

## A rare case of recurrent Guillain Barre Syndrome

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### ABSTRACT

After the reduction of poliomyelitis, Guillain–Barre syndrome (GBS) has become the most common cause of acute flaccid paralysis in the developed and developing countries. Although it is a monophasic illness, GBS might reoccur in 1–6% of patients, after an asymptomatic period of several months to years. Risk factors for recurrent GBS (RGS) include age less than 30, milder symptoms, and history of Miller-Fisher syndrome variant in the first episode. A recurrence was defined as two or more episodes that fulfilled the National Institute of Neurological and Communicative Disorders and Stroke criteria for GBS, with a minimum time between episodes of 2 months (when fully recovered in between) or 4 months (when only partially recovered). There appears to be no significant difference between GBS and RGS episodes with respect to clinical symptoms and triggering events. We report a 12-year-old girl who presented with a second attack of GBS after an interval of 6 years with similar signs and symptoms. Nerve conduction study in both the episodes was of acute motor axonal neuropathy variety.

**Key words:** *Acute flaccid paralysis, Acute motor axonal neuropathy, Recurrent Guillain Barre Syndrome*

After the reduction of poliomyelitis, Guillain–Barre syndrome (GBS) has become the most common cause of acute flaccid paralysis in the developed and developing countries. Its incidence is 0.6–4 individuals per lakh population per year [1]. It is a post-infectious polyneuropathy involving mainly motor nerves, but sensory and autonomic nerves might also be involved. Most patients have demyelinating neuropathy, but few patients might have primary axonal degeneration [2]. Although it is considered a monophasic illness, GBS can recur in 1–6% of patients. It has been reported to occur in 1–10% of patients after an asymptomatic period of several months to years [3–5]. Risk factors for recurrent GBS (RGS) include age less than 30, milder symptoms, and history of Miller-Fisher syndrome (MFS) variant in the first episode [6].

A recurrence was defined as two or more episodes that fulfilled the National Institute of Neurological and Communicative Disorders and Stroke criteria for GBS, with a minimum time period of 2 months between episodes (when fully recovered in between) or 4 months (when only partially recovered) [7]. There appears to be no significant difference between GBS and RGS episodes with respect to clinical symptoms and triggering events. It is important to know that there is always a possibility of recurrence in GBS and parents should be counseled accordingly.

### CASE REPORT

A 12-year-old girl presented with a second attack of GBS after an asymptomatic period of 6 years. During the first attack,

she was admitted to our hospital with the chief complaint of weakness and difficulty in walking since morning. She had a history of upper respiratory tract infection (URTI) 15 days back which was cured. Examination revealed acute-onset quadriparesis with areflexia with a power of 2/5 in upper and 1/5 in both lower limbs. The rest of the systemic examination along with bowel and bladder movements was normal. Hemogram, liver function tests, renal function tests, and electrolytes were within normal limits. She was diagnosed as a case of GBS and treated with intravenous immunoglobulin (IVIg) (2 gm/kg) over 5 days. Nerve conduction study was suggestive of acute motor axonal neuropathy (AMAN). Cerebrospinal fluid (CSF) study showed albuminocytological dissociation (2 cells, all lymphocytes, sugar 75 mg/dl, and protein 80 mg/dl). She was discharged after 14 days of hospitalization with a power of 3/5 in all limbs. She was able to walk without support after 3 months. Since then, she has been normal without any residual disability.

After 6 years during the second episode, she presented with similar complaint of difficulty in walking and swallowing. She had a history of URTI 15 days back. Examination revealed quadriparesis (power 0/5 in all limbs). Her single breath count was 8 and there was pooling of secretions in the mouth. Subsequently, she was intubated and ventilated. She was diagnosed as a case of RGS and was treated with IVIg (2 gm/kg) over 5 days, along with supportive therapy. Nerve conduction velocity study showed severe AMAN. Her CSF study showed an albuminocytological

dissociation (1 cell, all lymphocytes, sugar – 77 mg/dl, and protein – 126 mg/dl).

In view of slow improvement and anticipated prolonged ventilation, tracheostomy was done after 10 days. She remained on the ventilator for 75 days and tracheostomy was closed after 90 days. At the time of discharge, power in both upper limbs was 4/5 and lower limbs was 3/5. Compared to the previous episode, this episode was more severe with respiratory involvement requiring mechanical ventilation, tracheostomy and prolonged intensive care unit stay, and slow response to IVIg.

## DISCUSSION

GBS is an acute, immune-mediated inflammatory polyradiculoneuropathy involving the peripheral nervous system. Onset is preceded by an antecedent event in two-thirds of the patients, usually a URTI or a diarrheal illness [1,2], where the causative agent is assumed to trigger an immune response against the gangliosides and glycolipids distributed along the myelin sheaths and peripheral nervous system. This results in marked inflammation of the peripheral nerves, resulting in demyelination and defective impulse propagation. It is a heterogeneous group of disorders which involves motor, sensory, and autonomic nervous systems to varying degrees depending on the subtype; (1) acute inflammatory demyelinating polyneuropathy, (2) AMAN, (3) acute motor-sensory axonal neuropathy, (4) MFS, (5) acute pan-autonomic neuropathy, and (6) pure sensory GBS.

RGBS is a rare entity that has been reported in about 1–6% of GBS patients [3]. This is the first case of RGBS from our unit in 20 GBS patients in the past 5 years. There are only a few published case studies that include children with RGBS [1,3-5]. The most comprehensive study reported by Dr. R. H. Kennedy and colleagues at Mayo Clinic in 1978, retrospectively followed 40 GBS patients for up to 42 years after the first attack, and only one individual had the second episode of GBS, which occurred 4 years after the initial one [6].

The time lag between two episodes of GBS was 4 months–10 years in a study done by Das *et al.* [3] and a mean of 7 years with a range from 2 months to 37 years as described by Kuitwaard *et al.* [2]. A case report by Md Mohabub from Bangladesh had a lag period of 17 years, whereas in our case, it was 6 years. In a study of 49 RGBS patients by Kuitwaard *et al.*, one-third of the patients presented with same, one-third with more, and one-third with less huge disability score [2]. Our patients presented with similar signs and symptoms, but with an increase in the severity and respiratory involvement requiring ventilation. Recovery from the second episode was very slow as compared to the first episode.

In the same study by Kuitwaard *et al.*, 65% of patients had similar and 35% had different antecedent infections in recurrent episodes, which indicated that there is some genetic basis for the disease [2]. Studies by Taly *et al.* and Hayashi *et al.* have reported different antecedent events in individual RGBS patients [7,8]. In contrast, Wijdicks and Ropper described

similar antecedent illnesses in individual RGBS patients [9]. In our patients, URTI was the preceding infection in both the episodes. Most of the patients had either pure motor or sensory-motor symptoms in subsequent episodes. None of the patients initially had GBS in one episode followed by MFS in a subsequent episode. In contrast to the case report by M. Dy *et al.*, where the patient developed different variant, our patients had similar motor axonal neuropathy with more severity [10]. RGBS is more common in patients with demyelinating variety, but rarely with axonal alone. In our case, it is axonal variant. Nerve conduction study are similar in demyelinating variety of RGBS but one case report of Japanese patient showed sensory variation.[11]

There is no specific predictor of RGBS but age less than 30, milder symptoms, and history of MFS variant in the first episode are the few risk factors [12]. Most patients with RGBS develop one episode, but patients with two or more episodes are described with recurrence occurring months to years after the first episode [2]. Pathogenesis of RGBS is not clear, but an immunological and genetic basis is suspected because different types of the trigger result in similar presentation, recurrence is more common with MFS variant (have anti-GQ 1 antibody). In the study by Kuitwaard *et al.*, three patients of RGBS had different autoimmune diseases, which suggest a genetic basis for the disease.

## CONCLUSION

To the best of our knowledge, this is the first case report of axonal phenotype RGBS in a child from eastern India. This case highlights that GBS is not a monophasic illness and recurrence of GBS, though rare, might be seen in children. RGBS is an underrecognized and underdiagnosed entity in pediatric patients. Future researches are needed to further study the epidemiology and pathophysiology of the disease.

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