Diagnosis and Treatment of Chronic Cerebrospinal Venous Insufficiency in Multiple Sclerosis

Preauthorization is not required but is recommended if, despite this Protocol position, the physician feels this service is medically necessary.

The following Protocol contains medical necessity criteria that apply for this service. The criteria are also applicable to services provided in the local Medicare Advantage operating area for those members, unless separate Medicare Advantage criteria are indicated. If the criteria are not met, reimbursement will be denied and the patient cannot be billed. Please note that payment for covered services is subject to eligibility and the limitations noted in the patient’s contract at the time the services are rendered.

Description
Chronic cerebrospinal venous insufficiency (CCSVI) may be associated with multiple sclerosis (MS), although this is controversial and an active area of research. Correction of CCSVI has been attempted via percutaneous venoplasty. The intent of this procedure is to relieve MS symptoms by improving venous drainage of the central nervous system. Correction of CCSVI by this method may be referred to as the “Liberation Procedure.”

Summary of Evidence
The association of chronic cerebrospinal venous insufficiency (CCSVI) with multiple sclerosis (MS) is uncertain. The rate of CCSVI in MS patients varies widely in the literature for unclear reasons, from 0% to 100%. Some studies report higher rates of CCSVI in patients with MS compared with non-MS patients, but others do not. If there is an association between MS and CCSVI, it is not known whether this is a causative factor for MS or a secondary result of the disease. It also appears that CCSVI can occur in other disorders and is not specific for MS.

Treatment of CCSVI with endovascular interventions has been attempted. Some currently available studies report improvement in patient-reported symptoms following treatment, but this evidence is not sufficient to establish efficacy. A prospective, double-blind, sham-controlled randomized controlled trial (RCT) of venous angioplasty in MS patients (N=20) with CCSVI published in 2014 showed no significant differences in venous outflow characteristics between the treated and control groups, nor any significant improvements in clinical disease scores among treated patients compared with controls. The results of this RCT are limited by the small number of patients. However, the failure to show a beneficial effect of venous angioplasty on blood flow or symptoms supports a lack of efficacy for this treatment.

Adverse events occur at a low overall rate, but serious adverse events can occur, and the U.S. Food and Drug Administration issued an alert in 2012 concerning the potential for serious adverse events with treatment of CCSVI.

Policy
The identification and subsequent treatment of chronic cerebrospinal venous insufficiency (CCSVI) in patients with multiple sclerosis is considered not medically necessary.
Background

MS is generally considered a chronic inflammatory demyelinating disease of the central nervous system (brain, spinal cord, optic nerve) believed to be triggered by an autoimmune response to myelin. However, in part due to the periventricular predilection of the lesions of MS, vascular etiologies (CCSVI) have also been considered. The core foundation of this vascular theory is that venous drainage from the brain is abnormal due to outflow obstruction in the draining jugular vein and/or azygos veins. This abnormal venous drainage, which is characterized by special ultrasound criteria, is said to cause intracerebral flow disturbance or outflow problems that lead to periventricular deposits. In the CCSVI theory, these deposits have a similarity to the iron deposits seen around the veins in the legs of patients with chronic deep vein thrombosis. Balloon dilatation, with or without stenting, has been proposed as a means to treat the outflow problems, thereby alleviating CCSVI and MS complaints.

The following five criteria were defined by Zamboni et al as features of CCSVI.¹ To make the diagnosis of CCSVI, at least two of the five criteria need to be present:

1. Reflux constantly present (for a duration greater than 0.8 s) in the supine and upright positions at the level of an internal jugular or vertebral vein. This parameter was evaluated during a short breath-hold following normal breathing and not under Valsalva maneuver.
2. Reflux at the level of veins of the deep cerebral system (for a duration greater than 0.5 s). This was evaluated with the patient in the sitting and supine positions, and venous flow was enhanced by inviting the patient to breath in.
3. Stenosis (less than 0.3 cm), valve abnormalities and septa on B-mode imaging.
4. Absence of flow at the level of the internal jugular or vertebral vein, despite numerous deep inspirations.
5. No increase in the diameter of the internal jugular vein when changing from an upright to a supine position (lack of Δ-).

Regulatory Status

Endovascular correction of CCSVI is a surgical procedure and as such is not subject to FDA approval. However, in 2012, FDA issued an alert concerning the potential for serious adverse events with the treatment for CCSVI.

Services that are the subject of a clinical trial do not meet our Technology Assessment Protocol criteria and are considered investigational. For explanation of experimental and investigational, please refer to the Technology Assessment Protocol.

It is expected that only appropriate and medically necessary services will be rendered. We reserve the right to conduct prepayment and postpayment reviews to assess the medical appropriateness of the above-referenced procedures. Some of this Protocol may not pertain to the patients you provide care to, as it may relate to products that are not available in your geographic area.

References

We are not responsible for the continuing viability of web site addresses that may be listed in any references below.


