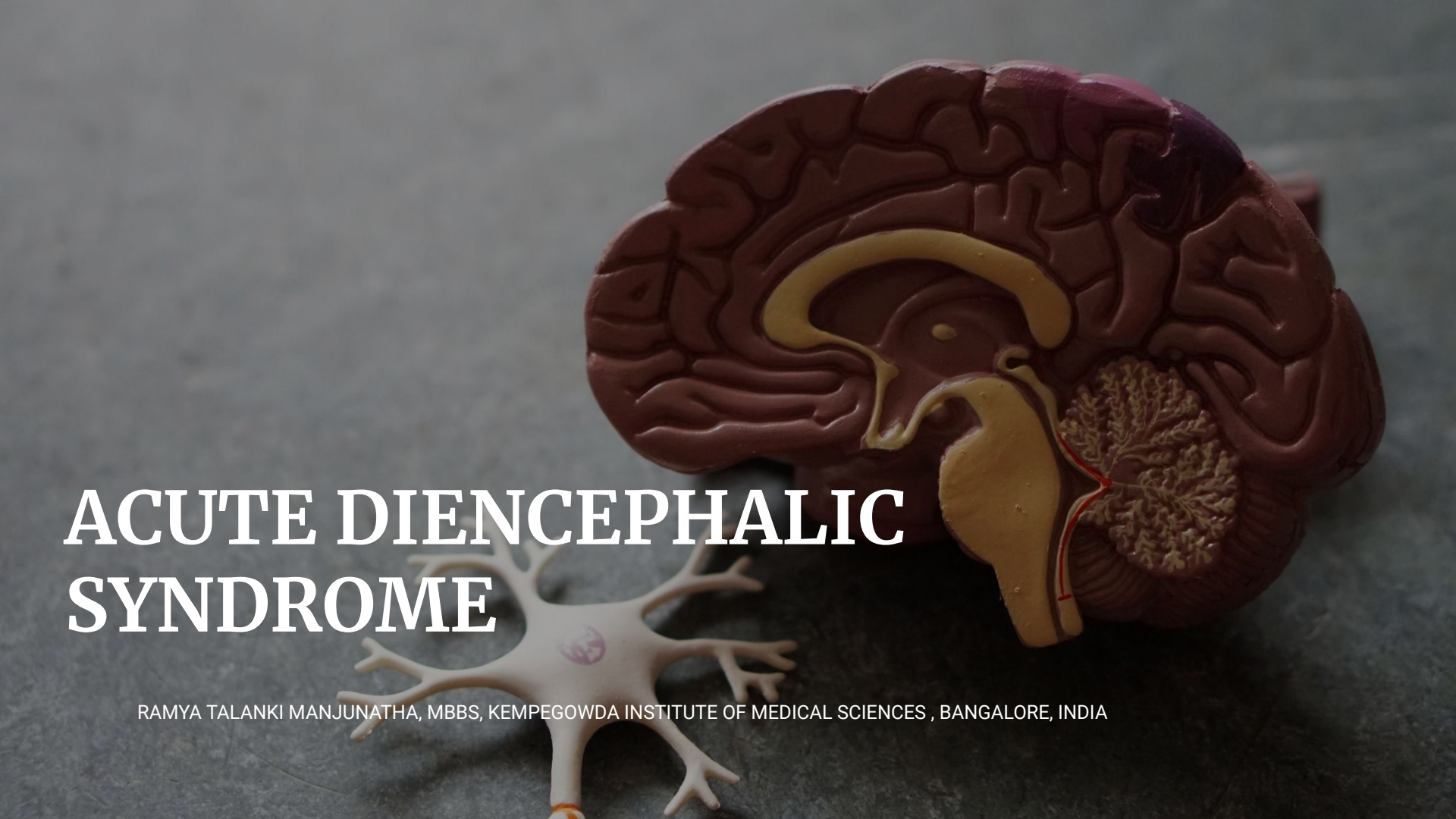


ACUTE DIENCEPHALIC SYNDROME

RAMYA TALANKI MANJUNATHA, MBBS, KEMPEGOWDA INSTITUTE OF MEDICAL SCIENCES , BANGALORE, INDIA

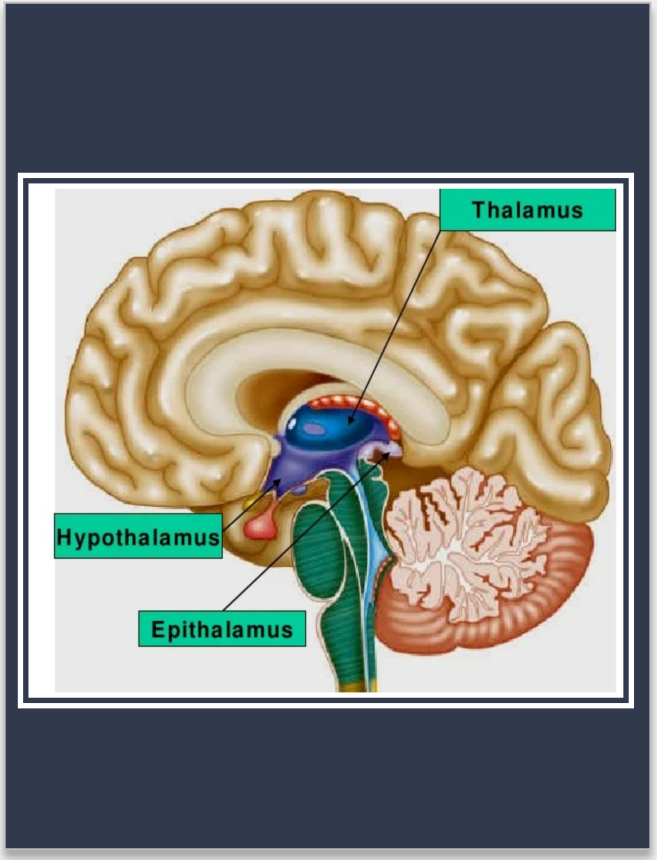
DIENCEPHALON

Thalamus

Hypothalamus

Epithalamus

Subthalamus



CLINICAL FEATURES

Hypothermia

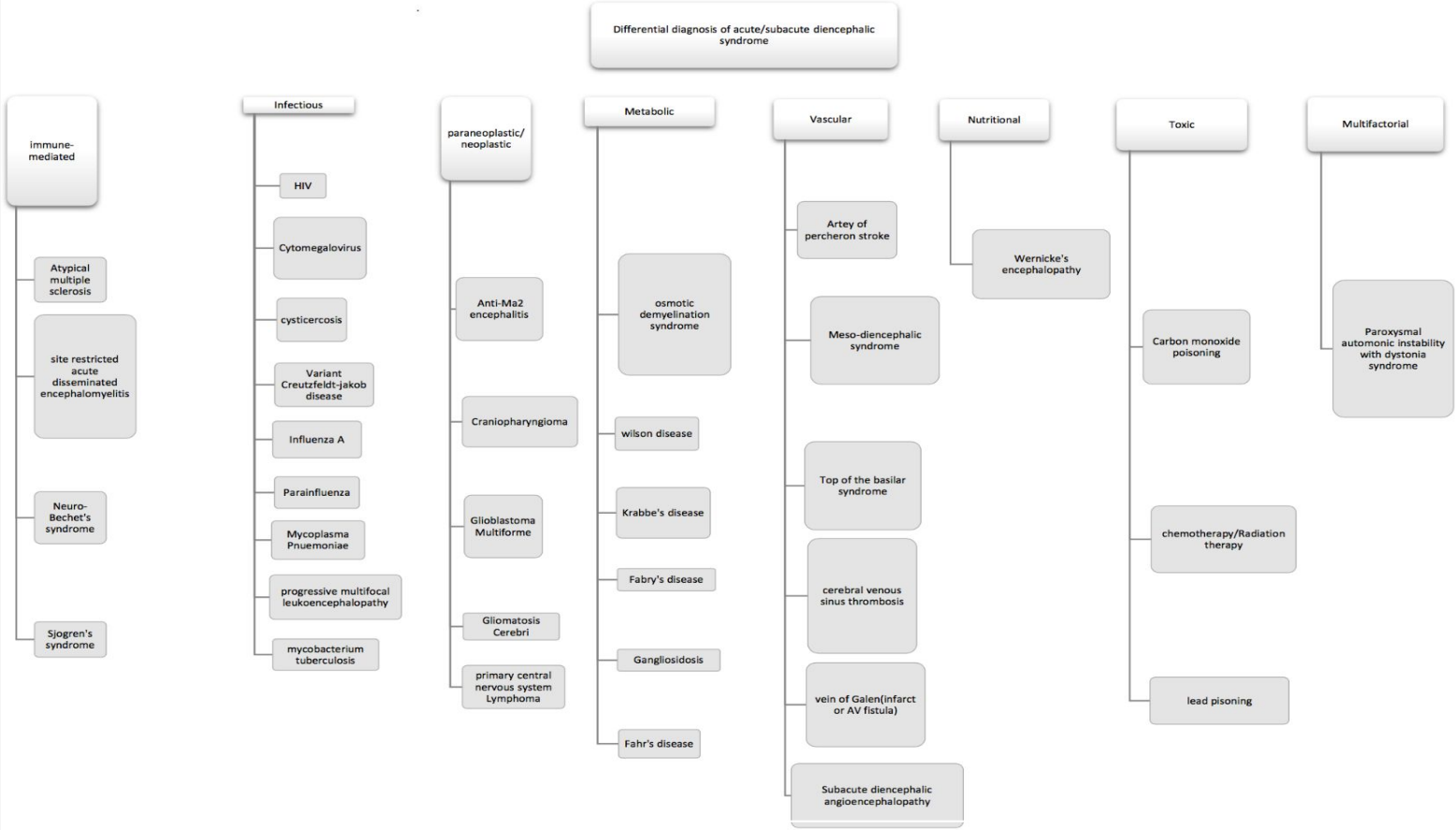
Hypotension

Hypersomnolence or narcolepsy-like syndrome

Bradycardia

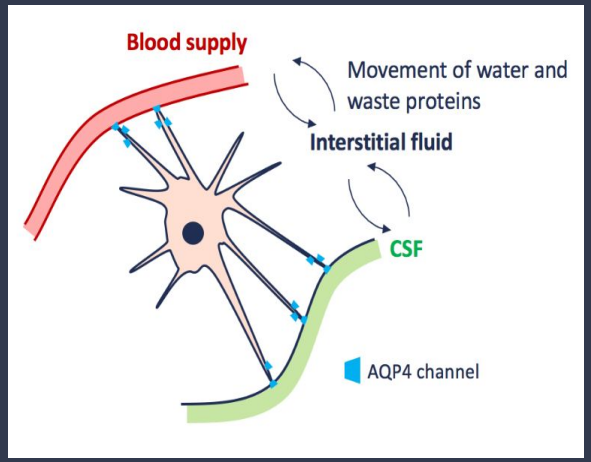
Anorexia with substantial weight loss

Endocrinological symptoms - SIADH, irregular menstruation, hyperprolactinemia

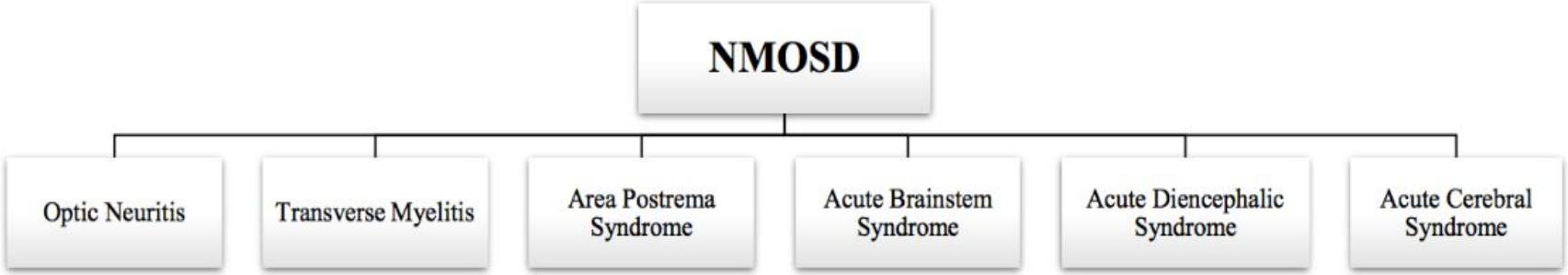


Jim Thomas, Medical graduate @ Houston Medical Clerkship

NMOSD



- Neuromyelitis optica spectrum disorders (NMOSD) are inflammatory disorders of the central nervous system characterized by severe, immune-mediated demyelination and axonal damage predominantly targeting optic nerves and the spinal cord.
- 90% cases present with relapsing - remitting NMOSD.
- Target antigen is AQP4 which forms transmembrane channel that primarily facilitates the movement of water across cell membranes in response to osmotic gradients.
- AQP4 is highly expressed in the periependymal regions. Periependymal lesions surrounding the third ventricle usually present as diencephalic syndrome.



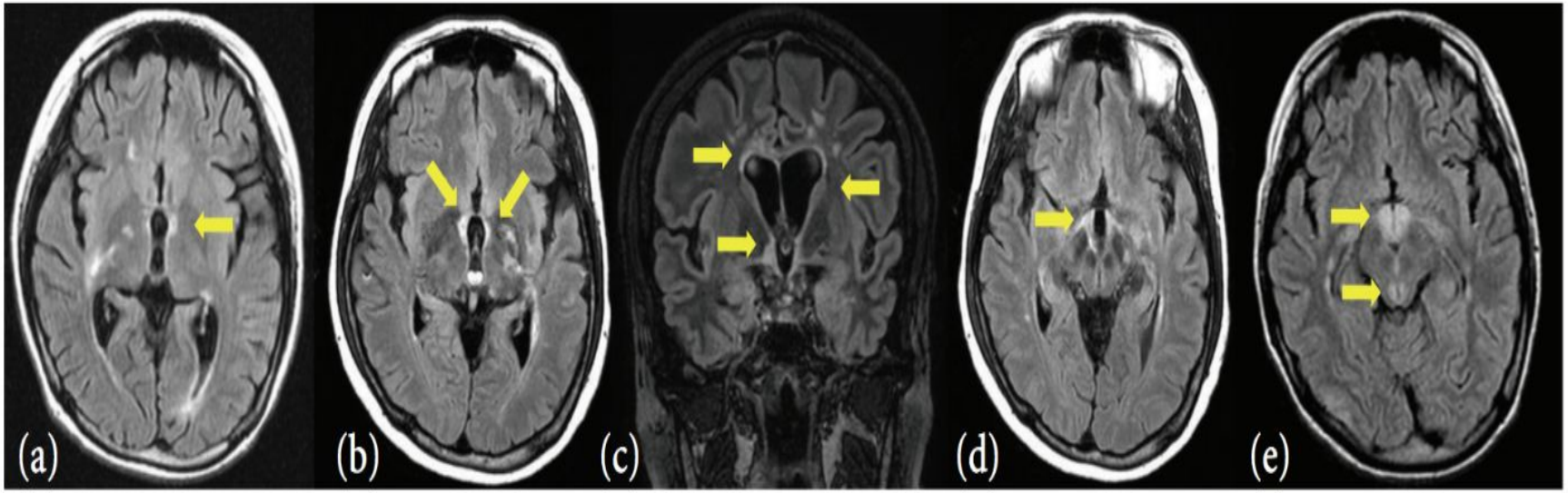
AQP4-IgG positive = 1 core clinical characteristic required, exclusion of alternate diagnosis

AQP4-IgG negative = 2 core clinical characteristic required, exclusion of alternate diagnosis

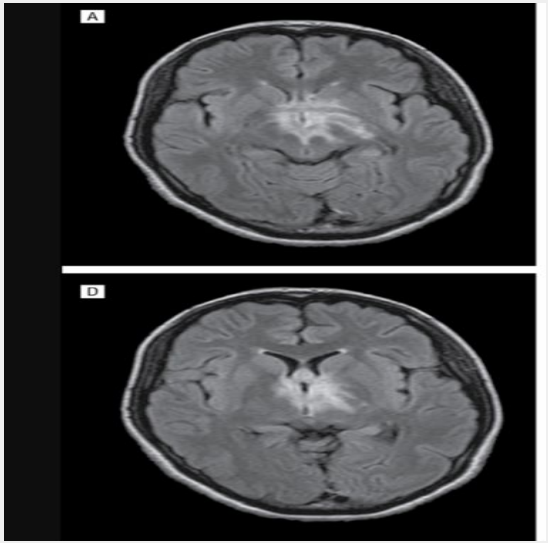
Note: 1 of 2 core clinical characteristic must be Optic Neuritis, Transverse Myelitis* or Area Postrema syndrome

*must be LETM

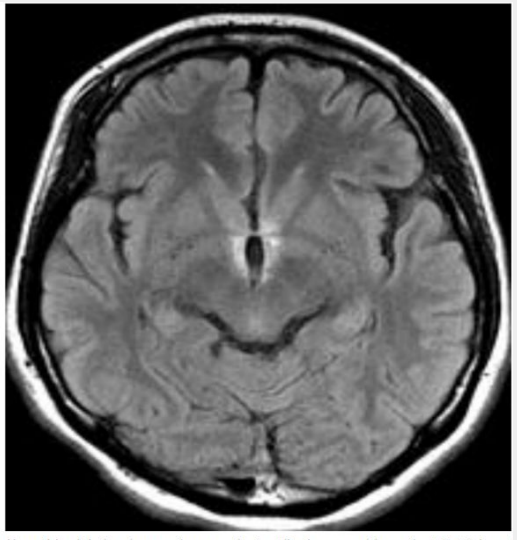
By: Dr. M. Faisal Khan & Dr. Neha Sharma (2020) @ Houston Medical Clerkship



Kim W, Kim SH, Huh SY, Kim HJ. Brain abnormalities in neuromyelitis optica spectrum disorder. Mult Scler Int. 2012;2012:735486. doi: 10.1155/2012/735486. Epub 2012 Dec 3. PMID: 23259063; PMCID: PMC3518965.



Suzuki K, Nakamura et al. Hypothermia, hypotension, hypersomnia, and obesity associated with hypothalamic lesions in a patient positive for the anti-aquaporin 4 antibody: a case report and literature review. Arch Neurol. 2012 Oct;69(10):1355-9. doi: 10.1001/archneurol.2012.300. PMID: 22777080.



Akaishi, Tetsuya; Nakashima, Ichiro; Sato, Douglas Kazutoshi; Takahashi, Toshiyuki; Fujihara, Kazuo (2017). *Neuromyelitis Optica Spectrum Disorders. Neuroimaging Clinics of North America, 27(2), 251–265.* doi:10.1016/j.nic.2016.12.010

MULTIPLE SCLEROSIS

Multiple sclerosis (MS) is a progressive inflammatory and degenerative disease of the human CNS that leads to demyelination and neuronal/axonal loss.

MS was considered to be a disease of the white matter but can studies have shown involvement grey matter from early stages of MS .

Thalamic involvement in MS presents with a wide range of symptoms from fatigue and movement disorders to pain syndromes and cognitive decline.

Thalamic lesions are usually more visible on MRI than cortical lesions, likely because the thalamus is normally more densely myelinated.

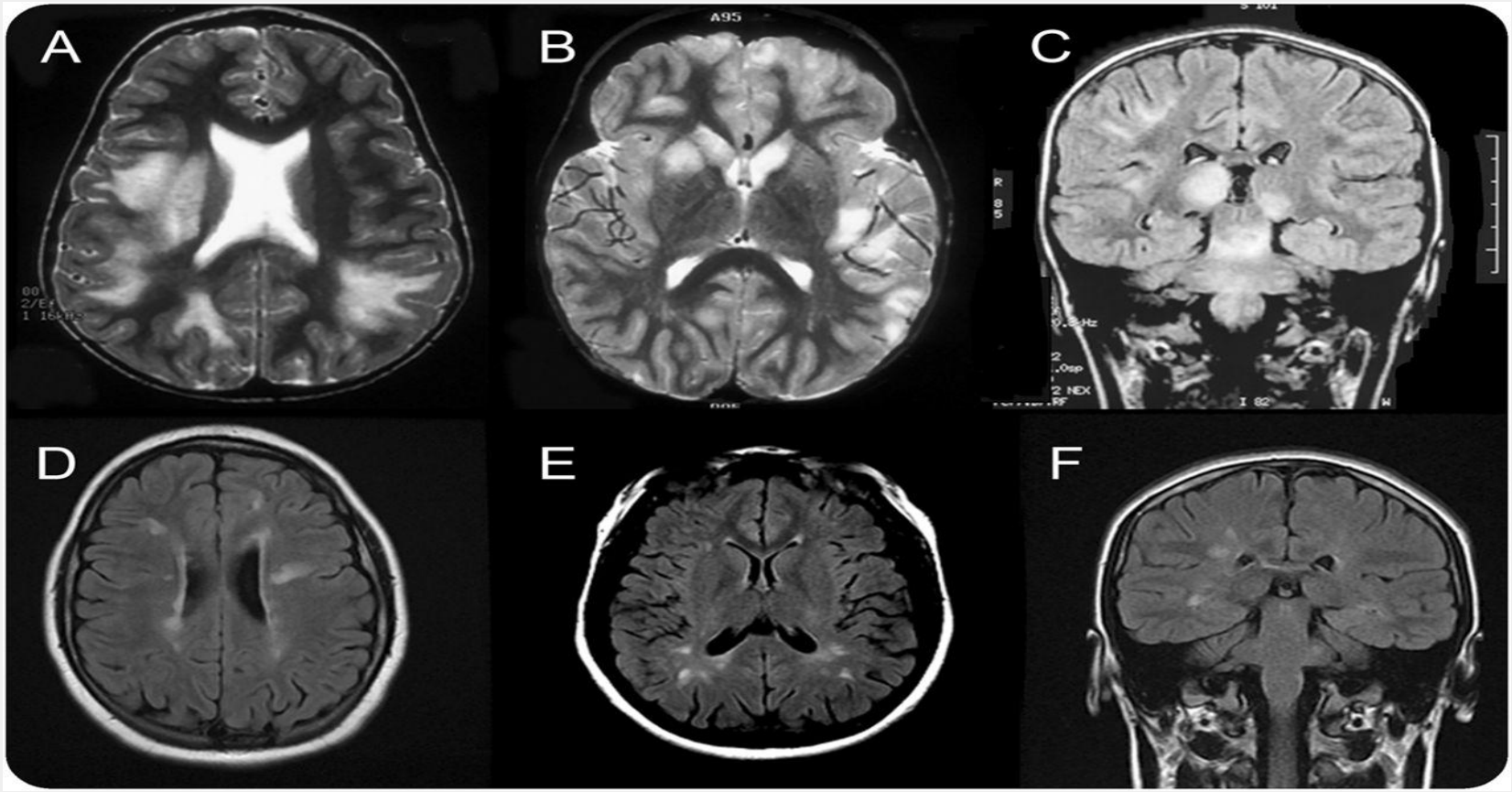
ACUTE DISSEMINATED ENCEPHALOMYELITIS

Acute disseminated encephalomyelitis (ADEM) is a rare kind of severe demyelinating disorder that affects the brain and spinal cord, usually in children(< 10 years).

The disease usually appears soon after a viral illness. It presents with fever, headache, seizures, confusion and vomiting. It is a monophasic disorder.

On MRI lesions are seen as multiple, widespread, asymmetric bilaterally throughout the brain with indistinct margins. CSF may disclose a mild lymphocytic pleocytosis and elevated albumin levels.

40% of ADEM lesions can present with thalamic involvement. 36-54% of patients have positive MOG antibodies.



Daniela Pohl et al. Neurology Aug 2016, 87 (9 Supplement 2) S38-S45;
DOI:10.1212/WNL.0000000000002825

NEURO-BEHCET DISEASE

Behcet's disease is a chronic relapsing inflammatory disorder of unknown etiology that involves many organ systems.

CNS is affected in 4%–49% of patients with Behcet disease, which has a predilection for men. Has predilection for meso-diencephalic region.

Neurological involvement in BD is classified into parenchymal and non-parenchymal group. Parenchymal NBD is more common and presents with attack of hemiparesis, cognitive changes, sphincter disturbance and possible fever. NBD involves blood vessels.

Typical radiologic finding of Neuro-Behcet's disease is involvement of the brainstem. Lesions are hyperintense on T2 weighted MR images, are hypointense on T1-weighted images with vasogenic edema.

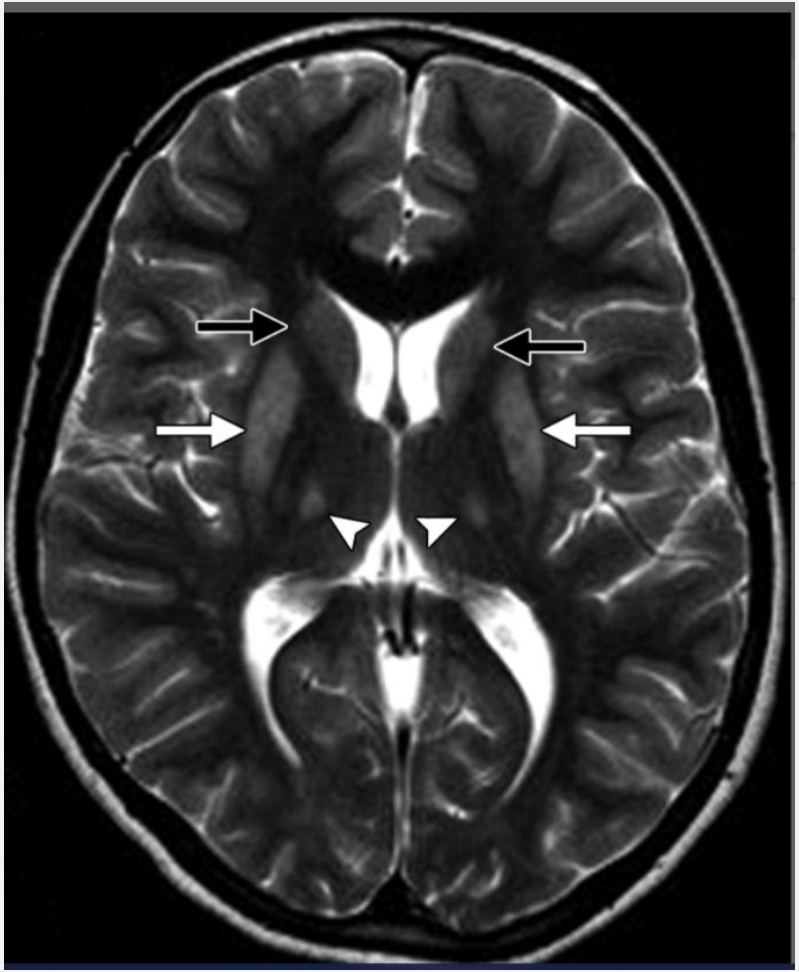
WILSON'S DISEASE

It is an autosomal recessive inborn error of copper metabolism due to ceruloplasmin deficiency. It is also known as hepatolenticular degeneration.

The symptoms vary and include dysarthria, dystonia, tremors, ataxia, parkinsonian symptoms, and psychiatric problems. The characteristic finding is KF rings in the cornea.

MR images show symmetric T2 hyperintensity of the deep gray matter: putamen, globus pallidus, caudate nuclei, and the thalami. Thalamic involvement is confined to ventrolateral aspect.

Treatment is with penicillamine which has been shown to reverse MRI findings. If not treated the condition can be fatal.



ACQUIRED METABOLIC DISORDERS

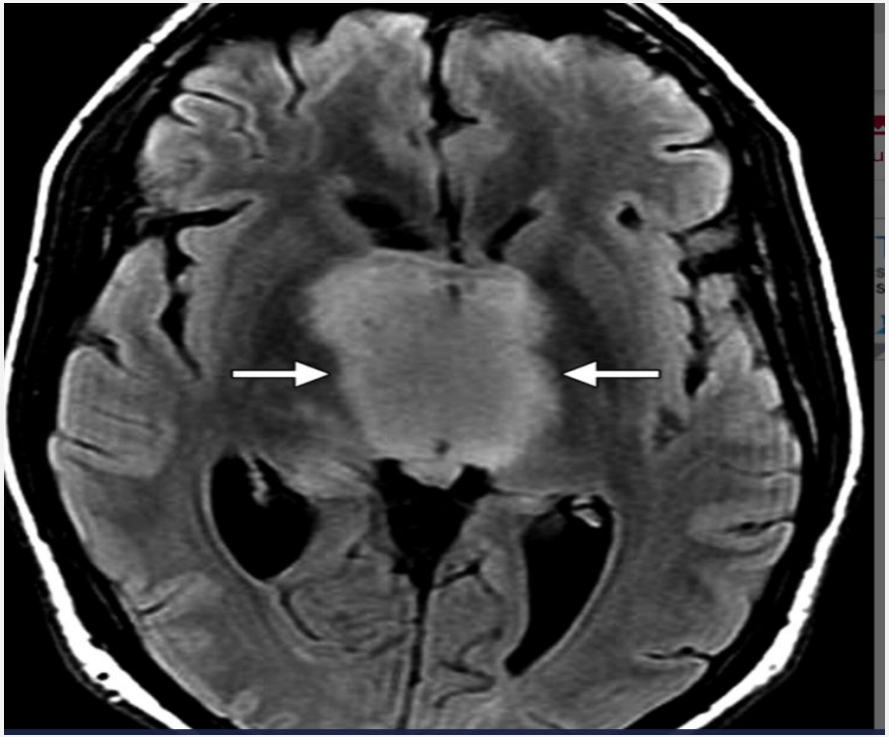
Thiamine deficiency encephalopathy (Wernicke's encephalopathy)	FLAIR, PD/T2	Hyperintense signal in the medial part of the thalami, in the mamillary bodies, in the hypothalamus, and in the periaqueductal gray matter
	T1	Hypointense signal in the affected areas
	T1 + Gd	Mamillary bodies show contrast enhancement
	DWI	Diffusion restriction
Fahr's disease	CT	Excessive bilateral calcifications of the basal ganglia, the thalami, the cerebellum, and the supratentorial white matter
Pontine und extrapontine myelinolysis (osmotic demyelinization syndrome)		*Pontine myelinolysis:* confluent hyperintense lesion in the central pons, with spar ing of the periphery and the corticospinal pathways
		Extrapontine manifestations: typically affect basal ganglia and white matter; involvement of thalami (lateral geniculate bodies) less common
	FLAIR, PD/T2	Hyperintense signal of the affected areas
	T1	*Acute state:* mild to moderate hypointense signal in the affected areas, isointense signal intensity less common
		Subacute state: lesions might be hyperintense
	T1 + Gd	Lesions only rarely show contrast enhancement
	T2*-GRE	Might show small hemorrhages
	DWI	Slight restriction of diffusion
Profound hypoxia of the newborn	DWI	Restriction of diffusion within the first 24 h of life
	T1	T1 shortening within 2–3 days
	T2	T2 prolongation within the first 24 h, T2 shortening within 6–7 days
	MRS	Elevated lactate (within few hours after birth)

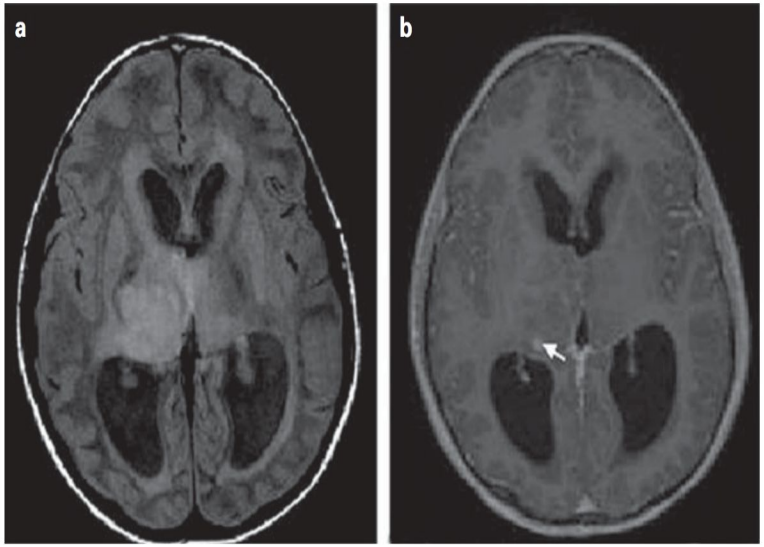
NEOPLASTIC

GLIOMAS

- About 1% of CNS tumours are thalamic. The typical clinical symptoms of bithalamic astrocytomas are personality changes and mental decline, but without any essential motor or sensory impairment.
- Astrocytomas can be low grade or high grade (glioblastoma multiforme).
- MRI reveals hyperintensity on T2-weighted images and hypointensity on T1-weighted images that is not associated with contrast enhancement.

Other neoplasms which may involve thalami include primary CNS lymphoma, dysgerminoma and teratoma, craniopharyngioma.





Figures 7a and 7b. Bithalamic astrocytoma, grade III, in a 14-year-old male adolescent with Recklinghausen's disease. The tumor is hyperintense in the FLAIR sequence (a) and shows small, focal uptake of contrast medium in the contrast-enhanced T1-weighted image (b, arrow). Besides, a hydrocephalus is present.

INFECTIONS

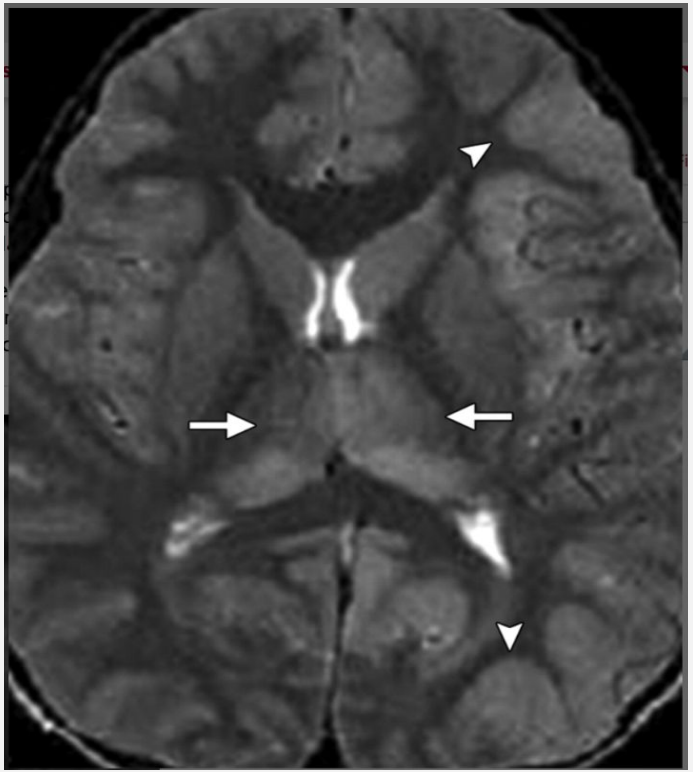
VIRAL - West Nile encephalitis, Japanese encephalitis, Murray Valley encephalitis, Eastern equine encephalitis, Rabies, EBV, HIV-1

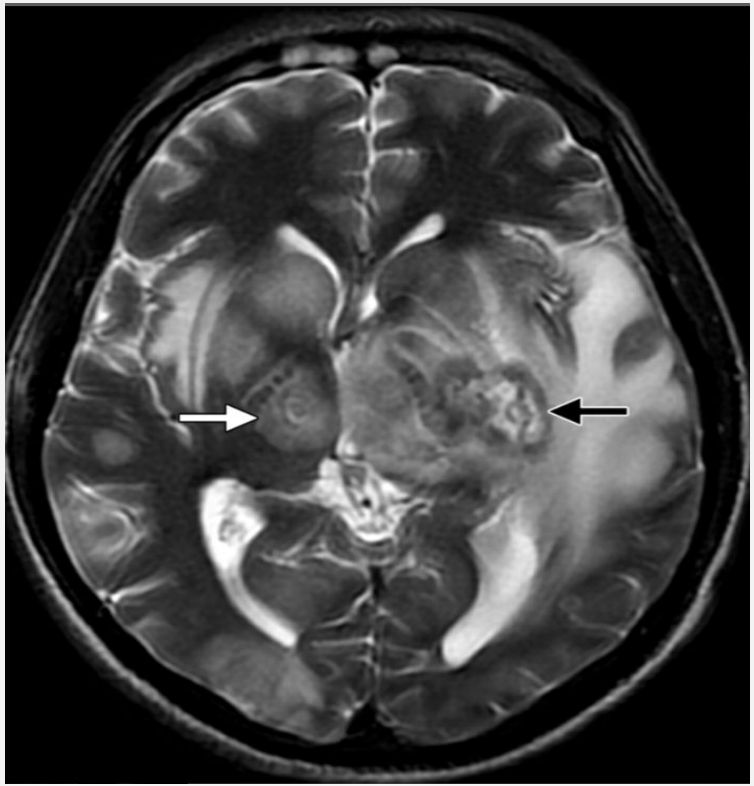
The clinical presentation typically involves a prodromal phase of fever, rigors, headache, rashes, and body aches followed by CNS symptoms that include dyskinesia, dystonia, tremors, drooling, dysarthria, altered consciousness, seizures, and coma. The most characteristic MR imaging finding of Japanese encephalitis is T2 hyperintensity, typically with bilateral involvement of the posteromedial thalamus.

BACTERIAL - Rare cases of TB meningoencephalitis

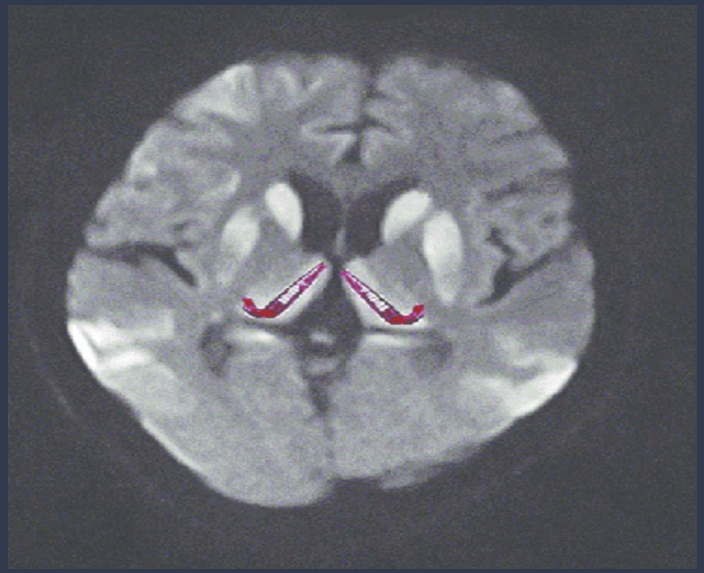
FUNGAL - Histoplasma, Cryptococcus

PARASITIC - Toxoplasmosis, Malaria

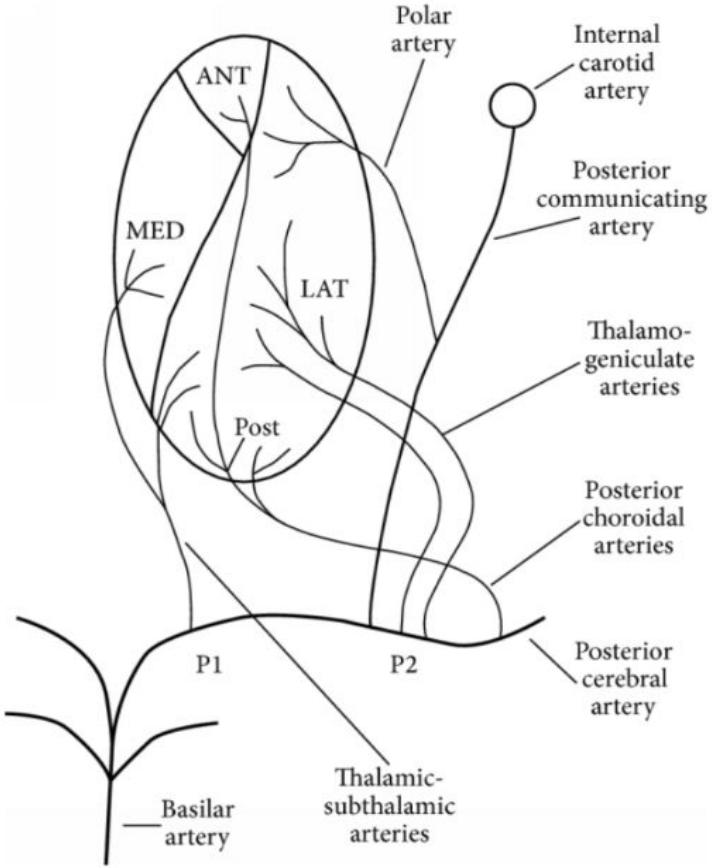




CREUTZFELDT–JAKOB DISEASE

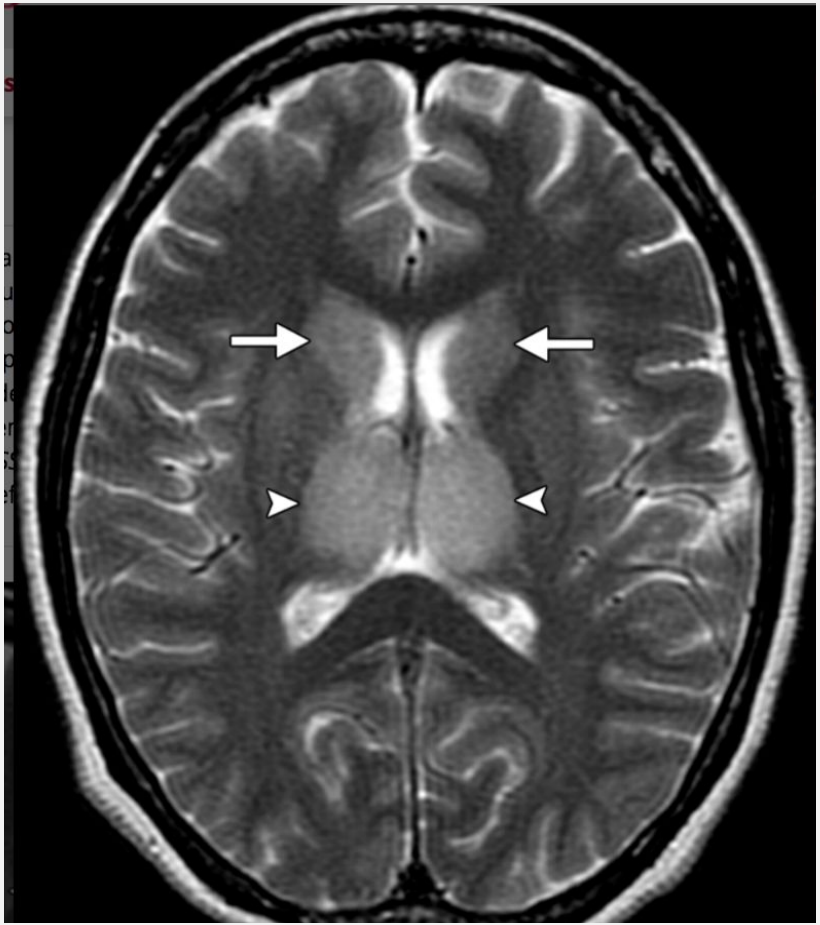


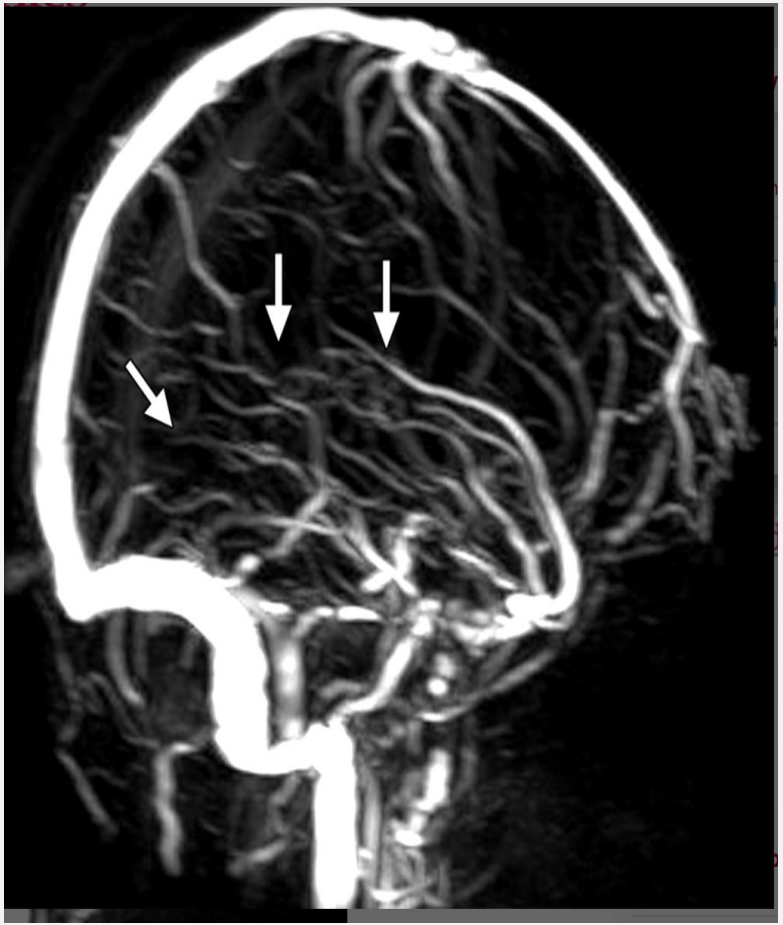
- Spongiform encephalopathy with a rapidly progressive fatal course.
- Patients with CJD typically present with rapidly progressive dementia, myoclonus, and multifocal neurologic dysfunction.
- MRI may reveal T2 prolongation and reduced diffusion in the basal ganglia, thalami, and cortical ribboning.
- A key imaging finding seen in variant CJD is pulvinar or hockey stick sign with typical bilateral lesions in the pulvinar nuclei of thalami.


Polar artery
Internal carotid artery
ANT
Posterior communicating artery
MED
LAT
Thalamo-geniculate arteries
Post
Posterior choroidal arteries
P1
P2
Posterior cerebral artery
Thalamic-subthalamic arteries
Basilar artery

CEREBRAL VEIN THROMBOSIS

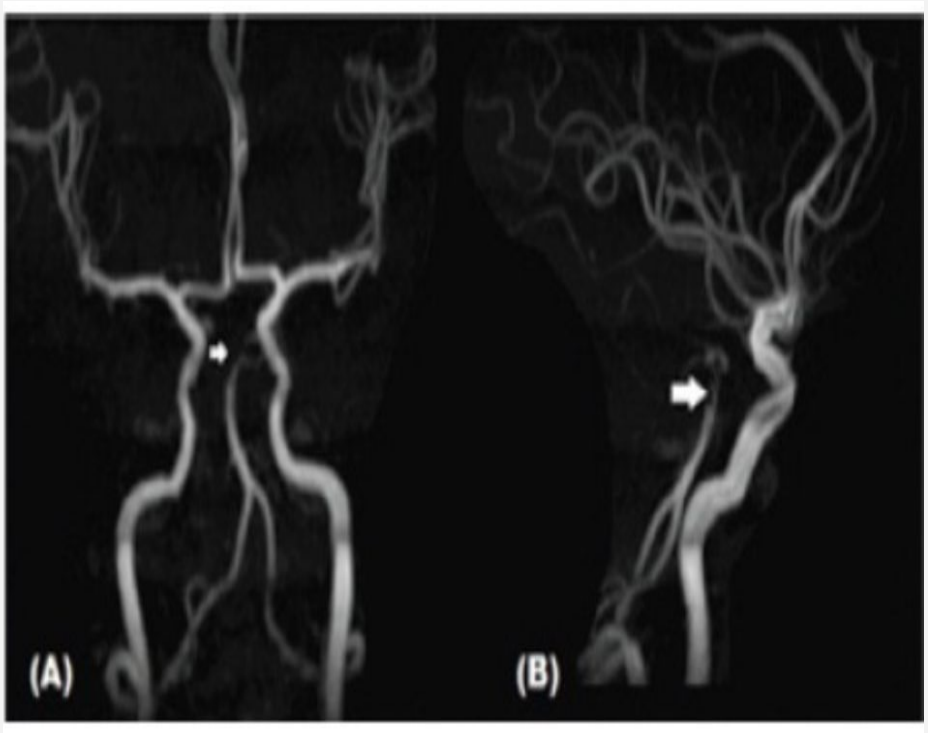
- CVT of the deep vein system(vein of galen, straight sinus, internal cerebral vein) results in B/L symmetric involvement of the thalami and basal ganglia.
- The causes include pregnancy, oral contraceptives, infection, trauma, and dehydration, cause is unknown in 20–25% of patients.
- Patients present with acute headache, nausea, vomiting, seizures, and altered mental status. Focal neurologic deficits, coma, and death may occur in severe cases.
- Hyperdense vein may be seen on CT scan. CTA shows filling defects in the affected vein. MRI shows T2 hyperintense lesions and phase contrast MRV can reveal absence of normal blood flow in the veins.

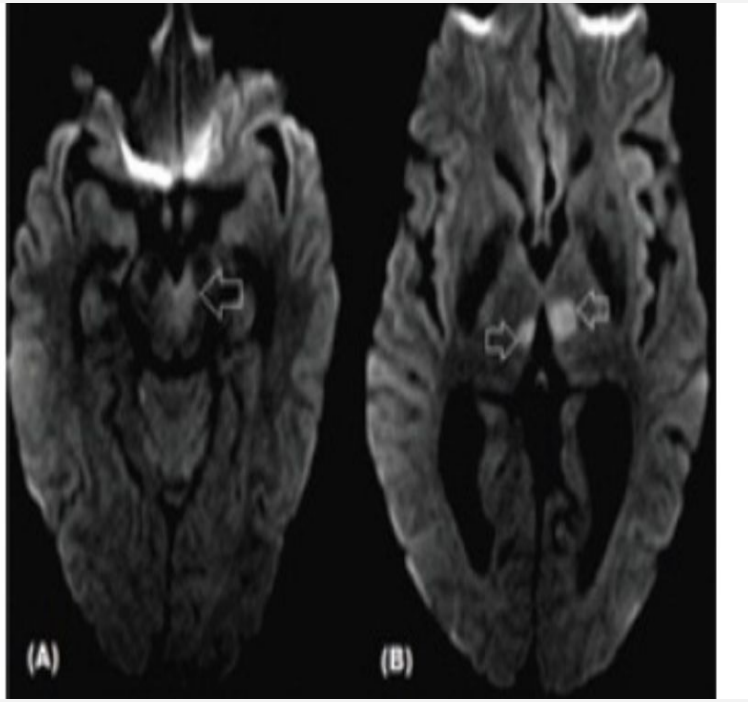




TOP OF BASILAR ARTERY SYNDROME

- Top of the Basilar Artery Syndrome is also known as a Rostral Brainstem Infarction.
- The cause of this typical bilateral infarct pattern is commonly a thrombosis at the top of the basilar artery or an embolic occlusion of the common main trunk of the posterior thalamic perforating arteries from the proximal P1 segment of a PCA.
- Patients with thalamic infarction typically present with agitation, obtundation or coma, memory dysfunctions, and various types of ocular and behavioral changes.
- CT shows a hyperdense basilar artery. Angiography can be used to confirm the finding by demonstrating filling defects.

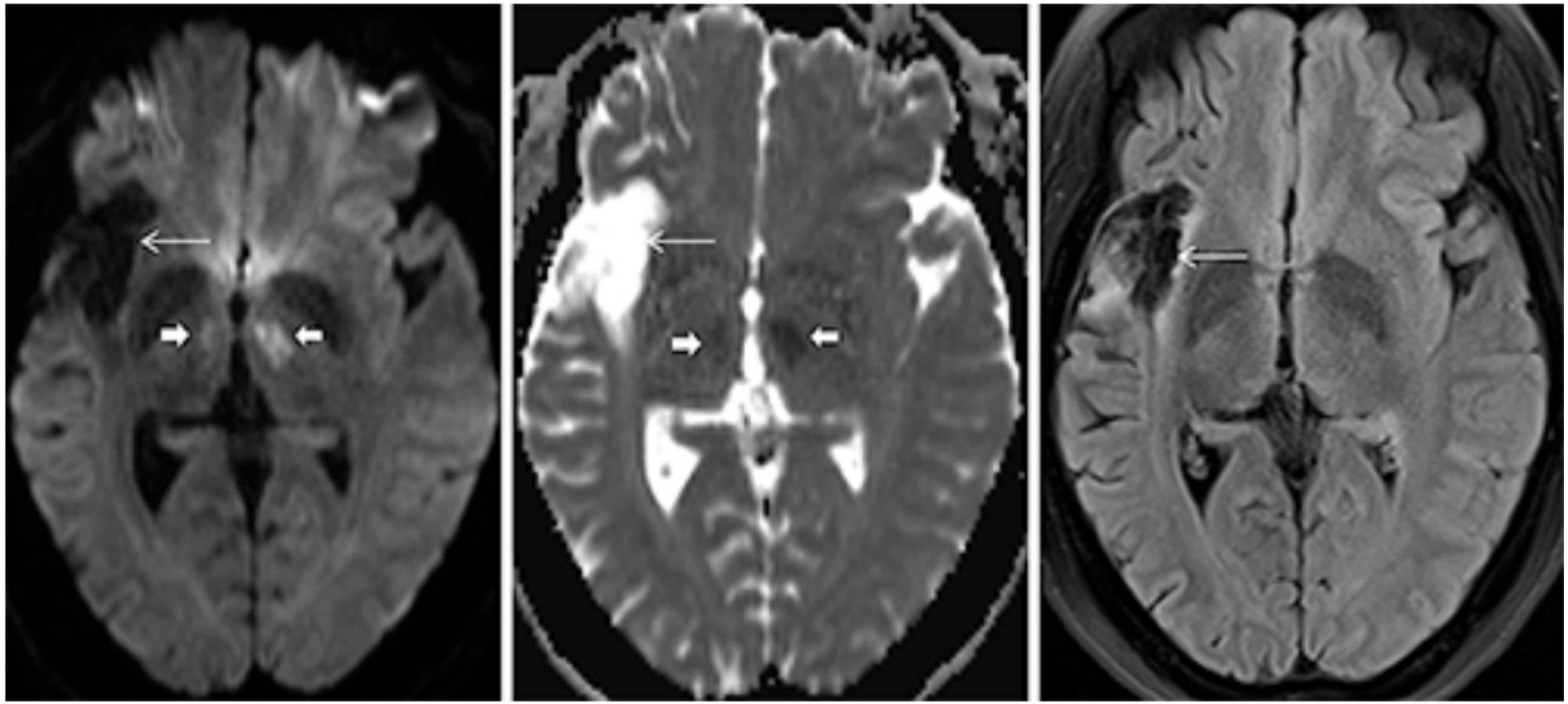




Suthar PP, Rana PJ, Mehta C, Patel NA. Top of Basilar Artery Syndrome. *J Clin Diagn Res*. 2015;9(7):TJ01. doi:10.7860/JCDR/2015/12250.6193

ARTERY OF PERCHERON STROKE

- The artery of Percheron, a variant, is a solitary arterial trunk arising from the proximal segment of the posterior cerebral artery and supplying the paramedian thalami and rostral midbrain bilaterally.
- Infarct in the AOP territory presents with non-specific symptoms such as memory loss, fluctuating level of consciousness, and altered mental status.
- A diffusion-weighted MRI of the brain in the early hours is the investigation of choice.



Sasi S, Ahmed A, Yousuf W, Vattoth S: Artery of Percheron Infarct: A Rare Presentation of Acute Ischemic Stroke in a High-Risk Antiphospholipid Syndrome Patient. Case Rep Acute Med 2020;3:46-52. doi: 10.1159/000509569

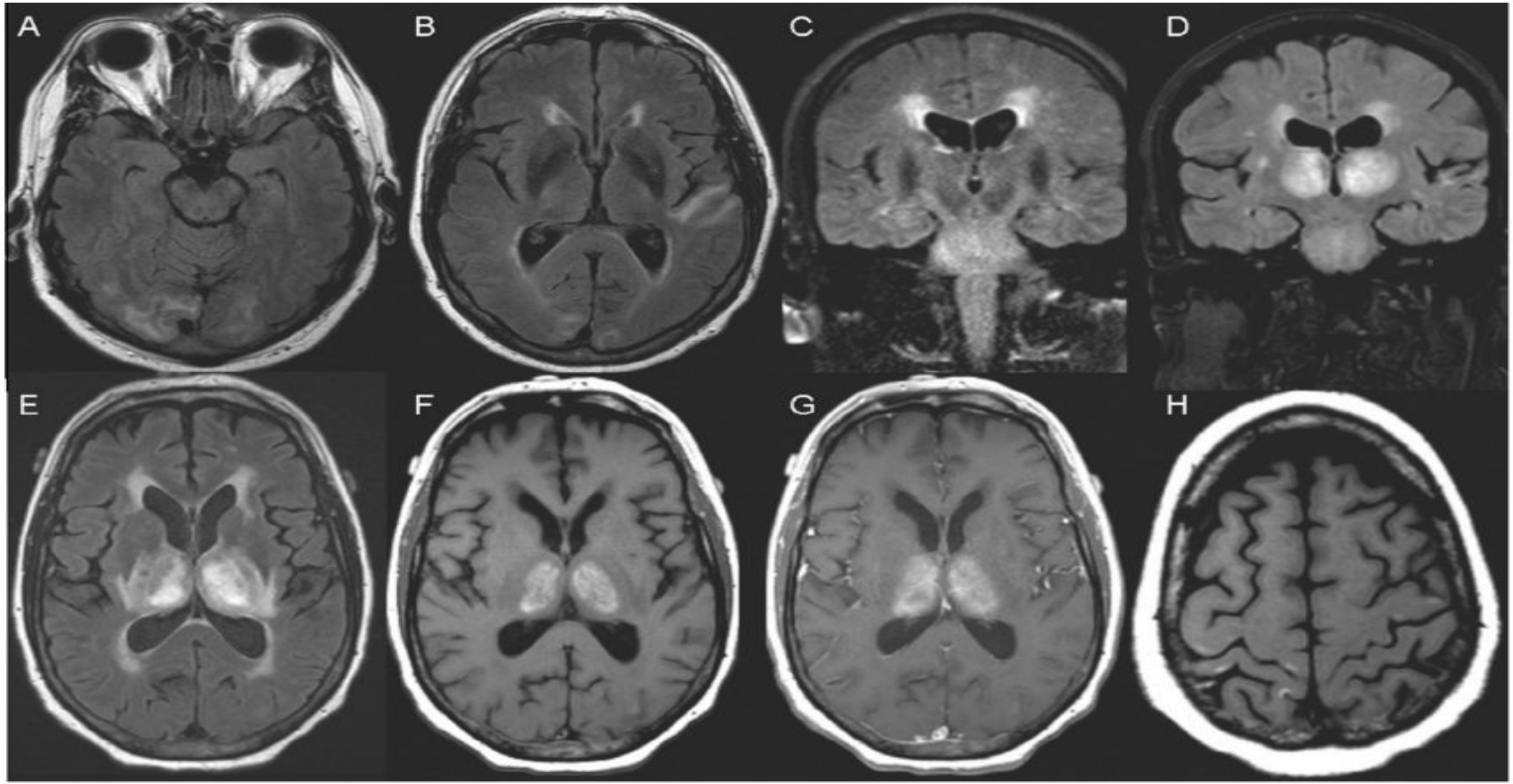
SUBACUTE DIENCEPHALIC ANGIOENCEPHALOPATHY (SDAE)

- Subacute diencephalic angioencephalopathy (SDAE), first described by DiGirolami el al. in 1974 is an extremely rare and fatal condition with an unknown etiology. Elderly male with hypertension are at the risk of developing SDAE.

- SDAE presents as a rapidly progressive encephalopathy characterized by progressive intellectual, motor and vegetative impairment. The disease can present with subacute confusion, impaired consciousness,headaches, dysarthria, language deficits, gait abnormalities ultimately leading to death.

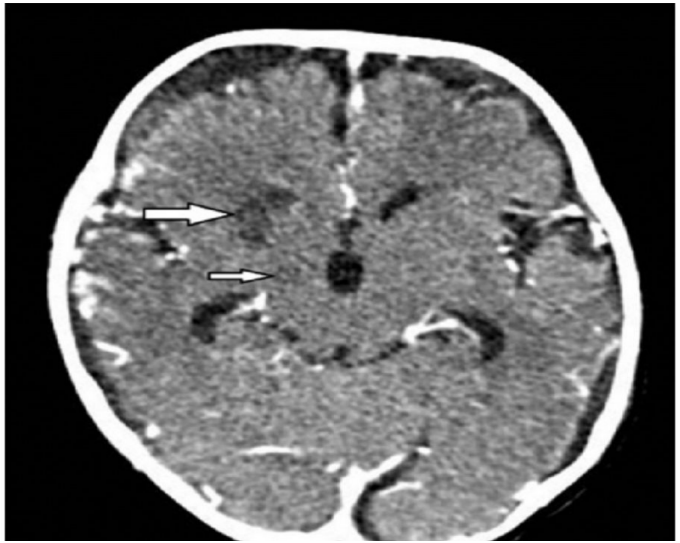
- Histopathology reveals patchy necrosis with dystrophic calcification, neuronal loss, gliosis and hyalinization of small arteries with mononuclear cell infiltration.

- MRI brain shows T1 and T2 hyperintensities in bilateral thalami with unremarkable vascular imaging. CSF analysis revealed a normal cell count with increased protein .



Graffeo et al. 2015. *Expanding the spectrum of subacute diencephalic angioencephalopathy. Journal of Clinical Neuroscience, (), S0967586815003549–.* doi:10.1016/j.jocn.2015.06.016

PAROXYSMAL AUTONOMIC INSTABILITY WITH DYSTONIA



Paroxysmal autonomic instability with dystonia (PAID) syndrome, a subset of dysautonomia, is characterized by paroxysms of marked agitation, diaphoresis, hyperthermia, hypertension, tachycardia and tachypnea accompanied by hypertonia and extensor posturing.

PAID is associated with trauma, anoxic injury, tuberculous meningitis, interpeduncular tuberculoma, pneumococcal meningoencephalitis, intracerebral hemorrhage and paraneoplastic limbic encephalopathy.

The pathophysiology is still uncertain, but best explained by dysfunction of autonomic centres in the diencephalon or their connections to cortical, subcortical and brainstem loci that mediate autonomic function.

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