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DURAL ARTERIOVENOUS FISTULA — THE RARE CAUSE OF A PULSATING NOISE IN THE EAR

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The article describes the clinical manifestations of dural arteriovenous fistula, which is an abnormal communication between the arteries of the dura mater and venous sinuses or cortical veins. The information on the etiology and pathogenesis of such a malformation in the domestic literature is limited to a few publications. The diagnosis is based on the identification of visual (pulsation of the earlobe) and acoustic phenomena in the patient, as well as the presence of a shunt between the posterior auricular artery (a branch of the external carotid artery) and the dural venous sinuses revealed by neuroimaging, in particular MR angiography. The best treatment method is a neurosurgical intervention using endovascular surgery.

Keywords: *cerebrovascular malformations, dural arteriovenous fistula, tinnitus, pulsating noise.*

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BACKGROUND

Ringing in the ear or in both ears (tinnitus, from Latin *tinnire* — "to ring") represents a sound sensation that is not associated with an external acoustic stimulus [1]. Patients describe it as a sound emanating from one or both ears, inside the head, or as a distant external noise that "hoots, rings, hisses, pulsates," or imitates other familiar sounds. The noise occurs intermittently or is felt constantly, but consistently reduces the quality of life of patients [2].

Subjective noise is felt only by the patient, while objective noise is perceived by others and can be registered by a doctor using a phonendoscope or a special device for recording otoacoustic emission [3], since it has an internal sound source. The latter is often abnormal blood flow in the vessels (often the petrous part of the carotid artery) located in the immediate vicinity of the middle ear, which causes the pulsating feeling in the patient, especially if the sound volume exceeds the hearing threshold in this ear [4]. The most dangerous causes of pulsating noise can be arteriovenous malformations and arteriovenous fistulas of the dura mater, dural fistulas [5, 6].

DURAL ARTERIOVENOUS FISTULAS

Dural arteriovenous fistula (DAVF) is an abnormal communication between the arteries of the dura mater and venous sinuses or cortical veins [1]. Among supratentorial and infratentorial vascular malformations, they account for 6% and 35%, respectively. Until recently, DAVFs were considered a relatively rare pathology, however, due to the increased frequency of neuroimaging methods (computed tomography, magnetic resonance imaging), in recent years, they are diagnosed in a greater percentage of cases [1–7]. According to P. Jabbour et al. [8], the prevalence of DAVF is 0.17 cases per 100 thousand population.

Most arteriovenous fistulas are solitary formations; however, multiple DAVFs have also been described [2–9]. Malformations can develop at any age, including childhood, but they are more often revealed in patients aged 50–60 years old [7–9]. There are congenital and acquired DAVFs [7–11]. Congenital ones are much less common; as a rule, they develop in the first trimester of pregnancy as a result of persistent communication between future arterial and venous segments of the primary vascular plexus or underdevelopment of the intermediate capillary network [7]. Birth trauma, intrauterine infections, and venous thrombosis can have a negative effect on angiogenesis [3–12].

The latter factor is important in the formation of acquired dural fistulas. In one of the studies, such comorbidity was found in 72% of cases [10]. The development of thrombosis of the venous sinus and cortical veins is promoted by hormonal changes associated with pregnancy, childbirth, or, conversely, with the use of oral contraceptives; procoagulant state of the blood; deficiency of proteins C and S, antithrombin III, mutations in genes II, V of coagulation factors; head trauma, intracranial surgery, compression and occlusion of the

venous sinus by a tumor (most often meningioma), otitis media, and sinusitis [9–13].

There are two hypotheses for the formation of DAVF under conditions of sinus thrombosis. According to one of them, an increase in venous pressure associated with thrombosis and obstruction of venous outflow leads to opening of pre-existing physiological shunts in the dura mater with gradual hypertrophy of the sinus wall and the formation of DAVF [13]. Changes in intracranial homeostasis in presence of thrombosis underlie another hypothesis. Prolonged cerebral venous hypertension causes local chronic hypoperfusion of the brain, the markers of which are low cerebral perfusion pressure and a decrease in regional cerebral blood flow. Under these conditions, increased expression of the hypoxia-inducible factor 1-alpha (HIF-1α) and vascular endothelial growth factor (VEGF) leads to aberrant angiogenesis and the formation of DAVP [14].

Classification

Among neurologists and neurosurgeons, Borden and Cognard classifications of dural anastomosis (1995) are the most recognized [9–11]. Both of them emphasize the assessment of retrograde leptomeningeal and cortical venous reflux, the characteristics of which are important for choosing the management of a DAVF patient [10–15]. In 2015, the Standard and Guidelines Committee for the Society of Neurointenventional Surgery proposed a simplified classification (SNIS S&G Classification) according to which DAVF is divided into type 1 (non-aggressive, without cortical venous reflux) and type 2 (aggressive, with cortical venous reflux). Each of them can be asymptomatic and symptomatic, requiring urgent surgical intervention [16].

Clinical presentation

In the presence of DAVF, patients most often complain of pulsating noise in the ear (81% of cases) and headache (15%) associated with irritation of the receptors of the meningeal vessels [9]. The sudden onset of headache and cerebral symptoms is a sign of intracranial hemorrhage (10%) [10].

The aspects of other symptoms that arise in the onset and during the progression of the disease are largely determined by localization of the fistulas. Impairment of venous outflow from the ophthalmic veins with carotid-cavernous fistula, localized in the anterior cranial fossa, can lead to intraocular hypertension and periocular edema. This causes retroorbital pain, periorbital edema, chemosis, ptosis, diplopia (due to external ophthalmoplegia), and decreased visual acuity [7, 13], as well as and retinopathy and glaucoma in rare cases [9].

Pulsating noise in the ear may be the only symptom of fistula of the transverse or sigmoid sinuses, which account for up to 80% in DAVF of the middle cranial fossa. The noise is often unbearable and exhausting for the patient. Its occurrence is associated with an increase in the volume of blood flowing directly under high pressure from the feeding arteries to the venous sinuses, which causes turbulent blood flow in them, synchronized with the systolic phase of the heartbeat [13, 15]. In 40% of cases, it is heard by a doctor during auscultation [17]. The occipital artery, which is involved in the blood supply to the DAVF in this area, is usually hypertrophied. Pressing it against the mastoid process reduces noise during physical examination [15]. The presence of a pulsating noise in the ear, heard by the patient and/or the doctor during auscultation over the mastoid process, is an indication for neuroimaging studies [10].

Infratentorial DAVF is a significant risk factor for the development of brain stem infarction and cervical myelopathy [10]. Isolated lesions of the cranial nerves are caused by their compression by hypertrophied feeding arteries or draining veins [13].

Rupture of cortical veins, depending on their location, leads to intracerebral, subarachnoid, or subdural hemorrhage. In addition, a powerful shunt flow, regardless of the presence or absence of venous sinus thrombosis, can cause local or systemic venous hypertension distal from the lesion site, and the development of venous hemorrhagic infarction. With an increase in venous pressure, intracranial pressure also increases, and as a result of this, in the decompensation phase, the clinical manifestations of the disease may resemble those in case of space-occupying lesion (pseudotumor symptoms), ranging from headache, nausea/vomiting, decreased visual acuity due to edema of the optic papilla, to focal neurological disorders, including hemihyperesthesia, hemiparesis, and aphasia. In severe cases, cognitive dysfunction, local or generalized seizures, stupor, and coma occur [7, 9, 11].

Some malformations remain asymptomatic or retain a stable clinical and angiographic presentation for many years, undergo spontaneous involution with stabilization or reduction of neurological symptoms. The factors predisposing to the disease regression remain unknown [8].

Diagnostics

At the initial stage, for the diagnostics of vascular malformations, it is advisable to conduct magnetic

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resonance (MR) angio- and veno-sinusography. Time-of-flight (TOF) MR angiography reveals the aspects of the vascular system rearrangement in DAVF, namely an increase in the number and size of the feeding arteries, expansion of the draining veins, the presence of a vasculature in the dural sinus or meninges, the state of collateral circulation and venous outflow. Susceptibility weighted imaging (SWI) MR images show signs of cortical venous hypertension or minor hemorrhage [15, 18–20].

Non-contrast computed tomography (CT) of the brain is effective only in diagnostics of adverse outcomes of DAVF such as hemorrhages and cerebral edema; however, CT angiography is able to identify altered arteries and veins, pronounced vasculature in the dural sinus or meninges, and venous sinus thrombosis. In some cases, abnormal restructuring of the vasculature can be hidden by artifacts protruding from the skull bones. The study disadvantages can be eliminated by using reconstruction algorithms to remove bone structures in images [12, 21]. However, digital subtraction angiography remains the gold standard for diagnostics of DAVF [2, 10]. Due to the high spatial and temporal resolution, catheter angiography provides information about the location and anatomical aspects of the fistula, and enables to identify important characteristics of DAVF, such as the presence of cortical reflux, obstruction of venous outflow, and aneurysm [9, 15, 19].

Ultrasound methods, in particular duplex scanning of brachiocephalic vessels, transcranial Doppler imaging and transcranial duplex scanning of intracranial vessels, are additional diagnostic methods for DAVF, especially for patients with nonspecific manifestations. The main advantages of ultrasound methods are non-invasiveness, widespread use, and relatively low cost compared to other angio-imaging diagnostic methods [22]. In DAVF patients, when performing ultrasound methods, lower peripheral resistance indices (Gosling and Purselo indices) are recorded in the feeding arteries, including the external carotid artery and its branches (for example, in the occipital artery). It has been proven that the value of the peripheral resistance indices in the feeding arteries correlates with the treatment efficiency and the clinical evolution of DAVF. In addition, in DAVF patients, transcranial methods reveal a high blood flow velocity, retrograde blood flow, and an abnormal Doppler waveform (arterialization of blood flow) in the cerebral veins and sinuses, and ophthalmic vein [23].

Regardless of the initial diagnostic results or treatment options, long-term angiographic and/or ultrasound follow-up is recommended for all DAVF patients [24].

Treatment

The optimal treatment for DAVF is complete elimination of the fistula. In recent years, endovascular interventions in the form of transarterial, transvenous or combined embolization have been the most preferred. In technically complicated cases, endovascular intervention followed by microsurgical resection can be performed [25]. In case of ineffectiveness or impossibility of using endovascular approaches, intraoperative embolization of meningeal arteries or veins, and resection of the dura mater are performed [6]. Stereotactic radiosurgery is used at a high risk of surgery or in cases when transvenous or transarterial fistula embolization is impossible [12, 25]. With the use of a gamma knife, linear accelerators or proton beams, the elimination of the fistula is achieved due to the targeted delivery of a certain dose of intense radiation to the area of its localization [26].

The following is a clinical case of a patient with a dural arteriovenous fistula as the cause of a pulsating noise.

CLINICAL CASE

Patient information

Patient D., 64 years old, complained of a constant pulsating noise behind the left ear, which intensified in silence, mainly at night, due to which D. began to experience difficulty falling asleep.

The disease history reveals that over the past two years, the patient noted a moderate hearing loss in both ears, was consulted by an otorhinolaryngologist with a diagnosis of sensorineural hearing loss. He periodically noted a pulsating noise in the left ear, which he heard while falling asleep, however, a change in posture or turn of the head helped to eliminate it. Since September 2019, he started to note the pulsation regardless of the time of day with an increase in the absence of ambient noise (early morning hours, a quiet room, noise suppressing, etc.). The night sleep was disturbed, the patient could not fall asleep for a long time, the change in the position of the body and head did not give the desired effect. He became anxious and irritable due to sleep deprivation and obsessive sensations. He was treated in a neurological hospital, referred by the polyclinic, and received a course of vascular, neurometabolic therapy which had no effect. He applied to a neuropsychiatric dispensary, where he was diagnosed with senestopathic disorder, and anti-anxiety and sleeping pills were prescribed.

The anamnesis vitae shows that for many years, the patient suffered from a paroxysmal atrial fibrillation, degree 1 arterial hypertension; constantly took anticoagulants, and received antihypertensive therapy. The patient denied having had diseases of the hearing organs, trauma, surgical interventions in the head, or the history of infectious diseases.

Admission studies and final diagnosis

Examination of the patient revealed a pulsation of the earlobe on the left; during auscultation of the postaural region above the mastoid process, an intense pulsating noise was heard, which coincided with the sounds of the heart. Comparison of the anamnesis and clinical data enabled to suspect the presence of vascular malformation, which requires confirmation using ultrasound and/or neuroimaging research methods.

Duplex scanning of the head vessels revealed signs of arteriovenous malformation in the system of the left external carotid artery with arterialization and increased blood flow through the internal jugular vein. Intensification of the blood flow velocity along the posterior parotid artery suggested it to be a "feeding" vessel (Fig. 1). Contrast MR angiography confirmed the presence of an arteriovenous fistula with involvement of the transverse and, partially, sigmoid sinuses on the left (Fig. 2).

The patient was consulted by a neurosurgeon; and endovascular treatment of the fistula is planned.

CONCLUSION

Dural arteriovenous fistulas represent a rare form of pathology. Knowledge and correct assessment of its symptoms are necessary for adequate interpretation of clinical data, the choice of methods for examining the patient, determining the disease prognosis and treatment approach.

INFORMED CONSENT

Written voluntary informed consent was obtained from the patient to publish the description of the clinical case (date of signing 12/23/2019).

ADDITIONAL INFORMATION

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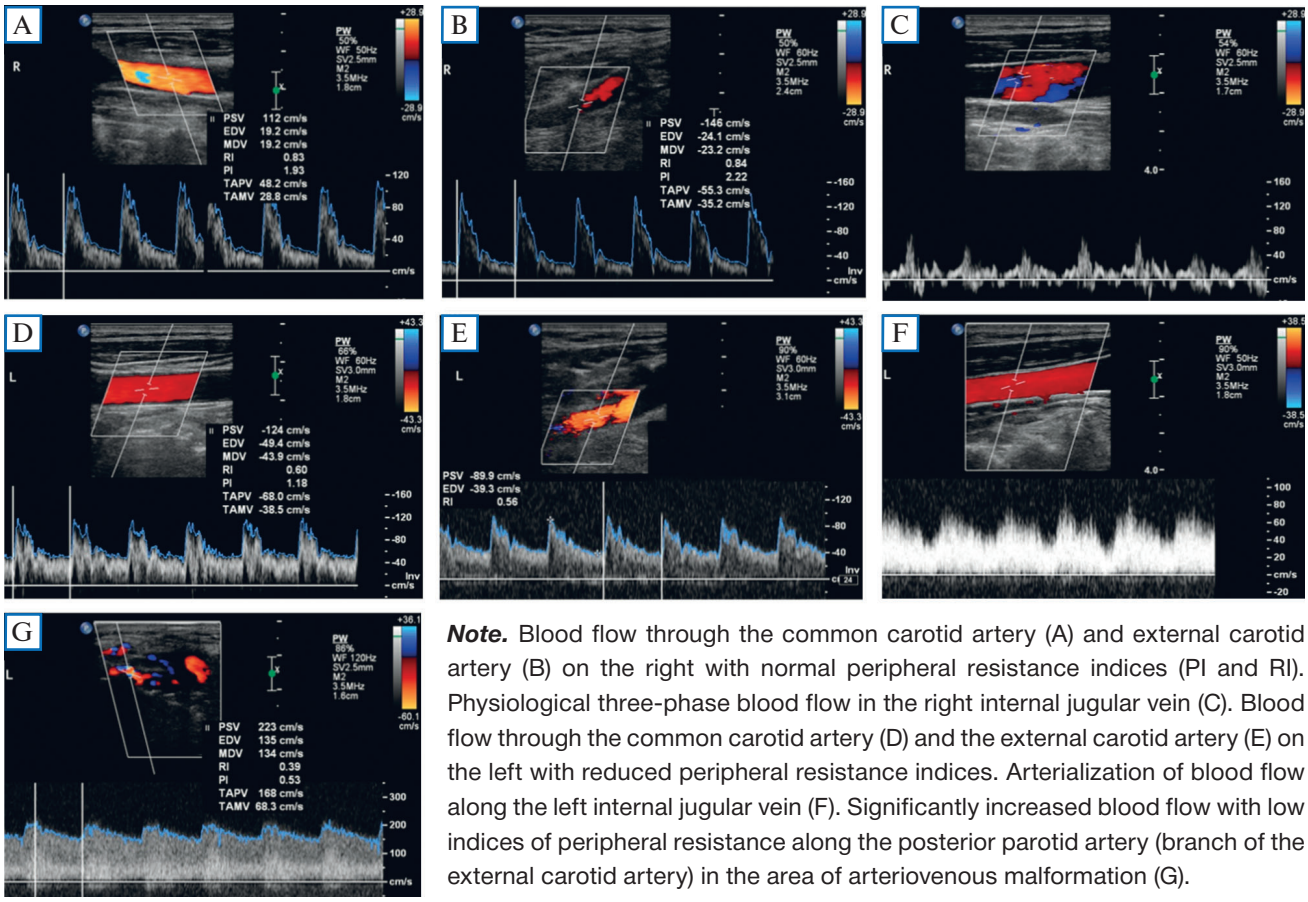
Competing interests. The authors declare no conflict of interest.

Fig. 1. Duplex scanning of neck vessels



Note. Blood flow through the common carotid artery (A) and external carotid artery (B) on the right with normal peripheral resistance indices (PI and RI). Physiological three-phase blood flow in the right internal jugular vein (C). Blood flow through the common carotid artery (D) and the external carotid artery (E) on the left with reduced peripheral resistance indices. Arterialization of blood flow along the left internal jugular vein (F). Significantly increased blood flow with low indices of peripheral resistance along the posterior parotid artery (branch of the external carotid artery) in the area of arteriovenous malformation (G).

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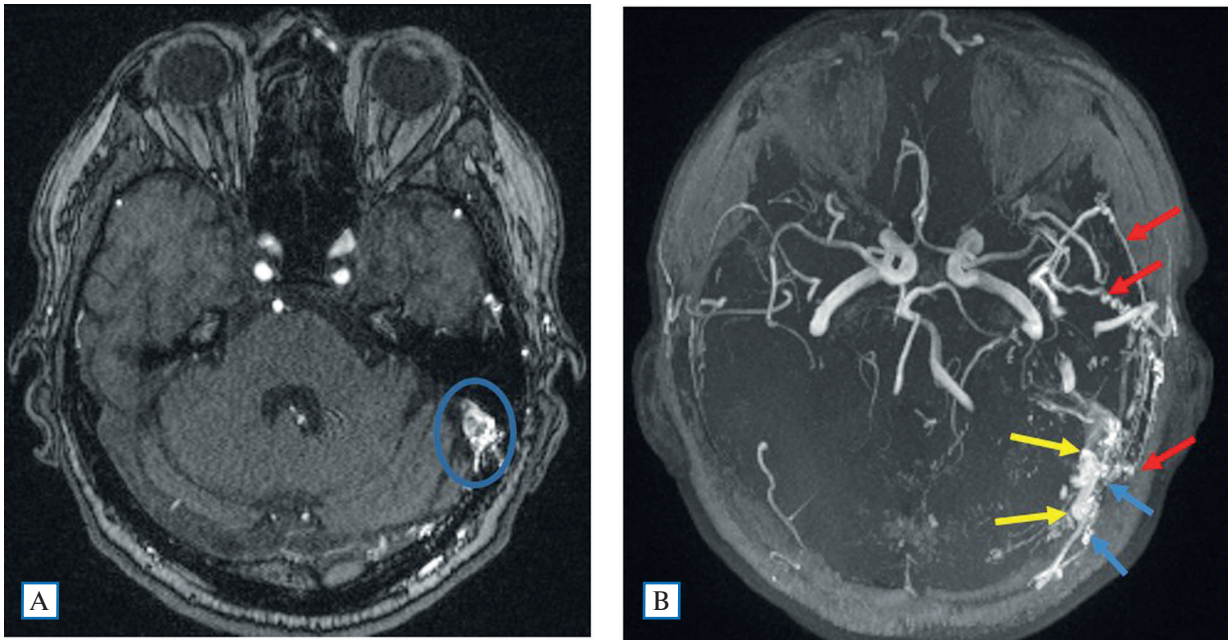
Fig. 2. MR-angiography (3D TOF-angiography)



Note. A — shunting of blood from the branches of the external carotid artery into the left transverse sinus. B — axial MIP-reconstruction: normal arteries of the Willis circle are visualized; on the left, dilated and convoluted branches of the external carotid artery (red arrows), from which blood is discharged into the transverse and sigmoid sinuses (yellow arrows), tortuosity and dilatation of the dural veins due to plethora (blue arrows).

AUTHOR CONTRIBUTIONS

All authors made a significant contribution to the preparation of the article, read and approved the final version before its publication.

REFERENCES

1. Kolesnikov VN, Anokchina EA, Lapin MA. Subjective tympanophonia. *Glavnyy vrach Yuga Rossii.* 2017;2(54):22–24. (In Russ).

2. Ahmad N, Seidman M. Tinnitus in the older adult: epidemiology, pathophysiology and treatment options. *Drugs Aging*. 2004;21:297. doi: 10.2165/00002512-200421050-00002.

3. Gunenkov AV, Kosyakov SYa. Subjective tympanophonia. The current concepts of therapy. *Vestnik otorinolaringologii.* 2014;(3):72–75. (In Russ).

4. Fortune DS, Haynes DS, Hall JW. Tinnitus. Current evaluation and management. *Neur Clin North Am*. 1999;83:153. doi: 10.1016/s0025-7125(05)70094-8.

5. Boiko NV. Diagnosis and treatment of tinnitus. *Vestnik otorinolaringologii.* 2018;83(3):82–87. (In Russ). doi: 10.17116/otorino201883382.

6. Kalinin MN, Khasanova DR, Ibatullin MM, et al. The unusual cause of stroke: cerebral dural arteriovenous fistula. *Nevrologiya, neyropsikhiatriya, psikhosomatika.* 2015;7(4):37–41. (In Russ). doi: 10.14412/2074-2711-2015-4-37-41.

7. Chaichana KL, Coon AL, Tamargo RJ, Huang J. Dural arteriovenous fistulas: epidemiology and clinical presentation. *Neur Clin North Am.* 2012;23:7–13. doi: 10.1016/j.nec.2011.09.001.

8. Jabbour P, Tjoumakaris S, Chalouhi N, et al. Endovascular treatment of cerebral dural and pial arteriovenous fistulas. *Neur Clin North Am*. 2013;23(4):625–636. doi: 10.1016/j.nic.2013.03.010 1052-5149.

9. Elhammady MS, Ambekar S, Heros RC. Epidemiology, clinical presentation, diagnostic evaluation, and prognosis of cerebral dural arteriovenous fistulas. *Handbook Clin Neurol*. 2017;143(3):99–105. doi: 10.1016/B978-0-444-63640-9.00009-6.

10. Lv X, Jiang C, Li Y, et al. Transverse-sigmoid sinus dural arteriovenous fistulae. *World Neurosurg.* 2010;74(2-3):297–305. doi: 10.1016/j.wneu.2010.02.063.

11. Nagm A, Horiuchi T, Kanaya K, Hongo K. Dural arteriovenous fistula could be due to hemodynamic disturbance in dural physiological shunts? *World Neurosurg.* 2016;90:699.e11–699.e18. doi: 10.1016/j.wneu.2016.02.036.

12. Gandhi DJ, Chen M, Pearl J, et al. Intracranial dural arteriovenous fistulas: classification, imaging findings, and treatment. *Am J Neuroradiol*. 2012;33(6):1007–1013. doi: 10.3174/ajnr.A2798.

13. Tsai L-K, Hon-Man L, Jeng J-S. Diagnosis and management of intracranial dural arteriovenous fistulas. *Exp Rev Neurotherapeutics.* 2016;16(3):307–318. doi: 10.1586/14737175.2016.1149063.

14. Uranishi R, Nakase H, Sakaki T. Expression of angiogenic growth factors in dural arteriovenous fistula. *J Neurosurg.* 1999;91(5):781–786. doi: 10.3171/jns.1999.91.5.0781.

15. Serulle Y, Miller TR, Gandhi D. Dural arteriovenous fistulae imaging and management. *Neur Clin North Am*. 2016;26(2):247–258. doi: 10.1016/j.nic.2015.12.003/1052-5149/16.

16. Lee SK, Hetts SW, Halbach V, et al. Standard and guidelines: intracranial dural arteriovenous shunts. *J Neurointervent Surg.* 2017;9(5):516–523. doi: 10.1136/neurintsurg-2015-012116.

17. Javadpour M, Wallace MC. Surgical management of cranial dural arteriovenous fistulas. *Schmidek and Sweet Operative Neurosurgical Techniques*. 2012. P. 959–976. doi: 10.1016/b978-1-4160-6839-6.10080-2.

18. Nakagawa I, Taoka T, Wada T, et al. The use of susceptibility-weighted imaging as an indicator of retrograde leptomeningeal venous drainage and venous congestion with dural arteriovenous fistula: diagnosis and follow-up after treatment. *Neurosurg*. 2016;72(1):47–55. doi: 10.1227/NEU.0b013e318276f7cc.

19. Mossa-Basha M, Chen J, Gandhi D. Imaging of cerebral arteriovenous malformations and dural arteriovenous fistulas. *Neur Clin North Am*. 2012;23(1):27–42 doi: 10.1016/j.nec.2011.09.007.

20. Van Asch CJ, Velthuis BK, Rinkel GJ, et al. Diagnostic yield and accuracy of CT angiography, MR angiography, and digital subtraction angiography for detection of macrovascular causes of in-

tracerebral haemorrhage: prospective, multicentre cohort study. *BMJ.* 2015;351:h5762. doi: 10.1136/bmj.h5762.

21. Lee CW, Huang A, Wang YH, et al. Intracranial dural arteriovenous fistulas: diagnosis and evaluation with 64-detector row CT angiography. *Radiology.* 2010;256(1):219–228. doi: 10.1148/radiol.10091835.

22. Yeh SJ, Tsai LK, Jeng JS. Clinical and carotid ultrasonographic features of intracranial dural arteriovenous fistulae in patients with and without pulsatile tinnitus. *J Neuroimaging.* 2010;20(4):354–358. doi: 10.1111/j.1552-6569.2009.00379.x.

23. Zakharkina MV, Chechetkin AO, Krotenkova MV, Konovalov RN. Ultrasound diagnostics of a spontaneous arteriovenous fistula of the head and neck. *J Ultrason.* 2017;17:217–221. doi: 10.15557/JoU.2017.0032.

24. Brzozowski K, Narloch J, Piasecki P, et al. Are type I dural arteriovenous fistulas safe? Single-centreexperience of endovascular treatment of dural arteriovenous fistulas. *Pol J Radiol.* 2019;84:e179–e184. doi: 10.5114/pjr.2019.84602.

25. See AP, Raza S, Tamargo RJ, Lim M. Stereotactic radiosurgery of cranial arteriovenous malformations and dural arteriovenous fistulas. *Neur Clin North Am.* 2012;23(1):133–146. doi: 10.1016/j.nec.2011.09.011.

26. Youssef PP, Schuette AJ, Cawley CM, Barrow DL. Advances in surgical approaches to dural fistulas. *Neurosurgery.* 2013;74(2):32–41. doi: 10.1227/NEU.0000000000000228.

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