Palliative care in demyelinating disorders

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مراقبت‌های حمایتی و تسکینی چیست؟

طبق تعریف سازمان بهداشت جهانی مراقبت های حمایتی و تسکینی رویکردی است که به ***بهبود کیفیت زندگی بیماران و خانواده آنان در مواجهه با مشکلات ناشی از بیماری‌های تهدیدکننده حیات می‌پردازد.***

این مراقبت‌ها به تشخیص و کنترل چالش‌های جسمی، روانی، اجتماعی و معنوی در بیمار و خانواده می‌پردازد. از سوی دیگر، سازمان جهانی بهداشت مؤلفه‌های اصلی در کنترل بیماری های سخت درمان را پیشگیری، تشخیص زود هنگام، درمان و مراقبت‌های حمایتی و تسکینی معرفی می‌کند.

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نگرش و فعالیت مراقبت‌های حمایتی و تسکینی

بطور کلی، نگرش و فعالیت مراقبت‌های حمایتی و تسکینی به شکل زیر تعریف می‌شود.
درد و دیگر علائم آزاردهنده بیماری و درمان را تسکین می‌دهد؛
به زندگی بها می‌دهد و مرگ را یک فرآیند طبیعی تلقی می‌کند؛
قصد تسریع مرگ یا به تعویق انداختن آن را ندارد؛
شامل مراقبت‌های روانی و معنوی می‌شود؛
به دنبال ایجاد فرصت فعال زیستن تا آخرین لحظهٔ زندگی برای بیمار است؛
از خانوادهٔ بیمار در مراحل مراقبت از بیمار و سوگواری پشتیبانی می‌کند؛
می کوشد به تمامی نیازهای بیمار و خانواده با رویکردی تیمی (از جمله مشاوره در مورد مرگ و سوگ) پاسخ دهد.
کیفیت زندگی بیمار و خانواده را افزایش می‌دهد و همچنین ممکن است بر طول عمر بیمار و عوارض آن نیز تأثیر مثبت داشته باشد؛
می تواند از ابتدای دوره بیماری و در کنار درمان‌های افزایش دهندهٔ طول عمر، مانند شیمی درمانی و پرتو درمانی، به بیمار کمک کند؛
شامل بررسی‌های مورد نیاز برای درك و مدیریت بهتر علائم و عوارض بیماری و درمان آن می‌شود.

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مراقبت های حمایتی و تسکینی

به هر نوعی از کارهای پزشکی که با هدف کاهش علائم بیماری، کنترل عوارض جانبی درمان، افزایش کیفیت زندگی و حمایت بیمار و خانواده‌ی اوانجام می‌شود، مراقبت‌های تسکینی گفته می‌شود.

سازمان جهانی بهداشت، سلامت را در چهار بعد جسمانی، روانی، اجتماعی و معنوی تعریف می‌کند. بنابرین هیچ چالش آزاردهنده‌ای در مواجهۀ بیمار و خانواده با بیماری نیست که پرداختن به آن از چارچوب مراقبت‌های تسکینی خارج باشد.

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برخلاف دیدگاه‌های سنتی که ارائه مراقبت های حمایتی و تسکینی را از نظر زمانی محدود به هفته‌های پایانی عمر بیمار، یعنی از زمان قطع درمان‌های علاجی تا زمان مرگ، می‌دانستند، امروزه این مراقبت‌ها از بدو تشخیص برای بیمار و خانواده آغاز شده و حتی پس از مرگ بیمار با مراقبت‌های سوگ برای خانواده ادامه می‌یابد.

چنانچه در شکل مشاهده می‌شود با پیشرفت بیماری از بدو تشخیص، نیاز بیمار به مراقبت‌های علاجی کاهش و به مراقبت های حمایتی و تسکینی افزایش می‌یابد.

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زمان مناسب برای استفاده از مراقبت‌های حمایتی و تسکینی

زمانی که بیماری وارد مرحلهٔ نهایی می‌شود، درمان‌های علاجی متوقف می‌شود؛ در حالی که نیاز به مراقبت‌های تسکینی به اوج خود می‌رسد و بیمار و خانواده در روزهای پایانی زندگی تحت پوشش خدمات ویژه حمایتی و قرار می‌گیرند.

مرحله انتهایی مراقبت های حمایتی و تسکیتی تحت عنوان مراقبت‌های پایان عمر به مرحله‌ای از بیماری سخت درمان اتلاق می‌شود که درمان علاجی دیگر سودمند نیست و از نظر علمی بیماری علاج‌ناپذیر ارزیابی شده و شرایط بیمار به صورت فزاینده‌ای رو به وخامت است.

پس از مرگ نیز پشتیبانی از خانواده تا بازگشت آنان به شرایط طبیعی ادامه می‌یابد. معمولا نقطهٔ آغاز این مراقبت‌ها با گفتن خبر بد ابتلای فرد به بیماری سخت درمان به بیمار و خانوادهٔ او در نظر گرفته می‌شود.

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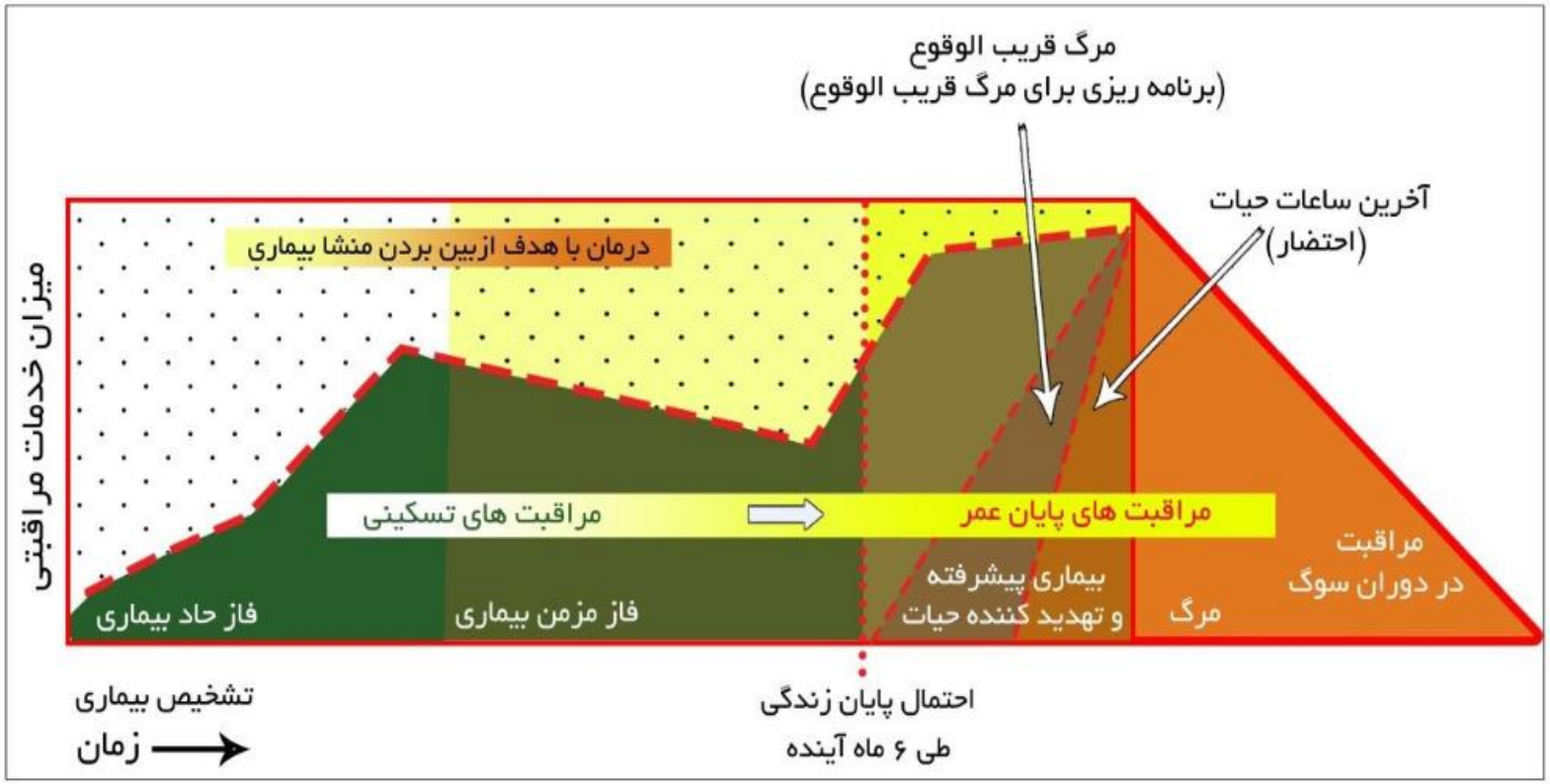

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Multiple Sclerosis: What You Need to Know About the Disease

What does MS look like?

- **Narges** – a 35yo white married mother of 3 who is exhausted all the time and can't drive because of vision problems and numbness in her feet
- **Asghar** – a 25yo Iranian man who stopped working because he can't control his bladder or remember what he read in the morning paper
- **Zahra** – a 10yo girl who falls down a lot and whose parents just told her she has MS
- **Najmeh** – a 47yo white single woman who moved into a nursing home because she can no longer care for herself

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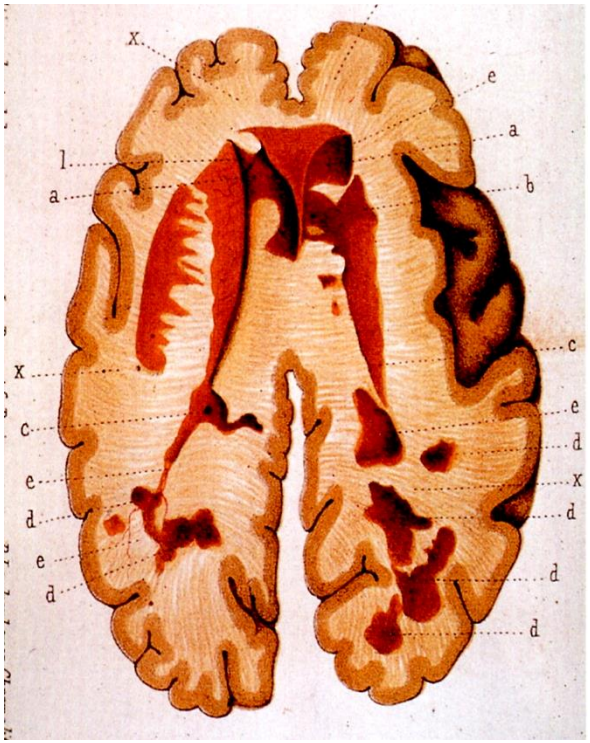
19th Century Highlights



MS-related central nervous system pathology—Jean Cruveilhier, c 1841

Jean-Martin Charcot (1825–1893) described features of MS

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What MS *Is*:

- MS is thought to be a disease of the immune system – perhaps **autoimmune.**
- The immune system attacks **the myelin** coating around the nerves in the central nervous system (CNS – brain, spinal cord, and optic nerves) and the nerve fibers themselves.
- Its name comes from the *scarring* caused by inflammatory attacks at ***multiple*** **sites** in the central nervous system.

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What MS *Is Not:*

- MS is not*:*
  - Contagious
  - Directly inherited
  - Always severely disabling
  - Fatal—except in fairly rare instances
- Being diagnosed with MS is not a reason to*:*
  - Stop working
  - Stop doing things that one enjoys
  - Not have children

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Who gets MS?

- Usually diagnosed between 20 and 50
  - Occasionally diagnosed in young children and older adults
- More common in women than men (2-3:1)
- Most common in those of Northern European ancestry
  - More common in Caucasians than Hispanics or African Americans; rare among Asians
- More common in temperate areas (further from the equator)

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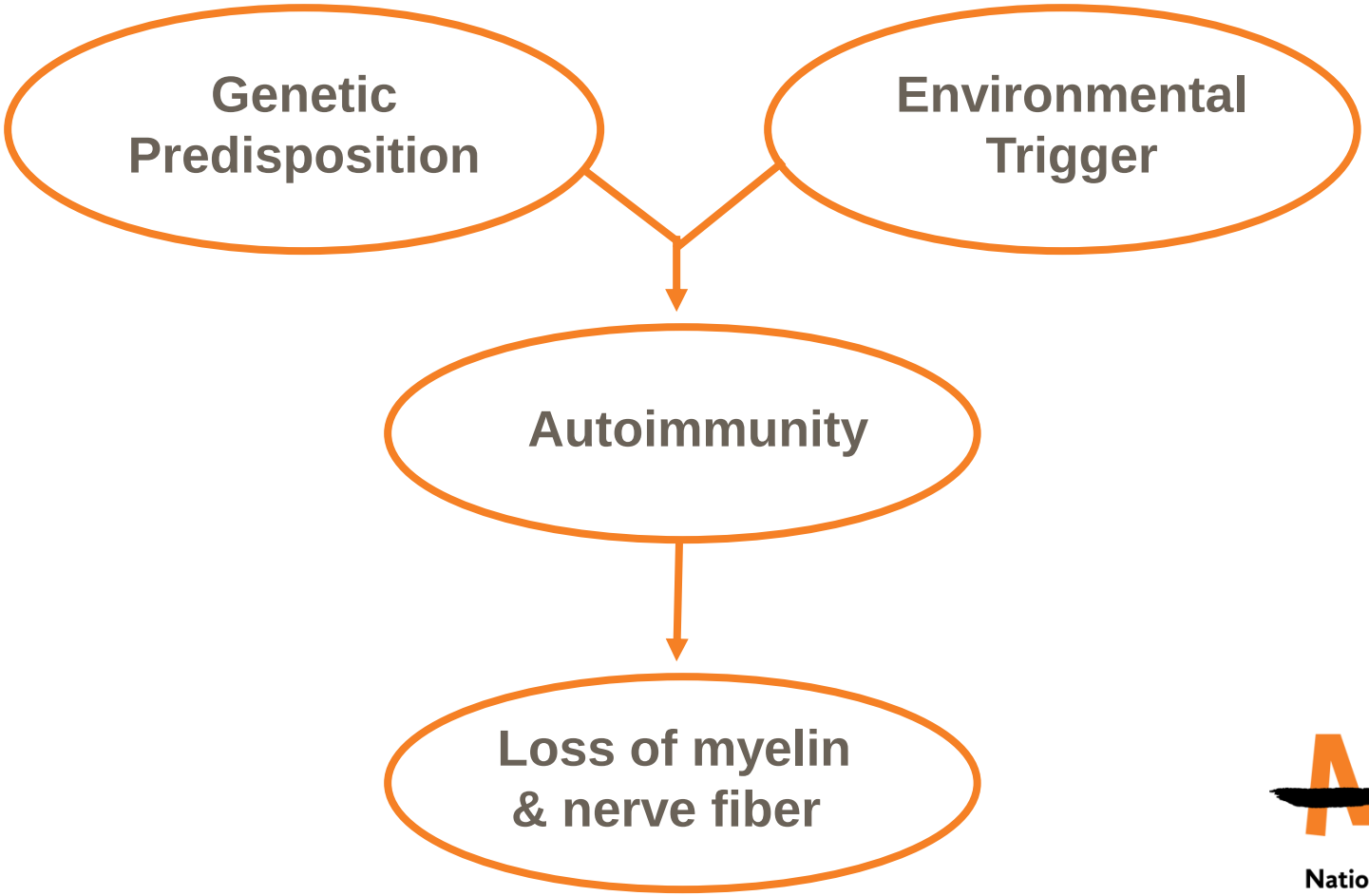
What Causes MS?



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What is the genetic factor?

- The risk of getting MS is approximately:
  - 1/750 for the general population **(0.1%)**
  - 1/40 for person with a close relative with MS **(3%)**
  - 1/4 for an identical twin **(25%)**
- 20% of people with MS have a blood relative with MS

The risk is higher in any family in which there are several family members with the disease **(aka *multiplex families*)**

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What is the prognosis?

- One hallmark of MS is its ***unpredictability.***
  - Approximately 1/3 will have a very mild course
  - Approximately 1/3 will have a moderate course
  - Approximately 1/3 will become more disabled
- Certain characteristics predict a better outcome:
  - Female
  - Onset before age 35
  - Sensory symptoms
  - Mono focal rather than multifocal episodes
  - Complete recovery following a relapse

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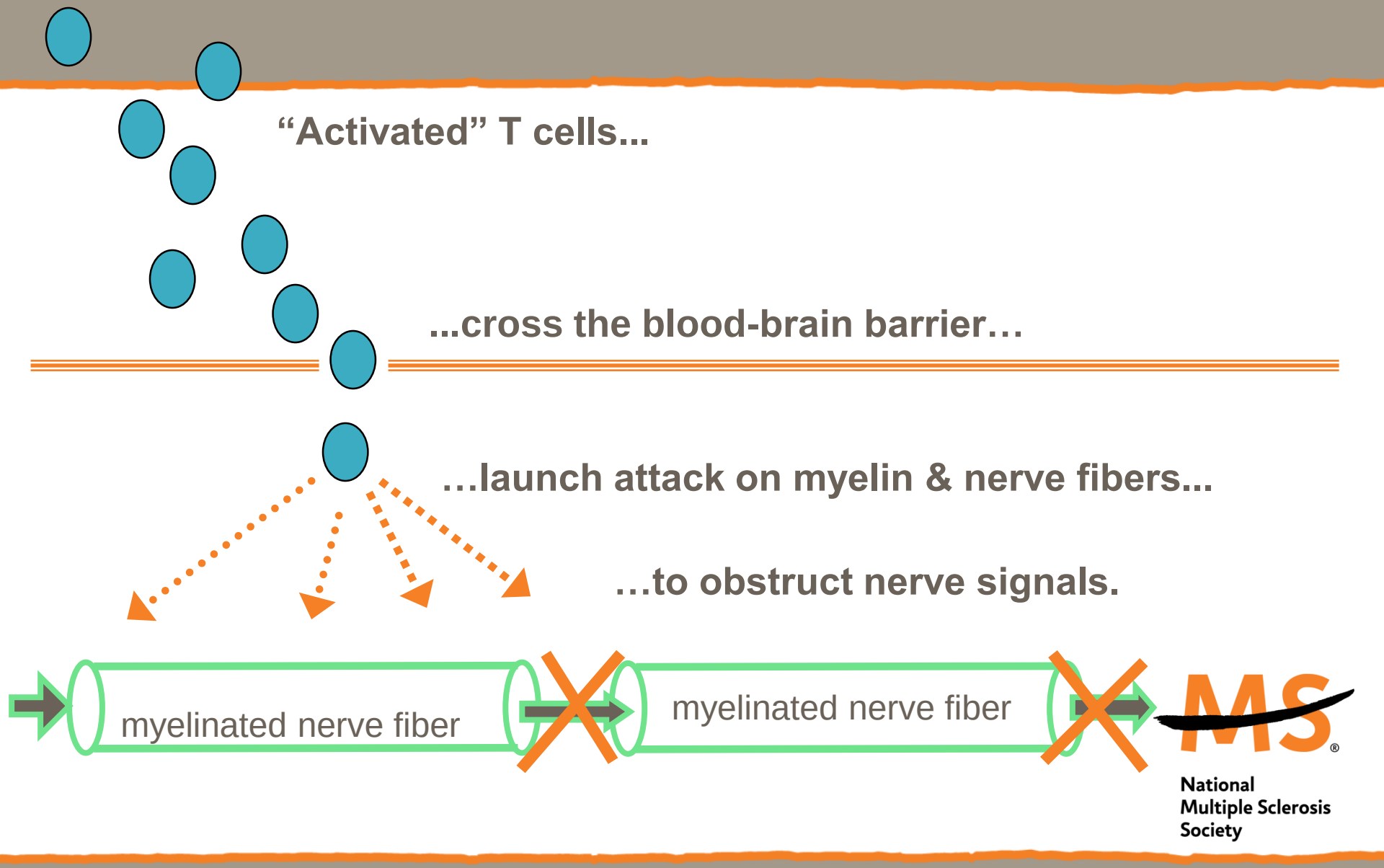
What happens in MS?



What happens to the myelin and nerve fibers?

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What are *possible* symptoms?

- MS symptoms vary between individuals and are unpredictable
  - Fatigue (most common)
  - Decreased visual acuity, diplopia
  - Bladder and/or bowel dysfunction
  - Sexual dysfunction
  - Paresthesias (tingling, (numbness, burning)
  - Emotional disturbances (depression, mood swings)
  - Cognitive difficulties (memory, attention, processing)
  - Pain (neurogenic)
  - Heat sensitivity
  - Spasticity
  - Gait, balance, and coordination problems
  - Speech/swallowing problems
  - Tremor

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How is MS diagnosed?

- MS is a **clinical diagnosis**:
  - Signs and symptoms
  - Medical history
  - Laboratory tests
- **Requires dissemination in time and space:**
  - Space: Evidence of scarring (plaques) in at least two separate areas of the CNS (space)
  - Time: Evidence that the plaques occurred at different points in time
- There must be no other explanation

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Making the Differential Diagnosis

- Infection (Lyme, syphilis, PML, HTLV-1
- Degenerative spinal disease
- Motor neuron disease
- Metabolic (B12 deficiency, familial diseases)
- CNS Lymphoma
- Inflammatory (SLE, Sjogren's, vaculitis, sarcoidosis)

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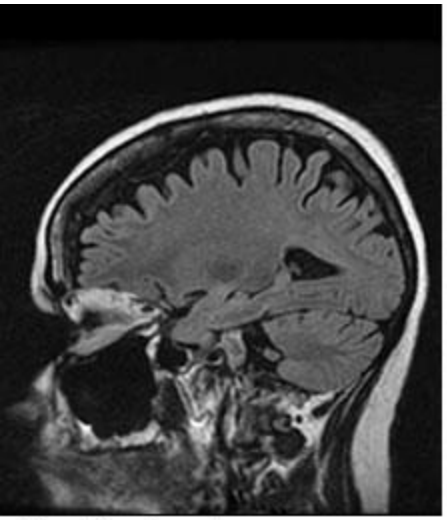
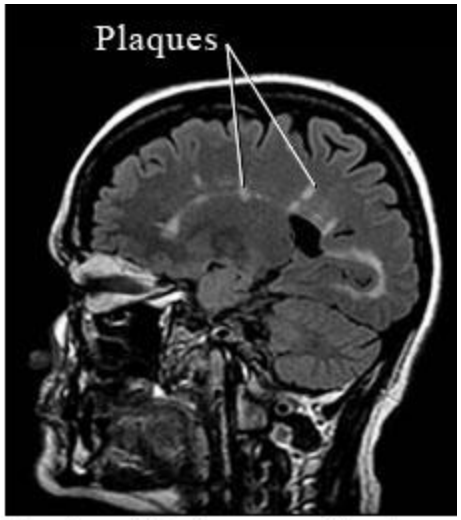
What tests may be used to help confirm the diagnosis?



Healthy brain



Brain with damage (lesions or plaques) caused by MS

- Magnetic resonance imaging (MRI)
- Visual evoked potentials (VEP)
- Lumbar puncture

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What are the Different Patterns (courses) of MS?

- Relapsing-Remitting MS (RRMS)
- Secondary Progressive MS (SPMS)
- Primary Progressive MS (PPMS)
- Progressive-Relapsing MS (PRMS)

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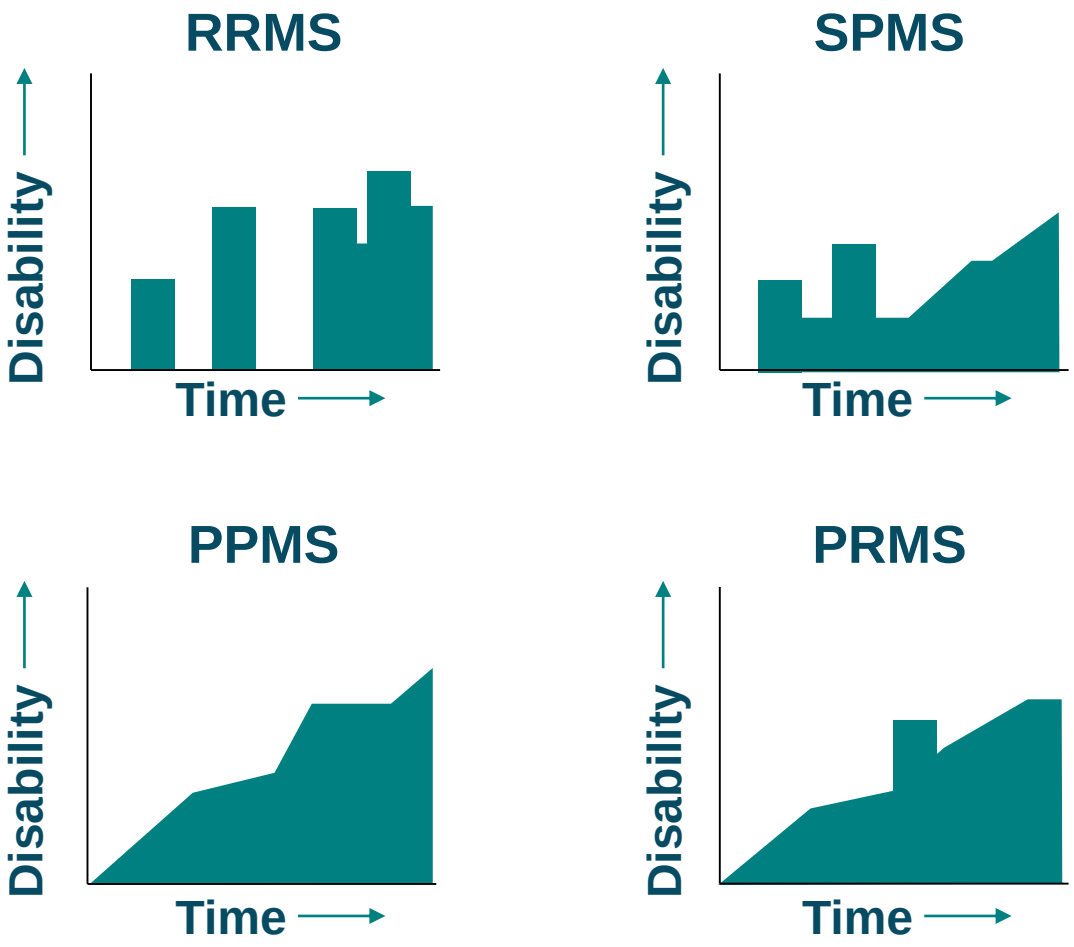
Disease Courses in MS



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The Nurse's Role in Caring for the Newly-Diagnosed MS Patient

- Familiarity with the normal immune system and the pathological mechanisms of MS
- Ability to educate and support patients and families
- Readiness to assist patients in making well-informed treatment decisions

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What are the treatment strategies?

- Gone are the "*Diagnose and Adios*" days of MS care
- Management of MS falls into **five general categories**:
  - Treatment of relapses (aka exacerbations, flare-ups, attacks—that last at least 24 hours)
  - Symptom management
  - Disease modification
  - Rehabilitation (maintain/improve function)
  - Psychosocial support

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How are relapses treated?

Not all relapses require treatment

- **Mild**, sensory sx are allowed to resolve on their own.

Sx that **interfere with function** (e.g., visual or walking problems) are usually **treated**

3-5 day course of IV methylprednisolone—with/without an oral taper of prednisone

High-dose oral steroids used by some neurologists

Rehabilitation to restore/maintain function

Psychosocial support

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MS Symptoms vs. Relapses… How Are They Different?

- MS symptoms are **chronic** or ongoing indicators of MS lesion damage in the brain, spinal cord, and/or optic nerve

- **MS relapses** are sudden flare-ups of disease activity (including new or worsening symptoms) that typically last several days to several weeks or months

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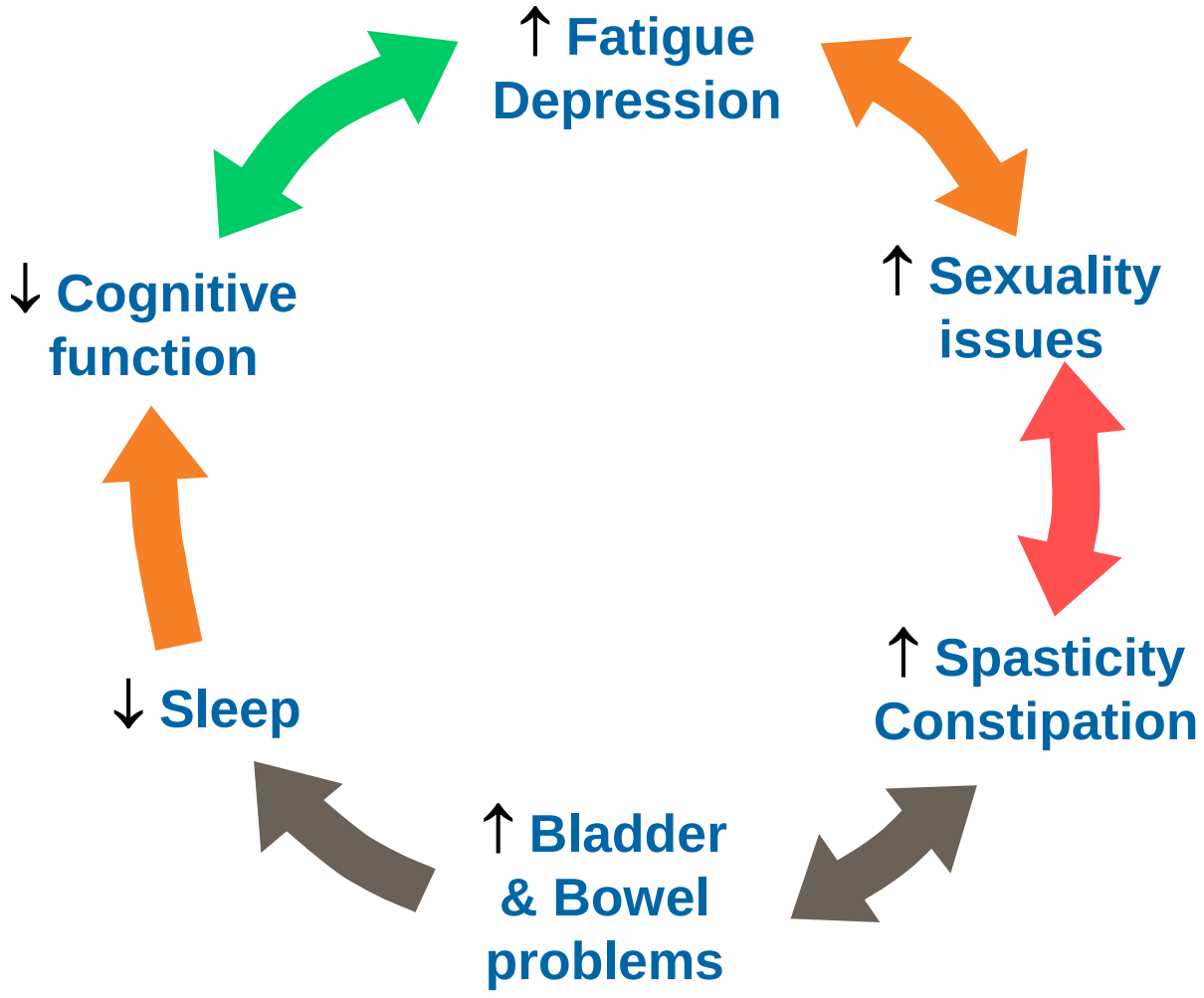
Cycle of MS Symptoms: Related and Interdependent



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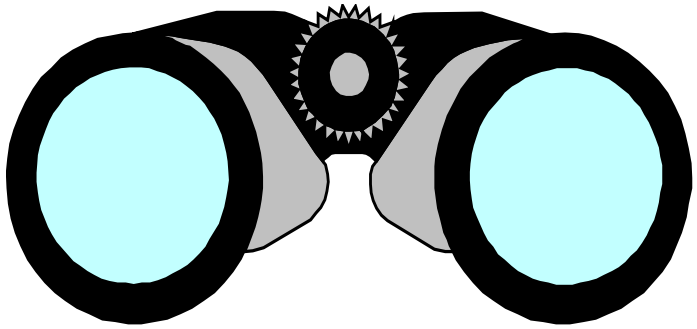
MS Symptoms

- Sort out / prioritize
- Not always MS – rule out other causes
- Any symptom can be related to side effects of medications
- Refer to appropriate discipline as needed



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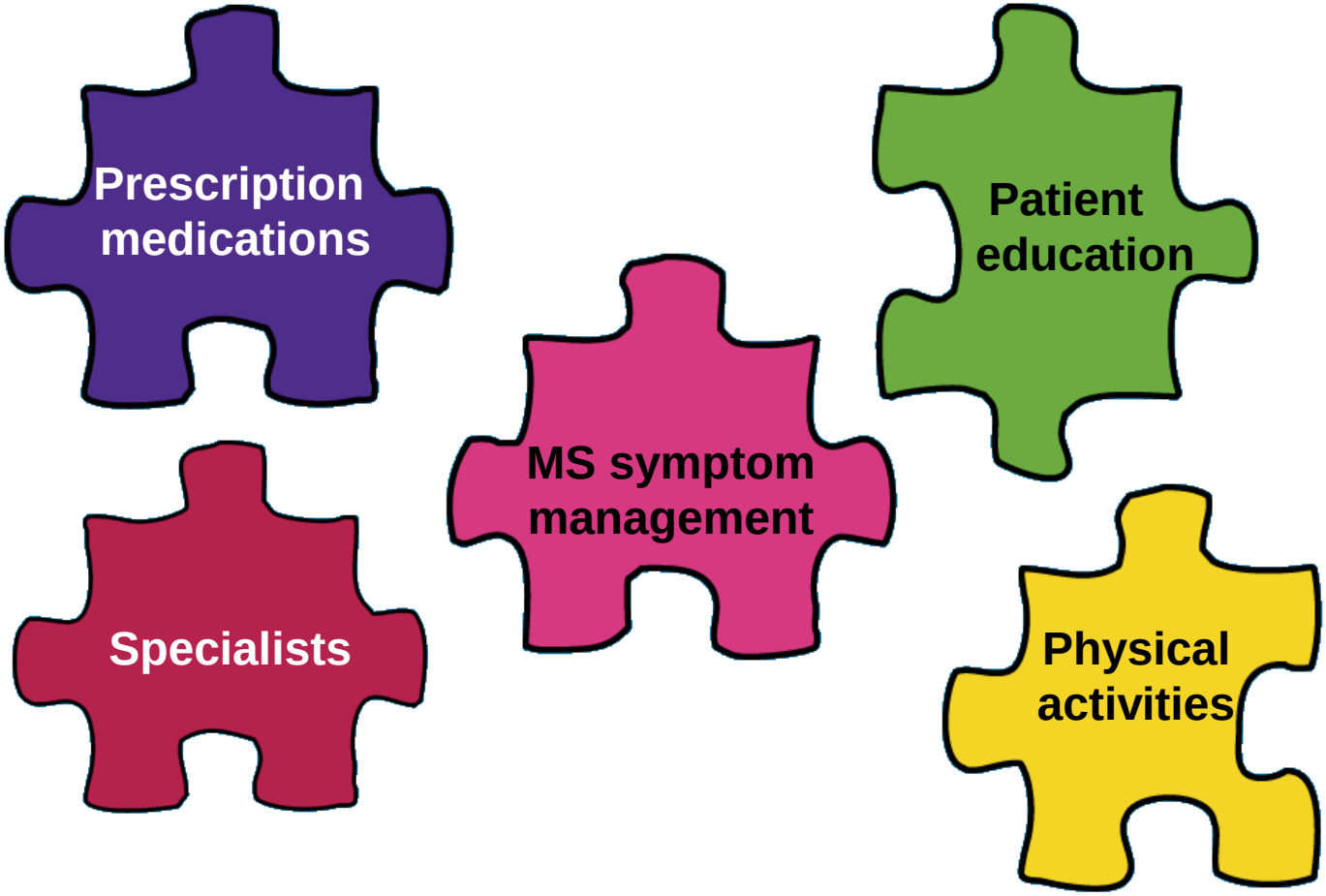
The Recommended Approach to Managing MS Symptoms



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Managing MS Symptoms

SYMPTOM	PHARMACOLOGICAL TX	NURSING INTERVENTIONS
Fatigue	•CNS stimulants: eg, modafinal •SSRIs: eg, fluoxetine	•Assist pt w/dosing; titrate up •Counsel re: naps, work simplification, use of assistive devices (eg. electric scooter), moderate aerobic activity •Referral to OT
Pain	•Anticonvulsants: carbamazepine, gabapentin, phenytoin •Duloxetine hydrochloride	•Assist pt w/dosing; titrate up •Assess for sedation, ↑fatigue •Monitor outcomes

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Managing MS Symptoms

SYMPTOM	PHARMACOLOGICAL TX	NURSING INTERVENTIONS
Cognitive dysfunction	•No symptomatic medications have been shown to be beneficial	•Screen for depression (one of the most common symptoms of MS) •Refer for neuropsychological testing, cognitive rehabilitation, •Consider computer-mediated memory exercises •Encourage regular exercise and healthy sleeping habits

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Managing MS Symptoms

SYMPTOM	PHARMACOLOGICAL TX	NURSING INTERVENTIONS
Bladder dysfunction	•Anticholinergic/antispasmodic: eg, oxybutynin, tolterodine	•Counsel re: behavior modification: regular voiding, eliminate irritants (caffeine, alcohol), encourage fluids •Determine if UTI is present •Monitor retention •Teach ISC
Bowel dysfuntion	•Constipation: stool softeners, bulk-forming agents, rectal stimulants, mild laxatives •Fecal incontinence: anticholinergics (for hyperreflexive bowel)	•Encourage adequate dietary fiber, fluids, exercise, regular pattern of elimination •Provide bowel program, diet counseling (too much fiber?)

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Managing MS Symptoms

SYMPTOM	PHARMACOLOGICAL TX	NURSING INTERVENTIONS
Mobility impairment (eg, balance, weakness, spasticity)	•Dalfampridine (Ampyra) to improve walking (speed; weakness) •See below for spasticity tx	•Refer to PT for exercise program (strengthen muscles & minimize atrophy), assistive devices (canes, braces) •Education re: mobility aids
Spasticity	•GABA agonists (oral or intrathecal baclofen) •α- Agonists (tizanidine) •Anticonvulsants (gabapentin, clonazepam, diazepam) •Botulinum toxin	•Time doses, titrate up •Asses for sedation, weakness •Intrathecal baclofen requires surgical implantation of programmable pump and assoc teaching

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The Nurse's Role In Symptom Management

- Recognize symptoms
- Encourage communication about symptoms
- Discuss treatments and options
- Set realistic expectations
- Follow-up to assess treatment outcomes

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Who is on the MS "Treatment Team"?

- Neurologist
- Urologist
- Nurse
- Physiatrist
- Physical therapist
- Occupational therapist
- Speech/language pathologist
- Psychiatrist
- Psychotherapist
- Neuropsychologist
- Social worker/Care manager
- Pharmacist

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How is the disease course treated?

- Thirteen disease-modifying therapies are FDA-approved for relapsing forms of MS:
  - glatiramer acetate (Copaxone®; Glatopa™ - generic equivalent) [inj.]
  - interferon beta-1a (Avonex®, Plegridy™, Rebif®) [inj.]
  - interferon beta-1b (Betaseron® and Extavia®) [inj.]
  - dimethyl fumarate (Tecfidera™) [oral]
  - fingolimod (Gilenya™) [oral]
  - teriflunomide (Aubagio®) [oral]
  - alemtuzumab (Lemtrada™) [inf]
  - natalizumab (Tysabri®) [inf]
  - mitoxantrone (Novantrone®) [inf]

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What do the disease-modifying drugs do?

- All reduce attack frequency and severity, reduce lesions on MRI, and probably slow disease progression.
- These medications are not designed to:
  - Cure the disease
  - Make people feel better
  - Alleviate symptoms

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How important is early treatment?

- The Society's National Medical Advisory Committee recommends that treatment be considered **as soon as** a dx of relapsing MS has been confirmed.
  - Irreversible damage to axons occurs even in the earliest stages of the illness.
  - Tx is **most effective** during **early**, inflammatory phase
  - Tx is **least effective** during **later**, neurodegenerative phase
- No treatment has been approved for primary-progressive MS.

Approximately 60% of PwMS are on Tx

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What is a *clinically-isolated syndrome* (CIS)?

- **First neurologic episode** caused by demyelination in the CNS
- May be monofocal or multifocal
- May or may not go on to become MS
  - CIS accompanied by MS-like lesions on MRI is more likely to become MS than CIS without lesions on MRI
- All four injectable medications delay second episode

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Treatment Adherence Issues

- Patient readiness is key
- Factors affecting adherence include:
  - Lack of knowledge about MS
  - Unrealistic expectations
  - Denial of illness
  - Side effects
  - Cultural factors
  - Lack of support (medical team, family)
  - Distrust of medical community

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The Nurse's Role as Patient Advocate

- Educate Patients
- Empower Patients
  - educate patients on insurance coverage
  - educate patients how to achieve optimal benefits from healthcare team
- Connect patients with community programs
- Assist with life planning
  - advance directives, living wills, healthcare proxies

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So What Do We Know About MS?

- MS is a chronic, unpredictable disease.
- The cause of MS is still unknown
- **MS affects each person differently**; symptoms vary widely.
- MS is **not fatal**, contagious, directly inherited, or always disabling.
- Early diagnosis and treatment are important:
  - Significant, irreversible damage can occur early on
  - Available treatments reduce the number of relapses and may slow progression
- Treatment includes: relapse management, symptom management, disease modification, rehabilitation, emotional support.

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What can people do to feel their best?

- Reach out to their support system; *no one needs to be alone in coping with MS*.
- Stay connected with others; avoid isolation.
- Become an educated consumer.
- Make thoughtful decisions regarding:
  - Disclosure
  - Choice of physician
  - Employment choices
  - Financial planning
- Be aware of common emotional reactions.

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National MS Society Resources for Your Patients

- Nationwide network of chapters around the country
- Web site (www.nationalmssociety.org)
- Access to information and referrals (1-800-344-4867)
- Educational programs (in-person, online)
- Support programs (self-help groups, peer and professional counseling, friendly visitors)
- Consultation (legal, employment, insurance, long-term care
- Financial assistance

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National MS Society Resources for You

- **Professional Resource Center**
  www.nationalMSsociety.org/PRC
  healthprof_info@nmss.org
  - **Clinical consultations with MS specialists**
  - **Literature search services**
  - **Professional publications**
  - **Professional education programs (medical, rehab, nursing, mental health)**
  - **Consultation on insurance and long-term care issues**

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