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Case Report

# Resolution of Papilledema Associated with Chiari I Malformation with Ventriculoperitoneal Shunting

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## Abstract

Chiari malformation type 1 (CMI) usually presents with cervical pain and suboccipital headache, among other symptoms. Patients with CMI describe symptoms that are clearly correlated with CMI for an average of 3.1 years before diagnosis. We present a case of a patient with bilateral papilledema and CMI but with no long-standing CMI symptoms. She was initially diagnosed with a concussion but developed unremitting intense occipital headaches 4 days later which prompted an evaluation for an alternative diagnosis. Treatment of this case was ventriculoperitoneal shunting, which may serve as an alternative to posterior fossa decompression under certain circumstances.

**Keywords:** *Chiari I malformation, papilledema, ventriculoperitoneal shunting*

## Introduction

Chiari malformation type 1 (CMI) usually presents with cervical pain and suboccipital headache and may be associated with hydrocephalus, papilledema, and other cranial neuropathies. Patients with CMI describe the presence of symptoms, that correlate to CMI, for an average of 3.1 years before diagnosis.[1] We presented a case of a patient with bilateral papilledema and CMI but with no long-standing CMI symptoms. Ventriculoperitoneal shunting (VPS), as an alternative to posterior fossa decompression (PFD), is described, and long-term outcome is discussed.

## Case Report

A 21-year-old woman of normal body habitus and health (Charlson Comorbidity Index 0-Charlson Comorbidity Score = 0) presented with diplopia and progressive bilateral visual loss shortly after a head-to-head collision. She was initially diagnosed with a concussion but developed unremitting intense occipital headaches 4 days later which prompted an evaluation for an alternative diagnosis. Initially, she did not endorse visual changes, diplopia, or associated nausea, vomiting, photophobia, or phonophobia. However, 4 days after the injury, the patient developed horizontal binocular double vision, blurred vision out of the left eye, intermittent pulsatile tinnitus, and transient visual loss when standing. She denied antecedent antibiotic, steroid, or Vitamin A derivative use, and there was no recent increase in her weight. There was no prior exposure to ticks or cats.

On initial presentation to her local ophthalmologist, she had visual acuity of 20/25 in the right eye and 20/40 in the left eye. Moderate optic disc swelling was observed in the left eye, while the right optic nerve appeared normal. Magnetic resonance imaging (MRI), performed without contrast, revealed a CMI. She was referred for neuro-ophthalmic evaluation. On examination, the patient was afebrile, had normal vital signs, and her body mass index (BMI) was 24.42 kg/m$^2$. Her visual acuity was 20/20 in the right eye and 20/30 eccentrically in the left eye. The color vision was full in both eyes, and there was no afferent pupillary defect. Testing of extraocular motility revealed bilateral mild abduction deficits. Orthoptic examination revealed a 30 prism diopter esotropia in primary position that worsened in horizontal gaze to either side. Slit lamp examination was normal, but examination of the posterior segment revealed Frisen Grade 5 papilledema of both eyes, obscuration of vessels, cotton wool spots, disc hemorrhages, associated macular thickening worse in the left eye, and

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tortuosity of retinal veins [Figure 1a and b: initial fundus examination]. Formal visual field testing demonstrated severe global depression with arcuate visual field loss involving fixation bilaterally [Figure 1c and d: initial visual field examination].

The constellation of bilateral severe optic disc swelling and bilateral sixth nerve palsies necessitated additional investigations to elucidate a cause of raised intracranial pressure (ICP). MRI with gadolinium demonstrated nonspecific scattered fluid-attenuated inversion recovery signal abnormalities in the subarachnoid spaces of the frontoparietal lobes bilaterally as well as CMI with low-lying cerebellar tonsils terminating 10 mm below the foramen magnum and resultant effacement of cerebrospinal fluid (CSF) spaces at craniocervical junction. CINE CSF flow sequence revealed markedly reduced CSF flow in the dorsal column at the craniocervical junction [Figure 2]. MRI further demonstrated optic nerve distention consistent with papilledema. MR venography revealed narrowing of the transverse sinuses but excluded a venous sinus thrombosis. Cervical spine MRI did not demonstrate syringomyelia. A small volume (6cc) lumbar puncture was performed after neurosurgical consultation was sought, and an opening pressure of 42 cm $H_2O$ was obtained with normal CSF constituents and no evidence of infection.

The patient was treated with intravenous methylprednisolone as well as acetazolamide 500 mg BID for 3 days with no improvement in headache, papilledema, or visual function.

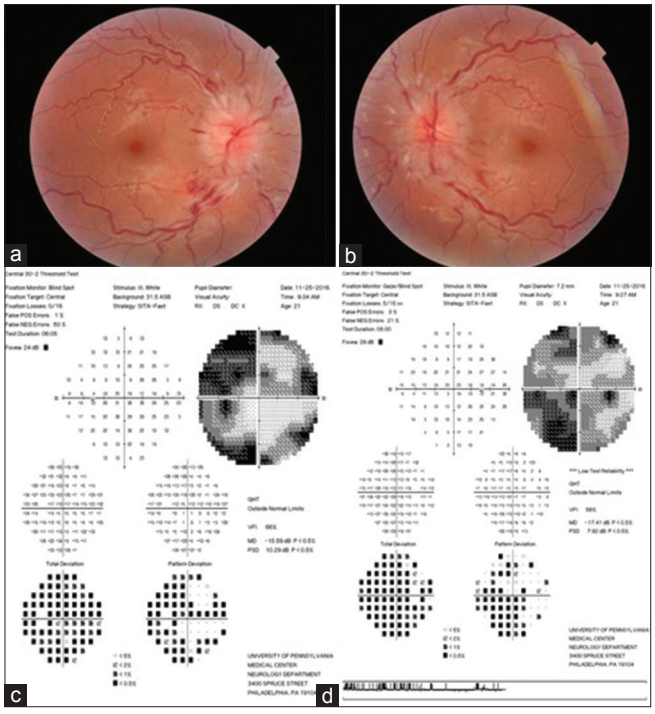


**Figure 1: Fundus photography (a and b) and formal visual field testing (c and d) at presentation. At presentation, our patient had evidence of severe papilledema with retinal hemorrhages, cotton wool spots, and retinal venous tortuosity (a and b). Humphrey 30-2 visual fields demonstrated severe global depression involving fixation bilaterally (c and d)**

Neurosurgical evaluation elucidated no long-term historical symptoms that could be attributed to the CMI. Given a lack of improvement in new symptoms or papilledema, surgical interventions were discussed, including VPS, PFD, or both VPS and PFD. The recommended intervention was VPS alone because of the reduced risk of surgical complication and the potential for improvement from a single intervention. Of additional benefit, if initial surgical management were to fail, VPS would not preclude subsequent PFD. The patient consented to VPS with the understanding that PFD might ultimately be necessary. She underwent successful insertion of the right frontal VPS, and her headache resolved immediately postoperatively. She experienced rapid improvement in her diplopia and normalization of her visual function and optic disc swelling over the subsequent weeks.

MRI performed 1 year after presentation demonstrated stable findings of CMI and effacement of CSF spaces with a well-positioned right VPS. There was a resolution of periventricular transependymal flow and papilledema. A CSF flow study (CINE) demonstrated stable decreased flow dorsally at the craniocervical junction. At a neuro-ophthalmic follow-up examination 16 months after her initial presentation, she denied symptoms that might be attributed to raised ICP. Her visual acuity was 20/20 in each eye with full-color vision. Fundus examination revealed mild gliosis of both optic nerves and complete resolution of papilledema with no appreciable pallor [Figure 3a and b: follow-up fundus examination]. Formal visual field testing revealed mild enlargement of the blind spots bilaterally

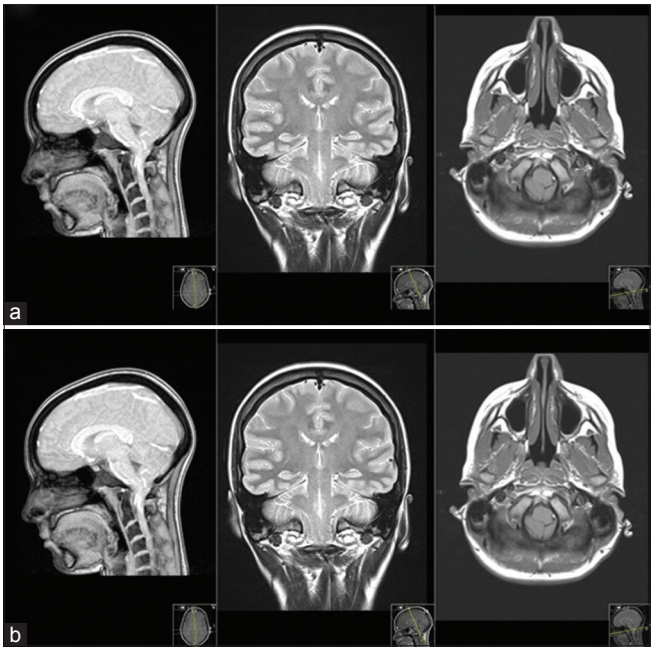


**Figure 2: Initial (a) and follow-up (b) magnetic resonance imaging demonstrating CMI. (a) First magnetic resonance imaging November 2016. (b) Second and f/u magnetic resonance imaging January 2018, no change in Chiari malformation, tonsillar herniation, or foramen magnum compression however with dramatic clinical improvement**

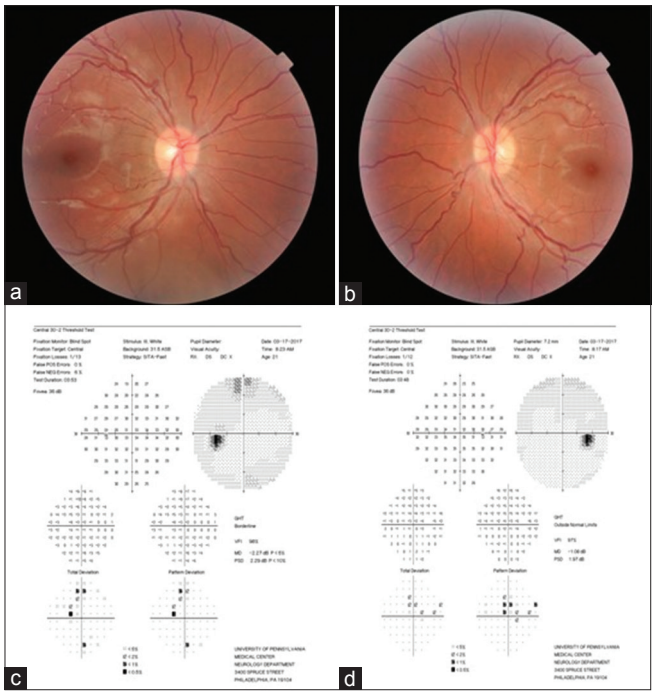

**Figure 3: Fundus photography (a and b) and formal visual field testing (c and d) after 16 months of follow-up. After 16 months of follow-up, there was a persistent resolution of papilledema with mild gliosis of both optic nerves (a and b). Humphrey 30-2 visual field demonstrated mild enlargement of both blind spots and nonspecific depression (c and d)**

with nonspecific depression [Figure 3c and d: follow-up visual fields].

## Discussion

Chiari I malformation (CMI) consists of a congenitally malformed hindbrain with herniation of the cerebellar tonsils below the foramen magnum and is observed in 0.24%–0.9% of adults undergoing MRI.[2] Many patients that present for clinical evaluation will also demonstrate syringomyelia on imaging of the cervical spine. Among patients with papilledema and evidence of CMI, the diagnostic possibilities include raised ICP secondary to pseudotumor cerebri syndrome (PTCS) or CMI. The distinction between these diagnoses is challenging because of overlapping demographics, age of presentation, and clinical symptoms. Even obesity, a well-described risk factor for PTCS, has also been shown to be more frequent among pediatric patients with CMI.[3] PTCS may be associated with MRI abnormalities including optic sheath distention and tortuosity, posterior globe flattening, empty or partially empty sella sign, and transverse venous sinus stenosis on neuroimaging but is not associated with ventricular dilatation.[4] While in most cases CMI with raised ICP will present with ventriculomegaly, CMI may cause raised ICP without associated ventricular dilation. The absence of ventriculomegaly and hydrocephalus does not reliably distinguish between the two possibilities.

The diagnostic challenge is made more complex by the reported association between PTCS and CMI. Banik *et al*. identified inferior tonsillar displacement in 24% of patients diagnosed with PTCS, with 10% meeting the criteria for CMI.[5] In this population, it is uncertain whether CMI was the primary cause of raised ICP, secondary to PTCS, or an incidental finding. The identification of restricted CSF flow at the level of the foramen magnum using dynamic CSF flow studies, such as the CINE sequence, supports the possibility that CMI is the cause of raised ICP.[6] Vaphiades and Braswell reported an obese patient with papilledema and CMI on neuroimaging, whose CSF flow studies demonstrated normal CSF flow through the posterior fossa at the level of the foramen magnum, suggesting PTCS was the most likely underlying etiology. The patient experienced a remarkable resolution of CMI on a subsequent MRI study after treatment with acetazolamide alone, further supporting the hypothesis that CMI may have been secondary to PTCS.[7]

In our patient's case, clinical and radiographic evidence were used to postulate about the underlying mechanism for raised ICP. She had a normal BMI and no history of provocative medications or medical conditions associated with PTCS. Her symptoms were suggestive of raised ICP which may be seen in both PTCS and CMI, but she did not endorse typical symptoms of CMI such as cervical pain, suboccipital headache, cerebellar dysfunction, or myelopathy which typically persist for months to years before presentation.[1] The radiographic identification of impaired CSF flow at the level of the craniocervical junction using dynamic CSF flow (CINE) MRI supported the mechanism of CMI-related raised ICP.

Identifying the underlying etiology for raised ICP has implications for successful management. PTCS is typically treated medically with lifestyle measures which promote weight loss and ICP lowering medications such as acetazolamide; CSF shunting procedures or optic nerve sheath fenestration are reserved for refractory or severe progressive cases.[8] In contrast, symptomatic CMI is traditionally treated with neurosurgical intervention, specifically with PFD. The neurosurgical literature contains many cases of successful management of papilledema secondary to CMI with PFD; however, no randomized trials have been performed comparing PFD and VPS.[9-13] While PFD is successful in many retrospective case series of patients with CMI and papilledema, recurrence of symptoms or papilledema postoperatively has also been reported. Five patients described by Alnemari *et al*. underwent PFD for CMI but experienced a recurrence of symptoms within days to 3 years postoperatively despite restoration of CSF flow at the craniocervical junction. Subsequent VPS was successful in providing symptom relief to all five patients.[14] In the senior authors' experience, some patients who fail to respond to PFD, particularly when healing problems and CSF leak are associated, will subsequently be found to have ventriculomegaly and clinical hydrocephalus and respond to CSF diversion through VPS.

To the best of our knowledge, this is a unique report of successful treatment of CMI-related raised ICP and papilledema with VPS alone. Before undergoing insertion of VPS, our patient experienced progressive deterioration of visual function from papilledema despite maximal medical therapy and enjoyed rapid resolution of headache, diplopia, and papilledema after VPS insertion, with stable follow-up for 16 months. The potential morbidity associated with acute brainstem herniation from lumbar punctures or lumboperitoneal shunting procedures among patients with CMI must be emphasized; however, VPS is unlikely to cause this dreaded complication because the site of diversion is rostral to CMI.[12,13] Through successful management with VPS alone, the patient avoided a prolonged postoperative recovery and the additional surgical morbidity/risk of complications with PFD, particularly when ICP is elevated, as well as a longer postoperative recovery.[15,16] PFD remains a surgical option should our patient experience a recurrence of symptoms in the future. VPS may, therefore, be considered as an option for initial surgical intervention among patients with headache and papilledema with radiographic evidence of CMI who do not endorse significant symptoms of CMI and who fail to respond to maximal medical management.

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### Conflicts of interest

There are no conflicts of interest.

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