When the Main Vein from the Brain Won’t Drain, Rooke thinks it Might Cause Pain … … Is he Insane?

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The head and neck can be affected, too … including intracranial … Cerebral Sinus Thrombosis

Cerebral Venous Sinus Thrombosis (CVST)
Posted by Rathachai Kaewlai, M.D

Why not still more effects? First in 2008
J Neurol Neurosurg Psychiatry 2009;80:392-399
Chronic cerebrospinal venous insufficiency in patients with multiple sclerosis
P Zamboni, et. al.

Venous obstruction / reflux has predictable effects …

Renal vein (“nutcracker”) with gonadal vein reflux

The head and neck can be affected, too …

Other effects
Transient global amnesia (TGA) is one of the most striking syndromes in clinical neurology … Studies with … venous duplex … confirmed the importance of jugular-vein-valve insufficiency.

… headache probably results from stretching of the … veins that drain into the sinuses, although elevated ICP may also play a role. In addition to headache, clouding of consciousness and seizures are common …

Why not still more effects?
http://missinglink.ucsf.edu/lm/ids_104_demyelination/didactic/msmicropath.htm

http://www.nature.com/neuro/journal/v3/n6/fig_tab/nn2138_F1.html


THE LANCET Neurology

http://missinglink.ucsf.edu/lm/ids_104_demyelination/didactic/msmicropath.htm
Multiple Sclerosis

Journal of Cerebral Blood Flow & Metabolism (2009) 29, 1587–1596. 3 September 2009
Anomalous venous blood flow and iron deposition in multiple sclerosis
Ajay Vikram Singh and Paolo Zamboni

Venous Ulcer


Chronic Cerebro-Spinal Venous Insufficiency (CCSVI)

Venous Ultrasound

... venous outflow anomalies were dramatically associated (with MS) (OR 43, p<0.0001).

“... MS is strongly associated with CCSVI, a scenario that has not previously been described...”

Focus on Obstruction

The “Liberation Procedure”

“... CCSVI endovascular treatment significantly improved MS clinical outcome measures ... the rate of relapse-free patients changed from 27% to 50% postoperatively ...”

“The results of this pilot study (65 pts) warrant a subsequent randomized control study.”


Meta-Analysis of the Correlation Between Chronic Cerebrospinal Venous Insufficiency and Multiple Sclerosis

Vascular and Endovascular Surgery 47(6) 620-624

2013

Odds Ratio = 2.6

This triggered an effusive world-wide response. For example ...

Cardiovascular and Interventional Radiological Society of Europe Commentary on the Treatment of Chronic Cerebrospinal Venous Insufficiency


Cardiovascular and Interventional Radiology

“... there is increasing worldwide acceptance of CCSVI ... even though there is no supporting scientific evidence. ... most of the information we have comes from one source only.
The treatment is called “liberation treatment,” and the results of the treatment can be watched on YouTube. The current forum for the reporting of success ... seems to be the Internet.
There are ... testimonies by MS patients who have gained improvement ... (but not) ...from patients who underwent unsuccessful treatments ..."
PLoS One. 2013;8(2) Italy
Chronic cerebrospinal venous insufficiency is not associated with MS and its severity: a blind-verified study.

Neurology. 2011 Aug 30;77(9):844-50. Italy
Progressive MS is not associated with chronic cerebrospinal venous insufficiency.

... CCSVI is not associated with disability.

BMC Med. 2013 Jul 18;11:167 Buffalo, NY
Chronic cerebrospinal venous insufficiency is not associated with MS and its severity: a blind-verified study.

... CCSVI was not associated with MS... nor its severity.

We find no evidence of an association between the presence and severity of CCSVI in patients with MS.

Neurology. 2013 Aug 30;77(9):844-50. Italy
Chronic cerebrospinal venous insufficiency is not associated with cognitive impairment in MS.

We find no evidence of an association between the presence and severity of CCSVI in patients with MS.

Mult Scler. October 2013 vol. 19 no. 11 1508-1517 multicenter Italy
Observational case-control study of the prevalence of chronic cerebrospinal venous insufficiency in multiple sclerosis: results from the CoSMo study

1874 subjects aged 18–55.
Color...duplex sonography

The primary (goal) is to compare the prevalence of CCSVI in patients with MS versus patients affected by other neurodegenerative diseases (OND) and healthy volunteers.

Conclusions: CCSVI is not associated with MS.

Prevalence of extracranial venous narrowing on catheter venography in people with multiple sclerosis, their siblings, and unrelated healthy controls: a blinded, case-control study.

Extracranial venous narrowing > 50% is a frequent finding in patients with MS... (and) controls.

The significance of venous narrowing to MS symptomatology remains unknown...
But ... could venous insufficiency (obstruction) produce other (non-MS) neurological/head symptoms?

What about?:
- Headache
- Visual changes
- Speech difficulties
- Imbalance
- Cognitive issues
- Elevated IC pressure
- Others

These are Very Common Problems

# of migraine sufferers ... in the US = 12 million


Prevalence of tension-type headache = 78%.


How often does venous impairment play a role?

Mechanism of “Venous” HA?

Intracranial blood flow increases

If the venous blockage is not “severe” ...

More on this later ...

#1 Problem — How do we determine if a patient has CCSVI?

Very Complicated

Simpler?

Validity of the diagnostic criteria for chronic cerebrospinal venous insufficiency and association with multiple sclerosis

CMAJ June 2, 2014

120 patients with multiple sclerosis and 60 healthy controls.

“... We detected no differences in the proportion of venous outflow abnormalities between patients with multiple sclerosis and healthy controls.”

“Perhaps the most striking finding in our study was the range of venous outflow anomalies detected that did not reflect pathology, but instead demonstrated the large natural variance in intracranial and extra-cranial venous anatomy ...”

variants of “normal” causing venous compression (often asymptomatic) are common

Popliteal vein entrapment

Normal?

Thoracic outlet syndrome
Movement of the neck is much more complicated.

FINDINGS: “MR venography the cervical and intracranial vasculature is negative. The dural venous sinuses are widely patent. No evidence of obstruction of the internal jugular veins in the neck.”

Case #1

“Pathology?” Or just a “Variant?”

Dilemma

Collateral vein? Or congenital variant?

Is this congenitally “small” transverse sinus “too small?” Is flow obstructed?

Decisions based entirely on anatomical appearance +/- catheter-assessed pressure gradient

• Stenosis?
• Compression?
• Normal variant?

Case #2

78 year-old woman with chronic, severe headache

“No Dx” “Nothing to do”

Gradient 2-3 mmHg

Dilatation of jugular stenosis X 3 sessions. Each helped ... for 3-6 weeks. Then symptoms – and stenosis – returned.

Finally treated with stent. Complete resolution of symptoms. No recurrence.

Case #3

38 year old male. Chronic visual “fatigue,” headache, “brain fog.”

Did we miss something?

Stenosis

Collaterals

MRV findings may have been positional? Or “overcall?”

Case #4

68 year-old man. DVT (Factor V positive). Subdural hematoma on Coumadin.

Hematoma evacuated X 2. Still unstable.

Bleeding

Shift

Small transverse sinus. Possible obstruction?
Dominant jugular is occluded?

Do you attempt to open the jugular?

What we need is a “functional test ...”

Ultrasound seems like a good place to start. It’s the basis for many other hemodynamic / functional vascular tests ...

But can we see the area of interest with Duplex?

“Normal”

How to Test?

- Increase inflow (vasodilator)
- Outflow obstructed (Limits overall flow)

This produces:

- Symptoms?
- Increased venous velocity
- Increased ICP
- Brain “stiffness”?
- EEG / PET scan?
- Other changes?
78 year old female
Worsening “myoclonus”

(Most often, myoclonus occurs as a result of a nervous system (neurological) disorder, such as epilepsy, or of a metabolic condition, or as a reaction to a medication.

The cause of essential myoclonus is often unexplained (idiopathic) or, in some cases, hereditary.)

http://www.mayoclinic.org/diseases-conditions/myoclonus/basics/definition/con-20027364

**Case Example?**

Work-up = negative
Attempted dobutamine echo (to assess possible CAD) – stopped because of severe exacerbation of myoclonus.

Does she have physiologically significant venous obstruction?
Is it contributing to her myoclonus?

**Chronic (Cervical) Venous Occlusive Disease:**

- Probably exists and is under-recognized
- Does not appear to cause MS?
- May cause HA and other CNS symptoms

However ...

- Diagnostic criteria are lacking (And thus all the controversy)
- There is a need for better anatomic / hemodynamic / functional testing