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Jugular Entrapment, Dilated ventricles, Intracranial Hypertension (**JEDI** Syndrome): a new clinical entity?

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Zamboni²

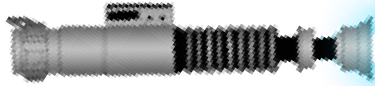
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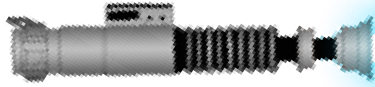


Introduction



- Patients with idiopathic intracranial hypertension (IIH) are more often obese women with normal or slit ventricles.
- IIH is often correlated with an extracranial increase of venous pressure (venous sinuses thrombosis, obesity...)
- high pressure hydrocephalus is acute or subacute and often has an intracranial etiology (like bleeding, infection, trauma)
- Here we present a unique and previously undescribed case that could represent a new nosological entity

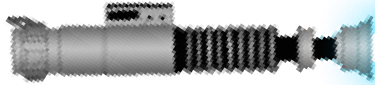
Case presentation: history



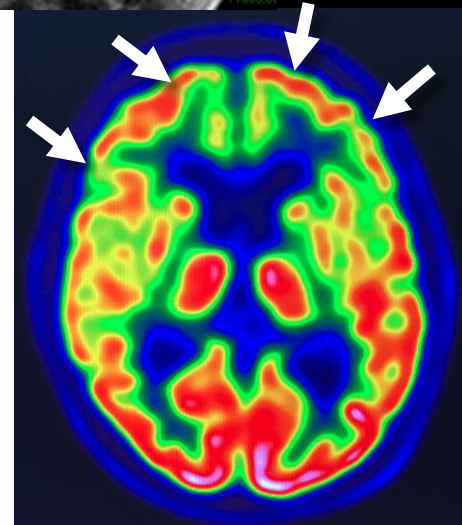
- June 2016: A 63 year old former nurse with clinical and radiological signs of intracranial hypertension.
- Symptoms and signs: headache, pulse synchronous tinnitus, grade 4 Frisen Papilledema and a mild cognitive slowing.



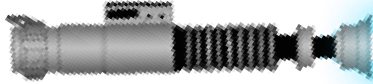
Case presentation: imaging



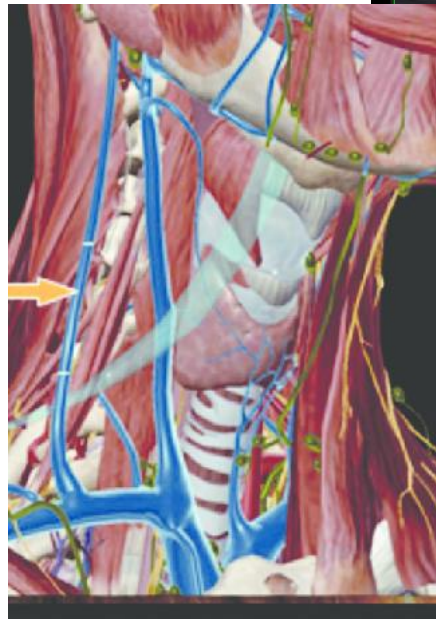
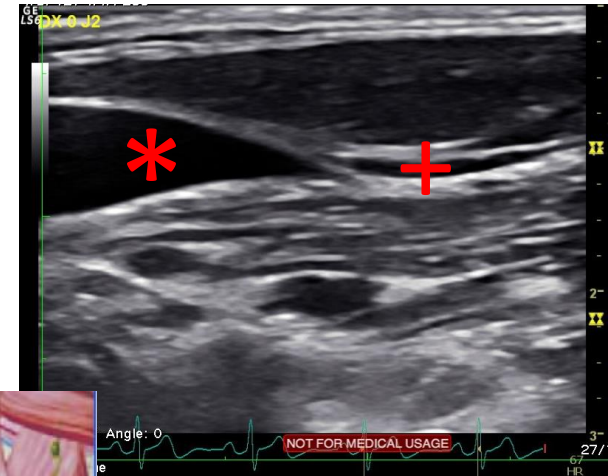
- Brain MRI: increase of brain ventricles (Evans Index 0.36).
- Brain MRI venography excluded thrombosis and revealed an apparent turgor of brain cortical veins.
- FDG-PET: diffuse hypomethabolism.



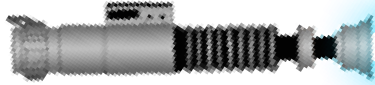
Case presentation: ultrasound



- Intra and extracranial Doppler ultrasound showed a bilateral external compression of the omohyoid muscle on the internal jugular veins: this corresponded to blocked flow.



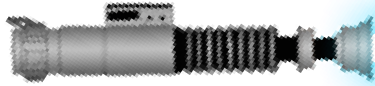
Treatment: VPS or ETV



But.....patient refused to be “neuro”-treated



Multidisciplinary team



Dept. Neurosurgery: Pasquale De Bonis

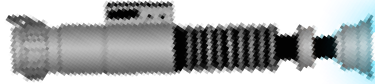
Dept. Traslational Surgery: Paolo Zamboni

Dept. Vascular Surgery: Francesco Mascoli

Dept. Physics: Francesco Sisini

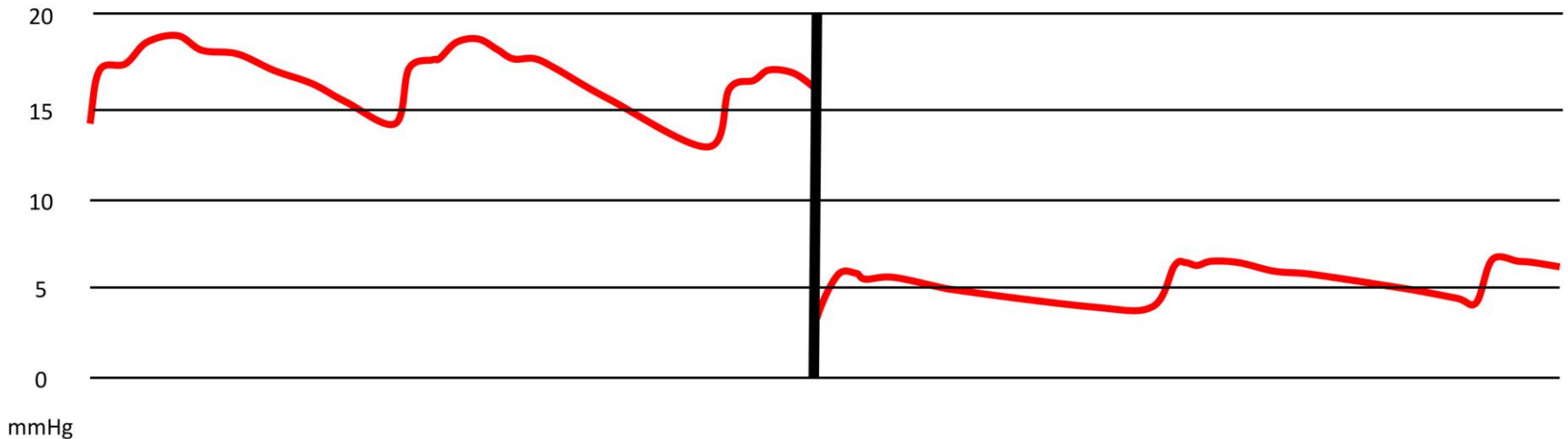
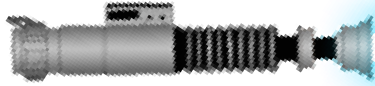


New Treatment modality



- We therefore decided for an “etiological” therapy.
- Right frontal EVD: baseline intracranial 16-18mmHg, with a pathological waveform.
- While on intracranial pressure continuous monitoring, a bilateral omohyoid muscles transection was therefore performed.

New Treatment modality



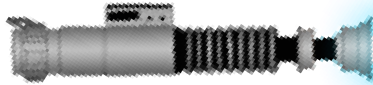
Baseline ICP (left): 16-18mmHg with pathological wave.

ICP After omohyoid muscle transection (right): 6mmHg.

Green line is set at 15mmHg.

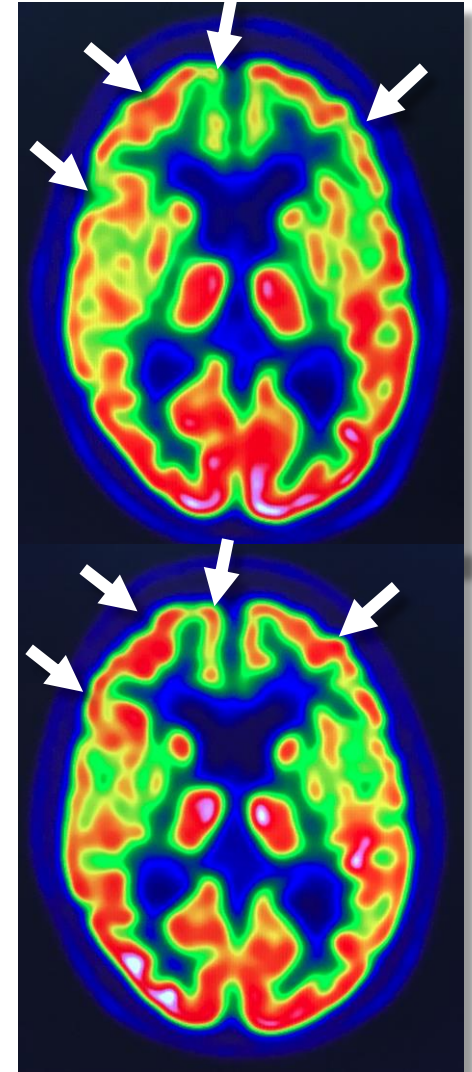
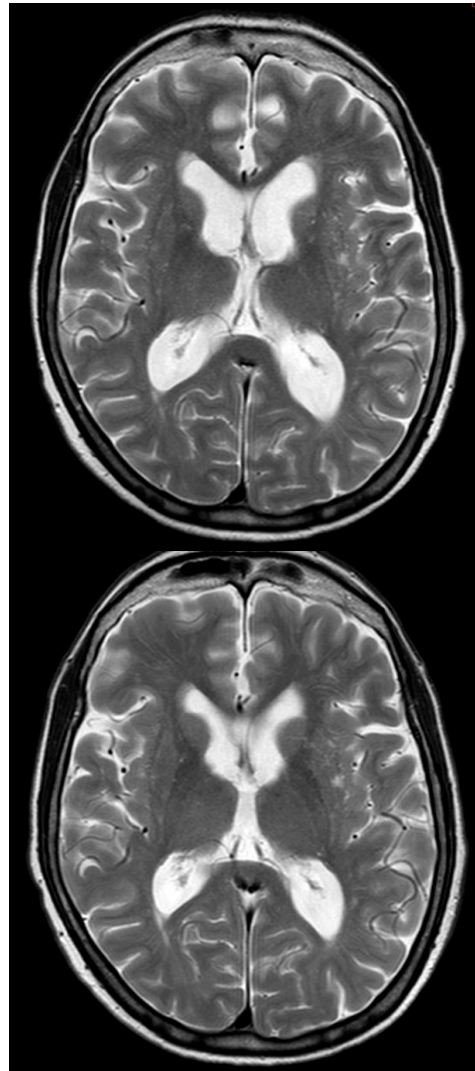
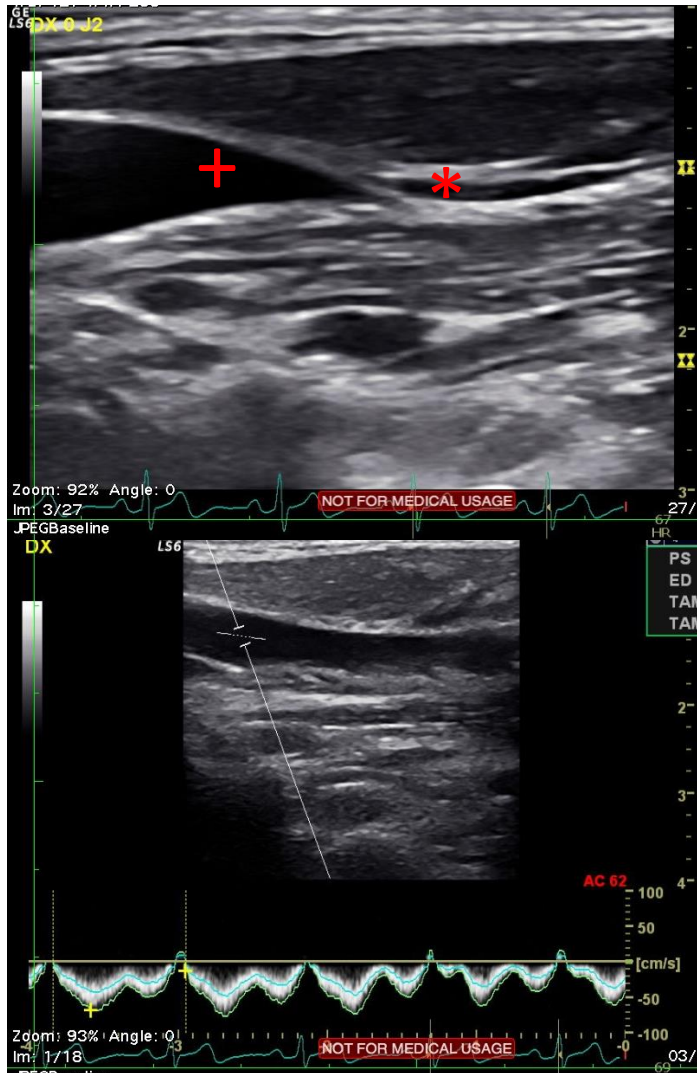
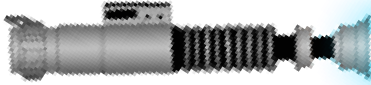
The right frontal catheter was therefore removed.

Outcome

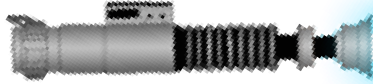


- Headache and tinnitus disappeared soon after surgery.
- Visual acuity improved, fundoscopic examination (12 and 24-month follow-up): no papilledema.
- MRI (6, 12 and 24 month): decreased ventricles (Evans Index 0.31) with more evident convexity CSF spaces.
- PET: improved of brain metabolism

Outcome



Conclusion: a new syndrome



- Our patient presented
 - Jugular **E**ntrapment
 - **D**ilated ventricles (Hydrocephalus)
 - **I**ntracranial hypertension
- We called this syndrome **JEDI**: these findings can represent a new syndrome, a novel form of high pressure hydrocephalus that can be treated without a CSF shunt.

Your attention
thank you for!





JEDI (jugular entrapment, dilated ventricles, intracranial hypertension) syndrome: a new clinical entity? A case report

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Abstract

Patients with idiopathic intracranial hypertension are frequently obese women with normal/slit ventricles. Patients with high-pressure hydrocephalus, instead, present enlarged ventricles. We describe a 63-year-old woman with signs and symptoms of intracranial hypertension. Brain MRI revealed hydrocephalus. Venous Doppler ultrasound showed external compression of the omohyoid muscles on the internal jugular veins. During jugular vein decompression, intracranial pressure dropped from 18 to 6 mmHg. Patient is asymptomatic at 2-year follow-up, with decreased brain ventricles. These findings could represent a novel form of high-pressure hydrocephalus that can be successfully treated without a CSF shunt. We called this syndrome JEDI (jugular entrapment dilated ventricles intracranial hypertension).

Keywords Intracranial hypertension · Hydrocephalus · Jugular veins