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CONTRIBUTION OF TRANSCRANIAL DUPLEX DOPPLER SONOGRAPHY TO THE DIAGNOSIS OF GREAT CEREBRAL ARTERY STENOSIS IN A CHILD

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SUMMARY – The contribution of pulsating duplex Doppler ultrasonography to the diagnosis of middle (MCA) and anterior (ACA) cerebral artery obstruction in one patient is reported. A 10-year-old boy was admitted to the hospital for pulsating headaches (especially pronounced on physical training). He had no neurologic disabilities. His EEG and brain CT scan were normal, and so were his funduscopic examination, lumbar puncture, and laboratory tests. Transcranial color duplex Doppler ultrasonography showed very high velocities in both ACA and right MCA as a sign of suspected stenosis or spasm. Bilateral subtraction cerebral angiography performed after several months of recurrent headaches and unchanged Doppler ultrasonography findings produced an image of high degree stenosis of A1 segment of both ACA and right MCA, with signs of 'steal syndrome' through the posterior cerebral circulation. MRI performed one year later, after episodes of transient ischemic attacks, showed ischemic infarction in the right temporo-occipital region. The etiology of stenosis was supposed to include vasculopathy, i.e. early stage of moyamoya syndrome. Other vasculopathies were excluded by laboratory tests and clinical elaboration. It is concluded that transcranial Doppler ultrasonography is a very helpful method for detection and follow-up of the degree of stenosis of great cerebral arteries in children, and that it correlates well with cerebral angiography, yet it is not useful in diagnosing the etiology of stenosis.

Key words: *Cerebral arteries, ultrasonography; Ultrasonography, Doppler, duplex; Moyamoya syndrome, etiology; Child*

Introduction

According to Broderick, the incidence of cerebrovascular diseases (CVD) in children is 2.72/100,000 children *per* year, while in young adults it is 14-62/100,000 *per* year. The etiology of CVD differs between children and adults. In adults, atherosclerosis and hypertension are the main risk factors for cerebrovascular insult, while in children the etiology includes cardiologic, hematologic and systemic diseases[1].

The prognosis of large and multiple lesions is poor, and in minor and isolated lesions it is better. Children with lesions of the same grade and localization have better prognosis than adults. It is so because of the 'brain plasticity' in children, i.e. the possibility that in a developing brain the healthy brain regions can 'take over' the function of the damaged ones. Therefore, the younger the child, the better the recovery[2,3].

CVD in children are divided into several groups: AVM and aneurysms, arterial thrombosis, sinovenous thrombosis, thromboembolism, intracranial hemorrhage (ICH), and transient ischemic attacks (TIA). Generally, CVD can also be divided into two large groups: cerebral hemorrhage and cerebral ischemia[4].

Cerebral ischemia is mainly caused by embolism, which is the most common non-traumatic lesion in children, and is caused by heart diseases. The symptoms of cerebral ischemia occur suddenly. The neurologic deficit

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reaches its maximal point right after the onset, and the recovery is quick and dramatic but often incomplete[4].

Risk factors for brain ischemia are numerous and include congenital and acquired heart diseases, systemic vascular diseases, vasospastic, hematologic and coagulation diseases, vasculitis and vasculopathies, structural anomalies, and trauma[1,4-15].

The diagnosis of ischemic stroke is usually made by cerebral angiography. However, in 1982 Aaslid introduced a high-energy pulsed-Doppler system, transcranial Doppler sonography (TCD), and it has since been suggested and even indicated for examination and follow-up of patients with vasoconstriction of whatever cause, for vasospasm after subarachnoid hemorrhage (SAH), and for the diagnosis of stenosis of great cerebral arteries[16-19].

We present a child with pulsating headaches in whom stenosis of great cerebral arteries was diagnosed by transcranial color-coded Doppler (TCCD).

Objectives, Methods and Results

A 10-year-old boy was admitted to the hospital for pulsating headaches, especially pronounced on physical training. His personal medical history showed neonatal jaundice, episodes of exertional dyspnea in early childhood, head trauma at the age of eight, and serous meningitis at the age of nine. The boy had suffered headaches from that time on.

His physical examination showed normal findings, without any neurologic disabilities. His EEG and brain computed tomography (CT) scan were normal, and so were funduscopic examination, lumbar puncture, routine blood tests and coagulation tests (PT, APTT, fibrinogen, TT, fibrinomeres, fibrinolysis, protein C and protein S, factor V Leiden, factor VIII, factor XII). Serologic tests were normal, and so were immunologic and rheumatologic tests. HLA-B12 and B27 were positive. Metabolic findings (homocysteine, vitamin B12 and folic acid) were within the normal range. Cardiac and renal examinations were normal.

TCCD showed very high velocities in the left middle cerebral artery (MCA) (Fig. 1a), attenuated flow in the right MCA (Fig. 1b) and both anterior cerebral arteries (ACA) (Fig. 2), with abnormal spectral velocity waveform and turbulent flow sound as a sign of vascular stenosis.

Carotid duplex Doppler showed normal spectral frequencies with mildly increased velocities in the right carotid siphon.

Bilateral subtraction cerebral angiography (SCA) was performed after several months of repeated pulsating headaches and unchanged TCCD findings, and showed an image of high-grade stenosis of A1 segment of both ACA and right MCA (Fig. 3), with a sign of 'steal syndrome' along posterior cerebral circulation.

Magnetic resonance imaging (MRI) was performed one year later, after a number of clinical episodes of transient ischemic attacks (TIA). MRI revealed ischemic infarction in the right temporoparieto-occipital region (Fig. 4). The treatment prescribed was low-dose aspirin.

We suppose that the etiology of stenosis included an early stage of moyamoya disease, as other vasculopathies were ruled out by laboratory tests and clinical work-up.

Discussion

A 10-year-old boy was hospitalized for pulsating headaches caused by stenosis of great cerebral arteries, detected by TCCD and confirmed by SCA. Detailed clinical and laboratory examinations excluded some types of vasculitis, i.e. granulomatous vasculitis (by normal cerebrospinal fluid finding), polyarteritis nodosa (by absence of abdominal aneurysms, mononeuritis and hypertension), systemic lupus erythematosus (by negative results of biochemical and rheumatologic tests), Wegener's granulomatosis and sarcoidosis (by absence of respiratory complications). It probably was neither Takayasu's arteritis (although cerebral arteries may be affected in type I) nor fibromuscular dysplasia (because of normal renal circulation and normotension). Angiographic findings were similar to those characteristic of moyamoya syndrome[18,19].

In 1957, Takeuchi was the first to describe an adult patient with telangiectatic vascular network at the base of the brain and distal occlusion of the internal carotid artery (ICA). The term 'moyamoya disease' was introduced later, in Japanese meaning "hazy, like a puff of cigarette smoke drifting in the air"[20,21].

In 1965, Leeds and Abbott reported on the same findings in two American-born Japanese children. Cases have also been reported in non-Japanese children[4]. Since 1957, approximately 3,900 cases of moyamoya disease have been reported in Japan and more than 1,000 cases elsewhere[22]. In Japan, the prevalence of moyamoya disease is 3.16/100,000, with an incidence of 0.35/100,000. As a family history of the disease is also found in 10% of patients, some authors have suggested that multifactorial inheritance plays a role in some cases. Anyway, moyamoya disease

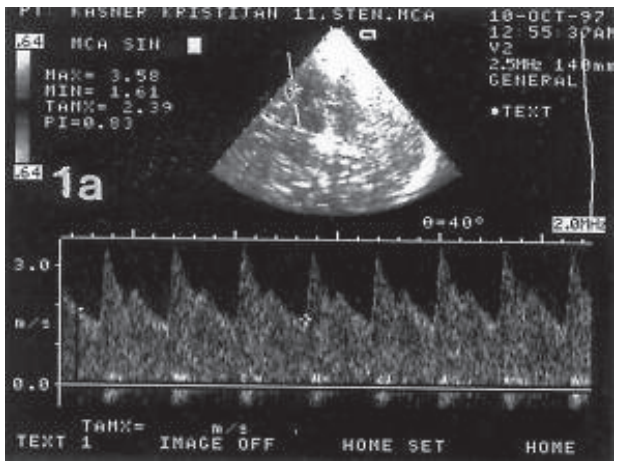


Fig. 1. TCCD findings: (a) very high velocities in the systole and diastole in the left MCA; (b) attenuated flow in the right MCA.

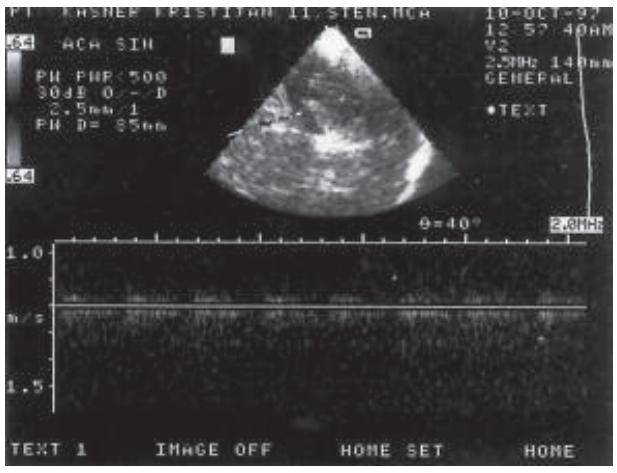
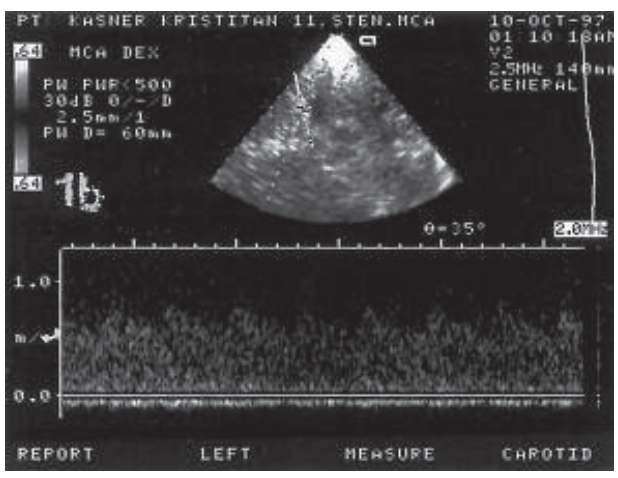
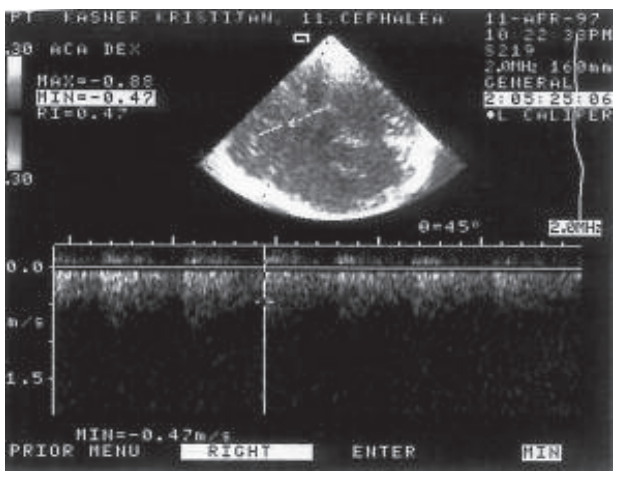


Fig. 2. TCCD finding: attenuated flow in ACAs.





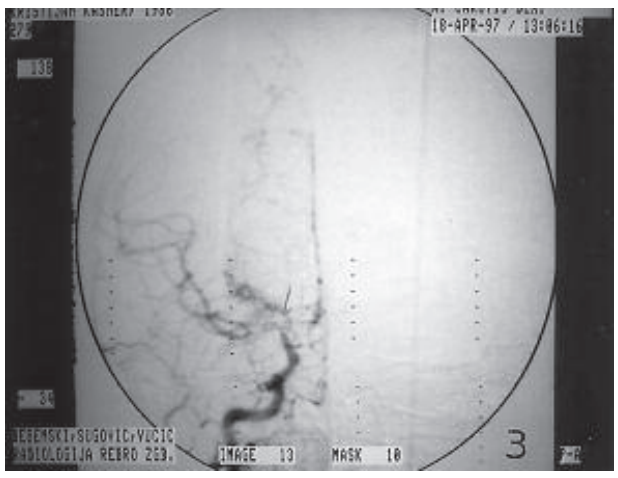


Fig. 3. Subtraction cerebral angiography: stenosis of A1 segment of the left ACA and MCA (arrow).

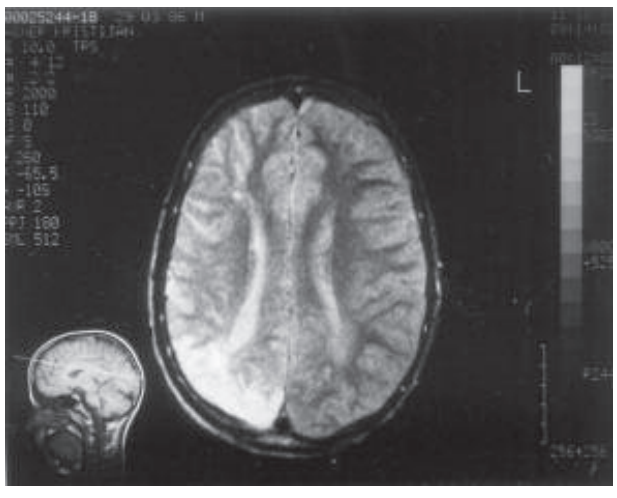


Fig. 4. Brain MRI: ischemic infarction in the right temporoparieto-occipital region.

remains a CVD of as yet unknown etiology[22]. It is a clinical entity characterized primarily by angiographic findings of bilateral stenosis or occlusion at the terminal portion of ICA and/or at the proximal portion of ACA and/or MCA, with abnormal vascular networks in the vicinity of these lesions. In this case, the diagnosis is definitive, however, in case of unilateral involvement, the diagnosis of moyamoya disease is probable, or the term 'moyamoya syndrome' can be used[20-24].

The symptoms and course vary, ranging from no symptoms (incidental findings), a transient disorder, or fixed neurologic deficits of a mild or severe degree. Cerebral ischemia predominates in children, while ICH is more common in adults. In children, hemiparesis, monoparesis, sensory impairments, involuntary movements, headaches, or convulsive seizures often recur, occasionally on alternating sides. Mental retardation or persistent neurologic deficits may also be observed[22]. Our patient suffered only pulsating headaches at first, while episodes of TIA occurred later, with MRI signs of cerebral ischemia.

As the etiology of the disorder is still unknown, different CVD and conditions such as atherosclerosis, autoimmune disease, meningitis, brain neoplasm, trauma, irradiation to the head, Down syndrome, and Recklinghausen's disease should be ruled out[25,26]. Therefore, we performed detailed clinical, biochemical, immunologic and metabolic examinations in our patient. All these findings were normal, thus excluding the above conditions (except for head trauma and meningitis as the probable etiology of this vasculopathy). Fibromuscular dysplasia has the same clinical signs and symptoms but different and typical angiographic findings ('string of beads')[27,28].

In our patient, multiple stenoses of great cerebral arteries were detected by TCCD. TCCD showed increased flow velocities in MCAs and ACAs. When a vessel narrows, irrespective of the cause, the velocity of blood flow increases to allow for the same volume of blood to pass the narrowed lumen. This 'law of continuity' is the basis for the compensatory flow velocity increase found in vascular spasm after SAH. The velocity also increases when there is an augmentation due to collateral contribution to other vessel territories[17,21]. Mild to moderate stenosis increases flow velocity, and this increase inversely correlates with the residual lumen diameter. Large stenosis or occlusion causes decreased velocities or no more flow in this (occluded) vessel[17].

TCCD has proved highly beneficial in the assessment of circulation in the main cerebral vessels in moyamoya patients[17]. It can also be used to establish an optimal treatment plan, including operative anastomotic procedures to prevent stroke or future hemorrhagic events[22].

Conclusion

TCCD is a useful, noninvasive diagnostic method for detection, analysis and follow-up of the degree of stenosis of great cerebral arteries in children, before they develop complications such as stroke. TCCD correlates well with cerebral angiography, but is not useful in the diagnosis of stenosis etiology.

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Sažetak

DOPRINOS TRANSKRANIJSKE DUPLEKS DOPPLEROVE SONOGRAFIJE DIJAGNOSTICI STENOZE VELIKIH MOŽDANIH ARTERIJA U DJETETA

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Prikazan je slučaj 10-godišnjeg dječaka koji je primljen na Kliniku zbog pulzirajućih glavobolja koje su se najčešće javljale za vrijeme tjelesnog napora. Dječak je bio urednog somatskog i neurološkog statusa. Njegov EEG i CT mozga bili su uredni, kao i pregled očnog dna, likvora i laboratorijske pretrage. Transkranijski obojeni dupleks Doppler pokazao je izrazito velike brzine u objema prednjim moždanim arterijama (ACA) i u desnoj srednjoj moždanoj arteriji (MCA), što je moglo odgovarati stenozi krvnih žila. Subtrakcijska cerebralna angiografija učinjena je nakon nekoliko mjeseci opetovanih glavobolja i nepromijenjenog doplerskog nalaza. Pokazala je veći stupanj stenoze prednjeg segmenta obiju ACA i početnog dijela desne MCA, sa znacima 'sindroma krađe' kroz stražnju moždanu cirkulaciju. MRI (učinjena godinu dana kasnije, nakon ponavljanih epizoda prolaznih ishemijskih napadaja) pokazala je ishemijski infarkt temporookcipitalno desno. Etiologija bolesti ostala je otvorenom. Pretpostavljeno je da se radi o vaskulopatiji, tj. ranom stadiju bolesti *moyamoya*. Ostale vaskulopatije isključene su laboratorijskim i kliničkim ispitivanjem. Zaključuje se kako je transkranijski obojeni dupleks Doppler vrlo dobra metoda za otkrivanje i praćenje stupnja stenoze moždanih arterija u djece i dobro korelira s cerebralnom angiografijom, ali još ne pomaže u otkrivanju etiologije stenoze.

Ključne riječi: *Cerebralne arterije, ultrasonografija; Ultrasonografija, Doppler, dupleks; bolest moyamoya, etiologija; Dijete*