Guillain—Barre Syndrome in 2016: The Centenary Advances

Bhawna Sharma and Madhuparna Paul

Guillain—Barre syndrome (GBS) is the most common and severe acute paralytic polyradiculoneuropathy. Since its initial description by Guillain, Barre and Strohl in the year 1916 there has been a huge expansion in the knowledge of this potentially treatable disorder. 2016 marks the centenary year of GBS. It was conventionally described as an acute onset ascending pure motor demyelinating illness with areflexia. It has an annual incidence of 1/100000 across several studies. It can occur at any age with a slight male preponderance and with seasonal variations. However with ever growing knowledge in last 100 years the clinical spectrum under this umbrella has also expanded and several subtypes based on histopathology and neurophysiology have emerged.

The various forms of GBS are Acute Inflammatory Demyelinating Polyneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN), Acute Motor and Sensory Axonal Neuropathy (AMSAN), Miller Fisher Syndrome(MFS). AIDP is the more common in the western world while AMAN is more common in Asian subcontinent (in Japan and China). Other variants like pandysautonomia, pure ataxic GBS, pharyngeal- cervical-brachial GBS, bibrachial onset GBS and isolated bulbar palsy have also been described. Few cases may have retained reflexes, positive Babinski sign, papilloedema and transient bladder involvement.

Two third of cases are preceeded by antecedent infections either bacterial or viral, Campylobacter jejuni (C. jejuni), Epstein Bar virus (EBV) or Cyto-megalovirus (CMV). Hepatitis E, Mycoplasma and recently Zika virus are responsible organisms. Immunizations (swine flu or rabies vaccine), insect bites, pregnancy, surgery, cancer, autoimmune diseases, spinal anaesthesia may also act as trigger. It is an immune mediated process and the basic pathogenesis involves autoimmune and complement activation.

HLA subtypes probably also play role. Molecular mimicry with cross reactivity between peripheral nerve ganglioside and antibody against lipooligosaccharides of infectious agents act as a trigger to initiate aberrant immune reactions causing destruction of peripheral nerves. About half of the cases has autoantibodies against peripheral nerve gangliosides. The syndrome evolution is quite characteristic with onset within 7–21 days of an acute respiratory tract infection or gastroenteritis with ascending areflexic paralysis with or without subjective sensory symptoms. The disease reaches its peak within 2–4 weeks followed by a plateau phase and then recovery. The syndrome often mimics other conditions like Hypokalaemic periodic paralysis, Acute Demyelinating Encephalomyelitis, Toxic neuropathy, Acute transverse myelitis in shock stage, Cauda equine syndrome, HIV radiculitis and Critical illness neuropathy. Acute onset CIDP and Treatment Related Fluctuations (TRF) also need differentiation and appropriate management. Diagnostic criteria laid by Asbury and colleagues is still of utmost important. CSF cell count is increased with cell count often less than 50/ccmm. Seen after first week of infection and Nerve Conduction Study shows demyelinating or axonal abnormality based on the subtypes. Both of these are supportive tools for correct diagnosis. Recently Brighton criteria have been used for diagnostic purpose.

There is various grading scale for prognostication of GBS based on which therapeutic strategies are planned. Some commonly used are Medical Research Council (MRC) score, the Erasmus GBS outcome score (EGOS), Erasmus GBS respiratory insufficiency score (EGRIS) and Hughes et al grading system, of which MRC grading and Hughes grading are most commonly used. Patients should be carefully monitored for respiratory involvement which occurs suddenly. About 25–30% of Patients are subjected to mechanical ventilation. Dysautonomia, in milder form seen in three quarter of patients, is another dreadful complication. Hughes functional grading scale for GBS is used to assess the functional disability of the patients. Grade 0 normal functional state, Grade 1 able to run with minor signs and symptoms, Grade 2 able to walk 5 m independently, Grade 3 able to walk 5 m with aid, Grade 4 bed or chair bound and Grade 5 require assisted ventilation. Dysautonomia occurs suddenly. About 25–30% of Patients are subjected to mechanical ventilation. Dysautonomia, in milder form seen in three quarter of patients, is another dreadful complication. Hughes functional grading scale for GBS is used to assess the functional disability of the patients. Grade 0 normal functional state, Grade 1 able to run with minor signs and symptoms, Grade 2 able to walk 5 m independently, Grade 3 able to walk 5 m with aid, Grade 4 bed or chair bound and Grade 5 require assisted ventilation. Poor prognosis is also seen with older age, rapid progression of disease, severe disease indicated by MRC score, preceding diarrhoea, positive serology for C jejuni and CMV, early cranial nerve involvement. Being an autoimmune disease immunotherapy is the mainstay of treatment. Intravenous Immunoglobulin (Ivlg), Plasma Exchange (PE) and Intravenous pulse Methyprednisolone (Iv MPS) have been tried. Ivlg (0.4 gm/kg over 5 days) is the gold standard treatment in view of its ease of administration and efficacy. The only drawback is its cost. PE is
cumbersome procedure in many setups and has various adverse effects. Though IvMPS has been refuted by many researchers, but in author’s experience in developing countries like India, it may be a good therapeutic option. Newer drugs like Eculizumab, a humanized monoclonal antibody that binds C5 complement component is still underway. About 10% patients who received IvIg may show deterioration in first 8 weeks (Treatment Related Fluctuations) and they often require repeat IvIg. Thirty five percent patient recovers completely, 35% has minimal residual motor deficits, 30% has moderate to severe residual paresis. Mortality ranges from 5–10% in most studies, the cause being respiratory involvement.

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CONFLICT OF INTEREST
There is no conflict of interest.

REFERENCES