

# Anatomical Differences of Variant Intracranial Cysts

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## Abstract

Cavum vergae, cavum septum pellucidum and cavum veli interpositi are brain midline embryological developmental cysts. They are rarely seen after trauma. Generally, they do not constitute clinical findings. These cysts sometimes become enlarged and become symptomatic. Enlarged cysts cause severe neurological dysfunction. During normal fetal development, the development of the adjacent structures of the limbic system and the septum pellucidum are synchronized. It is thought that dysgenesis in these adjacent structures may affect the lamina fusion of septum pellucidum and cause cavum septum pellucidum. Anterior one of the cavities that arise when septum pellucidum laminae do not join after birth is called "cavum septum pellucidum" (CSP). The one in the posterior is called "cavum vergae" (CV). Velum interpositum (VI) is a potential cavity below the corpus callosum splenium and sometimes presents as a cyst. Cavum veli interpositi (CVI) is located in the pineal area, below the columna fornicis and above the tela choroidea of the 3rd ventricle. Because of its rarity, the incidence of CVI cyst is uncertain. The CV obliterates from the front to the front and is seen with the cavum septum pellucidum. CSP and CV are cavities that are present in fetal life but are considered as variant intracranial defects that do not close 6 months after birth. CVI is rarely seen embryo logically in fetal life in children over 2 years and adults. These cysts are rare in adults. In this study, anatomical features and clinical reflections of CSP, CV and CVI were reviewed.

**Keywords:** Cavum septum pellucidum; Cavum vergae; Cavum veli interpositi; Lamina septi pellucidi; Septum pellucidum

## Introduction

CSP, CV and CVI, which are embryological closure defects, are highly variant and rare midline lesions. The treatment, diagnosis and development of these cysts are still not fully elucidated.<sup>[1-3]</sup> Often these cysts do not matter in the clinic<sup>[4,5]</sup> When we look at the literature, a few cases of symptomatic septum pellucidum enlargement is encountered which are treated.<sup>[1,3,5,6]</sup> Septum pellucidum (SP) is a thin compartment consisting of two thin laminae which is a compartment stretched between the concave face of the front half of the corpus callosum which forms the medial wall of the lateral ventricle and the right-left columna fornicis.

### Lamina of Septum Pellucidum

Each lamina of the chamber is called "lamina septi pellucidi" and on the 6th month of fetal development, the posterior half of these two laminates is completely fused, but the anterior half joins in the 3rd-6th month after birth.<sup>[7]</sup>

When the septum pellucidum laminae do not join, a cavite image appears. Called the cavum septum pellucidum, this cavity is associated with some psychotic disorders such as tourette syndrome and schizophrenia. CSP, which was first described by Franciscus De Le Boe in 1671, was seen as an abnormal element of the ventricular system and was defined as the 5th Ventricle. The embryological development of Septum pellucidum is synchronous with the development of other neighboring limbic

system elements. Therefore, embryonal development defects in other adjacent limbic structures should be considered in the closure defects of the septum pellucidum.<sup>[6-8]</sup>

The hippocampus and corpus callosum grow during fetal development and push the lamina septi pellucidi forward to close from the posterior to the anterior. The malformation of these limbic system structures adjacent to the septum pellucidum is thought to cause CSP by disrupting the laminar junction of the septum pellucidum.<sup>[7,9,10]</sup>

Franciscus De Le Boe first described CSP as an abnormal component of the ventricular system in 1671 and named CSP the fifth ventricle. The CV, first described by the Italian anatomist Vergae in 1851, was later considered the sixth ventricle.<sup>[2,6]</sup> However, the definition of ventricles in both cavities is incorrect<sup>[6]</sup>. Astrocytes, glial fibers and residual arachnoidal cells form the walls of these cavities.<sup>[2,6]</sup> The source of the liquid in the CV and CVI cavities is uncertain. According to some researchers, arachnoidal cells form the source of the fluid.<sup>[11]</sup>

CSP and CV are basically the same structures, and one is in

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front of the other behind the imaginary vertical line made by the column of the fornix. CVS is in front of the imaginary vertical line, CV is behind.

### Corpus callosum

It is the most basic way of connecting the two hemispheres of the cerebrum.

It provides the cognitive, motor and sensory performance of the brain by connecting stimuli originating from the cortex to the contralateral hemisphere. The embryological development of the corpus callosum begins with the formation of primitive lamina terminalis in the fifth week of fetal life. The thickened primitive lamina terminalis forms the commissural plate and the glial cells merge to form a junction path. In the seventeenth week of fetal life, the corpus callosum becomes mature. If agenesis occurs in the development of the corpus callosum, commissural fibers cannot cross the midline. The corpus callosum has four sections: rostrum, genu, truncus, and splenium.<sup>[12]</sup>

The corpus callosum has superior and anterior gyrus cinguli and posterior is the isthmus of gyrus cinguli. In the inferior of the corpus callosum, there are septum pellucidum and lateral ventricles [Figure 1]. There are limbic system structures around it.<sup>[13]</sup> Therefore, the corpus callosum is thought to be indirectly associated with the limbic system.<sup>[14]</sup>

### Septum pellucidum

Septum pellucidum (formerly septum lucidum): It is a vertical thin chamber made of white and gray ore, both sides covered with ependym. Septum pellucidum clings to the corpus callosum above, below clings to fornix [Figure 2]. It is located between rostrum and truncus corporis callosi in the front. In fact, it is in the form of two leaves called lamina septi pellucidi. Sometimes it fuses with each other in the form of a single layer, sometimes there is a narrow gap between them called the cavum septi pellucidi.<sup>[4,5]</sup> This cavity has no connection with the ventricular cavity.

Although CVP is found in newborns, it closes in adults and not infrequently. It serves as a responsible center for the dissemination of information occurring in and out of the limbic system. In the destruction of septum pellucidum have been occurred some clinical picture.<sup>[7,8]</sup>

### Plexus choroideus ventriculi lateralis

The plexus choroideus ventriculi tertii located in the third ventricle passes through the foramen interventricular (Monro hole) and proceeds in the pars centralis and cornu temporale [inferius] of the lateral ventricles. Its surface is approximately 40 cm<sup>2</sup>. Plexus choroideus ventriculi lateralis consists of two layers of pia mater and ependymal cells lining the ventricular face. Plexus choroideus is not found in the subsequently developing cornu frontale [anterior] and cornu occipitale [posterior]. Two pia mater leaves covering the lower face of the fornix above, the ceiling of the third vent and the upper face of the thalamus below, extend through the fissura choroidea under the name of interlining choroidea ventriculi tertii and extend to the outer wall



**Figure 1:** Image of septum pellucidum and adjacent structures on plastinated cadaver (A: Anterior, P: Posterior, 1: Septum pellucidum).



**Figure 2:** Image of septum pellucidum and adjacent structures on plastinated cadaver (1: Septum pellucidum, 2: Corpus callosum genu, 3: Corpus callosum splenium, 4: Thalamus).

of the pars centralis. Meanwhile, it carries the ependymal layer that lays the ventricular cavity. The staminate between these two pia mater leaves and directed towards the lateral is called fissura choroidea, and the vein-rich structure at the lateral end of this staminate is called plexus choroideus ventriculi lateralis. According to this statement, plexus choroideus ventriculi lateralis is a vein-rich cauliflower structure consisting of pia mater and ependym cells.

The veins are added to the plexus choroideus ventriculi lateralis between the two pia mater leaves (fissura choroidea). A. choroidea anterior, a branch of a. carotis interna, is inserted into the plexus at the tip of the cornu inferius. A. choroidea posterior, branch of a. cerebri posterior, is enters to plexus passing under the splenium corporis callosi. Its veins (v. choroidea) show a meandering course. The v. cerebri interna on both sides merge into the midline and v. creates magna cerebri. This vein opens

to the place where the sinus sagittalis inferior meets the sinus rectus.<sup>[15]</sup>

### **Cavum septum pellucidum and cavum vergae**

In the anterior part of the cerebrum, there are two potential cavities in the midline. These are CSP and CV. These cavities are formed in the median line by septum pellucidum leaves during cerebrum morphogenesis. Septum pellucidum is a thin membranous structure consisting of two transparent laminar layers. Different views have been proposed regarding the embryogenesis of CSP and CV.<sup>[12]</sup>

### **Literature Review**

Some authors suggest that the third month of pregnancy occurred as a result of the rapid expansion of the lamina terminalis,<sup>[16]</sup> while a group of researchers suggested that the formation of the interhemispheric fissure did not merge,<sup>[17]</sup> but the formation mechanism has not been clarified yet. Embryological development of CV and CSP; It depends on the embryological development of SP. Development of SP 10-12 of gestation begins in the weeks. It reaches adult form with corpus callosum in the 17th week of gestation. SP, which has a solid structure in the early stages of fetal life, later turns into a cavum as a result of interstitial cleavage of the “commissural plate”. The prevalence of CSP decreasing with age decreases in all preterm babies. The pellucidal leaves that make up the CSP merge and its top remains towards the occipital lobe and its base remains a septum directed towards the frontal lobe. With this junction, the cavum disappears, the triangular septum pellucidum remains.

SP is a transparent structure containing glial cells, nerve fibers and plexus choroideus related veins. It extends from the lamina terminal to the splenium of the corpus callosum. SP occurs by adhesion of two membranous laminates.<sup>[18]</sup>

Each lamina is located in the central part of the lateral ventricles and in the medial wall of the cornu anterior. The inner face of the lamina leaves is covered with pia mater and the ventricular face with the ependymal layer. SP is fed by the anteromedian ganglionic branches of a. cerebri anterior. For the first time in 1671, the anatomist named slyvus described the expansion between the SP leaves. CV was named Vergae because it was described by Andrea Vergae in 1851. CV is an expansion just behind CSP.

It is usually found with CSP and has been proposed as a posteriorly located CSP for some time. Considering that there is an imaginary vertical line made by columna fornicis, the cavity remaining at the anterior of the vertical line is CSP and the cavity remaining on the posterior side is CV. CSP and CV are actually formations with the same structure. There are corpus callosum genu in the anterior of the CSP, the forearm of the fornix in the posterior, the truncus of the corpus callosum, the rostrum corpus callosum and commissura anterior in the inferior, and the lamina septi pellucidis in the lateral.

The superior of the CV includes the truncus corpus callosum, the anterior legs of the fornix in the anterior, the splenium

corpus callosum in the posterior, and the residues on the tela choroidea and the commissura hippocampi in the inferior. Schunk found the width dimensions of CSP as 2 mm, 3 mm, and 5 mm. Schwidde, which calculates CSP dimensions as vertical 7.5 mm and transverse 3.6 mm; When CV and CSP are available together, they calculated their average dimensions as CV: 25 mm, CSP: 6.9 mm.

The content of the liquid in CSP and CV is a controversial issue. Because, although the cavities have no relation with the ventricular system, it has been determined that they can be connected to the ventricles with the windows formed for different reasons. Both cavities have connections with each other, the name of the connection is the aqueducta septi. They rarely differ from each other with columna fornicis. According to some circles, the structure of lamina leaves is suitable for communication with the ventricles. According to some circles, they think that this layer can secrete a liquid since the cell layer of the lamina leaves is covered with arachnoid and pia residues.

Septum pellucidum should not be considered as a stretched thin membranous structure only between the fornix and corpus callosum. It should be considered as an important element of the limbic system, as it is a part that transfers information between the hypothalamus and the hippocampus.<sup>[7]</sup>

### **Cavum veli interpositi**

The velum interpositum cavity is a subarachnoid space that is associated with fornix and fornix's respective plexus choroideus. This plexus choroideus forms roof of III. ventricle from the back.<sup>[19,20]</sup>

Cavum veli interpositi is a potential space between the double layer interlining choroidea layers of the third ventricle.<sup>[19]</sup>

If this gap is enlarged, then it is called as the cavum veli interpositi. Sometimes, this potential space is filled with CSF and can be seen sonographically as an interhemispheric cyst. Dilatation of this area with fluid accumulation creates a midline cyst above the apex of the third ventricle, forward and below the splenium of the corpus callosum, in front of the pineal gland.<sup>[19,21]</sup>

The incidence of CVI cysts is uncertain because of its rarity, and the results of affected individuals are variable.<sup>[22,23]</sup>

Contrary to rare pre-natal diagnoses, 3% of children up to 2 years of age had CVI cysts.<sup>[22-24]</sup>

When diagnosed before birth, these cysts are usually harmless, as they do not disrupt the brain parenchymal structures.<sup>[21]</sup>

However, attention should be paid to the excessive expansion of CVI cysts, since their increase in size may cause the foramen monron to close, thus causing hydrocephalus by partially blocking the flow of cerebrospinal fluid.<sup>[22,24,25]</sup>

Laroche and Baudey showed that CSP and CV are present in almost all newborns born with normal brain structures and that midline formations start from CV and close from back to front.<sup>[26]</sup> It is very rare to see CV and CSP together. While CV begins

to close in the 6th month of gestation, CSP starts to close in the eighth month. The brain midline structures begin to close from the posterior to the anterior. Compared to CSP, the CV in the rostral disappears earlier.<sup>[26,27]</sup>

As a result of pathological studies, CV prevalence was found to be 100% in 6 month old fetuses and 40% in newborns born on time. CV is usually seen with CSP. It is a rare condition to see CV alone. Insulating CV was not found in 1032 autopsy studies. CV ratios in the brain CT examination involving 1050 children aged 0-14 years by Nakona et al. <1 year was 0.6%, 1% in 1 year old, 0% in 10-14 years old. CV rate was determined as 0.4% at all ages. The rate of CSP was 97% in the first week in a normal newborn, 85% in the first 30 days, 45% in sixty days, 15% in the 3-6 months period, and 12% under the age of 16. Fast adhesion in the 2nd and 6th months after birth is remarkable.<sup>[28]</sup>

According to the CSP definition made by Shaw and Alvord; although the septum is defined as a space with a minimum distance of 1 mm or more from the top and bottom surrounded by two lamellar leaves, in some studies its evaluation as cavum in the cavities as much as the pinhead causes variation in the determination of CSP prevalence.<sup>[16]</sup>

While the incidence of CSP was 12% in 374 patients without neurological disorders between 6 months and 16 years; In the study carried out by Schwidde in 1952, the incidence of CSP was calculated as 20.34% in the study in which the human brain belonging to 1032 different age groups was examined. Shunk, on the other hand, found this rate as 60.2% in his study on 307 neurological disorders.<sup>[29]</sup> The reason for such a big difference in the autopsy study results is that the measurement parameters in the cavum areas are handled differently. However, there are differences not only in autopsy studies but also in cavum measurements made with imaging methods. While CSP prevalence ranged from 0.1% to 0.4% in a cavum calculation study with a PEG.

While CSP prevalence ranged from 0.1% to 0.4% in a cavum calculation study with a PEG,<sup>[29]</sup> in another imaging study, CSP prevalence was calculated between 0.21% and 1.7%.<sup>[30]</sup> In the CT examination study conducted by Nakona et al. on 1050 cases, the CSP prevalence was calculated as 3.9% under one age and 2.2% in all age groups. Sarwar found this rate as 3.3% in his study and determined that 1.6% of 633 cases had CSP and CV at the same time.<sup>[7]</sup>

CSP, which was first considered as a ventricular system element in 1671, was defined as the 5th Ventricle by Franciscus De Le Boe.<sup>[6]</sup> The CV was first described by the Italian anatomist Vergae and considered the 6th Ventricle.

The wall cavities of CSP and CV are composed of astrocytes, arachnoidal cell residues and glial fibers.<sup>[2,6]</sup> Therefore, the 5th and 6th ventricle definitions; It is not correct since CSP and CV walls do not contain ependymal and plexus choroideus cells.<sup>[6]</sup>

In some sources it has been suggested that the source of the liquid in CSP and CV is now arachnoidal cells.<sup>[11]</sup>

There is no clear concept for CSP and CV naming. Although

some scientists call ‘‘CSP and CV’’ as cysts, according to some scientists, they define CSP and CV as an enlarged cavite.<sup>[1,5,6,11,31]</sup>

It has been reported in the literature that CV is less common than CSP. It has been reported that CV cyst is usually seen with CSP.<sup>[1,5,16,32]</sup> The number of CV cyst cases alone is quite low in the literature.<sup>[2,11,33]</sup> CVI is a condition where the cisterna velum interpositum is dilated.<sup>[19]</sup> This cavum cyst usually occurs in newborns. CVI, which is expected to close after the age of two; Its incidence is 3% before 2 years of age.<sup>[22-24]</sup>

These midline cysts mentioned usually do not have any symptoms, so no treatment is given. In studies conducted, cases where midline cysts are symptomatic can be listed as convulsive disorders, growth and development retardation, and some neurological findings.<sup>[28]</sup>

Treatment of symptomatic cysts; if another pathological condition is not accompanied by opening a cyst wall and shunting.

In complicated cases leading to hydrocephalus attention should be paid to the obstruction in CSF flow and this should be considered in treatment.<sup>[34,35]</sup>

## Results and Discussion

The embryological development of CSP is closely related to the corpus callosum (CC). The absence of CSP is associated with an extremely broad spectrum of neuroanatomic malformation: these are hydranencephaly and deadly alobar holoprosencephaly; potentially serious but not fatal schizencephaly, porencephaly, basilar encephaloceles, and severe hydrocephalus.<sup>[36,37]</sup>

Routine neuropsychiatric tests should be performed in patients with CSP lesions.

Because neuropsychiatric diseases can remain hidden unless appropriate tests are performed. During embryological development, CV and CSP formed in the midline of the brain between the SP leaves can be visualized with today’s imaging techniques. While these cavities can be imaged with ultrasound in newborns; are the spaces that can be detected by CT and MRI in the following periods. CSP, the prevalence of which decreases with age, is present in all premature. It is very rare to see CV alone. The prevalence of the CV seen with CSP is 3-18%. The prevalence of CSP decreases rapidly by 85% in the first month and the first 6 months in the next 6 months, decreasing to 15% and 12% in adolescence. Although CV and CSP were named as the 5th and 6th ventricles, it was understood that they had a different embryological structure than the ventricles in later embryological studies. These midline cavities are associated with important parts of the limbic system. It has direct and indirect connections with hypothalamus, hippocampus, gyrus cinguli, frontal cortex, brainstem reticular formation.<sup>[38-40]</sup>

## Conclusion

Cavums, which are potential cavities, must have a significant expansion in order to be accepted as a cyst. Generally, CV and CSP do not produce findings, they are detected by chance. In the

clinic, these cysts usually manifest with emotional behavioral disorders. Pathological and asymptomatic cysts, which are divided into two, are asymptomatic, 1% observed in all cranial CT images. Pathological ones are much rare and commonly occur with cerebral diseases. Shunt operations are often used in treatment. According to some researchers, the appearance of CV in adults without CSP is one of the brain midline developmental defects. Clinical variations of CV and CSP and their relationship with neuropsychological diseases are still under investigation.

### Competing Interests

The authors declare that they have no competing interests.

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