

Frequency and Factors Contributing to Respiratory Failure in Childhood Guillain–Barre Syndrome

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ABSTRACT

Introduction: Guillain-Barre Syndrome (GBS) is a common cause of acute flaccid paralysis (AFP) in children globally, and in the advanced world it has emerged as the commonest cause of AFP after eradication of poliomyelitis

Objectives: Objective of the study was to find out frequency of respiratory failure and factors contributing to respiratory failure in cases of GB syndrome.

Study Design: It is a prospective descriptive study.

Patients and Methods: Detailed account of patients presenting with diagnosis of G.B. syndrome was recorded in pre-designed proforma. This study was conducted over a period of 06 months in Department of Paediatrics Children's Hospital & The Institute of Child Health, Lahore. Statistical analysis was done using SPSS 14.

Results: There were a total of 110 patients with age range of 05 to 15 years. 43.6% were male and 56.4% were female. 26.4% developed respiratory failure, out of them 48.3% had bulbar involvement and 62.1% had autonomic involvement. Out of 81 patients who did not develop respiratory failure only 25.9% had bulbar involvement and 34.6% had autonomic involvement. A p value of 0.018 for autonomic dysfunction and 0.047% for bulbar involvement was found. Both p values are significant.

Conclusion: Bulbar involvement and autonomic dysfunction are contributory factors for respiratory failure in cases of severe GBS. Addressing these factors can bring an improvement in management.

Key words: Childhood Guillain–Barre syndrome, Respiratory failure, Autonomic Dysfunction, Bulbar involvement, Intravenous Immunoglobulin

INTRODUCTION

Guillain-Barre Syndrome (GB Syndrome) is one of the common causes of acute flaccid paralysis (AFP) in children ; is an autoimmune, post infectious poly neuropathy which mainly involves motor but sometime autonomic and sensory nerves^{1,2}In children incidence is 0.34 to 1.34 per 100,000, about 30% do develop respiratory failure and mortality rate is 3-5%^{3,4}. Spectrum of disease may vary, ranging from mild disease which only requires observation to severe disease with respiratory failure which needs intubation and mechanical ventilation. Commonest clinical presentation is symmetrical ascending paralysis which may be accompanied by pain and paraesthesias. Certain percentages also have autonomic and cranial nerve involvement⁵. Pathogenesis involves complex interplay of autoimmunity (secondary to infections) and host factors along with emerging evidence of polymorphism in immune response genes^{6,7}.

Immune responses may primarily damage myelin or may cause axon damage or sometime is mixed pattern^{8,9} Four forms of GB syndrome have been described in literature.

1. Commonest variety is Acute Inflammatory Demyelinating Polyneuropathy (AIDP)¹⁰.
2. Pure motor form without any involvement of cranial nerves, also called Acute Motor Axon Neuropathy (AMAN)¹¹.
3. Third variety, Miller Fischer, which accounts for 5% of the cases, is milder form¹²
4. Pharyngo-Cervico-Brachial (PCB) variant is characterized by difficulty in speech and swallowing often associated with facial nerve involvement¹³. Significant overlap of manifestations is found in different variants. Some degree of axonal injury is found in many cases of GB Syndrome, probably as part of "bystander" injury during the events of demyelination¹⁴. Pain is present in 50% and autonomic involvement in different series ranges from 16% to more than 50%^{15,16}.

Objectives:

1. To determine the frequency of respiratory failure in childhood GBS
2. To find out contributory factors leading to respiratory failure in childhood GBS.

PATIENTS AND METHODS

This was a prospective descriptive study ,conducted at Children’s hospital & ICH, Lahore and patients enrolled from Emergency Department , Medical Intensive Care Unit and Department of Pediatric Neurology over six months period(from06- 02- 2014 until 06-08 02014). Sample size was 110 patients, calculated with 95% confidence interval, taking 7% margin of error and expected prevalence of autonomic involvement of 16%.

Inclusion criteria:

1. All patients, both sex, admitted with diagnosis of GB Syndrome having ascending symmetric paralysis, minimal sensory involvement, decreased or absent deep tendon reflexes with supportive evidence on cerebrospinal fluid (CSF) examination, electromyography (EMG) and nerve conduction studies (NCS).

Patients having GBS, bulbar involvement, autonomic dysfunction and respiratory failure were diagnosed on following **Diagnostic criteria:**

1. **Guillain-Barre Syndrome:** It is post infectious acute demyelinating polyneuropathy presenting as symmetrical acute flaccid paralysis with evidence of demyelinating polyneuropathy on EMG and NCS.
2. **Bulbar involvement:** Involvement of IX, X, XI and XII cranial nerves resulting in paralysis of muscles of deglutition and speech, evident by absent/weak gag reflex or nasal speech found on clinical examination.
3. **Autonomic involvement** is defined as having any of the following manifestations i.e urinary or fecal incontinence or retention, bradycardia or tachycardia(heart rate <60 or >130 beats/min) and labile blood pressure(BP)(difference in diastolic BP 20 mm of Hg taken on the same day)during hospital stay prior to development of respiratory failure
4. **Respiratory failure:** Patients diagnosed as having GB Syndrome using accessory muscles of respiration, pCO₂(partial pressure of carbon dioxide) more than 60 or pO₂ (partial pressure of oxygen) less than 60on arterial blood gases during hospital stay was considered as criteria for labeling as respiratory failure

Exclusion criteria:

1. Patients with GBS with concomitant chest infection resulting in respiratory compromise
2. Patients with GBS who had already received IVIG or plasmapheresis
3. Patients with history of recent diphtheria infection or lead exposure
4. Patients with serum potassium level <3.5 mmol/dl
5. Unvaccinated patients with asymmetric paralysis

Patients meeting study criteria were enrolled, informed consent was taken from their parents, EMG, NCS and CSF examination was carried out. Information which was noted in pre-designed proforma included patients’ name, age, medical record number and frequency of children who developed respiratory failure. Frequency of factors like bulbar involvement and autonomic dysfunction was noted in patients with and without respiratory failure.

Statistical analysis

SPSS 14 was used for data and analysis, Qualitative variables like gender, respiratory failure, bulbar involvement and autonomic involvement were presented by frequency and percentages. Quantitative data like age was presented by mean ± standard deviation. Continuous variables were expressed as mean± standard deviation. For categorical variables, the chi-square test was used. Values for *P*<0.05 were considered statistically significant.

RESULTS

A total of 110 patients, age range 5-15 years were included in this study over a period of six months. Majority were between 5-10 years (75.5%) while 24.5% were between age of 11-15 years. Mean age was 8.23 years ±2.89 years. Gender distribution revealed that 43.6% were males and 56.4% were females (Table No 1).

Respiratory failure was observed in 29 cases (26.4%), while 73.6% did not develop respiratory failure. Out of those 29 cases which developed respiratory failure, bulbar involvement was found in 48.3% and autonomic involvement in 62.1%.

Of the 81 patients who did not develop respiratory failure only 25.9%had bulbar involvement and 34.6% had autonomic dysfunction. P value for autonomic dysfunction and bulbar involvement are 0.018 and 0.047 respectively.

ORIGINAL ARTICLE

Table 1: Distribution of patients by age & gender.

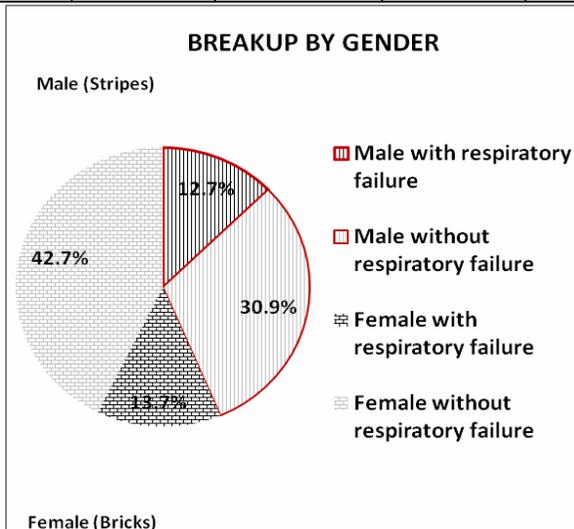
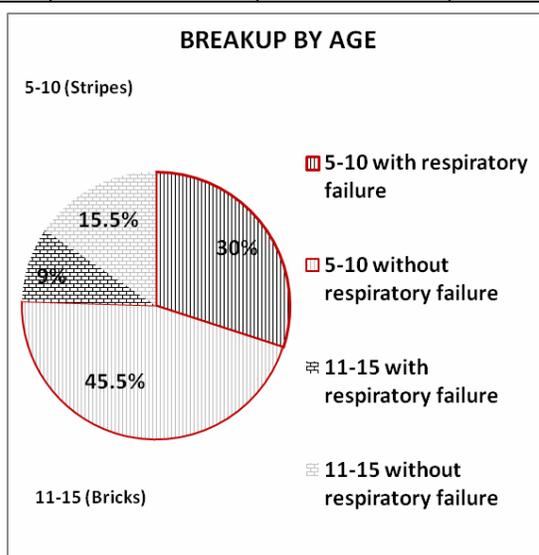
Age	Number	Percentage	Gender	Number	Percentage
5-10 Y	63	75.5%	Male	48	43.6%
11-15 Y	27	24.5%	Female	62	56.4%
Total	110	100.0		110	100.0

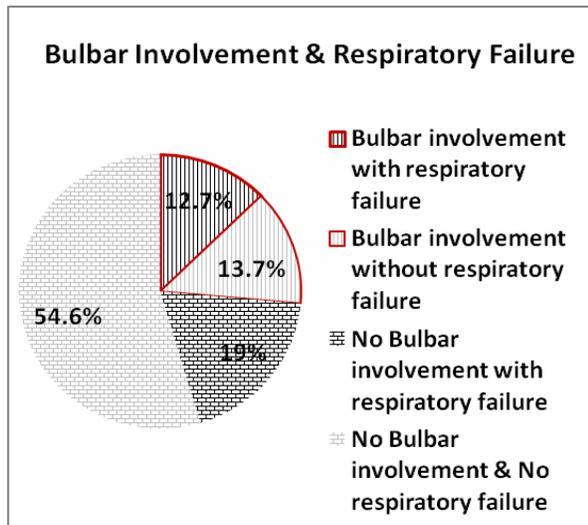
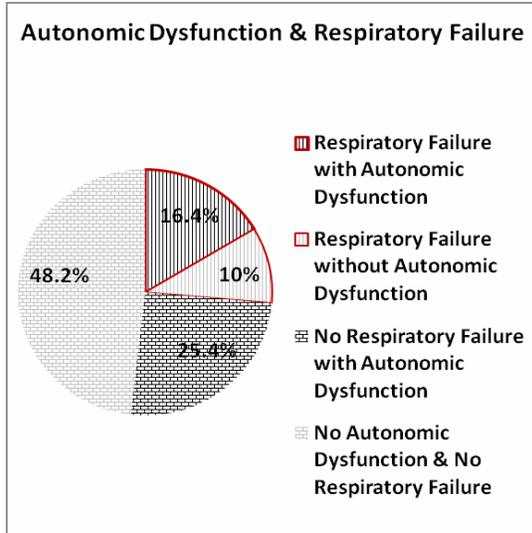
Table 2: Stratification of age and gender with regard to respiratory failure

Bulbar involvement	Respiratory failure		Total	Autonomic involvement	Respiratory failure		Total
	Yes	No			Yes	No	
Yes	14(48.3%)	21(26%)	35 (100%)	Yes	18(62%)	28(34.5%)	46
No	15(51.5%)	60(74%)	75 (100%)	No	11(38%)	53(65.4%)	64
Total Chi square= 4.917 P value= 0.047	29	81	110	Total Chi square= 6.63 P value=0.018	29	81	110

Table 3: Bulbar involvement and autonomic involvement leading to respiratory failure.

Age	Respiratory failure		Total	Gender	Respiratory failure		Total
	Yes	No			Yes	No	
5-10 Y	19(22.8%)	64(77.1%)	83 (100%)	Male	14(29%)	34(71%)	48(100%)
11-15 Y	10(37%)	17(63%)	27 (100%)	Female	15(24%)	47(76%)	62(100%)
Total	29	81	110		29	81	110
			Chi square= 62.100 P value= 0.231				Chi square= 0.345 p value= 0.712





DISCUSSION

Since the virtual elimination of poliomyelitis, in western world GBS is now the most common cause of acute flaccid paralysis.¹⁷In both developing and the developed world it remains an important entity to be recognized early as prompt intervention can be lifesaving.

Diagnosis of GB syndrome is chased by suggestive history, characteristic physical findings and supported by investigations which are findings in nerve conduction studies and abnormal findings on CSF examination. albumino-cytological dissociation (raised CSF proteins and normal cell count) is characteristic finding in GB syndrome^{18,19}. Patients developing respiratory failure frequently have associated feature of rapid disease progression, bulbar involvement and facial nerve

involvement at presentation, autonomic involvement and axonal variety²⁰. Treatment modalities with proven efficacy are intravenous immunoglobulins (IVIG) and plasma exchange; these have been found effective in shortening duration of ailment as well as need for ventilation in patients with respiratory failure.²¹

Autonomic dysfunction (AD) occurs in almost two- thirds of patients of GBS, can be potentially life threatening, is often associated with derangements of cardiovascular, vasomotor, bowel and bladder functions. It does involve both sympathetic and parasympathetic systems.^{22,23}

Results of present study did not show any significant disparity in gender distribution, however, literature describes an increased prevalence in males.²⁴GBS can affect any age group, however it is more frequent after the age of 40, and, usually affects children after the age of three years.^{25,2} Our study revealed respiratory involvement in 26.4% which is close to the result of studies by McGrogan et al and Shubhangi et al^{3,26}While another study has shown respiratory involvement of 10%.²⁷

Amongst 29 cases which developed respiratory failure, bulbar involvement was found in 48.3% and autonomic involvement in 62.1% with significant p value (table No 3). Among patients who did not develop respiratory, 21 patients (25.9%) had bulbar involvement and 28 patients (34.6%) had autonomic dysfunction. In one study on 114 patients 38 had autonomic dysfunction out of which 22 needed mechanical while 12 did not. In the same study 69 patients had bulbar dysfunction out of which 49 needed mechanical ventilation while 20 did not.²⁸

P values for autonomic dysfunction and bulbar involvement are 0.018 and 0.047 respectively. This shows that both factors have significant contribution to respiratory failure in patients with GBS. Similar evidence has been generated in other studies^{29,30}

Autonomic dysfunction and bulbar involvement pose a significant risk factor for development of respiratory failure in patients with GBS. Early detection and management of AD and bulbar involvement can lead to better outcome and avoidance of complications

CONCLUSION

In conclusion, respiratory failure occurred in 26.4% of patients in GB s and affected both male and female gender. A statistically significant difference was found; frequency of autonomic dysfunction

and bulbar involvement was more in cases which had respiratory failure. Both these factors contributed towards respiratory failure. Understanding these risk factors of severe GBS will provide better treatment strategies and improve the outcome

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