

GUILLAIN BARRÉ SYNDROME

Introduction

Guillain Barré syndrome (GBS) is a post-infectious inflammatory disorder affecting the peripheral nerves.

Table 1. Clinical pearls on GBS in children

- weakness may begin in the face or upper limbs, or may be asymmetrical at onset. However, at full evolution is almost always bilateral, symmetrical, proximal predominant limb weakness.
- sensory symptoms, e.g. limb pain and hyperesthesia, are common
- bladder and bowel involvement may occasionally be seen, but is *never* present at onset and *never* persistent (if so, think of spinal cord disorder)
- CSF protein level and nerve conduction studies *may be normal* in the first week of illness.
- clinical variants of GBS include:
 - Miller Fisher syndrome, characterised by ophthalmoplegia, pupillary dilatation, ataxia and areflexia
 - Pharyngo-Cervico-Branchial (PCB) syndrome, characterised by facial, bulbar (swallowing) and neck weakness
 - Autonomic GBS, which presents with motor weakness and prominent autonomic symptoms, such as dilated cardiomyopathy, postural hypotension and diarrhoea

Management

The principle of management is to establish the diagnosis and anticipate / pre-empt major complications.

- a **clinical** diagnosis can be made by a history of progressive weakness (< 4 weeks) with areflexia, and an elevated cerebrospinal fluid protein level and normal cell count (“protein-cellular dissociation”)
- nerve conduction study is **confirmatory**

Anticipated complications and management

- **Respiratory compromise**
 - monitor PEFR regularly
 - give oxygen, keep NBM breathless
 - provide respiratory support early with either BiPAP or mechanical ventilation.
- **Progressive severe weakness**

Table 2. Hughes Functional scale for GBS

- 0 - normal
- 1 - minor symptoms, capable of running
- 2 - able to walk up to 10 meters without assistance but unable to run
- 3 - able to walk 10 meters with assistance of one person, or a walker
- 4 - unable to walk
- 5 - requires assisted ventilation

Specific measures

- Intravenous Immunoglobulins (IVIG) 2 gm/kg total over 2 - 5 days in the first 2 weeks of illness has been shown to :
 - reduce disease severity (by 1 functional level)
 - shorten time on a ventilator
 - shorten time taken to reach functional level 2
- IVIG is as efficacious as Plasma exchange in both children and adults, and is safer and technically simpler
- 10 % of children with GBS may suffer a relapse of symptoms in the first weeks after improvement from IVIG. In these children, there is anecdotal evidence of benefit from a second dose of IVIG.

General measures

- prophylaxis for deep vein thrombosis should be considered for patients ventilated for GBS, especially if recovery is slow
- liberal pain relief, with either paracetamol, NSAIDs, gabapentin or opiates.