Benign Cavum Septum Pellucidum Cystic Lesions: A Case Series

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Abstract

A cavum Septum Pellucidum is a cerebrospinal fluid-filled (CSF) cavity between the lateral ventricles and is considered a normal anatomic variant sporadically seen on neuroimaging. While a cavum septum pellucidum is a relatively uncommon incidental neuroimaging finding, symptomatic cysts of the cavum septum pellucidum are very rare, with only a few cases reported in the literature so far. This study comprises of 6 patients 1 female and 5 males, age ranging from 1-70 years old treated at 2 clinics between 2021-2024 who were being managed by an ongoing process of monitoring through follow-ups and medical measures. These cases would guide future diagnosticians who come across this pathology in patients with limited resources and reluctance for non-pharmacological interventions since it can be difficult to treat something with very little evidenced literature.

Keywords: *Cavum septum pellucidum cyst, imaging, Hydrocephalus, intervention, symptomatic.*

INTRODUCTION

The cavum septum pellucidum is defined as a normal cavity present during the growth of a fetus, its embryological origin takes place when two leaflets of the septum pellucidum are formed which contains a cerebrospinal fluid (CSF) filled cavity in between them, in 85% of cases the leaflets fuse by three to six months postnatally in a caudal to rostral direction ending the existence of this structure but in a rare occurrence it might persist even after that. It is present in as much as 12% of children between 6 months and 16 years [1-3]. Other than the congenital persistence of CSP it has also been reported as an acquired cause after repetitive trauma as seen in professional contact sport athletes such as retired boxers and American football players [4, 5].

Previously the CSP was also known as the fifth ventricle but that term was labelled inappropriate as it is deprived of its choroid plexus and is non-continuous with the rest of the ventricular system [6]. It is most commonly observed as an innocuous incidental finding on an MRI and in some uncommon cases, the persistence of this cavity may lead to the formation of an expanded Cyst which could be labeled as asymptomatic or symptomatic depending on the presentation [3]. The proper criteria for these cysts are still ill-defined but multiple authors state that it can be characterized by lateral bowing of the walls with membrane distance at least 10mm or more with the main symptomatic presentation being headaches, nausea or vomiting, loss of consciousness, and psychiatric disturbances. A cyst large enough can also produce neurological impairment by compression of the neighboring neural structures with the patient presenting with specific symptoms such as seizures, visual disturbances, gait changes, vertigo, and cognitive impairment [4, 7, 8].

This study aims to highlight the array of Cavum Septum Pellucidum Cyst cases we have encountered, managed and followed up with and compare it with the previously available literature to add a new insight to its treatment.

CASE DESCRIPTION

CASE 1

We present the case of a 20-year-old male who had his first episode of generalized tonic-clonic fits five years prior in 2018 and was treated at the emergency department of a secondary care hospital. The fits were treated with intravenous diazepam. During the presentation to our clinic, the patient had no active complaints. His physical examination was unremarkable, and there were no focal neurologic deficits. The results of additional laboratory tests were negative, which aided in ruling out differentials. The patient was born through normal vaginal delivery and had no history of obstructive or traumatic birth. He denied the use of any recreational drugs or a history of traumatic injuries to the head. In 2020, he presented to the emergency department with generalized tonic-clonic seizures. After symptomatic management an MRI with and without contrast Gadolinium diethylenetriamine penta-acetic acid (Gd-DTPA) was performed, which showed a cavum septum pellucidum cyst measuring 17mm by 22mm as well as cavum vergae were also visualized (**Fig. 1**). Later on, the patient reported his third episode of tonic clonus seizures in 2022, he was then prescribed oral sodium valproate (500mg) twice daily, with which he has been compliant to date. The patient has been asymptomatic ever since, and he

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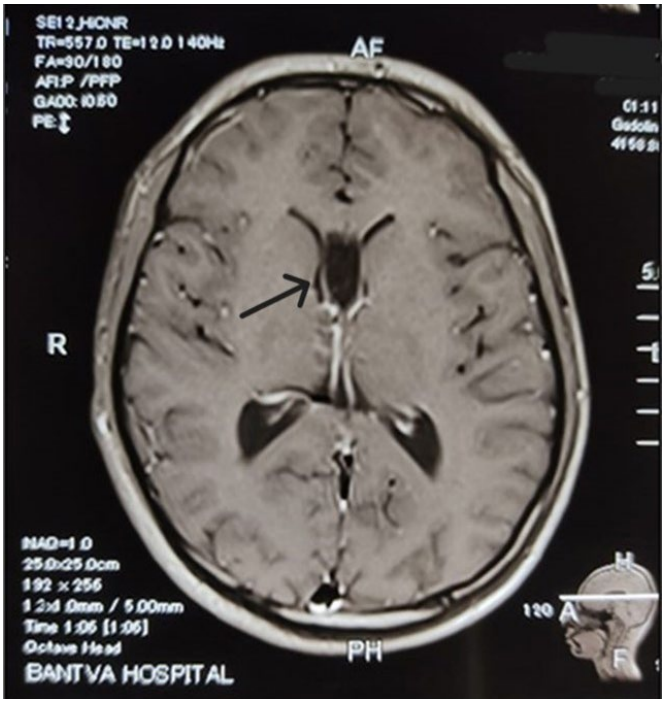

Fig. (1): T1 Weighted MRI in axial view showing CSP cyst (arrow).

has not reported any new episodes of seizures, on telemedical inquiry he is now able to perform his daily activities including his job without any problem, he does experience episodes of slight headache once a week but according to him, it is manageable. Now it has been more than a year, and he is still on Sodium Valproate 500mg once daily just before bed.

CASE 2

A 13-month-old female presented with 2 episodes of seizures. Physical Examination did not reveal any findings. An MRI of the head with and without contrast (Gd-DTPA) was performed and we came across a CSP cyst (**Fig. 2**) the patient was treated symptomatically and was given anti-epileptic levetiracetam dosed according to her body weight, and her symptoms subsided and she did not report any future complications or recurrence on follow-up after 3 months.

CASE 3

A 16-year-old male patient presented with only symptoms of gradual continuous headache, Examination was

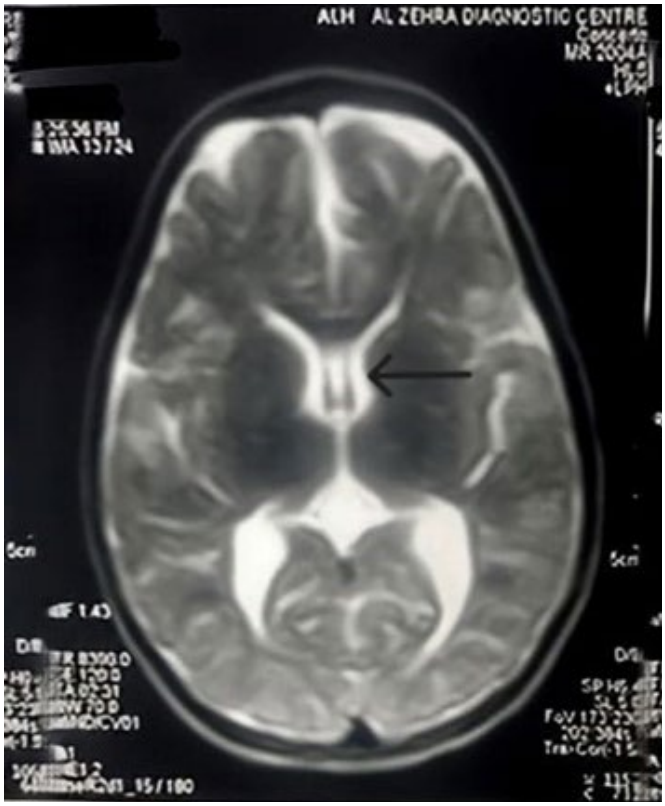

Fig. (2): T2 Weighted MRI of the patient showing cavum septum pellucidum cyst (arrow).

unremarkable for any focal neurological deficits we performed an MRI, and an incidental Cavum septum pellucidum cyst was observed with posterior extension, cavum vergae along with concomitant communicating hydrocephalus (**Fig. 3**) which we believed to be the cause of his headaches. Ventricular diversion procedure was advised at that time with proper risk-benefit counseling, along with symptomatic treatment for headaches. However, the patient was lost on follow-up.

CASE 4

A 4-year-old male patient presented with headache, difficulty in walking, and signs of raised intracranial pressure (papilledema was evident through fundoscopy). There was a birth history of cesarean section along with vascular insult and hypoxia. The patient had an MRI scan done beforehand and after having a look at his imaging

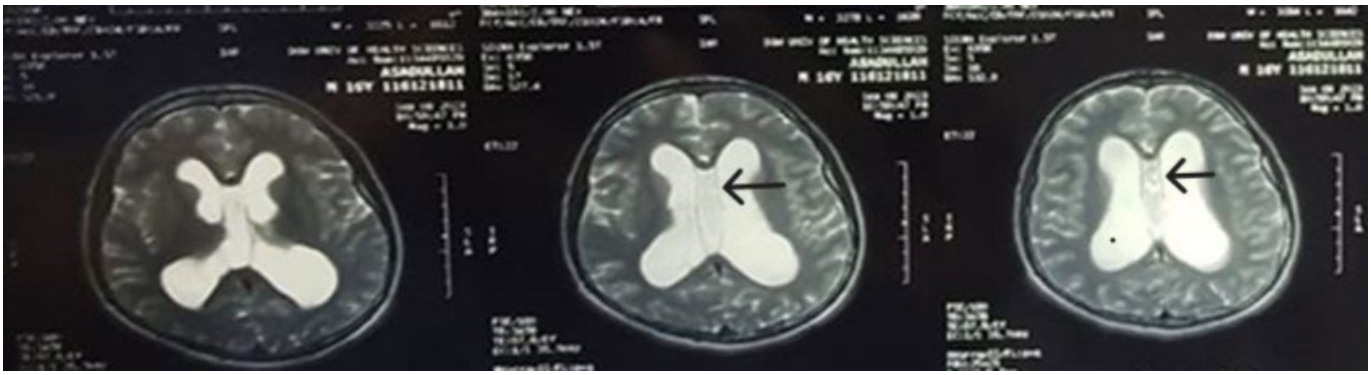

Fig. (3): T2 Weighted MRI showing CSP cyst (arrow) cavum vergae, along with hydrocephalus.

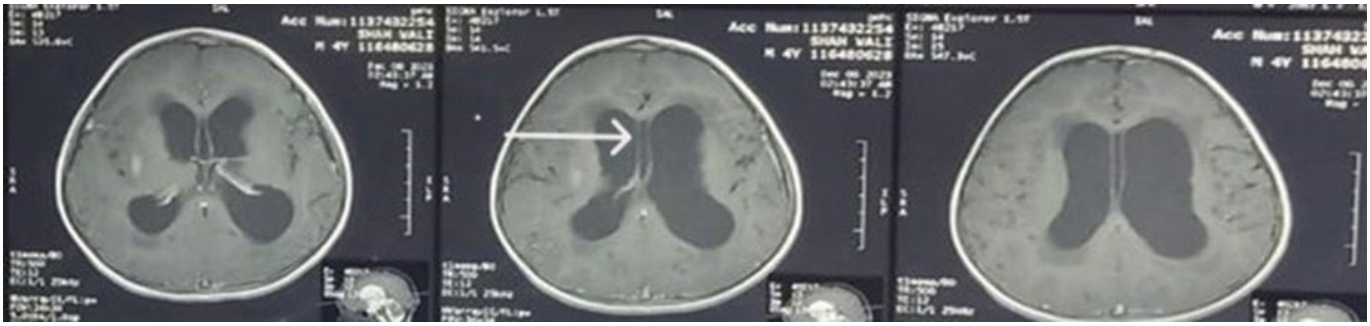

Fig. (4): T1 weighted MRI axial view showing incidental finding of cavum septum pellucidum cyst (arrow) and communicating hydrocephalus.

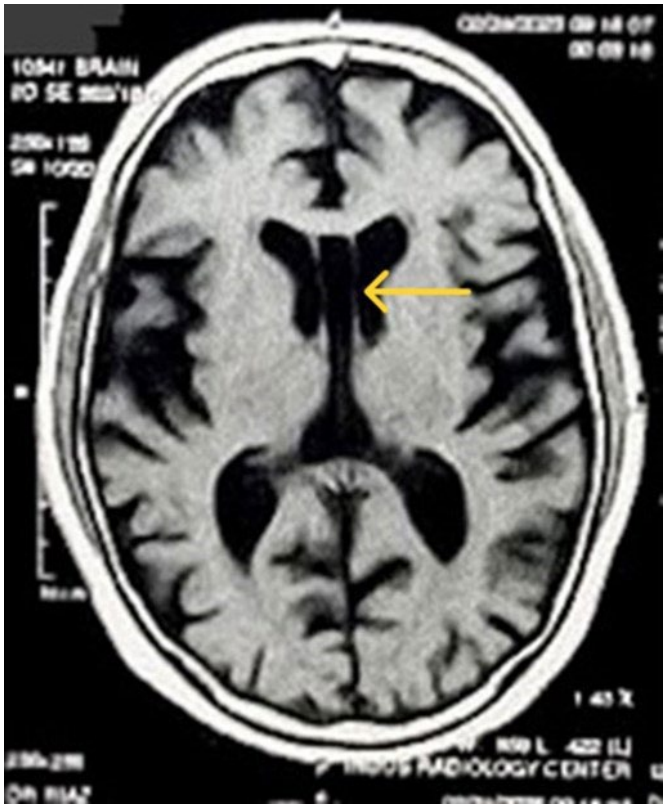

Fig. (5): T1 weighted axial MRI visualizing CSP cyst (arrow).

which revealed a communicating hydrocephalus along with CSP and CV (**Fig. 4**) he was offered decompression and ventricular shunt placement but the patient's attendants were not willing for any kind of intervention hence, conservative management with osmotic diuretics and antibiotics was suggested. Surprisingly the patient became asymptomatic on follow-up visits at the interval of one and three months respectively.

CASE 5

A 70-year-old male presented with a history of more than 2 episodes of syncope along with some degree of walking difficulty. The patient was a retired military officer who had no known comorbidities and went to multiple doctors to get all his tests done which revealed no significant findings. We performed an MRI with and without contrast which revealed a CSP cyst (**Fig. 5**). The patient was advised observation as he was uncertain

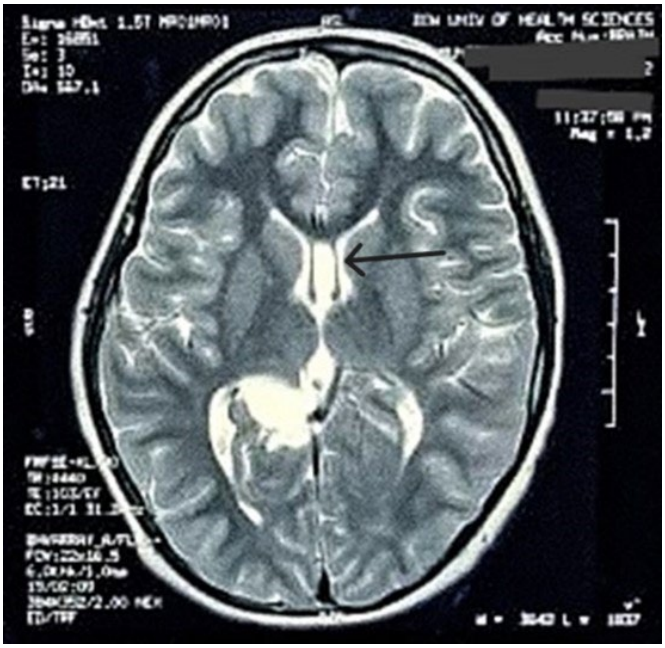

Fig. (6): T2 weighted MRI of the patient at 11 years of age showing CSP cyst (arrow) and a peculiar posterior extension.

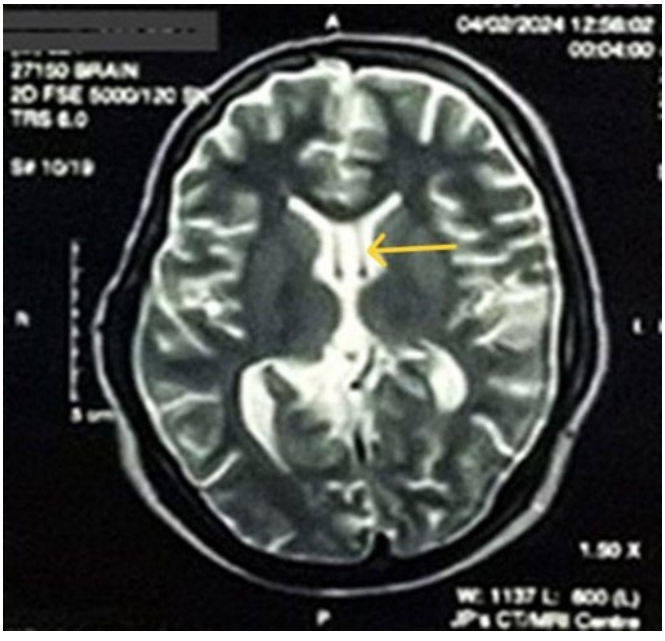

Fig. (7): T2 weighted MRI images done at 22 years of age showing CSP cyst (arrow) and its posterior extension.

about taking medications. However, he was also lost on follow-up.

CASE 6

11-year-old male patient with a history of delayed milestone development and hypoxia. Presented with sudden onset of urinary incontinence, decreasing cognition, and gradually increasing difficulty in walking. On MRI this patient also had a CSP cyst with posterior extension (**Fig. 6**) we preferred conservative management in this patient and observation was decided, the symptoms subsided as he got older and a follow-up MRI (**Fig. 7**) of the patient at 22 years of age revealed that the cyst was present but did not increase in size.

DISCUSSION

Incidental (asymptomatic) CSP cysts are considered of no harm if one comes across them on imaging. Pathological (symptomatic) CSP cysts although rare can present with a variety of symptoms, there are plenty of case reports highlighting the cause-and-effect relationship of this disease with the presentation [2, 9-12]. Although relatively easy to discern on imaging, the variety in symptoms of this disease can obstruct a diagnostician's view to look for it and if found, ruling out other causes is deemed necessary as a protocol. Over the years symptomatology of this disease has always been a topic of amusement as to how an anatomical variant not having a specific size can produce such a range of presentations, Authors have deduced surgical management as the prompt treatment of a cyst with non-specific symptoms [7, 10].

The reviewed literature elaborates endoscopic fenestration of the cyst as the surgical treatment of choice in symptomatic individuals, mainly taking adults into account, as it is believed that symptoms arise due to pressure effects of the cyst on the surrounding areas of the brain relieving the pressure will fix the symptoms. Along with that, it has also been reported that conservative management was the preferred strategy for pediatric neurosurgeons which included monitoring of ICP and radiological follow-up within 3-15 months because spontaneous regression of the cyst has been observed in pediatric patients and should be taken into consideration when dealing with a patient of that age [7-9].

The progression of a cyst is important for determining the severity of a clinical outcome, as these cysts are known to expand in size if left as they are [13], regular follow-ups and proper communication with the patient play a huge role. However, the question that needs answering is what should be the decided protocol for it. If we shine a light on the cases described above it is clear that the symptomatic presentation can be controlled through pharmacological means (Cases 1, 2, 4, and 6).

Case 4 is a prime example of the most common presentation of CSP Cyst a concomitant Hydrocephalus, a ventriculoperitoneal shunt procedure was advised but non-compliance of the patient led us to strategize with pharmacological therapeutics and the patient became asymptomatic, this could be added as a step of non-surgical intervention as a guideline for similar cases. There was an absence of deterioration of symptoms in any of the above cases excluding the ones we lost for a follow-up (Cases 3 and 5). This facilitates the idea of how important observation/monitoring can be to prevent unnecessary surgical intervention because there is evidence that such benign cystic lesions may remain of the same size even in the time frame of 10 years (Case 6).

As far as cause and effect understanding and relationship with management strategy is concerned, it is well documented in the literature that CSP cyst has two categories, one has a communication with the ventricular system while the other has no communication [14]. Hence the phenomenon of improvement of symptoms can be linked to this characteristic feature of communication with the ventricular system and size of the CSP cyst. Table **1** shows a clinical summary of the cases we have dealt with to provide a better understanding of the variation in age, symptoms, interventions, and follow-up time frame. Differentiation of this pathology from other intracranial midline cysts requires detailed radiological assistance, the peculiarity of this cyst includes its complete homogeneity and the visualization of two separate boundaries of the septum

Table 1: Clinical summary of the cases.

Cases	Age	Gender	Symptoms	Interventions	Result on Follow-Up	The Time Frame between Interventions and Follow-Ups
Case 1	20 years	Male	Tonic-clonic seizures	Sodium valproate twice daily	Asymptomatic	1 year
Case 2	13 months	Female	Tonic-clonic seizures	Levetiracetam	Asymptomatic	3 months
Case 3	16 years	Male	Headache	Ventricular diversion procedure advised	Lost on follow-up	-
Case 4	4 years	Male	Headache difficulty walking	Osmotic diuretic & antibiotics	Asymptomatic	3 months
Case 5	70 years	Male	Syncope	Observation	Lost on follow-up	-
Case 6	11 years	Male	Urinary incontinence decreased cognition walking difficulty	Observation	Asymptomatic on follow-up	11 years

pellucidum with CSF between them along with lateral bowing of its walls.

A larger study with a longer time frame will be more effective in deriving other unknown aspects of this rare incidental disease. By rule, it is worth mentioning that despite having expertise in minimally invasive interventions, resources, and advancements an asymptomatic and improving patient should avoid undergoing an unnecessary yet traumatic procedure.

CONCLUSION

Cavum septum pellucidum cyst is a rare disease of its type. Computed tomography and Magnetic resonance imaging of the brain are the main imaging modalities used to evaluate suspected disease. Most of the time it gets overlooked, clinicians usually do not consider it as an incidental finding. Although it is of significant importance to treat and monitor by interval imaging, to confirm size differences with the associated progression of hydrocephalus and cerebral edema and determine if it needs intervention. To the best of our knowledge, such cases can usually present with disconnection syndrome and seizure disorders well controlled on medications.

LIST OF ABBREVIATIONS

Gd-DTPA	Gadolinium Diethylenetriamine Penta-acetic Acid
MRI	Magnetic Resonance Imaging
CSP	Cavum Septum Pellucidum
CV	Cavum Vergae

CONSENT FOR PUBLICATION

Informed consent was obtained from the patients to use their clinical details and radiological studies for publication, for underage patients the consent was obtained from their guardians.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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AUTHORS' CONTRIBUTION

Study Concept and Design, SM, AAK, ALS; Initial Manuscript Drafting, SM.; Editing and Reviewing, SAH, MSH; Critical Review, SM. All authors have read and approved the manuscript of its publication.

REFERENCES

1. Das JM, Dossani RH. Cavum Septum Pellucidum. [Updated 2023 Apr 3]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024. Available from: https://www.ncbi.nlm.nih.gov/books/NBK537048/
2. Pillai B, Farooque U, Sapkota M, Hassan SA, Mechtler LL. Symptomatic cavum septum pellucidum cyst: a rare presentation. Cureus 2020; 12(9): e10395. DOI: https://doi.org/10.7759/cureus.10395
3. Shaw CM, Alvord Jr. EC. Cava septi pellucidi et verg æ: their normal and pathological states. Brain 19691; 92(1): 213-24. DOI: https://doi.org/10.1093/brain/92.1.213
4. Sarwar M. The septum pellucidum: normal and abnormal. Am J Neuroradiol 1989; 10(5): 989-1005.
5. Koerte IK, Hufschmidt J, Muehlmann M, Tripodis Y, Stamm JM, Pasternak O, *et al.* Cavum septi pellucidi in symptomatic former professional football players. J Neurotrauma 2016; 33(4): 346-53. DOI: https://doi.org/10.1089/neu.2015.3880
6. Alonso JR, Coveñas R, Lara J, Piñuela C, Aijón J. The cavum septi pellucidi: a fifth ventricle? Acta Anatomica 1989; 134(4): 286-90. DOI: https://doi.org/10.1159/000146704
7. Simonin A, Lind CR. Surgical management of symptomatic cavum septum pellucidum cysts: systematic review of the literature. Neurosurg Rev 2021; 44(5): 2425-32. DOI: https://doi.org/10.1007/s10143-020-01447-4
8. Bot GM, Constantini S, Roth J. Conservative treatment of cysts of the cavum septum pellucidum presenting in childhood: report of 3 cases. J Neurosurg Pediatr 2015; 16(3): 283-6. DOI: https://doi.org/10.3171/2015.3.PEDS14714
9. Tamburrini G, Mattogno PP, Narenthiran G, Caldarelli M, Di Rocco C. Cavum septi pellucidi cysts: a survey about clinical indications and surgical management strategies. Br J Neurosurg 2017; 31(4): 464-7. DOI: https://doi.org/10.1080/02688697.2016.1229743
10. Kryukov EV, Stanishevsky AV, Gavrilov GV, Gizatullin SK. Cysts of septum pellucidum, cavum vergae and cavum veli interpositi. Meta-analysis of 368 cases. Zh Vopr Neirokhir Im N N Burdenko 2020; 84(4): 111-8. DOI: https://doi.org/10.17116/neiro202084041111
11. Silbert PL, Gubbay SS, Vaughan RJ. Cavum septum pellucidum and obstructive hydrocephalus. J Neurol Neurosurg Psychiat 1993; 56(7): 820-2. DOI: https://doi.org/10.1136/jnnp.56.7.820
12. de Oliveira Ferreira DB, Medeiros JW, Cwajg E, Ferreira-Pinto PH, de Carvalho LU, Nigri F. Cavum septum pellucidum and vergae cyst: a symptomatic case with intracranial hypertension and multiple nerve involvement. Surg Neurol Int 2022; 13: 564. DOI: https://doi.org/10.25259/SNI_822_2022
13. Lancon JA, Haines DE, Raila FA, Parent AD, Vedanarayanan VV. Expanding cyst of the septum pellucidum: case report. J Neurosurg 1996; 85(6): 1127-34. DOI: https://doi.org/10.3171/jns.1996.85.6.1127
14. Van Wagenen WP, Aird RB. Dilatations of the cavity of the septum pellucidum and cavum vergae: report of cases. Am J Cancer 1934; 20(3): 539-57. DOI: https://doi.org/10.1158/ajc.1934.539