Imaging and Diagnosis in Chronic Cerebrospinal Venous Insufficiency

Nii-Kabu Kabutey[1], Ducksoo Kim[1], Sandy McDonald[2], Angela Lagace[2], Michael Stehling[1], Erica Menegatti[3], Paolo Zamboni[3]

[1]Boston University Medical Center, Dept. of Radiology Boston, MA. [2]Barrie Vascular Imaging, Barrie, ON. [3]University of Ferrara, Vascular Diseases Center, Ferrara, It

Chronic Cerebrospinal Venous Insufficiency

Chronic cerebrospinal venous insufficiency (CCSVI) is characterized by an impediment to the major extracranial venous outflow pathways, resulting in venous reflux and loss of postural regulation of cerebral venous outflow. The dynamic nature of extracranial cerebrospinal venous drainage due to the influence of respiratory mechanics and postural positioning, produce particularly difficult imaging challenges. In addition, the venous system is often asymmetric and more variable than arterial anatomy. The bony structure of the cranium and thoracic vault increase the difficulty of assessing the hemodynamic variability of the venous system with doppler ultrasound imaging alone.

In 1935, Dr. Tracy Putman of Boston City Hospital first proposed a relationship between venous obstruction and the formation of cerebral sclerotic plaques seen in multiple sclerosis (MS).[1] A strong association has been noted between CCSVI and multiple sclerosis (MS) a debilitating inflammatory condition of the central nervous system characterized by immune cell migration across the blood brain barrier and subsequent nerve sheath demyelination[2]. Multiple imaging modalites are currently being studied to diagnosis CCSVI and further delineate the relationship between CCSVI and multiple sclerosis.

Ultrasound Criteria for CCSVI Diagnosis

High resolution color Doppler ultrasound is a non-invasive method that has been used to assess five hemodynamic properties of CCSVI, which are absent in normal subjects[2]. At least two of five parameters must be present to diagnosis CCSVI.

1. B-Mode Structural Abnormalities
2. Failure of the internal jugular vein to dilate in the supine position compared to the erect position
3. Reflux in the internal jugular or vertebral veins
4. Obstructed flow in the internal jugular or vertebral veins
5. Reflux in the deep cerebral veins

The patient is examined in the supine and upright position. A linear 3.5-10 MHz probe is used for scanning the extracranial veins of the neck, a second transcranial 2.0-3.0 MHz probe is used for the interrogation of the deep cerebral veins.

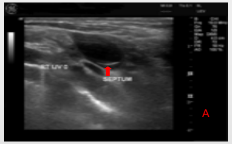
Ultrasound: B Mode Ultrasound Structural Abnormalities

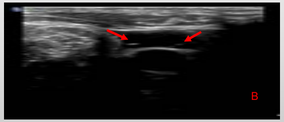
High resolution B-Mode ultrasound evidence of proximal internal jugular vein stenosis was witnessed in 37% of multiple sclerosis patients compared to 0.6% in a control population[2].

Figure A- Transverse ultrasonography demonstrating a valve leaflet (arrow) that has created an internal septum from the anterior to posterior wall of the jugular vein.

Figure B- Malaligned valve leaflets (arrows)

Figure C- Prominent echogenic abnormal valve leaflet (arrow)

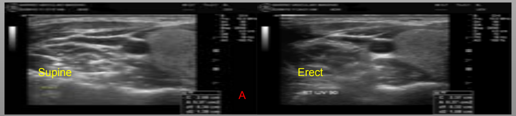


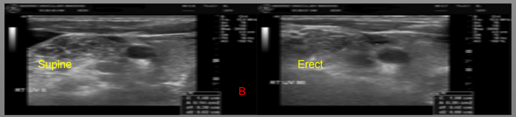


Ultrasound: Reverted Postural Control

The internal jugular vein (IJV) is the predominant extracranial venous drainage pathway in the supine position, this can be demonstrated by an increased cross sectional area (CSA) related to increased blood volume. Subtraction of the IJV CSA in the erect position from the IJV CSA in the supine position produces a positive value in normal subjects.

Figure A- ΔCSA in normal subject = CSA_{supine} 0.37 - CSA_{erect} 0.27 = 0.10
Figure B- ΔCSA in MS patient = CSA_{supine} 0.14 - CSA_{erect} 0.26 = -0.12





Ultrasound: Reflux in the Internal Jugular and Vertebral Veins

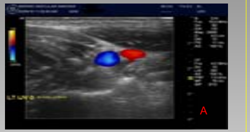
In normal subjects, flow within the IJV and vertebral veins (VV) is directed towards the heart, in both the supine and erect position. Abnormal reflux is considered a reversal of the flow from its physiological direction lasting longer than 1.5 s.

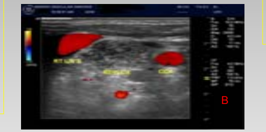
Figure A – Normal flow pattern with the direction of flow within the carotid artery and internal jugular vein in opposite directions.

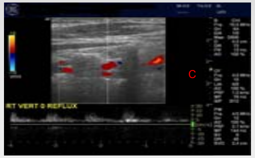
Figure B - Prolonged reflux noted in the internal jugular vein. The direction of flow in the IJV is noted to be in the same direction as the common carotid artery (CCA).

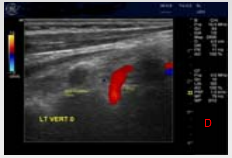
Figure C – Vertebral vein and artery displayed with spectral Doppler indicating flow in the same direction. Sustained reflux within the vertebral vein pattern is suggestive of azygous vein obstruction.

Figure D - Reflux of the intra-rachidian veins which run outside the vertebral column.







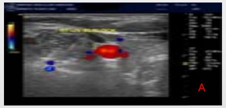


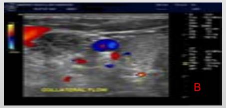
Ultrasound: Obstructed Flow in the Internal Jugular and Vertebral Veins

In approximately 52% of multiple sclerosis patients scanned Zamboni et al. were unable to detect flow in an IJV or VV despite numerous deep inspirations[2].

Figure A – No flow detected in the internal jugular vein indicative of obstruction.

Figure B - Excessive collateral patterns visualized near the thyroid gland and vertebral regions may also be indicative of IJV obstruction.





Transcranial Doppler Evaluation

Flow within the deep cerebral veins should be monodirectional. A transcranial color Doppler is used to assess the flow in at least one of the deep cerebral veins (great vein of Galen, basal vein of Rosenthal, internal cerebral vein).
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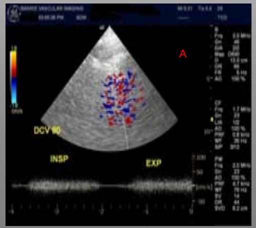
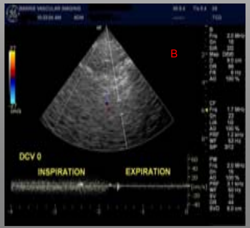
Figure A – Flow in the deep cerebral veins with inspiration and expiration.

Figure B – Bidirectional or no flow detected on expiration.



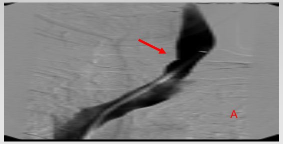


Internal Jugular Venography

Selective invasive venography of the extracranial outflow circulation has demonstrated significant stenotic lesions within the cervical and thoracic levels of venous drainage. IJV lesions were noted unilaterally or bilaterally in over 90% of patients with multiple sclerosis. Venous pathology frequently noted upon venography included; venous twisting, septum formation, venous atresia, valvular anomalies and bone compression. A small control population of normal patient did not exhibit venous pathology.

Figure A – Left IJV venography performed from a femoral approach. Digital subtraction image demonstrates a venous stenosis (arrow) within the proximal vein at the level of the clavicle. Flow through a dilated inferior thyroid vein is noted

Figure B - Balloon angioplasty at the level of the clavicle demonstrates a persistent circumferential waisting of the balloon highly suggestive of a restrictive lesion.



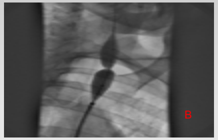
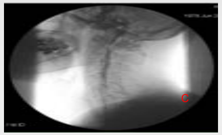
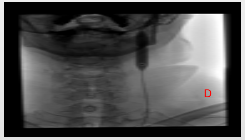


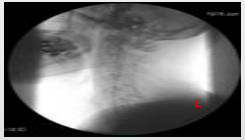
Figure C – Venography demonstrating a greater than 50% stenosis of the distal IJV.

Figure D – Balloon angioplasty of the lesion showing Persistent narrowing at the level of the mandible.

Figure E – Self-expanding stent placement within the Distal IJV





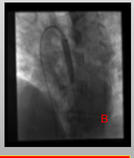


Azygous Venography

In patients with multiple sclerosis azygous vein stenosis was present in 86% of cases.

Figure A – Venogram of the azygous vein with mild proximal stenosis (arrow).

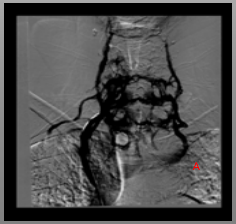
Figure B – Balloon angioplasty of the azygous vein lesion.



Vertebral Venography

The vertebral venous anatomy is a complex valveless system compromised of multiple segmental intercommunicating systems. In the erect position the vertebral veins are the primary outflow tract. The system communicates with the deep thoracic veins, lumbar veins, intercostal veins and the azygous vein. Stenosis within the azygous vein may precipate reflux and venous hypertension within the intraspinal circulation[3].

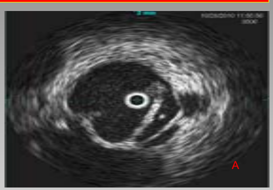
Figure A - Contrast venography demonstrating complex intercommunication of the vertebral venous system.

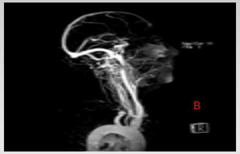


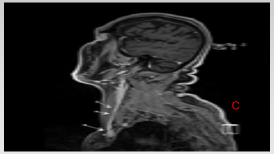
Future Direction of CCSVI Imaging

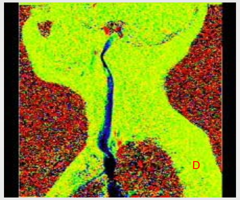
Both ultrasonography and venography have inherent imaging limitations. Alternative imaging modalities may help to further delineate the physiologic and structural abnormalities of CCSVI. Intravascular ultrasound may provide further information regarding valvular dysfunction, stenosis and obstruction.

Figure A – Intravascular ultrasound image within the inferior vena cava showing a lobulated vessel with intraluminal septum (arrow).









MR imaging allows for 3D structural visualization of the head, neck and azygous vein anatomy. MR venography has the added benefit of being able to see the extensive venous collateralization patterns. Flow calculations of the extracranial cerebral circulation can also be performed.

Figure B -3D MIP imaging demonstrating grade stenosis of the upper segment IJV (arrow).

Figure C – Sagittal MR venogram post gadolinium showing diffuse stenosis of the lower segment of the left IJV (arrows) .

Figure D - MR venogram showing with color coding demonstrating focal stenosis of the upper IJV.

Figure A – Sagittal plane MRV using both respiratory and cardiac gating techniques to clearly visualize ayzgous vein (arrow) anatomy.

Conclusion

There is an incomplete understanding of the anatomic variability of the extracranial venous drainage system, and how subtle venous malformations may relate to pathologic conditions. The proposed correlation between CCSVI and MS has gained worldwide momentum within the MS community. Despite a paucity of clinical trials MS patients have been seeking imaging and treatment options with balloon angioplasty and stenting. Further research is required to fully understand the cerebrospinal venous pathology that has been noted in patients with MS.

The diagnosis of CCSVI is entirely based on imaging characteristics. It is imperative that radiologist take a leading role in understanding the physiological and anatomical relationships of the extracranial venous circulation.

Reference

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