



GBS

Guillain – Barre Syndrome

A brief overview

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Historical Background

- ✧ In 1859, a French neurologist, Jean-Baptiste Landry, described ten patients who, over days to two or so weeks, developed ascending weakness and paralysis of, in sequence, the legs, arms, neck and breathing muscles of the chest (**Landry, 1850**).
- ✧ In 1916 three physicians, Georges Guillain, Jean Alexander Barre, and Andre Strohl reported elevated protein levels in spinal fluid with normal cell counts.
- ✧ Neurologists call this cytoalbuminologic dissociation.

What is GBS?



- ❧ GBS is a rare disease where the body's immune system attacks the peripheral nerves. This affects 1 in 100,000.
- ❧ GBS is a rapid developing disease that starts with weakness. The weakness is accompanied by abnormal sensations in the hands and feet.
- ❧ This sensation progresses to paralysis within hours to days. Some of the paralysis extends to the arms, face, and may affect the breathing. Some patients require ventilators.

What does it look like?



- ❧ Studies show that this disease can affect any of the peripheral nerves, motor – movement, sensory – temperature, pain, joint position and autonomic nerves- heart rate, blood pressure, urge to void.
- ❧ GBS patients recovery starts spontaneously.
- ❧ Most patients fully recover as long as they received the support when first diagnosed (ventilator if necessary)
- ❧ GBS occurs usually once. Additional episodes are rare.
- ❧ The main problem in GBS patients is the damage to the myelin covering the peripheral nerves.

Nerve Damage



- ❧ The body's own immune system attacks the patient's tissue. This is why GBS is considered an autoimmune disorder.
- ❧ After the peak of damage to the nerves the nerves are remyelinated or repaired. The patient usually regains strength and sensation.
- ❧ In some cases recovery can be slow and incomplete resulting in long term weakness particularly if the nerve axons are damaged.

Nerve Damage



- ❧ When sensory nerves are damaged the patient may experience abnormal sensations, pain, and poor balance.
- ❧ Various disorders differ by their onset, duration, symmetry of clinical findings, and whether the damage is to the peripheral nerve fibers of the motor, sensory, or autonomic function.
- ❧ Accurate diagnosis of these disorders is important because treatment and outcomes vary.

Other names for GBS



- ☞ GBS is also known as acute inflammatory demyelinating polyneuropathy. Acute – sudden onset. Inflammatory – inflammation in the nerves. Demyelinating – damage to the outer coating of the nerves. Polyneuropathy – involving many nerves.
- ☞ Acute idiopathic polyneuritis – inflammation of many nerves do to unknown cause.

CIDP



- ❧ Chronic Inflammatory Demyelinating Polyneuropathy.
- ❧ It is a rare disorder affecting 1 in a million.
- ❧ It can affect children and adults of any age.
- ❧ It commonly affects people in their 60's and 70's
- ❧ The disorder is more common in men than in women.

What is CIDP



- ❧ Slowly progressing autoimmune disease affecting the peripheral nerves.
- ❧ Sensory loss in a patient's legs and arms that develops in a steady fashion over more than eight weeks.
- ❧ The longer progression distinguishes it from Guillain-Barré Syndrome.
- ❧ CIDP is not selflimiting and, if untreated, about 30% of patients will progress to wheelchair dependence.

What is CIDP



- ❧ Some patients are able to continue to work but with progressing disabilities.
- ❧ Diagnosing and treating CIDP early can avoid a significant amount of this disability. Patients with CIDP may have different treatments.

Different Forms



- ❧ A progressive form can extend over several years.
- ❧ In the recurrent form patients have multiple episodes of disease that may be separated by months or even years.
- ❧ In the third type, a single episode or monophasic disease process extends over one to three years without recurrence.

How do you get it?



- ✧ Certain patients may be genetically more prone to CIDP than others.
- ✧ There is some evidence of a genetic basis for susceptibility to the more chronically progressive form of CIDP but there is no evidence of transmission between parent and child since only very rarely do cases of CIDP affect more than one family member.

What happens?



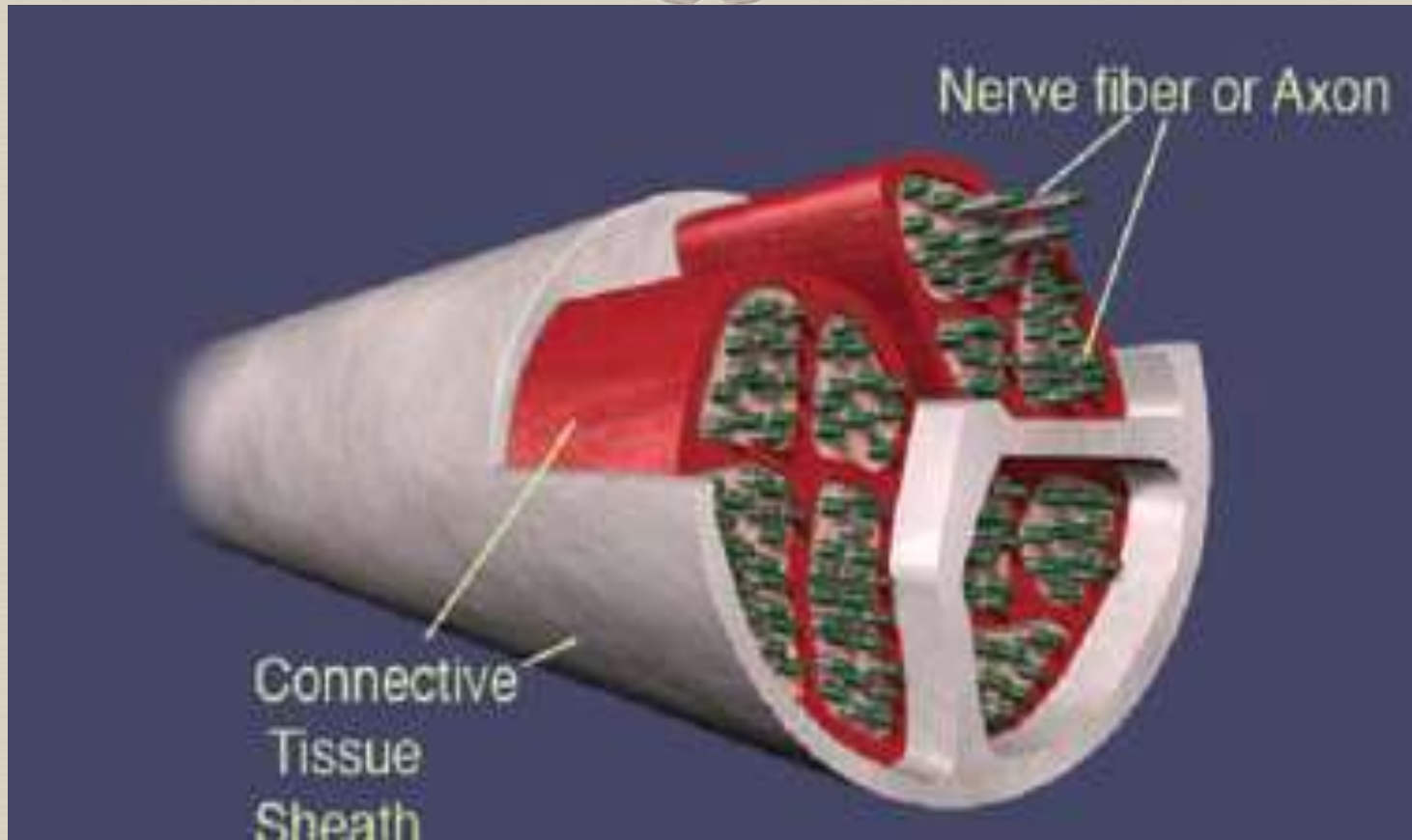
- ❧ The myelin sheath surrounding the motor and sensory nerves is destroyed. Myelin is a fat-rich covering that surrounds the nerve fiber or axon. The Schwann cell produces the myelin that wraps around a segment of the nerve fiber; and helps electrical current flow along the axon or fibermyelin.
- ❧ It is similar to insulating material around an electrical wire. Destruction of the myelin segments results in the loss of ability of the nerve to conduct an electrical impulse and leads to muscle weakness and altered ability to feel different sensations.

Peripheral Nerves

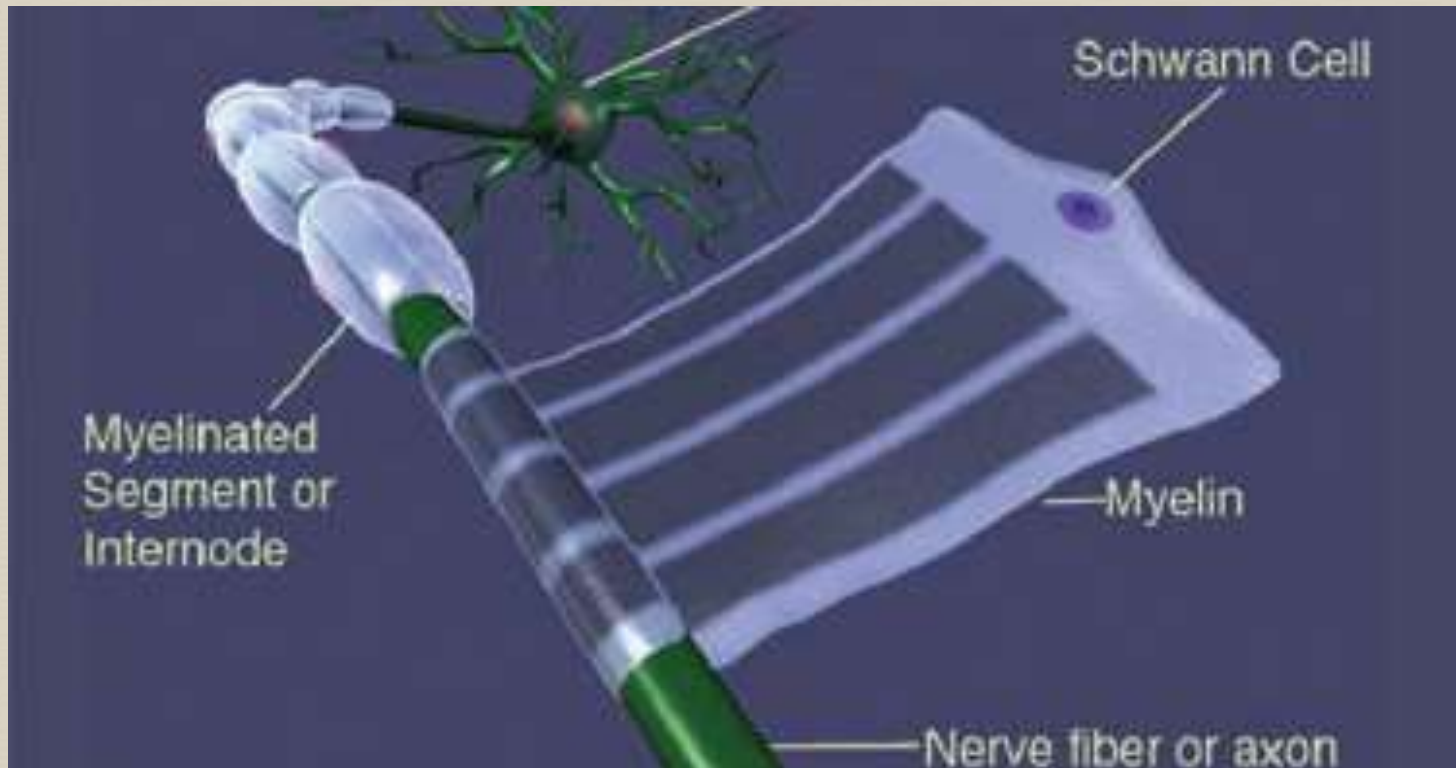


- ❧ They carry electric like impulses to muscles causing them to shorten or contract.
- ❧ Relay sensations from skin and other organs such as pain, hot and cold back to the brain.
- ❧ Damage to these nerves produces Weakness, muscle wasting, poor balance and numbness.

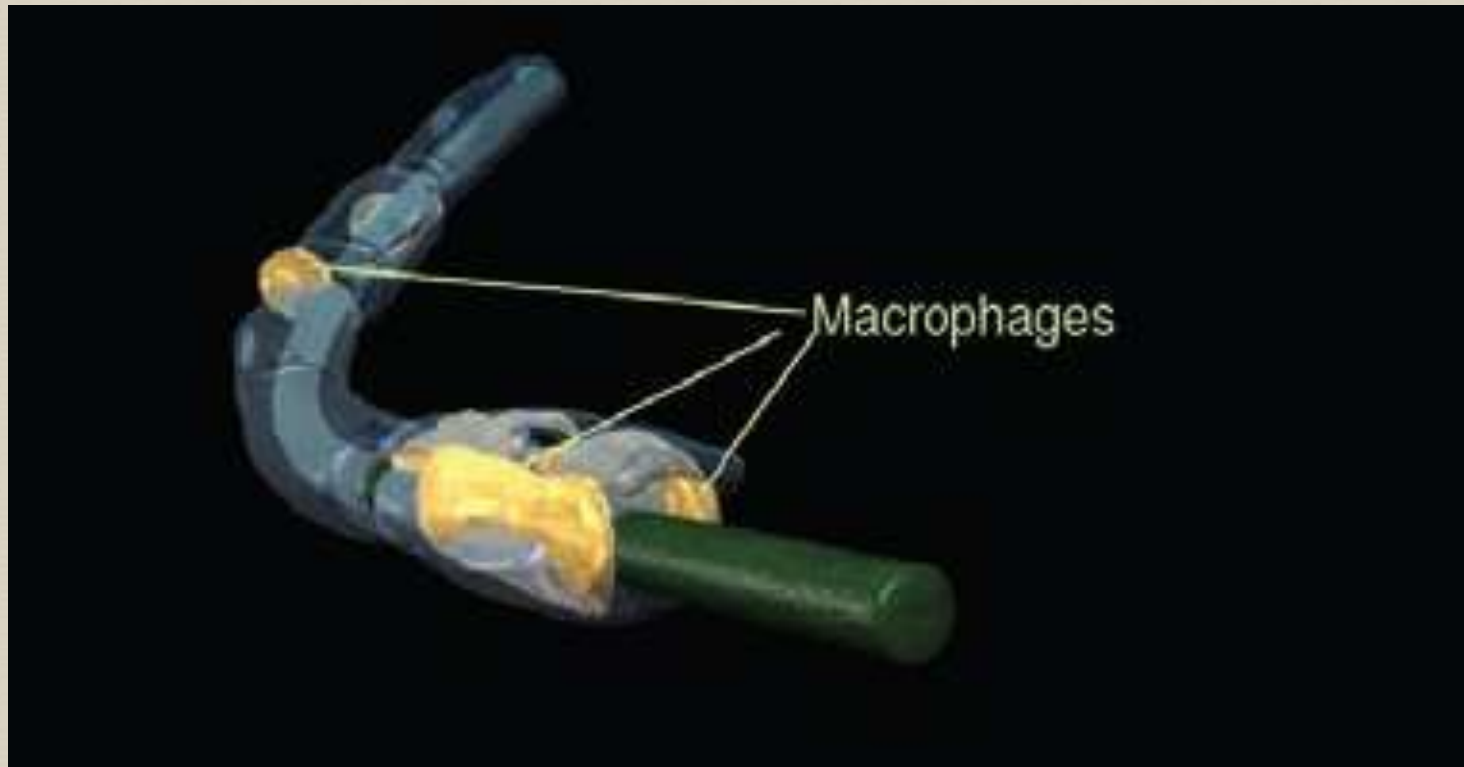
Peripheral Nerve



Myelinated nerve fiber



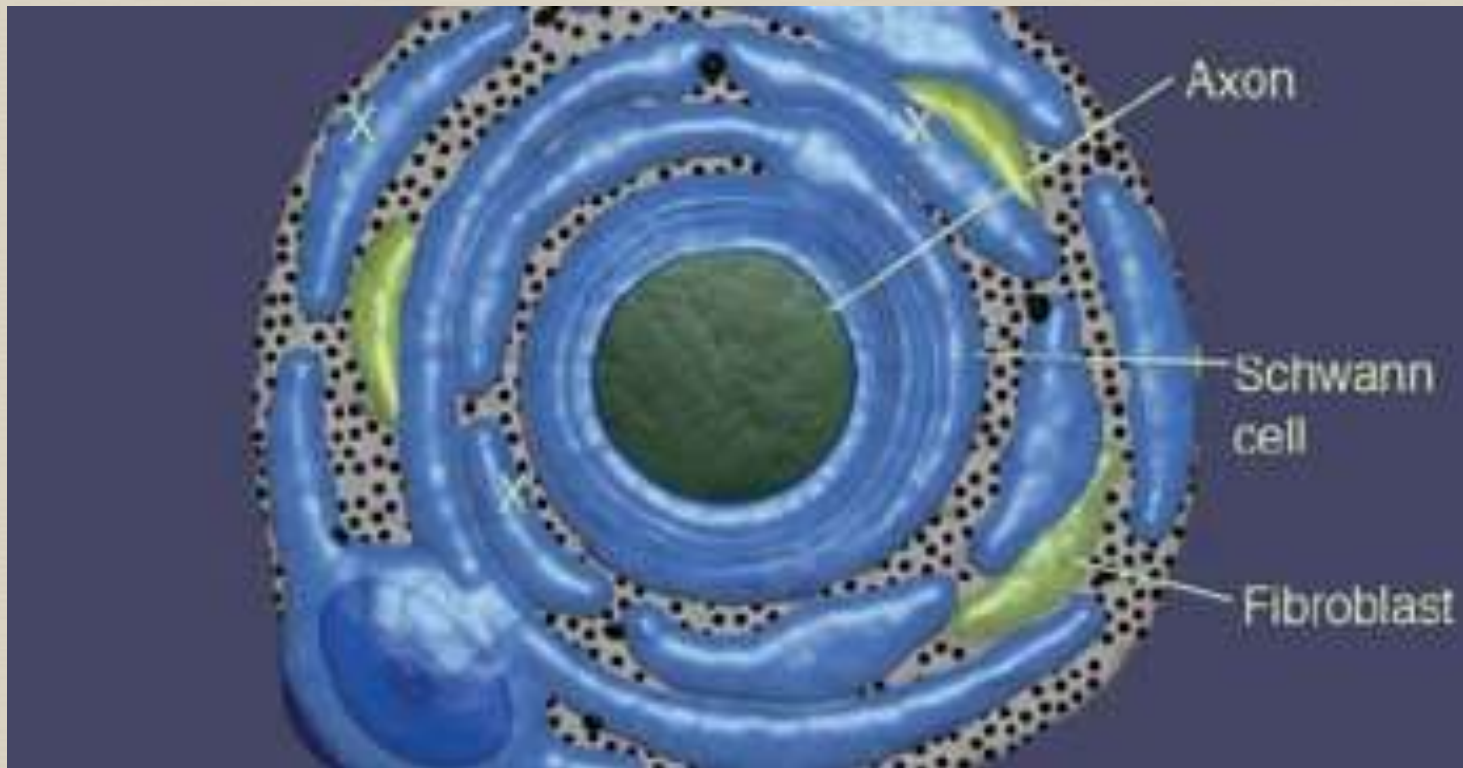
Nerve Fiber demyelination



What happens.

- ❧ With time and severity of disease the nerve fiber or axon itself can be damaged, leading to poor recovery even with adequate treatment.
- ❧ During this process the peripheral nerve continually tries to repair itself by regrowing the damaged nerve fibers and repairing the myelin sheath.
- ❧ Over time, with repeated episodes of myelin damage and attempted repair, layers of the myelin-producing Schwann cells and fibroblasts that form fibrous connective tissue produce a swelling called an onion bulb. Onion bulb formation limits the ability of the nerve to rapidly carry an electrical impulse.

Onion Bulb



How CIDP presents itself



- ❧ Most patients complain of difficulty climbing stairs or lifting their arms up to perform daily living skills. These symptoms indicate muscle weakness.
- ❧ Numbness, pins and needles sensation, unsteadiness or poor balance are signs of sensory nerve involvement.

What Causes CIDP?



- ❧ The factors that cause patients to develop CIDP are still unknown.
- ❧ The nerve is damaged in CIDP is not fully understood.
- ❧ The bodies own Immune system attacks itself.
- ❧ The immune system, by mistake, reacts against parts of our bodies causing disease.

How is CIDP Diagnosed ?

- ❧ No definitive diagnostic test for this disorder.
- ❧ The diagnosis is based on a patient's clinical history and various diagnostic test performed by a neurologist.
- ❧ EMG –Electromyography with nerve conduction velocity (NCV).
- ❧ Spinal Tap
- ❧ Blood Tests to determine if there is inflammation.
- ❧ Sometime a nerve biopsy

Symptoms

- ☞ The symptoms the patient experiences and the neurologist confirms by examination include:
- ☞ Muscle weakness
- ☞ Loss of deep tendon reflexes
- ☞ Poor balance
- ☞ Distal loss (feet or hands > calves or forearms) of sensation including the ability to feel pain, light touch, and vibration
- ☞ Decreased perception of the position of a body part relative to the space around.

Treatments



- ☞ Treatments shown to work in CIDP are corticosteroids
- ☞ Plasmapheresis – a blood plasma exchange
- ☞ IVIG - intravenous immunoglobulin
- ☞ Other drugs that decrease immune function or inflammation are used in the treatment of CIDP

Immunosuppressive drugs



- ❧ immunosuppressive drugs are used to suppress the immune system.
- ❧ cyclophosphamide, cyclosporin, tacrolimus, mycophenolate mofetil, azathioprine and type 1 interferon (INF-1?) and IFN-beta can be used to limit corticosteroid and immunoglobulin use.
- ❧ May be indicated because of disease progression or poor response despite aggressive treatment with established therapies.

Therapy



- ❧ Many patients will require rehabilitative care including intensive physical and occupational therapy.
- ❧ The rehabilitation process itself does not improve nerve regeneration.
- ❧ The major goal of rehabilitation is to assist the patient in optimal use of muscles as their nerve supply returns.
- ❧ Patients need to adapt to a lifestyle within their functional limitations.

Principles of Rehabilitation for the GBS Patient.

- ❧ Strength usually returns in a descending pattern, so that arm and hand strength usually returns before leg strength.
- ❧ During the rehabilitation process, certain issues are unique to GBS patients. Most rehabilitation patients are exercised to maximum ability, to fatigue. This should be avoided in GBS patients as exhaustion requires some time to resolve and will delay the rehabilitation process without benefitting the patient.
- ❧ Therapist should be cognizant of the potential for substitution and customize exercises to strengthen weak muscles.
- ❧ Nerve pain can limit the patient's ability to tolerate rehabilitation and should be recognized and treated.

Q&A

