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Mortality of Guillain Barre Syndrome and Myasthenic Crisis in Haji Adam Malik Central General Hospital Medan January 2021 - December 2022

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ABSTRACT

Introduction: Neuromuscular disorders are disorders of the neuro system that impact the muscular system. Among them are Guillain Barre Syndrome and Myasthenic Crisis. Guillain Barre syndrome is an autoimmune disorder of the nervous system that is polyradiculoneuropathic, characterized by weakness that radiates from the legs to the arms, torso, and face. Myasthenic crisis is a state of acute respiratory failure due to the exacerbation of Myasthenia Gravis. These two diseases are the most common types of neurodiseases found in hospital ICU patients.

Objective: Determining the mortality rate of Guillain Barre Syndrome and Myasthenic Crisis at Haji Adam Malik Central General Hospital Medan January 2021-December 2022.

Methods : This study was a descriptive study with data obtained from medical records. The subjects were inpatients and outpatients with Guillain Barré Syndrome and Myasthenia Crisis, with as many as 129 patients. The subjects were selected by purposive sampling and met the inclusion and exclusion criteria.

Results: A total of 40 Guillain Barre Syndrome patients were studied, and 6 of them died. The majority were patients aged 36-50 years, and the male and female sexes were equally significant. A total of 89 patients with Myasthenic Crisis were studied, and 1 of them died, namely patients with the age category of 36-50 years and female.

Conclusion: Guillain Barre Syndrome mortality was 15%, and Myasthenic Crisis was 1.12% at Haji Adam Malik Center General Hospital Medan January 2021-December 2022.

Keyword : Guillain Barre Syndrome, Mortality, Myasthenic Crisis

1. Introduction

The Neuromuscular disorders are disorders in the nervous system that affect the muscular system. Some examples include Amyotrophic Lateral Sclerosis (ALS), Charcot-Marie-Tooth, Multiple Sclerosis, Lambert-Eaton Myasthenic Syndrome (LEMS), Guillain-Barre syndrome (GBS), Myasthenia Gravis (MG) and Spinal Muscular Atrophy (SMA).[1] Guillain Barre syndrome is an

autoimmune disorder of the nervous system characterized by polyradiculoneuropathy with progressive muscle weakness and reduced or nonexistent muscle reflexes. This disorder is found worldwide, with a prevalence of 1.9 cases per 100,000 inhabitants yearly. The disease has two peak ages, 15-35 and 50-75. The ratio of men and women is 3:2.[2] Guillain Barre syndrome can be managed with Plasma Exchange (PE) and Intravenous Immunoglobulin (IVIG). Despite implementation, about 3% of patients with this syndrome will die. The average duration of hospitalization ranges from 7 days, with about 25% of patients requiring intubation and mechanical ventilation.[3]

Myasthenia gravis is a peripheral nerve autoimmune disorder in which circulating antibodies work against components on the motor end plate of nicotinic acetylcholine (AChR) receptors, resulting in a decrease in the number of acetylcholine receptors and the potential for action in the synaptic membrane.[4] The peak age at the onset of the first symptoms of myasthenia is bimodal, i.e., 20 to 30 years in women and 50 to 60 years in men.[5] The cardinal sign of myasthenia gravis is weakness in the volunteer muscles or skeletal muscles fluctuating, especially in the muscles innervated by the motor nerve of the brainstem. The clinical symptom of a myasthenic crisis is a weakness of the respiratory muscles, which interferes with the airway and is life-threatening.[6] Plasma exchange therapy (PE) and intravenous immunoglobulin (IVIG) are patient-specific therapies. Patient mortality in the last four decades has fallen to 4.5% from 75%. Advances in patient management that combine cholinesterase inhibitor agents, immunosuppressive drugs, plasmapheresis, immunotherapy, and supportive care at ICUs give almost all patients a good life expectancy.[7]

2. Method

The study used a descriptive research design with data derived from the patient's medical records at the Haji Adam Malik Central General Hospital Medan. The research was carried out immediately after obtaining the approval of the ethics committee of the Medical Faculty of the Universitas Sumatera Utara with the issuance of an ethical clearance on August 14, 2023. The research was carried out at the Haji Adam Malik Central General Hospital Medan; subjects were obtained using a non-probability sampling technique with a purposive sampling method, i.e., taking subjects based on specific considerations made by researchers based on characteristics or characteristics of populations already known. The criteria for inclusion of the subjects are inpatients and outpatients with Guillain Barre syndrome and Myasthenic Crisis in January 2021-December 2022. The exclusion criteria were subjects with missing or incomplete data, diagnosed in the past time, and patients with other concomitant diseases that may cause death, who were not counted as the study subjects. Subjects meeting the inclusion and exclusion criteria were 129 patients. The analysis in this study uses a univariate analysis technique, which is a technique that aims to describe the condition being studied.

3. Result and Discussion

The data was carried out from August to September 2023 with patients consisting of 20 inpatients and 20 outpatients with Guillain Barre Syndrome, 8 Myasthenic Crisis inpatients, and 81 Myasthenic Crisis outpatients. The results of the study are presented in a characteristic and frequency in **Table 1**.

In a total of 40 Guillain Barre Syndrome patients, the majority were aged 51-75 years, as many as 11 patients (27.5%). The peak age in these patients corresponds to one of two peak age rates

of Guillain Barre Syndrome patients epidemiologically, namely in the age group of 51-75 years. As we age, the immune function also declines such as the immune system becomes slower in responding to pathogens, autoimmune diseases are more likely to develop, the healing process becomes longer, and the ability of immune cells to detect and correct damaged cells also decreases.[8] Of the patients, 22 (55%) were men, and 18 (45%) were women, with a ratio of 1.2: 1. This figure shows the predominance of Guillain Barre Syndrome patients in men, also by epidemiological data of this disease. The reason why men are more susceptible to Guillain Barre Syndrome is still unknown. Still, several studies reveal that men with advanced age are more vulnerable to immune diseases, so diseases related to immunity are dominated by men.[9] However, the ratio of male to female sex is slightly lower than the epidemiological comparison, which is 3:2.

Table 1. Demographic Characteristics of Research Subjects.

Characteristics	Guillain Barre Syndrome						Myasthenic Crisis					
	Total		Inpatients		Outpatients		Total		Inpatients		Outpatients	
Age	n	%	n	%	n	%	n	%	n	%	n	%
<15 years	10	25,0	4	20	6	30	0	0	0	0	0	0
15-35 years	8	20,0	3	15	5	25	31	34,8	2	25,0	29	35,8
36-50 years	10	25,0	7	35	3	15	29	32,6	5	62,5	24	29,6
51-75 years	11	27,5	5	25	6	30	28	31,5	1	12,5	27	33,3
>75 years	1	2,5	1	5	0	0	1	1,1	0	0	1	1,2
Sex												
Man	22	55,0	13	65	9	45	27	30,3	1	12,5	26	32,1
Woman	18	45,0	7	35	11	55	62	69,7	7	87,5	55	67,9
Total	**40**	**100**	**20**	**100**	**20**	**100**	**89**	**100**	**8**	**100**	**81**	**100**

Of 89 Myasthenic Crisis patients, the majority of patients aged 15-35 were 31 (34.8%). The peak age in these patients corresponds to one of two epidemiologic peak age rates of Myasthenic Crisis patients, namely in the age range of 20-30 years. In the study, 62 patients (69.7%) were women, and 27 patients (30.3%) were men, with a ratio of 2.3: 1. This figure shows the dominance of Myasthenic Crisis patients in the female sex by the epidemiological data of this disease. However, this ratio is slightly higher than the epidemiological ratio of 3:2.

Table 2. Clinical Characteristics of Guillain Barre Syndrome Patients.

Characteristics	Total		Inpatients		Outpatients	
	n	%	n	%	n	%
Weakness both legs	9	22,5	4	20	5	25
Weakness four limbs	26	65	14	70	12	60
Headache	3	7,5	0	0	3	15
Hard to breath	2	5	2	10	0	0
Onset						
<1 month	25	62,5	17	85	8	40
1–6 months	10	25	3	15	7	35
>6 months –1 year	2	5	0	0	2	10
>1-3 years	2	5	0	0	2	10
>3 years	1	2,5	0	0	1	5
Hospitalized period						
<1 week	8	40	8	40	0	0
1-2 weeks	6	30	6	30	0	0
>2-3 weeks	4	20	4	20	0	0
3 weeks-1 month	1	5	1	5	0	0
>1 months	1	5	1	5	0	0

Women at the age of 20-30 years are women in the fertile period, and some women at this age who have HLA-DR3 and B8 genes are known to make the body more susceptible to autoimmune disorders.[10] HLA-DR3 is often associated with an increased risk of developing non-organ-specific autoimmune diseases.[11] HLA-B8 is related to the early onset of myasthenia gravis and thymus hyperplastic events in Caucasian races.[12]

Table 3. Clinical Characteristics of Myasthenic Crisis Patients

Characteristics	Total		Inpatients		Outpatients	
	n	%	n	%	n	%
Eyelid weakness	51	57,3	0	0	51	63,0
Leg weakness	1	1,1	0	0	1	1,2
Limb weakness	8	8,9	0	0	8	9,9
Double vision	5	5,6	0	0	5	6,2
Hard to talk	2	2,2	0	0	2	2,5
Whistle sound	9	10,1	0	0	9	11,1
Hard to swallow	4	4,4	1	12,5	3	3,7
Hard to breath	9	10,1	7	87,5	2	2,5
Onset						
<1 month	18	20,2	6	75,0	12	14,8
1–6 months	30	33,7	1	12,5	29	35,8
>6 months–1 year	8	8,9	0	0	8	9,9
>1-3 years	13	14,6	1	12,5	12	14,8
>3-5 years	5	5,6	0	0	5	6,2
>5 years	15	16,8	0	0	15	18,5
Hospitalized period						
<1 week	4	50,0	4	50,0	0	0
1-2 weeks	3	37,5	3	37,5	0	0
>2-3 weeks	1	12,5	1	12,5	0	0
3 weeks-1 month	0	0	0	0	0	0
>1 months	0	0	0	0	0	0

In inpatient and outpatient Guillain Barre Syndrome patients, the majority of complaints experienced by patients that cause them to come to the hospital are limb weakness. This weakness initially arises from weakness in both legs, then spreads to the upper part of the body (ascending), and is symmetrical so that both hands.[13] It is experienced by 70% of inpatients and 60% of outpatients. Onset occurs within <1 month in 85% of inpatients and 40% of outpatients. The length of stay of Guillain Barre Syndrome patients is generally <1 week, as much as 40%. This is by the length of hospitalization of Guillain Barre Syndrome patients in general, which is about one week. The results of the study are presented in a characteristic table in **Table 2**.[3]

In hospitalized Myasthenic Crisis patients, the majority of complaints and onset experienced by patients to cause coming to the hospital are hard to breathe in 75% of patients and beginning for <1 month in 75% of patients. Shortness of breath is caused by widespread muscle weakness that occurs in the body to hit the respiratory muscles. In addition to respiratory failure, other complaints commonly found in Myasthenic Crisis patients are ptosis, diplopia, dysphagia, and weakness of the four limbs.[6] While in outpatients, the majority of complaints were eyelid weakness in 63% of

patients and onset for 1-6 months in 35.8% of patients. This weakness is a specific complaint in patients. In addition, typical weakness can also occur in the muscles of the movement of the eyes, face, mouth, throat, and neck.[4] The length of hospitalization of Myasthenic Crisis patients is generally <1 week, as much as 50%. This figure is slightly lower than the Myasthenic Crisis patients' typical hospitalization length of 8-12 days.[14] This is due to improved management of crisis patients.[15]

Table 4. Patient's Mortality Frequency.

Mortality	Total		Guillain Barre Syndrome		Myasthenic Crisis	
	n	%	n	%	n	%
Alive	21	42,8	14	70	7	87,5
Dead	7	32,1	6	30	1	12,5
Total	**28**	**100**	**20**	**100**	**8**	**100**

From 20 Guillain Barre Syndrome inpatients, the mortality result of this disease in inpatients was 30%. This figure shows that 6 out of 20 or 1 in 3 hospitalized patients die as seen in **Table 4.** The majority of patients who died were in the age category of 36-50 years, as many as three patients (50%) as seen in **Table 5**. This shows that patients in the age group of 36-50 contribute the highest mortality rate, younger than the risk factors for the age group of Guillain Barre Syndrome patients who experience death, which is >60 years.

This may be due to clinical symptoms already severe upon hospital admission, such as disability in the limbs.[16] Patients with advanced age are prone to death because they quickly experience physical deterioration, comorbidities, and side reactions to drugs that are more severe than patients with a young age.[17] Overall, mortality was 15%. This figure shows that 6 out of 40 Guillain Barre Syndrome inpatients and outpatients at Haji Adam Malik Central General Hospital Medan from January 2021 to December 2022 died. This figure is higher than the epidemiological data on mortality of this disease, which is about 3%. This is due to the patient's clinical symptoms at hospital admission that are already severe and the still unavailability of PE and IVIG therapy as Guillain Barre Syndrome specific treatments. In general, things that can cause a poor prognosis in Guillain Barre Syndrome patients include old age, severity of clinical symptoms when rushed to the hospital, history of surgery, autonomic dysfunction, disorders of cranial nerves, elevated levels of liver enzymes, high levels of CSF protein, gastroenteritis, presence of complications, use of ventilators, and low MRC (Medical Research Council) scores.[18]

Of the 8 Myasthenic Crisis inpatients, the mortality result of this disease in inpatients was 12.5%. This figure shows that 1 in 8 hospitalized patients die. These patients are 36-50 years old, according to the scope of risk factors for the age of Myasthenic Crisis patients to experience mortality, which is 40 years old. In general, things that can cause poor prognosis in Myasthenic Crisis patients include age <50 years, smoking history, thymomas, positive repetitive nerve stimulation, and positive acetylcholine receptor antibodies (AchR Ab).[19]

The patient is also female, corresponding to most patients' sexes in this disease. This is because women during their fertile period who have HLA-DR3 and B8 genes are known to make the body more susceptible to autoimmune disorders, and the resulting prognosis is worse.[10] Overall, mortality was 1.12%. This figure shows that 1 in 89 Myasthenic Crisis inpatients and outpatients at Haji Adam Malik Central General Hospital Medan in January 2021-December 2022 died. This figure is lower than the epidemiological mortality data for this disease, which is 4.5%. Mechanical

ventilation and crisis patient management have improved the quality of life and reduced mortality rates for these diseases.[20]

Table 5. Demographic Characteristics of Patients Who Died

Characteristics	Total		Guillain Barre Syndrome		Myasthenic Crisis	
Age						
<15 years	1	14,2	1	16,7	0	0
15-35 years	0	0	0	0	0	0
36-50 years	4	57,1	3	50,0	1	100
51-75 years	2	28,5	2	33,3	0	0
>75 years	0	0	0	0	0	0
Sex						
Man	3	42,8	3	50	0	0
Woman	4	57,1	3	50	1	100
Total	**7**	**100**	**6**	**100**	**1**	**100**

In general, the decreased Myasthenic Crisis mortality rate is due to advances in patient management that combine cholinesterase inhibitor agents, immunosuppressive drugs, plasmapheresis, immunotherapy, better diagnostic methods, and supportive care in the Intensive Care Unit, giving almost all Myasthenic Crisis patients a good life span.[7]

4. Conclusion

The mortality rate of Guillain Barre Syndrome at Haji Adam Malik Central General Hospital Medan January 2021-December 2022 was 15%, which is dominated by patients in the age category of 36-50 years (50%) and equal sex (50%) in men and women. The overall mortality rate for Myasthenic Crisis patients at Haji Adam Malik Central General Hospital Medan from January 2021 to December 2022 is 1.12%, which are a female patient in the age category of 36-50 years.

5. Acknowledgements

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6. Conflict Of Interest

We declare that there is no conflict of interest in this study.

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