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Gullian – Barre Syndrome Variant with Unilateral Facial Weakness

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Abstract

It is a postinfectious polyneuropathy due to alteration of protein component of myelin (p2 neurotogenic peptide) leading to demyelination because of autoimmune mechanism. Neurological manifestation begins after 2 to 4 weeks of viral or bacterial infection. Clinical expression includes an acute onset symmetrical ascending weakness (both proximal & distal) with unilateral facial weakness and respiratory weakness and autonomic dysfunction. The diagnosis depends on clinical picture, electrophysiological findings and CSF examination. Immunotherapy is the main stay of treatment. IVIG & Plasmaphrersis done within 2 to 4 weeks of symptoms onset is recommended. Treatment is warranted in non ambulatory patient (Modified Hughes GBS disability scale). The patient who hav not responded to initial IVIG treatment may benefit from second course of IVIG. General supportive care includes cardiorespiratory care, physiotherapy, nutritional management, management of neuropathic pain, bladder–bowel care and prevention of deep vein thrombosis.

Keywords: Acute inflammatory demelinating polyradiculoneuropathy (AIDP); Acute motor axonal neuropathy (AMAN); Acute flaccid paralysis; Clinical neurophysiology; Immunotherapy

Abbreviations: AIDP: Acute Inflammatory Demelinating Polyradiculoneuropathy; AMAN: Acute Motor Axonal Neuropathy; NCS: Nerve Conduction Study; IVIg: Intravenous Immunoglobulin

Introduction

Gullian – Barre syndrome is also known as acute inflammatory demyelating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN). AIDP is the predominant subtype in North America & Europe while AMAN is commonly reported subtype in Asia including India & Central & South America. About 65% of children report preceding upper respiratory tract & gastrointestinal tract infection. Immunopathogenesis involve molecular mimicry & formation of cross reacting antiganglioside antibodies. The GBS has several variants depending upon distribution of motor, sensory, cranial, autonomic or cebellar involvement variant, among them AIDP is the most common [1]

Discussion

Clinical expression

- Muscle pain, difficulty while walking or refusals to walk are often the first presenting symptoms (50%)
- Distal limb weakness which ascending & symmetrical (20%) Areflexia.
- Respiratory muscle weakness & facial weakness (20%)
- Sensory symptoms including painful parathesia, backache & meningismus (50% -80%)
- Transient bladder involvement can occur in few children.
- Approximately 25% develop respiratory insufficiency requiring artificial ventilation & 75% have autonomic dysfunction. The course is monophasic in most of the children. 80 % reach maximum severity within 2 weeks & 97 % in 4 weeks.This phase is followed by a relatively static 'plateau phase' ranging from 2 days to 6 months before recovery begins [1].

Diagnosis

The diagnosis of GBS is clinical & is supported by a few investigations. Characteristic CSF finding are of Albumino-Cytological dissociation i.e. a combination of elevated CSF protein & normal cell count. Nerve conduction studies help in diagnosis of different sub types of GBS. In early phase of GBS motor & sesory nerve conduction study (NCS) is normal. In such situation diagnosis supported by prolonged F waves latencies. NCS abnormalities

tend to peak by 2 weeks of illness. Children with GBS should be managed in PICU during initial phase (Table 1) [2,3].

Table 1: Common differential diagnosis of Gullian – Barre syndrome [2].

Muscle Disorders	Neuromuscular Junction Disorders	Neuropathies	Central Nervous System Disorders
Inflammatory myopathy	Mysthenia gravis	Diphtheritic neuropathy	Acute myelopathy
Periodic paralysis	Botulism	Porphyria	Poliomyelitis
hypokalemia	Eaton – lambert syndrome	Traumatic neuritis	Brainsem stroke
infection		Vaculitis neuropathy	Brainstem encephalitis

Clinical neurophysiology

Earliest abnormalities is a drop in the amplitude of the evoked muscle action potential & conduction blocks. Marked slowing of nerve conduction can be recorded in about 50% patients. Reduced compound motor unit potential amplitude is the most frequent finding. Absence or prolongation of F wave is common. Proximal blocks can be detected by measuring F wave. Conduction studies improve slowly over a period of several months. Spontaneous fibrillation may detected on electromyography during recovery phase after 2 or 3 weeks [1].

Pathogenesis

AIDP is multifocal noninfective inflammatory process causing demyelinaton or axonal degeneration of peripheral nerves. ADIP is generally due to T cell mediated immune myelin damage. Increased incidence of several axonal degeneration in GBS following *C. jejuni* infection is known with more severe involvement. The mechanism of axonal damage is different molecular mimicry due to shared epitopes with gangliosides [1] (Figure 1).

Figure: 1: Recovery of upper limb and respiratory weakness after immuno therapy in three week admitted in my Civil Hospital Ahmadabad. India (Oct 2016).

Prognosis

Recovery is common & often complete in the majority.Mortality is now ≤ 5%. Most of the death in childhood are due to preventable respiratory complication. Acute axonal neuropathy, with good pgognosis.Acute axonal and sensory axonal neuropathy generally causes poor prognosis. Miller Fisher variant in which cerebellar signs, cranial nerves are involved also has a poor prognosis. The disabilities included foot drop, pes cavus and postural tremor & persisting weakness of the hands [1].

Treatment

Supportive symptomatic treatment is the mainstay of therapy in the majority. The child should be closely observed in hospital, objective assessment of respiratory function, regular measurement of vital capacity is performed. Ventilatory support should be considered if there is evidence of respiratory insuffiency [3,4].

- Dysphagia if present necessitates nasogastric feeding.
- Chest and limb physiotherapy should be initiated early and carried out carefully. Bladder and bowel function should be attended to.
- Plasmapheresis has been shown to be effective in decreasing severity and shortening the non ambulatory phase. No significant complication ensue fron well conducted plasmapheresis. Several paediatric studies support the results.
- Intravenous immunoglobulin (IVIg) is atleast as effective as Plasma exchange. IVIg is now become the preferred treatment due to ease of administration. Dose of IVIg 0.4 g/kg body weight daily given on five successive days or two successive doses of 1 g/kg may be given active treatment of impending/ respiratory failure is imperative. Indication of immunotherapy includes a hughes GBS disabilities scale ≥3 or when patient is unable to walk unaided for 10 meters [5-7].

Conclusion

Acute flaccid paralysis in children is a medical emergency. AFP is a clinical syndrome with array of differential diagnosis.The common causes of AFP are Gullian – Barre syndrome, Anterior horn cell myelitis and Acute transverse myelitis. Rapid evolution of the weakness can lead to respiratory failure. Hence a child with AEP Should be managed in PICU in the initial few days. Immunotherapy is a main stay treatment.

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