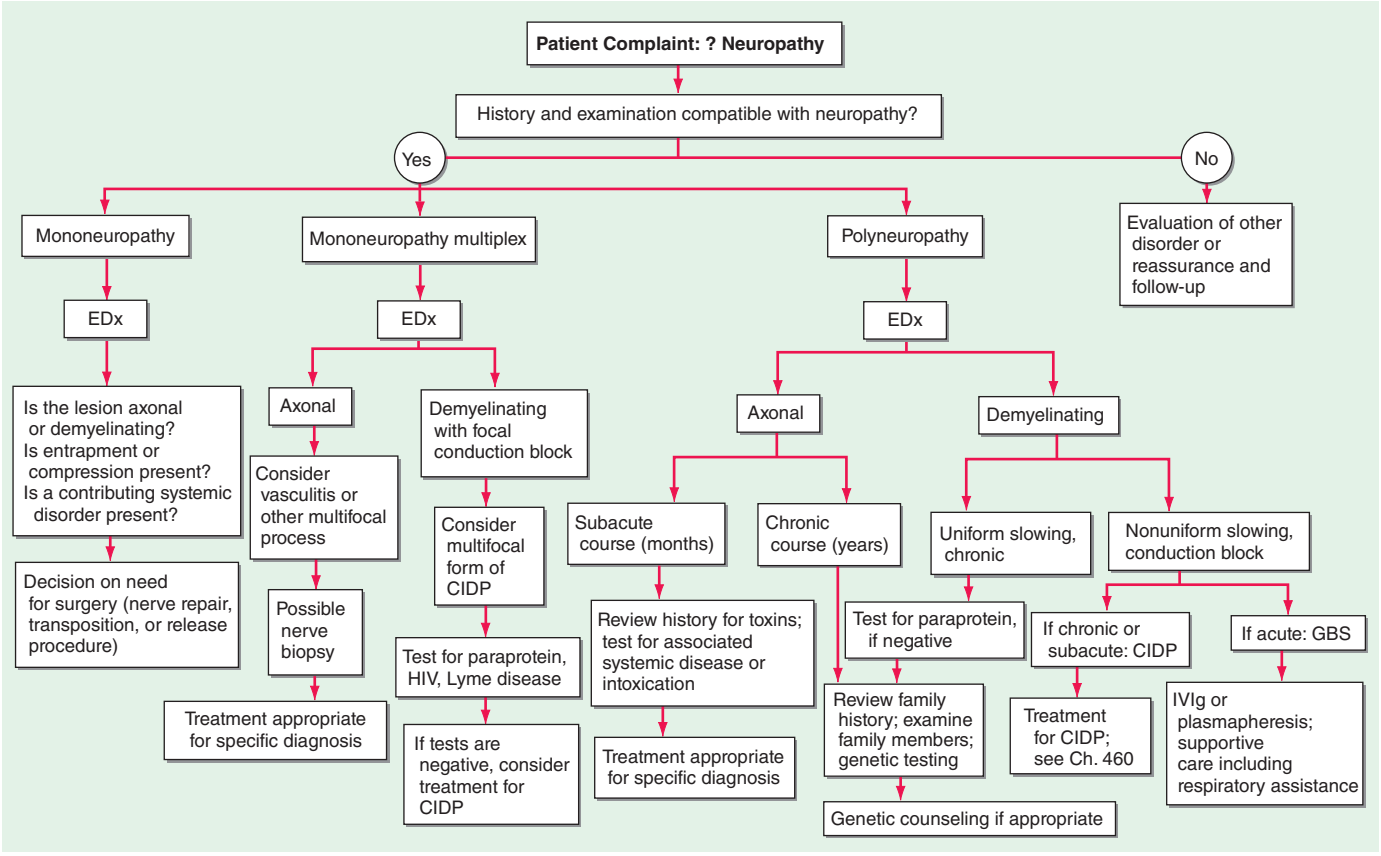

FIGURE 459-1 Approach to the evaluation of peripheral neuropathies. CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; EDx, electrodiagnostic; GBS, Guillain-Barré syndrome; IVIg, intravenous immunoglobulin.

alert the clinician to the possibility of amyloid polyneuropathy. Rarely, a pandysautonomic syndrome can be the only manifestation of a peripheral neuropathy without other motor or sensory findings. The majority of neuropathies are predominantly sensory in nature.

TABLE 459-1 APPROACH TO NEUROPATHIC DISORDERS: SEVEN KEY QUESTIONS

1. What systems are involved?
- Motor, sensory, autonomic, or combinations

2. What is the distribution of weakness?
- Only distal versus proximal and distal
- Focal/asymmetric versus symmetric

3. What is the nature of the sensory involvement?
- Temperature loss or burning or stabbing pain (e.g., small fiber)
- Vibratory or proprioceptive loss (e.g., large fiber)

4. Is there evidence of upper motor neuron involvement?
- Without sensory loss
- With sensory loss

5. What is the temporal evolution?
- Acute (days to 4 weeks)
- Subacute (4–8 weeks)
- Chronic (>8 weeks)
- Monophasic, progressive, or relapsing-remitting

6. Is there evidence for a hereditary neuropathy?
- Family history of neuropathy
- Lack of sensory symptoms despite sensory signs

7. Are there any associated medical conditions?
- Cancer, diabetes mellitus, connective tissue disease or other autoimmune diseases, infection (e.g., HIV, Lyme disease, leprosy)
- Medications including over-the-counter drugs that may cause a toxic neuropathy
- Preceding events, drugs, toxins

2. What is the Distribution of Weakness? Delineating the pattern of weakness, if present, is essential for diagnosis, and in this regard two additional questions should be answered: (1) Does the weakness only involve the distal extremity, or is it both proximal and distal? and (2) Is the weakness focal and asymmetric, or is it symmetric? Symmetric proximal and distal weakness is the hallmark of acquired immune demyelinating polyneuropathies, both the acute form (acute inflammatory demyelinating polyneuropathy [AIDP], also known as Guillain-Barré syndrome [GBS]) and the chronic form (chronic inflammatory demyelinating polyneuropathy [CIDP]). The importance of finding symmetric proximal and distal weakness in a patient who presents with both motor and sensory symptoms cannot be overemphasized because this identifies the important subset of patients who may have a treatable acquired demyelinating neuropathic disorder (i.e., AIDP or CIDP).

Findings of an asymmetric or multifocal pattern of weakness narrow the differential diagnosis. Some neuropathic disorders may present with unilateral extremity weakness. In the absence of sensory symptoms and signs, such weakness evolving over weeks or months would be worrisome for motor neuron disease (e.g., amyotrophic lateral sclerosis [ALS]), but it would be important to exclude multifocal motor neuropathy that may be treatable (Chap. 452). In a patient presenting with asymmetric subacute or acute sensory and motor symptoms and signs, radiculopathies, plexopathies, compressive mononeuropathies, or multiple mononeuropathies (e.g., mononeuropathy multiplex) must be considered.

3. What is the Nature of the Sensory Involvement? The patient may have loss of sensation (numbness), altered sensation to touch (hyperpathia or allodynia), or uncomfortable spontaneous sensations (tingling, burning, or aching) (Chap. 31). Neuropathic pain can be burning, dull, and poorly localized (protopathic pain), presumably transmitted by polymodal C nociceptor fibers, or sharp and lancinating (epicritic pain), relayed by A-delta fibers. If pain and temperature perception are lost, while vibratory and position sense are preserved along with