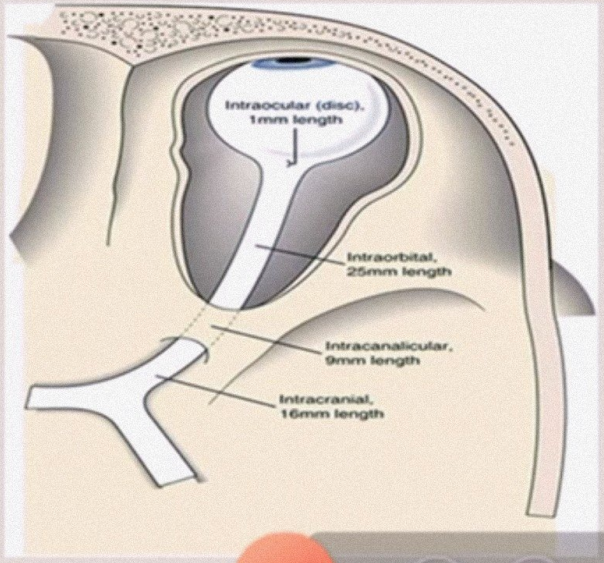
# WHAT IS OPTIC NEURITIS?

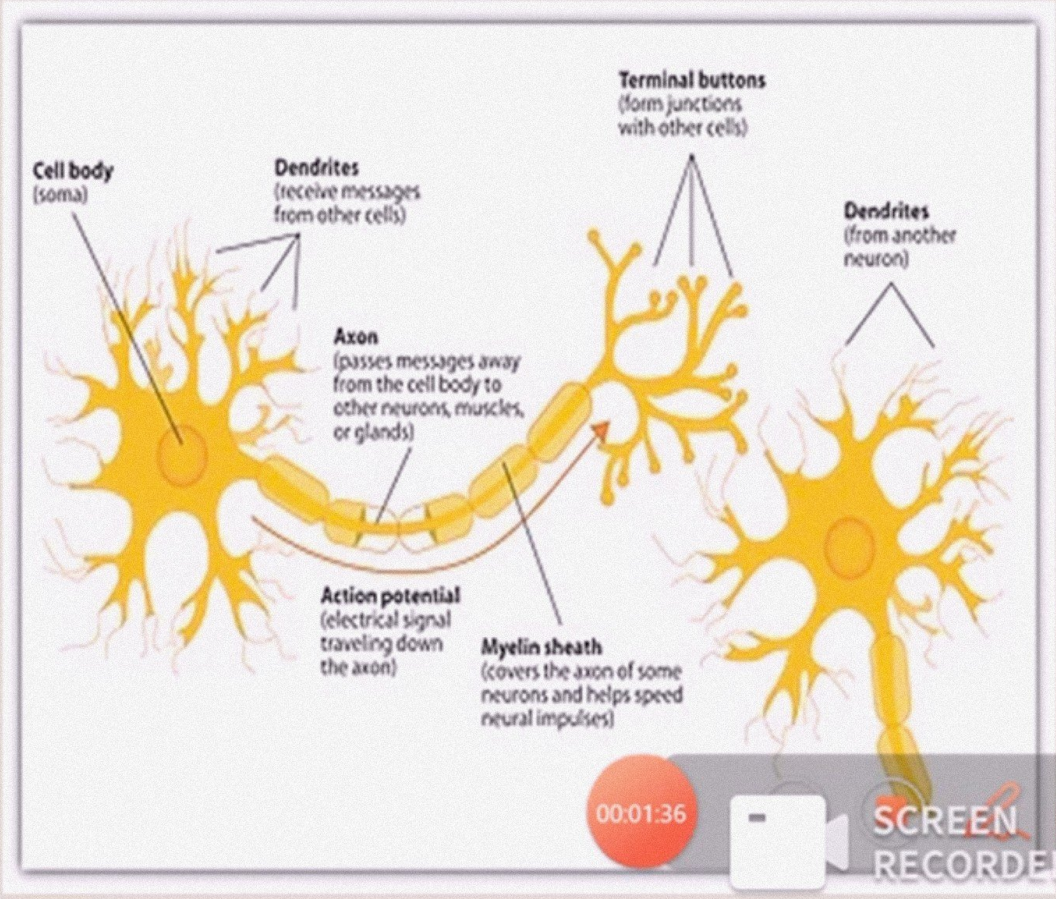
- A demyelinating inflammation of the optic nerve is known as optic neuritis. The optic nerves may be affected by inflammation in any part of its course.

- **PAPILLITIS**
- **NEURORETINITIS**
- **RETROBULBAR NEURITIS**



# PATHOPHYSIOLOGY

**DEMYELINATION**

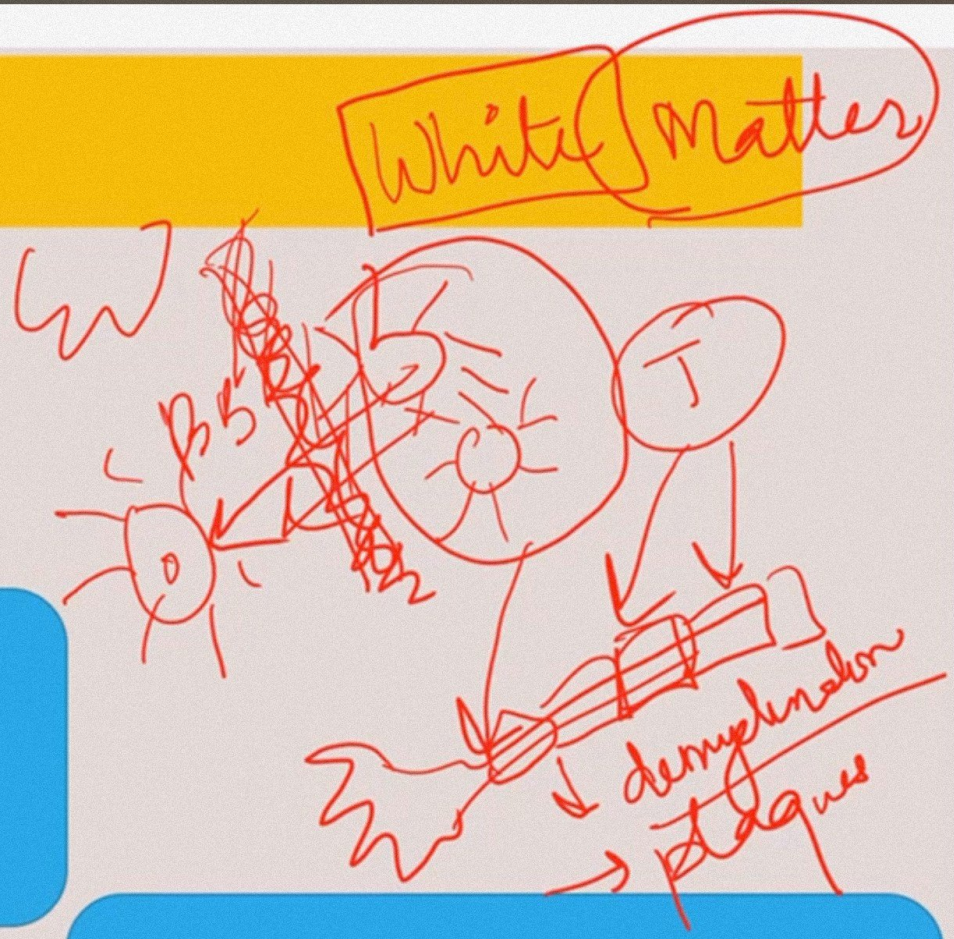


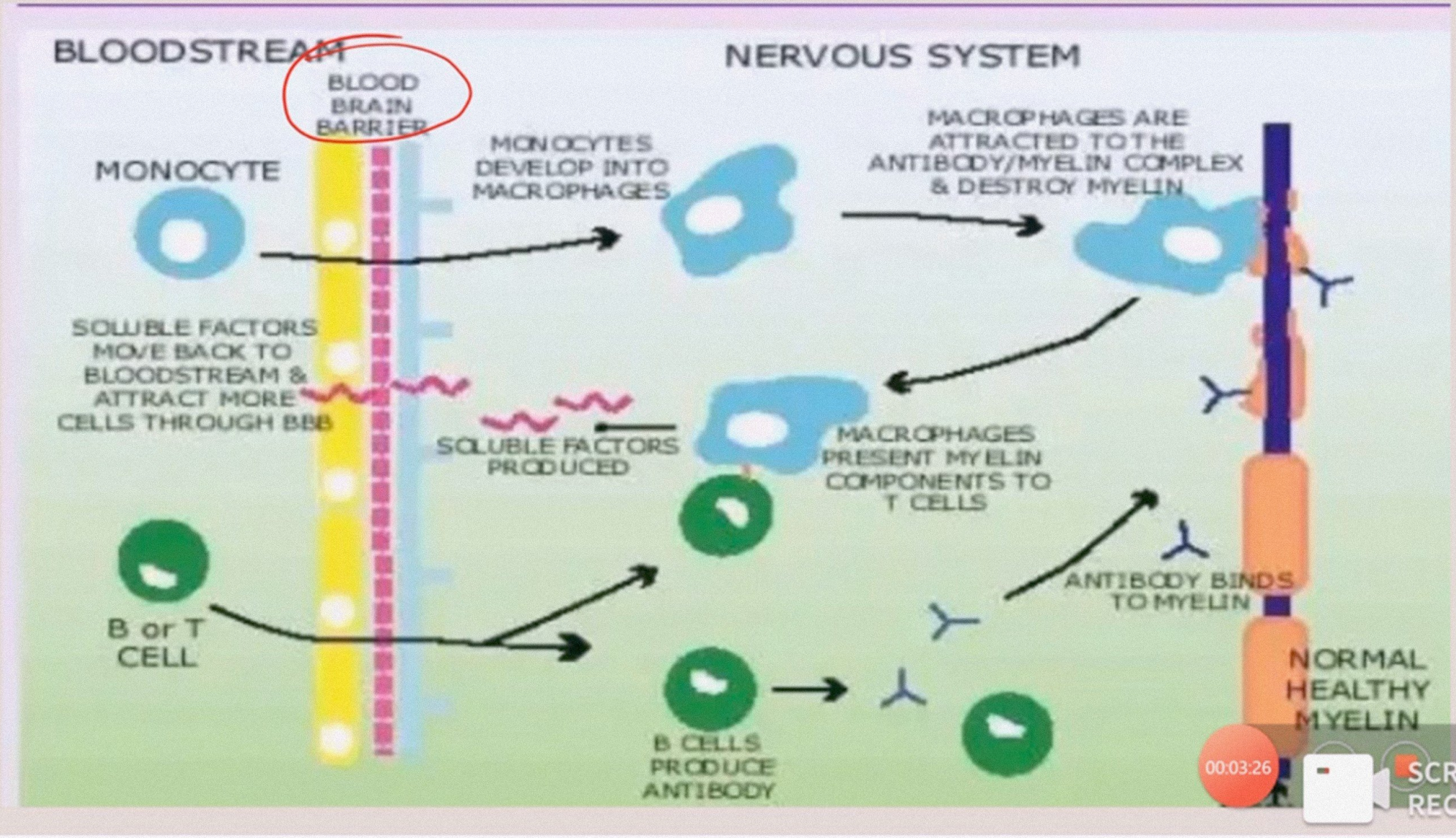
# PATHOPHYSIOLOGY

BLOOD BRAIN BARRIER BREAKDOWN

AUTOIMMUNOLOGY

INFLAMMATION




BLOODSTREAM
NERVOUS SYSTEM
BLOOD BRAIN BARRIER
MONOCYTE
MONOCYTES DEVELOP INTO MACROPHAGES
MACROPHAGES ARE ATTRACTED TO THE ANTIBODY/MYELIN COMPLEX & DESTROY MYELIN
SOLUBLE FACTORS MOVE BACK TO BLOODSTREAM & ATTRACT MORE CELLS THROUGH BBB
SOLUBLE FACTORS PRODUCED
MACROPHAGES PRESENT MYELIN COMPONENTS TO T CELLS
B or T CELL
ANTIBODY BINDS TO MYELIN
NORMAL HEALTHY MYELIN
B CELLS PRODUCE ANTIBODY
00:03:26
SCREEN RECORDER

# PATHOGENESIS

- Demyelination in  varying degrees, which could be axial or peripheral.
- Histopathological

- **PERIVASCULAR CUFFING, T LYMPHOCYTES AND PLASMA CELLS.**

- **EDEMA OF THE MYELINATED NERVE SHEATHS**

- **MYELIN BREAKDOWN**

# DEMYELINATING DISORDERS

- Isolated
- Associated with multiple sclerosis
- Neuromyelitis Optica (DEVICS DISEASE)
- ACUTE DISSEMINATATED ENCEPHALOMYELITIS

# IMPORTANT NEW ANTIBODIES

## AQP4 - IgG

ANTIBODIES AGAINST ASTROCYTE AQUAPORIN -4 WATER CHANNELS

2.1.

## MOG-IgG Ab

ANTIBODIES AGAINST THE MYELIN OLIGODENDROCYTES GLYCOPROTEINTHAT RESULTS IN DAMAGE TO MYELIN INSULATION AROUND CNS AXONS

15.1.

# Associated with infections

- Associated with infections

## Local

- Endophthalmitis
- Orbital cellulitis
- Sinusitis
- Contiguous spread from meninges, brain, base of skull

# SYSTEMIC INFECTIONS

- **Viral***—Influenza, measles, mumps, chicken pox, herpes zoster, infectious mononucleosis, cytomegalovirus
- **Bacterial**—Tuberculosis, syphilis (perineuritis), cat-scratch disease (Bartonella, Rochalimaea henselae and R. quintana), Lyme disease (borreliosis)
- Fu**ngal**—Cryptococcosis, histoplasmosis (Histoplasma capsulatum)
- **Protozoal**—Toxocariasis (Toxocara canis), toxoplasmosis (Toxoplasma gondii), malaria (Plasmodium), pneumonia (Pneumocystis carinii)
- **Parasitic**—Cysticercosis (Cysticercus cellulosae)

# AUTOIMMUNE VASCULITIS

- **SLE (SYSTEMIC LUPUS ERYTHMATOSIS)**
- **POLYARTERITIS NODOSA**
- The pathogenesis is related to ischemia, which may produce demyelination alone, axonal necrosis, or a combination of the two. The clinical profile includes acute optic neuritis (both papillitis and retrobulbar neuritis), acute ischemic optic neuropathy and chronic progressive visual loss.

- **METABOLIC CAUSES**

- ANEMIA,
- B12 deficiency
- DIABETIS,
- PREGNANCY,
- STARVATION,

# CLINICAL FEATURES

Typical ON

- Loss of vision which typically deteriorates over hours to days and reaches a trough about 1 week after the onset.
- The visual loss can be subtle or profound (there may even be complete blindness in a few patients)
- It is usual unilateral
- 18 and 45 years of age.
- It is accompanied by deep orbital, retroocular or brow pain usually aggravated by eye movement and is increased by pressure upon the globe

# CLINICAL FEATURES

- Other visual functions such as loss of colour vision (typically red desaturation) and reduced perception of light intensity
- Occasionally, patients may observe an altered perception of moving objects (**Pulfrich phenomenon**)
- worsening of symptoms with exercise or an increase in body temperature **(Uhthoff sign).**

# COURSE OF TYPICAL OPTIC NEURITIS

- Vision starts to improve in the second or third week and by the fourth to fifth week visual acuity returns to normal or near normal (6/18 to 6/12; 20/60 to 20/40).
- Subsequently, vision slowly and steadily improves over several months and is ultimately usually restored to 6/6 (20/20).
- Color vision, contrast sensitivity and visual fields take longer to recover (6–12 months or so) and may never return completely to normal

TYPICAL OPTIC NEURITIS

AGE : YOUNG ADULTS 18-45 years

MILD PAIN THAT WORSENS ON EYE MOVEMENT

NORMAL DISC OR MILD DISC EDEMA

TYPICAL OPTIC NEURITIS

PROGRESSION OVER HOURS TO DAYS

STARTS IMPROVING IN 2-4 WEEKS

IMPROVEMENT IRRESPECTIVE OF STEROID TREATMENT

CONTINUES TO IMPROVE AFTER STEROID WITHDRAWAL

# ATYPICAL OPTIC NEURITIS

- Outside Typical age range
- No pain on eye movements or severe pain
- Poor vision persisting beyond 2 weeks from onset,
- Progressive diminution of vision beyond the first week
- Recurrence after stopping steroids
- Bilateral involvement
- Severe disc edema with hemorrha

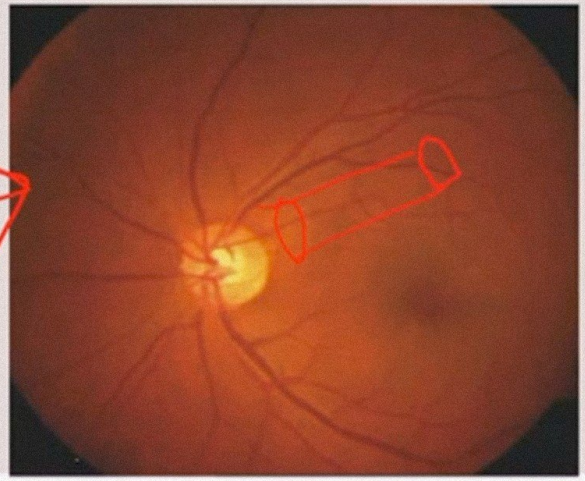
indications for specific further investigations.

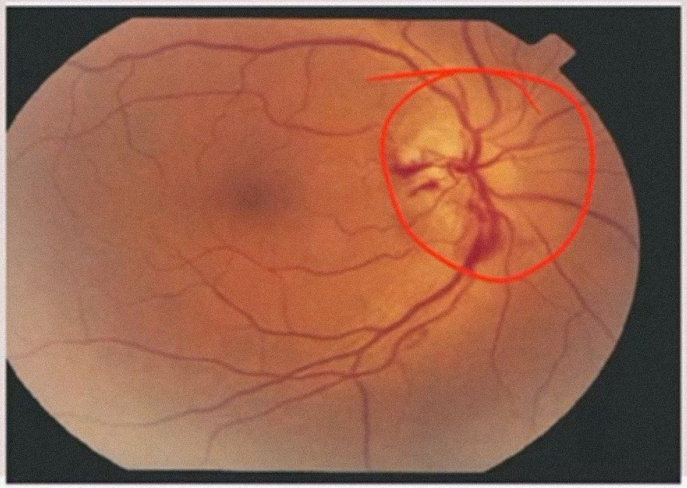
# SIGNS

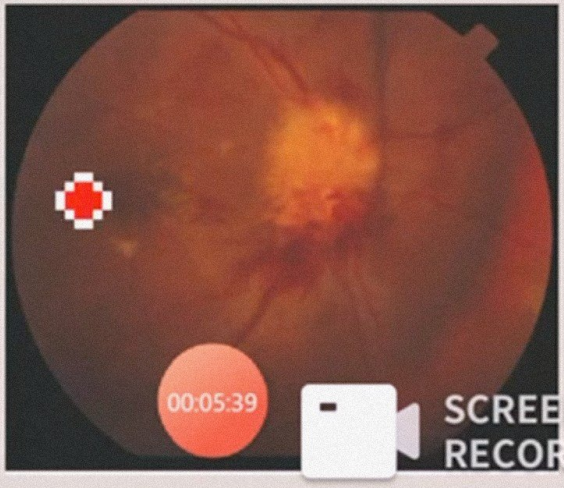
- variable degree of decreased visual acuity
- decreased colour vision,
- abnormal contrast sensitivity,
- decreased stereoacuity
- visual field defects which could be central, centrocaecal, arcuate, sectorial, altitudinal focal pattern defects or a generalized non-specific depression in retinal sensitivity
- Presence of a relative afferent pupillary defect or Marcus Gunn pupil
- Prolonged latency is seen on testing the visual evoked potentials (VEP)

# OPHTHALMOLOGICAL SIGNS

- the disc could be normal in **retrobulbar neuritis** which is more common in adults.
- It may be hyperaemic and swollen with or without peripapillary flame-shaped haemorrhages in **papillitis** , which is most commonly seen in children and young adults.
- 
- It may be inflamed with involvement of the neighbouring retina showing a stellate pattern of retinal exudates in **neuroretinitis**, which is commonly due to an infectious aetiology, secondary or atypical optic neuritis or in children and is **not** seen in multiple sclerosis.

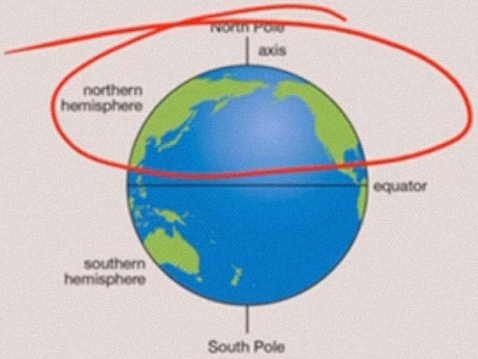






# RISK OF MULTIPLE SCLEROSIS WITH OPTIC NEURITIS

- AGE OF ONSET : YOUNGER THE PATIENT ; MORE IS THE RISK OF MS
- GENDER : Women 3 times greater risk
- RACE : CAUCASIANS
- GEOGRAPHICAL AREA : Northern LATITUDES
- CLINICAL FEATURES : RETROBULBAR, BILATERAL or SECOND EYE INVOLVEMNT WITHIN 2 WEEKS



# INVESTIGATIONS

- **MRI ORBIT AND BRAIN : to evaluate optic nerve enhancement and cerebral demyelination**

TYPICAL OPTIC NEURITIS is UNILATERAL and short segment involving ( <3mm)
FAT SUPPRESSED T2 weighted images HIGH T2 signal in ON
OPTIC NERVE ENHANCEMENT

# DEMYELINATION PLAQUES

Prognostic assessing MS

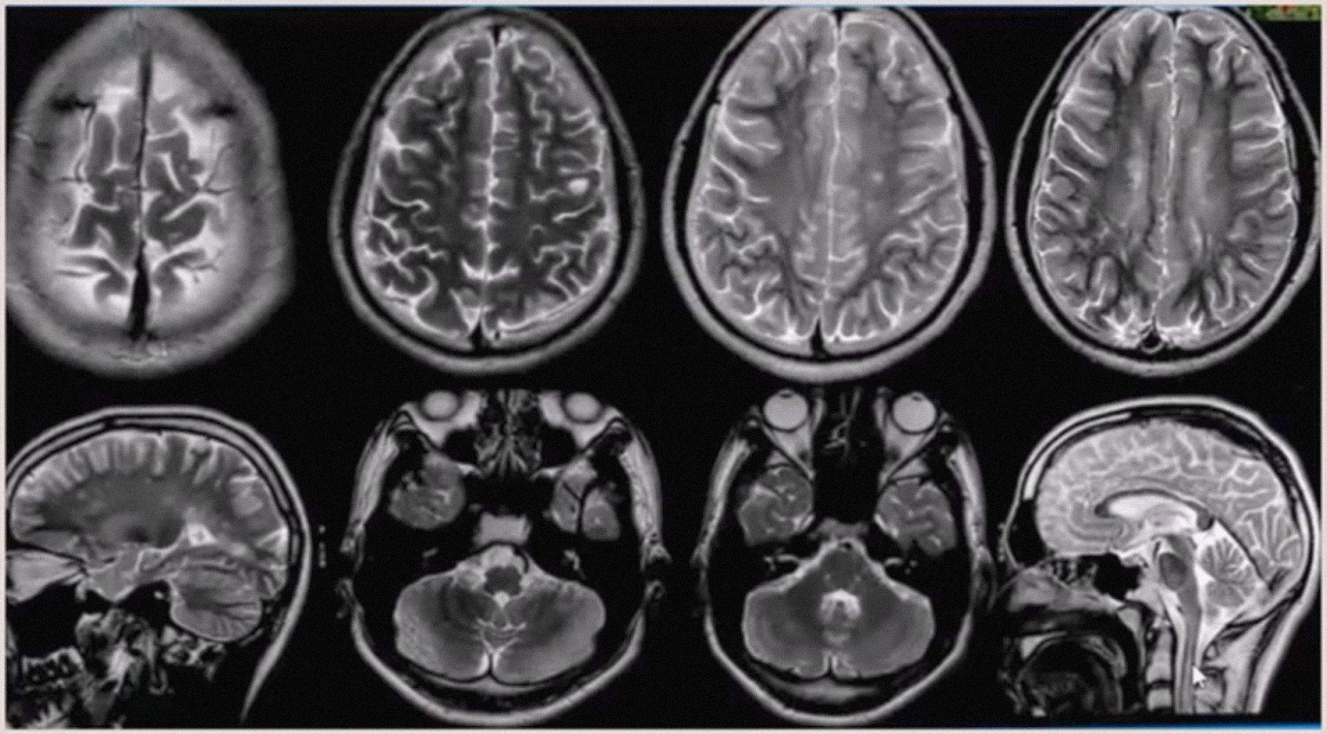
- If no lesions : 25 % risk of MS over 15 years
- If 1 lesion :60 % risk
- 2 lesions : 68 %
- 3 lesions 78%

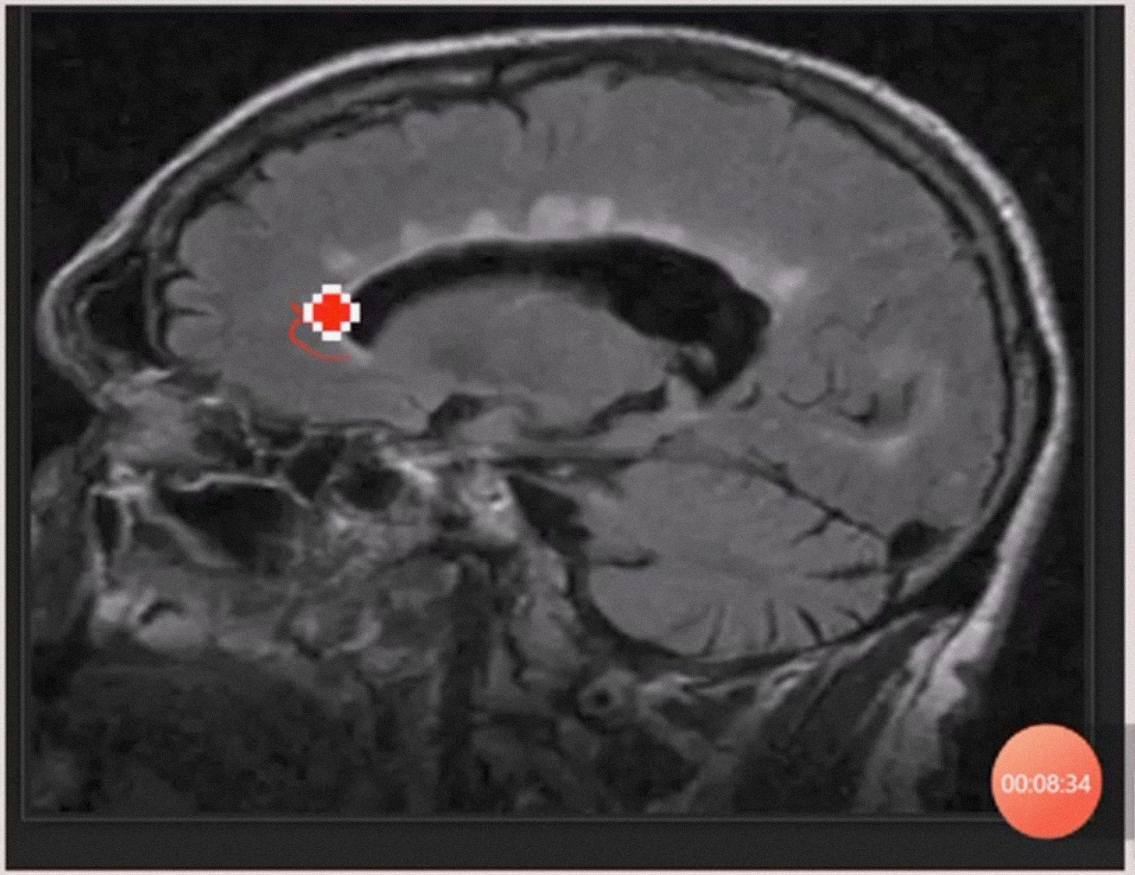
DISSEMINATION IN SPACE

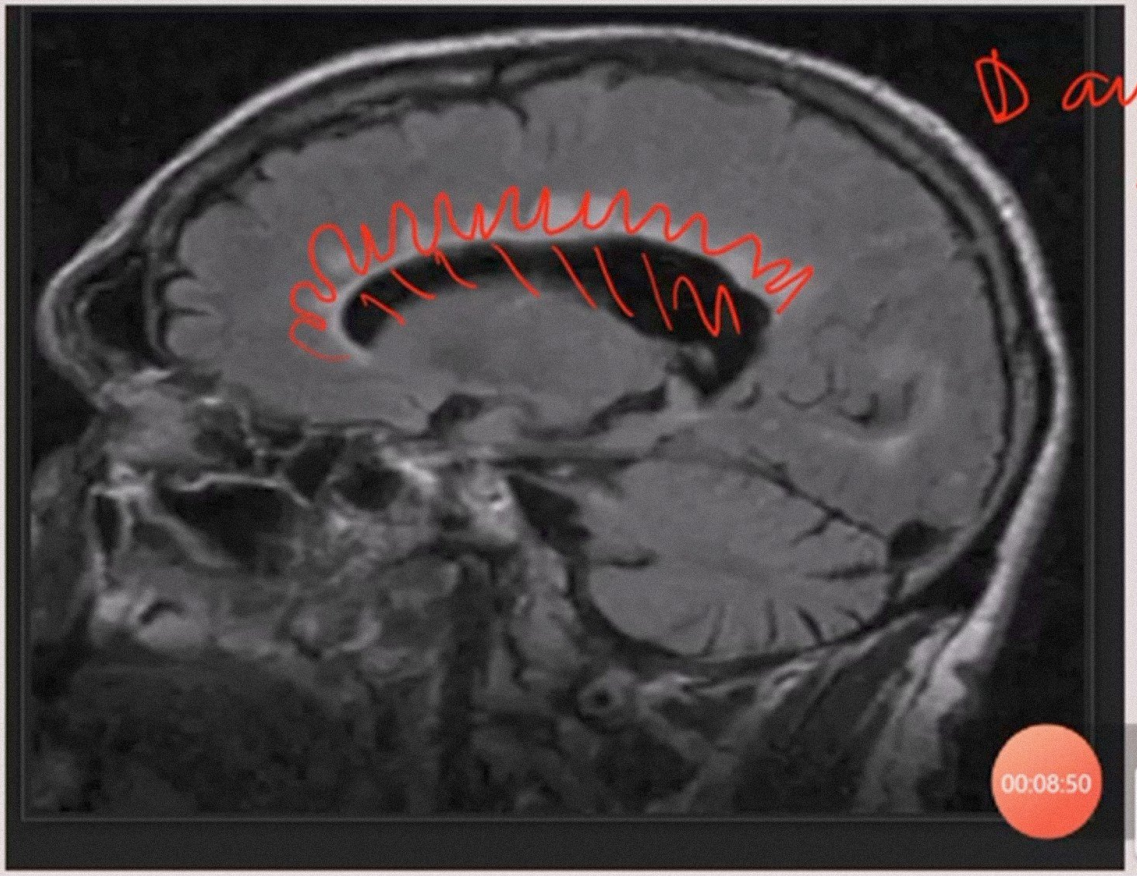
- At least One T2 hyperintense lesions in ATLEAST two of four CNS LOCATIONS
- periventricular, cortical or juxtacortical, infratentorial or spinal cord lesions

DISSEMINATION IN TIME

- Gadolinium enhanced and non-enhanced lesions appearing simultaneously .
- Or NEW lesions appearing on recent imaging compared to older imaging ( IRRESPECTIVE OF TIME SINCE BASELINE )



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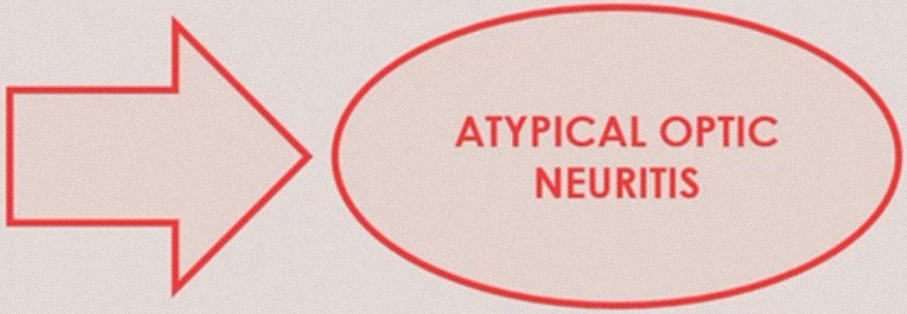
- OPTIC NERVE LESIONS >3mm
- BILATERAL OPTIC NERVE INVOLVEMENT
- OPTIC NERVE SHEATH ENHANCEMENT
- 
- EXTENSION TO CHIASM



# CSF ANALYSIS

1. USUALLY not necessary for diagnosis
2. PRESENCE OF OLIGOCLONAL BANDS correlated with later development of MS

3. HIGH IGg INDEX

# SERUM LABS FOR ANTIBODIES

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| | NMOSD-ON | MOG-ON | MS - ON |
|---|---|---|---|
| **Distribution of ON lesions** | Bilateral | Bilateral | Unilateral |
| **Segment involvement** | Intracranial, chiasmal, optic tract | Retrobulbar | Retrobulbar and canalicular |
| **Length of lesions** | Longitudinally extensive | Longitudinally extensive | Short segment/focal |
| **Degree of ON swelling** | Mild | Severe | Mild |
| **Location of postcontrast enhancement** | Optic nerve | Optic nerve and perineural | Optic nerve |
| **Presence of brain MRI lesions** | Commonly observed | Infrequently observed | Frequently observed |
| **Location/characteristics of brain lesions** | Hypothalamic lesions more common than MOG-ON and MS-ON; posterior fossa and periaqueductal gray | Large, tumefactive lesions; cortical and subcortical lesions | Periventricular, ovoid lesions; subcortical and juxtacortical lesions |

**MOG –Ab** associated optic neuritis-→ MORE RESPONSIVE TO THE STEROIDS with good recovery

BUT

More relapses

ROLE OF LIFELONG IMMUNOSUPPRESSION

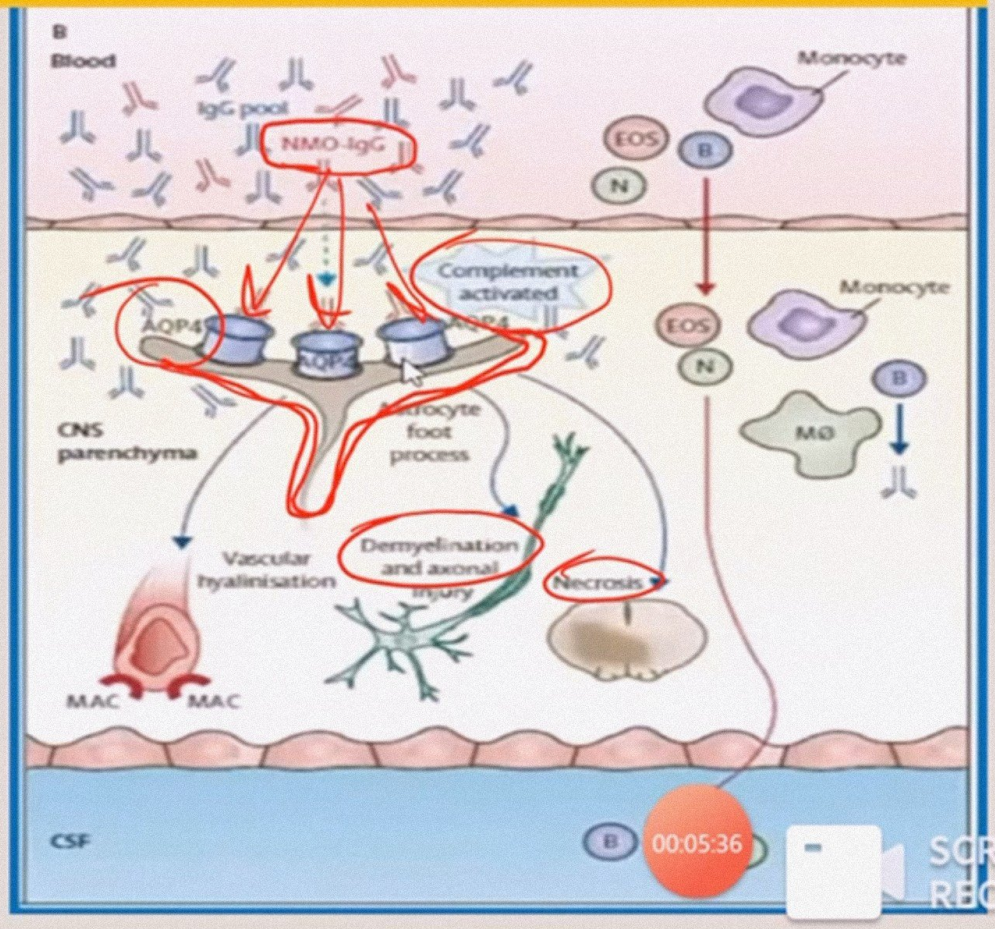
**AQP4 –Ab** has worse prognosis and is MINIMALLY RESPONSIVE TO STEROIDS

PLASMA EXCHANGE
LOMG TERM IMMUNOSUPRESSION ( rituximab, Azathioprine

# NMO :diagnostic criteria

- HISTORY OF OPTIC NEURITIS
- HISTORY OF ACUTE MYELITIS
- Two of the following three :

1. MRI spinal cord > 3 segments
2. NMO- IgG antibody
3. Brain MRI not consistent with MS



# POST NEURITIC OPTIC ATROPHY

- The ophthalmoscopic picture is indistinguishable from that following papilloedema—the disc margins are blurred, the floor has a dirty grey colour and is filled in with organized tissue which extends onto the constricted arteries as perivascular sheaths.

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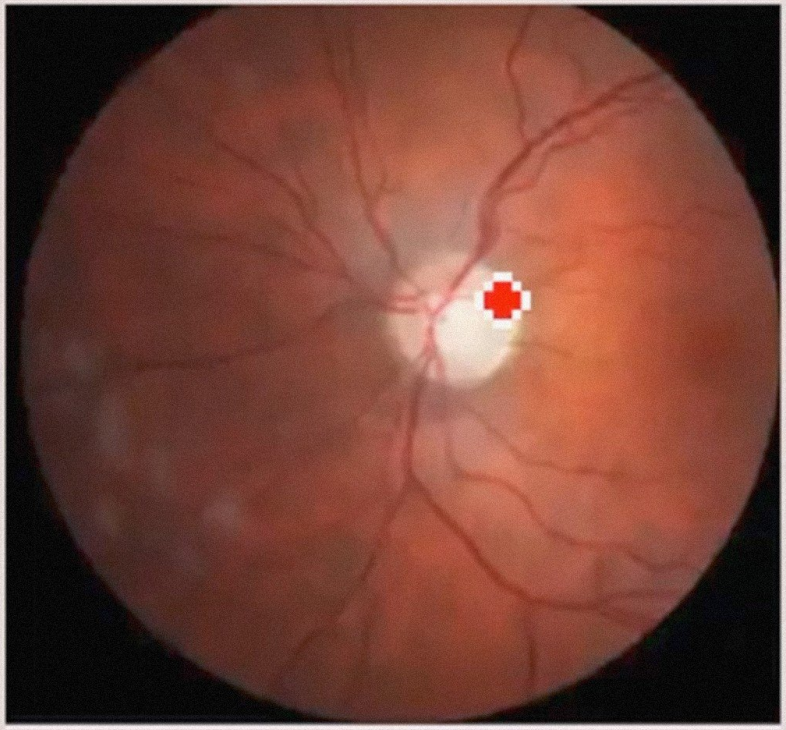
- Acute retrobulbar neuritis produces no ophthalmoscopically visible changes, unless the lesion is near the lamina cribrosa when some signs of papillitis may be seen with distension of the veins and attenuation of the arteries.



→ Retrobulbar

# ADDITIONAL TESTING IN ATYPICAL CASES

- complete blood count
- Eestimation of rapid plasma reagin
- CRP and ESR
- fluorescent treponemal antibody absorption (FTA-ABS) test
- antinuclear antibody (ANA) test.
- For the first episode and in every atypical case, magnetic resonance imaging (MRI) of the brain and orbits with gadolinium enhancement is recommended. The scan helps in predicting the likelihood of multiple sclerosis and ruling out a space-occupying lesion masquerading as optic neuritis
- Patients with demyelination of the central nervous system on MRI or an abnormal neurological examination should be referred to a neurologist for evaluation and management of possible multiple sclerosis.