Review

Chronic Pediatric Headache as a Manifestation of Shunt Over-Drainage and Slit Ventricle Syndrome in patients harboring a Cerebrospinal Fluid Diversion System: A Systematic Literature Review

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Chronic Pediatric Headache as a Manifestation of Shunt Over-Drainage and Slit Ventricle Syndrome in patients harboring a Cerebrospinal Fluid Diversion System: A Systematic Literature Review

Dimitrios Panagopoulos [1*], Maro Gavra [2], Efstathios Boviatsis [3], Stefanos Korfias [4] and Marios Themistokleous [5]

[1] Pediatric Hospital of Athens, 'Agia Sophia', Director of Neurosurgery Department
[2] Pediatric Hospital of Athens, 'Agia Sophia', email: mmgavra@yahoo.com
[3] University of Athens, Medical School, 2nd University Neurosurgical Department, General Hospital of Athens 'Attikon', email: eboviatsis@gmail.com
[4] University of Athens, Medical School, 1st University Neurosurgical Department, General Hospital of Athens 'Evangelismos', email: skorfias@otenet.gr
[5] Pediatric Hospital of Athens, 'Agia Sophia', email:mthemistocleous@gmail.com
* Correspondence: dimpanayop@gmail.com

Abstract: Headache is currently considered to be a common neurological manifestation in the pediatric population, along with one of the leading causes of disability and discomfort worldwide. Notably, when the pediatric and adolescent subpopulations of patients are taken into consideration, headache is undoubtedly the main culprit for a noteworthy impact on daily physical and psychological issues. The main point is that definitive, high-quality evidence is limited and that not all subtypes of headache have been analyzed in details. The main aim of the current review is to elucidate all aspects, as well as to highlight the main characteristics of a specific subtype of the spectrum of headaches, which is related with the shunt overdrainage and slit ventricle syndrome, in pediatric patients harboring an implanted shunt device for the management of hydrocephalus. This problem is generally underestimated, although the entity of shunt overdrainage is a well-known complication in the management of hydrocephalus. This is partly due to the absence of universally agreed-upon diagnostic criteria, as well as due to a misunderstanding of relationships among the implicated pathophysiological mechanisms. A lot of attempts have been performed to propose an integrative model, aiming toward the determination of all the offending mechanisms of the shunt overdrainage syndrome, as well as to the clinical determination of the characteristic symptomatology that accompanies the relevant headaches. It seems that this subcategory of headaches, named postural dependent headache, are associated with nausea, and vomiting and/or radiological signs of slim ventricles and/or subdural collections. The ultimate goal of our review is to draw clinician's attention, especially of those who are managing pediatric patients with permanent, long-standing, ventriculoperitoneal or, less commonly, ventriculo-atrial shunts. This specific subgroup of patients may eventually suffer from severe, intractable headaches which may negatively impair their quality of daily living. In the absence of any other clinical condition that could be attributed as the cause of the headache, shunt overdrainage should not be overlooked but, on the contrary, should be seriously taken into consideration in the therapeutic armamentarium of such cases, that are difficult to be handled.

Keywords: over-drainage; slit ventricle syndrome; anti-siphon device; programmable valve

1. Introduction

A recently published meta-analysis centered on primary headache epidemiology in children, manifested that the prevalence of migraine in children and adolescents was 11% overall [1]. Existing literature data support the concept that the incidence of pediatric headache peaks at 13 years of age [2]. The term primary pediatric headaches, including the entity of migraine, constitutes a

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constellation of commonly reported neurological entities that are frequently encountered in clinical practice. When the pediatric and adolescent population is being considered, they are associated with a very disabling, albeit common, form of clinical disorder. Another main issue that is intimately related with this clinical condition is underlined by the fact that the overall prevalence is unknown and underestimated, as a lot of cases are underdiagnosed. Apart from that, the treatment options are restricted, as the pathophysiology that accompanies this spectrum of disorders is not fully elucidated. The main reason for this confusion comes from the limited number of relevant comprehensive epidemiological studies, dedicated on the prevalence and incidence of primary headaches in the pediatric age group. Moreover, the existing ones are frequently heterogeneous, and this is an intimate characteristic of the intrinsic characteristics of the studies. These include age range, sex, social and economic background, the utilized methodologies (e.g., school-based questionnaires, clinician interviews, phone surveys), along with the different diagnostic criteria applied, which occasionally may not be specific to developmental age [3]. So, compared to primary headaches in adults, Due to all of these restrictions, a limited number of epidemiological studies are available in children and adolescents, in comparison with their adult counterparts· based on bibliographic data, the estimated prevalence of headache and migraine is up to 58% and 7.7% [4] respectively. In children and adolescents, their quality of life is substantially impaired by headaches, causing negative feedback on their daily living [5]: elimination of their social activities, physical activity and school absenteeism, weaker learning outcomes, a higher risk of dropping out of school, and a negative effect on parent's careers [5,6].

The main purpose of this review is to analyze an aspect of the entity of pediatric headaches that is not primary in origin, as migraine is, but, on the contrary, could be regarded as secondary to the iatrogenic management of pediatric hydrocephalus. We have collected relevant data regarding the entity of chronic shunt over-drainage and slit ventricle syndrome and tried to investigate their pathophysiological association with the development of secondary, and often refractory to medical treatment, headaches.

2. Materials and Methods

2.1. Search Strategy

We executed a title-specific search using Thomson Reuters Web of Science database to identify the articles (reviews, case reports, original research, technical notes) that were related with shunt overdrainage, slit ventricle syndrome and headache on ependymomas till March 2024. We used these terms as our search criterion without setting any restrictions regarding publication dates. Afterwards, we reviewed the results in order to clarify that they were relevant to the purposes of our research. The papers that were chosen were further analyzed in order to extract any conclusions regarding the existence of any association between shunt overdrainage and slit ventricle syndrome, with reluctant headaches.

3. Discussion

3.1. The Entity of Shunt Overdainage and its Association with Headache

The term shunt over-drainage is been utilized in order to delimit a well-known complication that is causally related with excessive drainage of cerebrospinal fluid in patients harboring a CSF shunt system. The term "over-drainage" was first utilized in bibliographic series in 1968 [7,8] and is been increasingly accepted and adopted since the 1990s [9,10]. It is world widely known that over-drainage represents one of the most frequently encountered complications that is secondary to CSF shunting procedures [11]. It may be associated with all types of CSF diversion procedures and is not restricted to any specific pediatric age-group, but is most commonly encountered with valve-bearing shunt systems [12]. The clinical equivalent of this pathology is named as postural headache and is manifested radiologically with a slender ventricular system ("slit ventricle syndrome"). Due to the common coexistence of these two entities, postural headache is currently been considered as a clinical

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observation that is frequently recorded in combination with over-drainage [13–16]. Shunt over-drainage is manifested with a wide spectrum of pathological manifestations, such as subdural hematoma [13,17–19] and premature closure of cranial sutures (in infants) [9,20], as well as low ICP syndrome [21] · this could be managed as another aspect or definition of the entity of shunt over-drainage. There is a wide discrepancy regarding the bibliographic estimation of the exact incidence of over-drainage, as it varies from 2 to 71%. The most accepted explanation for this marked fluctuation regarding the statements for this syndrome could be based upon the non-well-specified diagnostic criteria, the heterogeneity of the investigated populations, as well as the different policies for follow-up after shunting [15,16,22]. Moreover, it is also corroborated that over-drainage may be underreported and underestimated due to the lack of consensus regarding the definition criteria of this entity, as well as due to an incomplete knowledge of the pathophysiology [16,23]. Consistent with our statements is a survey that was executed among American pediatric neurosurgeons, which documents the lack of consent and the existing uncertainty regarding the understanding and management of overdrainage-associated complications [22]. Apart from that, normal reference values range for ICP and probably postural CSF pressure/volume regulation are age-dependent. All of these data imply that the risk of overdrainage, its clinical manifestations, and treatment modalities and protocols may differ between children and adults, and, even so, between toddlers and young teenagers [24–27]. The majority of researchers agree that the remarkable variation in reported incidence is caused by the absence of a clear definition, resulting in lack of clinical consensus and uncertainty about diagnosis.

3.2. Evolution of Concepts and Current Pitfalls in Shunt Overdrainage Syndrome

There are several early, even sparse, previously reported bibliographic reports, in the form of historical cases, of excessive drainage of cerebrospinal fluid [8,16,28–33]. In the contemporary era of neurosurgery, Fox and coworkers were the first that attempted to report ICP monitoring findings in shunted patients. Their data were extracted from 18 patients suffering from normal pressure hydrocephalus· their relevant mean cerebrospinal fluid pressure values about −220 mm H2O for ventriculoperitoneal shunts and about −190 mm H2O for ventriculoarterial shunts, when they were obtained in the upright position. These findings have initially been attributed to the siphoning effect of shunts. Siphoning might be considered as the main culprit for any disabling postural headaches. The initial management option that was adopted was the incorporation of higher-pressure valves, along with VAS, especially for patients who are expected to be upright for much of their waking period [8]. Portnoy contributed to this “mechanistic model” by developing an antisiphon device, aiming at prevention of the effect of siphoning [31,34]. ICP characteristics of siphoning related to postural changes were confirmed in 1990 by Chapman, who utilized a telemetric device in patients with VPS, VAS and ventriculo-pleural shunts. Initial investigations centered on the definition of the role of ASD revealed that they were, in general terms, effective in the restoration of “normal pressures” in the upright position [35].

3.3. Clinical Manifestations in Shunt Overdrainage- Postural Headache

Overdrainage of CSF may appear in an acute manner, but this complication is not intimately related with the development of chronic refractory headaches [36,37]. Whenever chronic overdrainage is been evaluated and suspected, we should always take into consideration the possibility that a silent period of asymptomatic overdrainage of variable, and unknown, duration pre-exists. A minority of patients may not even manifest any symptomatology after the adoption of low values of intracranial pressure [38]. When a constellation of symptoms appears, they usually start as a “low-pressure headache”, headache of postural characteristics or “spinal headache”. This is clinically manifested with the patient been unable to tolerate a sitting up position. The constellation of symptoms may also include nuchal or upper back pain, nausea, vomiting, dizziness, fatigue, irritability, gait disturbance, diplopia, seizures and lethargy [39,40]. Symptomatology associated with low intracranial pressure may eventually evolve to intermittent, disabling headaches. The next step to the evolution of this clinical syndrome is related to chronic pathological entities, which include

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developmental delay, decline in school performance and social withdrawal. When the clinical records of these patients are carefully reviewed, multiple episodes of shunt revisions are frequently registered, that are in accordance with episodes of severe and intractable headaches.

3.4. Manifestations of Overdrainage

The concept of excessive drainage of CSF has been introduced by Dandy in 1932. In 1968, Becker et al. utilized the term "overdrainage" in order to explain the pathophysiologic substrate of the mechanism by which the over-drainage can induce depression of the fontanelle, as well as overriding sutures, craniosynostosis, low ventricular pressure and, finally, small ventricles [7,8]. This sequence of events involves only the infant population. Pudenz et al. first published a review article centered on overdrainage that is causally related with insertion of a shunt device in 1991 [10]; they concluded that premature closure of cranial sutures and skull deformities (in infants), stenosis or occlusion of the aqueduct, SVS, and low ICP syndrome are all included in the constellation of manifestations that constitute the entity of overdrainage.

In 2018, Ros et al. published a review centered on the entity of shunt overdrainage syndrome, attempting to specify the constellation of symptoms that constitute the entity of over-drainage. These include headache, with or without associated vomiting and neurological signs or symptoms plus different degrees of altered consciousness, in association with radiological evidence of small ventricular size and subdural collections of blood or fluid [15]. Current evidence suggests that overdrainage can manifest with a wide variety of clinical manifestations, including postural headache, subdural hygromas/hematomas, stenosis/occlusion of the aqueduct of Sylvius, craniosynostosis, SVS (characterized by intermittent headache, small ventricles, and slow refilling of the ventricular shunt reservoir), and obstruction of the ventricular catheter. Though the phenomenon "overdrainage" has been recognized as a complication related to shunt surgery for several decades, the lack of a strict definition and thus consistent terminology to describe overdrainage throughout the literature is evident.

3.5. Prevention of Headaches Associated with Shunt Overdrainage: The entity of CSF Over-Drainage.

According to a recently published data base [41,42], the underlying pathophysiologic mechanism in about 3% of cases of shunt revision procedures was reported to be CSF overdrainage. Nevertheless, the actual relevant rate is rather underestimated, with experts raising this percentage in the rate of 20% of cases. Several techniques aiming towards the reduction of the rate of CSF drainage have been reported, incorporating the use of high-pressure non-programmable fixed differential pressure valves, flow valves, as well programmable differential pressure valves [11,43,44]. A major drawback that is inherently associated with these cases is related to the fact that CSF drainage may not be as is required when the patient adopts a vertical position. On the contrary, any attempt to solely increase the opening pressure of the valves was not associated with satisfactory results in several published series [11,43,45,46]. These observations forced scientists to develop new mechanistic models related to the pattern of shunt drainage protocols. The main representative of these, newly developed drainage systems was included under the term of antisiphon systems. The main aim associated with their development was the prevention of gravitational related over pull of CSF when the patient is attempting the upright position.

Several antisiphon systems exist, all of which aiming towards the prevention and management of the entity of CSF overdrainage. The common concept that underlies their function is that they are supposed to be able to adapt to changing clinical situations or physical conditions, such as the change from the supine to the erect posture [11,47].

Another pathophysiologic mechanism that is inherently related with the development of intractable headaches, especially in the pediatric population, is related with the entity of the slit ventricle syndrome. This is widely recognized as one of the potential complications of CSF overdrainage and its pathophysiologic explanation is mainly associated with the acquisition of a pathologically reduced cerebral compliance with a typical leftward shift of the curve in the pressure/volume graph. The collapsed ventricular configuration represents the most typical

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radiographic feature of SVS. This feature by no means could be considered as been pathognomonic of SVS, as many patients may not exhibit any clinical symptoms. The exact prevalence of a collapsed ventricular system is not universally accepted, although it has been reported in the range of 10%-85% of all shunted patients [48]. A wide variety of clinical symptoms have been related with the entity of SVS and a world-wide unanimity does not exist, regarding its definition. Nevertheless, Classic SVS clinical features consist of severe and persistent or recurrent headaches, often related or provoked by positional changes. The constellation of symptoms, apart from headaches, include vomiting, weakness, ataxia, seizures, cranial nerve deficit, bradycardia, and systemic hypertension, especially in more compromised patients [49,50]. Referring to the patient's medical history, we often find records which mention that several therapeutic attempts using medication, such as intravenous fluids, analgesics, and steroids, were tested. Moreover, the referring physicians have attempted several positional changes, along with valve upgrade as recommended by several literature reviews [51–54]. Several patients have undergone repeated procedures aiming toward valve revision (using valves without any antisiphon system). Nevertheless, no one of these interventions have provided permanent relief of the symptomatology of the affected individual.

Apart from small ventricular size, patients suffering from SVS may present with several indirect radiologic signs of overdrainage. These include a small sized posterior fossa, hyperostosis of the calvarium, dolichocephalic disproportion, suture sclerosis in proximity to the skull base, parenchymal calcifications, and/or sinus hyperpneumatization [55–57]. MRI may prove to be a useful diagnostic modality, as it may offer valuable details about the ventricular and cistern anatomy [51,58–60]. Relevant, albeit not usual, MRI findings include the existence of epidural venous plexus engorgement [51,61], along with lumbar canal stenosis [11,62,63]. Other, anecdotally reported findings include the existence of pneumocephalus, as well as isolated ventricles [64], along with extra-axial collections of fluid or blood [50,64–66].

The proposed treatment algorithm for these group of patients varies greatly [67,68]. Regarding the less severe cases, the current trend is the selection of a conservative management protocol [51–54]. In general terms, the management of SVS should aim to restore the pathologically reduced cerebral compliance. Several treatment modalities have been proposed, thus reflecting the inhomogeneity and complexity of the implicated pathophysiologic mechanisms, as no one individual pathogenetic theory could explain the wide variety of clinical manifestations of this syndrome. Other treatment modalities, such as ETV, lumbar drainage, and cranial expansion have been utilized in refractory cases [69]. Nevertheless, the treatment of SVS may be associated with a wide range of complications and failures to manage it successfully· the exact prevalence of all these complications is unclear, as the relevant literature is mainly based on case reports, rather than clinical series [50,69,70].

3.6. Management of SVS

The most commonly utilized therapeutic measurement as the first step of our treatment algorithm regarding SVS is related to valve upgrade to higher opening pressure values. This maneuver is potentially effective in the management of non-complex clinical cases. Although it is technically easy, the overall handling of cases that are managed in such a manner is generally demanding [70,71]. The clinical experience that we have gained with the management of such cases has pointed out the significance of the incremental titration of the valve pressure settings, which means that a one level setting adjustment at a time is the only safe and acceptable strategy. In clinical practice, this means that the valve settings have to be re-regulated for several times, and that several single-level adjustments are the most appropriate therapeutic intervention. According to most centers recorded data, this option offers the most effective alleviation of the relevant symptomatology in the vast majority of patients who are suffering from mild range of symptoms. This is especially true for the pediatric cohort of patients, and this seems to be due to the lesser disturbance of the curvature that follows the cerebral compliance, as the time course of the disease is sooner and the diagnosis is relatively earlier registered. On the contrary, we have realized that the more severe or more chronic is the clinical equivalent of the syndrome, the lesser are the chances that a positive and

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long-lasting respond will occur or, more importantly, will be permanent. Reinforcing this view is the fact that is based on a recent relevant study [41], which enrolled a subgroup of 16 patients that were severely affected, who improved by valve reprogramming.

Another subgroup of patients has failed these conservative measurements and it requires surgical treatment. The current trend is to initially attempt externalization of the existent shunt. This therapeutic manipulation offers us the opportunity to obtain a valuable and measurable evidence of the initial opening pressure, as well as the possibility to monitor the fluctuations of the ICP values. These variations in the measurements of ICP could be used as a guidance when we attempt to increase the reservoir height. There are reports that have proposed that the spontaneous ICP fluctuations, along with the ICP variations to progressive changes of reservoir height could be considered as the basis for planning of the ongoing treatment options [37]. Several relevant studies [72–75] have adopted a treatment protocol which is based upon the ICP values. More precisely, when patients with normal or high ICP are been managed, even when the ventricular system is considered to be small, the initial management option was direct shunt replacement using programmable differential pressure valves, which incorporated an antisiphon system. On the other hand, for patients with low ICP measurements an EVD was the treatment modality of choice. Following that, the ICP was gradually increased by a progressive increasement of the reservoir height. For these patients who demonstrated ventricular enlargement concurrently with an increase in ICP values, the proposed management option was an ETV, based on the hypothesis that it could improve the cerebral compliance and equilibrate the pressure gradient between ventricles and subarachnoid spaces. There are reports who support the efficacy of this treatment modality [75].

Another subgroup of patients includes those cases which demonstrate a ventricular system whose dimensions remained unchanged, despite the increases in ICP measurements. They received a new Programmable differential pressure valve, with an incorporated antisiphon system. There are data which support that upgrading of the valve opening pressure prevented or at least delayed surgical treatment in one third of cases. Moreover, according to published studies, an initial attempt based on conservative treatment constitutes a reasonable suggestion [43,68,74,76]. It is widely accepted that young patient age and the utilization of an antisiphon system are factors that are associated with a significantly favorable outcome.

In conclusion, we have mentioned several studies which state that VPS replacement constitutes the treatment of choice for SVS· the incorporation of an antisiphon device, in association with valve/substitution is strongly recommended [11,43,46,47].

In the subgroup of patients which share in common a markedly reduced cerebral compliance, it may be beneficial to incorporate a programmable antisiphon system in conjunction with the valvular mechanism. This combination offers the possibility to gradually change either the ICP or the drainage modalities or both [11,43,47].

3.7. Clinical and Radiologic Outcome

Despite considerable improvements in our knowledge regarding SVS and technical advances in the embio-mechanics of the shunt systems, the prognosis of SVS remains unknown in many patients. According to a recently published series [41], no more than half of the participants demonstrated complete resolution of their findings, in terms of clinical and radiologic improvement. We would like to mention for one more time that children were associated with significantly better outcomes than their adult counterparts and a negative association is established as patient's age increases.

A major drawback when our therapeutic armamentarium regarding SVS treatment is been considered is related with the fact that most cases have been anecdotally reported and large case series with previously reporting specific results and treatment outcomes are lacking [15,41]. Current treatment targets are mainly centered on the control of CSF over-drainage and on the improvement of cerebral compliance.

We have concluded that patients suffering from hydrocephalus and have initially been treated with a programmable differential pressure valve were associated with a lesser chance to develop SVS.

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Nevertheless, it seems that the initial placement of antisiphon systems could not provide any protective effect against the development of SVS.

Children are generally associated with a better natural history than their adult counterparts. Apart from that, there is consensus that a prompt, proper, and, eventually, a more aggressive treatment may lead to better control of the syndrome in all age-groups.

Another important notice is related with the fact that an immediate and appropriate diagnosis remains of utmost importance. This is explained by the assumption that a protracted clinical course stands for more protracted periods with poor quality of life. The current trend stands for the importance of the utilization of modern and more sophisticated programmable valves, which offer a new therapeutic armamentarium in our attempt to prevent and manage this entity. Nonetheless, SVS remains a problem and we have to assume that all treatments modalities have failed in a significant percentage of patients. We hope that ongoing technical innovations will essentially help the prevention, diagnosis, and treatment of the SVS.

4. Conclusions

The exact knowledge of the epidemiology of migraine and other headache disorders, especially in the pediatric population, has lagged behind developments compared to other areas of neuro-epidemiology. The overall effect of headache disorders on individual patients as well as on the society itself is extremely difficult to be elucidated with clarity and constitutes a target for public health interventions that is difficult, albeit important, to be achieved. Although there is a widespread disability intimately associated with the entity of pediatric headache, this disorder remains under-diagnosed and, most importantly, under-treated and not appropriately managed.

The main aim of this manuscript is to highlight the importance of the recognition by the scientific community of the entity of headache that is related with shunt overdrainage and slit ventricle syndrome, which is secondary to the surgical management of pediatric hydrocephalus, especially in infants. It is common consent that excessive CSF drainage following the insertion of a ventricular shunt is a well-known complication that is intimately related with the treatment of hydrocephalus. Nevertheless, the absence of a world widely accepted definition in the literature is evident, as well its consequences. There is no consensus regarding the relevant diagnostic criteria, and, because of that, the exact incidence remains uncertain. The overall impact of this uncertainty is reflected on the absence of recommendations dedicated to the prevention, management, and treatment of this condition. Since no agreement has been reached for several decades, we suggest that a definition of OD should rely, not on a single expert opinion, but on consensus reached by several OD researchers. The majority of researchers have concluded that a consensus could be established, based on an agreement that OD manifests as a condition with either clinical symptoms or radiological signs, or a combination of clinical symptoms and radiological signs. CSF overdrainage following implantation of a ventriculoperitoneal shunt is characterized as a persistent condition with postural dependent headache, nausea, and vomiting, and/or radiological signs of slim ventricles and/or subdural collections (>3.6 mm width).

Moreover, the entity of SVS is intimately related with persistent and difficult to manage headaches in the pediatric population. The main issue that we have to overcome regarding SVS is that its treatment is currently based primarily on sparse anecdotal reports as, to the best of our knowledge, we are not aware of case series which report results and treatment outcomes based on a widely accepted treatment algorithm. Nowadays, our main goal is centered on attempts to control CSF over-drainage and improve cerebral compliance. Nonetheless, SVS remains an intractable problem as its management has proved to be insufficient in a significant percentage of patients, which is a fact that cannot be ignored. A promising technical advancement, that will help us to associate the clinical manifestations of the slit ventricle syndrome with the underlying pathology (reduced cerebral compliance), is the innovation of telemetric systems for ICP measurement. We wish that these devices will offer us the possibility of dynamic ICP monitoring in the near future, thus improving our therapeutic armamentarium in terms of prevention, diagnosis, and treatment of the SV

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The entities of shunt overdrainage and slit ventricle syndrome should always be included in our differential diagnostic plan, whenever we are in front of a patient with intractable headaches, that may resemble the inherent characteristics of migraine, in a patient that harbors a ventriculo-peritoneal or a ventriculo-atrial shunt, especially from the infantile period. In cases that the diagnostic work-up is unable to underline another pathological substrate, we should have a suspicion that our patient could fulfill the diagnostic criteria in order to be considered under the term of shunt overdrainage and slit ventricle syndrome.

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Abbreviations

CSF: cerebrospinal fluid
OD: over-drainage
VPS: ventriculo-peritoneal shunt
SVS: slit ventricle syndrome
VAS: ventriculoatrial shunt
ASD: Anti-siphon device
ETV: Endoscopic third ventriculostomy
EVD: External ventricular drainage
ICP: intracranial pressure

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