NHS
Greater Glasgow and Clyde

Guidance for Clinicians Referring for Neurology Outpatient Advice and Assessment

"Right Place, Right Time, Right Clinician"

Contents

Introduction

Thank you for consulting this document which is aimed at supporting clinicians referring to the neurology outpatient department. More than ever, it is important that the patient pathway follows the most appropriate route from the outset so that unnecessary delays can be avoided, and patients are assessed by the appropriate clinician without duplication of care.

In this document, we outline some common neurological disorders and presentations. We offer some advice as to how to recognise different presentations and how to either seek advice from the vetting clinicians, or to refer to the neurology outpatient department. We include, where appropriate, advice on useful investigations and considerations to be made by the referring clinician prior to review by a neurologist. As such we hope this is a useful document for GPs and hospital doctors alike. Not all neurological presentations and disorders are covered here and therefore this document is not a substitute for clinical judgement. Some conditions, for example, TIA and stroke, are not covered in this document as there are other well established services to deal with these conditions.

This document applies to patients aged 16 years or older, who are resident in the NHS Greater Glasgow and Clyde catchment area & the Regional West of Scotland area.

If the referring clinician has a particular concern about a patient, they may wish to discuss the patient with the on call neurology team available through the QEUH switchboard (0141 201 1100) on a 24/7 basis.

Acute Referrals

Please note this document provides advice regarding outpatient referrals. If the referring clinician is concerned about more acute problems, telephone neurology advice is available on a 24 hour basis from the neurology on call registrar (under the supervision of an on call consultant). The on call neurology registrar can be reached via the QEUH switchboard.

If admission to hospital is required this should generally be through the acute medical admissions pathways via the local hospital.

Cognitive Disorders

Definition

Cognition is a term referring to mental processes including remembering, producing language, understanding, thinking and problem solving.

Cognitive impairment is when a person has trouble remembering, learning new things, concentrating, or making decisions that affect their everyday life. Cognitive impairment ranges from mild to severe.

Cognitive disorders that may be seen in neurology include:

- Alzheimer's disease with young onset or atypical features
- Dementia with Lewy bodies
- Fronto-temporal dementia and motor neuron disease
- Huntington's disease
- Corticobasal degeneration
- Progressive supranuclear palsy
- Creutzfeld Jakob disease (CJD)
- Vascular dementia with atypical features

Referral

Recommended Actions prior to referral:

Please ensure that medical and psychiatric causes of cognitive impairment, and other recommended pathways for referrals have been considered (see below).

- Clinical history of age of onset of cognitive symptoms, symptoms at onset, nature of cognitive symptoms, pace of evolution, impact on work and family life, issues with safety.
- Clinical history of other neurological symptoms such as weakness, swallowing and speech difficulties, muscle wasting, ataxia, seizures, tremor.
- Discuss issues regarding safety with patient and family (for example, driving and the DVLA regulations, work environment).
- History of past and current use of alcohol and recreational substances.
- History of diet and nutritional status.
- History of vascular risk factors.
- Family history of neurological illness, dementia, psychiatric illness.
- Neurological examination to document presence of weakness, muscle wasting, tremor, extra-pyramidal features of Parkinsonism.
- Blood tests including full blood count, ESR, CRP, B12, folate, renal, liver and thyroid parameters.
- List of current medications. Please review and consider withdrawal of medications that may impair cognition (for example, amitriptyline, opiates, methadone).
- Details of prior brain imaging if available.

Who to Refer (please see section below for referrals for suspected dementia in those >65y, and for suspected young onset dementia <65y)

Patients resident in NHS Greater Glasgow and Clyde.

New onset cognitive decline with associated neurological symptoms including seizures and neurological disability.

Rapid onset cognitive decline not due to mental health disease and medications.

Younger patients with cognitive decline with atypical features. These patients will be often be assessed by mental health services before referral to neurology.

Please note alternate referral pathways for cognitive disorders

Suspected Dementia in patients ≥ 65y: Patients over the age of 65 are assessed by memory services within community mental health services and Old Age Psychiatry.

Suspected Dementia in patients < 65y: Patients under the age of 65 with suspected dementia may be assessed by the Greater Glasgow and Clyde Young Onset Dementia service (please see appendix below).

Patients with a family history of Huntington's disease: such patients may be assessed by the mental health services.

Patients with prior head injury: such patients may be referred to the Head Injury service.

Patients with a prior stroke: such patients may be referred to the Stroke service for assessment by the Stroke psychology service.

Patients resident in other health boards are assessed in their local services.

How to Refer

GPs: please refer via SCI Gateway (with all clinical information as recommended above)

Others: (e.g. referrals from hospital)

Vetting Consultant

Cognition Clinic in Neurology

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Patients with significant mental health symptoms or patients already assessed or under the care of mental health services. You may wish to liaise with the clinicians in the mental health services for further advice.

Support Resources for Patients

Alzheimer Scotland

https://www.alzscot.org/

Rare Dementia Support:

https://www.raredementiasupport.org/

Professional Guidelines for Clinicians

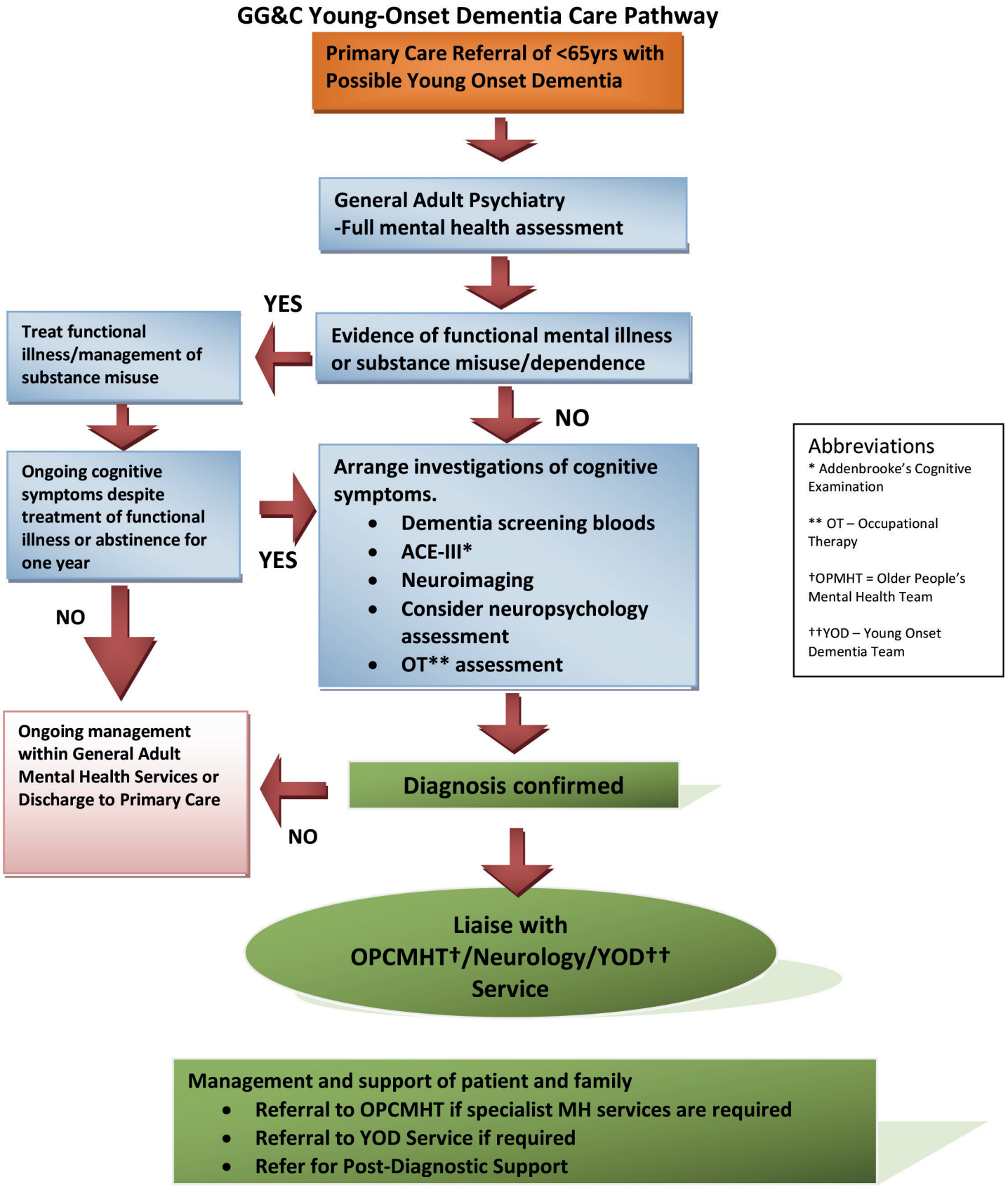
SIGN Guidance 86 Management of Patients with Dementia

http://umh1946.edu.umh.es/wp-content/uploads/sites/172/2015/04/Management-of-patients-with-dementia-NHS.pdf

Appendix

NHS Greater Glasgow and Clyde Young Onset Dementia Care Pathway

(please see Appendix below).



Epilepsy

Definition (adapted from ILAE 2014)

A seizure is a transient neurological symptom or sign resulting from abnormal excessive or synchronous neuronal activity. Seizures may be focal seizures (involving a part of the brain) or generalised seizures (involving bilateral cerebral hemispheres with alteration of consciousness, tonic and clonic features).

Epilepsy is an enduring predisposition to seizures. Epilepsy may be diagnosed if an individual has 2 or more unprovoked seizures in 10 years.

Infrequently, epilepsy can be diagnosed after a single seizure if risk of recurrence is high (for example in the presence of a brain tumour).

Provoked seizures due to alcohol or recreational substance misuse, or due to certain medications, are typically treated by addressing the provocative factor.

Typical Treatments

The aim of treatment in epilepsy is to reduce the risk of further seizures and help maintain normal lifestyle and function. Approximately 60-70% of people with epilepsy will achieve a prolonged period of seizure freedom. The choice of treatment is dictated by the type of epilepsy, age, gender (e.g. women of childbearing age) and associated comorbidity.

Commonly used medications include Lamotrigine, Levetiracetam and Carbamazepine but there are many other medications that may be considered.

Referral

Recommended Actions Prior to Referral:

- Clinical history of episode including prodrome or aura, alteration of consciousness, duration of episode, presence of tonic or clonic features, associated seizure markers such as lateral tongue injury, time to recover, post-ictal phase.
- Family history of seizures and epilepsy.
- Prior history of seizures.
- Use of alcohol and recreational substances (lead to provoked seizures).
- Current medications (several medications e.g. Tramadol can lead to provoked seizures).
- Routine bloods
- 12 lead ECG
- Advise patient and family regarding first aid measures if further episodes were to occur, safety at work and at home and lifestyle.
- If the patient is a driver, the patient must be advised to stop driving in keeping with the DVLA regulations.

Who to Refer:

New presentations

- Any patient with a suspected first seizure should be referred to the First Seizure clinic for further assessment.
- Patients with multiples seizures or a rapidly escalating frequency of seizures may be discussed with the on-call neurology team at the Queen Elizabeth University Hospital.

Patient with an existing diagnosis (not currently under routine follow-up)

- Patients with deterioration of pre-existing epilepsy
- Women with epilepsy who are considering starting a family.
- Patients with epilepsy wishing to stop their medication.
- Women of child-bearing age on teratogenic medication e.g. Valproate.

Patient with an existing diagnosis currently under routine follow-up

- Please contact the patient's named neurologist or epilepsy nurse specialist.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital) please address correspondence to:

Vetting Consultant
Epilepsy Service
Department of Neurology
Institute of Neurological Sciences
Queen Elizabeth University Hospital
1345 Govan Road
Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Patients with syncope are not assessed by neurology.

Patients with a confirmed diagnosis of epilepsy who are clinically stable and do not wish any change in treatment.

If patients with epilepsy are seizure free and clinically stable, they may be discharged for ongoing care to take place in primary care with the support of secondary care services if required. At times, patients may be offered Patient Initiated Review (PIR) for a period of time.

Patients with provoked seizures solely due to alcohol and recreational substance misuse, who have been previously assessed by neurology but need assessment and advice via the addiction services.

Support Resources for Patients

Epilepsy Scotland

https://www.epilepsyscotland.org.uk

Epilepsy Society

https://www.epilepsysociety.org.uk

Epilepsy Action

https://www.epilepsy.org.uk

Professional Guidelines for Clinicians

SIGN Guidelines

https://www.guidelines.co.uk/neurology-/sign-epilepsy-guideline/252618.article

NICE Guidelines

https://www.nice.org.uk/guidance/cg137

International League Against Epilepsy

https://www.ilae.org

Migraine

Definition

Features of Migraine Headache

(Adapted from www.ichd-3.org)

A.Headache has at least two of the following four characteristics:

1. unilateral location
2. pulsating quality
3. moderate or severe pain intensity
4. aggravation by or causing avoidance of routine physical activity (e.g., walking or climbing stairs)

B. During headache at least one of the following:

1. nausea and/or vomiting
2. photophobia and phonophobia

C. Not better accounted for by another diagnosis

Episodic migraine is defined when patients with migraine have less than 15 headache days per month. Chronic migraine is defined as headache on 15 or more days per month, for 3 months, at least 8 of which have migraine features.

Approximately 30% of patients have aura, most commonly visual aura.

Typical Treatments

Acute Abortive Therapies

Typical treatments for acute attacks include:

- Non-steroidal anti-inflammatories or high dose aspirin
- Anti-emetics such as metoclopramide
- Triptans (see Appendix)

Abortive therapies should be limited to 2 days per week. People with migraine are at risk of medication overuse headache (MOH) if abortive therapies are taken more frequently. For example, use of simple analgesia (NSAIDS, paracetamol) on 15 or more days per month, triptans on more than 10 days per month, and any use of opioid analgesia (especially if used more than 8 days per month). Opioid analgesia should be avoided.

Preventative Therapies

For those people with frequent disabling migraine, preventative therapies may be considered. The choice of therapy should take account of co-morbidities, the use of contraceptive agents, and drug interactions. Agents may include

- Propranolol
- Amitriptyline
- Topiramate
- Candesartan

For people with chronic migraine (15 or more headache days per month) who do not respond to the treatment above, further treatments issued from the headache specialist clinic may include

- Flunarizine
- Botox therapy delivered as part of a chronic migraine protocol, delivered 3 monthly
- Injections of monoclonal antibodies directed towards the calcitonin gene related peptide (CGRP)

Referral

Recommended Actions Prior to Referral:

- For patients with new onset headache at 50 years or older, consider performing CRP, ESR and FBC – if temporal arteritis is suspected, institute appropriate therapy and refer urgently to local acute medical or rheumatological services
- For patients with headache >3 months in whom cranial imaging is felt to be required, GPs may request this via "GP Access CT scan service", where available

Who to Refer

Examples of people who may benefit from being referred to the headache clinic

- People with a headache where there is **diagnostic uncertainty,** including those people with red flag features OR
- People with disabling migraine for whom **3 preventative medications** have been tried and have either been ineffective after 2 months at target dose, or have not been tolerated. In some patients with co-morbidities and medication contra-indications, we acknowledge that it may not be possible to try as many as 3 medications prior to referral. Trials of at least 3 preventative medications from the following list (see Appendix for further details) should have been attempted or considered, if safe to prescribe
  - » propranolol
  - » amitriptyline
  - » topiramate
  - » candesartan

Patients with thunderclap headache should not be regarded as having migraine. Patients with a thunderclap headache should be considered for admission via local medical receiving pathways to exclude serious causes including subarachnoid haemorrhage. Selected cases of patients with thunderclap headache occurring more than 2 weeks prior to referral may be appropriate for urgent outpatient investigation.

How to Refer

GPs: please refer via SCI Gateway, indicating clinical features of the headache (e.g. location, onset, frequency, nature) and which medications have been tried and for how long, and the reason for discontinuation.

Others: (e.g. referrals from hospital) for patients fulfilling the criteria outlined above in the "Who To Refer" section please address correspondence to:

Vetting Consultant

Headache Clinic

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road, Glasgow G51 4TF

Support Resources for Patients

NHS

https://www.nhs.uk/conditions/migraine/

Migraine Trust

https://www.migrainetrust.org

Professional Guidelines for Clinicians

SIGN 155 – Pharmacological Management of Migraine

https://www.sign.ac.uk/assets/sign155.pdf

British Association for the Study of Headache (BASH) (All aspects of primary headache management)

http://www.bash.org.uk/downloads/guidelines2019/01_BASHNationalHeadache_Management_SystemforAdults_2019_guideline_versi.pdf

NICE Guidance- "Headaches in over 12s: diagnosis and management

https://www.nice.org.uk/guidance/cg150/ifp/chapter/Treatments-for-migraine

Appendix – Guidance on Medications for Headache

Triptans

Please consider a prescription of a triptan, as per BNF. As a first option we recommend sumatriptan 50mg orally, as per SIGN 155.

Types of triptan: There are seven different triptans in total – sumatriptan, zolmitriptan, naratriptan, rizatriptan, almotriptan, frovatriptan, and eletriptan. Response to different triptans is variable, and people who fail to respond to one triptan may respond to another type of triptan. Therefore if the patient does not respond to one type of triptan after use in three separate attacks, please consider a trial of another triptan. Note that naratriptan and frovatriptan have a slower onset but a longer half-life and are therefore useful if patients describe a rebound headache with another triptan. All preparations come in tablet form. Sumatriptan comes as a subcutaneous injection also, and sumatriptan and zolmitriptan come in nasal spray preparations (useful if prominent nausea) and rizatriptan also comes in a melt preparation.

Side effects: Patients should be warned that triptan sensations may occur. Symptoms may include tightness in the jaw, throat, chest, and pins and needles in the face.

Cautions and contraindications: Triptans are contraindicated in coronary heart disease, vascular disease, or those with a history of stroke, and are cautioned in those with Raynaud's phenomenon. They should not be used in patients with a history or moderate or severe hypertension, or mild uncontrolled hypertension. Do not prescribe if blood pressure measurements are consistently above 140/90mmHg. Triptans are not licensed for adults greater than 65 years old and should be used with caution in this age group, especially in those with vascular risk factors.

How to Take Triptans: Triptans should be taken at the onset of the headache pain, and are more effective when taken earlier in an attack. They should be limited to two days per week – more frequent use can result in a rebound medication overuse headache. If the first dose is ineffective, a second dose should not be taken for the same attack. If there is response to the first dose, but symptoms recur, a second dose may be taken provided there is a minimum of 2 hours between doses of sumatriptan, rizatriptan, zolmitriptan, almotriptan, eletriptan, frovatriptan, and 4 hours between doses for naratriptan.

Drug Interactions (non-exhaustive list)

Triptans should not be combined with monoamine oxidase inhibitors. In patients taking propranolol, limit rizatriptan to the 5mg dose.

Please check BNF for drug interactions in those taking antibiotics, antifungal agents, cimetidine, antiretroviral agents, and verapamil – interactions vary between triptans

Migraine Prophylactic Drugs

General Prophylaxis Advice

Migraine prophylactics may take many weeks to work. Judgment of efficacy should be made after being on the target dose or highest tolerated dose at 3 months. If the migraine prophylactic is ineffective at 3 months, please wean over approximately 2 weeks. If it is effective (i.e. reduces headache days by 30-50%) consider weaning the drug after 6 to 12 months. If side effects are experienced after a dosage increase please decrease to previous dose and then attempt a dosage increase after 2 weeks. If patients are drowsy they should be warned to refrain from driving. Driving should be avoided when starting medications, and around the time of each dosage increase in case of side effects.

Migraine prophylaxis is generally not recommended in pregnancy, and certain medications are contraindicated in pregnancy.

Amitriptyline

Week 1 – 10mg nocte

Week 2 – 20mg nocte

Week 3 – 30mg nocte

Week 4 – 40mg nocte

Week 5 – 50mg nocte

Patients should be warned about side effects which include constipation, difficulty with micturition, arrhythmias, syncope, confusion, nausea, dry mouth, drowsiness and weight gain. Patients should seek immediate medical attention if they are unable to micturate of experience visual blurring.

Alternatives include nortriptyline or dosulepin.

Propranolol

Please prescribe propranolol 20mg bd, gradually up-titrating to 80mg bd.

When the dose reaches 80mg bd the tablets could be converted to a modified release preparation in order to minimise side effects.

Beta blockers are contraindicated in a number of conditions including asthma, heart failure, PVD, depression and should not be used in patients taking verapamil.

Side effects include bradycardia, hypotension, fatigue, sexual dysfunction, wheezing.

Topiramate

Please prescribe topiramate as below

Increase the dose until benefit is achieved or there are intolerable side effects, as below.

Week 1 – 25mg nocte

Week 3 – 25mgs twice daily

Week 5 – 25mgs mane, 50mg nocte

Week 7 – 50mgs twice daily

It should not be used in patients who have a history of glaucoma or renal stones or anorexia nervosa. Caution should also be exercised in patients with a history of depression. There may be interactions with digoxin, metformin , carbonic anhydrase inhibitors, and thiazide derivatives. There is a potential for serious interaction with sodium valproate and concomitant use is not recommended.

Sexually active women of child bearing age should take adequate contraceptive precautions since this medication is teratogenic. Owing to enzyme inducing effects (at any dose) the recommended contraception from the Faculty of Sexual and Reproductive Healthcare (2018) is either the levonorgestrel intrauterine system (LNG-IUS), or the progestogen-only injectable: depot medroxyprogesterone acetate (DMPA).

Side effects include acute glaucoma, peripheral paraesthesias, fatigue, nausea, diarrhoea or weight loss, taste change, concentration difficulties, word finding difficulties, insomnia, anxiety, and depression

Candesartan

Start at 4mg per day, increasing by 4mg every week to a maximum of 16mg.

Avoid in patients with renal artery stenosis, hypotension, renal impairment, or history of angioneurotic oedema.

Candesartan should not be used in those patients receiving lithium therapy, and is cautioned for use in those who are taking medications which increase serum potassium such a spironolactone

Candesartan should not be used in pregnancy, and should be discontinued before planning a pregnancy. Women of child bearing age should ensure appropriate contraception is in place. Candesartan is not recommended during breast feeding.

Side effects include hypotension, renal impairment and cough **During dose titration of Candesartan, monitoring of serum creatinine, potassium is recommended in those with renal impairment.**

Cluster Headache

Definition

Features of Cluster Headache

(Adapted from www.ichd-3.org)

A. At least 5 attacks fulfilling criteria B-D

B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes (when untreated). Some attacks may be less severe, and shorter/longer

C. Either or both of the following:

1. at least one of the following symptoms or signs, ipsilateral to the headache:
   - » conjunctival injection and/or lacrimation
   - » nasal congestion and/or rhinorrhoea
   - » eyelid oedema
   - » forehead and facial sweating
   - » miosis and/or ptosis
2. a sense of restlessness or agitation

D. Occurring with a frequency between one every other day and 8 per day (may be less frequent at times)

E. Not better accounted for by another diagnosis

*Most patients have episodic cluster headache pattern. However, if attacks occur for more than 1 year without remission for more than 3 months, the pattern is termed 'chronic cluster headache'

Typical Treatments

Acute Abortive Therapies

Typical treatments for acute attacks include

- Sumatriptan 6mg subcutaneously, can be repeated once in a 24 hour time period if further attacks occur, with a minimum separation of 1 hour between doses. Zolmitriptan (intranasal) 5mg is a less effective alternative.
- 100% oxygen at 12-15l/min, for around 15 minutes. Typically prescribed from the headache clinic

Bridging Therapies

- Greater occipital nerve (GON) blocks with local anaesthetic and steroid (if appropriate) may be administered at the start of a cluster bout, to allow other preventative therapies to take effect, or to reduce the length of the bout. GON blocks are also effective in some patients with chronic cluster headache

Preventative Therapies

- Verapamil is the treatment of choice for patients whose cluster bouts last for many weeks. This is typically suggested from the headache clinic and stringent ECG monitoring is required.
- If verapamil is ineffective, other agents are sometimes recommended by the headache clinic, including lithium (with monitoring), topiramate, sodium valproate, gabapentin, and melatonin.

Other Therapies

- Non -invasive vagal nerve stimulation (Gammacore ®) may be effective in some patients for acute and preventative treatments. This device is sometimes arranged through the headache clinic.

Referral

Who to Refer

- Most patients with suspected cluster headache should be referred to the headache clinic.
- However, **for patients presenting with a first presentation of cluster headache, consideration should be given to investigation for secondary causes** and should be discussed with the on call neurology team prior to referral to the headache clinic. Mimics of cluster headache include temporal arteritis, glaucoma, acute sinusitis, tumours, stroke, arteriovenous malformations and dissection of the carotid or vertebral arteries. Other rarer trigeminal autonomic cephalgias may also be considered (e.g. paroxysmal hemicrania and SUNCT).

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital) please address correspondence to:

Vetting Consultant

Headache Clinic

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Support Resources for Patients

NHS

https://www.nhs.uk/conditions/cluster-headaches/

OUCH UK

https://ouchuk.org

Professional Guidelines for Clinicians

British Association for the Study of Headache (BASH) (All aspects of primary headache management)

http://www.bash.org.uk/downloads/guidelines2019/01_BASHNationalHeadache_Management_SystemforAdults_2019_guideline_versi.pdf

NICE

https://cks.nice.org.uk/topics/headache-cluster/

Idiopathic Intracranial Hypertension

Definition

Definition:

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a disorder of increased intracranial pressure that occurs mainly in **overweight women of childbearing years,** often in the setting of weight gain.

Presenting Features:

Typically, patients present with persistent headaches that may have features of raised intracranial pressure (worse on lying down, better on standing, worse with straining, coughing, laughing and sneezing). Patients may also describe pulsatile tinnitus (pulse synchronous), worse in the lying down position. Transient visual obscurations may be experienced and are described as loss of vision for < 30 seconds, with full recovery. It is common to have migrainous headaches. Some may have diplopia due to sixth cranial nerve palsy. Patients may experience blurring of vision.

None of the above features are pathognomonic.

Diagnostic criteria:

- Papilloedema (signs of raised intracranial pressure)
- No localising neurological signs (except those that can be solely explained by raised pressure only such as 6th nerve palsy)
- No structural abnormality of brain and dural sinuses – confirmed by adequate imaging including MRI or CT brain with venography (to ensure there is no other cause for raised intracranial pressure)
- CSF pressure >25 cm (to be interpreted in the appropriate clinical context - up to 30 cm can be seen in normal population)
- CSF constituents normal

Typical Treatments

- Look for other causes and treat accordingly. Please stop medications such as tetracycline, retinoic acid preparations and anabolic steroids.
- Weight loss - generally aim for 10% body weight loss over 4 to 6 months (refer to a weight loss programme).
- Acetazolamide.
- An alternative to acetazolamide is topiramate (can render hormonal contraception ineffective due to an enzyme inducing effect. Please see guidance on interactions from the Faculty of Sexual and Reproductive Health)
- Do not use lumbar puncture solely to treat headache in IIH. If the vision is at risk then lumbar punctures, lumbar drains and CSF shunt procedures may be considered.
- Do not use corticosteroids as a treatment.
- Treat chronic headache according to headache presentation (e.g. migraine, medication overuse headache).

Referral

How to Refer

GPs:

- For patients who are well and asymptomatic and have suspected papilloedema, please refer to an optician for assessment. If there are further concerns please refer to an ophthalmology service urgently.
- For patients with headache with suspected papilloedema, please refer for hospital admission immediately, via local receiving pathways.

Please do not refer patients with suspected papilloedema to neurology out-patients, as this is likely to cause delay in assessment.

Others: (e.g. referrals from hospital)

- For current inpatients, management should be discussed with the on-call neurology team. If neurology inpatient review is required, referral to the local neurology liaison service may be advised
- Patients should be discussed with the on-call neurology team prior to discharge and if it is agreed that it is appropriate should be referred to the IIH clinic as below:

  Vetting Consultant

  IIH Clinic

  Department of Neurology

  Institute of Neurological Sciences

  Queen Elizabeth University Hospital

  1345 Govan Road

  Glasgow G51 4TF

Support Resources for Patients

IIH Support UK

https://www.iih.org.uk

Professional Guidelines for Clinicians

BMJ Best Practice

Idiopathic intracranial hypertension - Symptoms, diagnosis and treatment |

https://bestpractice.bmj.com/topics/en-gb/1070

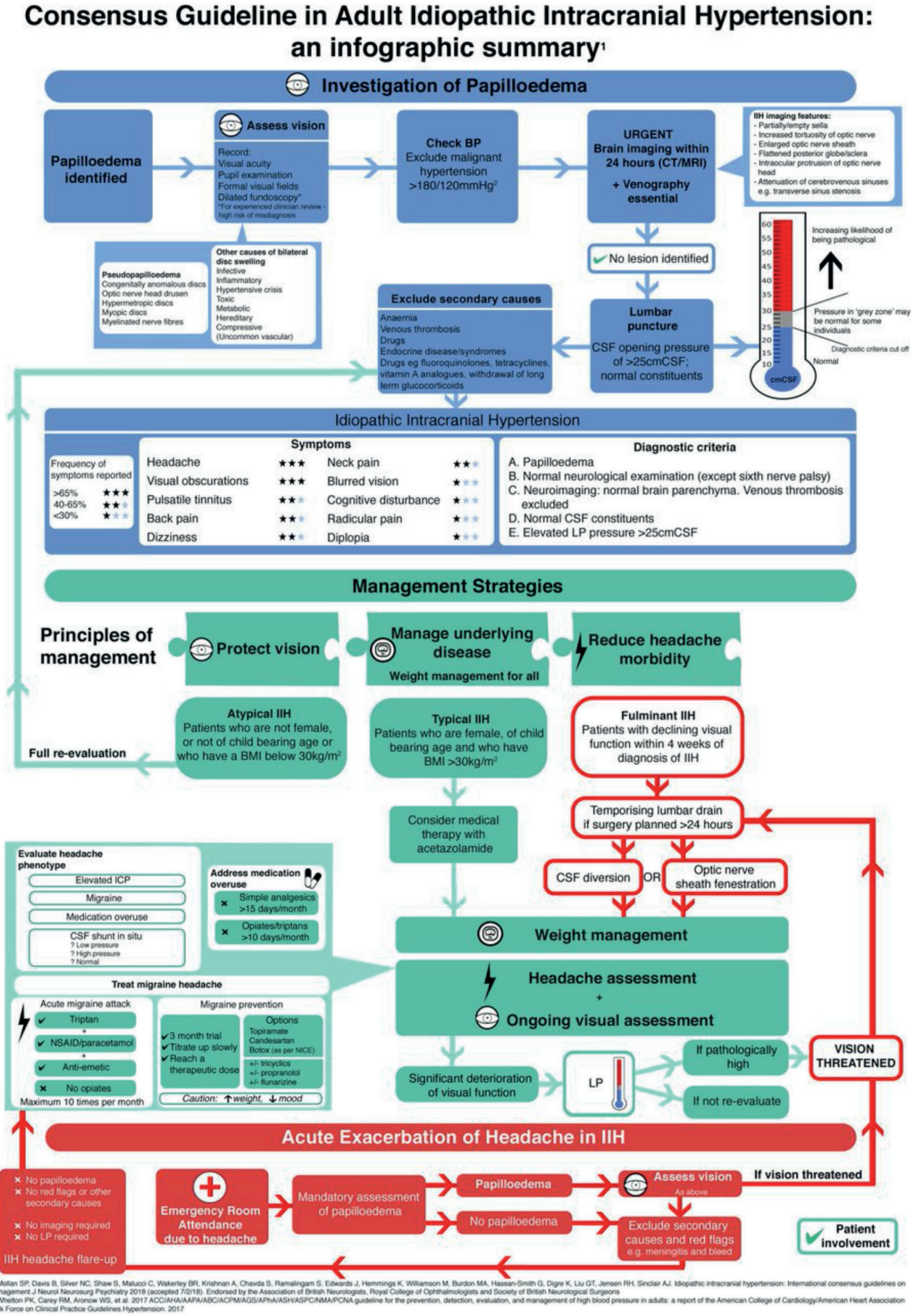
Consensus Guidelines for IIH

Evaluation and management of adult idiopathic intracranial hypertension
http://dx.doi.org/10.1136/practneurol-2018-002009

Interactions document from the Faculty of Sexual and Reproductive health, describing interactions between medication and contraception

fsrh-guidance-drug-interactions-hormonal-contraception-jan-2019.pdf

Appendix



Reproduced from Mollan SP, et al. Pract Neurol 2018;18:485–488.
http://dx.doi.org/10.1136/practneurol-2018-002009

Motor Neurone Disease

Definition

Motor neurone disease (MND) is a neurodegenerative disorder.

It is characterised by progressive weakness typically affecting a limb or limbs, speech and swallowing function. This is typically not associated with loss of feeling or pain.

MND most commonly occurs in the mid-sixties affecting males more than females and classically evolves over months. However, it can occur in any age group.

There can be a family history of the condition in ten percent of patients. Neurological findings on examination that are consistent with MND include a combination of upper and lower motor neurone signs in multiple anatomic zones, for example limbs and bulbar musculature (early symptoms and signs may be confined to one zone). Cognitive and behavioural disturbance can co-exist with weakness. Breathlessness and respiratory failure can develop or rarely be the presenting symptom. Examination features that would be against the diagnosis of MND include ptosis, limitation of down gaze, tremor, autonomic involvement and sphincter dysfunction.

Typical Treatments

There are no known curative treatments. Riluzole is the only licensed drug treatment in the United Kingdom that can slow the evolution of the condition. It does not stabilise or reverse the disorder. It is given as a tablet twice a day and can rarely affect the liver and or blood count necessitating blood monitoring.

Supportive approaches tailored to the individual with respect to mobility aids and optimising their living environment is an important focus of therapy. In addition, symptom control of muscle cramp, saliva drooling, difficulty with breathing, eating and drinking can be addressed on an individual basis.

Referral

Recommended Actions Prior to Referral:

Is there an imminent choking risk? If yes, consider admission to hospital via local receiving pathways.

Is there imminent respiratory failure? If yes, consider admission to hospital via local receiving pathways

Who to Refer

Patients with progressive painless weakness. See the “Think MND” document in the appendix.

How to Refer

GPs: If MND is suspected, an urgent referral to Neurology should be made via SCI Gateway.

Others: (e.g. referrals from hospital)

If MND is suspected during a hospital admission, please contact the on-call neurology service at the Institute of Neurological Sciences via switchboard. A ward review by the visiting neurologist as an inpatient may be advised, or alternative routes of review suggested.

For other outpatient referrals please refer to:

Vetting Consultant

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road, Glasgow G51 4TF

Support Resources for Patients

MND association

https://www.mndassociation.org/

MND Scotland.

https://www.mndscotland.org.uk/

Professional Guidelines for Clinicians

NICE guidelines for MND

https://www.nice.org.uk/guidance/ng42

Problem solving approach to MND published by MND Scotland:

https://www.mndscotland.org.uk/media/2930/problem-solving-booklet-2020.pdf

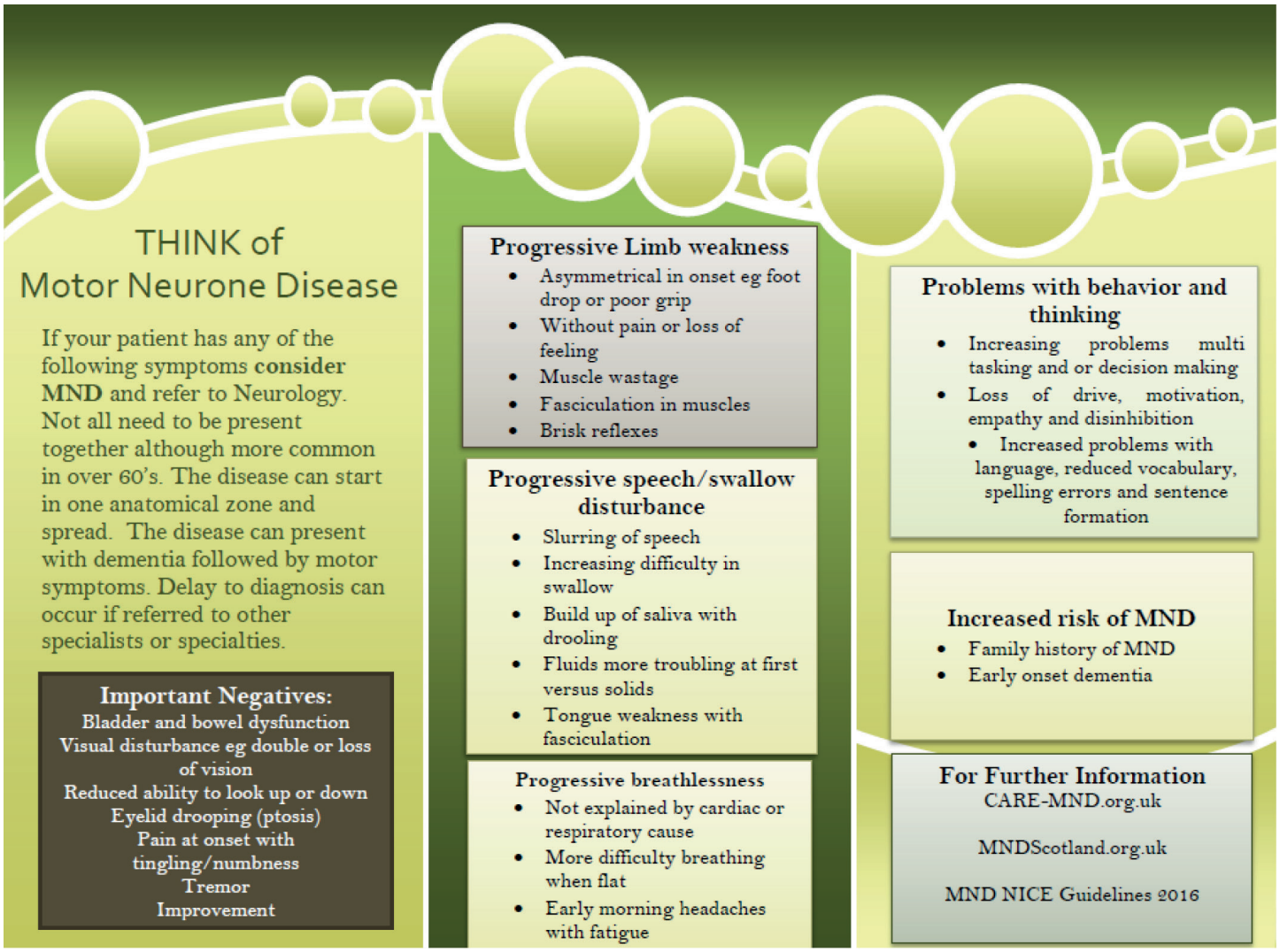
Saliva management guidelines GGC:

http://www.staffnet.ggc.scot.nhs.uk/Info%20Centre/PoliciesProcedures/GGCClinicalGuidelines/GGC%20Clinical%20Guidelines%20Electronic%20Resource%20Direct/Motor%20Neurone%20Disease%20-%20Saliva%20Control%20Guidance.pdf

Palliative care

https://www.palliativecareguidelines.scot.nhs.uk/

Appendix



From Ann Rowling Regenerative Neurology Clinic

Movement Disorders - Dystonia

Definition

Dystonia is a movement disorder in which muscles contract involuntarily, causing repetitive or twisting movements. The condition can affect one part of the body (focal dystonia), two or more adjacent parts (segmental dystonia) or most of the body (generalised dystonia). The muscle spasms can range from mild to severe.

Types of dystonia include

- Cervical dystonia
- Blepharospasm
- Oromandibular dystonia
- Task specific dystonia such as writer's cramp or musician's dystonia
- Generalised dystonia e.g. due to cerebral palsy or due to an underlying genetic disorder
- Tardive dystonia secondary to neuroleptics or metoclopramide or prochlorperazine
- Dystonic tremor
- Functional dystonia
- Dopa-responsive dystonia
- Hemi-facial spasm (this is not actually a type of dystonia, but is often treated in a similar way with botulinum toxin)
- Paroxysmal dystonia and dyskinesias

Typical Treatments

Botulinum toxin may be considered for certain types of focal dystonia such as cervical dystonia or blepharospasm. It may be considered in task specific dystonia.

Oral treatments include medications such as anticholinergics (e.g. trihexyphenidyl), benzodiazepines (e.g. clonazepam) and antispasmodics (e.g. baclofen). Combinations may be considered in some types of generalised dystonia but may not be helpful in focal dystonia.

Levodopa based medications are useful for patients with dopa-responsive dystonia (which is very rare).

Deep Brain Stimulation. Some patients with medically refractory dystonia may be considered for Deep Brain Stimulation.

Physiotherapy, occupational therapy or speech and language therapy may be of benefit.

Referral

Recommended Actions Prior to Referral

- Clinical history including age of onset, nature and pace of onset, location of symptoms, functional impairment, presence or absence of pain.
- Prior medical history.
- Family history of dystonia or neurological illness.
- Please review medications in case the patient is on medications that could be causing symptoms.
- Blood tests including full blood count, ESR, B12, folate, thyroid function, liver and renal parameters, creatine kinase.

Who to Refer

Please refer patients with suspected dystonia to the neurology service for assessment and management.

Please consider alternate services

In many areas, patients with blepharospasm are managed by ophthalmologists and could be referred to those services.

Patients with dystonia related to cerebral palsy may be referred to their local neurorehabilitation team rather than to neurology.

Patients with post-stroke dystonia should be referred to the stroke services.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital)

Please address correspondence to:

Vetting Consultant

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Patients who have been previously seen by a specialist and have trialled and failed with medications or other types of management (such as physiotherapy) may not benefit from re-referral to the s ervice, and may benefit more from referral to rehabilitation services or allied healthcare professionals such as physiotherapy.

Support Resources for Patients

Dystonia UK is a national patient charity, with local self-help groups

www.dystonia.org.uk

NHS website

www.nhs.uk/conditions/Dystonia or

www.nhsinform.scot/illnesses-and-conditions/muscle-bone-and-joints/conditions/dystonia

National Institutes of Health (USA) website

www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Dystonias-Fact-Sheet

Professional Guidelines for Clinicians

Dystonia: A Guide to Best Practice. For Health and Social Care Professionals

https://www.dystonia.org.uk/Handlers/Download.ashx?IDMF=2b634984-6d69-4612-9c53-b0d9c81b20ea

Movement Disorders - Essential Tremor

Definition

Essential tremor is the most common tremor disorder causing rhythmic shaking of the arms and less often voice, face, neck and lower limbs.

It is often bilateral in the upper limbs (but may be asymmetrical) and predominantly on maintaining posture and during movement (arms outstretched and moving between goals) and so may cause problems with holding cups or cutlery.

Fifty percent of patients may improve on drinking alcohol or taking beta-blockers. There is often family history of a tremor.

The differential diagnosis includes medication related tremor (e.g. due to beta agonists, valproate, lithium, steroids, thyroxine or certain antidepressants), dystonic tremor (which may have additional dystonic posturing of the limbs or of the neck such as torticollis), and parkinsonism (which is often asymmetrical at onset, with tremor at rest, with associated bradykinesia and rigidity).

Typical Treatments

Non-medical management of Essential Tremor

Cessation of caffeine, reduction in alcohol, management of any exacerbating anxiety, withdrawal of any medications that may cause or exacerbate tremor, metabolic screen (thyroid, renal, glucose, calcium), use of heavier cutlery or pen or cup, drinking through a straw, typing instead of writing or for more severe functional impairment, a community occupational therapy referral. Patients may gain further information from the National Tremor Foundation website www.tremor.org.uk

Medical management of Essential Tremor

If the tremor is functionally impairing and there are no contraindications then consider medications. Around 50% of patients respond to propranolol. Response to other medications varies, and can be disappointing and patients should be counselled regarding expectations of treatment versus side effect profiles. The choice should take into account co-morbidities such as asthma and the use of contraceptive agents. In patients with co-morbidities (especially cognitive impairment or in those who are unsteady) it is our usual practice to avoid any medications that are known to cause sedation (such as primidone and clonazepam). We also avoid clonazepam in those who are at higher risk of developing addiction (such as those with previous addictions or addictive behaviour).

Some patients will benefit from occasional use e.g. of propranolol to manage tremor which is worse in specific situations. Otherwise treatment should be trialled for at least 8 weeks at maximally tolerated dose before moving onto any other appropriate medication.

Agents may include; (see Appendix)

- Propranolol
- Topiramate
- Primidone
- Clonazepam
- Gabapentin

Sometimes these can be used in combination (e.g. propranolol and primidone)

Surgical treatment

In those with very severe tremor, brain surgery techniques such as deep brain stimulation may be considered. This is appropriate only in those who are medically well, severely disabled by their tremor and those who are refractory to medical treatments.

Referral

Recommended Actions Prior to Referral

- Clinical history including age of onset, duration of symptoms, location of symptoms, symmetrical or asymmetrical tremor, exacerbating and relieving factors, presence or absence of functional impairment, impairment of gait.
- Family history of tremor or neurological illness.
- Please review any medication that may cause tremor. Withdraw medication or reduce dose if possible.
- Blood tests including full blood count, ESR, B12, folate, renal, liver and thyroid parameters, bone profile, serum glucose.
- Please give advice to patient on non-medical management.
- Consider trial of medications for essential tremor if there is reasonable diagnostic security for essential tremor.

Who to Refer

If there is significant diagnostic doubt about a tremor diagnosis, or new signs emerge that aren't in keeping with a benign tremor. Please note cogwheeling and gait imbalance can occur in severe essential tremor present for decades.

If medical treatments have been tried but tremor remains functionally impairing, and the patient may be suitable for surgical treatments.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital)

Vetting Consultant

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4T

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

If a patient has mild bilateral kinetic tremor (without any other neurological signs) with no functional impairment, monitoring in primary care would be reasonable in the first instance, after exclusion of metabolic causes and drug induced causes.

If a patient has previously been seen regularly in the movement disorders clinic but has a refractory tremor and has significant co-morbidities such as COPD or polypharmacy that would prevent use of beta-blockers or cognitively impairing drugs, patients are unlikely to benefit from medications or surgical treatments and should have supportive care including community occupational therapy.

If there is diagnostic security for essential tremor in a patient with a dominant family history of tremor and clinically has bilateral postural tremor without other signs, most patients do not require referral to neurology specialist services.

Support Resources for Patients

National tremor Foundation website

www.tremor.org.uk

The National Deep Brain Stimulation Service NHSGGC

www.nhsggc.org.uk/your-health/health-services/deep-brain-stimulation

Professional Guidelines for Clinicians

Essential Tremor, deceptively simple. Practical Neurology 2007, Vol 7, issue 4

https://pn.bmj.com/content/7/4/222.short?rss=1&ssource=mfc

MDS Evidence-Based Review of Treatments for Essential Tremor. Movement Disorders, Vol. 34, No. 7, 2019

https://pubmed.ncbi.nlm.nih.gov/31046186/

Appendix

- Propranolol (licensed). If no contraindications please start propranolol 20mg twice daily (lower in elderly) up to 80 - 160mg total daily dose (if BP and pulse allow). Higher doses may be used in selected patients. It is contraindicated in a number of conditions including asthma, heart failure, PVD, depression and should not be used in patients taking verapamil. Side effects include bradycardia, hypotension, fatigue, sexual dysfunction, wheezing.
- Primidone (licensed). Please start at 50mg once daily increasing on a 2 weekly basis if tolerated to 250mg three times daily (rarely tolerated at this level). Common side effects include sedation, nausea, unsteadiness, confusion and shouldn't be used in the elderly or those with significant comorbidities. Check interactions in the BNF - it is an enzyme inducer of CYP3A4 and interferes with oral contraceptive pill.
- Topiramate (unlicensed for tremor). Please start at 25mg nocte increasing by 25mg every 2 weeks (divided dose). Up to 100mg twice daily was shown to be useful in clinical trials but these higher doses are more likely to cause cognitive side effects. Side effects include acute glaucoma, peripheral paraesthesias, fatigue, nausea, diarrhoea, appetite suppression and weight loss, concentration difficulties, word finding difficulties, insomnia, anxiety and depression, propensity to develop renal stones, changes in taste. Sexually active women of child bearing age should take adequate contraceptive precautions since this medicine is teratogenic. Owing to enzyme inducing effects (at any dose) the recommended contraception from the faculty of Sexual and reproductive Healthcare (2018) is either levonorgestrel intrauterine system (LNG-IUS), or the progestogen-only injectable - depot medroxyprogesterone acetate (DMPA).

  Check interactions in BNF, interactions include with digoxin, metformin, carbonic anhydrase inhibitors, thiazide derivatives. There is a potential for serious interaction with sodium valproate and concomitant use is not recommended.
- Clonazepam (unlicensed for tremor). Please consider 0.5mg nocte increasing to maximum of 0.5mg twice daily. Side effects include sedation, potential for abuse, anxiety and depression.
- Gabapentin (unlicensed for tremor). Please start 300mg daily as per the BNF, and titrate to 600mg twice daily. Side effects include lethargy, fatigue, dyspnoea, decreased libido, dizziness, anxiety, increased appetite.

Movement Disorders - Parkinsonism

Definition

Parkinsonism is an umbrella term that describes a group of disorders with signs of bradykinesia plus one or more of rest tremor, rigidity or postural instability. Parkinson's disease is the most common form of parkinsonism and is often unilateral in onset and progressive.

Types of Parkinsonism include;

a. Parkinson's disease

b. Drug induced parkinsonism (eg due to neuroleptic or antiemetic medications such as prochlorperazine or metoclopramide or due to sodium valproate)

c. Lewy body dementia

d. "Atypical parkinsonism" or "Parkinson's Plus conditions"

    i. Multiple System Atrophy (MSA)

    ii. Progressive Supranuclear Palsy (PSP)

    iii. Corticobasal Degeneration (CBD)

e. Vascular parkinsonism

Typical Treatments

Treatments depend on the underlying cause of parkinsonism. Withdrawal of any precipitating drugs may cause a complete improvement although can take many months to do so.

Treatments for people with Parkinson's Disease are dependent on symptoms and may include referral to various allied health professionals. Exercise is very important. Medications include levodopa based medications Co-careldopa (Sinemet), or Co-beneldopa (Madopar), or dopamine agonists (such as Pramipexole, Ropinirole or Rotigotine), MAO-B inhibitors (Rasagiline, Selegiline) or COMT inhibitors (Entacapone, Opicapone). These should be started by a specialist. Deep Brain Stimulation (DBS) Surgery may be appropriate for some patients who have symptoms which are refractory to medication.

For those with atypical parkinsonism and Parkinson's plus, the treatments may include medications used for Parkinson's, although these are less effective. Supportive management is required.

People with Lewy Body Dementia should be under the care of the local memory team which is usually run by psychiatry and may benefit from cognitive enhancers and access to a community psychiatric nurse (CPN). Treatments for any accompanying movement disorder may not be required, may be less effective and are more likely to give neuropsychiatric side effects.

There are no specific drug treatments for vascular parkinsonism apart from supportive care and secondary prevention of cerebrovascular disease.

Referral

Recommended Actions Prior to Referral:

- Clinical history including age of onset, duration and nature of symptoms, location of symptoms, symmetrical or asymmetrical symptoms, functional impairment, impairment of gait and balance.
- Please enquire regarding impairment of cognition.
- Please discuss safety at home in case of impaired balance and falls or if impaired cognition.
- Family history of tremor or neurological disease.
- Please review medications in case the patient is on any medication that could be causing symptoms (e.g. neuroleptic agents, metoclopramide, prochlorperazine, sodium valproate).
- Please assess for possible postural hypotension.
- Please do not start medications for Parkinson's Disease prior to referral.

Who to Refer

Consider referral for patients who have suspected parkinsonism other than those where drug induced parkinsonism is suspected and symptoms resolve after cessation of offending medications.

Patients <65 years old are referred to the neurology service and those who are older than 65 years are typically referred to the local Department of Medicine for the Elderly services.

Patients from NHS Ayrshire and Arran and NHS Lanarkshire are typically referred to their local services.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital) for patients fulfilling criteria outlined above in "who to refer" section please address correspondence to:

Vetting Consultant

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Note that patients with longstanding essential tremor may normally have evidence of mild cogwheel rigidity and usually do not need to be referred for assessment.

Patients already diagnosed with vascular parkinsonism are unlikely to benefit from being re-referred if they already been assessed and investigated. Such patients should be given supportive care.

Patients with symptoms suggestive of Lewy Body Dementia (e.g. those early and more prominent cognitive symptoms, early hallucinations and mild motor symptoms) are likely to require input from the memory team for diagnosis and may not require neurology input.

Support Resources for Patients

Parkinson's disease

NHS website on Parkinson's Disease

https://www.nhs.uk/conditions/parkinsons-disease

Parkinson's UK charity is a national resource for patients and caregivers and has local advisors

https://www.parkinsons.org.uk

Atypical parkinsonism resources

Multiple System Atrophy

https://www.msatrust.org.uk

Progressive Supranuclear Palsy Association (this charity is also for those with Corticobasal degeneration)

https://pspassociation.org.uk

Professional Guidelines for Clinicians

SIGN guidelines for Parkinson's (2010)

https://www.sign.ac.uk/our-guidelines/diagnosis-and-pharmacological-management-of-parkinsons-disease/

NICE guideline (2017): Managing Parkinson's in adults

https://www.nice.org.uk/guidance/NG71 **and**

https://cks.nice.org.uk/topics/parkinsons-disease

Restless Legs Syndrome

Definition

Restless legs syndrome (RLS) is defined by the International RLS Study Group as

A. An **urge to move the legs** usually but not always accompanied by, or felt to be caused by, uncomfortable and unpleasant sensations in the legs.

B. The urge to move the legs and any accompanying unpleasant sensations **begin or worsen during periods of rest or inactivity** such as lying down or sitting.

C. The urge to move the legs and any accompanying unpleasant sensations are **partially or totally relieved by movement,** such as walking or stretching, at least as long as the activity continues.

D. The urge to move the legs and any accompanying unpleasant sensations during rest or inactivity **only occur or are worse in the evening or night** than during the day.

E. The occurrence of the above features are not solely accounted for as symptoms primary to another medical or a behavioral condition (e.g., myalgia, venous stasis, leg oedema, arthritis, leg cramps, positional discomfort, habitual foot tapping).

Restless legs syndrome can occur in the arms. It is also commonly associated with periodic limb movements of sleep (repetitive and involuntary limb movements over night occurring about every 20 - 40 seconds).

In all patients it is important to exclude secondary causes and advise them of lifestyle changes and ways to manage during an attack (see below in non-medical management).

If mild then an explanation and reassurance may be all that is necessary.

If moderate to severe, and symptoms have not improved with lifestyle changes, then medications can be considered.

Non-medical management of RLS

Assess for secondary RLS

- Pregnancy: drug treatment during pregnancy is not advised
- Iron deficiency anaemia: treat low ferritin with iron supplements even in absence of overt anaemia
- Renal failure, B12 and folate deficiency, hypothyroidism or hyperthyroidism
- Review medications for potential causes
  - » This is a common cause of worsening symptoms e.g. all antidepressants (especially SSRI and SNRI classes of antidepressants), antipsychotics, metoclopramide, prochlorperazine.
- RLS can sometimes be due to an underlying neurological condition such as a neuropathy or myelopathy. This is screened for with a neurological examination of limbs.

Lifestyle Changes

- Reducing or stopping alcohol, caffeine and nicotine
- Moderate regular exercise
- Good sleep hygiene

Symptom management

- Walking, stretching limbs, applying heat, relaxation exercises, distracting the mind, massaging the affected limbs.

Medical Management of RLS (for moderate to severe symptoms)

First line treatment is often with either a non-ergot dopamine agonist (pramipexole, ropinirole, or second-line with rotigotine), or an alpha-2-delta ligand (pregabalin or gabapentin — both off-label indications).

A weak opioid (such as codeine) is an alternative for people with painful symptoms, and can be used intermittently. Clinicians must take into account the risk of opioid dependence.

A short or intermittent course of a hypnotic drug (such as a "z-drug") may be considered for people with significant sleep disturbance due to RLS. See CKS topic on insomnia **https://cks.nice.org.uk/topics/insomnia/**

Further cautions regarding dopamine agonists

Although dopamine agonists may be effective treatment for RLS, patients should be aware of the risk of augmentation (an overall increase in severity of RLS symptoms with earlier onset of symptoms during the day, faster onset of symptoms when at rest, spreading of symptoms to the upper limbs), impulse control disorders (such as gambling, overeating, overbuying, sexual activities), and sleepiness which can sometimes be quite problematic, and more rarely sudden onset of sleep. These usually necessitate drug withdrawal. Patients should be monitored for these. Family members should preferably be informed of the risk of impulse control disorders as in some cases patients may not have insight to this occurring.

Referral

Recommended Actions Prior to Referral:

- Clinical history including age of onset, duration of symptoms, location and nature of symptoms, diurnal variation of symptoms, presence or absence of functional impairment.
- Neurological examination including assessment of muscle strength and tone, deep tendon reflexes, plantar responses, sensory examination including pinprick sensation and joint position sensation, balance and gait.
- Family history of neurological disease.
- Current and prior medications. Please withdraw or reduce dose of medications that may contribute to symptoms.
- History of alcohol use
- Blood tests including full blood count, ESR, ferritin, B12, folate, glucose, renal, liver, thyroid and bone parameters.

Who to Refer

Patients in whom there is diagnostic uncertainty.

Patients with treatment failure or significant side effects from treatment such as augmentation.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital)

Vetting Consultant

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Mild restless legs or those with moderate to severe restless legs controlled on treatment

The diagnosis of restless legs is usually made in primary care and a referral to specialist care is not required for confirmation in most cases unless there is significant diagnostic uncertainty.

Support Resources for Patients

NHS website on restless legs

www.nhs.uk/conditions/restless-legs-syndrome/

Restless Legs Charity

www.rls-uk.org/

Professional Guidelines for Clinicians

NICE guidelines

https://cks.nice.org.uk/restless-legs-syndrome

BMJ review article (2012)

https://www.bmj.com/content/344/bmj.e3056

Multiple Sclerosis (MS)

Definition

Multiple Sclerosis (MS) is an autoimmune demyelinating condition of the central nervous system, which can affect optic nerves, brain and the spinal cord. The typical age at diagnosis is in the third and fourth decade of life. Women are 2 to 3 times more affected than men.

MS is a progressive neurological condition and one of the commonest causes of disability in the young. The first attack of MS is termed "clinically isolated syndrome" (CIS). Each subsequent attack is termed a relapse. Such clearly defined relapses are characterised by either full recovery, or residual deficit. MS usually presents initially as a relapsing-remitting phenotype, which can subsequently transform into a secondary progressive phase. However, MS can also present in a primary progressive form in approximately 10% of people, and presents with progressive disease form onset.

An MS relapse is defined as following:

A patient with a known diagnosis of MS or CIS, who develops new or acutely worsening neurological symptoms which are:

A. Consistent with inflammation and demyelination and

B. Last more than 24 hours and are

C. Separated by at least 30 days from onset of last relapse and are

D. Not related to an infection, fever or other stresses and

E. Have no other explanation

Typical Treatments of Multiple Sclerosis

Disease Modifying Therapy (DMT)

In patients with a confirmed diagnosis of MS and significant illness, disease modifying therapy (DMTs) may be considered. These include oral, injectable and infusion therapies, with the choice of agent depending on the clinical characteristics and needs of an individual patient. This includes consideration of age, gender and comorbidity.

A wide and expanding range of DMTs are now available. They are all associated with the risk of significant adverse effects, in particular related to the modulation of the immune system. Therefore, patients on DMTs are counselled and monitored for the same by the MS service.

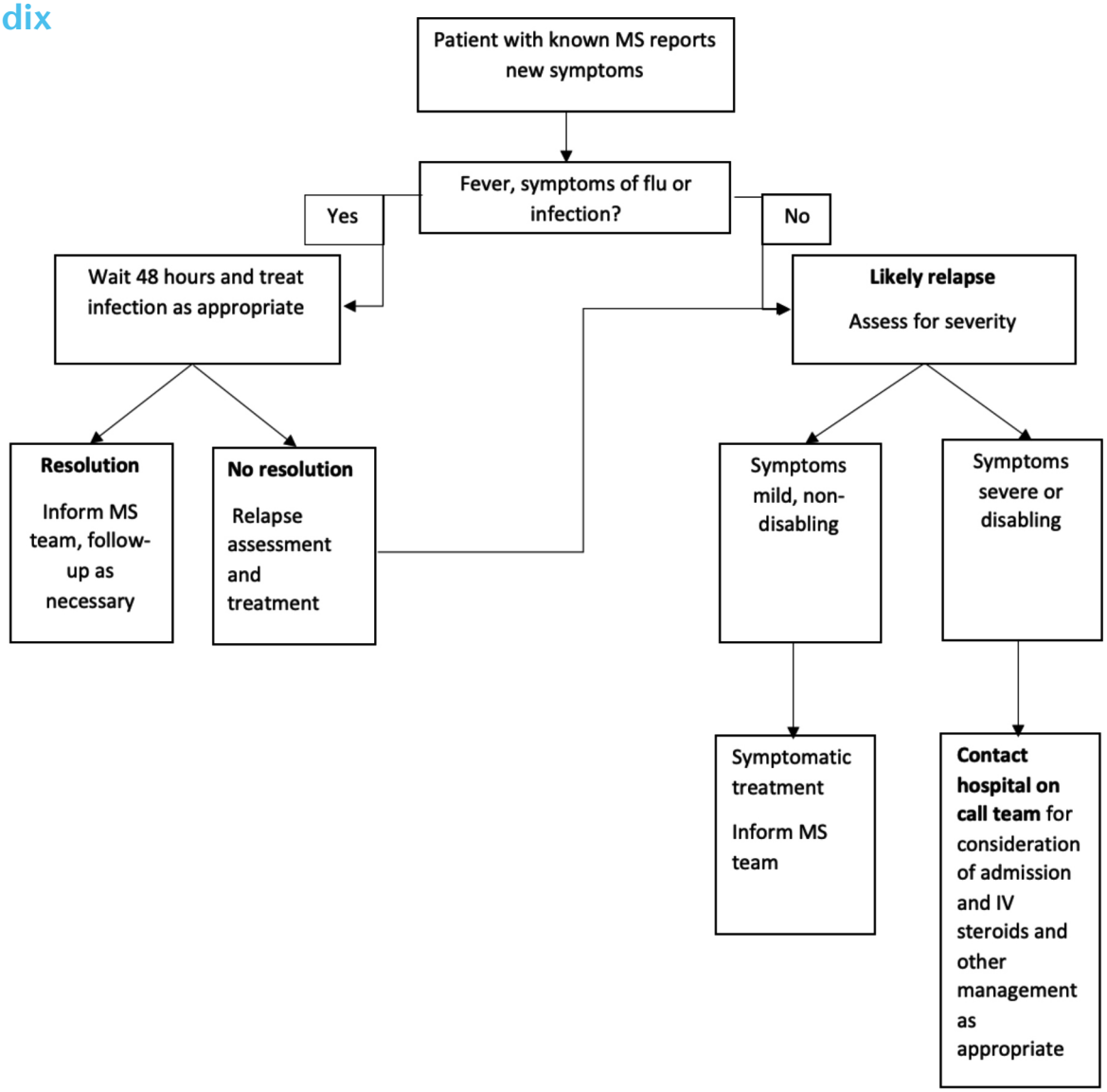
Access to the MS service for patients with milder side effects and queries regarding their medications is typically via the MS Clinical Nurse Specialist team at the Queen Elizabeth University Hospital.

Relapses

Acute attacks may be treated with steroids if symptoms are functionally disabling. Such treatment in selected patients is typically after advice by the on-call neurology team at the Queen Elizabeth University Hospital or the Multiple Sclerosis team via consultants or MS nurse specialists. Such steroid treatment is often given intravenously.

For patients with known MS in whom a relapse is suspected, the diagram below illustrates a suggested management pathway.

Appendix



Referral

Recommended Actions Prior to Referral:

- Clinical history including age of onset, nature and duration of symptoms, location of symptoms, motor, sensory and visual symptoms, functional impairment, gait and balance impairment.
- History of cognitive symptoms.
- History of disturbance of bladder and bowel habits.
- Family history of neurological disease.
- Current and prior medications.
- Neurological examination including assessment of eyes, muscle strength and tone, deep tendon reflexes, plantar responses, sensory examination including pinprick sensation and joint position sensation, balance and gait.
- Blood tests including full blood count, ESR, ferritin, B12, folate, glucose, renal, liver, thyroid and bone parameters.

Who to Refer

Suspected MS

All clinically stable patients with a suspected diagnosis of MS should be referred to a general neurology clinic for prompt review and investigations.

The urgency of the referral and mode of referral should be guided by the nature and severity of the neurological syndrome.

Patients with acute or significantly disabling symptoms may be discussed with the acute medical receiving teams and could be admitted to their local hospital via acute medical pathways if appropriate. For example, a spinal cord syndrome may require urgent referral to the Emergency Department and admission to hospital. A person with suspected optic neuritis can initially be referred to the eye casualty

For urgent queries, please discuss with the on-call neurology team at the Queen Elizabeth University Hospital.

Known MS (under active follow up of MS service)

In patients with a confirmed diagnosis of MS and under active follow please communicate with the named responsible MS consultant and the MS Clinical Nurse Specialist team at the Queen Elizabeth University Hospital.

Known MS (not under active follow up of MS service)

In patients with known MS not under active follow up, please refer to the MS service.

How to Refer

GPs: Please refer via SCI Gateway.

Others: (e.g. referrals from hospital)

Consultant Neurologist

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Support Resources for Patients

MS Trust

https://mstrust.org.uk

National MS Society

https://www.nationalmssociety.org

Professional Guidelines for Clinicians

NICE Guidelines

https://www.nice.org.uk/guidance/cg186

MS Trust

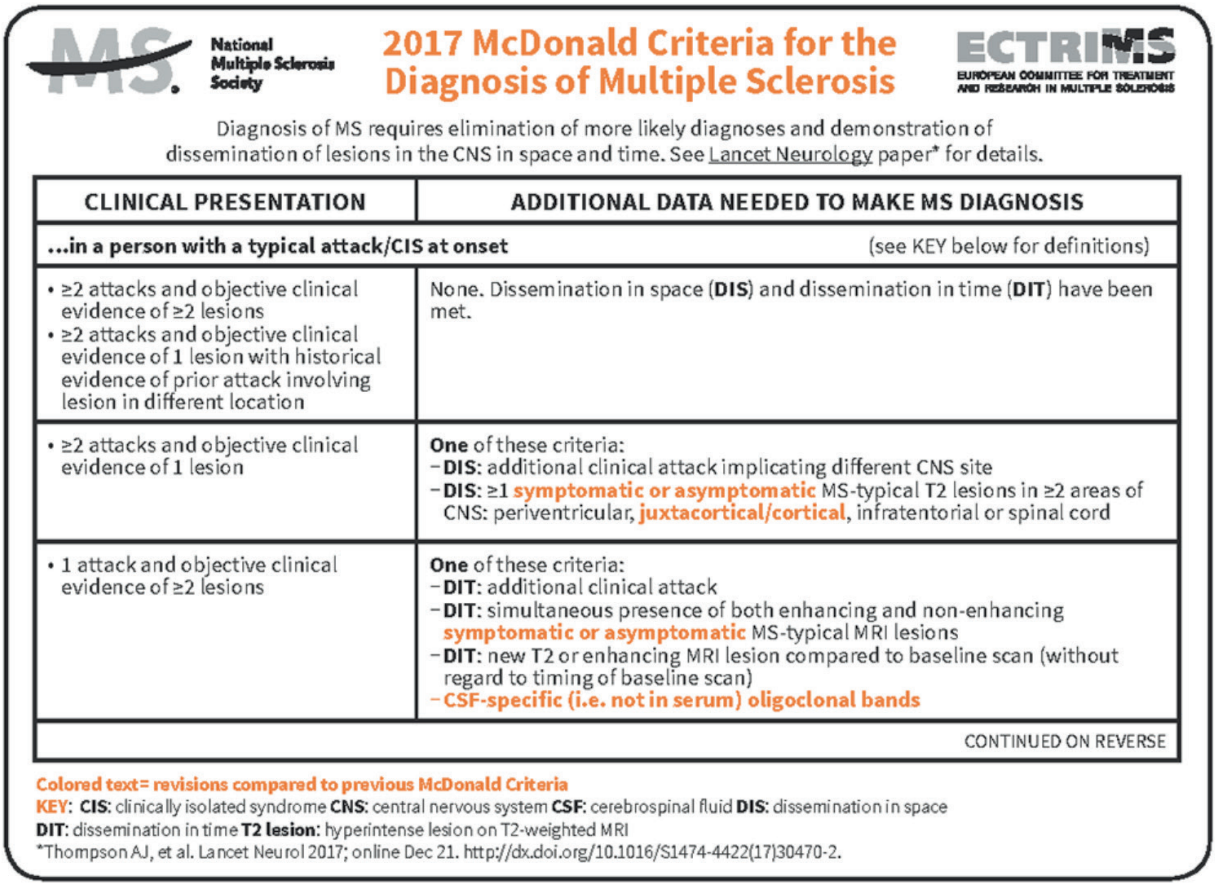
https://mstrust.org.uk

National MS Society

https://www.nationalmssociety.org/For-Professionals/Physicians

Appendix

The diagnostic criteria for MS are detailed below.



National Multiple Sclerosis Society

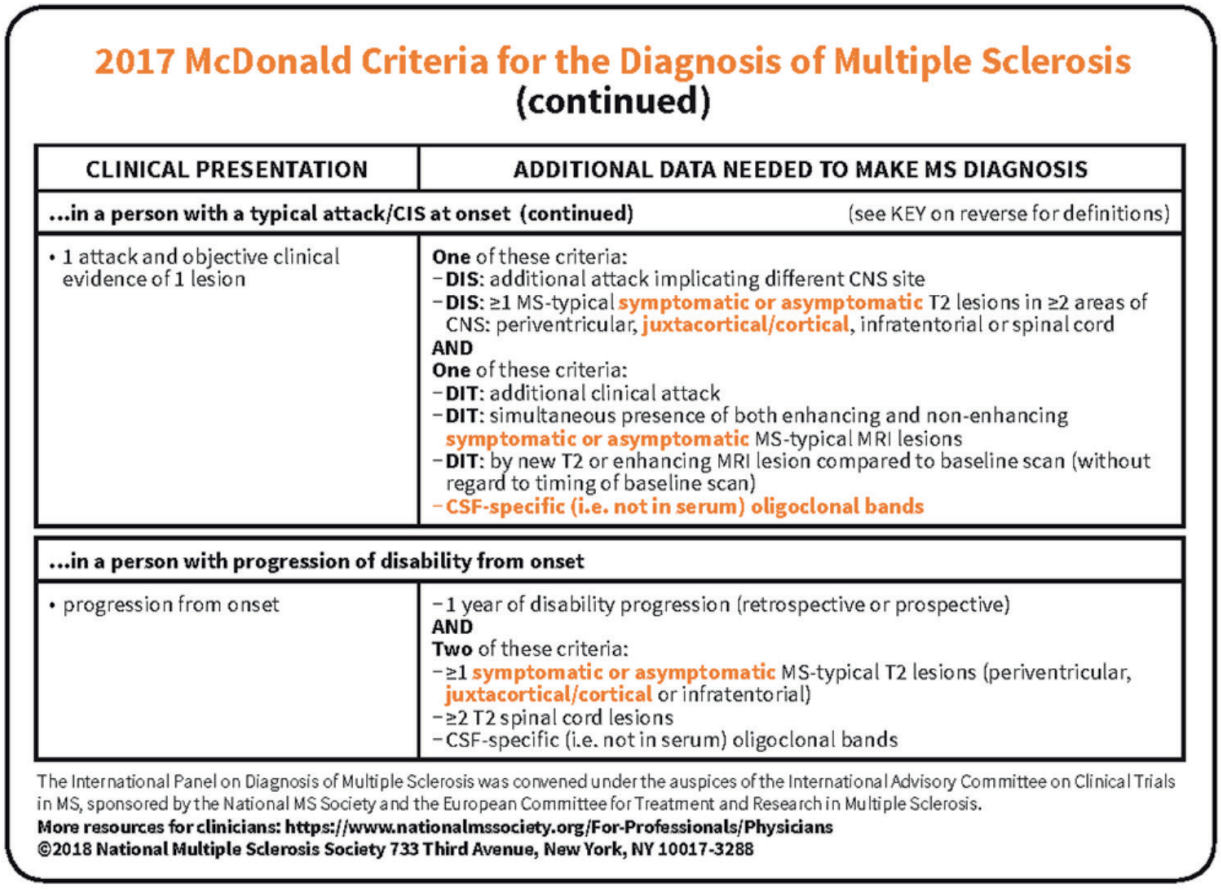
2017 McDonald Criteria for the Diagnosis of Multiple Sclerosis

ECTRIMS
EUROPEAN COMMITTEE FOR TREATMENT AND RESEARCH IN MULTIPLE SCLEROSIS

Diagnosis of MS requires elimination of more likely diagnoses and demonstration of dissemination of lesions in the CNS in space and time. See Lancet Neurology paper* for details.

CLINICAL PRESENTATION	ADDITIONAL DATA NEEDED TO MAKE MS DIAGNOSIS
...in a person with a typical attack/CIS at onset	(see KEY below for definitions)
• ≥2 attacks and objective clinical evidence of ≥2 lesions • ≥2 attacks and objective clinical evidence of 1 lesion with historical evidence of prior attack involving lesion in different location	None. Dissemination in space (**DIS**) and dissemination in time (**DIT**) have been met.
• ≥2 attacks and objective clinical evidence of 1 lesion	**One** of these criteria: – **DIS:** additional clinical attack implicating different CNS site – **DIS:** ≥1 **symptomatic or asymptomatic** MS-typical T2 lesions in ≥2 areas of CNS: periventricular, **juxtacortical/cortical**, infratentorial or spinal cord
• 1 attack and objective clinical evidence of ≥2 lesions	**One** of these criteria: – **DIT:** additional clinical attack – **DIT:** simultaneous presence of both enhancing and non-enhancing **symptomatic or asymptomatic** MS-typical MRI lesions – **DIT:** new T2 or enhancing MRI lesion compared to baseline scan (without regard to timing of baseline scan) – **CSF-specific (i.e. not in serum) oligoclonal bands**
	CONTINUED ON REVERSE

Colored text= revisions compared to previous McDonald Criteria
KEY: CIS: clinically isolated syndrome **CNS:** central nervous system **CSF:** cerebrospinal fluid **DIS:** dissemination in space **DIT:** dissemination in time **T2 lesion:** hyperintense lesion on T2-weighted MRI
*Thompson AJ, et al. Lancet Neurol 2017; online Dec 21. http://dx.doi.org/10.1016/S1474-4422(17)30470-2.



2017 McDonald Criteria for the Diagnosis of Multiple Sclerosis (continued)

CLINICAL PRESENTATION	ADDITIONAL DATA NEEDED TO MAKE MS DIAGNOSIS
...in a person with a typical attack/CIS at onset (continued)	(see KEY on reverse for definitions)
• 1 attack and objective clinical evidence of 1 lesion	**One** of these criteria: – **DIS:** additional attack implicating different CNS site – **DIS:** ≥1 MS-typical **symptomatic or asymptomatic** T2 lesions in ≥2 areas of CNS: periventricular, **juxtacortical/cortical**, infratentorial or spinal cord **AND** **One** of these criteria: – **DIT:** additional clinical attack – **DIT:** simultaneous presence of both enhancing and non-enhancing **symptomatic or asymptomatic** MS-typical MRI lesions – **DIT:** by new T2 or enhancing MRI lesion compared to baseline scan (without regard to timing of baseline scan) – **CSF-specific (i.e. not in serum) oligoclonal bands**
...in a person with progression of disability from onset	
• progression from onset	– 1 year of disability progression (retrospective or prospective) **AND** **Two** of these criteria: – ≥1 **symptomatic or asymptomatic** MS-typical T2 lesions (periventricular, **juxtacortical/cortical** or infratentorial) – ≥2 T2 spinal cord lesions – CSF-specific (i.e. not in serum) oligoclonal bands

The International Panel on Diagnosis of Multiple Sclerosis was convened under the auspices of the International Advisory Committee on Clinical Trials in MS, sponsored by the National MS Society and the European Committee for Treatment and Research in Multiple Sclerosis.
More resources for clinicians: https://www.nationalmssociety.org/For-Professionals/Physicians

From:

https://www.nationalmssociety.org/For-Professionals/Clinical-Care/Diagnosing-MS/Diagnosing-Criteria

Muscle Disorders

Definition

Muscle disorders are a very **heterogenous group of conditions.** In the clinic we see a wide range of muscle conditions including genetic and acquired muscle disorders.

Examples of **acquired muscle disorders** include the inflammatory myopathies, e.g. dermatomyositis, necrotising myositis secondary to statins or cancer. Treatments for these conditions include immunosuppressive therapies. In contrast, some other acquired muscle disorders such as sporadic inclusion body myositis do not respond to immunosuppression. Myopathies may also develop secondary other medical conditions e.g. endocrine disease.

Genetic **muscle disorders** include muscular dystrophies, congenital myopathies, myotonic dystrophy, mitochondrial disorders, myofibrillar myopathies and channelopathies (including myotonia congenita and periodic paralysis). Some genetic muscle conditions only become apparent later in life, for example myofibrillar myopathies and facioscapulohumeral muscular dystrophy. Emerging therapies for different genetic muscle disorders underscores the importance of genotyping.

Patients with **rhabdomyolysis**, either as a single or a recurrent event, are often assessed in the muscle clinic. Similarly patients with **metabolic myopathies** presenting with exercise intolerance are investigated for a genetic basis contributing to their phenotype.

Involvement of cardiac and respiratory muscles varies depending on the specific muscle condition. When the disorder is not genetically characterised, we advise surveillance of both systems at least for a period of time. The importance of physical therapy, adequate nutrition and psychosocial wellbeing remains crucial in the management of these disorders.

Referral

Recommended Actions Prior to Referral:

- Clinical history of age of onset, duration of symptoms, nature and location of symptoms, motor and sensory symptoms, functional impairment.
- History of balance and gait impairment.
- Presence of difficulties with swallow and speech.
- Family history of neurological illness.
- Prior medical history including metabolic illness, cancer and cancer treatments.
- Please review current medications and consider if they may be contributing to symptoms e.g. statins.
- Ask about cardiac symptoms including palpitations, syncope, presyncope and exercise intolerance.
- Ask about respiratory symptoms including orthopnoea and daytime hypersomnolence.
- Neurological examination including assessment of eyes, muscle strength and tone, deep tendon reflexes, plantar responses, sensory examination including pinprick sensation and joint position sensation, balance and gait.
- Blood tests including full blood count, ESR, ferritin, B12, folate, glucose, renal, liver, thyroid and bone parameters.
- Please perform ECG.

Who to Refer

- Patients who have had rhabdomyolysis, have persistent weakness and may have red flags on examination or past medical history.
- Patients with a history of exercise intolerance and muscle symptoms suggestive of a problem secondary to energy production (a metabolic myopathy)
- Patients developing new-onset muscle weakness (proximal or distal or both, with or without facial and/or axial involvement).
- Patients who have evidence of respiratory muscle involvement, and where an underlying myopathy requiring further investigation is a possibility.
- Patients with familial cardiomyopathy where a generalised myopathic disorder is being questioned.
- Patients with longstanding muscle weakness, which is likely to be genetic and where a definite molecular diagnosis has not yet been reached.
- Patients attending the paediatric service with a known muscle disorder which requires transitioning to adult services (in conjunction with paediatric neurologists)
- Patients with episodic muscle weakness characteristic of a channel dysfunction.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital)

Vetting Consultant

Muscle clinic

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Patients with a defined muscle diagnosis where investigations have been completed and who may have progressed as a result of the natural history of their condition but no active therapies are available and travel to hospital for reassessment is unlikely to add further. It would be useful to discuss the case with the consultant neurologist to consider if referrals to other services, e.g. rehabilitation medicine, occupational therapy, WESTMARC, may be useful.

Support Resources for Patients

Muscular Dystrophy UK

https://www.musculardystrophyuk.org/

Scottish Muscle Network

https://www.smn.scot.nhs.uk/

Muscular Dystrophy Association

https://www.mda.org/

Treat NMD Neuromuscular Network including the Fascioscapulohumeral Muscular Dystrophy UK Patient Registry

https://treat-nmd.org/

https://www.fshd-registry.org.uk/

Spinal Muscular Atrophy UK

https://smauk.org.uk/

Duchenne UK

https://www.duchenneuk.org/

National Organization for Rare Disorders (USA)

https://rarediseases.org/

Orphanet ("The portal for rare diseases and orphan drugs")

https://www.orpha.net/consor/cgi-bin/index.php

Professional Guidelines for Clinicians/Resources

Scottish Muscle Network

https://www.smn.scot.nhs.uk/

A useful website for professionals from the Neuromuscular Disease Centre, Washington, St Louis, USA

https://neuromuscular.wustl.edu/

National Organization for Rare Disorders (USA)

https://rarediseases.org/

Orphanet ("The portal for rare diseases and orphan drugs")

https://www.orpha.net/consor/cgi-bin/index.php

Muscular Dystrophy UK

http://www.musculardystrophyuk.org/wp-content/uploads/2015/04/The-diagnosis-and-management-of-duchenne-muscular-dystrophy.pdf

Myasthenia Gravis

Definition

Myasthenia Gravis (MG) is an autoimmune neuromuscular junction disorder characterised by fatiguable weakness.

Most patients with MG present initially with ptosis, which is fatiguable and variable, and usually asymmetrical. Patients may develop blurred vision with or without diplopia. Sometimes patients are not aware of double vision until they are examined.

Most patients' symptoms evolve within 6 months of onset of ocular symptoms. They may extend to involve other muscles including facial, neck and axial muscles, bulbar muscles with symptoms of dysarthria, dysphagia and chewing fatigability, and limb weakness (arms more than legs).

Myasthenic crisis with respiratory muscle involvement is rare. It is more likely to occur in patients who are evolving rapidly, particularly at presentation. It may also occur in patients who are undertreated, or in patients where clinical evolution is quicker than anticipated out-pacing treatment titration.

Most patients with MG can be initiated on treatment in the community. However, inpatient management is considered in patients with bulbar symptoms, especially in older patients with speech or swallow symptoms and in patients with significant head drop.

Typical Treatments

Treatments are initiated under the supervision of specialist neurology services.

Symptom control

- Pyridostigmine (oral)
- Neostigmine (subcutaneous) only in special circumstances

Induction of remission

- Corticosteroids in dose discussed with neurology
- Steroid-sparing immunosuppression in some patients e.g. Azathioprine, Mycophenolate mofetil, Methotrexate, Ciclosporin and Tacrolimus.

Rescue treatments for myasthenic crises

In inpatients with care discussed with neurology

- Intravenous immunoglobulin
- Plasmapheresis
- Rituximab

Treatment Resistant Disease

- Rituximab

Referral

Recommended Actions Prior to Referral from Primary Care

- If patient has new symptoms and is unwell, please discuss with the on-call neurology team at the Queen Elizabeth University Hospital.
- If patient is clinically stable and well, please refer to the outpatient Myasthenia Gravis clinic
- Please request anti-acetylcholine receptor antibodies, thyroid function, creatine kinase, renal, liver and haematological indices, diabetes mellitus screen.
- Please monitor patient whilst waiting for the neurology outpatient clinic appointment.

Recommended Actions Prior to Referral from Hospital

- If patient has clinical features of myasthenia gravis, please discuss with the on-call neurology team at the Queen Elizabeth University Hospital.
- Please request anti-acetylcholine receptor antibodies.
- Please request CT scan of the chest and mediastinum to look for thymoma.
- Please consider MRI brain scan to assess brainstem structures and to rule out alternative pathology in brainstem and orbits.
- Please monitor respiratory and bulbar function.

Who to Refer

- All patients with clinical features of Myasthenia Gravis.
- All patients with symptoms or signs of ocular Myasthenia Gravis with or without anti-acetylcholine receptor antibodies
- All patients with anti-acetylcholine receptor antibodies.
- Patients with thymoma and positive acetylcholine receptor antibodies, with or without weakness.

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital)

Vetting Consultant

Myasthenia Clinic

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Support Resources for Patients

Myaware – this is the only UK-based charity for myasthenia gravis

https://www.myaware.org/

Myasthenia Gravis Foundation of America - offers useful resources including podcasts for patients.

https://myasthenia.org/

Consider giving the patient an information leaflet which can be downloaded from the NHS GGC website below

http://www.staffnet.ggc.scot.nhs.uk/Acute/Regional%20Services/Neuro-sciences%20Institute/Neurology/Documents/Patient%20Information%20Leaflet%20on%20Myasthenia%20Gravis.pdf

http://www.staffnet.ggc.scot.nhs.uk/acute/regional%20services/neuro-sciences%20institute/neurology/Documents/Forms/AllItems.aspx

Scottish Muscle Network (Fatigue Management)

https://www.smn.scot.nhs.uk/wp-content/uploads/2018/11/Fatigue_MG_PIL-MI-version.pdf

Professional Guidelines for Clinicians

Information on Pyridostigmine

http://www.staffnet.ggc.scot.nhs.uk/Acute/Regional%20Services/Neuro-sciences%20Institute/Neurology/Documents/Pyridostigmine.pdf

Information on Myasthenia gravis

http://www.staffnet.ggc.scot.nhs.uk/Acute/Regional%20Services/Neuro-sciences%20Institute/Neurology/Documents/Patient%20Information%20Leaflet%20on%20Myasthenia%20Gravis.pdf

Medicines that can affect myasthenia gravis

http://www.staffnet.ggc.scot.nhs.uk/Acute/Regional%20Services/Neuro-sciences%20Institute/Neurology/Documents/Myasthenia%20Gravis%20or%20Lambert-Eaton%20Myasthenia%20Syndrome.pdf

This includes information on all treatments prescribed in MG

http://www.staffnet.ggc.scot.nhs.uk/acute/regional%20services/neuro-sciences%20institute/neurology/Documents/Forms/AllItems.aspx

Treatment guidelines

https://pn.bmj.com/content/practneurol/15/3/199.full.pdf

https://n.neurology.org/content/neurology/87/4/419.full.pdf

Treatment approaches used in West of Scotland Myasthenia Service

https://www.frontiersin.org/articles/10.3389/fneur.2020.00604/full?&utm_source=Email_to_authors_&utm_medium=Email&utm_content=T1_11.5e1_author&utm_campaign=Email_publication&field=&journalName=Frontiers_in_Neurology&id=552451

Neuropathy

Definition

Peripheral neuropathies encompass disorders of the peripheral nerve cells and fibres, which manifest secondary to a wide range of pathology.

Typical Treatments

Treatment depends on the aetiology of the peripheral neuropathy.

In patient with neuropathy due to diabetes mellitus or impaired glucose control, lifestyle measures e.g. weight loss, and strict diabetic control are advised.

In some neuropathies with an inflammatory basis, immune modulating and immunosuppressive treatments may be indicated.

Symptomatic treatments are sometimes used in patients with troublesome painful paraesthesias.

- First line agents include Amitriptyline and Gabapentin
- Pregabalin and Duloxetine are further options in patients fail on first line therapies.
- Topical agents such as a menthol cream, e.g. Dermacool or Capsaicin, may help in patients with localised neuropathic pain.

Referral

Recommended Actions Prior to Referral:

- Clinical history of onset and pace of evolution, symmetrical or asymmetrical symptoms, motor and sensory symptoms, presence or absence of bladder and bowel symptoms.
- History including diet and nutritional status, recent weight loss, smoking, alcohol use, current and prior medications, history of cancer and cancer treatments.
- Family history of neurological illness.
- Neurological motor examination to document presence and pattern of weakness, e.g. proximal or distal weakness, muscle wasting, presence or absence of deep tendon reflexes, plantar responses.
- Sensory examination including joint position sensation and ataxia.
- Blood tests including full blood count, ESR, CRP, B12, folate, renal, liver and thyroid parameters, creatine kinase, HbA1c.
- Myeloma screen including serum electrophoresis and immune fixation, serum light chains and urine for Bence Jones protein.
- Consider a glucose tolerance test if you suspect metabolic syndrome or diabetes.

Who to Refer

Patients developing weakness.

New onset sensory ataxia.

Neurological examination consistent with neuropathy.

In patients with rapidly evolving or significant motor weakness, please consider hospital admission via local acute medical admission pathways.

Please note alternate referral pathways for Entrapment Neuropathies

Patients with suspected or confirmed Carpal Tunnel Syndrome or Ulnar Neuropathy – please refer to Trauma and Orthopaedics – Wrist and Hand Service

Patients with other suspected nerve entrapment syndromes – please refer to Trauma and Orthopaedics

Patients with traumatic or post-traumatic neuropathy – please refer to Trauma and Orthopaedics

Patients with suspected or confirmed nerve root entrapment and cervical or lumbo-sacral degenerative disease – please refer to Trauma and Orthopaedics or Neurosurgery

How to Refer

GPs: please refer via SCI Gateway

Others: (e.g. referrals from hospital)

Vetting Consultant

Neuropathy Clinic

Department of Neurology

Institute of Neurological Sciences

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Examples of Patients Who May Be Managed in Primary Care / Unlikely to Benefit from Specialist Referral

Patients with a previously confirmed genetic neuropathy e.g. Charcot Marie Tooth Disease, with clinical progression due to natural history of the neuropathy, but no available active therapies. Travel to hospital for reassessment is unlikely to add further in such patients. It could be useful to discuss the case with the responsible consultant in case referrals to other services may be helpful e.g. rehabilitation medicine, physiotherapy, WESTMARC, occupational therapy.

Genetic Testing of Families with Inherited Neuropathies

Familial or cascade testing of relatives of patients with confirmed genetic neuropathies is best performed by Clinical Genetics. Please refer directly to:

Vetting Consultant

Department of Clinical Genetics

Queen Elizabeth University Hospital

1345 Govan Road

Glasgow G51 4TF

Diabetic neuropathy

Patients with a history of diabetes mellitus with gradual onset symmetrical distal sensory neuropathy do not typically need further assessment in neurology. Blood tests as recommended above may be useful.

Support Resources for Patients

Scottish Muscle Network

https://www.smn.scot.nhs.uk/

Guillain Barre CIDP Foundation

https://www.gbs-cidp.org/

Charcot-Marie Tooth UK

https://cmt.org.uk/

Diabetes UK

https://www.diabetes.org.uk

Professional Guidelines for Clinicians

A useful website for professionals from the Neuromuscular Disease Centre, Washington, St Louis, USA

https://neuromuscular.wustle.edu/

Chronic Non Malignant Pain Neuropathic Pain Guideline accessed via staffnet:

http://www.staffnet.ggc.scot.nhs.uk/Info%20Centre/PoliciesProcedures/GGCClinicalGuidelines/GGC%20Clinical%20Guidelines%20Electronic%20Resource%20Direct/Chronic%20Nonmalignant%20Pain%20Neuropathic%20Pain%20Guideline.pdf