

ORIGINAL RESEARCH PAPER

PAEDIATRIC

CAVUM SEPTUM PELLUCIDUM IN VARIOUS NEUROLOGICAL DISORDERS AMONG CHILDREN - A DESCRIPTIVE STUDY

KEY WORDS: septum pellucidum, cavum pellucidum.

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OBJECTIVE: To study the association of persistence of cavum septum pellucidum in various neurological disorders among children above one year of age attending pediatric neurology outpatient department of Stanley Hospital.

DESIGN: Hospital based descriptive study.

SETTING: Paediatric Neurology Department of Stanley Hospital.

PARTICIPANTS: 40 children between age 1 and 12 years presenting to Paediatric Neurology outpatient clinic, Stanley Hospital. **PROCEDURE:** Children who had undergone MRI Brain study for their clinical condition and had findings consistent with cavum septum pellucidum (CSP) were included in the study. Detailed history, clinical examination and diagnosis were noted and analyzed for significant association.

RESULTS: cavum septum pellucidum was more commonly associated with seizures (67%) followed by developmental delay (16%) and breath holding spells (16%). There was no significant association with behavioural abnormalities.

CONCLUSION: The common embryological origin of the septum pellucidum, corpus callosum and limbic system explains the anatomical basis for their concurrent involvement in disease processes. Thus CSP can be considered as a possible marker for limbic system dysgenesis.

INTRODUCTION:

The Septum Pellucidum is a thin, vertically placed partition consisting of two laminae separated in the greater part of their extent by a narrow interval called the cavity of the septum pellucidum. It is a part of the limbic system and plays an important role in the linkage between the hypothalamus, hippocampus, amygdala, habenula, and brainstem reticular formation. The septum pellucidum extends between the anterior portion of the corpus callosum and the body of the fornix [3]. Thus the septum pellucidum is a possible regulatory key station between the hippocampus and the diencephalon. Very early in development, the septum pellucidum is solid, then it cleaves to form a fluid-filled cavity along its length, the cavum septi pellucidi. In normal foetal development, these two leaflets fuse back together near the time of birth. Thus when the two leaves of the septum pellucidum fail to fuse as the fetal brain matures, so called cavum septi pellucidi (CSP) results [3]. CSP is a normal variant which is usually asymptomatic. Cavum septum pellucidum is present in 100% of foetuses and premature infants, but the posterior half of the leaves are normally fused by the age 3-6 months. [1-2] Persistence of CSP is seen as a part of a wide spectrum of abnormalities of the central nervous system. The presence of a CSP later in life might reflect developmental abnormalities of the structures bordering the septum pellucidum, such as the corpus callosum and hippocampus. Thus, the CSP can be considered a marker of limbic system dysgenesis $^{\scriptscriptstyle{[5,\ 6]}}$. CSP is associated with schizophrenia [4,8], post-traumatic stress disorder, traumatic brain injury, chronic head ache, Cognitive defects etc.

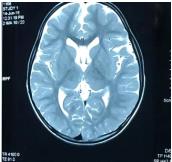


IMAGE 1: T2 sequence showing cavum septum pellucidum.

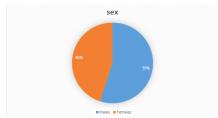
METHODS:

This is a descriptive study which was conducted over a period of 1 year from March 2017- March 2018 after obtaining Ethical committee approval. It was conducted at the out-patient department of Paediatric Neurology department of Stanley Hospital. Children attending pediatric neurology OPD between 1-12 years of age with MRI brain finding consistent with that of cavum septum pellucidum were included in the study. Infants and those who did not consent to the study were excluded. Children who had undergone MRI Brain study for their clinical condition and have findings consistent with cavum septum pellucidum (CSP) were included in the study. Detailed history, Clinical examination and diagnosis of each patient was noted. The complete data collected was transferred into a master chart which was then subjected for analysis. Association between Cavum Septum Pellucidum (CSP) and various neurological disorders were studied from the data collected.

RESULTS:

Out of 40 children who participated in the study, 55% (n=22) were boys and 45% (n=18) were girls. Majority of them presented with seizures 67% (n=26), while others presented with developmental delay 16% (n=7) and breath holding spells 16%(n=7). At birth, 90% (n=36) were term infants. 70% (n=28) had an uneventful birth history while 30% (n=12) had birth asphyxia. There was no association found between CSP and chronic headache, movement disorders or autism in our study population. 35% (n=14) were found to have microcephaly. In addition to CSP, 35% (n=14) had additional MRI abnormalities such as corpus callosal agenesis, signal abnormalities etc.

FIGURE 1: Gender distribution in the study group



Males:22, Females: 18

FIGURE 2: Age distribution in the study group

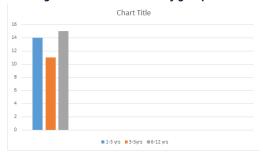
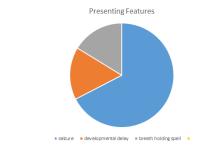
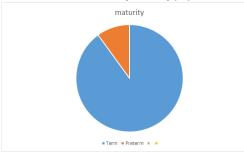


FIGURE 3: Presenting features in the study population



Seizure: 26, Developmental delay: 7, Breath holding spell: 7

FIGURE 4: Gestational maturity in study population



Term: 36, Preterm:4

DISCUSSION:

The septum pellucidum refers to a thin, vertically placed partition consisting of two laminae separated in the greater part of their extent by a narrow interval called the cavity of the septum pellucidum or cavum septum pellucidum. It forms the medial wall of lateral ventricles, extending from the lamina terminalis to the splenium of corpus callosum. The lateral aspect is triangular in shape, with its base lying anteriorly and apex lying posteriorly. Cavum septum pellucidum is also known as the fifth ventricle, as it is filled with cerebrospinal fluid. But the cavity has no connection with the leptomeningeal or ventricular spaces. Very early in development, the septum pellucidum is solid, which then cleaves to form a fluid-filled cavity along its length, the cavum septi pellucidi. In normal foetal development, these two leaflets fuse back together near the time of birth starting from the posterior part at around the 6th month of gestation. It remains as a slit but usually closes completely around the 2nd month as the brain develops.9 Thus when the two leaves of the septum pellucidum fail to fuse as the fetal brain matures, so called cavum septi pellucidi (CSP) results [3].CSP is a normal variant which is usually asymptomatic. Cavum septum pellucidum is present in 100% of foetuses and premature infants, but the posterior half of the leaves are normally fused by the age 3-6 months. [1-2] Septum pellucidum is part of the limbic system and acts as a relay station connecting the hypothalamic autonomic system to the hippocampus, amygdala, habenula and brainstem reticular formation. 10,6. The presence of a CSP later in life might reflect developmental abnormalities of the structures bordering the septum pellucidum, such as the corpus callosum and hippocampus. Thus, CSP can be considered a marker of limbic system dysgenesis ^[5, 6]. CSP is associated with schizophrenia [4, 8], post-traumatic stress disorder, chronic head

ache, cognitive defects etc. According to our study, 26 out of 40 study subjects(67%) presented to us with seizures. out of 26 cases with seizures, one child had Tuberous Sclerosis and one child had a mitochondrial disorder. In a similar study done by Ki-Young Choi et al, 8.2% of the patient group had CSP compared to 1.6% in the study group. Our study found no association between prevalence of CSP and prevalence or symptomatology of autism. In our study, 7 out of 40 subjects (16%) with CSP presented with developmental delay, one child with developmental delay had Downs Syndrome. In a study done by John Bodensteiner et al on a group of 249 children, a similar association of (15.3%) was found between prevalence of CSP and developmental delay/mental retardation. Though CSP is more commonly associated with premature infants, as found in a study on 108 normal newborns which showed a higher prevalence of CSP with lower gestational age (69% at 36 weeks and 36% at 40 weeks)12, the prevalence was higher in term deliveries (90% where n= 36) in our study population. In the current series, no association was found between CSP and asphyxia at birth. Bryun had postulated that the reason for increased incidence of epilepsy and psychosis in epilepsy patients was probably disruption of a circuit connecting SP-fornixcingulate-hippocampal region by the cystic dilatation of the septum pellucidum.¹¹ Thus abnormalities of SP could be expected to cause symptoms like disturbance in emotions and behavioural functions of the limbic system. But our study could find only an association of 0.5% between CSP and behaviour disturbances.

CONCLUSION:

In conclusion, the patients with cavum septum pellucidum in this study were found to have seizures, developmental delay and breath holding spells as the most common presentations. Though CSP has been linked with behavioural abnormalities, such an association could not be made in our study group. We also had a higher prevalence of CSP in term neonates. further studies involving larger study population are needed.

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