Guillain Barre Syndrome [created by Paul Young 29/11/07]

general

- aka acute inflammatory demyelinating polyradiculoneuropathy
- typically a motor greater than sensory peripheral neuropathy with subacute onset, monophasic course and nadir within 4 weeks

aetiology

- precise aetiology is unknown but GBS is immune mediated and related to antibodies directed against peripheral nerve components
- numerous antecedents have been implicated but the more frequent are:
(i) upper respiratory tract infections
(ii) Campylobacter jejuni enteritis
(iii) CMV infection
(iv) EBV infection
(v) hepatitis A, B and C
(vi) HIV infection
- the association with antecedent infections suggests that certain agents may elicit immune responses involving antibodies that cross react with peripheral nerve gangliosides
- most patients suffer a demyelinating neuropathy but in about 5% of cases the condition is a primary axonopathy
- GM1 antibodies are present in axonal forms of GBS and GQ1b antibodies are present in the Miller Fisher variant of GBS

clinical manifestations

- initial findings are usually subacute and progressive weakness that is most marked in the legs associated with sensory complaints but without objective signs of sensory dysfunction
- deep tendon reflexes are often significantly reduced or absent at presentation

autonomic dysfunction:
- most typically presents as a hypersympathetic state and is often heralded by unexplained sinue tachycardia; patients may rarely experience bradycardic episodes that require pacing
- blood pressure may fluctuate wildly
- autonomic surges during tracheal suction or due to distended viscus may be dramatic

eye complications:
- corneal ulcers may complicate poor lid closure

diagnostic criteria

diagnostic criteria for typical Guillain Barre Syndrome
1. features required for diagnosis
(i) progressive weakness in both arms and both legs
(ii) areflexia
2. features strongly supportive of the diagnosis
(i) progression over days to 4 weeks
(ii) relative symmetry of symptoms
(iii) mild sensory symptoms or signs
(iv) cranial nerve involvement (particularly bilateral weakness of facial muscles)
(v) recovery beginning 2-4 weeks after progression ceases
(vi) autonomic dysfunction
(vii) absence of fever at onset
(viii) high CSF protein with fever than 10x6cells/L
(ix) typical electrodiagnostic features
3. features excluding diagnosis
(i) botulism, polio, myaesthenia or toxic neuropathy
(ii) abnormal porphyrin metabolism
(iii) recent diptheria
(iv) history or evidence of lead intoxication
(v) pure sensory syndrome without weakness

investigation

CSF:
- typically reveals elevated protein content without pleocytosis
- the nucleated cell count is <10 cells/mm3
- a CSF lymphocytosis of 10-20 cells/mm3 may suggest the possibility of HIV

nerve conduction studies:
- may be normal initially but often reflect segmental nerve demyelination with:
(i) multifocal conduction blocks
(ii) temporally dispersed compound muscle action potentials
(iii) slowed conduction velocity
(iv) prolonged or absent F waves

imaging:
(i) CT or MRI of c-spine should be considered to exclude a high cervical lesion

treatment

general:
- components of treatment for patients with GBS are as follows:
(i) management of ventilatory failure
(ii) management of autonomic dysfunction
(iii) management of painful peripheral neuropathy
(iv) meticulous nursing care
(v) psychologic support
(vi) physical and occupational therapy
(vii) prevention of deep vein thrombosis
(viii) nutritional support
(ix) early planning for rehabilitation
(x) immunotherapy for underlying immune process

respiratory failure:
- patients with GBS and evolving respiratory failure should generally be intubated when the vital capacity falls to about 15ml/kg or when difficulty with secretions begins because the recovery is generally slow [suxamethonium should be avoided]
- many patients become too weak to trigger the ventilator requiring mandatory modes
- weaning patients with GBS must wait for adequate strength to return [an improvement in vital capacity to >15ml/kg and in negative inspiratory force to greater than 25cmH2O usually suggests that a patient has improved enough to begin weaning; a formula using a combination of ventilatory and gas exchange variables has also been designed for this purpose]
- the majority of patients require support for less than 4 weeks but as many as 20% require more than 2 months

autonomic dysfunction:
- treatment of autonomic dysfunction is difficult with marked lability in blood pressure responses

painful peripheral neuropathy:
- may be extremely difficult to treat

nursing care:
- similar to care of other paralysed and ventilated patients but particular care needs to be taken to remember that GBS patients are lucid
- in addition to carefully explaining procedures, arranging distractions during the daytime and adequate sleep at night is very important
- in concert with physiotherapists, passive exercise should be performed frequently throughout the day
- eye patching may be helpful
- meticulous pressure care is required

DVT prophylaxis:
- a significant danger for patients with GBS requiring DVT prophylaxis and a low threshold for investigation of potential DVT or PE

nutritional support:
- should begin as soon as the patient is admitted with appropriate concern for the risk of aspiration
- most patients can be nasogastrically fed without difficulty; however, autonomic neuropathy sometimes complicates feeding

Immunotherapy
- overall, summary is that both plasma exchange and IVIG are effective and equivalent and no additional benefit is conferred by combined treatment
- patients with heart disease, renal insufficiency, hyperviscosity or IgA deficiency may be more susceptable to the complications of IVIG whereas labile BP, septicaemia and venous access problems may preclude plasma exchange

(i) plasma exchange:
- the efficacy of plasma exchange has been reviewed in a Cochrane review of six class II trials comparing plasma exchange alone with supportive care. Most of the trials employed up to five plasma exchanges of 50ml/kg over 2 weeks. This metanalysis demonstrated a more rapid recovery in ventilated patients treated with plasma exchange within 4 weeks of onset of symptoms
- the optimal number of exchanges has been assessed in patients with mild, moderate and severe GBS by the French cooperative group. On the basis of this trial, two exchanges are better than none in mild GBS, four are better than two in moderate GBS and six are no better than four in severe GBS
- albumin is the preferred replacement fluid

(ii) IVIG
- the efficacy of IVIG has also been examined by Cochrane review with three randomised controlled trials demonstrating its equivalency with plasma exchange

steroids:
- corticosteroids are not effective in GBS and are therefore not recommended

drugs associated with autonomic instability in GBS

Exaggerated hypotensive response
Phentolamine
Nitroglycerin
Hexamethonium
Edrophonium
Thiopentone
Morphine
Furosemide
Exaggerated hypertensive response
Phenylephrine
Ephedrine
Dopamine
Isoprenaline
Arrhythmias
Suxamethonium
Cardiac arrest
General anaesthesia