

Calcifications of Cerebral Medullary Veins Associated with Carotid-Jugular Fistula

Wen C. Yang, M.D.¹
Jay Castle, M.D.²
Harvey Lozman, M.D.³
Moses Nussbaum, M.D.³

Departments of Radiology,¹ Neurology,²
and Surgery,³ Beth Israel Medical Center,
New York, New York

Unusual calcification of the cerebral medullary veins is noted in a 34-year-old mentally retarded female who had a right carotid-jugular anastomosis 30 years earlier. The patient was hospitalized because of a right neck mass, proptosis, and the recent onset of left flaccid hemiplegia. Arch aortography and CT of the head and neck were performed. Surgery to oblate the fistula provided dramatic relief of venous engorgement in the orbits and face. The finding of medullary vein calcification is emphasized because it is a hitherto unreported entity.

Calcifications have always generated great interest in radiology because they are easily demonstrable on roentgenograms. In many disease processes calcifications exhibit characteristic features that are essential in diagnostic analysis. This is particularly true in intracranial calcified lesions. In the past few decades, numerous reports have been published to provide better understanding of almost every conceivable type of intracranial calcification (1). However, with the advent of computed tomography (CT), a new perspective for further discovery has evolved.

Recently we had the opportunity to use CT to study a patient with a long-standing carotid-jugular fistula. A new entity—medullary vein calcification—was discovered and is detailed in this paper.

CASE REPORT

A 34-year-old female was admitted to the Beth Israel Medical Center in October, 1980, for evaluation of a large neck mass, facial disfiguration, exophthalmos, and cerebrovascular accident. The patient had been

Address reprint requests to Wen C. Yang, M.D., Department of Radiology, Beth Israel Medical Center, 10 Nathan D. Perlman Place, New York, NY 10003.

mentally retarded since birth. At the age of 4 years she underwent a right common carotid artery-internal jugular vein anastomosis, a procedure then believed to improve the cerebral circulation in mentally retarded individuals. There was no apparent improvement of her mental status.

Six years prior to this admission swelling of the right side of the neck and right eyelid developed and progressed to a state of facial disfiguration and exophthalmos. One month prior to admission, she developed a left hemiplegia.

Examination on admission revealed a large pulsatile right neck mass with an audible bruit. Vascular engorgement and a pulsatile thrill were noted in both upper eyelids, the right more than the left. There was entropion of the lower eyelids with trichiasis. The right conjunctiva and sclera were congested. The retinae and maculae were normal. Tests for extraocular muscles revealed possible right VIth cranial nerve palsy. There was flaccid paralysis of the left extremities.

Arch aortography revealed a right common carotid artery-internal jugular vein fistula with marked dilatation of the internal jugular vein, which accounted for the neck mass (Figure 1). There was engorgement and early filling of the lateral, straight, and cavernous sinuses as well as the orbital and internal cerebral veins.

CT of the head and neck demonstrated the enlarged venous channels. The orbital veins on the right were more dilated than those on the left. There was no CT evidence of infarct, but striking calcifications of the deep medullary veins were noted, predominantly in the right cerebral hemisphere (Figures 2, 3).

The patient underwent corrective surgery to ablate the arteriovenous fistula. Postoperatively there was dramatic relief of the venous engorgement of the orbits and face.

DISCUSSION

In 1949 Beck et al. (2, 3) introduced a surgical procedure with which they hoped to improve the cerebral

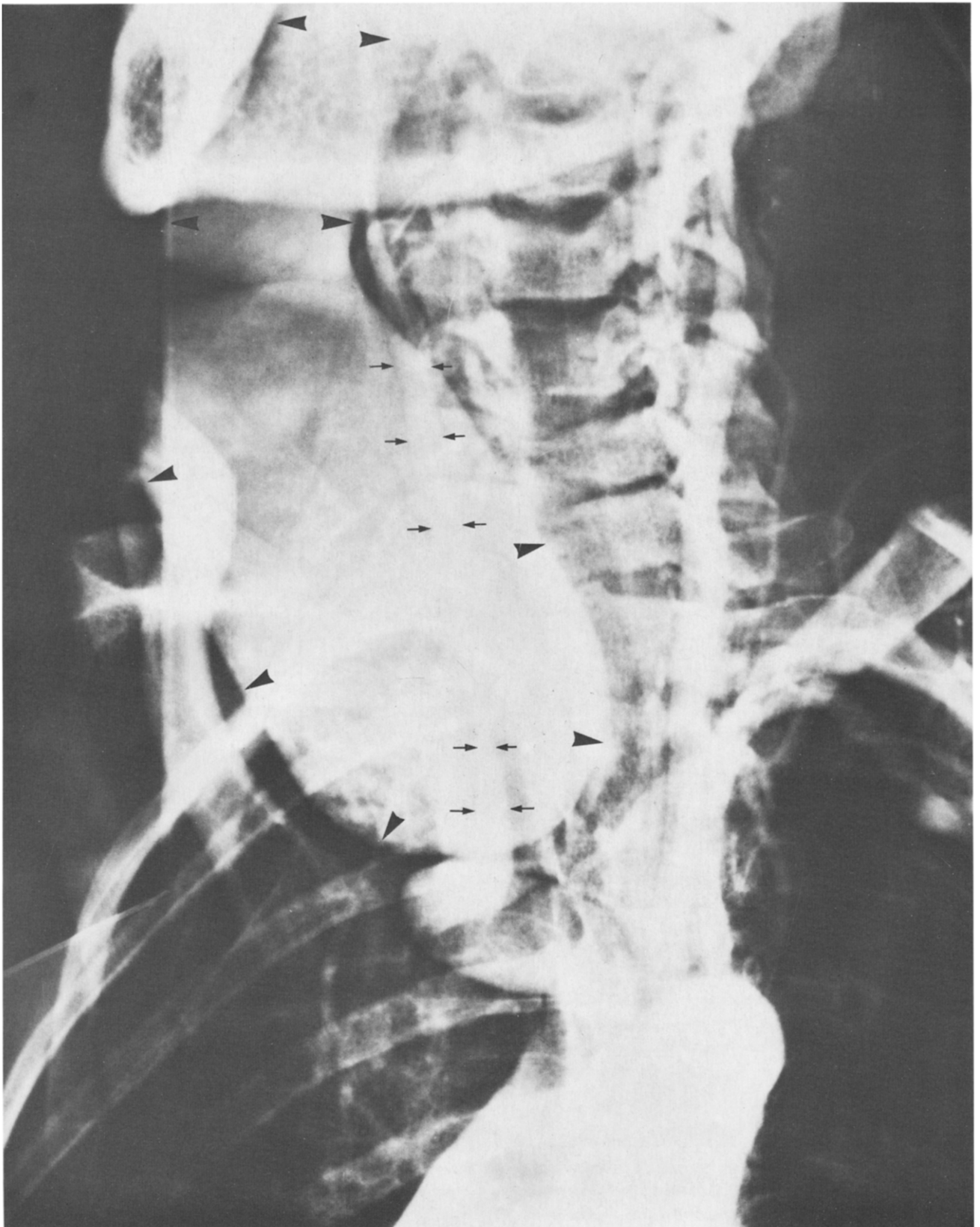


Figure 1. Arch aortogram. Note the marked enlargement of the right internal jugular vein (*large arrowheads*) with a blind pouch inferiorly giving a large sausagelike appearance. The anastomotic site is at the middle segment of the common carotid artery (*arrows*).

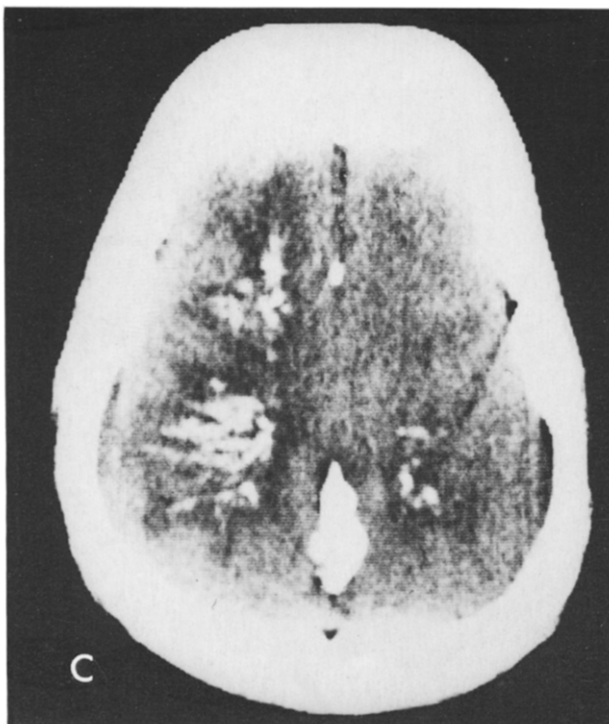
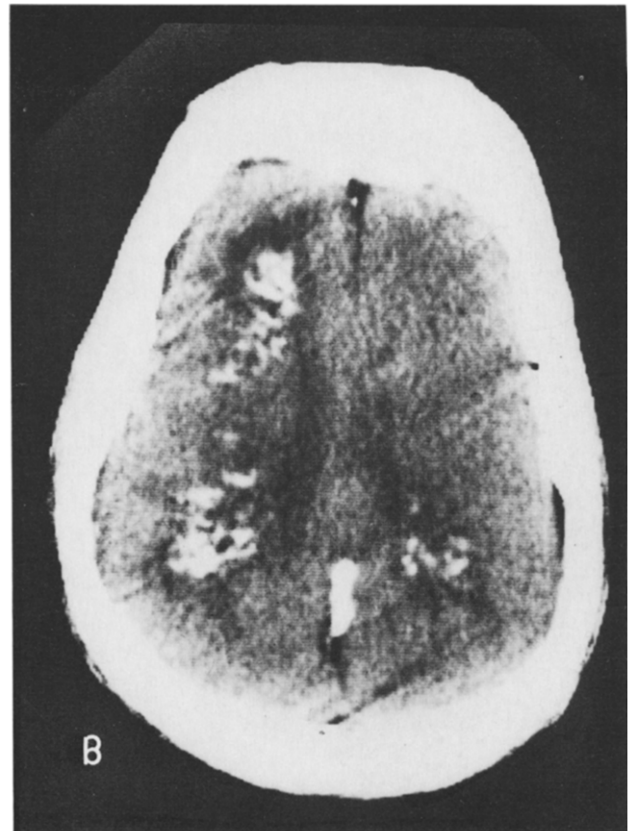
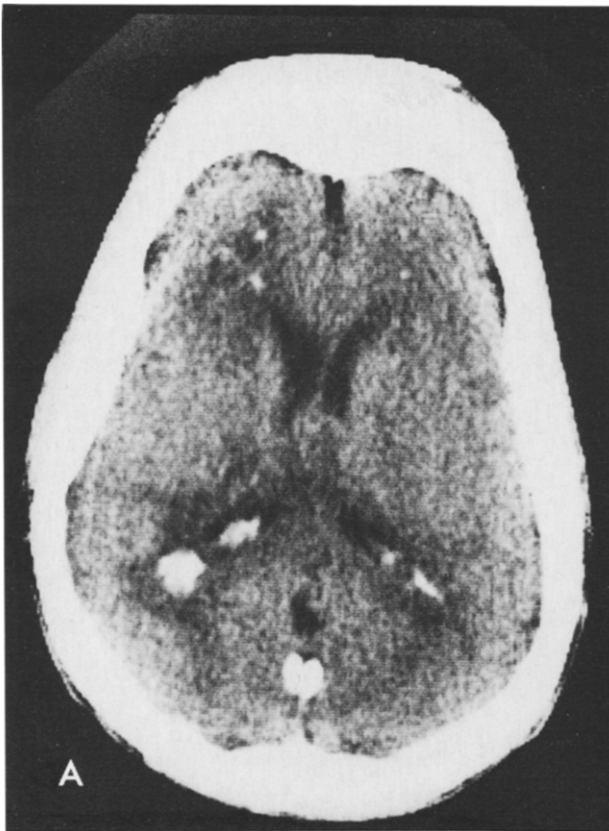
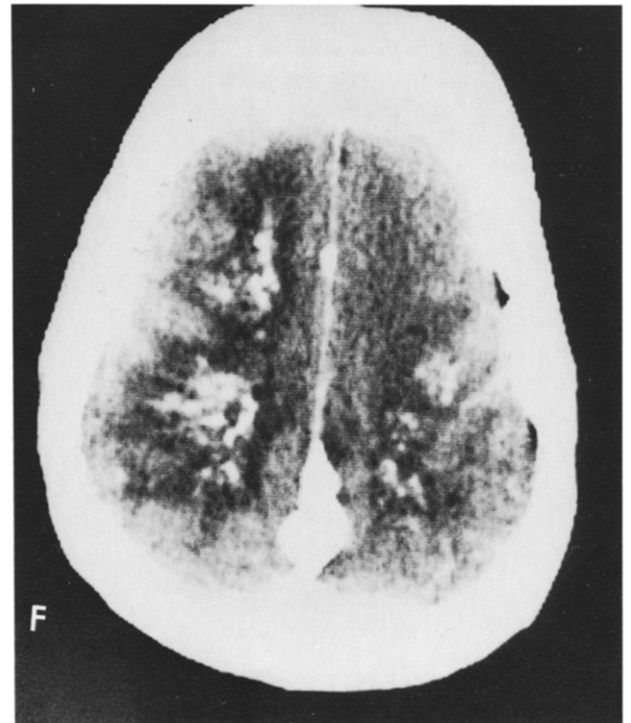
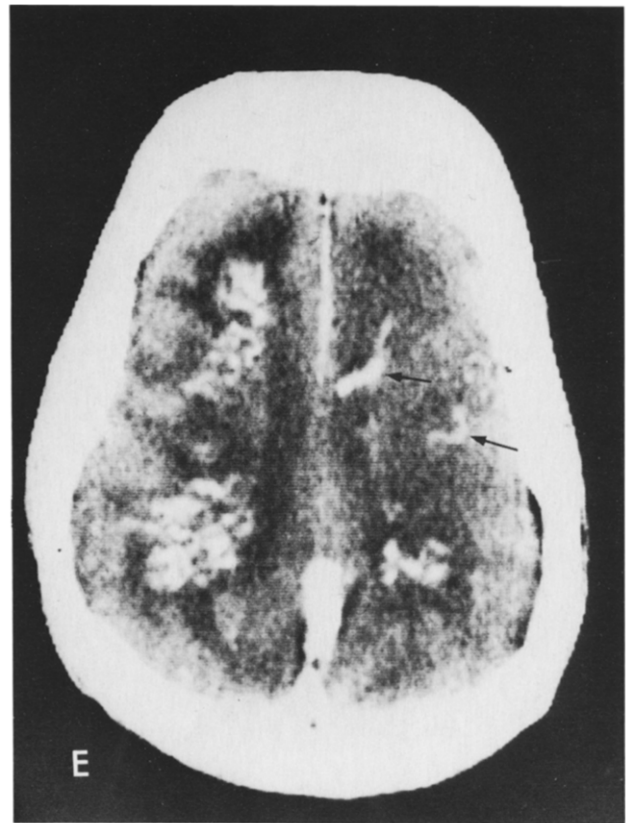
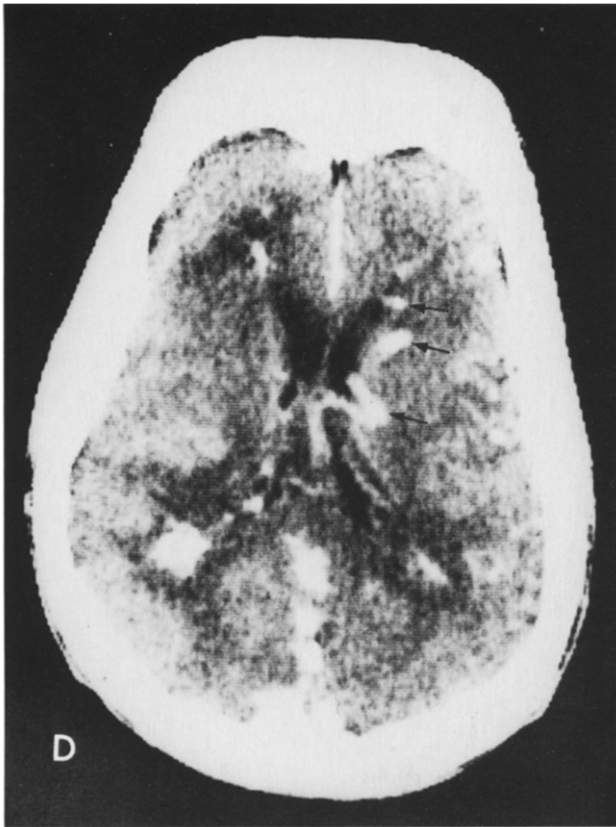


Figure 2 A, B, & C. Precontrast CT scans reveal linear calcifications of various lengths and widths in the white matter of cerebral hemispheres (more on the right) converging toward the lateral angles of the lateral ventricles.

circulation for mentally retarded or epileptic patients by establishing a fistula between the common carotid artery and the internal jugular vein. Several years later accumulated experience led to the conclusion that the carotid-jugular anastomosis lacked any therapeutic value and, when remaining open, created complications (4-8). The procedure was therefore abandoned.

Aside from the venous engorgement in the neck and orbit as presented clinically, a new finding of unusual cerebral calcifications in the courses of the medullary veins is noted. To our knowledge, intracranial calcification of this type has not been previously reported.

Medullary veins are venous channels of white matter in the cerebral hemispheres and constitute part of the deep cerebral venous system. Detailed anatomic studies with angiographic correlation have been reported by Kaplan (9), Huang and Wolf (10), and Hooshmand et al. (11). Depending on the destination of drainage, these veins may be divided into superficial or deep medullary veins. Superficial medullary veins begin 1 to 2 cm beneath the cortical gray matter and stream toward and through the cerebral cortex to empty into the superficial cortical veins. Deep medullary veins start from the depth of the white matter and drain into the subependymal veins at various locations of the lateral ventricle (10).



On a normal cerebral angiogram deep medullary veins are more easily identified and can be observed in more than 50% of cases in magnified angiograms (12), whereas the superficial medullary veins are usually obscured and difficult to distinguish from the superficial cortical veins. In pathologic conditions of the white matter such as infiltrating glioma or arteriovenous malformation the deep medullary veins are usually dilated and may appear as relatively straight, crowded, and converging vessels giving a conical "spray" appearance (10). Dilatation of the medullary veins has also been observed in reticulum cell sarcoma, cerebral hemorrhage, inflammatory disease, and superficial venous thrombosis. No calcification of medullary veins has ever been mentioned (12).

High-resolution CT makes possible the visualization and identification of the intracranial vessels. The dilated vascular channels that characteristically converge toward the lateral angles of the lateral ventricles are dilated deep medullary veins (Figure 2E, F). On CT scans, the multiple linear calcifications with "wedge" or conical "spray" appearance (Figures 2, 3) converging toward the lateral angles of the lateral ventricles are therefore in the courses of the medullary veins. It is not surprising that plain skull roentgenography failed to show these calcifications, since CT is 5 to 15 times more sensitive than plain skull films in the detection of intracranial calcifications (13).

Figure 2 D, E, & F. Postcontrast CT scans at corresponding levels. In **D** a few dilated subependymal veins (*arrows*) are noted along the wall of the left frontal horn. In **E** dilated deep medullary veins (*arrows*) are enhanced in the white matter of the left cerebral hemisphere.

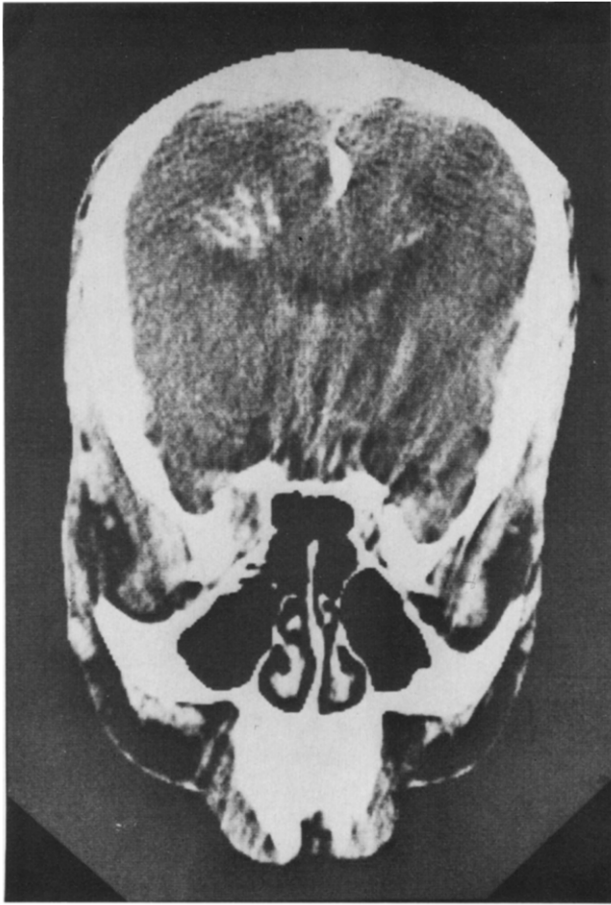


Figure 3. Direct coronal postcontrast CT scan. The cluster of linear calcifications has the same "wedge" or conical "spray" appearance as that exhibited by deep medullary veins collectively.

The pathogenesis of these calcifications is not clear. There might exist some degree of venous stasis at the levels of the medullary veins secondary to retrograde filling in the deep venous system as a result of arteriovenous fistula. This postulation is substantiated by the finding that a few dilated deep medullary veins were still noted in the white matter of the left cerebral hemisphere (Figures 2E, F). A 30-year time interval in our patient may have contributed to the occurrence of these venous calcifications.

CONTINUING MEDICAL EDUCATION QUESTIONS (CALCIFICATIONS OF CEREBRAL MEDULLARY VEINS ASSOCIATED WITH CAROTID-JUGULAR FISTULA)

True or false:

1. Cerebral medullary veins are venous channels of white matter in the cerebral hemispheres.
2. Dilated cerebral medullary veins can be seen in infiltrating glioma, reticulum cell sarcoma, inflammatory disease and arteriovenous malformation.
3. Carotid-jugular anastomosis was a surgical procedure intended to improve cerebral circulation for the mentally retarded and has been proven to have some therapeutic value.

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