Surgical management of symptomatic cavum septum pellucidum cysts: systematic review of the literature

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Abstract
cavum septum pellucidum (CSP) and cavum vergae (CV) cysts are commonly found incidentally. They are usually asymptomatic but may present with symptoms related to obstructive hydrocephalus. There is no consensus about the management of symptomatic CSP and CV cysts. We present, to the best of our knowledge, the first systematic review of the different treatment options for symptomatic CSP and CV cysts. We conducted a literature review using PubMed database, searching for cases of symptomatic CSP and CV cysts managed surgically, and published until April 2019. Preoperative characteristics, surgical procedure, and postoperative outcome were analyzed using SPSS® software (Statistical Package for Social Sciences, IBM®). We found 54 cases of symptomatic CSP and CV cysts managed surgically (34 males, 20 females, 1.7/1 male to female ratio). Mean age was 24.3 ± 20.1 years. The most common presentation was headaches (34 patients, 62%), followed by psychiatric symptoms (27 patients, 49.1%). Preoperative radiological hydrocephalus was present in 30 patients (54.5%). The most common surgical procedure was endoscopic fenestration (39 patients, 70.9%), followed by shunting (10 patients, 18.2%), open surgery (3 patients, 5.5%), and stereotactic fenestration (1 patient, 1.8%). Complete resolution of symptoms was achieved in 36 patients (65.5%) and partial resolution in 7 patients (12.7%), and symptoms were unchanged in 2 patients. The present review suggests that surgical treatment could provide resolution of the symptoms in most of the cases, regardless of the procedure performed. Although mean follow-up was short among the studies, recurrence rate was low.

Keywords Cavum septum pellucidum · Cavum vergae · Endoscopic fenestration

Introduction
cavum septum pellucidum (CSP) is a common incidental finding, defined as a midline cerebrospinal fluid (CSF) space delimited superiorly by the crus of the fornices and inferiorly by the tela choroidea of the third ventricle [1]. It is anatomically distinct from cavum vergae (CV) which is a CSF space extending posteriorly to the columns of the fornix. However, CSP and CV cysts are used interchangeably in the literature and may co-exist in many cases [1–5]. In this manuscript, we will use the terminology cavum septum pellucidum and vergae (CSP and CV) cyst. Although considered as an incidental finding by most neurosurgeons, they may present with symptoms related to hydrocephalus, like headaches, nausea or vomiting, loss of consciousness, or psychiatric disturbances [1–10]. There is no consensus about the management of symptomatic CSP and CV cysts, and various procedures (endoscopic or stereotactic fenestration, shunting, open fenestration, etc.) have been proposed [1, 3–5, 8, 11–13]. Although most of the authors report good results, there is currently no review of the literature concerning the surgical management of this controversial condition. The aim of the present study is to review the preoperative characteristics, surgical procedures, and postoperative outcome of symptomatic CSP and CV cysts treated surgically and reported in the literature.

Methods
We conducted a literature review using PubMed database, searching for cases of symptomatic CSP and CV cysts...
managed surgically and published until April 2019. We used the search term “cavum septum pellucidum” to perform the research. Three hundred and forty-five articles were found, and we considered 37 articles eligible for our study [1–35] based on the criteria that the management of a symptomatic cyst was detailed in the article. Twenty-three articles reported cases managed surgically [1–3, 6, 8, 10, 14, 15, 17, 18, 21–27, 32–35], and a total of 54 patients were finally identified. We created a database using Microsoft Excel® and SPSS® software (Statistical Package for Social Sciences, IBM®). Preoperative characteristics (Table 1) were summarized with patient gender, age, clinical presentation, and presence of preoperative hydrocephalus. Management (type of surgical procedure), clinical outcome, radiological outcome, complications, follow-up, and recurrences were also recorded (Table 2). We used SPSS® software (Statistical Package for Social Sciences, IBM®) descriptive statistics to analyze age and follow-up means and standard deviation. Frequency statistics were used to analyze gender (male/female), headaches (yes/no), psychiatric symptoms (yes/no), loss of consciousness (yes/no), seizures (yes/no), nausea/vomiting (yes/no), papilledema (yes/no), preoperative hydrocephalus (yes/no), surgical procedure (endoscopic, stereotactic, open surgery, shunting), type of endoscopic procedure (frontal, parietal, transcavum), shunt (definitive, transient, or none), complications (yes/no), clinical outcome (resolution of symptoms/improvement/unchanged/died/unknown), radiological outcome (significant decrease of the cyst/persistent enlargement/unknown), and recurrences (yes/no). We used an unpaired Student’s t test to look for significant differences in clinical presentation and outcome between male and female patients. Unpaired Student’s t test was used to look for independent variables between males and females and between children and adults and to identify independent predictors of good clinical outcome (resolved of improved symptoms), recurrence, and good radiological outcome (decrease of the cyst size on postoperative imaging).

**Results**

We found 54 cases of symptomatic CSP and CV cysts managed surgically (34 males, 20 females, 1.7/1 male to female ratio). There were 23 children and 21 adults (18 years old). Patients’ characteristics are presented in Table 1. Mean age was 24.3 ± 20.1 years. The most common presentations were headaches (34 patients, 62%), followed by psychiatric symptoms (27 patients, 49.1%). Preoperative radiological hydrocephalus was present in 30 patients (54.5%). Different surgical approaches were performed and are detailed in Fig. 1. The most common surgical procedure was endoscopic fenestration (39 patients, 72.2%), followed by shunting (10 patients, 18.5%), open surgery (3 patients, 5.5%), and stereotactic fenestration (1 patient, 1.8%). Complete resolution of symptoms was achieved in 36 patients (65.5%) and partial resolution in 7 patients (13%), and symptoms were unchanged in 2 patients. Complications occurred in 5 patients (9.1%), including 1 death. Recurrence of the cyst occurred in 2 patients (5%). Mean follow-up was 2.8 ± 4.3 months. Comparison between children (n = 23) and adults (n = 21) revealed statistically significant differences in clinical presentation. There were more headaches in adults (89%) than children (48%), p = 0.004. Psychiatric disturbances were more common in children (76%) than adults (33%), p = 0.006. The other presenting features did not statistically differ between the two age groups, neither the surgical outcome. Between males (n = 34) and females (n = 20), only loss of consciousness almost reached significance (only 8% of females, but 33% of males presented with loss of consciousness, p = 0.053). Independent predictors of good clinical outcome (resolution or improvement of symptoms) were preoperative radiological hydrocephalus (96% in the good outcome group vs 4% in the bad outcome group, p < 0.0001), presence of nausea/vomiting (27% in the good outcome vs 0% in the bad outcome group, p = 0.003), and papilledema (13% in the good outcome group, vs 0% in the bad outcome group, p = 0.04). Open surgery/shunting procedures (n = 13) were associated with poor clinical outcome (p = 0.02) compared with endoscopic/stereotactic procedures (n = 40). There were no differences between types of endoscopic or stereotactic procedures performed (frontal, parietal, transcavum fenestrations, stereotactic or endoscopic). Recurrence (n = 2) was associated with older age (mean = 52 ± 13 years old) than the group without recurrence (n = 52) (mean = 22 ± 17 years old), p = 0.016. Shunt procedures were also associated with recurrence (50% of recurrence, p = 0.027) compared with the other procedures. Good radiological outcome (decrease of the cyst size) was achieved in all patients, except one (who died). This mortality was attributed to bleeding and infection related to a surgically placed external shunt.

**Discussion**

Cavum septum pellucidum and cavum vergae cysts are potential space filled with CSF between the leaflets of the tela choroidea of the third ventricle [1]. As stated in the introduction, CSP is delimited superiorly by the crus of the fornices and inferiorly by the tela choroidea of the third ventricle [1]. CV extends posteriorly to the columns of the fornix. Finally, cavum velum interpositum (CVI) is also anatomically distinct, because it surrounds the internal cerebral veins, whereas CV and CSP lie above them. As CSP and CV cysts were mostly used interchangeably in the literature, we decided to study them together. Usually considered as an incidental finding, and mostly managed conservatively, they may however present with symptoms. Wang et al. [34] found that 22 of 54,000
| Patient | Sex | Age (years) | Clinical presentation | Duration of symptoms (months) | Preoperative hydrocephalus | Reference |
|---------|-----|-------------|-----------------------|-------------------------------|---------------------------|-----------|
| 1       | Male| 3           | Developmental delay, irritability, macrocephaly | Yes                           |                           | 1)        |
| 2       | Female| 13        | Psychiatric symptoms (eating disorder, mood disturbances, anxiety) | No                            |                           | 1)        |
| 3       | Male| 42          | Postural headaches, disorders of consciousness | 48                            | Yes                       | 2)        |
| 4       | Male| 61          | Postural headaches, disorders of consciousness, dizziness, ataxia | 2                            | Yes                       | 2)        |
| 5       | Male| 46          | Postural headaches, disorders of consciousness | No                            |                           | 2)        |
| 6       | Female| 60         | Postural headaches | No                            |                           |           |
| 7       | Male| 0.5         | Developmental delay | Yes                           |                           | 2)        |
| 8       | Male| 12          | Headaches, vomiting | 24                            | Yes                       | 3)        |
| 9       | Female| 13         | Macrocephaly, irritability | 24                            | No                        | 3)        |
| 10      | Female| 26         | Headaches, blurring of vision | Yes                           |                           | 3)        |
| 11      | Male| 4.5         | Impaired mental function with vomiting, seizures | Yes                           |                           | 20)       |
| 12      | Female| 1.9        | Impaired mental function with hydrocephalus, papilledema, paraparesis | 12                            | Yes                       | 22)       |
| 13      | Female| 2.9        | Impaired mental state with hydrocephalus and ataxic gait | Yes                           |                           | 23)       |
| 14      | Male| 6           | Behavioral disturbance with impaired gait | Yes                           |                           | 24)       |
| 15      | Male| 1.5         | Hydrocephalus and dysnea with mental changes | Yes                           |                           | 25)       |
| 16      | Male| 13          | Headaches, dizziness, behavioral disturbance | Yes                           |                           | 26)       |
| 17      | Female| 23         | Headaches, behavioral disturbance | 12                            | Yes                       | 26)       |
| 18      | Female| 19         | Epilepsy | 2                             | Yes                       | 26)       |
| 19      | Male| 60          | Headaches, unstable gait, papilledema | 36                            | Yes                       | 26)       |
| 20      | Female| 32         | Headaches, dizziness, papilledema, behavioral disturbance | 12                            | Yes                       | 26)       |
| 21      | Male| 18          | Epilepsy, behavioral disturbance | 24                            | Yes                       | 26)       |
| 22      | Male| 34          | Headaches, dizziness, epilepsy, behavioral disturbance | 30                            | Yes                       | 26)       |
| 23      | Male| 3           | Headaches, vomiting | 26                            | Yes                       | 26)       |
| 24      | Male| 8           | Headaches, behavioral disturbance | 16                            | Yes                       | 26)       |
| 25      | Male| 3           | Signs of hydrocephalus | 12                            | Yes                       | 26)       |
| 26      | Male| 8           | Headaches, behavioral changes, syncopal attacks | Yes                           |                           | 27)       |
| 27      | Male| 0.5         | Headaches, syncopal episodes, neuropsychological disturbances | Yes                           |                           | 28)       |
| 28      | Male| 42          | Headaches, syncopal episodes, impairment of memory | Yes                           |                           | 28)       |
| 29      | Male| 17          | Uncontrolled seizures, cognitive impairment | Yes                           |                           | 6)        |
| 30      | Female| 24         | Headaches, vomiting, mental dulling, drowsiness | Yes                           |                           | 8)        |
| 31      | Male| 44          | Postural headaches | Yes                           |                           | 10)       |
| 32      | Male| 14          | Headaches, vomiting, syncope, decreased concentration and attention | Yes                           |                           | 14)       |
| 33      | Male| 12          | Headaches, disorders of consciousness, rigidity | Yes                           |                           | 15)       |
| 34      | Male| 44          | Sudden headaches, loss of consciousness | Yes                           |                           | 17)       |
| 35      | Male| 17          | Wilson disease, seizures, cognitive impairment | Yes                           |                           | 18)       |
| 36      | Male| 9           | Motor and mental retardation, seizures | No                            |                           | 19)       |
| 37      | Male| 31          | Intermittent headaches, nausea, vomiting | Yes                           |                           | Our series |
| 38      | Female| 24         | Postural headaches, nausea | Yes                           |                           | Our series |
| 39      | Female| 22         | Headaches, dizziness, nausea, vertigo | Yes                           |                           | Our series |
| 40      | Female| 22.5       | Explosive headache | 24                            | 34)                       |
| 41      | Male| 46.8        | Explosive headache, nausea, vomiting, behavioral disturbance | 4                             | 34)                       |
| 42      | Male| 31          | Explosive headache | 9                             | 34)                       |
| 43      | Male| 2           | Progressive macrocephaly, unclosed anterior fontanelle, delayed psychomotor development | 6                             | 34)                       |
| 44      | Male| 13.5        | Progressive behavioral deterioration, uncontrollable mood swings, declining school performance | 23                            | 34)                       |
| 45      | Female| 69         | Progressive deterioration of gait, quadriaparesis, headache | 8                             | 34)                       |
| 46      | Female| 9          | Learning difficulties, unable to concentrate, emotional changes, memory loss, epilepsy, vomiting, loss of consciousness, declining school performance | 72                            | 34)                       |
| 47      | Female| 22.8       | Headache, vertigo, visual disturbance, memory loss | 17                            | 34)                       |
| 48      | Male| 12.1        | Explosive headache accompanying collapse, visual disturbance | 6                             | 34)                       |
| 49      | Male| 33          | Progressive headache | 60                            | 34)                       |
| 50      | Female| 11         | Progressive headache, mental retardation | 60                            | Yes                       | 35)       |
| 51      | Female| 36         | Disturbance of eye-movements, diplopia | 0.25                          | Yes                       | 35)       |
| 52      | Male| 63          | Diplopia, headaches, confusion | 24                            | Yes                       | 35)       |
| 53      | Female| 82         | Short-term memory deficits and gait instability, falls, urinary incontinence | Yes                           |                           | 37)       |
| 54      | Female| 41         | Short-term memory deficits and gait instability, falls, urinary incontinence | Yes                           |                           | 37)       |
| Patient | Management | Clinical outcome | Radiological outcome | Complication | Follow-up (years) | Recurrence |
|---------|------------|------------------|---------------------|--------------|------------------|------------|
| 1       | Endoscopic fenestration (right parietal) | Improvement of development, unchanged macrocephaly | Decrease of the cyst | 0.5 | No |
| 2       | Endoscopic fenestration (right parietal) | Symptoms unchanged | Decrease of the cyst | 2 | No |
| 3       | Open transcortical approach, fenestration | | | | |
| 4       | Unsuccessful fenestration, ventriculo-cysto-atrial shunt | | | | |
| 5       | Stereotactic fenestration | Died | Persistent enlargement | Died | 3 | No |
| 6       | Failed ventriculo-peritoneal shunt | Improved | Decrease of the cyst | 4 | No |
| 7       | Endoscopic fenestration with navigation | Head size stabilized | Decrease of the cyst | 3 | No |
| 8       | Endoscopic fenestration with navigation | Improved | Decrease of the cyst | 3.6 | No |
| 9       | Open transcallosal fenestration | Complete resolution | | | |
| 10      | Open transcortical fenestration | Complete resolution | | 0.33 | No |
| 11      | Stereotactic cysto-ventricular shunt | Marked improvement | | 1 | No |
| 12      | Stereotactic cysto-peritoneal shunt | Improvement of symptoms | | | |
| 13      | Cystoperitoneal shunt | Died | | | |
| 14      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms, intraventricular hemorrhage | Significant decrease of the cyst | 5 | No |
| 15      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 4.5 | No |
| 16      | Endoscopic fenestration with navigation, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 4 | No |
| 17      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 4 | No |
| 18      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 4 | No |
| 19      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 3 | No |
| 20      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 1.5 | No |
| 21      | Endoscopic fenestration, transitory external drainage | Resolution of symptoms | Significant decrease of the cyst | 1 | No |
| 22      | Cysto-peritoneal shunt | Resolution of symptoms | Significant decrease of the cyst | 1 | No |
| 23      | Endoscopic fenestration | Resolution of symptoms | Decrease of the cyst | 1.5 | No |
| 24      | Endoscopic fenestration | Decrease of the cyst | 1 | No |
| 25      | Endoscopic fenestration | Resolution of symptoms | Decrease of the cyst | 1.5 | No |
| 26      | Stereotactic fenestration | Resolution of symptoms | Decrease of the cyst | 1 | No |
| 27      | Endoscopic fenestration | Resolution of symptoms | Decrease of the cyst | 0.75 | No |
| Patient | Management | Clinical outcome | Radiological outcome | Complication | Follow-up (years) | Recurrence |
|---------|------------|------------------|---------------------|--------------|------------------|------------|
| 32      | Endoscopic fenestration | Resolution of symptoms | Decrease of the cyst |              | 0.5 | No |
| 33      | Endoscopic fenestration | Resolution of symptoms | Decrease of the cyst |              | 1 | No |
| 34      | Endoscopic fenestration | Resolution of symptoms | Decrease of the cyst |              | 1.5 | No |
| 35      | Endoscopic fenestration (right parietal) | Symptoms unchanged | Decrease of the cyst |              | 0.03 | No |
| 36      | Endoscopic fenestration with navigation | Resolution of symptoms | Decrease of the cyst |              | 0.25 | No |
| 37      | Endoscopic fenestration with navigation | Resolution of symptoms | Decrease of the cyst |              | 0.5 | No |
| 38      | Endoscopic fenestration with navigation | Resolution of symptoms | Decrease of the cyst |              | 0.09 | No |
| 39      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 5.5 | No |
| 40      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 7.08 | No |
| 41      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 5.83 | No |
| 42      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 4.17 | No |
| 43      | Endoscopic fenestration with navigation | Symptoms unchanged | Significant decrease of the cyst | | 2.58 | No |
| 44      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 3.5 | No |
| 45      | Endoscopic fenestration with navigation | Complete recovery | Decrease of the cyst | | 1.75 | No |
| 46      | Endoscopic fenestration with navigation | Marked improvement | Significant decrease of the cyst | | 0.5 | No |
| 47      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 0.25 | No |
| 48      | Endoscopic fenestration with navigation | Resolution of symptoms | Significant decrease of the cyst | | 0.5 | No |
| 49      | Internal shunting | Improvement of symptoms | Significant decrease of the cyst | | 3 | No |
| 50      | Internal shunting | Resolution of symptoms | Significant decrease of the cyst | | 0.5 | No |
| 51      | Internal shunting | Improvement of symptoms | Significant decrease of the cyst | | 3 | No |
| 52      | Internal shunting | Improvement of symptoms | Significant decrease of the cyst | | 1 | No |
| 53      | Endoscopic fenestration with navigation | Improvement of symptoms | Significant decrease of the cyst | | 1 | No |
| 54      | Endoscopic fenestration with navigation | Improvement of symptoms | Significant decrease of the cyst | | 1 | No |
patients (0.04%) having an MRI had a dilated cyst of the CSP. According to Shaw et al. (1969) [2], cysts may be classified into two groups: incidental (asymptomatic) or pathological (symptomatic). The incidence of symptomatic CSP and CV cysts is hard to define. To the best of our knowledge, the current paper is the first review of the literature concerning symptomatic CSP and CV cysts. Symptomatic CSP and CV cysts are rare and usually present with specific symptoms, such as headaches, behavioral disorders, or cognitive impairment [4]. However, there may be signs and symptoms related to hydrocephalus secondary to the occlusion of the Monroe foramina by the leaflets of the cyst [4, 26]. Moreover, most of the cases reported in the literature presented with radiological evidence of hydrocephalus, as well as regression of the cyst on postoperative imaging1. Regarding the size of the cyst, several authors argue that a CSP is defined as a cyst having a width of 10 mm or more between the ventricles [1–5, 19–25]. It may not be reasonable to consider a surgical fenestration for smaller cysts. In our review, the main presenting symptom was headaches (67%), which may be related to the potential implication of hydrocephalus in the development of a symptomatic CSP and CV cyst. Interestingly, psychiatric symptoms, mainly behavioral disturbances, were the second most common finding (56%). We use the generic term “psychiatric symptoms,” because of the heterogeneity of symptoms reported in the literature: behavioral changes, eating disorder, mood disturbances, and anxiety have been reported by several authors [1, 3, 5–10]. These symptoms may be difficult to correlate with CSP and CV cysts. However, an improvement was observed in most of the cases, which may suggest that stretching of midline structures by the cyst could be related to psychiatric disturbances [1]. Our results show that preoperative hydrocephalus was present in most of the patients and was an independent predictor of good clinical outcome. Absence of hydrocephalus at presentation may suggest that the correlation between the cyst and the presenting symptoms is unclear. Conservative management could be offered in those cases, although some reports suggest that symptoms may be improved [1, 3, 5, 6]. Most of the papers reviewed did not specify the duration of symptoms for each patient. However, most of the cases had 1 month to 3 years of symptoms before fenestration [25]. Different procedures have been proposed in the papers we reviewed. The most common approach was endoscopic fenestration of the cyst. This involves a burr-hole craniotomy, most commonly performed in the right frontal region, to fenestrate the cyst to the lateral ventricle. However, other approaches have been described, including parietal cystostomy, or direct transcavum interforniceal endoscopic fenestration, as described by the authors of this manuscript elsewhere (in press, Operative Neurosurgery). Preoperative and postoperative images of a patient that benefited from this procedure are illustrated in Fig. 2. Endoscopic and stereotactic approaches may be superior to open or shunting procedures. However, there was no statistically significant difference between the types of endoscopic approach performed regarding the outcome, complications, or recurrence rate. Most of the authors recommend an endoscopic cyst fenestration through a frontal burr-hole, with neuronavigation. Regardless of the technique, most of the cases presented with a reduction of the cyst size after fenestration (Table 2). We advocate a transcavum interforniceal approach to restore more anatomically the flow of CSF to the third ventricle, because it creates a communication between the lateral ventricles and the third

![Flowchart showing the different approaches used in the literature review cases](image-url)
ventricle (like the foramen of Monro). Moreover, it avoids midline structures (fornices, internal cerebral veins) that are displaced laterally by the cyst.

**Conclusion**

This review of the literature suggests that surgical treatment may be an option for the treatment of symptomatic CSP and CV cysts. Resolution or improvement of the symptoms was achieved in most of the cases. Endoscopic or stereotactic fenestrations seem to be superior to open or shunting procedures, with better clinical outcome and less recurrence. Operative management may be considered for symptomatic CSP and CV cysts, especially when associated with hydrocephalus.

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**Compliance with ethical standards**

**Ethical approval** This manuscript is in accordance with the ethical standards of the Sir Charles Gairdner Hospital Human Research Ethics Committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent was given by both patients operated in the Sir Charles Gairdner Hospital.

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