Journal of Health Care and Prevention

Review Article Open Access

The Rehabilitation of Patients with Anatomical Variants of the Circle of Willis and Ischemic Stroke in a Multidisciplinary Context

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Abstract

Introduction: Any changes in the morphology of the constituent arteries of the circle of Willis (CoW) could lead to the appearance of different vascular insufficiency syndromes, the capacity of blood distribution in the arteial circle depending on the state of its component vessels.

Material and Method: There was made a scientifical research of medical data from articles written in the last 16 years, from several international medical libraries and databases such as Google Scholar, and PubMed. There were used the following keywords combinations: “circle of Willis, variants, ischemic stroke”, “ischemic stroke, neurological, circle of Willis” and, rehabilitation, stroke, circle of Willis”.

Results: The study shows that the anatomical changes that lead to the impossibility of this anastomotic structure to maintain an adequate flow at the cerebral level could be classified into four main category types. The variants of the CoW could be ischemic stroke risk associated. Beside the pharmacological treatment, patients who suffered an ischemic stroke or a transient ischemic attack (TIA) episode require a special care within a complex interdisciplinary team of professional practicioners (physicians, nurses, psychologists, speech therapists, physiotherapists and occupational therapists) that would work together for their highest benefit.

Conclusion: The early rehabilitation of such patients should be seen as a priority within the healthcare group of professionals and should include therapy in a multidisciplinary context.

Keywords: Morphology; Anatomical changes; Healthcare

Introduction

The arterial circle of Willis (CoW) represents the main arterial anastomosis of the brain, which provides communication between two arterial systems: carotid and vertebro-basilar. In the cases presenting symmetry of the arterial circle, in patients with stenosis or severe occlusion of the Internal Carotid artery (ICA), the blood can be reallocated to the ischemic area cerebral perfusion preservation. Thus, there is prevented the onset of ischemic events and their progression. Any changes in the morphology of the constituent arteries of the circle of Willis could lead to the appearance of different vascular insufficiency syndromes, the capacity of blood distribution in the arterial circle depending on the state of its component vessels [1]. Patients suffering from ICA occlusion but have an arterial circle of Willis with effective collateral circulation have a lower risk of ischemic or hemorrhagic stroke than those without such collaterals [1]. More and more articles reporting various variations from the original definition have appeared in the literature. The autopsic or imagistic studies published so far show that a normal CoW occurs only in 16.6% or, respectively in 42.8% of the population, meaning in less than half of the cases [2], [3].

Material and Methods

There was made a scientifical research of medical data from articles written in the last 16 years, from several international medical libraries and databases such as Google Scholar, and PubMed. There were used the following keywords combinations: “circle of Willis, variants, ischemic stroke”,“ischemic stroke, neurological, circle of Willis” and „rehabilitation, stroke, circle of Willis”. Out of a total of 40 abstracts related to our topic, we selected only 17 most relevant full articles published between 2006 and 2022. Thus, we realized a descriptive review on the topic of neurological rehabilitation of patients with ischemic stroke and anatomical variants of the circle of Willis.

Results

Principal types of variants of the circle of Willis

The anatomical changes that lead to the impossibility of this anastomotic structure to maintain an adequate flow at the cerebral

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Received: 9-Aug-2023, Manuscript No: jhcpn-23-109301, **Editor assigned:** 11-Aug-2023, PreQC No: jhcpn-23-109301 (PQ), **Reviewed:** 25-Aug-2023, QC No: jhcpn-23-109301, **Revised:** 29-Aug-2023, Manuscript No: jhcpn-23-109301(R) **Published:** 5-Sep-2023, DOI: 10.4172/jhcpn.1000212

Citation: Dumitrescu AM, Costea CF, Cucu A, Trandafir LM, Gavrilescu CM, et al. (2023) The Rehabilitation of Patients with Anatomical Variants of the Circle of Willis and Ischemic Stroke in a Multidisciplinary Context. J Health Care Prev, 6: 212.

level are of the following type: 1. the absence (absence) of some of the vessels that compose this structure; 2. the abnormal origin of some of the branches, found to be obviously different to the right and to the left of this structure (in vessels other than the classical one); 3. the presence of additional arteries (duplication, triplication); 4. the presence of a smaller diameter of an artery compared to the contralateral artery (hypoplasia) [4]. When reporting was done only on the number of cases of CoW with variations, the duplicated Anterior Communicating Artery (AcoA) was ranked first among variations of this artery. Kardile et al. (2013) conducted a study in India focusing on investigation mode and number of brains investigated, but took into consideration only the analysis of ACoA variations [5]. They analyzed 100 formalinized brains and found a total of 38 CoWs showing various abnormalities and/or anatomic variants of the component arteries. At the ACoA level, they reported vessel absence in 21.05% of cases, duplication in 26.31%, triplication in 2.63%, hypoplasia in 15.79% and plexiform appearance in 7.89% of all cases with variations [5]. In the study by Sinha et al. (2014), ACoA was duplicated in 22.5% of all variants and hypoplastic in 5.6% of cases, respectively. It was noted if an anatomical variant was present in one of the analyzed arteries, i.e. absence, hypoplasia, duplication, fenestration, difference in length and difference in origin and place of discharge, compared to the opposite segments [6].

Types of stroke related to the variants of the coW

Having the correct knowledge of the anatomical variants of the CoW in adults with cerebrovascular diseases is of great importance in the assessment of cerebrovascular morbidity, especially for neurosurgeons when planning surgery for ischemic strokes, when they need to know the hemodynamics of cerebral circulation.The Anterior Cerebral Artery (ACA) is a terminal branch of the ICA and forms the anterior component of the cow together with the ACoA. It supplies blood to the orbital-frontal and medial hemispheric portions of the brain, including the paracentral lobe and the outer superior part of the frontal lobe. The A1 segment hypoplasia of the ACA represents an uncommon fetal variant of CoW. The frequency of this congenital variation is 1–13%, as derived from angiograms and autopsy reports in various experimental studies [7]. Among all ischemic strokes, strokes in the territory of the ACA constitute only 0.6–3% and are correlated with anatomical variations of the A1 segment of the ACA. The most common abnormalities of ACA are agenesis, hypoplasia, and duplication [7]. The A1 segment of the ACA is a major supplier of anterior collateral blood flow. This segment is also a source for numerous penetrating striatal arteries that supply the anterior hypothalamus, the septum pellucidum, and the anterior and inferior portions of the striatum. Hypoplasia of the A1 segment of ACA as an incompetent part of the CoW appears to be a risk factor for acute ischemic stroke. Most of these patients have small-vessel occlusive strokes, particularly within the striatum, as we also identified in our series. Such strokes occur because of poor collateral capacity, especially when the arteries entering the striatum show lesions of arteriolosclerosis or hypertensive hyalinosis. There is also poor clearance of thromboembolism within the striatum when there is defective collateral circulation. Ischemic stroke related to hypoplasia of the A1 segment is usually tolerable, predominantly presenting with sensory-motor deficit contralateral to the hypoplasia of the A1 segment [7]. Chuang et al. reported that the A1 segment of the ACA can show variations of up to 15% in cerebral magnetic resonance angiographic analyses. The effects of the presence of this anatomical variant are identified either predominantly in the form of ipsilateral lacunar striatal infarcts (in about 70% of the analyzed cases), or in the form of striatal infarcts as a result of the occlusion of penetrating arteries in the striatal area [8]. Pentyala et al. also reported that most of their cases (64.06%) with A1 segment hypoplasia caused poor blood supply to the ipsilateral or bilateral striatal areas, which are irrigated by the striatal branches, which are terminal branches of the ACA, leading to infarcts associated with the hypoplasic artery [9]. Generally, most hypoplastic or aplastic A1 ACAs are asymptomatic and not directly related to any particular neurological disease, but may cause insufficiency of the inter- or intra-hemispheric collateral circulation, which may represent increased risk of ischemic stroke in the area irrigated by the normal ACA. It appears that hypoplasia of the A1 segment of ACA in the pediatric population is predominantly associated with headaches and dizziness, which may play a role in the incidence of neurological disease later in life. The mechanism of these symptoms may be represented by progressive cerebral ischemia [9]. In general, arterial "fenestration" is described if the cerebral artery is doubled in one part of its trunk into two independent channels, which have their own tunica intima and tunica media, and sometimes even an adventitia tunica, and after the appropriate course these channels reform the original artery [10]. Fenestration of the A1 segment may occur due to the absence of fusion of the primitive plexiform anastomosis between the ACA and the primitive olfactory artery, normally present at the 18–43 mm embryonic stage [10]. On the other hand, the ACoA, as an unpaired artery on the midline of the brain base, is characterized by the occurrence of multiple fenestrations or duplications, but these variants have not been associated with cerebrovascular disease. Abnormalities identified in the posterior part of the arterial polygon of Willis result either from the persistence of vessels that should normally disappear during embryonic development, or from the absence of certain vessels that should have been present at birth [3]. The presence of efficient posterior communicating arteries is a vital factor in stroke patients, as these vessels link the anterior to the posterior cerebral circulation, providing a powerful source of plasticity in cerebral hemodynamics [11]. Posterior Communicating artery (PcoA) hypoplasia is also considered a predisposing factor for slow-flow hemispheric infarcts from ICA occlusions [12,13]. It can be asymptomatic except in cases where ipsilateral ICA stenosis occurs [14-16]. Many authors have suggested an increased risk of ischemic stroke in the presence of CoW variations. Chuang et al. (2008), for example, point out that PCoA hypoplasia is associated with an increased risk of infarction, particularly in the thalamic region [17]. Recently, in 2021, Dumitrescu et al. [18] analyzed the medical causes of death in patients with bilateral ACoP hypoplasia. The obtained data allowed the conclusion that extensive stroke can be correlated with bilateral hypoplasia of the ACoP, especially if there is also an associated systemic pathology, as well as occlusion of an important artery that supplies the brain, such as the ICA or the basilar trunk.

a. The intercorrelation between variants in the circle of Willis, additional risk factors and ischemic stroke development

In the last decades, it was demonstrated that obesity, type 2 diabetes mellitus, high blood pressure and also polyarteritis nodosa, due to its systemic manifestations, have been identified as common causes of morbidity and mortality in middle-aged patients [19-21] But neurological diseases, in particular ischemic or hemorrhagic stroke, can result in some of the most important disabilities, increased levels of co-dependency, and even mortality, particularly in the case of the elderlies. Within the pediatric population, there exist, as well, pathologies that have a significant impact on health, especially type 2 diabetes and obesity with insulin resistance, due to their severe complications [22,23], but also the hemato-oncological pathology [24]. Familiarity with the most common variations of CoW and their prevalence may be of vital importance to clinicians. Knowing the anatomical variants at the level of CoW and their frequency can help us understand the atypical patterns of strokes and predict their likelihood. There is also

a great heterogeneity of clinical manifestations of anatomical variants at the CoW level because they are correlated not only with the type of variants, but also with the risk factors that each patient has. The most significant risk factors are atherosclerosis and hypertension, to which is added insulin-requiring type II diabetes. This extends the effect of the first two risk factors, to which are added liver cirrhosis and drug coagulopathy, which also contribute to the occurrence of hemorrhagic stroke, as well as acute hemorrhagic pancreatitis, which, by inducing hypercoagulability status, causes ischemic stroke even in patients with a single anatomical variant, regardless of their age [25]. Therefore, each patient must be investigated in detail, on each organ, each pathology must be treated properly so that the clinical effects of the anatomical variants are as limited as possible.

a. Neurological rehabilitation in a multidisciplinary context

Beside the pharmacological treatment, patients who suffered a stroke episode require a special care within a complex interdisciplinary team of professional practicioners (physicians, nurses, psychologists, speech therapists, physiotherapists and occupational therapists) that would work together for their highest benefit. In the case of post-stroke cognitive impairment, there is continued uncertainty over the benefits and risks of actovegin and cerebrolysin for cognition [26]. The benefits of actovegin and cerebrolysin are considered to be rather limited and might correlate with important side effects. Putting into balance the risks and benefits in the case of using these therapeutical agents, they are mostly indicated in the cases of post stroke cognitive impairment. In the cases of post stroke dementia, there is little positive effect of cholinesterase inhibitors, when side effects should be taken into consideration, as well [27]. The occupational therapists' role becomes important in the case of patients who suffered a stroke, because of the great positive outcomes and impact they could bring in their lives. Focusing on improving the motor function, sensations, overall coordination, visual perception, and cognition of post-stroke patients becomes vital for the general management of their daily life activities. The occupational therapy focuses on remediating deficits, generally, in the case of these patients. It has the purpose and the role to engage them in meaningful activities, tasks, finding remedies to cognitive and motor deficits. The target of both occupational and physical therapies is improving independence, developing skills for task adaptation and environmental changes within a multidisciplinary team [28]. Memory and concentration deficits are usually intercorrelated with reading difficulties in post-stroke patients. Therefor, anomic aphasia can associate with short-term memory loss. In a recent publication, Aras et al. focus on the role of of speech and language therapy in post-stroke patients suffering from aphasia. With the help of magnetic resonance imaging, the authors indicated the affected artery in 100 patients and they also proceeded an aphasia test, realized with Gülhane Aphasia Test-2. They conclude that speech functions could be improved in patients that have the middle cerebral artery affected than in the case of patients with anterior or posterior cerebral artery affected [28]. A specific type of aphasia named anomic aphasia seems to be most often connected to PCA strokes. This particular type would reffer to difficulty in words finding. According to a recent study on the topic of post-stroke neurological rehabilitation, the recommendations for locomotor training and rebalancing exercises for walking recovery consist of: strength building, cognitive training, mobility training [28]. Besides the strength building and rebalancing interventions, the emotional support comes as an important component of the global rehabilitation. For the patients who have survived a stroke episode, there are indicated lifestyle-based interventions such as exercising, adjustment of diet, alcohol-intake and smoking reduction, weight loss [29, 30].

Conclusion

The symmetry and structure of the circle Willis is important for neurosurgeons, vascular surgeons, interventional radiologists and radiologists, because anatomical anomalies and variants affect the hemodynamics of blood flow especially during cardiac surgery with extracorporeal circulation, through the occlusion of one of the major cerebral arteries. The anatomical variants of CoW play an important role in the development of cerebrovascular diseases, with a focus on ischemic strokes, most often having clinical and prognosis-related implications, with a role in the occurrence of psychiatric disorders, as well. The detailed knowledge of the anatomical variants of CoW and the cerebrovascular diseases that can be determined by them have clinical applications, being useful for neurosurgeons in planning their operations, for radiologists in the interpretation of angiograms, and for neurologists, family practitioners and other medical specialists in the correct diagnosis of a stroke. According to the European Stroke Organisation, the early management and rehabilitation of such patients should become a priority within the healthcare group of professionals involved in neurological rehabilitation and include a multidisciplinary assessment and treatment.

Funding

This paper was co-funded from the European Social Fund - the Human Capital Operational Programme, Project/ Grant No: POCU/993/6/13/154722

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