ORIGINAL ARTICLE

Guillain-Barré Syndrome: A Retrospective Study in the Department of Critical Care of Pediatrics in a Tertiary Care Hospital

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Abstract:
The cross sectional, retrospective descriptive study was done to document the number of GBS patient needed critical care, pattern of their presentation, management, complications and outcome in Pediatric intensive care unit (PICU) of Dhaka Shishu Hospital from January 2011 to December 2013. Data was collected from the admission record file in PICU. A total of 30 patients aged 1 to 15 years diagnosed as Guillain-Barré syndrome were included in the study. Detailed history, clinical examination, investigations, treatment modality and outcome for GBS was recorded including age, gender, cause of transfer to ICU, antecedent illness, respiratory muscles and bulbar paralysis, cranial nerve involvement, sensory and autonomic dysfunction, CSF study. During the study period, a total of 214 patients were diagnosed as GBS in the hospital and 30 patients were transferred to PICU. Among those, 29 (96.7%) patient were classical GBS and 1 (3.3%) relapse GBS. Common cause of transfer to ICU was respiratory muscle and bulbar paralysis (27, 90%). Most patients were in the age range of 1-5 years (20, 66.7%). Autonomic dysfunction was present in 16 (53.3%) cases and cranial nerve involvement in 1 (3.3%) cases. In 22 (73.3%) patients the antecedent event was respiratory infections and diarrhea in 2 (20%) cases. CSF study was done in 25 (83.3%) patients and albuminocytological dissociation was found in 19 (76%) cases. Total 26 (86.7%) patients were treated with IVIG and rest were with steroid (4, 13.3%). 12 (40%) patients needed mechanical ventilation, respiratory muscle paralysis (9, 75%) was the commonest indication. Most common complication during treatment was aspiration pneumonia (14, 46.7%). Twenty-three patients (76.7%) were improved and transferred to ward, 7 (23.3%) expired. Bulbar involvement with respiratory failure is the most common indication of admission in ICU. IVIG is the effective treatment modality and need of mechanical ventilation indicates poor outcome.

Introduction:
Guillain-Barré syndrome is a post infectious polyneuropathy presumed to be immune mediated and manifests as acute flaccid paralysis (AFP) involving mainly motor, sometimes also sensory and autonomic nerves. It affects people of all ages including pediatric age group. The paralysis usually follows a nonspecific viral infection such as respiratory tract infection and acute gastroenteritis by one to two weeks. It is the commonest cause of acute flaccid paralysis (AFP) after eradication of poliomyelitis¹,²,³. An incidence of 0.5-5/100000 children/year has been reported worldwide⁴,⁵,⁶.

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muscle weakness and areflexia. Weakness usually begins in the lower extremities, progressively involves the trunk, the upper limbs and finally the bulbar muscles. Bulbar involvement occurs in about half of the cases that results respiratory insufficiency. This interferes with eating and increase the risk of aspiration. Respiratory effort must be monitored to prevent respiratory failure and respiratory arrest. Urinary incontinence or retention is a complication in about 20% of cases. The autonomic nervous system is also involved in some cases where cardiovascular monitoring is important. Patients in early stages of this acute disease should be admitted to the hospital for observation because the ascending paralysis can rapidly involve respiratory muscles during the next 24 hours and may need mechanical ventilation.

With regard to clinical course and prognosis, classical Guillain-Barré syndrome has to be differentiated from variants with accompanying central nervous system inflammation and from chronic inflammatory demyelinating polyneuropathy.

Diagnosis of GBS is usually clinical. In addition to routine investigations, CSF examination, electrophysiological and nerve conduction studies and plasmapheresis were usually done. CSF shows no pleocytosis and protein is variably normal or mildly elevated.

Rapidly progressive ascending paralysis is treated with intravenous immunoglobulin. Plasmapheresis and/or immunosuppressive drugs are alternatives if IVIG is ineffective. Supportive care such as respiratory support, treatment of secondary bacterial infection is important.

Prognosis is good with complete recovery in more than 95% patients with GBS but it usually takes weeks to months, 3% mortality due to respiratory and autonomic involvement. It is necessary to have the knowledge regarding practical scenario of critically ill patients of GBS, their treatment in ICU and overall outcome which will ultimately help to modify the overall management plan in limited critical care resource.

Materials and method:

The study was conducted at the pediatric intensive care unit (PICU) of Dhaka Shishu (Children) Hospital from January 2011 to December 2013. During this period 214 patients aged 1 to 15 years were admitted and diagnosed as GBS, out of those 30 patients were transferred to the PICU and included in this study. Guillain-Barré syndrome was diagnosed clinically and the patients with pseudo paralysis, CNS infections and encephalopathy, chronic flaccid paralysis and stroke were excluded.

Detailed history and clinical examination for distribution of weakness, cranial nerve involvement, sensory loss and autonomic dysfunction and involvement of respiratory muscles and bulbar paralysis was recorded. Routine investigations including CBC, ESR, serum electrolytes and random blood sugar were done in all patients. CSF examination was done in the second week of illness. Nerve conduction studies and electromyography were not done due to unavailable resources.

Treatment modalities including supportive, Intravenous Immunoglobulin (IVIG) and steroids were selected in patients with GBS depending upon indication and facilities available. Indications for IVIG were rapidly
progressive disease, paralysis or impending paralysis of respiratory muscles, dysphagia and involvement of autonomic nervous system. Data was collected in pretested questionnaire and results were analyzed using SPSS version 17.

Results:

A total of 215 patients were admitted in DSH with the diagnosis of acute flaccid paralysis, 214 were GBS and one transverse myelitis. Out of these, 14% patients were transferred to the PICU. Among these GBS patients, 29 (96.7%) were classical GBS and 1 (3.3%) was a relapsing case. Most patients in this study were in the age range of 3-5 years (12, 40%) and other age groups were 1-2 years 8 (26.7%), 6-10 years 8 (26.7%) and more than 10 years 2 (6.6%); no patient were below 1 year age (Table-I). In this study male female ratio was 1:1.

Common cause of transfer to ICU was respiratory muscle and bulbar paralysis 27, (90%), other causes were autonomic involvement, aspiration pneumonia and for close monitoring. In 22 (73.3%) patients the antecedent event was respiratory tract infections (RTI) and diarrhea in 2 (30%) cases. In 2 (6.7%) cases there was no significant preceding illness. Regarding clinical assessment, areflexia and paraesthesia was present in all 30, (100%) patients, autonomic dysfunction was present in 16 (53.3%) and cranial nerve involvement in 1 (3.3%) cases (Table-II).

Cerebrospinal fluid study was done in 25 (83.3%) patients and albuminocytological dissociation was found in 19 (76%) cases. The criteria for albuminocytological dissociation were CSF protein more than 80mg/dl and cells less than 10/mm³.

Out of 30 patients, 26 (86.7%) were treated with IVIG and rest were treated with intravenous steroid (4, 13.3%) as they could not afford IVIG. Mechanical ventilation in course of treatment was needed in 12 (40%) cases out of total 30. Respiratory muscle paralysis (9, 75%) was the commonest indication of mechanical ventilation, cardiac arrest (3, 25%) was another reason.

During treatment in ICU, 14 (46.7%) patients developed aspiration pneumonia, cardiac arrest 3 (10%) and autonomic involvement was 5 (16.7%) cases.

Out of 30 patients, 23 (76.7%) patients were improved and transferred to ward, there mean duration of ICU stay was 19.7 (SD: 10.2; median 21) days. On the other hand, 7 (23.3%) patients expired during treatment in ICU, there mean duration of ICU stay was 5.1 (SD: 1.6; median 6.0) days.

Table-I: Age-group distribution of the GBS patients admitted in ICU (n=30)

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<thead>
<tr>
<th>Age-group</th>
<th>Number</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>1-2 yrs</td>
<td>8</td>
<td>26.7</td>
</tr>
<tr>
<td>3-5 yrs</td>
<td>12</td>
<td>40.0</td>
</tr>
<tr>
<td>6-10 yrs</td>
<td>8</td>
<td>26.7</td>
</tr>
<tr>
<td>&gt;10 yrs</td>
<td>2</td>
<td>6.6</td>
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Table-II: Major Clinical Findings of the Enrolled Cases (n=30)

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<tr>
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<td>Areflexia</td>
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<td>30</td>
<td>100</td>
</tr>
<tr>
<td>Autonomic dysfunction</td>
<td>16</td>
<td>53.3</td>
</tr>
<tr>
<td>Cranial nerve palsy</td>
<td>01</td>
<td>3.3</td>
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Discussion:

Acute flaccid paralysis (AFP) in children is defined as acute onset of flaccid paralysis in one or more limbs or of bulbar paralysis in any child less than 15 years of age\(^1\). GBS is the commonest cause of AFP worldwide. Other causes include transverse myelitis, botulism, Tic bite paralysis and traumatic neuritis. In our study the commonest cause of AFP was also GBS. In a study from Australia common causes of AFP were GBS (47%) and transverse myelitis (19%) followed by acute disseminated encephalomyelitis, traumatic neuritis, Tic bite paralysis and infantile botulism.\(^1\) A study from Hong Kong showed GBS was 42% followed by transverse myelitis 15%, as the common causes of AFP in children\(^2\). Two different studies from Pakistan also describes GBS as the leading cause of AFP\(^{10,11}\).

Among the children with GBS 20, (67.7%) were under 5 years of age. It could be due to high incidence of infections in young children which is consistent with other studies from Hong Kong and Central America\(^{2,12}\). Male to female ratio in our study was 1:1 which also correlates with a study from Malaysia\(^13\) where this ratio was 1.3:1.

Involvement of respiratory muscles was present in 27 (90%) patients. It is much high compared to a study performed in Pakistan where this figure was 55.9%.\(^{14}\) Cranial nerve involvement was found in 3.3% children, which have been found to be 45% and 50% in other studies in pediatric patients\(^{18,19}\). Autonomic dysfunctions were noticed in 53.3% cases, which is comparable (51%) in a similar study\(^{15}\).

CSF albuminocytological dissociation was present in 76% of patients in our study while in another study it was found in 97.5% of patients\(^{16}\). CSF examination done in second week of illness and criteria for dissociation was protein >80 mg/dl and cells <10/mm\(^3\).

In this study, patients with GBS mortality was 23.3%. All these patients belong to mechanically ventilated group with or without other treatment modalities. Total 12 (40%) patient needed mechanical ventilation during treatment in ICU which was 15% to 20% in another study\(^{17,18}\).

It shows that the patients who have severe disease at onset and required mechanical ventilation had poor prognosis. The high mortality in this group also may be related to complications like infection, aspiration, autonomic dysfunction and cardiac arrest.

Data on the course of recovery were better than in the literature. Briscoe et al reported a mean time of recovery after reaching the maximum disability of the disease of 28 days\(^{19}\). where in this study it was 19.7 days (5-40 days). Mean duration of death cases were 5.1 days, which indicates either rapid progression of the disease or delayed transfer to ICU.

In this study 86.7% patient received IVIG and 13.3% patient was treated with steroid. Patients treated with IVIG improved significantly earlier than those with steroid which is comparable with the study of Gures et al\(^{23}\). In children, three cases with rapid improvement during administration of intravenous immunoglobulin within four to seven days have been observed which is comparable with other study\(^{20-24}\).

It was observed that the benefit of intravenous immunoglobulin is better in children. Although in our study corticosteroids were also shown to be of value, their effectiveness seems to be inferior to immunoglobulin.
Conclusion:

Bulbar involvement with respiratory failure is the most common indication of admission in ICU. IVIG is the effective treatment modality. Need of mechanical ventilation indicates poor outcome. Aspiration is one of the significant complications that lead to further respiratory deterioration. Overall improvement is satisfactory with well equipped supportive critical care.

References:


