An Introduction to Myasthenia Gravis

Neuro ICU

slido

What do you know about Myasthenia Gravis?

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What is Myasthenia Gravis?

From the Greek for ‘grave muscle weakness’

An autoimmune disease of variable severity.

Characteristised by weakness and fatigue of skeletal muscle groups.

- It commonly effects muscles that control the eyes, facial expressions, Chewing swallowing and speaking

MG affects the neuromuscular junction.

- Immune system develops antibodies to nicotinic acetylcholine receptors (postsynaptic) in 90% of cases.

Neuromuscular Junction

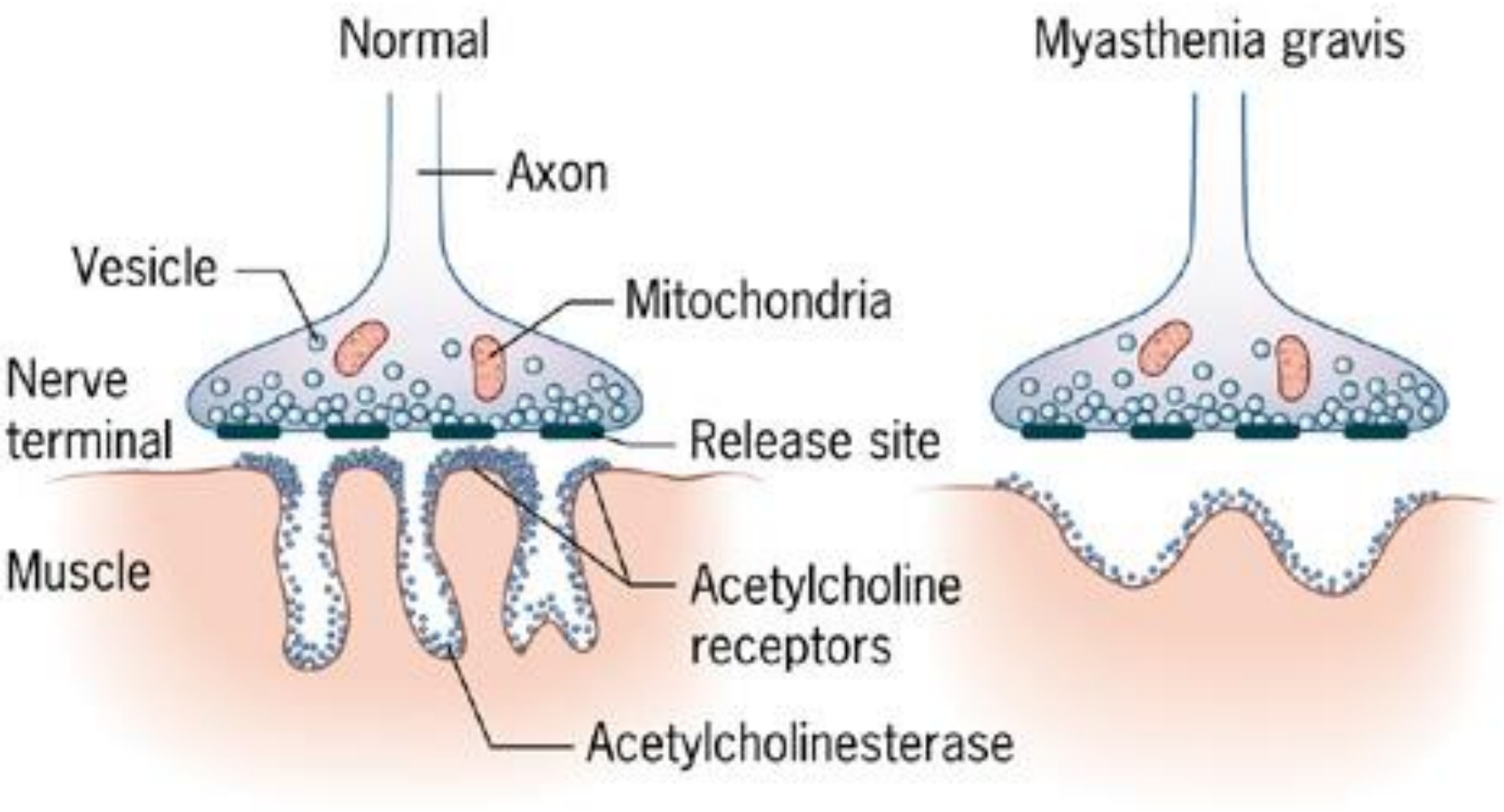
Acetylcholine (AcH) is released from vesicles in the pre-synaptic neurone.

AcH diffuses into the synaptic cleft and

binds to receptors on the post-synaptic membrane.

Muscle contraction occurs as a result.

The Neuromuscular Junction



In Myasthenia Gravis

In Myasthenia Gravis antibodies are made that damage the AcH receptor cells on the post synaptic neuron.

Reduced number of Ach receptors.

Decreased capacity for the NMJ to transmit impulse

The nerve transmission is impaired or blocked.

Myasthenia Gravis Statistics

Affects around 14-20 per 100,000 of population.

Anyone can be affected but more common in;

Women less than 40 years old affected most.

Males > 60 years old.

Periods of relapse and remission.

Causes

The thymus gland is implicated in 75% of cases.

- Thymus gland during childhood is responsible for development of immune system responses.
- MG patients may have benign thymomas in young women and hyperplasia in elderly men.

Genetic factors.

Diagnosis

- Based on history
- EMG (Electromyography)
- Antibody tests anti-Ach receptor
- MRI/CT to see if Thymus gland enlarged
- Anticholinesterase drug testing (Tensilon or Edroponium test)
- Nerve conduction studies

Symptoms

Vary depending on which muscles are affected.

Drooping of one or both eyelids (Ptosis).

Difficutly chewing, swallowing, speakng

Diplopia

Triedness after physical activity improving with rest.

Difficulty walking, holding head up.

Symptoms are made worse by;

- Physical over exertion
- Emotional stress
- Hot weather
- Menstruation
- Infection
- Alcohol
- Some household cleaners
- Chemical Lawn treatments

Treatment

Immunosuppressive therapy

Suppresses the production of antibodies Prednisolone, Azathioprin, Rituximab

Anticholinesterase Drugs

Medications used to treat disorder

- Slow the breakdown of acetylcholine at the neuro muscular junction, Therefore improving muscle strength
- Mestinon and Pyridostigmine

Plasmapheresis

- removes harmful antibodies
- usually for 5 days. Improvement within a week but effects last only 6-8 weeks.

Treatment

Thymectomy

- The Thyroid if often abnormal in patients with myasthenia
- A thymectomy will give long lasting remissions in 50% if cases

Immunoglobulin Therapy

- A concentrated dose of antibodies
- Pooled from many healthy donors
- It binds to the antibodies that cause myasthenia gravis and removes them from circulation
- given over 5 days
- with improvements within a week but effects last only 6-8 weeks. Gives immunosuppressant's time to work.

Myasthenic Crisis

Most likely cause of NICU admission due to respiratory decompensation.

Respiratory Failure

Bulbar weakness may also occur with hypophonic or nasal speech, difficulty chewing and swallowing, aspiration.

How do we treat these patients?

- Plasma Plasmapheresis
- Immunoglobulin
- Ventilation and chest physio

Giving medication to Myathenics

Withdrawal or increase in anticholinesterase.

- Unavoidable but be cautious
- Could cause a cholinergic crisis and worsen symptoms

Steroid commencement.

- A common treatment for myasthenia
- When they are started the myathenic symptoms may worsen for the first 2weeks

Antibiotics (Fluoroqinolones)

- Ciprofloxacin, Levofloxacin
- Can worsen symptoms

Antibiotics (aminoglycoside)

- Gentamycin, Amikacin, Clindamicin
- Can agrovate Pre existing MG

Giving medication to Myathenics…

Magnesium

- Can exacerbate Muscle Weakness
- IV can be potentially dangerous

Anaesthetic agents e.g. Vecuronium (non depolarising Blockers)

- even in small doses can have profound side effects If Sedating a myasthenic ICU must be on stand by,
- They may need More Paralysis than other patients as neostigmine is closely related to pridostigmine

Statins

- Increases muscle weakness and exacerbates MG

Iodinated contrast agents.

- Can exacerbate symptoms

Giving medication to Myathenics…

Botox

Quinine Used to treat muscle cramps,

- Quinine in tonic water can also bring on mild symptoms

Antihypertensives e.g. Betablockers,

- Shown to effect the neuromuscular junction

Calcium Channel Blockers

- can decrease neuromuscular transmission

Alpha blockers

- Methyldopa Causes fatigue

Non Potassium Sparing diuretics

- Furosemide

Precaution for Nurses

Pyridostigmine

- Ensure that gloves are worn when Handling
- Ensure you are in a well ventilated area
- Wear goggles to prevent powder getting into eyes
- Do Not Handle drug if you are pregnant

Check any new drug if safe to give

- Speak to your pharmacist

Symptoms of Cholinergic Crisis

- Drooping of one or both eyelids
- Blurred or double vision
- Changes in facial expressions
- Difficulty in swallowing
- Shortness of breath
- Difficulty talking and dyspagia

Myasthenic or Cholinergic Crisis?

Vital to detect if respiratory failure is due to Myasthenic or Cholinergic crisis.

Patients admitted may not be able to cope with Tensilon test and intubation should not be delayed.

Cholinergic Crisis is due to over medication with anticholinesterases.

Signs of a cholinergic crisis are abdominal cramps, diarrhoea, excessive pulmonary secretions (thick), salivation, urinary incontinence.

Table 2. Differentiating myasthenic from cholinergic crisis

	Myasthenic crisis	Cholinergic crisis
History of muscle weakness	Usually present	Usually present
Respiratory failure	Mostly	Mostly
Salivation, lacrimation, diarrhoea, urinary incontinence, abdominal cramps, fasciculations	Absent	Mostly
Heart rate	Tachycardia	Bradycardia
Pupils	Usually normal	Miosis
Bronchospasm	Absent	Present
Tensilon test	Positive	Negative (usually worsens symptoms)
Cholinesterase inhibitors	Possible non-compliance	Possible overdose

Jacob 2007

Specific Nursing Management Topics

- Neurology
- Respiratory
- Drug Therapy
- Supportive
- Psychological

Neurology

GCS to assess responsiveness while sedated.

Assess strength of all muscles involved

Extraocular muscles, is ptosis or diplopia present or improving.

Limb and muscle weakness strength tests ability to hand grip.

Cranial nerve assessment;

- Assess if there is diplopia, ptosis, facial weakness, bulbar function.

Respiratory Management

Indication for ventilation is a Vital capacity less than 15ml /kg (or less than 1L)

Ventilation and perfusion of lungs is normal so Oxygen saturation and ABG's often normal until late in the crisis.

Patient may be anxious and fearful but this may be masked due to facial muscle weakness.

Detection of aspiration pneuomonia by CXR, Sputum analysis.

Physiotherapy for secretion clearance. Cough may be ineffective.

- Sputum may be thick if in cholinergic crisis or if anticholinesterases are started.

Supportive

- NG tube for Nutritional support
- Monitor CVS
- Maintain normal blood chemistry.
- Observe for underlying infection, pyrexia.
- Elimination – if commenced on anticholinesterases may develop diarrhoea.
- Patients tire easily allow for specific rest periods.

Psychological

Support newly diagnosed patients as this is a lifelong illness with periods of remission.

Life changing illness due to symptoms, may require lifestyle adjustments, job, housing etc.

Thymectomy can offer cure in some cases but many patients will need teaching to manage the illness.

Sarah Jupp

With permission from sandy Jupp

First Admission

29/04/2010
Admitted to NICU

Myasthenia gravis for past 8 years. Admitted to ESH 12/4 in crisis.

PMH: Inoperable metastatic thymoma (had chemo).

28/4 trachy (ESH).

21/5/10 - Completed Plasmaphoresis therapy

25/5/10 - CT Pelvis, Abdo and Chest

Second Admission

08/12/2010 Admitted to NICU

SOB / low VC's. Intubated 12/12;

- 16/12 re-intubated.
- 20/12/10 - Percutaneous Tracheostomy

Inoperable metastatic thymoma - just completed palliative chemotherapy.

Third Admission

08/11/2014 Admitted to NICU

Norovirus,

ongoing chemotherapy

worsening SOB, diarrhoea

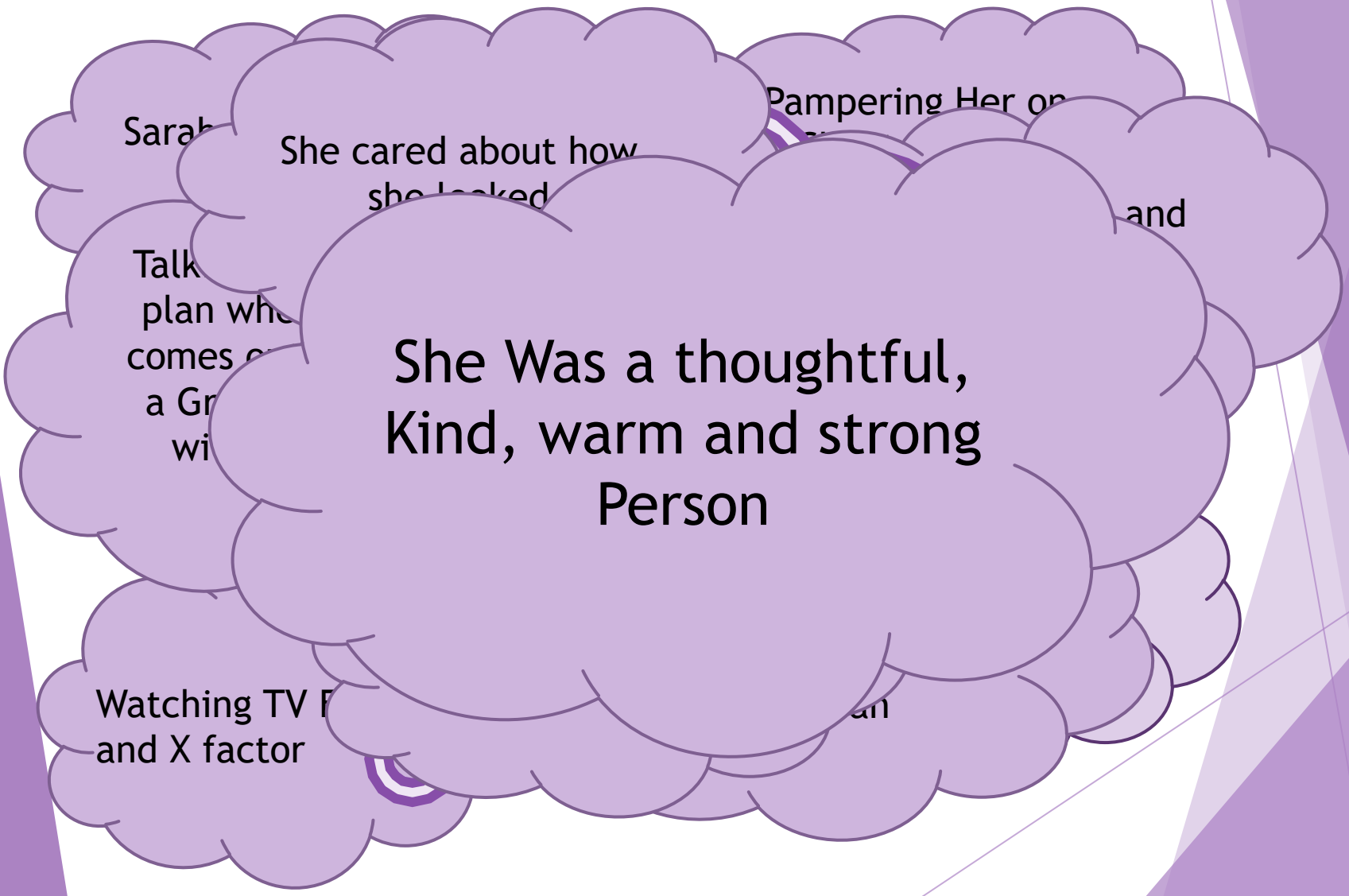
PMH: Malignant Thymoma, Myasthenia Gravis

CT: 21/11/14- progression of disease into left lung base, diaphram and peritoneum

plasma exchange, awaiting further chemo

Sarah Died 25th March 2015

Our Memories




The
Blyth Room

Teaching & Resources

- Information for patient and relatives regarding support organisations.

- Advise wearing of Medic Alert bracelet.
- Relapse of symptoms can be brought on by physical and emotional stress, extremes of temperature, menstruation, infections.

Resources

- Charity for patients and carers is;
- www.mgauk.org

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