Cavum septum pellucidum

AKA fifth ventricle, among others. The septum pellucidum is a membrane that separates the two lateral ventricles. Embryologically starts as two thin parallel leaves with fluid between them (the cavum septum pellucidum (CSP)) that usually fuse together by birth or shortly thereafter. Thus, CSP is present in ≈ all preemies. It is found in ≈ 10% of the adult population. Persistence of the CSP can be a normal variant, representing an asymptomatic developmental anomaly. However it may also indicate disruption of neurodevelopment and has been associated with neurodevelopmental and psychiatric conditions including bipolar disorder, Tourette’s syndrome, obsessive-compulsive disorder, and schizophrenia, among others. CSP may also result from trauma possibly as a result of tears in the membrane, and has been associated with TBIs especially in professional boxers suffering from chronic traumatic encephalopathy, and motor vehicle trauma.

Patients with large CSPs

The compartment is usually isolated, although some communicate with the third ventricle.

Cysts of the cavum septi pellucidi (CSP), cavum vergae (CV) and cavum velum interpositum (CVI) are anterior midline intracranial findings which are typically incidental.

They are persistent, primitive, or acquired, midline structures, fluid-filled, generally communicating located between the third ventricle and corpus callosum.

It is sometimes called the fifth ventricle.

Nonneoplastic cysts of the septum pellucidum are of two general forms. The first, the asymptomatic cavum septum pellucidum, is not a malformation or a true cyst and has been recognized since the time of Sylvius.

Embrologically the leaves of the septum pellucidum enclose a cavity, the cavum septum pellucidum. This space first appears during the 3rd month of intrauterine growth by secondary cleavage of the banks of fusion of the cerebral hemispheres.

Epidemiology
A CSP is present in the normal fetus, but over 85% of them fuse by 3-6 months of age meaning that a CSP persists in ~15% of the adult population.

**Gross anatomy**

The CSP commonly occurs, and is often confused with the cavum vergae, which is situated posterior to the anterior columns of the fornix. During development, these spaces obliterate postero-anteriorly - the cavum vergae followed by the cavum septum pellucidum - and it is not uncommon that both occur together.

**Boundaries**

- **anterior:** genu of the corpus callosum
- **superior:** body of the corpus callosum
- **posterior:** anterior limb and pillars of the fornix
- **inferior:** anterior commissure and the rostrum of the corpus callosum
- **lateral:** leaflets of the septum pellucidum

**Related pathology**

There have been various reports of their association with many behavioral and psychiatric disorders. Infrequently, they have been associated with an obstructive hydrocephalus-like picture. Although the structure and management of CSP has long been known, it has been an enigma as far as functional significance and management indications are concerned.

An absent cavum septum pellucidum in antenatal imaging is a concerning feature and is associated with significant CNS anomalies.

It is frequent among athletes with a history of repeated traumatic brain injury (TBI), such as boxers. Few studies of CSP in athletes, however, have assessed detailed features of the septum pellucidum in a case-control fashion. This is important because prevalence of CSP in the general population varies widely (2% to 85%) between studies. Further, rates of CSP among American pro-football players have not been described previously.

A retrospective study assessed retired American pro-football players presenting with cognitive/behavioral symptoms in whom structural MRI was available with slice thickness ≤2 mm (n=17). Each player was matched to a memory clinic control patient with no history of TBI. Scans were interpreted by raters blinded to clinical information and TBI/football history, who measured CSP grade (0-absent, 1-equivocal, 2-mild, 3-moderate, 4-severe) and length according to a standard protocol. Sixteen of 17 (94%) players had a CSP graded ≥2 compared with 3 of 17 (18%) controls. CSP was significantly higher grade (p<0.001) and longer in players than controls (mean length±standard deviation: 10.6 mm±5.4 vs. 1.1 mm±1.3, p<0.001). Among patients presenting to a memory clinic, long high-grade CSP was more frequent in retired pro-football players compared with patients without a history of TBI. ⁴
Clinical features

Only rarely do we encounter symptomatic cysts of this type. Only a quite small number of these cysts series have been published 9).

Differential diagnosis

Cavum vergae

Cavum velum interpositum.

Treatment

Symptomatic patients usually present aspecific symptoms. For this reason, the management of these patients is still debated.

TCD in the absence of other objective confirmatory studies, can aid in the diagnosis and provide information about the success of fenestration of the cavum septum 6).

Results suggest that in most of the patients with aspecific symptoms, clinical observation and eventually ICP monitoring are adequate to identify patients for surgery 7).

(1) Endoscopic fenestration of symptomatic CSP cysts is a safe treatment option. (2) Neurocognitive assessment is essential in the evaluation and outcome assessment of CSP 8).

Case series

In a retrospective study of 10 patients treated at 2 clinics between 2002-2018. 9 patients underwent surgery and 1 is under long-term monitoring. Apart from demographic data, the study analyzed symptoms, cyst size and progression over time, ventricle size, complications, and treatment modality.

CSP with CV was found in 8 cases with 1 case each of CSP and CVI. The study comprised 6 men and 4 women, including 4 children. The mean follow-up time was 43.4 months. The average cyst size was 20.4 mm in CSP and 19.8 mm in CV; the CVI was 33 mm. Headache was most commonly reported (70%) followed by behavioral disturbance (30%). Disturbance in memory, psychomotor development, school performance, visual acuity, and vomiting was variously noted in 20%. The prevailing symptom was headache in adults and behavioral and autonomic disturbance in children. Postoperatively, cysts had reduced by an average of 44.3% while the ventricles remained unchanged. Symptoms resolved in all cases with residual problems in patients presenting with memory loss. No complications were noted.

Endoscopic fenestration is the method of choice in the treatment of symptomatic midline cysts. We recommend that any further research focuses on precisely establishing their clinical presentation, particularly neuropsychological symptoms 9).
A retrospective analysis of 3 patients who underwent endoscopic fenestration for CSP with obstructive hydrocephalus between 2012 and 2014 was done in the Division of Paediatric Neurosurgery, Department of Neurosurgery, Amrita Institute of Medical Sciences and Research Centre, Kochi, India.

Data were analyzed for symptomatic clinical improvement in particular behavior.

Pre- and postoperative brain MRI showed a significant decrease in the size of the cyst as well as the ventricles. There were no recurrences during follow-up. All of the patients improved.

(1) Endoscopic fenestration of symptomatic CSP cysts is a safe treatment option. (2) Neurocognitive assessment is essential in the evaluation and outcome assessment of CSP.

Case reports

1997

The authors describe the morphological, histological and histo-immunological characteristics of an additional case of septum pellucidum-cavum Vergae cyst in a forty-year-old man who died the day following a ventriculo-peritoneal shunt.

1996

A giant CSP and CV cyst in an 18-month-old boy, extending to the posterior cranial fossa and causing hydrocephalus. The literature is reviewed, and the MRI and CT findings of the case are reported.

References

1) Dandy WE: Congenital cerebral cysts of the cavum septi pellucidi (fifth ventricle) and cavum vergae (sixth ventricle). Diagnosis and treatment. Arch Neurol Psychiatry 25: 44–66,1931
6) Bell RS, Vo AH, Dirks MS, Mossop C, Gilhooly JE, Cooper PB, Razumovsky AM, Armonda RA.


