GUILLAIN-BARRE SYNDROME Et. GBS usually follows nonspecific gastrointestinal or respiratory infection by ≈ 10 days. This include nonspecific viral infections, bacterial

infection e.g. Campylobacter jejuni, H. pylori, & M. pneumonia; or vaccines

e.g. rabies, influenza, & meningococcus. Path. GBS is an autoimmune disorder often considered as

postinfectious polyneuropathy due to demyelination & sometimes axonal degeneration involving mainly motor nerves, but sometimes also sensory and autonomic nerves. C.M. GBS affects people of all ages. The classic disease pattern called Landry ascending paralysis; initial symptoms include numbness and

paresthesia followed by symmetrical weakness in the lower limbs &

progressively involves trunk, upper limbs and finally the bulbar muscles (which occur in ≈ half of cases). Asymmetrical weakness occurs in ≈ 9% of cases. Onset usually gradual over days or weeks or may be abrupt → pain & tenderness of muscles initially then flaccid tetraplegia & respiratory muscles paralysis within 24hr. Dysphagia and facial weakness are often impending signs of respiratory failure and also they interfere with eating and ↑ the risk of aspiration. Some patients may exhibit symptoms of viral meningo-

encephalitis & others may have papilledema. Sensory nerves involvement → paresthesia; whereas autonomic

nervous system involvement → variation of BP or HR e.g. postural hypotension, episodes of bradycardia or asystole, as well as urinary incontinence or retention. ॐ Types (variants) of GBS:- 1. Classic type " Landry ascending paralysis" is the most common. 2. Miller-Fisher syndrome is characterized by ataxia, areflexia, &

ophthalmoplegia (mostly due to 6th cranial nerve involvement). 3. Chronic Relapsing (unremitting) Polyradiculoneuropathy is recur intermittently or do not improve for a period of months or years. 4. Congenital GBS is rare, improve gradually & do not require Rx.

D.Dx. Spinal cord lesions, Peripheral Neuropathies (toxic, infections, inborn errors of metabolism), or Neuromuscular junction disorders. Inv. CSF protein ↑ > twice of the upper limit of normal, other profiles are normal except WBC are <10/mm3. This dissociation between high CSF protein and lack of cellular response is diagnostic of GBS. NCV of motor nerves is greatly ↓, whereas in sensory nerves is slow. EMG show features of acute denervation of muscle. MRI (with enhancement) shows thickening of cauda equine & intrathecal nerve roots in >90% of cases; it also may be used to exclude other D.Dx. CK level is normal or mildly ↑.

Serologic tests; Antiganglioside antibodies are mainly ↑ with axonal neuropathy. Biopsy of nerve or muscle is usually not required. Rx.

Patients with slowly progressive disease may simply be observed in hospital for spontaneous remission with supportive therapy e.g. feeding, hydration, Px of bed sores & DVT, Rx of secondary infections, &

Rx of neuropathic pain by gabapentin or carbamazepine. Patients initially presented with rapidly progressive disease should be

admitted to the ICU for observation & stabilization; in addition to the supportive care, patients should be given the following:- IVIG in dose 0.4 g/kg/day for 5 consecutive days or 1 g/kg/day for 2 consecutive days. Alternative is Plasmapheresis +/_ Immunosuppressives.

Note: Corticosteroids are ineffective in GBS. Respiratory monitoring is mandatory by PEFR or PEEP with intervention by ETT & artificial ventilation if there is respiratory failure. Cardiovascular monitoring by continuous measurement of HR & BP with insertion of a temporary pacemaker if required.

Patients with Chronic Relapsing Polyradiculoneuropathy may be

treated with IVIG (which can be given subcutaneously), plasma

exchange (upto 10 times daily), immunosuppressives, or high-dose pulsed IV methylprednisolone. Pg. GBS is usually a benign disease, and spontaneous recovery begins

within 2-3 wk. Most patients regain full muscular strength, although some are left with residual weakness (especially with axonal

neuropathy).

Improvement usually follows a gradient inverse to the direction of involvement. Tendon reflexes are the 1st lost (although 10% may have normal reflexes) & usually the last one to recover. Long-term outcome after complete recovery may be easy fatigability; only ≈ 7% of patients suffer from relapse.

Factors associated with poor prognosis include: maximum disability at the time of presentation, cranial nerve involvement, & endotracheal

intubation. Death may occur in GBS due to either respiratory failure or

cardiovascular Cxs.