the FOUNDATION for PERIPHERAL NEUROPATHY®

Welcome!

FPN Webinar:

Idiopathic Neuropathy
with Norman Latov, MD, PhD

Wednesday, February 22, 2023

We will begin our presentation shortly.

the FOUNDATION for PERIPHERAL NEUROPATHY®

Moderator:

Lindsay Colbert
Executive Director
the Foundation *for* Peripheral Neuropathy

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Before We Begin

This presentation is being recorded. The recording link will be emailed to you so you can view it again later.

Submit your questions anytime via the Questions Box. We will try to answer them during this webinar.

If you are having trouble with the audio using your computer, you can dial in by phone (check your email for dial-in instructions).

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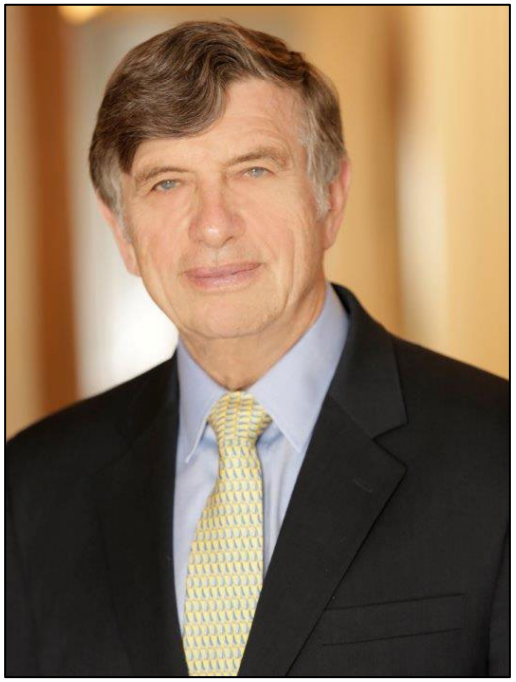


Norman Latov, MD, PhD
Professor of Neurology and Neuroscience
Weill Cornell Medical College

IDIOPATHIC NEUROPATHY

AND WHAT TO DO IF YOU HAVE IT

Norman Latov, MD,PhD
Weill Cornell Medicine

"You have a serious illness of an undisclosed nature."

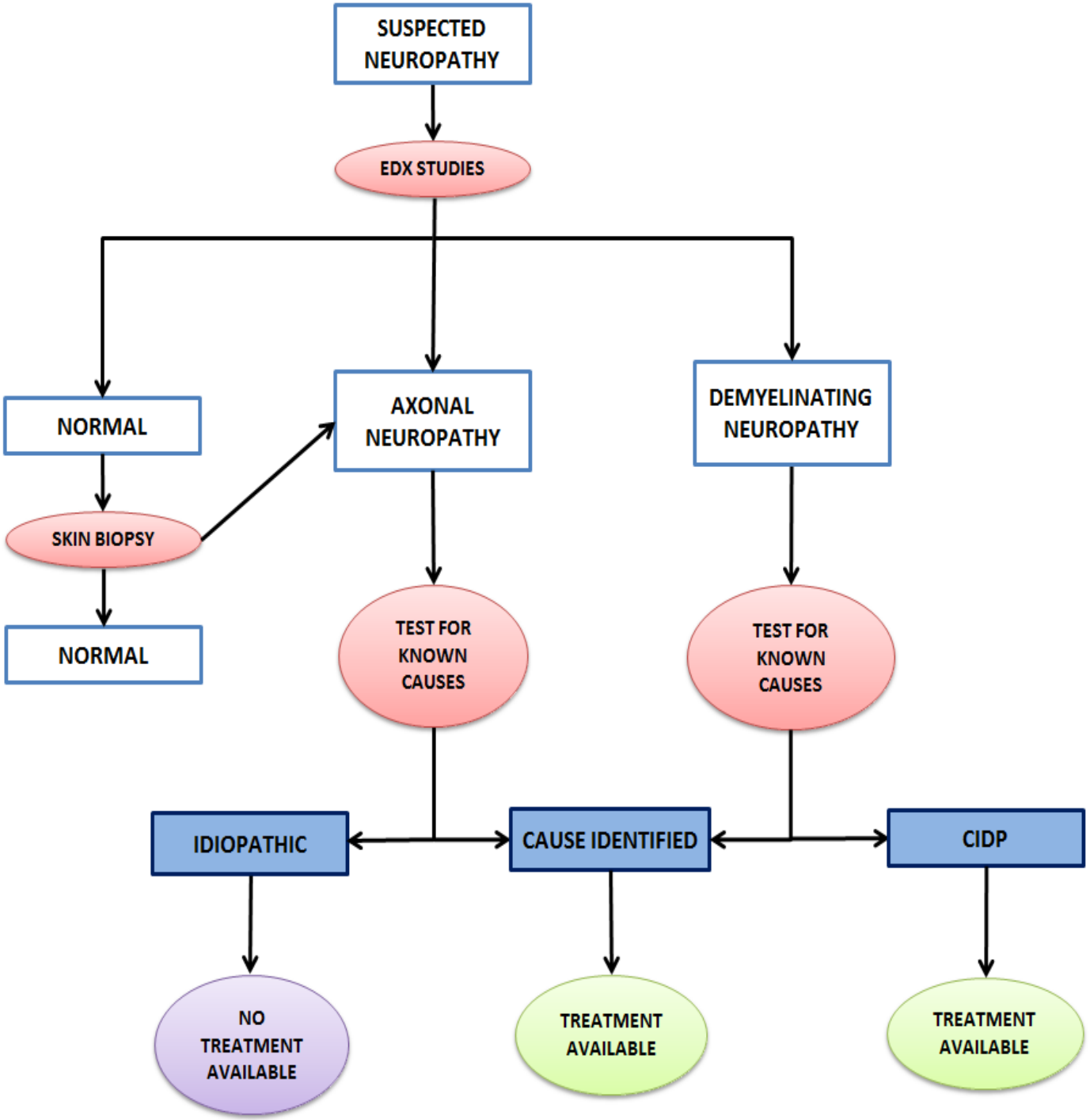
IDIOPATHIC - DEFINITION

- **MEDICAL - RELATING TO OR DENOTING ANY DISEASE OR CONDITION FOR WHICH THE CAUSE IS UNKNOWN.**

- **FROM THE GREEK (idios, one's own) and (páthos, suffering, i.e. disease), or "A DISEASE OF ITS OWN"**

- **CRYPTOGENIC – FROM THE GREEK (crypto, hidden and genic, origin, or "OF HIDDEN ORIGIN" (known or unknown)**

- **FROM A PATIENT – A BLEND WORD COMBINING "IDIOTIC" and "PATHETIC"**


SUSPECTED NEUROPATHY
EDX STUDIES
NORMAL
AXONAL NEUROPATHY
DEMYELINATING NEUROPATHY
SKIN BIOPSY
NORMAL
TEST FOR KNOWN CAUSES
TEST FOR KNOWN CAUSES
IDIOPATHIC
CAUSE IDENTIFIED
CIDP
NO TREATMENT AVAILABLE
TREATMENT AVAILABLE
TREATMENT AVAILABLE

CAUSES OF DEMYELINATING NEUROPATHY

- **IMMUNE MEDIATED**
  - ACUTE INFLAMMATORY DEMYELINATING POLYNEUROPATHY (GUILLAIN BARRE SYNDROME, GBS)
  - CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP)
  - MULTIFOCAL MOTOR NEUROPATHY (MMN)
  - MAG NEUROPATHY
  - POEMS SYNDROME
- **HEREDITARY DEMYELINATING NEUROPATHY**
- **DRUG INDUCED -** AMIODARONE

CAUSES OF AXONAL NEUROPATHY

- **ENDOCRINE AND METABOLIC -** DIABETES , HYPOTHYROID, RENAL FAILURE
- **INFECTIOUS -** LYME, HEPATITIS C, HIV-1/2, …
- **AUTOIMMUNE –** AXONAL GUILLAIN BARRE, SJOGREN, VASCULITIS, CELIAC DISEASE, SARCOID …
- **PARANEOPLASTIC –** ASSOCIATED WITH CANCER (LUNG, LYMPHOMA, MYELOMA, IgM MONOCLONAL GAMMOPATHIES
- **NUTRITIONAL –** ALCOHOL TOXICITY, B12 OR B1 DEFICIENCY, B6 DEFICIENCY OR TOXICITY
- **TOXIC –** LED, MECRCURY (SEA FOOD)…
- **DRUG INDUCED –** CHEMOTHRAPY, INH, CHECK POINT INHIBITORS…
- **HEREDITARY AXONAL NEUROPATHY**

WHAT ELSE CAN IT BE

- **IF STABLE**
  - RESIDUAL SYMPTOMS FROM A PAST INSULT
    - GUILLAIN BARRE SYNDROME, CIDP IN REMISSION, LYME DISEASE , ACUTE VIRAL ILLNESS, DRUG OR TOXIN INDUCED, OTHER INFLAMMATORY CONDITIONS, etc.
    - COMMON IN SMALL FIBER NEUROPATHIES
- **IF PROGRESSIVE**
  - DIAGNOSIS CAN BE MISSED IF NOT TESTED FOR
  - AN AS YET UNIDENTIED GENETIC MUTATION
  - NON-SYSTEMIC VASCULITIS OR SARCOIDOSIS
  - ATYPICAL CIDP NOT MEETING ELECTRODIAGNOSTIC CRITERIA
  - PRIMARY (LIGHT CHAIN) AMYLOIDOSIS

HOW TO TELL IF THE NEUROPATHY IS WORSENING (PROGRESSIVE)

- FLUCTUATING SYMPTOMS MOST OFTEN INDICATE A STABLE UNDERLYING NEUROPATHY; FLUCTURATIONS CAN BE DUE TO ENVIRONMENTAL OR OTHER FACTORS
- MOTOR FUNCTIONS ARE MORE RELIABLE MEASURES OF PROGRESSION THAN SENSORY SYMPTOMS.
- SENSORY SYMPTOMS ARE LESS CONSISTENT OR RELIABLE
- SUSPECTED PROGRESSION NEEDS TO BE CONFIRMED BY NEUROLOGICAL EXAMINATION AND ELECTRODIAGNOSTIC STUDIES

MEASURING MOTOR FUNCTIONS

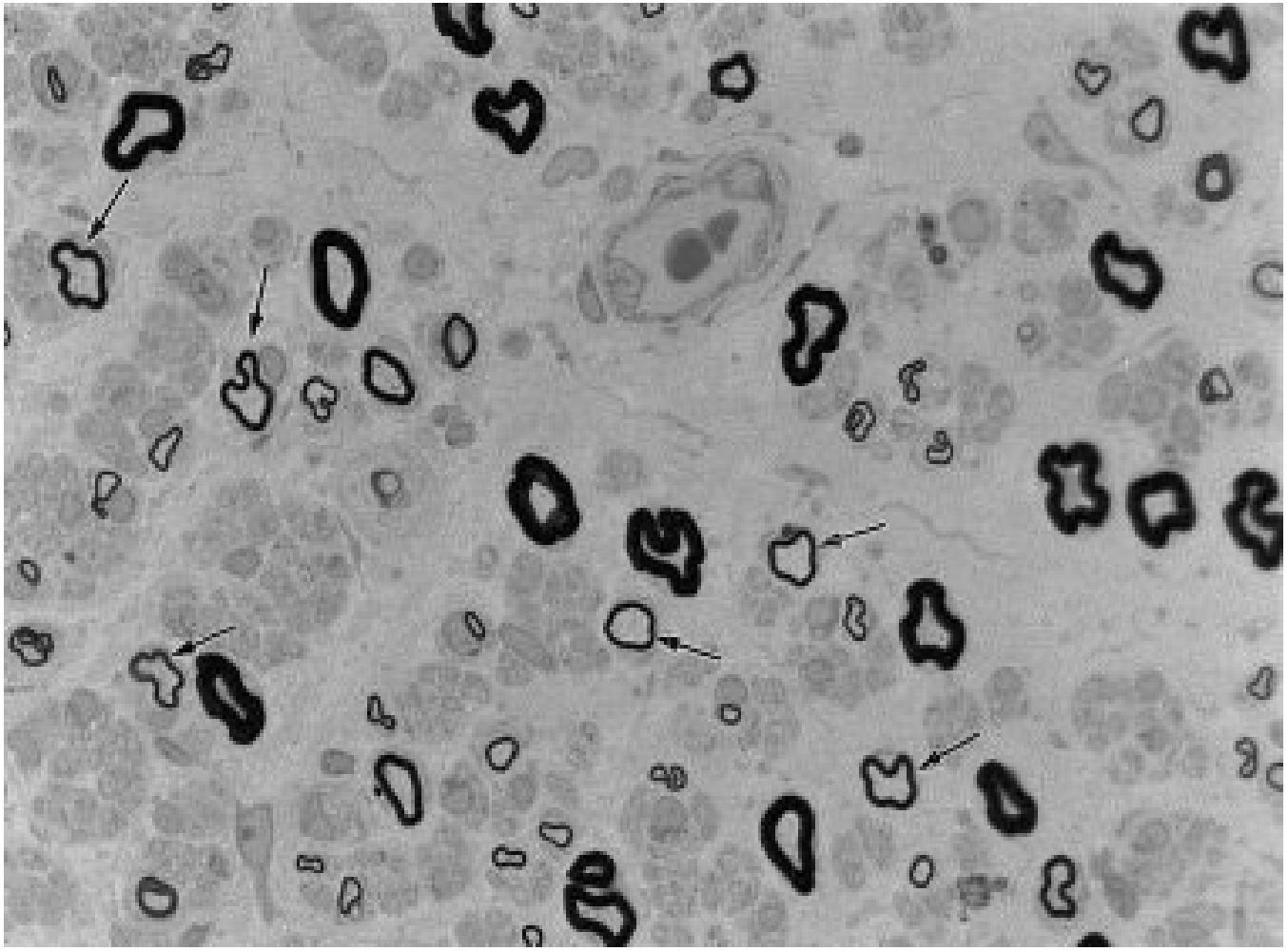
- GRIP STRENGTH
  - USE HAND- HELD DYNAMOMETER
- LEG STRENGTH – MOST SENSITIVE FUNCTIONS ARE THOSE THAT CAN BE DONE WITH SOME DIFFICULTY
  - WALKING ON THE HEELS OR TOES, GETTING UP FROM A CHAIR OR FROM A KNEELING POSITION WITH EITHER LEG, WALKING UP OR DOWN STAIRS, RUNNING
- WALKING SPEED
  - MEASURE ABOUT 25 ft, WALK IT AS FAST AS YOU CAN AND TIME YOURSELF, REPEAT 3 TIMES AND AVERAGE, LOG THE RESULTS, REPEAT ONCE OR TWICE A WEEK

GRIP STRENGTH

WHAT TO DO IF THE NEUROPATHY IS PROGRESSIVE

- REPEAT TESTING
  - EMG AND NC STUDIES MIGHT REVEAL DEMYELINATING CHANGES INDICATIVE OF CIDP THAT WERE PREVIOUSLY UNDETECTED
  - PREVIOUSLY BORDERLINE ABNORMALITIES MIGHT BECOME MORE OBVIOUS
  - IF UNCHANGED – CONSIDER MYELOPATHY OR MYOPATHY
- GENETIC TESTING, IF NOT ALREADY DONE, EVEN IF THERE IS NO FAMILY HX OF NEUROPATHY
- NERVE AND MUSCLE BIOPSY CAN REVEAL A DIAGNOSIS IN APPROXIMATELY 20% OF CASES
  - NON-SYSTEMIC VASCULITIS, ATYPICAL CIDP, PRIMARY (LIGHT CHAIN) AMYLOIDOSIS, NON- SYSTEMIC SARCOIDOSIS
  - NEEDS TO BE DONE AT SPECIALIZED REFERRAL CENTERS

Figure 1. Sural nerve biopsy. Significant loss of myelinated fibers. Several myelin sheaths are too thin with respect to the diameters of the axons (arrows). (Original magnification, ×40, Vallat et al, M&N 2003;27:478-485.)



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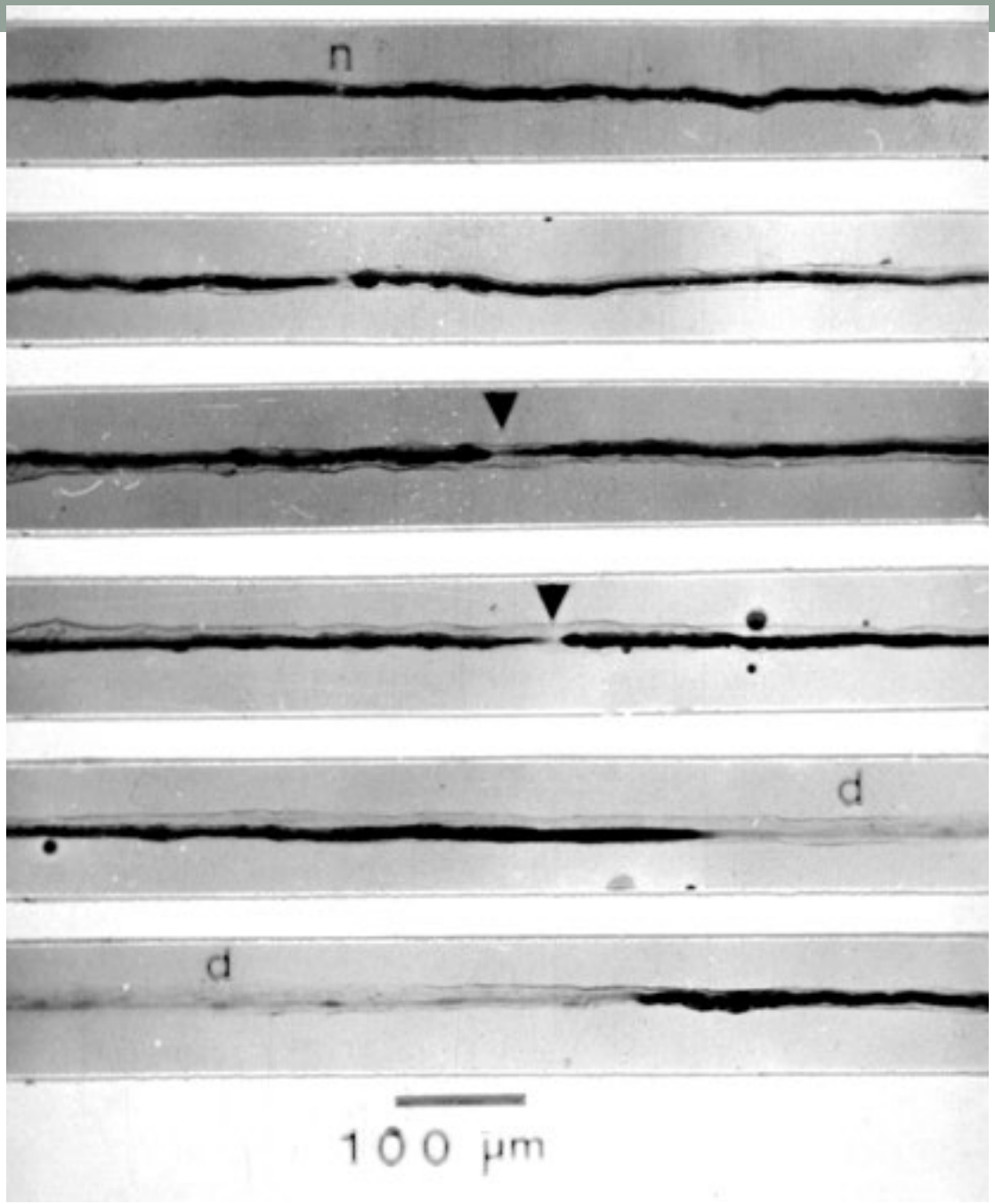
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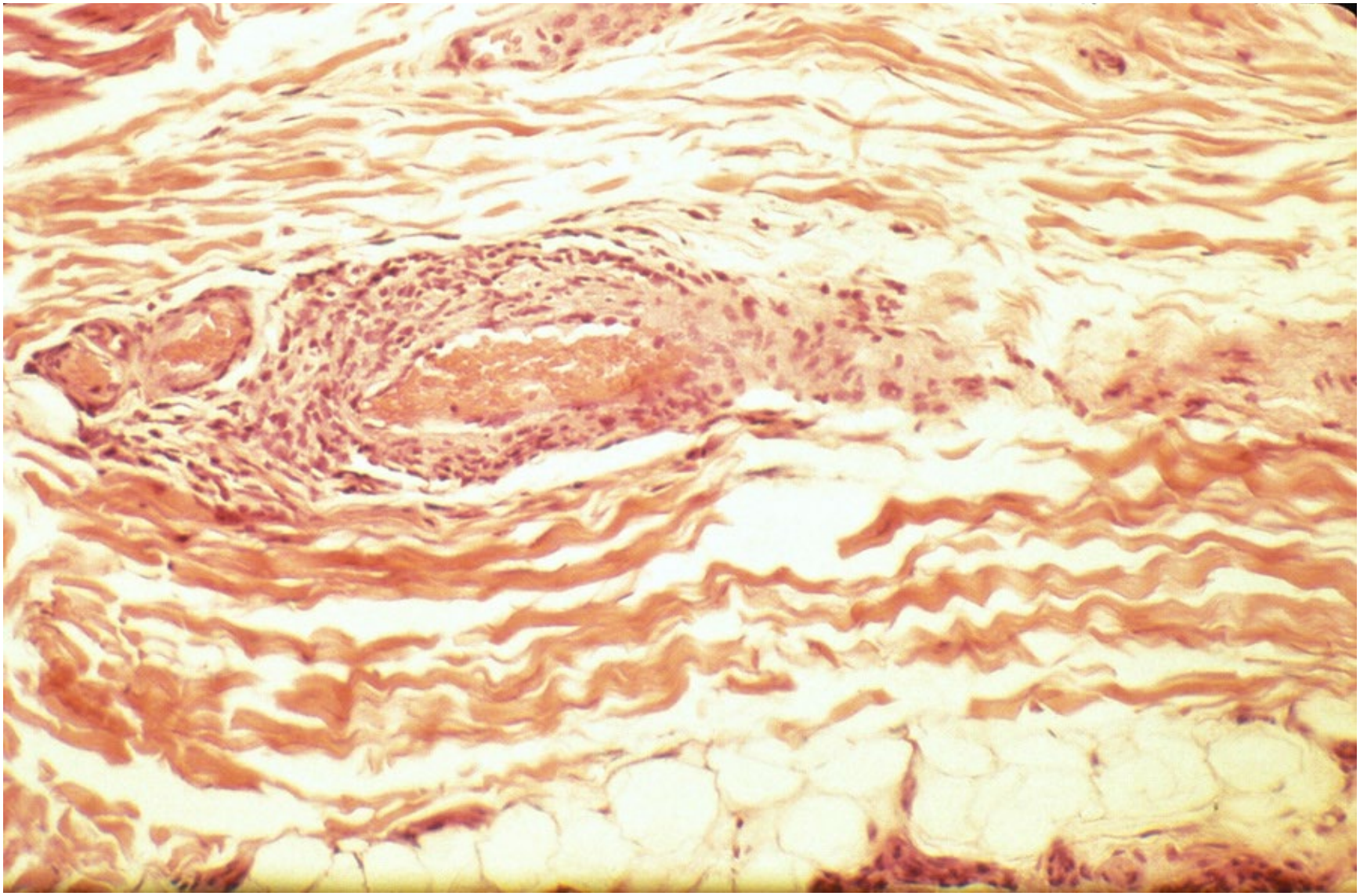
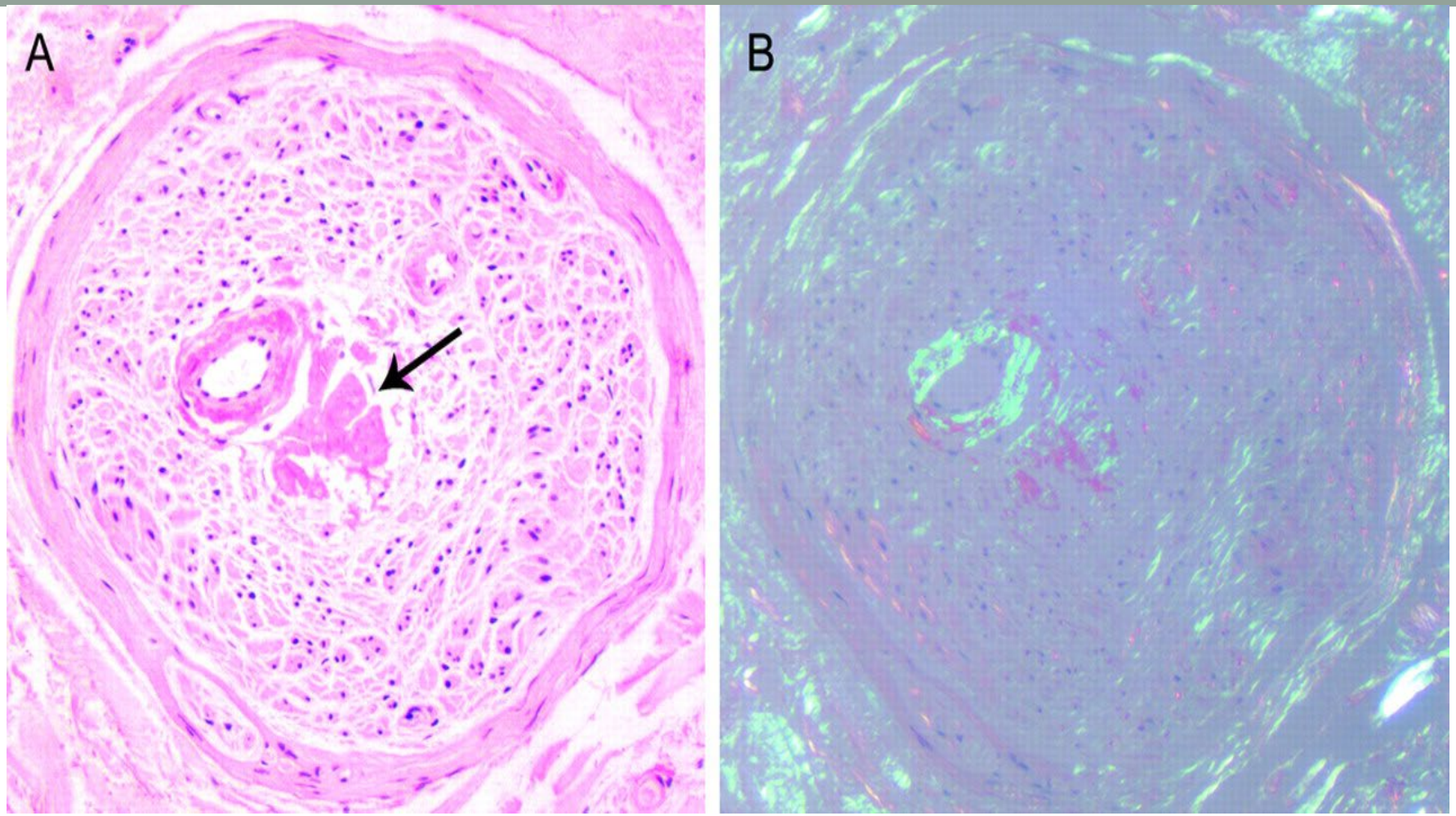
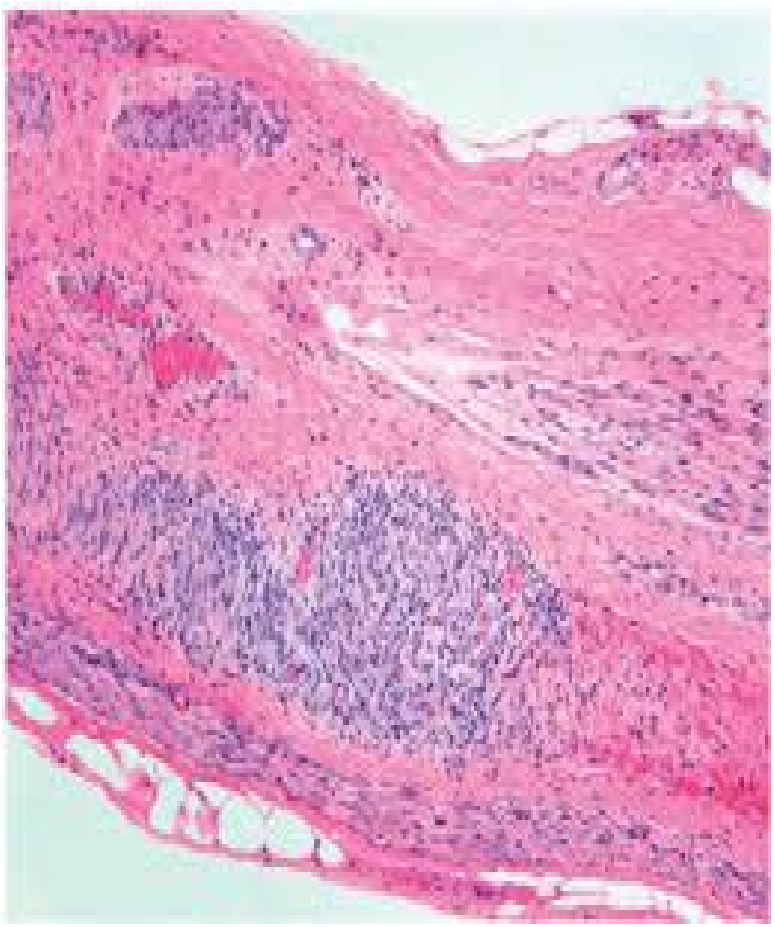
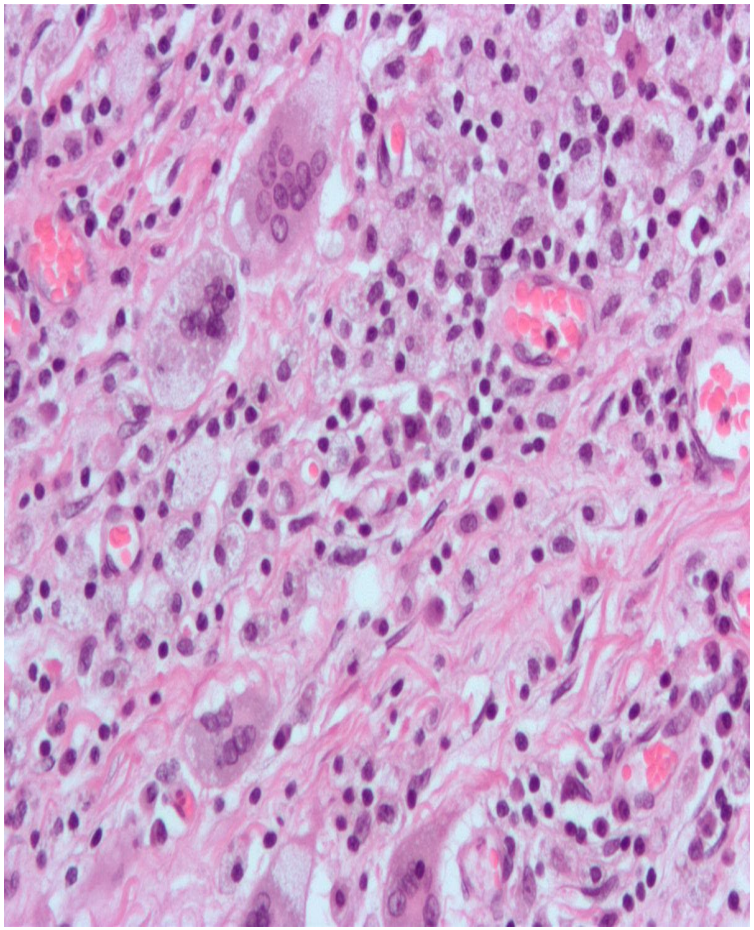
NERVE WITH VASCULITIS





AMYLOID NEUROPATHY





SARCOID GRANULOMA IN NERVE

WHAT TO DO IF THE NEUROPATHY IS PROGRESSIVE AND ALL TESTS INCLUDING NERVE BIOPSY ARE NEGATIVE

- **COULD IT BE AUTOIMMUNE ?**
  - THERE IS NO CONSENSUS
  - SOME PHYSICIANS WOULD CONSIDER “OFF LABEL” (EXPERIMENTAL) THERAPY WITH CORTICOSTEROIDS, IVIg, OR RITUXIMAB (ANTI B-CELL ANTIBODY), DEPENDING ON THE CLINICAL PRESENTATION (RAPID PROGRESSION, MULTIFOCALITY), RISK TOLERANCE, AND INSURANCE APPROVAL

ALTERNATIVE TREATMENTS

- THEY ARE CALLED “ALTERNATIVE” BECAUSE THEY ARE UNPROVEN
- MOST ARE NATURALLY OCCURING AND NOT PROPIETARY, SO THAT THEY ARE NOT REGULATED OR FUNDED FOR CLINICAL TRIALS
- IT WOULD BE USEFUL TO ESTABLICH AN ON-LINE DATA BASE WHERE RESPONSES CAN BE TABULATED AND COMPARED

WHAT ELSE CAN BE DONE

- **SUPPORTIVE THERAPIES**
  - **PHYSICAL THERAPY, ORTHOTICS, AIDS TO AMBULATION**
  - **TREATMENT OF NEUROPATHIC PAIN**
  - **TREATMENT OF AUTONOMIC SYMPTOMS**

- **RESEARCH IS HOPE**
  - **DIRECTLY SUPPORT RESEARCH**
  - **FORM OR JOIN POLITICAL ACTION COMMITTEES TO ADVOCATE FOR RESEARCH FUNDING BY THE NIH**
    - "ALL POLITICS ARE LOCAL" – Tip O'Neill
    - REQUEST THAT THE CONGRESSIONAL APPROPRIATION COMMITTEES DIRECT THE NIH TO REPORT ON FUNDING FOR NEUROPATHY RESEARCH

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Questions?

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