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STUDY ON THE PRESENTATION AND CLINICAL COURSE OF PAEDIATRIC GUILLAIN-BARRÉ SYNDROME

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ABSTRACT

Guillain-Barré syndrome has now emerged as one of the most important differential diagnoses in children presenting with acute flaccid paralysis. We studied the clinical profile of paediatric patients diagnosed with this condition. This is a retrospective study of patients admitted between November 2011 and May 2018. 14 patients were included in our study with 8 females and 6 males. Mean age was 7.8 years and a time to presentation of 5.5 days. Cranial nerve involvement was seen in 4 cases. CRP levels were elevated in 3 cases. Most patients were treated with IV Ig and recovered rapidly.Three patients required invasive ventilation. AIDP was common in the 4-9 years age group with slightly more females than males. Most presented within 7 days. Cranial nerve involvement and increased CRP correlated with severe disease. Children respond well to IV Ig therapy and rarely have long-term sequelae.

INTRODUCTION

Guillain-Barré Syndrome [GBS] is an acquired, acute, immune mediated, symmetrically progressive ascending peripheral neuropathy due to demyelination and/or axonal degeneration.[1][2] It can involve peripheral nerves, spinal sensory or motor nerve roots and the cranial nerves.[1]Up to two-thirds of patients report an antecedent infection.[3]The most common pathogen that has been implicated is Campylobacter jejuni. Other infections that have been implicated include Mycoplasma pneumoniae, Haemophilus influenzae, Cytomegalovirus infections, Epstein-Barr virus, Varicella Zoster and Zika virus infections.[4][5]It is believed that an abnormal immune response to an antecedent infection, immunisation or immune triggering event leads to activation of humoral and cellular immunity.The resultant damage to nerves may be a result of molecular mimicry, superantigenicity or by cytokine stimulation. It was initially considered as a one disorder but electrophysiological testing has shown that there are several subtypes. These subtypes are: i) Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP) ii) Acute motor Axonal Neuropathy (AMAN) iii) Acute Motor Sensory Axonal Neuropathy (AMSAN) iv) Miller Fisher Syndrome v) Acute Panautonomic Neuropathy.[1] These subtypes are characterised by varying degrees of dysfunction, they differ in patient demographics, pathological findings, clinical course and response to treatment. Diagnosis is primarily based on clinical findings supplemented by the presence of albumino-cytological disassociation in CSF and electrophysiological studies demonstrating slowing of nerve conduction or the presence of serum antibodies.Sequential examination is usually required to establish the diagnosis. The causes of mortality in GBS include respiratory failure due to either diaphragmatic involvement or Bulbar weakness, thromboembolism or due to dysautonomia with resultant arrhythmias.[6]With the eradication of Poliomyelitis in India, GBS has emerged as one of the most important differentials in a child presenting with acute flaccid paralysis. GBS is less common in children and when it does occur it has a relatively benign course.[6]

This paper will examine the clinical findings in children and their course at our hospital.

MATERIALS AND METHODS

This is a retrospective, record-based study which compiled data of patients admitted in our centre between November 2011 and May 2018.

Patients who were less than 18 years who had been diagnosed to have Guillain-Barré Syndrome were included in our study.

Patients with history of exposure to lead or hexacarbons, those with purely sensory symptoms, recent history of diphtheria infection or evidence of Neurotoxic conditions were excluded from the study.

Patient details such as age, sex, presenting complaints, duration of complaints, clinical finding, cranial nerve involvement, levels of CRP or CPK, findings of CSF examination if performed, results of Nerve conduction studies, treatment modality and time to recovery were gathered from case records. Most patients in our centre are treated with IV Ig in doses of 2 g/kg for 5 days. They are also administered physiotherapy. The need for intubation is assessed primarily on the basis of clinical findings (patient condition, rate and depth of respiration, use of accessory muscles, upper airway reflexes and the single breath count test) supplemented by ABG analysis

These details were then compiled and analysed. All the data were entered and analysed using Microsoft Excel 2016

RESULTS

Fourteen patients were included in our study. The average age of the patients in our study was 7.8 years (Median= 8 years, Range: 7 months-14 years). Of the 14 cases in our study there were 8 girls and 6 boys(Ratio: 1.33:1). Twelve of the fourteen patients presented with weakness and two patients complained of tingling and numbness of the limbs. One patient complained of breathlessness, two complained of difficulty in swallowing and two patients reported a change in their voice. The average time to presentation at our centre from symptom onset was 5.5 days with a median of 4 days (Range: 1-14 days). On examination, all patients showed at least some decrease in power but upper limb power was preserved in two patients. All patients showed areflexia or severe hyporeflexia. Four patients in our study showed involvement of the cranial nerves. Two of these had involvement of the IX and X nerve, one had involvement of the VI and VII nerve while another had isolated involvement of the VII nerve. Three patients in our study had signs of ataxia on examination. Three of the eight patients in whom CPK levels had been performed had elevated CPK levels (>120 IU/L). Three of the eight patients in whom CRP levels had been estimated had elevated CRP (>3mg/L). On nerve conduction studies, ten patients showed demyelinating

polyradiculoneuropathy, two patients showed motor axonal neuropathy and two patients had Motor-sensory axonal neuropathy. Of the 6 patients in whom CSF analysis had been done, three showed albumino-cytological disassociation. Of the other three, two had presented within 3 days of the onset of symptoms. Twelve patients were treated with intravenous immunoglobulin, one patient was started on plasmapheresis but developed a reaction to FFP and was switched to IV Ig therapy and one patient received only supportive care. All patients in our study made a swift recovery within days of initiation of therapy, with a range of 3 to 30 days. Three of the 14 patients in our study required intubation and mechanical ventilation.

DISCUSSION

Guillain-Barré syndrome is less common in children as compared to adults. Half the patients in our study were in the age group of 4-9 years. Peaks have also been observed in late adolescence and early adulthood.[1]One patient in our study was 7 months old at the time of presentation raising the possibility that this episode developed after immunisation. Although the causal nature of immunisation has not been established, anecdotal reports do suggest that this is a possibility.[7][8]There is no sex predilection in the occurrence of GBS and our study reflects that.[9]Most patients with GBS have predominantly motor symptoms with the sensory symptoms being prominent in AMSAN variant of GBS. Of the two patients whose chief complaint was tingling and numbness, one was found to have the AMSAN variant of GBS. The average time to presentation varied widely and to some degree is useful in predicting the severity of the disease. It has been shown that patients presenting within 7 days of onset of symptoms usually have a worse prognosis.[2]The one patient who presented within one day of symptom onset required mechanical ventilation and had a prolonged hospital stay (66 days). GBS is characterised by nearly symmetrical ascending neuropathy and hyporeflexia to areflexia is a pre-requisite for diagnosis although the AMAN variant of GBS may have preserved reflexes.[1][2]All the patients in our study had severe hyporeflexia or areflexia. Cranial nerve involvement is quite common in GBS and is known to occur in about 45-75 % of cases.[10]In our study 4 of the 14 patients had cranial nerve involvement. Two of these patients had involvement of the facial nerve and the other two had bulbar palsy. Cranial nerve involvement has been correlated with severity of the illness.[11]Three of these patients were found to have severe disease and two of these patients required mechanical ventilation. Further, bulbar palsy increases the risk of aspiration and resultant complications. Ataxia is a common finding in children with GBS.[12]Rhabdomyolysis has been documented in some cases of GBS and can complicate the condition.[13][14]CPK levels were elevated in three patients in our study. CPK levels are useful to rule out other conditions which may affect the muscle such as polymyositis. Three patients in our study had elevated CRP. All three patients required mechanical ventilation. Studies have shown that CRP levels correlates with severity of GBS.[15]

Demyelinating polyradiculoneuropathy was the most common finding on NCS and was seen in 10 of the 14 cases (71.4%). AMAN variant was found in two cases. The AMAN variant is more common in children, is sometimes associated with fever and haemorrhagic conjunctivitis, can have asymmetric illness with CSF pleocytosis and preserved Deep tendon reflexes.[1][2]The AMAN variant is more common in India. AMSAN is more common in adults, but two patients in our study had the AMSAN variant of GBS. Both of these variants are usually preceded by Campylobacter infection and are more severe due to the associated axonal degeneration. Despite its severity, AMAN is usually associated with a rapid recovery.[1]One of the patients with AMAN in our study recovered within 3 days of admission and initiation of therapy. Further, it is important to note that severity of the illness does not correlate with long term disability. Albumino-cytological disassociation was found in three of the 6 patients in whom CSF analysis had been done. It is important to note that the CSF may be normal initially; usually for 48 hours after symptoms onset. Two of the three patients with normal CSF findings presented within 3 days of symptoms onset. However, 90% of patients have abnormalities by the second week.[2]IV Ig therapy is usually preferred over plasmapheresis due to the lower associated risk of treatment related complications. However, a Cochrane review established that both treatments were equally effective. In children, most studies on GBS have only examined the effectiveness of IV Ig therapy and only a few studies have studied the role of plasmapheresis. Combined therapy is not usually recommended.[16]In our study, 21.4% of patients required intubation and mechanical ventilation which is similar to data from other studies.[17]With treatment, most patients can expect to make a good recovery. In fact, 90-95% of children affected by GBS make a full recovery in 6-12 months.[18]

CONCLUSION

The average age of the study population was 7.8 years with a slight female predilection (1.33:1). The most common complaint was weakness and mild sensory symptoms such as tingling and numbness with a mean time to presentationof 5.5 days. Cranial nerve involvement and elevated CRP levels are predictors of more severe disease. AIDP is the most common variant although AMAN is quite common in children as compared to adults. Most children with GBS respond well to IV Ig therapy and are expected to make a rapid recovery with little to no long-term disability.

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