v. 2, n. 8, 2022

International Journal of Health Science

HYDROCEPHALUS IN ADULTS AND ELDERLY, LITERATURE REVIEW

Alícia Calinne Melo Santos
http://lattes.cnpq.br/3583652855161857

Nathalia Costa Macedo Noronha
http://lattes.cnpq.br/1465849002467493

Erasmo de Almeida Júnior
http://lattes.cnpq.br/9488766148228030

Luana Rocha de Souza
http://lattes.cnpq.br/8471986302764645

Carmem Isaura Salles do Nascimento
http://lattes.cnpq.br/5915123975311655

Thayná Bezerra Lima
http://lattes.cnpq.br/2816789558890383

Marina Dantas Freire
http://lattes.cnpq.br/3532709223910275

Mikaela Rodrigues da Silva
http://lattes.cnpq.br/0368515680575492

Juliana Santos Feitosa
http://lattes.cnpq.br/1007156315823299

Alana Anjos da Rocha
http://lattes.cnpq.br/7346970290174218

Mateus Araújo dos Santos
http://lattes.cnpq.br/0489854484593731

Ana Clara Passos Melo
http://lattes.cnpq.br/2585590425937376

Abstract: Introduction and Purpose: Hydrocephalus is an active distention of the ventricular system resulting from inadequate passage of Cerebrospinal Fluid (CSF) from its point of production within the cerebral ventricles to the point of absorption into the systemic circulation. Several causes can motivate the onset of the disease, which occurs regardless of age or gender. The objective of this work was to carry out a systematic review of case reports involving hydrocephalus in adults and the elderly, focusing on epidemiological data, risk factors, clinical aspects, diagnostic methods and therapeutic approach. **Methods:** the bibliographic research was developed from the analysis of scientific articles obtained from the Scientific Electronic Library Online (SciELO), BIREME, PUBMED, LILACS, Google Scholar and Virtual Health Library (BVS) databases. The keywords were used: "hydrocephalus", "normal pressure hydrocephalus and the corresponding words in English, "hydrocephalus" and "normal pressure hydrocephalus". **Results:** There are several causes of Hydrocephalus, such as trauma, hemorrhages, tumors, cysts, infections, neoplasms and problems of a developmental nature, which can arise with senility. Its characteristic clinical aspect is the triad: gait disorder, progressive mental deterioration and urinary incontinence, among others. Regarding diagnostic methods, serial tap-test or lumbar puncture examination can collaborate to establish a more accurate diagnosis. The most recurrent treatment is ventriculoperitoneal shunt with implantation of a programmable valve. Conclusion: hydrocephalus, therefore, can affect people of all ages and genders, with a higher prevalence of some types in the elderly, mainly affecting those over 65 years of age.

Keywords: Hydrocephalus, normal pressure hydrocephalus.

NTRODUCTION AND OBJECTIVE

Hydrocephalus is caused by an imbalance in the production and absorption of cerebrospinal fluid (CSF) or obstruction of its pathways, resulting in ventricular dilation and increased intracranial pressure, which can cause damage to the brain. (AZEVEDO et al, 2017) Unlike pediatric hydrocephalus, its development in adults is often secondary to other pathologies. (Bir, Shyamal C et al, 2016).

With regard to disease investigation, imaging plays a crucial role in diagnosis, differential diagnosis and treatment planning. (Langner, Sönke et al, 2017) In many cases, the course of hydrocephalus can be significantly altered with neurosurgical intervention, thus offering patients a normal life expectancy; however, in those with cancer-related hydrocephalus, therapy options may be limited to palliative care. (Filis et al, 2017)

The objective of this article was to carry out a systematic review of hydrocephalus in adults and the elderly, focusing on epidemiological data, pathophysiology, clinical aspects, diagnosis, treatment and prognosis.

METHODOLOGY

This review article was constructed through a survey of scientific articles found in the literature. The bibliographic searches were carried out in the electronic databases LILACS (Latin American and Caribbean Literature on Health Sciences), Pubmed, BIREME, Google Scholar, Scielo (Scientific Electronic Library Online) and VHL (Virtual Health Library). A total of 6033 articles were found in the databases, and of these, 2272 were excluded, 50 were selected and 17 were used. The keywords used in the searches were: hydrocephalus; normal pressure hydrocephalus.

EPIDEMIOLOGY

Even with the advent of technology,

hydrocephalus remains an underreported disease.

PREVALENCE OF HYDROCEPHALUS IN THE ADULT AND ELDERLY POPULATION

According to Jaramillo (2019), epidemiological studies of the elderly population, ≥ 60 years, revealed that the prevalence of hydrocephalus for the age group between 70 and 79 years is 0.2% in relation to the world population, while individuals over 80 years reach almost 6%. In Brazil, hydrocephalus affects one to three people in every thousand births, 40% of which are in the elderly population, with a predominance of males (AZEVEDO, 2019).

According to Oliveira et al (2019), a trial was carried out using magnetic resonance imaging in Japanese elderly (≥ 61 years), reaching a prevalence of 6.46% of dilated ventricles, with only 0.51% of these individuals having specific symptomatology for hydrocephalus.

The exposed prevalence of hydrocephalus in the analyzes showed a 17-fold increase in adults to elderly. However, there is a high probability that a part of the adult population has a stable disease since childhood, underreported by health professionals. It is essential to emphasize that more assertive epidemiological data will provide appropriate guidance and movement of resources for patient care (ISAACS et al, 2018).

ETIOLOGY

Hydrocephalus generally has a congenital or acquired etiology. Congenital forms result from central nervous system malformations (cerebral aqueduct stenosis, Dandy-Walker syndrome, Chiari malformation, meningomyelocele), infection, intraventricular hemorrhage, genetic defects, trauma, and teratogens. Among the acquired etiologies, we highlight the development of hydrocephalus after a hemorrhagic process in the subarachnoid space or, less commonly, in the ventricular system, central nervous system (CNS) tumors, CNS infections and low pressure hydrocephalus. (RAMOS, Júlio César et al, 2019)

In adults there is a predominance of normal pressure hydrocephalus (HNP), a pathology in which the cerebral ventricles are enlarged, but intracranial pressure (ICP) is not elevated. This type of hydrocephalus can be idiopathic in which there is no proven etiology. Or, PHN may present secondary to impaired CSF absorption caused by intraventricular or subarachnoid hemorrhage (from aneurysm or trauma) and previous acute or chronic chronic meningitis). (OLIVEIRA et al, 2019; SALDARRIAGA-CANTILLO et al, 2020).

PATHOPHYSIOLOGY

The cerebrospinal fluid, protective of the Central Nervous System (CNS), distributor of nutrients and defense agents against infections, is produced by the choroid plexuses and absorbed by the bloodstream. To make the journey, the fluid circulates through the ventricles until it leaves through small holes located at the back of the spinal cord. If, for some reason, the total obstruction of the ventricular system occurs, the ventricles rapidly expand, making the CSF no longer able to circulate through the CNS and be absorbed.

Some hypotheses are involved in the elucidation of the pathophysiology of Hydrocephalus, among them, there is the "Hydraulic Pressure Hypothesis" which proposes that the pressor-hydraulic effect explains the increase in force that would occur on the surrounding brain tissue, when progressive dilatation of the muscles is observed. ventricles. According to this hypothesis, at the beginning of the disease

intracranial pressure is probably elevated and ventricular dilatation. Once dilated, the ventricles would then be maintained under the action of a lower pressure, since the dilatation causing an increase in the ventricular surface, leads to an increase in force on the surrounding brain tissue.

In cases where there are no antecedents of changes in CSF flow, other theories suggest that hypertension and atherosclerotic vascular disease alter the self-regulatory mechanisms, decreasing cerebral blood flow and producing a decrease in perfusion that would lead to ischemia and microinfarctions. These vascular changes reduce the tensile capacity of the brain, which would result in secondary dilation of the cerebrospinal fluid spaces. In addition, microinfarctions in the periventricular tissue limit CSF reabsorption.

CLINICAL ASPECTS

Hydrocephalus is an increase in the amount of cerebrospinal fluid inside the braincase, usually in the ventricular cavities, but it can also occur in the subdural space. The main immediate clinical consequence is intracranial hypertension (RAMOS, Júlio César et al, 2018; AZEVEDO, Amanda et al 2019). Underlying conditions contribute to increased intracranial pressure, such as tumors, choroid plexus cyst, infections, obstructions, obstruction of a previously installed ventriculoperitoneal shunt valve (RAMOS, Júlio César et al, 2018).

The condition consists of an acute phase, which is manifested through symptoms such as severe headache, convulsion, vomiting, drowsiness, syncope or rapid lowering of the level of consciousness (AGARWAL, Nitin et al, 2018). According to AGARWAL, Nitin et al (2018), headache is commonly described as pressure, tension or shunt headache. In addition, headache is usually accompanied by sensitivity to external stimuli, and is also described as migraine. In certain situations, Cushing's triad may be present, characterized by systemic arterial hypertension, bradycardia and irregular breathing pattern (RAMOS, Júlio César et al, 2018).

Regarding the symptoms of chronic hydrocephalus, there are chronic headache (with worsening after awakening), vomiting and lethargy. In the elderly, the chronic stage of hydrocephalus can mimic dementia, called Normal Pressure Hydrocephalus (RAMOS, Júlio César et al, 2018). The clinical presentation may consist of cognitive decline, urinary incontinence, and gait disturbances. Regarding cognition disorders, difficulties commonly involve multitasking, planning, organizing or directing functions (AGARWAL, Nitin et al, 2018).

Regarding gait changes, abnormalities are usually related to irregular surfaces or a slow pace, so that most patients have a history of falls or require assisted gait (AHMED, AK et al, 2018; AGARWAL, Nitin et al, 2018; AGARWAL, Nitin et al. al, 2018; RAMOS, Júlio César et al, 2018).

DIAGNOSIS

PNH represents a diagnostic challenge because it presents some differential diagnoses, such as Alzheimer's disease and microangiopathy, and even with aging itself. Therefore, in order to define the diagnosis, there must be converging evidence from the clinical history, physical examination, neuroimaging studies and dynamic CSF studies. Regarding the clinical evaluation, there is the symptomatic triad of PNH, where there must be findings of gait disturbance, at least one area of change in cognition and symptoms of urinary incontinence. Regarding gait disturbance, at least two of the following must be present: decreased step height, step length and gait rhythm, increased trunk balance, widened base, toes turned out when walking, turning in block and altered balance

when walking. Regarding cognition, one must have a documented change and/or decrease in performance on a cognitive test. As for the symptoms of urinary incontinence, one of the following must be present: episodic or persistent urinary incontinence, not attributed to primary urological diseases; urinary and fecal incontinence; urinary urgency and increased urinary frequency.

With a compatible clinic, the diagnostic investigation must be continued with neuroimaging, which includes computed tomography (CT) and magnetic resonance imaging (MRI). CT is useful as an initial screening, to exclude the diagnostic hypothesis, or to indicate further investigation. The MRI is

superior to CT, as it provides more precise information and allows the evaluation of specific parameters.

The main radiological parameters with strong diagnostic and prognostic value referenced in the literature are:

(1) Ventricular area: > 33.5 cm2, presents a direct correlation with the ventricular volume, allowing an easier calculation (BARONCINI et al, 2018, p. 227);

(2) Evans Index: It is considered altered when greater than 0.31 (ABDELRAZED et al, 2017, p. 35);

(3) Third ventricle width: measured in the axial plane, in the middle portion (ENGEL et al, 2018, p. 2);

(4) High convexity of the subarachnoid space: rated as 0 (normal), 1 (light pinching), 2 (moderate pinching) and 3 (severe pinching), presenting a predictive value of response to CSF derivation (BARONCINI et al, 2018, p.221);

(5) Sylvian fissure width: evaluated in cross-sections as 0 (narrowed), 1 (normal), 2 (slightly dilated) and 3 (severely dilated) (ENGEL et al, 2018, p. 2);

(6) Periventricular and deep white matter hyperintensities (ENGEL et al, 2018, p. 2);

After diagnostic confirmation by neuroimaging parameters, it becomes necessary to use supplementary tests in order to confirm the diagnosis and identify which patients are likely to improve after the surgical intervention. The most commonly used tests include the intracranial pressure monitoring test, the lumbar puncture test, and the CSF outlet resistance test. The first can be considered during the PNH diagnosis/ prognosis phase by measuring both static and pulsatile ICP. The presence of spikes in CSF pressure elevation (B wave) represents a positive surgical prognosis variable. The second, a lumbar puncture is performed and about 30-50 ml of CSF is removed. The patient is observed for changes in gait and/ or cognitive function approximately 30 to 60 minutes later. A positive response to the test guarantees a greater degree of certainty for a favorable response to the placement of a CSF shunt. The third consists of infusing saline or fluid into the ventricle or lumbar subarachnoid space with the aim of increasing CSF outflow resistance with subsequent increase in CSF pressure.

PROGNOSIS

The prognostic assessment of hydrocephalus in adults and the elderly is directly linked to the etiology, diagnosis and treatment. The mortality rate is reduced in cases of early diagnosis, generally obtaining a greater chance of successful treatment.

The poor prognosis is related to treatment complications, especially in relation to infections in cases of PVD and valve obstruction, in addition to cases with more prolonged symptoms.

It is therefore necessary to adequately prepare professionals, form multidisciplinary

teams and improve levels for the detection and treatment of neurological diseases.

TREATMENT

The therapeutic approach to hydrocephalus in adults and the elderly aims to restore functional ability and restore the patient's quality of life. Management depends on the causes and type of hydrocephalus the patient has. (RAMOS, Júlio César et al, 2018). Therefore, it can be treated temporarily or definitively, through invasive and non-invasive approaches. (Pereira RM et al, 2012).

In non-invasive transient management, drugs may be administered in order to reduce excessive CSF production or stimulate absorption. Acetazolamide and furosemide can reduce CSF production by 50-60% as carbonic anhydrase reducers. The recommended doses for acetazolamide are between 50 and 150 mg/kg/day and for furosemide it is 1mg/kg/day. However, these drugs have a limited effect on the control of hydrocephalus, in addition to generating side effects such as metabolic acidosis, demyelination and nephrocalcinosis. (Pereira RM et al, 2012).

The definitive treatment of hydrocephalus can be through the removal of obstructive processes (neoplastic, granulomatous, etc.), the implantation of extracranial shunts, such as PVD or ventriculoatrial (VAD), or through internal shunts with the use of neuroendoscopy, ducts in which there is a significant reduction in the cerebrospinal fluid volume effectively. (RAMOS, Júlio César et al, 2018).

The surgical technique of ventriculoperitoneal shunt (PVD) is the preferred indication for treatment. It is a procedure adopted many decades ago with rates of efficacy and safety above 80%. (ROCHA, Suyanne. Et al, 2018). It consists of placing a catheter in the cerebral ventricle, connected to a valve and another catheter, in which the liquor, through a system with a unidirectional valve, is diverted for absorption in another natural cavity of the body, usually the peritoneal cavity. (RAMOS, Júlio César et al, 2018).

The valve has the function of regulating the flow, opening whenever there is an increase in the ventricles and draining excess CSF. (ROCHA, Suyanne et al, 2018). Symptoms disappear completely soon after the procedure and recurrence rates are extremely low. However, it is contraindicated in elderly patients on anticoagulant medication, due to the risk of subdural hemorrhage. (Pereira RM et al, 2012).

The second alternative is VAD, with the implantation of the distal catheter in the right heart atrium. (Pereira RM et al, 2012). VAD is a more complex technique than DVP, with a lower incidence of complications, but of greater severity, such as venous thrombosis, endocarditis, septicemia, thromboembolism, nephritis and convulsions. (RAMOS, Júlio César et al, 2018).

There is also a procedure called neuroendoscopy, which makes it possible for the ventricle to communicate with the cistern, so that the CSF circulates more easily and minimizes cerebral pressure, in addition to having the advantage of the absence of a foreign material introduced into the body. (ROCHA, Suyanne et al, 2018).

CONCLUSION

It is certain, therefore, that hydrocephalus consists of dilatation of the ventricles with consequent elevation of intracranial pressure for different causes, in addition to causing brain damage.

Despite technological advances, hydrocephalus is still an underreported and little known disease, compared to other more popular ones. With greater knowledge about the disease, the chances of prevention and/or

faster identification would be greater.

The pathophysiology of hydrocephalus, as well as the entire regulatory system regarding cerebrospinal fluid, is quite complex and must be well investigated when there is suspicion, since the clinic allows a differential diagnosis with other diseases. The prognosis depends on the cause, early diagnosis or not and treatment. This can be medicated, surgical, temporary or definitive, according to each patient.

REFERENCES

AGARWAL, Nitin et al. **Observations from SocialMedia Regarding the Symptomatology of Adult Hydrocephalus Patients.** WORLD NEUROSURGERY, p. 1-8, 2018. Disponível em: file:///H:/ARTIGO%20%201%20HIDRO.pdf. Acesso em: 31 de janeiro de 2022.

AHMED, A.K. et al. "**Does the Presence or Absence of DESH Predict Outcomes in Adult Hydrocephalus?.**" American Society of Neuroradiology, p. 2022-2026, Nov, 2018. Disponível em: file:///H:/ARTIGO%202%20HIDRO.pdf. Acesso em: 31 de janeiro de 2022.

AZEVEDO, Amanda Santana; SANTO MENDES, Nathália Barbosa do Espírito. **Hidrocefalia: Aspectos clínicos, etiologia e fatores associados.** Biológica-Caderno do Curso de Ciências Biológicas, v. 2, não. 1, 2019.

BARRETO, Wilker John et al. **Hidrocefalia de pressão normal: perspectivas terapêuticas para o envelhecimento saudável**. Campina Grande. 2019.

BIR, Shyamal C et al. **"Epidemiology of adult-onset hydrocephalus: institutional experience with 2001 patients".** Neurosurgical focus vol. 41,3 (2016): E5. doi:10.3171/2016.7. FOCUS16188.

CASTRO, A. F. S.; VIEIRA, N. B. S. **Práticas Cirúrgicas no Tratamento da Hidrocefalia: Revisão Integrativa**. Brazilian Journal of Development, v. 7, n. 2, p. 11757-11774, 2021.

FILIS, Andreas K et al. **"Cerebrospinal Fluid and Hydrocephalus: Physiology, Diagnosis, and Treatment".** Cancer control: journal of the Moffitt Cancer Center vol. 24,1 (2017): 6-8. doi:10.1177/107327481702400102.

ISAACS, A. M. *et al.* **Age-specific global epidemiology of hydrocephalus: systematic review, metanalysis and global birth surveillance.** Public Library of Science Medicine, v. 13, n. 10, p. e0204926, 2018.

JARAMILLO, E. G. C. **Hidrocefalia y complicaciones en adultos tratados con derivación ventriculo peritoneal**. Tese de Doutorado. Universidad de Guayaquil. Facultad de Ciencias Médicas. Carrera de Medicina, 2019.

LANGNER, Sönke et al. **"Diagnosis and Differential Diagnosis of Hydrocephalus in Adults".** "Diagnostik und Differenzialdiagnostik des Hydrozephalus beim Erwachsenen." RoFo: Fortschritte auf dem Gebiete der Rontgenstrahlen und der Nuklearmedizin vol. 189,8 (2017): 728-739. doi:10.1055/s-0043-108550.

MUÑOZ-SUAREZ, D. R. *et al.* **Hidrocefalia De Presión Normal.** Revista Ecuatoriana de Neurología, v. 26, n. 1, p. 40-45, 2017.

NAKAJIMA, M. *et al.* "**Guidelines for Management of Idiopathic Normal Pressure Hydrocephalus (Third Edition): Endorsed by the Japanese Society of Normal Pressure Hydrocephalus."** *Neurologia medico-chirurgica,* vol. 61,2, 2021.

OLIVEIRA, Louise Makarem; NITRINI, Ricardo; ROMÁN, Gustavo C. **Hidrocefalia de pressão normal: uma revisão crítica.** Dementia e neuropsicologia, v. 13, p. 133-143, 2019.

PEREIRA RM, Mazeti L, Lopes DCP, Pinto FCG. **Hidrocefalia de pressão normal: visão atual sobre a fisiopatologia, diagnóstico e tratamento/ Normal pressure hydrocephalus: current view on pathophysiology, diagnosis and treatment.** Rev Med (São Paulo). 2012 abr.-jun.

ROCHA, Suyanne Aley Lima. Et Al. **Hidrocefalia em pacientes idosos: uma revisão integrativa.** Revista Científica Multidisciplinar Núcleo do Conhecimento. Ano 03, Ed. 08, Vol. 06, pp 190 – 208, agosto de 2018.

RAMOS, Júlio César et al. **Hidrocefalia aguda: uma revisão bibliográfica**. Revista Salusvita, n.4, p. 1019-1028, 2018.

SALDARRIAGA-CANTILLO, Alejandra; YESPE-GAVIRIA, Viviana; RIVAS, João Carlos. **Hidrocefalia de pressão normal: atraso diagnóstico**. Biomédica, v. 40, n. 4, p. 656-663, 2020.