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# Marchiafava Bignami Syndrome: A Review

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***Abstract***

*Marchiafava-Bignami syndrome (MBS) is a rare neurological disorder characterized by demyelination and necrosis of the corpus callosum, which is the main bundle of nerve fibers connecting the two hemispheres of the brain. MBS primarily affects individuals with chronic alcoholism, although cases unrelated to alcohol consumption have also been reported. The corpus callosum is affected in almost a pathognomonic way. The clinical presentation is based on neurological impairment, motor abnormalities, seizures, behavioral changes, and, in severe cases, coma. The diagnosis of MBS involves a combination of clinical assessment, neurological examinations, neuroimaging studies (such as magnetic resonance imaging), and, in some cases, biopsy can be done to assess necrosis, degeneration and gliosis. Management of MBS focuses on addressing underlying causes, providing supportive care, and managing symptoms. This includes alcohol cessation, nutritional support, seizure management, symptomatic treatment, rehabilitation therapies, and psychosocial support. However, the prognosis for MBS is generally poor, with a high mortality rate. Collaboration with the healthcare team and ensuring continuity of care are essential for providing comprehensive and coordinated nursing care for individuals with MBS.*

**Keywords:** Corpus callosum, hemisphere, demyelination, gliosis, syndrome

## INTRODUCTION

Marchiafava-Bignami syndrome (MBS) is a rare affliction characterized by primary degeneration of corpus callosum, associated with chronic consumption of alcohol. First described in 1903 by two, *Italian physicians, Marchiafava* and *Bignami,* the syndrome primarily affects individuals with a history of chronic alcoholism. However, non-alcoholic cases have also been reported. MBS is associated with significant neurological impairment and carries a poor prognosis. The exact pathophysiology of MBS is not fully understood, but several factors are believed to contribute to its development. Chronic alcohol abuse, nutritional deficiencies, impaired blood flow, and genetic susceptibility have been implicated. These factors can lead to damage and cell death in the corpus callosum, resulting in the characteristic manifestations of MBS [1]. Given the rarity and complexity of MBS, further research is needed to advance our understanding of its pathophysiology, improve diagnostic methods, and develop more effective treatments. Increased awareness among healthcare professionals and the general population is also important to facilitate early recognition and timely intervention for this debilitating syndrome.

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### Definition

Marchiafava-Bignami disease is a progressive neurological disease of alcohol use disorder, characterized by corpus callosum demyelination and necrosis and subsequent atrophy [2].

### Incidence

As of 2004, approximately 250 patients had been identified, 200 had died, 30 suffered from severe dementia, and only 20 experienced positive outcomes. It seems that with alcoholism, the prognosis is worse [3].

 

## REVIEW OF LITERATURE

A study was conducted to investigate the clinico-radiologic features of acute Marchiafava-Bignami disease (MBD) and its evolutionary process after effective treatment through subgroup comparison. The clinical and MRI data of 23 patients with acute MBD were retrospectively analyzed and divided into type A (12 cases, with entire callosal involvement) and type B (11 cases, with focal callosal involvement). The clinical assessments and MRI findings (before and after treatment) were compared between the two subtypes. Compared with type B, type A had lower MoCA (Montreal Cognitive Assessment) scores at admission ($16.50 \pm 1.73$ vs. $18.27 \pm 1.68$, $P = 0.021$) and were more common with extra callosal involvement (66.67% vs. 18.18%, $P = 0.036$) and longer illness duration ($18.3 \pm 2.1$ days vs. $15.6 \pm 2.4$ days, $P = 0.012$). During the treatment, the residual lesion in the splenium was more common in type A (58.33% vs 9.09%, $P = 0.027$). After treatment, the MoCa scores of both subtypes gradually increased ($P < 0.001$), and the callosal and extracallasal lesions disappeared completely. Clinico-radiologic typing of acute MBD is related to the severity of early symptoms, but not to the prognosis. Complete clinico-radiologic recovery is possible for both subtypes with combined treatment. The clinico-radiologic reversibility is helpful for accurate diagnosis and therapeutic evaluation [4].

A study was conducted for diagnosis and management of Marchiafava-Bignami disease by a review of CT/MRI confirmed cases. On review of 122 reports, containing data on 153 subjects with confirmed MBD that was associated with either alcoholism or malnutrition, and 20 reports with data on 53 subjects with conditions mimicking MBD. All the cases had been verified antemortem by brain imaging. Unconditional logistic regression was used to demonstrate factors that were associated with the outcome of MBD. The mimicking conditions were differentiated from MBD by the occurrence of solitary and rapidly disappearing splenial lesions; fewer signs and symptoms with exception of seizures, hemiparesis and tetraparesis; nystagmus; and rapid and complete recovery. MBD occurred most frequently among alcoholics, but it was also reported in 11 non-alcoholics (7.2% of all the MBD cases). A better outcome was observed among those who were treated within 2 weeks after onset of symptoms with parenteral thiamine ($p=0.033$). As thiamine deficiency is frequently associated with alcoholism, malnutrition and prolonged vomiting; the study recommend prompt treatment of MBD with parenteral thiamine in such subjects. Recovery should be followed by repeated neuropsychological and MRI examinations, preferably using diffusion tensor imaging [5].

## ETIOLOGY

The exact etiology of Marchiafava-Bignami syndrome (MBS) is not fully understood. However, several factors have been proposed to contribute to the development of this rare neurological disorder. Here are some potential etiological factors [6]:

1. **Alcohol toxicity:** Chronic alcohol abuse is strongly associated with the development of MBS. Alcohol can induce oxidative stress, impair cellular metabolism, and disrupt the balance of neurotransmitters, leading to damage and death of the cells in the corpus callosum.
2. **Nutritional deficiencies:** Malnutrition, specifically deficiencies in B vitamins (thiamine, niacin, folate), vitamin E, and trace elements, can contribute to the development of MBS. Alcoholism often leads to poor dietary intake and impaired absorption of essential nutrients, which can contribute to the vulnerability of the corpus callosum to injury and degeneration.
3. **Liver disease:** Liver dysfunction or cirrhosis, often associated with chronic alcohol abuse, may contribute to the development of MBS. Liver disease can lead to metabolic imbalances, impaired detoxification, and altered nutrient metabolism, which can affect the integrity of the corpus callosum.
4. **Other factors:** MBS has been reported in individuals without a history of alcoholism, suggesting that factors other than alcohol may contribute to its development. These factors include anemia, severe malnutrition, head trauma, and exposure to certain toxins.

## TYPES

There are two clinical subtypes of MBS [7]:

 

1. **Type A:** Stupor and coma will present. It affects the entire corpus callosum.
2. **Type B:** This type has mild impaired mental status and it cause only partial lesions in the corpus callosum.

## PATHOPHYSIOLOGY

The pathogenesis of tissue damage in Marchiafava-Bignami disease [8]:

- The corpus callosum consists of myelinated nerve fibers that facilitate communication between the two hemispheres of the brain.
- In MBS, the breakdown of myelin, which is the protective covering of nerve fibers, and damage to axons (the nerve fibers themselves) occur.
- This disruption of the myelin and axonal integrity in the corpus callosum leads to impaired communication between the hemispheres and disrupts the blood flow.
- Insufficient blood supply and oxygenation to the cells in the corpus callosum can contribute to their degeneration and necrosis.
- This damage to the brain can trigger an inflammatory response, involving activation of immune cells and release of pro-inflammatory molecules, which can further contribute to tissue damage and neuronal death.

## CLINICAL MANIFESTATIONS

The clinical findings of Marchiafava-Bignami disease are [9]:

1. **Neurological impairment:** This can include confusion, disorientation, impaired memory, and difficulty with concentration and attention. They may have difficulties with problem-solving, language, and executive functions.
2. **Motor abnormalities:** This can manifest as weakness, spasticity (muscle stiffness), and difficulty with coordination. They may have an unsteady gait, tremors, and difficulty performing fine motor tasks.
3. **Seizures:** Seizures can be focal (involving a specific area of the brain) or generalized (affecting both hemispheres of the brain). Seizures can further contribute to neurological dysfunction and impair overall functioning.
4. **Behavioral and psychiatric changes:** MBS can cause changes in behavior and mood. Patients may exhibit emotional instability, depression, apathy, irritability, and agitation. Personality changes and social withdrawal may also be observed.
5. **Coma and neurological deterioration:** In severe cases of MBS, individuals may progress to a state of coma and experience rapid neurological deterioration. This can be life-threatening and may result in significant disability or death.

## DIAGNOSTIC EVALUATION

The diagnostic evaluation of Marchiafava-Bignami syndrome (MBS) typically involves a combination of clinical assessment, neuroimaging, and laboratory tests [10].

1. **Clinical history and physical examination:** History including information about alcohol consumption, nutritional status, and any other underlying conditions.
2. **Physical examination** is mainly focused on to assess neurological function and check for signs and symptoms associated with MBS.
3. **Neurological examination:** To evaluate cognitive function, motor abilities, coordination, reflexes, and sensory perception.
4. **Imaging studies:** Magnetic resonance imaging (MRI) is the preferred modality and it reveals abnormalities such as signal changes, atrophy, or degeneration in the corpus callosum that are indicative of MBS. Additional imaging, such as computed tomography (CT) scans, may be performed to rule out other possible causes of neurological symptoms.
5. **Biopsy:** A small sample of tissue from the corpus callosum is examined to assess for characteristic pathological changes such as necrosis, demyelination, and gliosis.

 

6. **Laboratory tests:** To evaluate nutritional status, liver function, and rule out other underlying conditions. Blood tests may include complete blood count (CBC), liver function tests, vitamin levels (such as thiamine), and markers of inflammation.

## MANAGEMENT

The management of Marchiafava-Bignami syndrome (MBS) typically involves a combination of supportive care, addressing underlying causes, and specific interventions aimed at improving symptoms and promoting recovery [11].

1. **Alcohol cessation:** Complete abstain from alcohol consumption is essential to prevent further damage to the brain and allow for potential recovery.
2. **Nutritional support:** Ensuring proper nutrition and correcting any nutritional deficiencies may involve a well-balanced diet, vitamin supplementation (especially thiamine or vitamin B1), and close monitoring of nutritional status.
3. **Seizure management:** Appropriate antiepileptic medications such as Levetiracetam 500 mg may be prescribed to control and manage seizures. The choice of medication will depend on the specific type of seizures and individual patient factors.
4. **Symptomatic treatment:** Treatment may be provided to address specific symptoms and complications such as cognitive impairment, motor abnormalities, mood disturbances, or other neurological symptoms.
5. **Rehabilitation therapies:** These are beneficial in managing motor deficits, improving coordination, and enhancing cognitive function. Therapies include:
   - Physical therapy like ROM exercise, Strengthening exercise, Balance and coordination training.
   - Cognitive behavioral therapy.
   - Occupational therapy like ADL training.
   - Speech therapy.
6. **Psychosocial support:** Counseling, support groups, and other psychosocial interventions to cope with the emotional, behavioral, and social changes associated with the condition.

## NURSING IMPLICATIONS

Nursing care plays a critical role in supporting individuals with Marchiafava-Bignami syndrome (MBS) throughout their treatment and recovery process. Here are some nursing implications and considerations for caring for patients with MBS [12, 13]:

1. **Assessment:** Regular monitoring of vital signs, neurological functioning, and any changes in symptoms is essential**.**
2. **Nutritional support:** Monitoring the patient's dietary intake, promoting a well-balanced diet, and ensuring adequate thiamine and vitamin supplementation. Encouraging proper hydration and addressing any swallowing difficulties or dietary restrictions can also be important.
3. **Seizure management:** Monitoring the patient during a seizure episode, protecting them from injury, and documenting seizure activity. Nurses should also assist with the administration of antiepileptic medications as prescribed and educate the patient and family about seizure precautions and management.
4. **Rehabilitation support:** Nurses can collaborate with rehabilitation specialists to support the patient's participation in physical therapy, occupational therapy, and speech therapy.
5. **Psychosocial support:** Nurses can provide emotional support, actively listen to the patient's concerns, and facilitate access to counselling services or support groups.
6. **Collaboration and continuity of care:** Ensuring effective communication, accurate documentation, and smooth transitions of care between different healthcare settings is essential for providing comprehensive and coordinated care.

## CONCLUSION

 

MBS is a rare and complex neurological disease of alcohol use disorder characterized by corpus callosum degeneration and necrosis associated with chronic alcohol consumption. Due to the rarity of MBS, there are no specific guidelines or standardized treatment protocols available. Therefore, a multidisciplinary approach involving neurologists, rehabilitation specialists, nutritionists, and other healthcare professionals is often necessary to develop an individualized management plan based on the patient's specific needs and symptoms. Early recognition, appropriate diagnostic evaluation, and multidisciplinary management are essential for positive patient outcomes and enhancing quality of life for individuals with MBS.

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