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Original Research Paper

Proportion of Guillain Barre Syndrome and their outcome among children presenting with acute flaccid paralysis

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ABSTRACT:

Background: Guillain Barre syndrome (GBS) is a common cause of acute flaccid weakness in children with long term morbidity in term of poor functional outcome. **Aim**: To estimate the frequency of GBS and to determine contributing factors for poor functional outcome. **Methods**: Retrospective study. Two year data of children presented with acute flaccid paralysis (AFP) were retrieved from hospital records. Outcome measures of GBS were need for mechanical ventilation (MV), mortality and functional status at 6 month. **Results**: Out of 47 cases of AFP, 39 (83%) had GBS. Summer season witnessed lowest number of cases. Median age of GBS patient was 9 year, 64% males. At presentation 90% had disability score>=3. AMAN (49%) most common subtype. 28% patient needed MV. Mortality was 15%. Out of 33 survived patient,11 (33.33%) had poor functional outcome at 6 month. High disability score (p=o.o49), long duration between symptom onset and admission (p=0.047), rapid progression (p=0.03) and AMAN variety (p=0.0031) were associated with poor outcome. **Conclusion**: Long term disability is common in children having GBS. Atonal type of nerve injury is poor predictive factor both for mortality and functional outcome.

Keywords: Acute flaccid paralysis, Guillain Barre syndrome, Disability Score

INTRODUCTION:

Acute flaccid paralysis (AFP) is characterised by acute onset of weakness and paralysis with reduced muscle tone. Guillain Barre syndrome (GBS), Acute Transverse myelitis (TM), paralytic Poliomyelitis, traumatic neuritis repreaent the most common causes of AFP. The other causes are hypokalemia, encephalitis, peripheral neuritis, myasthenia, and other myopathies etc. [1]

GBS is the most common cause of AFP with an overall incidence of 1.1 - 1.8/ 100000/year.[2] The most common clinical presentation includes bilateral ascending weakness with absence of deep tendon reflexes. [3,4] Remission is seen in some patients after 7 - 14 days after disease onset, but life threatening complications and long term disabilities are commonly seen. [5] Long term outcome is different in adults and children. Recovery is faster and better in children than adults, still long term disability in children are significant and poorly defined. [3,6] The treatment modalities are administration of intravenous immunoglobulin (IVIG) and plasma exchange. [2] Effect of both modalities in halting disease progression and fastening the recoveryis equivalent. Studies on effect of treatment modalities on the outcome of GBS found no difference. [7,8] Hence other factors apart from treatment modality might be associated with functional outcome in children with GBS. [2] This study was planned to evaluate the frequency of GBS among children having AFP and to predict the contributing factors for poor outcome in children with GBS.

MATERIALS AND METHODS:

Study Design: A retrospective hospital based observational study was conducted in pediatric ward of tertiary care center. Data from the case records of patients having acute onset of flaccid paralysis attending pediatric OPD or admitted in pediatric ward from January 2022 to December2023 were collected. 44 cases were recruited in the study based on final diagnosis of acute flaccid paralysis. Ethical clearance was obtained from Institutional Ethical Committee.

Data collection: Acute flaccid paralysis (AFP) is a notifiable disease in our region. Etiological diagnosis for AFP was made based on clinical presentation, Nerve

conduction study (NCV), CSF examination, MRI brain/ spine finding. Daily record of clinical condition particularly neurological status was being maintained in a hospital case sheet for all patients. Patients were followed up in neurology clinic after discharge, where records of their clinical condition were maintained in register. Data pertaining to the study were retrieved from these records.

Accordingly details like socio demographic data, symptoms & signs on admission, time duration between symptom onset and admission, history of antecedent event, Hughe's Guillain- Barre syndrome (GBS) disability score [2,9], need for respiratory support, treatment during admission, condition on discharge and on follow up up-to 6 month after discharge were noted.

Outcome measures were need for respiratory support or mortality during hospitalization and time to achieve independent walking after onset of weakness in survived patient.

Statistical Analysis: Collected data were entered in MS Excel sheet and analysed by Open Epi online software. Categorical data were presented as frequency (%). Quantitative data were presented as mean and SD or median and IQR according to distribution of data. Chi Square testfor qualitative data and Mann Whitney U test for quantitative data was applied for statistical significance.

RESULTS:

Over a two years period, 47 patients had presented with acute flaccid paralysis. GBS constituted 83% of patients as a cause of AFP. Three patients (4.2%) had Acute Transverse Myelitis, out of which two had a preceding history of Mumps. Out of two patients of ADEM onehad preceding history of Measles. Three patients had Bell's palsy, were treated on OPD basis andwere not included in further statistical analysis. (Table 1)

Table 1: Etiological distribution of AFP (N=47)

Etiology	Frequency (%)
GBS	39 (82.9%)
Transverse Myelitis	3 (6.3%)
ADEM	2 (4.2%)
Bell's Palsy	3 (6.3%)

Figure 1: Cases were scattered over whole year with highest number of cases seen betweenJanuary to March and between September to December.

Figure 1 : Month wise distribution of AFP cases (N =44)

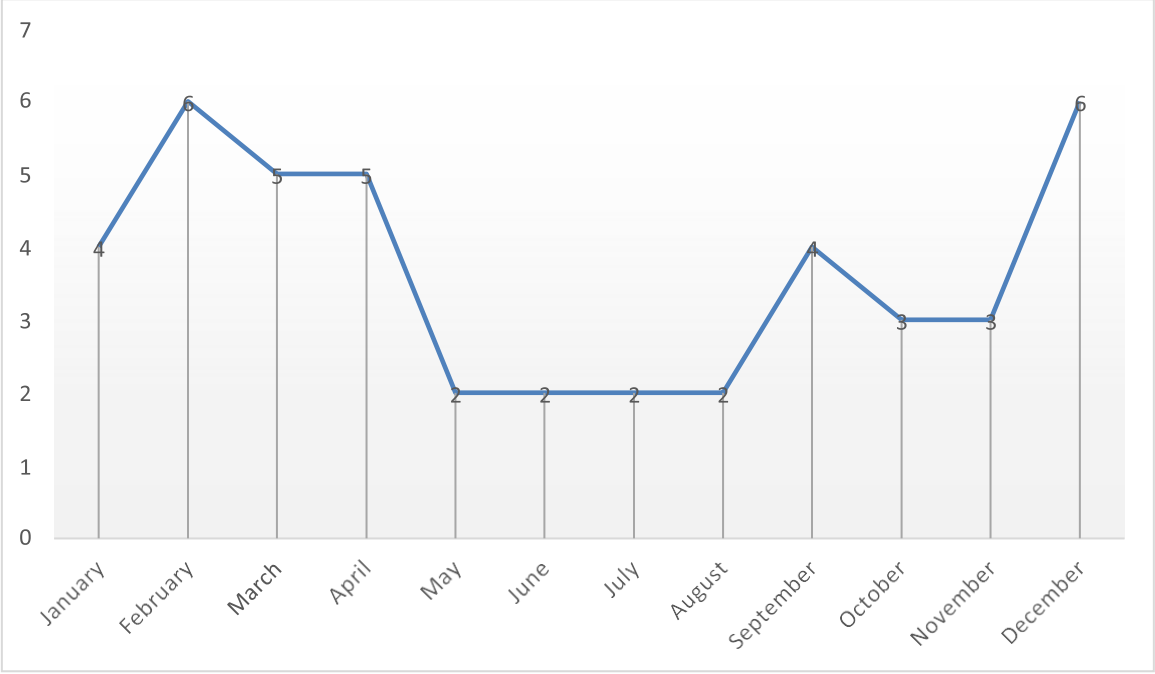

Table 2: Demographic & clinical characteristic (N =44)

Variable	GBS (n=39)	Non GBS (n=5)	p value (<0.05)
Age (median, IQR)	9,8	11, 4.5	0.5
Male Gender	26 (66.7%)	1(20%)	0.146839
Antecedent events (yes)			
Respiratory tract illness	7 (63.63%)	--	np
Diarrhea	2 (18.2%)	---	np
Other	--	3 (66.7%)	np
Total	11 (28.2%)	3 (60%)	0.1618
Duration between antecedent event &onset of weakness (median, IQR)	1, 5	4, 11	0.16853
Duration between weakness onset -admission (median, (IQR)	4, 5	3, 9.5	0.36317
Duration between weakness onset - nadir(median, IQR)	8,8	7, 21.5	0.43251
Neurological symptom at admission			
Pain	12 (30.8%)	1 (20%)	0.26272
Lower Limb weakness	28 (71.8%)	4 (80%)	0.489166
Arm weakness	19 (48.8%)	2 (40%)	0.481092
Sensory symptom	2 (5.1%)	1 (20%)	0.08648
Asymmetric weakness	4 (10.2%)	1 (20%)	0.264606
Cranial nerve involvement	3 (7.7%)	0	np
Autonomic dysfunction	0	0	np
Areflexia	30 (76.9%)	3 (60%)	0.13032
Bladder involvement	1 (2.6%)	2 (40%)	0.18352
NCV performed			
Total performed	39 (100%)	2 (40%)	0.00001
Abnormal NCV	39 (100%)	0	
MRI performed			
Total performed	6 (15.4%)	3 (60%)	0.129786
Abnormal	3 (50%)	1(33.3%)	
CSF Examination performed			
Total performed	1 (2.6%)	3 (60%)	np
CSF pleocytosis	0	1 (33.3%)	
Raised CSF Protein	0	0	
Length of hospital stay (median, IQR)	8, 6	5, 10.5	0.16354
Time to achieve independent walking indays (median, IQR)	100, 20	105, 100	0.40417

***np- not performed**

Table 3: Nerve conduction study characteristic related to GBS (N=39)

Nerve conduction study finding	Frequency (%)
AMAN	19 (48.7%)
AMSAN	05 (12.8%)
AIDP	12 (30.7%)
Mixed polyneuropathy	03 (7.6%)

Table 4: GBS Disability score on admission (N=39)

Score	Frequency n (%)
1	0
2	4 (10.25)
3	20 (51.28)
4	11 (28.2)
5	4 (10.25)

Table 5 : Characteristics according to need of MV & Outcome (N=39)

Variable	Need for MV(N=11)	No MV (N=28)	P value		Death (n=6)	Survived(n=33)	P value
Age (median, IQR)	11, 9	9,9	0.06944		9.5, 9	9, 9.5	0.26109
Male Gender (%)	4 (63.63%)	19 (67.8%)	0.403797		5 (83.3%)	21 (63.4%)	0.17981
Symtoms onset &nadir in days (median, IQR)	7, 10	10, 7.5	0.04947		8.5, 12	10, 8	0.31207
GBS subtype							
AMAN	7 (63.4%)	12 (42.9%)	0.08157		6 (100%)	13 (39.4%)	0.002676
AIDP	1 (9%)	11 39.3%)			0	12 (36.4%)	
ASMAN	3 (27.3%)	2 (7%)			0	6 (18.1%)	
Mixed	0	3 (10.7%)			0	3 (9%)	
Disability score onadmission (median, IQR)	4, 1	3, 0.5	0.040902		4, 1	3, 0.5	0.040902
Need for MV	--	--	---		6 (100%)	5 (15.15%)	0.00001
Duration of MV (median, IQR)	11, 12	--	np		11.5, 12	10, 22.5	0.261681

***Np – not performed, *mv- mechanical ventilation**

Table 6: Predictors of poor functional outcome at 6 months (n=33)

functional status Parameter	Good (Disability score 0 - 2) N= 22	Poor (Disability score >2) N= 11	p value (<0.05)
Age (median, IQR)	9, 9.5	10, 11	0.37448
Male Gender	8 (66.66%)	6 (54.54%)	0.22923
Disability score on admission(median, IQR)	3,0	4, 1	0.03144
Duration between antecedent event& onset of weakness	5, 8.5	0, 4	0.06552

(median, IQR)			
Duration between weakness onset -admission (median, IQR)	4, 6	7, 6	0.04746
Duration between weakness onset -nadir (median, IQR)	14, 8	8, 8	0.03673
Need for mechanical Ventilation	3 (13.63%)	1 (9.09%)	0.342443
Type of nerve injury			
AIDP	9 (40.9%)	3 (27.27%)	0.003124
AMAN	5 (22.72%)	8 (72.7%)	
AMSAN	5 (22.72%)	0	
MIXED	3 (13.63%)	0	

Table 2,3 & 4: The median age for GBS was 9 years, youngest patient was of one year. Males (64%) were predominant. Antecedent events were noted in 14 patients (32%), At presentation 51% children with GBS had disability score of 3 and 28% had score of 4. AMAN variety (49%)was the most common subtype followed by AIDP (31%). All patients of GBS were treated with intravenous immunoglobulin. The median duration to achieve independent walking was 100 days. Non GBS cases had median age of 11 years with male (25%) predominance, asymmetrical weakness in 60% (p = 0.0065), Bladder involvement at presentation in 40% (p = 0.00001) and CSF pleocytosis in 60% (p value 0.00001)

Table 5: Respiratory support in form of mechanical ventilation was needed in 28% of GBS patients. Patient requiring mechanical ventilation had higher disability score at presentation (p=0.040902) and shorter median duration between symptom onset to complete involvement (p=0.049). Mortality was observed in 6 (15%) cases. Statistically significant mortality was observed in AMAN subtype (p=0.0026).

Table 6: Out of 33 survived patients, eleven (33.33%) had median disability score >2 at the end of 6 months follow up. Statistical significance for poor functional outcome was noted with AMAN subtype (p=0.003124), longer duration between onset of weakness and admission (4 days Vs 7 days, p=0.047). High disability score on admission (p=0.03144) and rapid progression (14 days Vs 8 days, p=0.036).

DISCUSSION:

Present study showed GBS as the most common cause of AFP in children. Patients were scattered over the whole year, however summer months had lowest number of patients in current study. The median age was 9 years with a male predominance in current study which is similar to finding noted in previous studies. [3,6,9,10,11]. All patients presented with walking difficulties and 90% had disability score of >=3 at presentation. Shangab M et al noted 64.7% patient bed bound at presentation. [2,11,12]. Transverse Myelitis patients had bladder involvement (40%, p= 0.029) and asymmetrical weakness (60%, p= 0.0065) at presentation as compared to GBS patients. However other features like the demography, neurological sign/ symptom and clinical course were similar to patients with GBS which is similar to observation noted in other studies. [3]

We observed AMAN subtype (49%) as the most common form of GBS. The prevalence of GBS subtypes in other studies were: 46.6% AIDP, 30.2% AMAN, 6.8% AMSAN, 6.1% Miller Fisher (MF) and 7.9% unclassified.[3] However compared to western countries AMAN is more common in Eastern countries.[13]

In our study 28% required mechanical ventilatory support during hospitalisation. The need for ventilatory support is estimated between 20% to 30% in children suffering from GBS.[3] Different studies had reported need for ventilatory support in range of 3.7% to 24.4%.[3] Similar to our finding, patients with a higher disability score at presentation were more likely in need of ventilatory support.[14]

We observed mortality rate of 15% (6 out of 39 patients) in present study. The highest reported mortality in acute phase was 11.5% [15]. The observed overall mortality rate with long follow up of up to eleven years in other study was 2.6% [3]. All the patients who died were of AMAN subtype in our study. Two patients were referred from other centre, one on 12th day and other on15th day after onset of weakness. Four patients had > 10 days duration of ventilatory support The higher mortality rate in present study can be due to ventilatory associated complications andprolonged mechanical ventilation.

Time to achieve independent walking in our study was median duration of 100 days with a range of 20 to 200 days in GBS patients. The mean duration to achieve independent walking was 68.2 (16.8) days by Agrawal et al [16] and 2.97 (3.02) months by Barzegar et al [17]. Chaweekulrat et al developed prognostic scoring system in which score of 5 required mean duration of 34 days while score of zero required mean of 158 days to achieve independent walking over 8 years follow up [18].

Barzegar et al [17] determined disability score of >3 as a poor predictor of
independent walking. Similarly we also observed statistical significance for poor functional outcome in patients with a higher disability score at presentation. The other poor predictor for independent walking was rapid progression to maximal weakness and AMAN subtype in our study which is similar to finding observed by other authors.[2,19].
The limitation of the study is retrospective nature and relatively small number of patients. However the strength of the study being the inclusion of data up to 6 month follow up of theparticipants.

CONCLUSION:

GBS is the most common cause of AFP in children. Walking difficulty with areflexia was the most common clinical presentation. Higher disability score, axonal type of nerve injury, rapid progression to maximal weakness were poor predictors of functional outcomeat 6 month.

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