# MYALGIC ENCEPHALOMYELITIS

## CHRONIC FATIGUE SYNDROME

## FROM HARM TO HELP

DR CATHY STEPHENSON

# MYALGIC ENCEPHALOMYELITIS

**M.E. Myalgic Encephalomyelitis**
***also known as Chronic Fatigue Syndrome (CFS)***
***Tapanui Flu, or ME/CFS***

ME/CFS is a serious, chronic, complex and multisystem disease that frequently and dramatically limits the activities of affected patients.
(National Academy of Medicine 2015)

Classified in ICD 10 as a neurological condition

# THE DOCTOR'S EXPERIENCE

- **Medical training has little/no ME/CFS specific information**
- **Multisystem and highly variable condition (both within and between patients)**
- **No diagnostic test and no clear agreement about clinical criteria**
- **Multiple contributing, co-occurring and confounding conditions**
- **Medical system discourages integrating knowledge**
- **Autonomic dysregulation may be mistaken for anxiety**

***Doctors are perplexed. "If it can't be measured and I don't understand it, it must be mental health". ME/CFS is PUS (psychiatrically unexplained symptoms)***

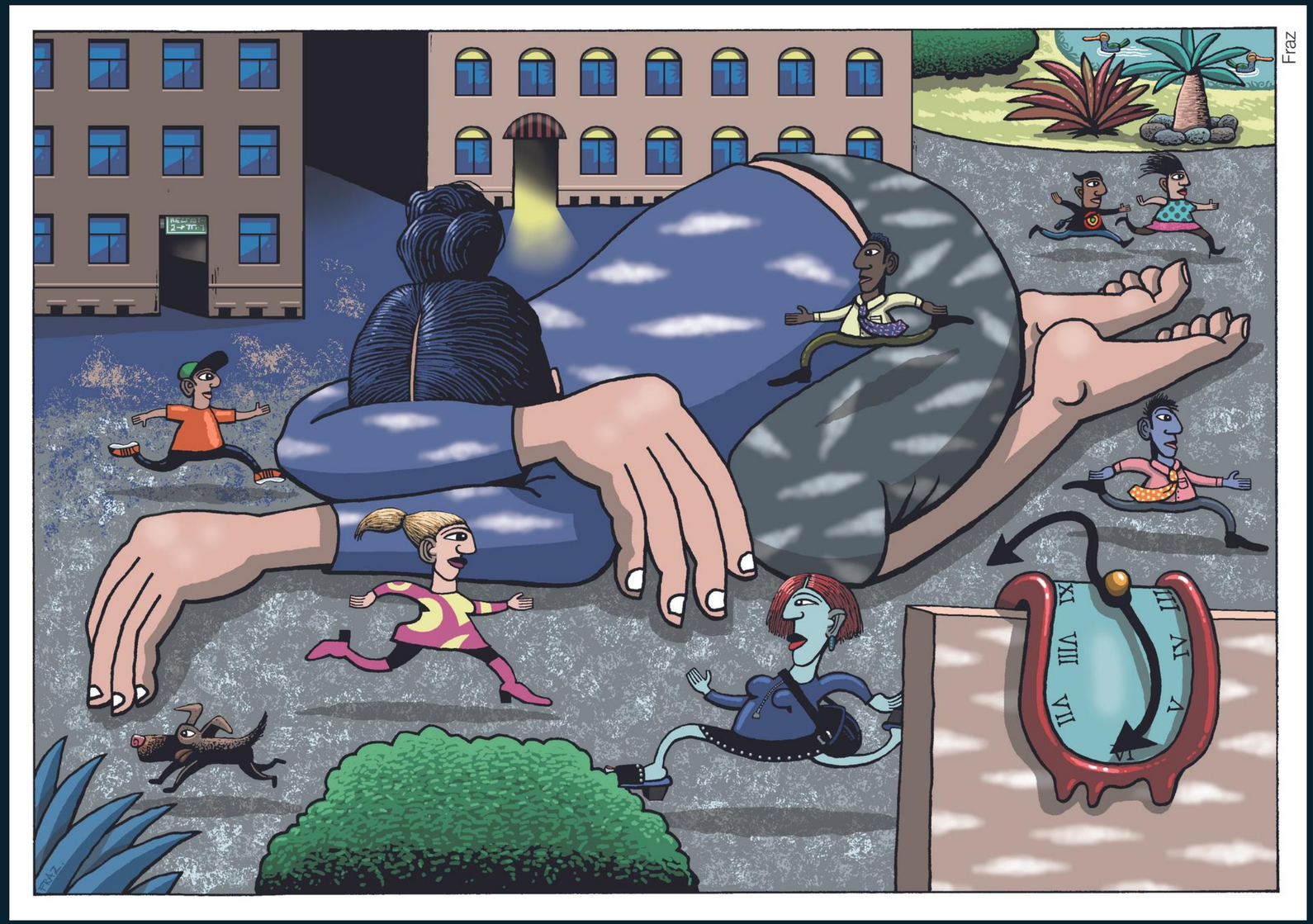
New Zealand Doctor Rata Aotearoa +Educate

# HOW TO TREAT ME/CFS

CME

+E DOCTOR

1 CR



# A PARADIGM SHIFT

**1969.**
**ME included (and remains) in the ICD as Neurological disease.**

**1988.**
**CDC renames ME as Chronic Fatigue Syndrome.**
**It becomes a 'syndrome of fatigue'.**
**Psychological models proliferate.**

**2011.**
**PACE trial erroneously concludes Graded Exercise and Cognitive Behavioural Therapy may be curative.**

**Recommendations included in all major guidelines.**

**2011 to current.**
**PACE trial acknowledged as methodologically flawed.**

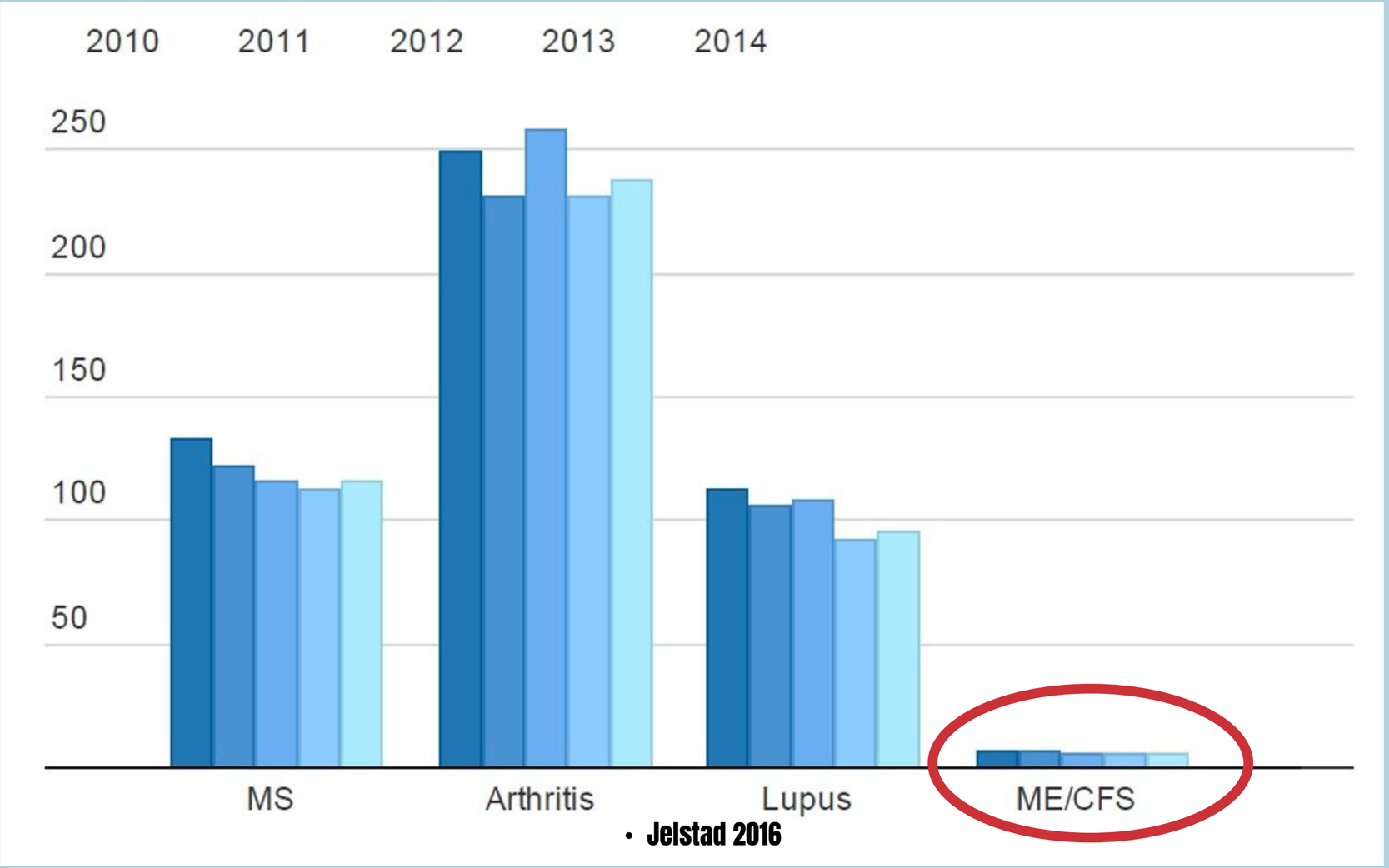
**CDC apologises.**

**2021 NICE guidelines redrafted.**

**Graded Exercise and Cognitive Behavioural Therapy are NO LONGER RECOMMENDED for ME and have been shown to be harmful.**
**No evidence exists for psychological models of ME.**

# NIH RESEARCH FUNDING 2010-2014

## One Year of Funding for MS would Fund 23 Years of Research into ME




- Jelstad 2016

# MISINFORMATION HAS DERAILED RESEARCH AND IMPACTED CARE OF PATIENTS

Test Results

Help

**Submission Date/Time:** 02/06/2020 12:42:43

**Status:** Declined/Reviewed

**Status Reason:** Declined - Other reason in notes

**Notes to referrer:** [redacted]: Patient has been well investigated and has a management plan - what input do you want . We avoid seeing patients like this because they block new patients and we have nothing to add . [redacted]

**Priority:**

**Speciality:** General Medicine

**Clinic Name:**

**Organisation Name:** [redacted]

Test Results

Help

NHI No: [redacted]

**NHI** [redacted]

**Submission Date/Time:** 06/05/2021 10:30:11

**Status:** Declined/Reviewed

**Status Reason:** Declined - Other reason in notes

**Notes to referrer:** [redacted] Chronic fatigue syndrome in 16 year old is bad news and needs to be addressed urgently and positively by Psych . [redacted]

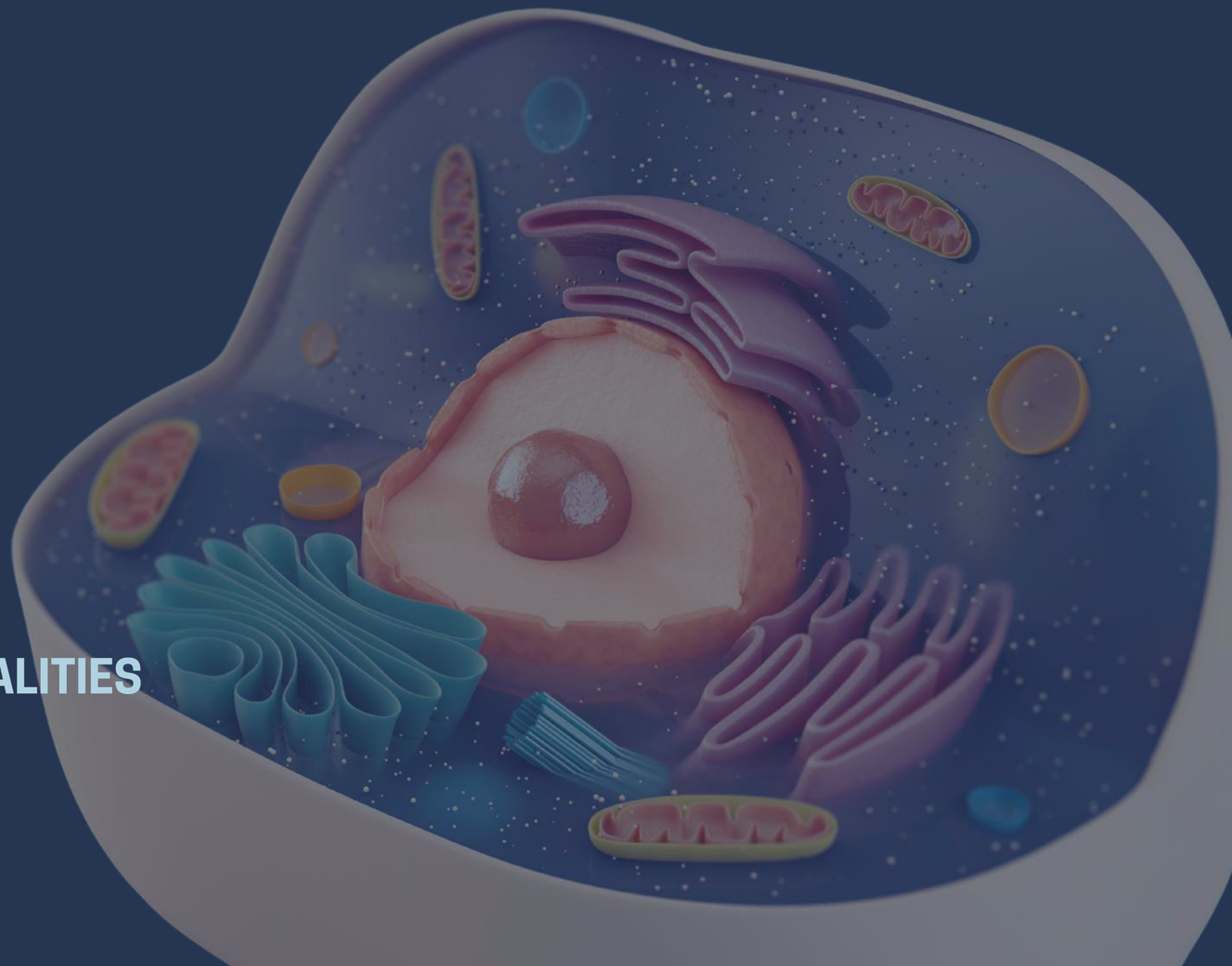
**Priority:**

**Speciality:** General Medicine

# PATHOPHYSIOLOGY

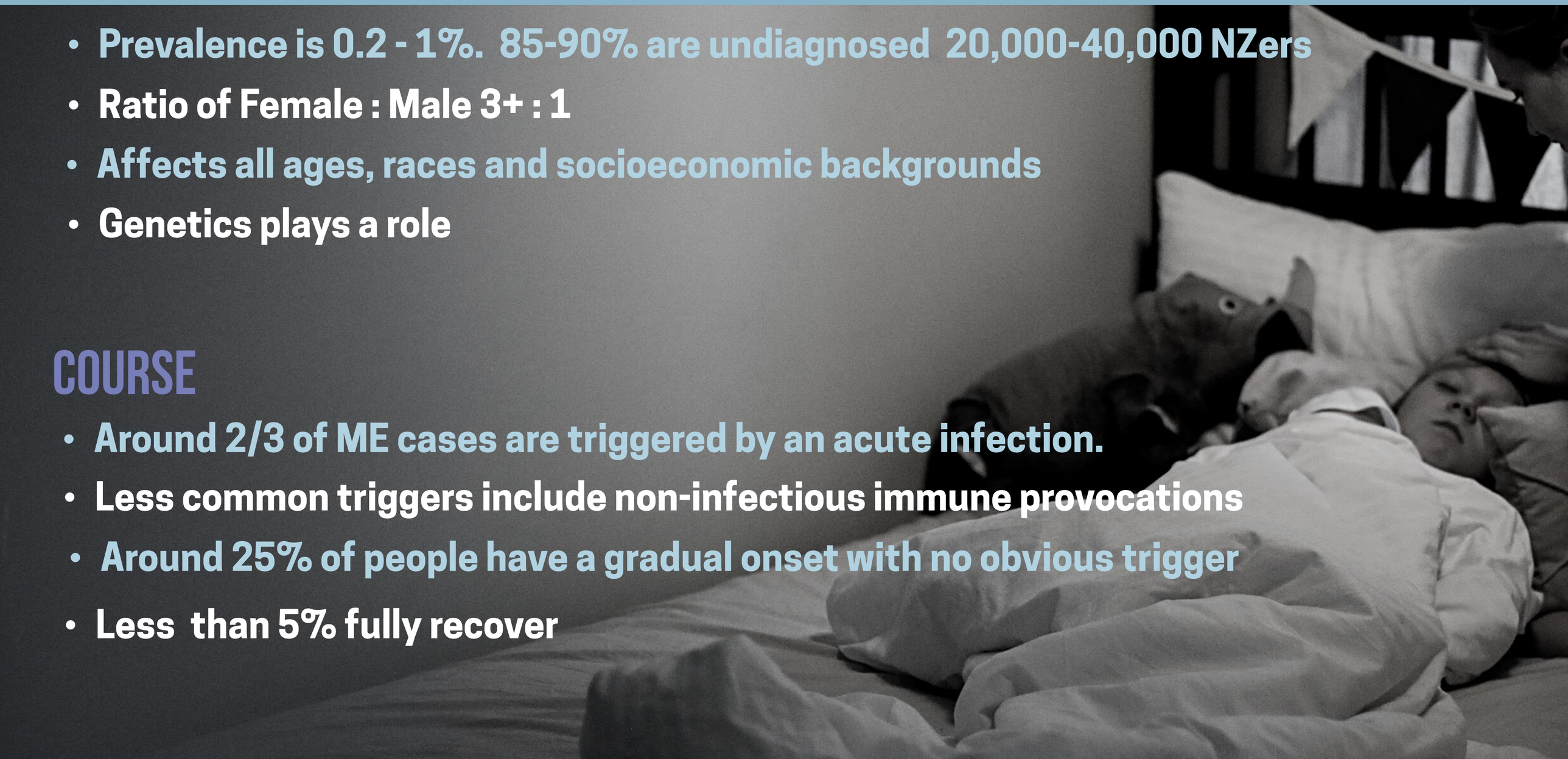
## RESEARCH STUDIES DESCRIBE PATHOPHYSIOLOGICAL CHANGES

- IMMUNE SYSTEM ABNORMALITIES
- CELLULAR METABOLISM ABNORMALITIES
- NEUROENDOCRINE DISTURBANCES
- BLOOD PRESSURE and HEART RATE ABNORMALITIES



# EPIDEMIOLOGY

- Prevalence is 0.2 - 1%.  85-90% are undiagnosed  20,000-40,000 NZers
- Ratio of Female : Male 3+ : 1
- Affects all ages, races and socioeconomic backgrounds
- Genetics plays a role



## COURSE

- Around 2/3 of ME cases are triggered by an acute infection.
- Less common triggers include non-infectious immune provocations
- Around 25% of people have a gradual onset with no obvious trigger
- Less  than 5% fully recover

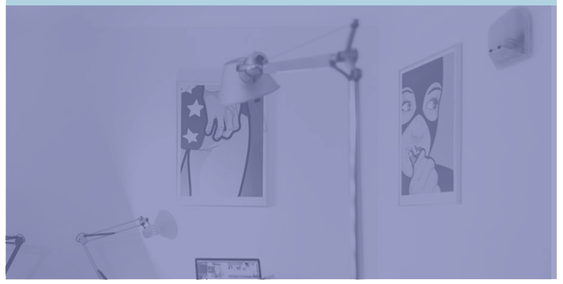
# SPECTRUM OF SEVERITY

**ME is a Spectrum Disorder from MILD to VERY SEVERE**

**‘Mild' cases involve the loss of at least 50% of normal function**

## MILD

**May work but only at expense of other areas of life.**



## MODERATE

**Mostly housebound**
**May use walking aid**

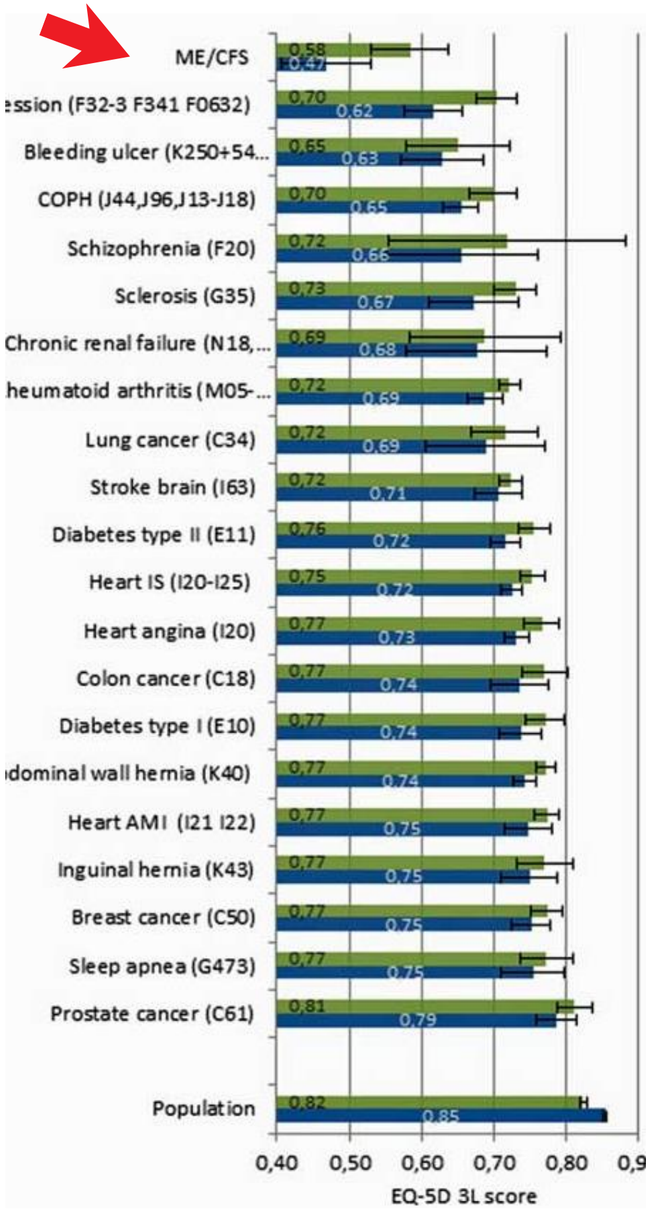
## SEVERE

**Mostly bed-bound.**
**May rely on wheelchair & carers.**

## VERY SEVERE

**Fully bed-bound.**
**Often can't speak.**
**Untreatable pain.**
**Often require nutritional support.**

# QUALITY OF LIFE




- **People with ME are often more functionally impaired than people with type 2 diabetes, multiple sclerosis, congestive heart failure and end stage kidney disease.**

- **They suffer a lower quality of life than people with cancer, stroke, kidney failure and schizophrenia.**

HVIDBERG ET AL 2015

# SYMPTOM PRESENTATION

FLU-LIKE SYMPTOMS
REDUCED FUNCTIONALITY
DYSPNEA PAIN MUSCLE PAIN
MUSCLE FATIGABILITY & WEAKNESS
TEMPERATURE DYSREGULATION VERTIGO
POST-EXERTIONAL MALAISE PHOTOPHOBIA
HYPERSOMNIA MCAS CHEMICAL SENSITIVITES
SWOLLEN LYMPH NODES ANAEMIA CHEST PAIN
GASTROINTESTIONAL DYSFUNCTION HYPERACUSIS
SLOWED SPEECH INSOMNIA PROFOUND FATIGUE
pOSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME
HEADACHES MOOD SWINGS FOOD INTOLERANCES
JOINT PAIN UNREFRESHING SLEEP DEPRESSION
VITAMIN D DEFICIENCY ALCOHOL INTOLERANCE
WEIGHT GAIN/LOSS ANXIETY HYPOGLYCAEMIA
POOR COORDINATION JOINT HYPERMOBILITY
COGNITIVE IMPAIRMENT BLURRED VISION
MIGRAINES SYNCOPE CCI ALLODYNIA
ORTHOSTATIC INTOLERANCE
TACHYCARDIA DYSPHASIA

# DIAGNOSTIC CRITERIA

FLU-LIKE SYMPTOMS

**REDUCED FUNCTIONALITY**

DYSPNEA PAIN MUSCLE PAIN

MUSCLE FATIGABILITY & WEAKNESS

TEMPERATURE DYSREGULATION VERTIGO

**POST-EXERTIONAL MALAISE** PHOTOPHOBIA

HYPERSOMNIA MCAS CHEMICAL SENSITIVITES

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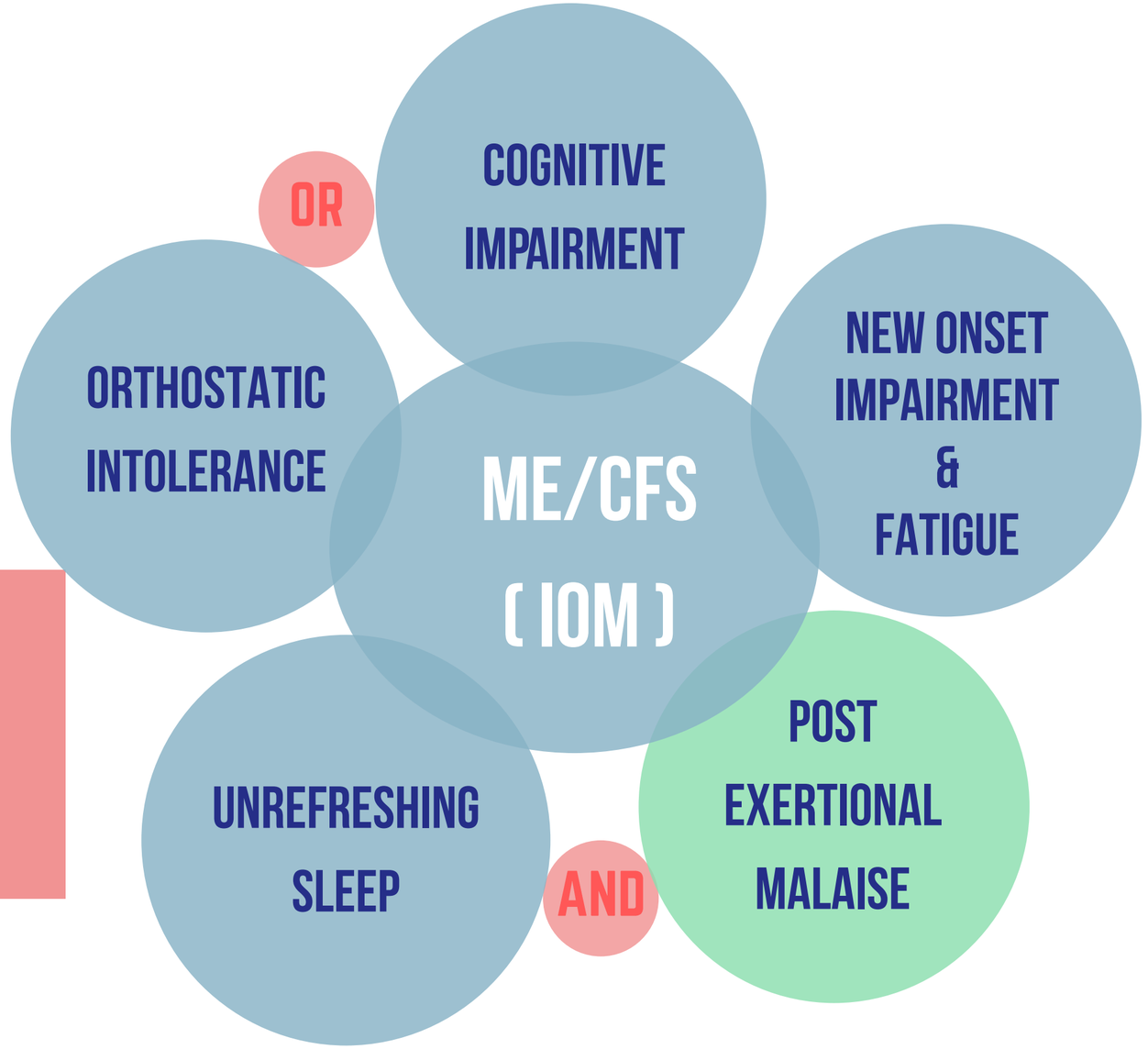
**ORTHOSTATIC INTOLERANCE**

TACHYCARDIA DYSPHASIA

# DIAGNOSIS




**Sx present**
**6 months**
**at least half the time**
**moderate-severe intensity**

**ASSESS**
**DIAGNOSE**
**INVESTIGATE**
**DIFFERENTIALS**
**REFER**

**Institute of Medicine (IOM/NAM) Criteria for ME/CFS 2015.**

# ROLE OF THE GENERAL PRACTITIONER

# WHAT IS POST-EXERTIONAL MALAISE (PEM)?

**It is defined by a worsening of symptoms (not just fatigue) in the period following exertion.**

**Is not the same as post-exertional fatigue or exercise intolerance.**

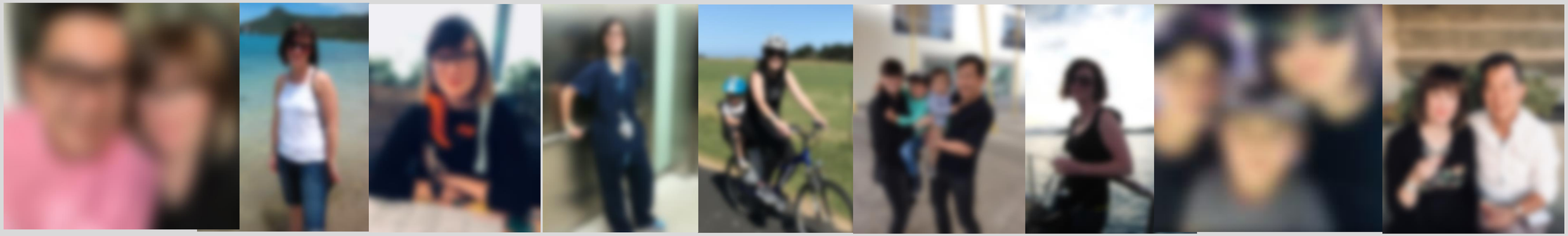
## POST- EXERTIONAL MALAISE (PEM)

- Is Unique to ME
- Is Required for a Diagnosis of ME
- Is Key to Distinguish ME From Other Diseases
- Is Delayed 24 - 72 hrs After Exertion
- May Last Days, Weeks or Much Longer
- Can Result in a Lowered Baseline

**Exercise physiology research has revealed the pathology of PEM**

- **Marked abnormalities on two day cardiopulmonary exercise testing.**
- **Characteristic deterioration in exercise capacity on the second day.**
- **ME is not caused by deconditioning**

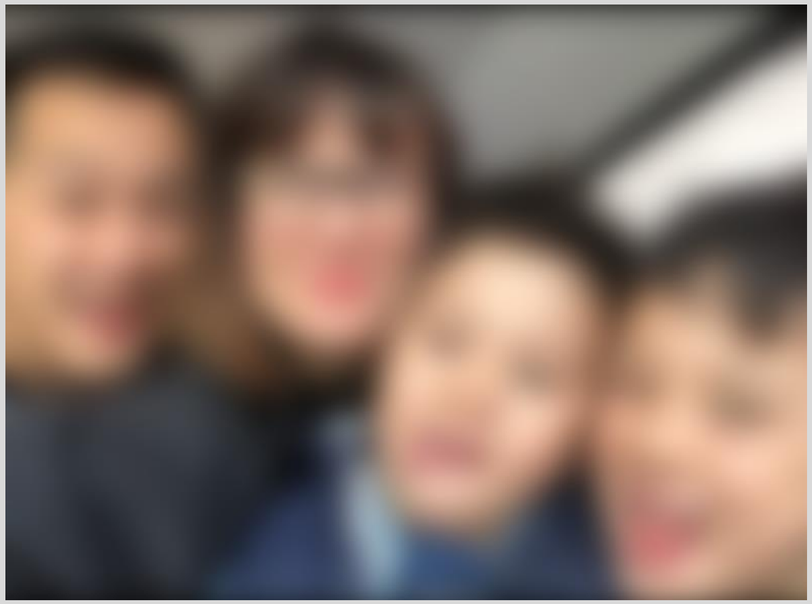
# PRE-ILLNESS LEVEL OF FUNCTIONING

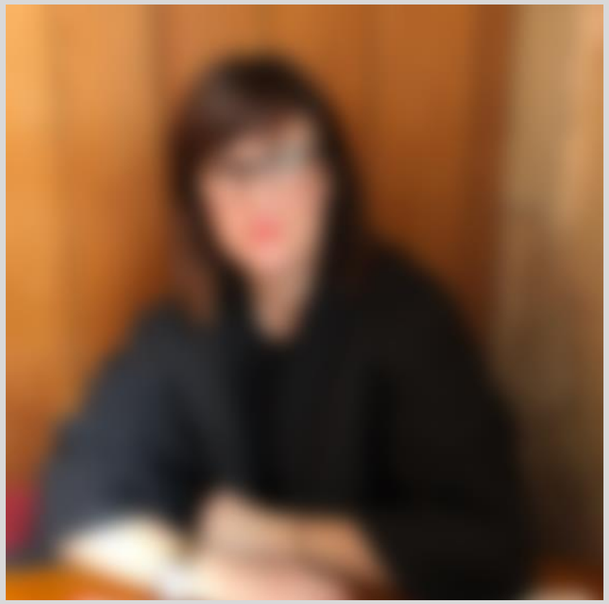


- Bachelor of Design with career as an Interior Architect
- Then ICU nurse with post graduate qualifications
- Mother of two
- Busy and high functioning
- 10 years of mild undiagnosed illness
- Pushed through and became severe in the context of overexercising.

# WILL I SEE PEM IN THE CLINIC?

## WHAT YOU MIGHT SEE....

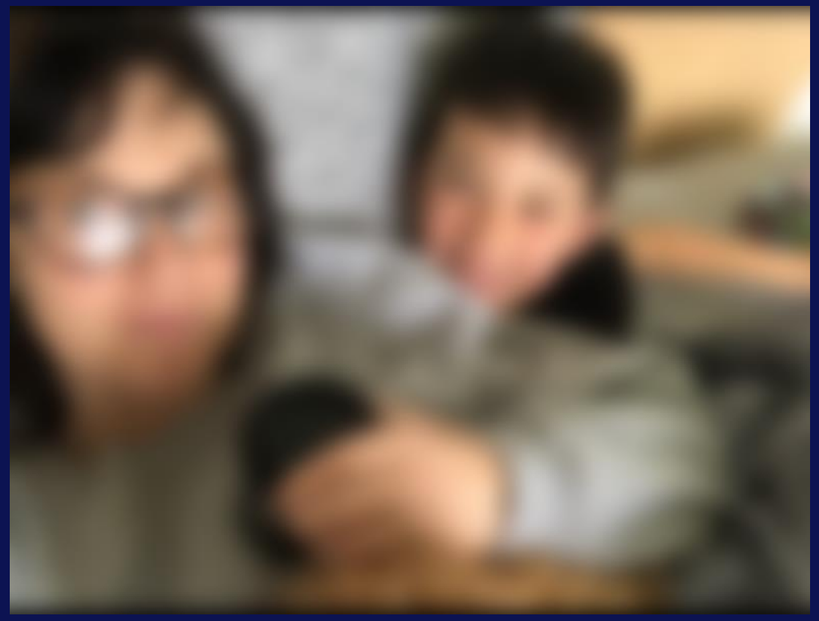
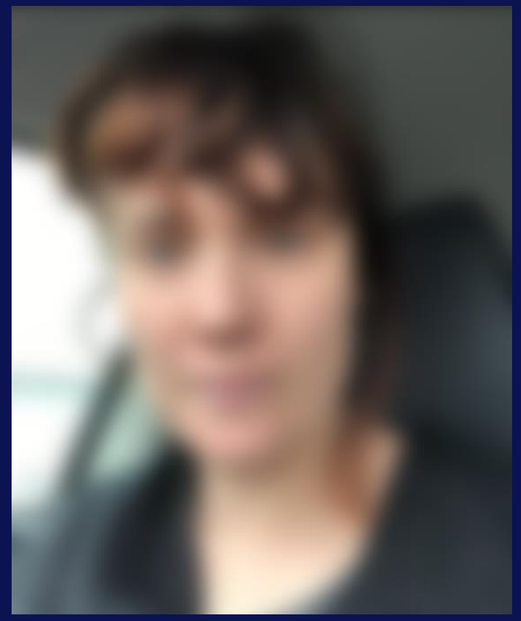
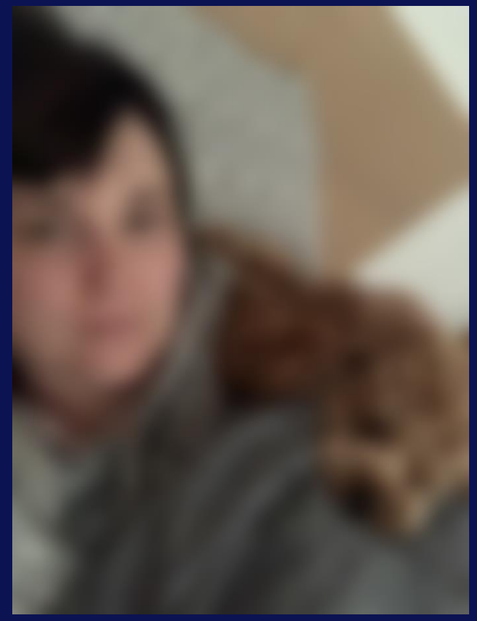
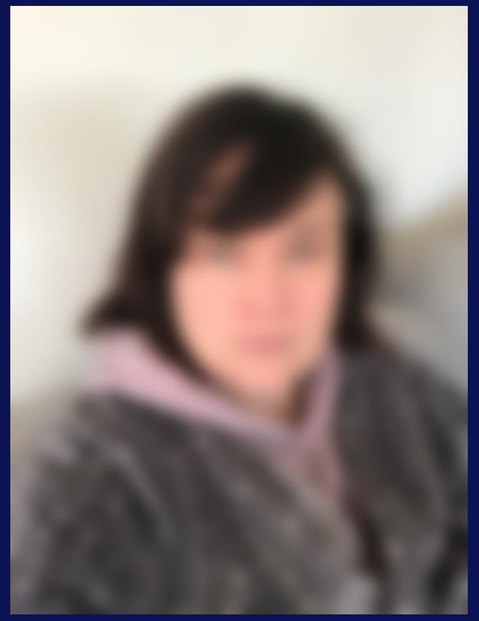




A PATIENT WITH MODERATE / SEVERE ME ON A GOOD DAY

OFTEN DOESN'T APPEAR UNWELL

# 2019 DIAGNOSED WITH ME/CFS

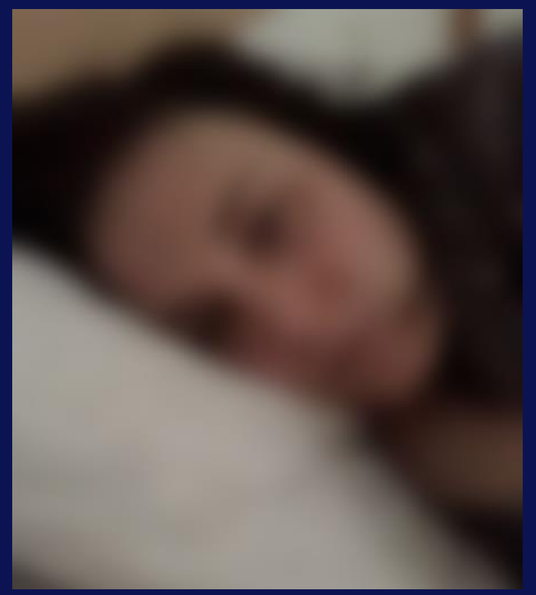
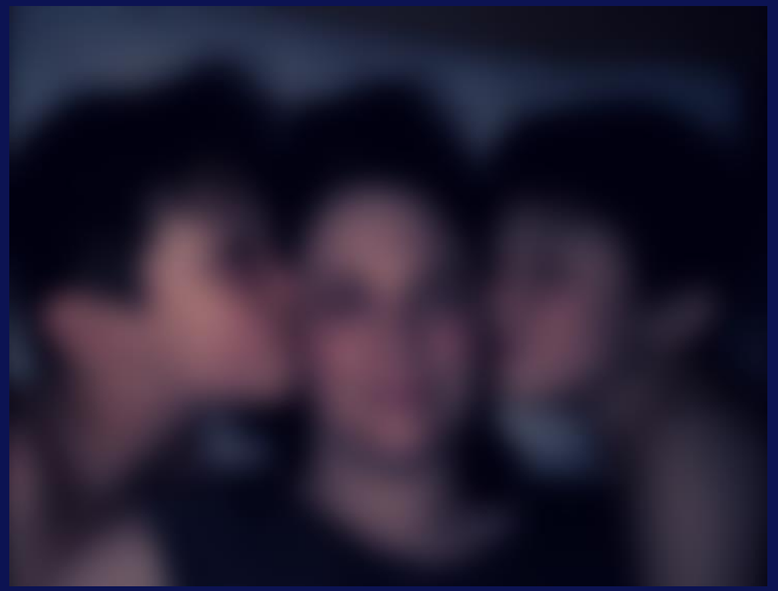
**After developing ME/CFS, this is new baseline**

- **Housebound 70% of time**
- **Unable to work**
- **Often unable to care for children**
- **Unable to cook/clean/household chores**
- **Often unable to wash hair/shower**
- **Use of e-scooter for mobility**
- **Unable to walk more than a few hundred metres at a time**

# AFTER OVEREXERTION.... SEVERE ME

## PEM

- **Profound fatigue**
- **Slurred speech**
- **Unable to leave bed**
- **Difficult to chew food**
- **Intense muscle fatigue with weakness**
- **Arrhythmias** (SVT, elevated rate, 2nd degree block - wenckebach)
- **Dizziness** (cannot tolerate being upright)
- **Shortness of breath**
- **Cognitive dysfunction**
- **Unable to tolerate light or sound**
- **Unable to care for self or children**

*"I feel like I'm drowning,*
*I don't know which way is up,*
*I can't see, I can't find my way out.*
*I can hear voices and movement*
*but can't make sense of them.*
*I want to find the surface for air*
*but I'm weighed down under this immense fatigue.*
*It can be terrifying".*
*Description of PEM from patient.*

# INVESTIGATIONS

ASSESS
INVESTIGATE
DIFFERENTIALS,
DIAGNOSE,
REFER

**Routine blood tests** – complete blood count, C-reactive protein, ferritin, electrolytes, renal and liver function tests, calcium, magnesium, blood glucose, thyroid function tests, vitamin B12, folate, vitamin D and coeliac antibodies

**Other clinically indicated tests**– immunoglobulins, antinuclear antibodies, rheumatoid titres, creatinine kinase and cortisol.

**If associated with a viral illness, consider** – Epstein–Barr virus, cytomegalovirus and HIV testing. Also consider testing for Lyme disease in patients who have travelled outside New Zealand, if symptoms indicate.

Covid 19 is now being shown to be  a significant  trigger.

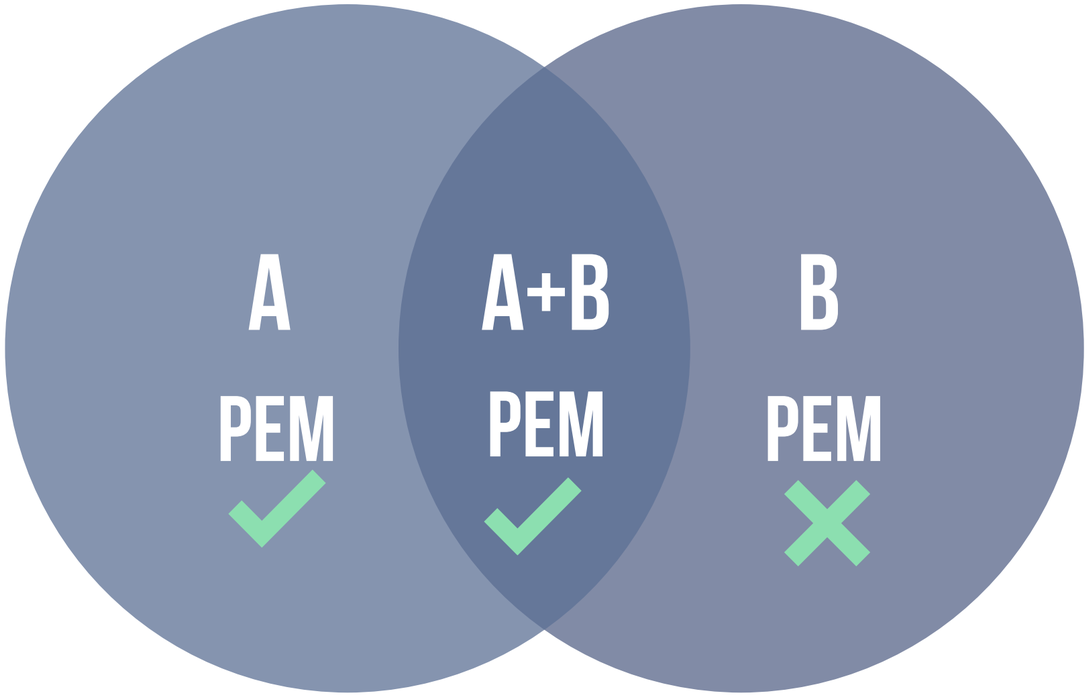
# DIAGNOSTIC CONSIDERATIONS - OVERLAPPING AND COMORBID CONDITIONS ARE THE NORM

**POST-EXERTIONAL MALAISE IS A DISTINGUISHING CRITERION SYMPTOM**

**ASSESS**
**DIAGNOSE**
**INVESTIGATE**
**DIFFERENTIALS**
**REFER**

## A ME/CFS

PEM
Loss of function/Fatigue
Unrefreshing sleep
OI and/or cognitive impairment




## B Overlapping Conditions (sample)

Autonomic Dysfunction (eg POTS)
Mast Cell Activation Syndrome
Fibromyalgia
Hypermobility Syndromes (eg EDS)
Cranio-Cervical Instability
Small Fiber Polyneuropathy
Other Autoimmune Conditions

*For a complete list of overlapping conditions, differential diagnoses and investigations refer to NZ Doctor ME/CFS*

# SYMPTOM MANAGEMENT

## STEPWISE SYMPTOM MANAGEMENT


- Together agree on a stepwise symptom hierarchy.
- List symptoms from most to least troubling.
- Tackle one thing at a time, in line with the symptom hierarchy.
- Sleep, pain and cognitive issues are common problems that GPs are well versed in managing.
- Approach symptom management for ME/CFS patients the same as for anyone else but remember these symptoms tend to be more persistent and resistant to treatment in this group.

# SYMPTOM MANAGEMENT

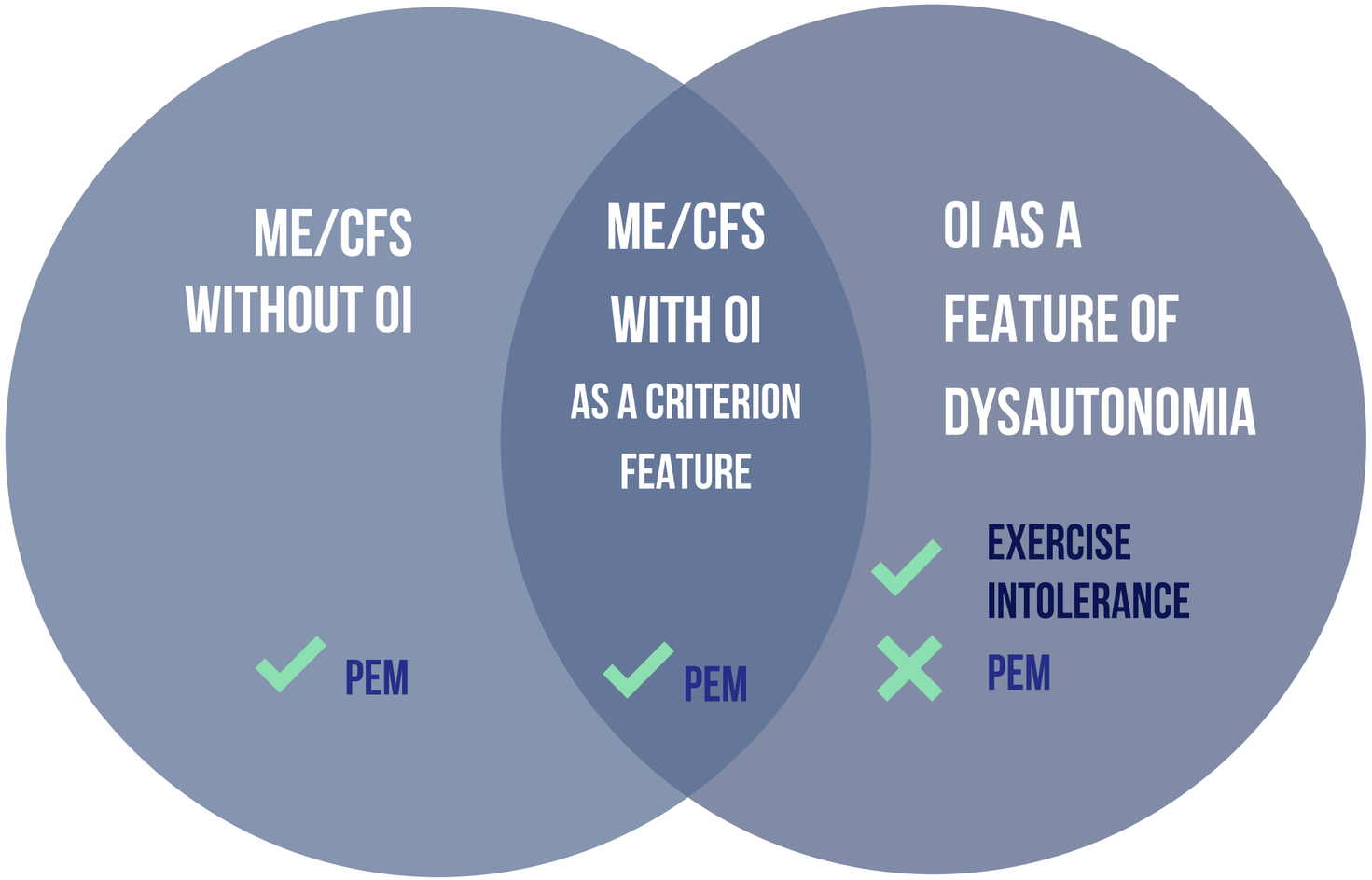
## Management Considerations

- Consider comorbid and differential conditions. LOTS of things can look like ME/CFS.
- Medication sensitivity is the rule. Go low and slow. Minimise polypharmacy.
- There is no definitive evidence for supplementation but B12 is commonly recommended.
- Food  intolerances and sensitivities are common.
- Patients may be aware of emerging evidence - be prepared to partner with them and experiment .

**SYMPTOM MANAGEMENT**
Focus on the patients top priority

**If ME/CFS is suspected commence pacing.**

# ORTHOSTATIC INTOLERANCE




**ASSESS**
**DIAGNOSE**
**INVESTIGATE**
**DIFFERENTIALS**
**REFER**

- Many conditions look and feel like ME/CFS.
- PEM must be present for a diagnosis of ME/CFS.

# ASSESSING POTS

Bateman Horne Center

RESEARCH | CLINICAL CARE | EDUCATION

**ASSESS**
**DIAGNOSE**
**INVESTIGATE**
**DIFFERENTIALS**
**REFER**

## Poor Man's Tilt / Nasa Lean Test / Passive Stand Test

Sustained increase in HR to >30 bpm over baseline (>40 bpm for children/adolescents) OR above 120 bpm.

# MANAGING POTS

## MANAGEABLE AS AN ASPECT OF ME

**SYMPTOM MANAGEMENT**

Agree on a hierarchy of need and focus on one symptom at a time

### First Line

- Rationalise medications that may be contributing
- Increase fluid above 2l
- Increase salt intake up to 8 gm
- Compression
- Physical manoeuvres

### Second line ( in consultation with secondary care)

- Fludrocortisone, midodrine, beta blockers, mestinon
- Ivabradine (very common use in USA and UK - available in NZ but delays due to demand from post covid POTS)

# PACING: REDUCE THE IMPACT OF PEM BY LEAVING SOME FUEL IN THE TANK

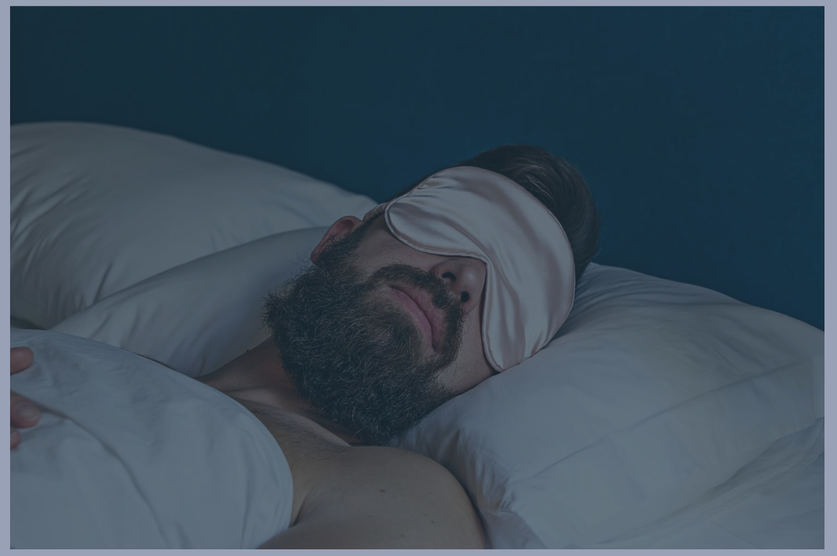
**PACING improves quality of life and reduces PEM.**

**PACING**

**ME/CFS Suspected? Continue investigation AND commence pacing.**

**PACING**

- **Expend  less energy than is available and stay within the Energy Envelope - below threshold that triggers PEM**
- **Energy expenditure includes physical, cognitive and emotional activity.**
- **Expect variability of energy envelope.**
- **Break activities down into short bursts.**
- **Intersperse with rest.**



**Rest means absolutely minimal activity and  minimal stimulation.**

*https://m.e.awareness.nz/a-guide-to-pacing-for-pwme*

# WHAT PATIENTS WANT US TO KNOW

- Accept that your patient may be the expert - partner with your patient.
- Emerging treatment may take decades to be approved. Within the limits of safety be prepared to experiment.
- Fatigue is a symptom but it is not the defining symptom.
- Provide flexible appointment such as phone consultations - coming to the office will trigger PEM for most.
- Assist with practical issues such as benefit applications, referrals to health school .
- Be vigilant for secondary disability - this is a hard road to walk
- ME is NOT a mental illness, patients are experiencing real and distressing physical symptoms.

# TAKE HOME MESSAGES

- Complex, multisystem disorder
- Diagnostic clarity and validity is improving
- GET is not curative and may be harmful
- CBT is supportive but not curative
- Symptomatic treatment can be effective (eg POTS, sleep, pain)
- Listen to your patients and partner with them – it might be a long journey