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# Arnold-Chiari Malformation-I Borderline Personality Disorder

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## ABSTRACT

**INTRODUCTION:** Arnold-Chiari Malformation I (AM-I) is a congenital anomaly that manifests with cerebellar dysfunction. There is a displacement of cerebellar tonsils into the foramen magnum. Several mood disorders, personality disorders, and intellectual disabilities are associated with AM-I. Borderline personality disorder (BPD) is characterized by symptoms of mood lability, impulsivity, extreme efforts of abandonment, splitting and dysfunctional relationships.

**CASE DESCRIPTION:** The patient is an early aged adult with a past medical history of AM-I, hypothyroidism, Wolff-Parkinson-White syndrome, and diabetes mellitus type II. The patient was admitted to the hospital after ingesting foreign bodies. He/she presented with mood lability, sad mood, anhedonia, insomnia, panic attacks, ruminative worries, feelings of emptiness, and recurrent suicidal gestures and threats. The patient was eventually diagnosed with borderline personality disorder. This case suggests a possible connection between AM-I and BPD.

**DISCUSSION:** Emerging from contemporary research involving the cerebellum, it is important to acknowledge that the current definition of control of fine motor and balance is inadequate. Symptoms associated with AM-I and BPD may be better explained by Cerebellar Cognitive Affective Syndrome (CCAS), a condition that ties the cerebellum with the higher cognitive functioning in the brain.

**KEYWORDS:** Arnold-Chiari Malformation, Borderline Personality Disorder, Cerebellar Cognitive Affective Syndrome, Impulse Control Disorder

## INTRODUCTION

Arnold-Chiari Malformation I (AM-I) is a congenital malformation caused by the displacement of cerebellar tonsils into the foramen magnum. This displacement can potentially cause disturbances in intracranial pressure, resulting in headaches as a common presenting symptom. Additionally, cognitive deficits are frequently an associated finding with AM-I. To date, there are few studies evaluating cognitive functioning associated with AM-I. A study of cognitive performance of 39 patients with AM-I demonstrated significantly lower scores in 9 cognitive domains, most notably executive functioning, and theory of mind.<sup>1</sup> Brill et al. described a study of 11 children with seizures, motor or language delay, or autism that all were found to have AM-I which concluded that AM-I should not be considered an “incidental finding, rather a marker for developmental disabilities.”<sup>2</sup> Borderline personality disorder (BPD) is characterized by symptoms of mood lability, impulsivity, extreme efforts to avoid abandonment, splitting, and dysfunctional relationships, with many patients having a history of abuse. A significant but minimally studied symptom in patients with BPD is foreign body ingestion, usually involving inanimate objects.

Numerous psychiatric disorders have been associated with AM-I including attention deficit hyperactivity disorder, autism spectrum disorders, schizophrenia, bipolar disorder, major depressive disorder, and anxiety disorders.<sup>3</sup> Despite there being evidence of co-occurring psychiatric disorders associated with AM-I, to our knowledge there is no known study connecting these two disorders. This case report will address a possible link between AM-I and BPD.

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## CASE PRESENTATION

To ensure anonymity, the age and sex of the patient is concealed. This patient is an early-aged adult with a past medical history of AM-I, hypothyroidism, Wolff-Parkinson-White syndrome, and diabetes mellitus type II. He/she has a significant psychiatric history with diagnoses of bipolar I, pica, post-traumatic stress disorder schizoaffective disorder, depression, and alcohol use disorder. He/she had corrective surgery for AM-I as a teenager when it was discovered as a result of frequent migraines. This patient has a significant family history of psychiatric illness including suicide.

This patient was initially brought to the emergency department for foreign body ingestion. He/she admitted to mood problems with symptoms of sad mood, anhedonia, insomnia, panic attacks, ruminative worries, and feelings of emptiness. He/she also reported symptoms of mood lability, irritability, poor impulse control, impaired judgment, and racing thoughts. Patient complained of occasional auditory hallucinations that tell him/her to “kill himself/herself.” There is also a history of referential and persecutory delusions related to exposure to stressful situations including altercations with his/her family and peers. However, the patient did not present with a gross thought disorder and the psychotic symptoms were not persistent throughout the hospital course. Patient admitted to feeling “empty” and bored. He/she acknowledged recurrent suicidal gestures and threats. Alcohol abuse was reported as an impulsive act and not persistent. Furthermore, he/she liked the repeated hospitalizations as these gave him/her the opportunity to have time away from the residential facility. The patient had engaged in repeated foreign body ingestions that reinforced these behaviors. The patient was diagnosed with pica but there was no chronic ingestion of non-nutritive substances. When interviewed, the patient was overwhelmingly pleasant to the physician, though just prior to the encounter was shouting at the nursing staff, which is a typical example of splitting. During this particular hospital stay, diagnoses of bipolar affective disorder type I with mixed features, as well as borderline personality disorder were made. In addition, he/she was diagnosed with PTSD and a mild to moderate intellectual disability because of chronic deficiencies in social and cognitive domains. While intellectual disability is associated with these behaviors, the patient denied swallowing foreign objects as a child and claimed these behaviors only started in early adult life. This impulsive, attention-seeking, and manipulative behavior has

worsened in the past three years.

He/she had persistently posed a danger to himself/herself, manifesting recurrent suicidal gestures. Consequently, providers kept him/her on a combination of Clozapine, Chlorpromazine, Fluoxetine, and Gabapentin. This patient had prior trials of Paroxetine, Olanzapine, Topiramate, Lithium, Sertraline, Trazodone, Venlafaxine, Lorazepam, and Haloperidol.

Abdominal CT confirmed the presence of radiopaque objects noted intraluminally in the gastric antrum. The patient was put on a behavioral plan to discourage self-harming behaviors throughout this hospital stay. After the endoscopic removal of the foreign bodies, the patient was discharged back to the residential facility.

## DISCUSSION

Emerging from contemporary research involving the cerebellum, it is important to acknowledge that the current definition of its control of fine motor and balance is inadequate. Instead, it is now thought that the cerebellum may have higher cognitive roles than once previously thought.<sup>4,5</sup> Interestingly, there have been reports of other personality and mood disorders, like generalized anxiety disorder and bipolar-I that co-occur with deformations of the cerebellum such as AM-I.<sup>6,7</sup>

First described in 1998 by Dr. Jeremy Schmahmann, Cerebellar Cognitive Affective Syndrome (CCAS) is characterized by injury to the cerebellum resulting in disturbances of executive functioning, impaired spatial cognition, personality change with blunted affect, and disinhibited or inappropriate behavior and linguistic difficulties.<sup>8</sup> The study concluded that there is known anatomical circuitry between the cerebellum and higher functioning regions of the brain including the cingulate gyrus, prefrontal cortex and limbic circuitry. Disruptions of these connections in patients with cerebellar lesions results in hindered cognitive and affective functioning, among other cerebellar roles. It is possible that the damage to the cerebellum in AM-I has the propensity to cause CCAS.<sup>8</sup>

Per the DSM-V, impulsivity consists of a lack of inhibition that results in spontaneous actions and responses that fail to account for consequences, often taking the form of self-harming behaviors. Impulsivity can be measured on the Barratt Impulsiveness Scale Version 11 (BIS-11) and it is measured using 34 questions graded on a 4-point scale assessing three factors: increased motor activity, decreased attention, and decreased planning.<sup>9</sup> Since

the BIS-11 is a self-reported scale, our patient was not able to complete the test. However, considering the clinical presentation, it was expected that he/she would score in the higher range, demonstrating increased impulsiveness. Impulsivity has a high comorbidity with bipolar disorder.<sup>10</sup> This patient has mild to moderate intellectual disability, previous mood disorder diagnoses, and impulsivity manifesting in foreign body ingestion. To say that there is an underlying personality disorder that classically exhibits self-harming behavior might be suitable, however patients with intellectual disabilities have been shown to have an association with pica which is the ingestion of non-nutritional, inanimate objects.<sup>11</sup> Whether this behavior is due to a personality disorder, intellectual disability, or impulse control dysregulation is unclear since AM-I has been associated with cognitive dysfunction, mood disorders, and other psychiatric disorders. Perhaps our patient could be demonstrating sequelae of CCAS due to cerebellar defect, or presenting with specific, overlapping symptoms shared between these various disorders.

The cerebellum and the limbic system share distinct, neuronal connections that are involved in detecting, integrating, and filtering interoceptive, autonomic, and emotional information.<sup>5</sup> In patients with AM-I specifically, hyperconnectivity in the posterior cingulate cortex results in hyperactive processing of pain, which is thought to have a distracting effect that impedes cognition.<sup>12</sup> Patients with BPD were shown to have reduced gray matter volume in the anterior cingulate, particularly in areas necessary for performing complex cognitive tasks like sustained attention, emotional control, and conflict resolution.<sup>13</sup> Alterations in the cingulate gyrus and other limbic structures may provide key information in connecting AM-I and BPD.

AM-I and BPD each have a genetic basis for disease, but environmental factors also play a role in the development of these diseases.<sup>14,15</sup> No specific gene has been found as the major cause for either disease, as both diseases are likely multifactorial in nature. Common genetic factors linking the two disorders were not identified in our review of the literature. Although alterations in the balance of neurotransmitters such as dopamine and serotonin frequently result in psychiatric illness, it is unclear which pathways are predominantly involved in these diseases. Further studies are needed to investigate whether neurotransmitters and other neurometabolites affect these disease processes.

## CONCLUSION

There appears to be several poorly established connections between the cerebellum and cognition. There are a substantial number of overlapping symptoms between AM-I and BPD. These similarities may be attributed to distinct neurocircuitry, anatomical commonalities, genetics, or specific neurometabolite levels. Further studies are needed to demonstrate a relationship between these two diseases, and a connection may be better explained by CCAS. When neurological and psychiatric impairments coexist in patients, an interrelation between these manifestations of disease should be investigated to provide accurate diagnoses that will help to guide and improve treatment outcomes.

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