Maldeveloped septum pellucidum associated with schizophrenia: a case report

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Abstract
The body of a patient of schizophrenia was donated to the Medical College. During routine dissection a small anteriorly placed cavum septum pellucidum was observed along with fenestration of septum pellucidum posteriorly. Also, the lateral ventricles were dilated, foramen of Monro was absent and the fornix was caudally displaced. The article discusses the correlation between schizophrenia and septum pellucidum variations from point of view of embryological development as well as points to a possible infective or traumatic causation.

Key words [septum pellucidum] [cavum septum pellucidum] [schizophrenia] [corpus callosum] [ventricle]

Introduction
The septum pellucidum (SP) is a thin sheet of nervous tissue consisting of both white and gray matter. It is a double membrane with a closed slit like cavity between the membranes. This cavity in the SP has been widely regarded in autopsy series as an incidental finding of little clinical importance. Cavum septum pellucidum (CSP) is a frequent finding in premature infants and its high incidence has been found in patients suffering from schizophrenia spectrum disorder [1]. It has also been found to be relatively frequent in boxers or cases of head trauma [2].

The SP is partly or completely absent in 2 or 3 per 100,000 individuals. This phenomenon rarely occurs in isolation, usually being associated with additional malformations of brain such as agenesis of corpus callosum, hydrocephalus, porencephaly or hydranencephaly [3].

Case Report
A 53-year-old female cadaver with a history of schizophrenia was donated to the Medical College by a local Mental Hospital. The cadaver was preserved in 10% formalin. During routine dissection variations were observed in the SP.

In a coronal section taken at the level of optic chiasm, SP showed two laminae with a central CSP (Figure 1). Subsequent coronal sections showed malformed SP. The SP was present in the form of thin fibrous tags, which at places were attached only to the corpus callosum above, or only to fornix below and most posteriorly attached both to corpus callosum and fornix in the form of clearly visible fibrous strands. The SP at the level of splenium of corpus callosum appeared in the form of fine bridge like strands (Figures 2, 3).

The two lateral ventricles communicated with each other across the midline through these fenestrations in the SP. The lateral ventricles were found to be dilated. The foramen of Monro was absent. The fornix was observed to be caudally displaced.

In all sections, the corpus callosum was well preserved. No variations were observed in the rest of central nervous system. Optic chiasm and optic tracts were well developed.

Discussion
The SP is a thin double membrane separating the two lateral ventricles stretching between the fornix and corpus callosum. Anteriorly it joins the rostrum with body of corpus callosum, inferiorly and posteriorly attached to the superior part of the fornix. If the 2 thin membranes are not in perfect apposition, or their attachments to either corpus callosum or fornix are not at the same point, a cavity results called as CSP. ‘Cavum vergae’ is the posterior drawn out portion of the CSP. SP consists of deep, midline, limbic structures [4, 5].
Understanding of development of SP is essential to understand the pathophysiology of the associated schizophrenia. Lamina terminalis is that part of the neural tube which closes the cranial end of the prosencephalon. The corpus callosum develops in the thickened lamina terminalis. The remains of lamina terminalis, between corpus callosum and the fornix become stretched to form a clear membrane, the SP. At twelfth week of gestation, a space forms between two laminae called CSP. At 20th week of gestation, the laminae fuse from posterior to anterior direction as a consequence of growth of corpus callosum and limbic system structures. The CSP fuses by age of 6 months. Altered central nervous system development in these regions such as dysgenesis of either hippocampus or corpus callosum could arrest the posterior to anterior fusion process, resulting in preservation of the CSP in adulthood [6,7].

Van Wagenen and Aird stated that while all fetuses and newborns showed presence of CSP, it was observed in only 4% of adult males and 9% of adult females [5] in an MRI study, detected CSP in 68% of healthy adults [8]. This difference in percentage of incidence reported by the two studies can be explained by the differences in methods of investigation [6]. Kim and Peterson state that "normal CSP" is not one that is completely closed but one that ranges around a size that is reasonably small [6].

In present case, there is coexistence of small anteriorly placed CSP and SP present in the form of thin fibrous tags posteriorly, in a patient with history of schizophrenia (Figures 1, 2, 3). Trzesniak et al. noted that a significantly higher incidence of large CSP was seen in cases suffering from schizophrenia spectrum disorders, while a small CSP could be considered as a normal neuroanatomical variation [1]. Liao et al. also observed an abnormally enlarged CSP with a cavum vergae in a case of resistant childhood-onset schizophrenia [7]. On the other hand, Kim and Peterson state that the CSPs in Tourette syndrome (inherited neuropsychiatric disorder)
are smaller than in controls and that smaller CSP could arise from disturbances in the development of CSP such as from its premature closure or failure of corpus callosum to develop properly [6].

While CSP is widely regarded as an incidental finding of little clinical importance, Lewis and Mezey observed a statistical association between CSP and functional psychosis in their study on psychiatric and neurological cases [9]. It was found that CSP might signal an underlying abnormality of function relevant to the pathogenesis of these illnesses. Similarly, though in the present case, the CSP was small, it was found in association with symptoms of schizophrenia.

Bogdanoff and Nater report that out of 9 men detected to be having CSP on CT scans, 6 were either boxers or had history of head trauma. They note that CSP may not essentially be a congenital variant but could also be an indicator of prior head trauma [2].

The phenomenon of absent SP rarely occurs in isolation [3]. It is usually associated with malformation of brain like septal dysplasia, agenesis of corpus callosum, porencephaly and hydranencephaly. Thus its absence could be a valuable due to the diagnosis of other malformations of the brain. Agenesis of SP has been associated with epileptic seizures [10]. Supprian et al. described a case of schizophrenic psychosis where isolated absence of SP was the only finding on MRI [3]. Since septal area is a part of limbic system, its pathology is said to play a key role in the development of schizophrenia.

Agenesis of SP is a part of symptoms seen in De Morsier’s syndrome (septo-optic-pituitary dysplasia) where patient may have absent SP, optic nerve hypoplasia and pituitary insufficiency [11]. However in the present case there was no such history of symptoms of pituitary illness or finding of any optic nerve anomaly.

Alcohol exposure also contributes to SP abnormality. Filipovic et al. state that CSP could be a marker of long-term alcohol abuse which causes demyelination due to the lipolytic effect of ethanol [12]. Partial or complete absence of SP is either a part of a ventral induction disorder or it can be due to a disruptive process leading to occurrence of remnants of SP [13].

Fenestration of SP has been reported by Cavalli et al. as a complication of hydrocephalus [14]. The authors state that obstruction of foramen of Monro causes pressure of cerebrospinal fluid to rise causing the fenestration. Since the foramen of Monro was absent in the present case and the lateral ventricles were also found to be enlarged, this could be the pathophysiologic mechanism behind the fenestration. Chun et al. also reported two autopsy cases that revealed partial absence of the SP with ventriculomegaly as seen in present case [15].

**Conclusion**

In the present study the patient was a case of schizophrenia whose body was donated to the medical college for dissection where SP variations were observed along with dilatation of the lateral ventricles and absence of foramen of Monro. The article discusses the correlation between schizophrenia and SP variations from point of view of embryological development as well as points to a possible infective or traumatic causation. Besides endorsing the correlation between SP variations and schizophrenia, the present case also can help a medical student develop better understanding of details of central nervous system development and the effects of any maldevelopment here on the well being of the human being finally formed.

**References**


