

## RESPONSE AND CLINICAL OUTCOME OF CRITICALLY ILL CHILDREN WITH GUILLAIN BARRE SYNDROME ADMITTED TO PICU OF CHILDREN'S HOSPITAL LAHORE

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### ABSTRACT

**Objective:** To analyze patients with Guillain Barre Syndrome (GBS) requiring intensive care unit (ICU) admission, their course, response to treatment and outcome.

**Methods:** A retrospective study conducted at ICU of The Children's Hospital Lahore in 194 children with diagnosis of GBS from June 2018 to May 2020. Demographic profile, clinical features, treatment, duration of mechanical ventilation, length of ICU stay and outcome was noted.

**Results:** Mean age of children was  $5.95 \pm 3.1$  years with male predominance 136 (70 %). Major proportion were in range of 1-5 years 93(47.9%). Mean duration of ICU stay was  $21.3 \pm 34.4$  days. Out of 194 patients 125(64%) received IVIGs while 112 (57.7%) required mechanical ventilation because of rapidly progressive weakness within 24 hours of admission and those who remained on mechanical ventilation for longer period showed AMSAN (Acute motor sensory axonal neuropathy) type of GBS (p value <0.001). Tracheostomy was performed in 22.7% of patients who were difficult to wean off from ventilator. Among nerve conduction studies AIDP (Acute inflammatory demyelinating polyneuropathy) was predominant 48% associated with p value <0.001 in those who showed complete recovery, followed by AMSAN 24%, AMAN (Acute motor axonal neuropathy) 13.4% and miscellaneous 13.9 %. Regarding outcome complete recovery was seen in 28%, minimal residual disability in 70% and 1.5% remained ventilator dependent.

**Conclusion:** Patients of severe GBS often require prolong respiratory support and ICU care. The indicators of poor outcome are rapidly progressive course, severe weakness and axonal neuropathy leading to longer duration on mechanical ventilation.

**KEYWORDS:** Guillain Barre Syndrome, Outcome, intensive care unit (ICU)

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### INTRODUCTION

Guillain – Barre syndrome (GBS) is one of the main cause of acute flaccid paralysis (AFP). It is an ascending form of symmetrical paralysis starting from lower limbs, then involving upper limbs with areflexia or diminished reflexes. There may or may not be the involvement of autonomic nervous system or sensory system. CSF findings suggestive of GBS are elevated CSF proteins

but without an elevation in white blood cells. Respiratory insufficiency because of respiratory muscle weakness is a very serious complication of GBS. The GBS patients, who are severely affected need careful observation in intensive care setting and ventilatory support can prove helpful in securing life of sick patients.<sup>1</sup>

The symptoms usually evolve over a period of 14 days and clinical nadir is reached within 28 days. The worldwide incidence figures of Guillain Barre Syndrome vary from 1.1-1.8/100,000/year, but statistics are not sufficient regarding its incidence in Pakistan.<sup>2</sup> Respiratory paralysis due to respiratory muscle insufficiency can lead to death if not managed timely.<sup>3</sup>

The course of GBS in children usually advances

quickly that can be hazardous for life. If the diagnosis is made on time with subsequent guidance for proper supportive treatment, the outcome will be better definitely. Patients with swiftly advancing course, cranial nerves involvement or respiratory insufficiency should be monitored in ICU setting for supportive care and early administration of IVIGs.<sup>4</sup>

Children with rapidly progressive weakness leading to respiratory paralysis in ICU settings show axonal variant of neuropathy. Around 50% of the patients usually not respond to definitive therapy despite mortality being low. Decrease muscle power in the initial phase of disease as well as quickly advancing course are the predictors of bad outcome.

While treating patients with GBS along with recognizing bad prognostic indicators will help in wise usage of the minimal funds.<sup>5</sup> However supportive therapy including mental wellbeing, nutritional rehabilitation, pain relief, and physiotherapy are also important aspects of management.<sup>6</sup>

The purpose of the study was to analyze critically ill patients with GBS, their progression, need for respiratory support, response to management, and their outcome. As there is scarcity of studies on severe GBS patients requiring ICU admission so this study can determine the prognostic outcome in critically ill GBS patients.

## METHODS

This was a retrospective observational study, carried out in Pediatric ICU of The Children’s Hospital &The Institute of Child Health (ICH), from June 2018 to May 2020 after the approval from Institutional Review Board. We used a Proforma for data collection. Non-probability convenience sampling method was used. As the exact prevalence of GBS in the developing countries is not known, data of 2 years 194 patients of GBS presented to ICU was taken. Children of both gender with age range of 1 year to 16 years who were diagnosed as GBS on the clinical basis were included. While patients with anoxic brain insult, those who left against medical advice (LAMA) and those in which the final diagnosis was AFP other than GBS were excluded from the study. In all included patients, information about demographic profile, clinical presentation & subtypes of GBS were determined. The need for respiratory support with or without mechanical ventilation, duration of respiratory support and ICU

stay, tracheostomy need requiring ventilation for more than 14 days, NCS findings, administration of IVIGs in a dose of 2g/kg body weight over 3-5 days in patients with rapidly progressive muscle weakness within 24 hours of admission and subsequent response in the form of motor power improvement were evaluated. Motor power assessment was made on daily basis and outcome mentioned as complete recovery; those who will be weaned from ventilator & being able to walk, Minimal residual disability; those who will be weaned from ventilator but still needs some assistance and Ventilator dependent who were not weaned off from the ventilator. Data was analyzed by SPSS version 25. Descriptive statistics were used to describe the demographic details like age, gender, duration of ICU stay and duration of ventilation as mean and standard deviation (SD). Qualitative variables like need for respiratory support / mechanical ventilation, tracheostomy, IVIGs given and outcome presented by calculating frequency and percentages. A p-value of less than 0.05 was considered significant.

## RESULTS

A total of 194 patients with GBS fulfilling the inclusion criteria were included. The results of our study showed male predominance 136 (70.1%) over female 58 (29.9%) with mean age of 5.95 ± 3.16 years. Major proportion of patients were in range of 1-5 years constituting 93(47.9%) of total patients (Table I).

Table- I Demographics and clinical characteristics

Characteristics	Number (%)
Age	
1-5 Years	93 (47.9%)
5-10 Years	77 (39.7%)
>10 Years	24 (12.4%)
Sex	
Male	136 (70.1%)
Female	58 (29.9%)
Type of Respiratory Support	
None	69 (35.9%)
Nasal Cannula/FM	13 (6.7%)
Mechanical Ventilation	112 (57.7%)
Tracheostomy	44 (22.7%)
IVIGs	
Given	125 (64.4%)

Table II: Duration of Respiratory Support vs NCS

Duration of Respiratory Support	NCS				Total	p-value
	AIDP	AMSAN	AMAN	Miscellaneous		
1-14 days	71 (63%)	5(4.4%)	14(12.5%)	22(19.6%)	112(57.7%)	<0.001
15-28 days	21 (44%)	16(34%)	8(17%)	2(4.2%)	47(24.2%)	<0.001
29-56 days	2 (7.1%)	19(67.8%)	4(14.2%)	3(10.7%)	28(14.4%)	<0.001
>57 days	0(0%)	7(1%)	0(0%)	0(0%)	7(3.6%)	<0.001
Total	94(48.4%)	47(24.2%)	26(13.4%)	27(13.9%)	194	

Mean duration of ICU stay was  $21.37 \pm 34.46$  days. Out of 194 patients 125(64%) received IVIGs while 112 (57.7%) required mechanical ventilation because of rapidly progressive muscle weakness within 24 hours of admission and those with longer duration on mechanical ventilation showed AMSAN variety of NCS associated with significant p value of  $<0.001$  (Table II).

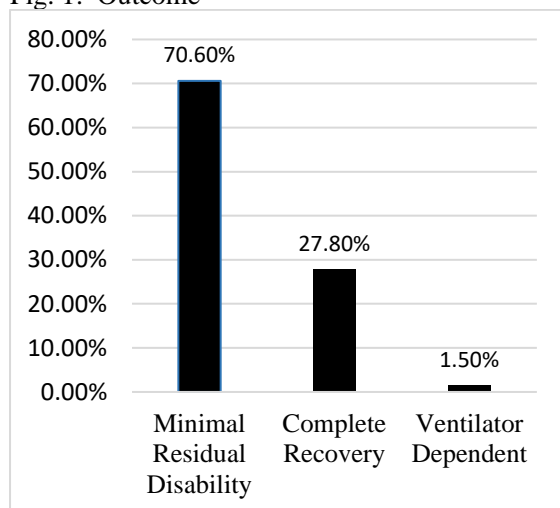
Tracheostomy was performed in 22.7% of patients who were difficult to wean off from ventilator. Among nerve conduction studies AIDP was commonest 48.5% and it was associated with complete recovery showing significant p value  $< 0.001$ , followed by AMSAN 24.2%, AMAN 13.4% and miscellaneous 13.9 % (Table III).

Table III: Outcome vs NCS

Outcome	NCS				Total	p-value
	AIDP	AMSAN	AMAN	Miscellaneous		
Complete Recovery	18(33.3%)	13(24%)	16(29.6%)	7(12.9%)	54(27.8%)	$<0.001$
Minimal Residual Disability	76(55.4%)	31(22.6%)	10(7.2%)	20(14.5%)	137(70.6%)	$<0.001$
Ventilator Dependent	0(%)	3(1%)	0(%)	0(%)	3(1.5%)	$<0.001$
Total	94(48.4%)	47(24.2%)	26(13.4%)	27(13.9%)	194	

Regarding outcome 27.8% underwent complete recovery, 70.6% with minimal residual disability and 1.5% remained ventilator dependent and there was no death (fig 1).

Fig. 1: Outcome



## DISCUSSION

GBS in children occasionally follows a rapidly advancing course often leading to respiratory paralysis that can be hazardous for life. Although clinical course of GBS in every patient is variable and difficult to predict but the role of supportive therapy could be beneficial. In this study 194 patients admitted to PICU of The Children's Hospital Lahore with severe GBS were enrolled. The mean age of the patients was  $5.95 \pm 3.1$  years that is compatible with research done by Asmat et al.<sup>2</sup> A mean age of 6.7 years was given by a study from Pakistan.<sup>7</sup> Major proportion of the patients were in range of (1-5 years) 93(47.9%) of the total patients and results were comparable to the study from Iran by Ali Akbar Momen et al. which showed 52.2% of children younger than 5 years.<sup>8</sup> It could be due to increased risk of infection and likelihood of peripheral myelinated nerves to demyelination in younger age

group. Similar results were found in studies from Iraq.<sup>9</sup> The results of our study also showed a male preponderance and that was comparable with other studies too. However, reason for this preponderance is unclear yet.<sup>2</sup>

Mean duration of ICU stay was  $21 \pm 34.4$  days. In a study Nikki Van Leeuwen et al. reported that 26 (31%) patients stayed in ICU, of which 17 (65%) were ventilated with median length of stay in ICU for 12 days. Among those with longer duration of stay had axonal neuropathy, rapidly progressive course and severe weakness at presentation leading to financial burden for hospital.<sup>10</sup>

Involvement of respiratory muscles leading to respiratory insufficiency usually found in 20% of patients with GBS and time period on ventilator may differ among patients. It fluctuates from weeks to months and in some cases could be in years.<sup>11</sup> In our study out of 194 patients 112(57%) of patients needed mechanical ventilation. In a research conducted by Christa Walgaar et al. at Netherland reported 27% of patients at ICU needed mechanical ventilation for a period of 4 weeks, while 71% of them needed ventilation for  $> 14$  days.<sup>12</sup> Tracheostomy should be considered when duration of ventilation expected to exceed for  $> 14$  days.

In this study 125(64%) of patients received IVIGs as a part of treatment 31.2% underwent complete recovery, 66.4% underwent minimal residual disability and 2.4% remain ventilator dependent showing p-value  $>0.05$  as statistically non-significant. It was generally observed that response to IVIG is favorable in demyelinating and disappointing with axonal neuropathy as said by a study published by Sarmad Hamdani et al. reported that results are still unclear regarding response of IVIG which is contrary to a research conducted by G Kanra et al. who reported considerable efficacy in treating children with GBS.<sup>13-14</sup>

NCS findings in our study showed AIDP as the most frequent subtype of GBS (48.5%), followed by

AMSAN 24.2%, AMAN 13.4% and miscellaneous 13.9 %. A latest study at Children's Hospital Lahore also showed AIDP as most common type with frequency of 67%. Similar results were reported by Prem Chand et al. as AIDP (58%) and results were comparable with study done by Asmat et al.<sup>2, 7, 15</sup>

Statistics regarding outcome were 28 % underwent complete recovery, 70 % with minimal residual disability and 1.5% remained ventilator dependent. Generally, GBS has a good prognosis in children with recovery in 85% of cases. Supportive care and rehabilitation is mandatory for achieving more rapid and global improvement. On the whole prognosis and outcome is good among younger children as reported by Jasem et al. that chances of residual paraparesis were greater in children above 5 years. In a study done by Korinthenberg et al. reported that most of the children 96% were only had minimal symptoms or even without any symptoms at the end of 41 weeks of diagnosis.<sup>(9)</sup> In a study conducted at Cairo University by Hafeez M. Bazara et al. showed favorable outcome as patient can take steps without support at discharge were documented in 58% of cases and severe neurological disability was reported in 13% by Rees et al.<sup>4,16-17</sup>

### STRENGTH AND LIMITATIONS

It is among the few studies in our setup about the demographics and outcome of the GBS patients admitted to pediatric intensive care unit. Limitation of the study is that it is a retrospective study, long-term prognosis of GBS could not be assessed.

### CONCLUSION

Prognosis of GBS in children is generally good. Patients of severe GBS often require prolong respiratory support and ICU care. However, indicators of poor outcome are rapidly progressive course, severe weakness and axonal neuropathy leading to longer duration on mechanical ventilation. Despite protracted course, results of our study showed that prognosis is good.

### ETHICAL APPROVAL

The study was approved by the Ethical Review Committee of The Children Hospital & the Institute of Child Health, Lahore, Pakistan. Vide reference No. 2020-154 dated October 14, 2020.

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**AUTHORS' CONTRIBUTION:**

**AJ:** Conceived, designed, manuscript writing

**NS:** Supervision and contribution in manuscript writing

**MS:** Critical Review

**FK:** Data Collection

**GS:** Review & suggestions

**UWA:** Data analysis