

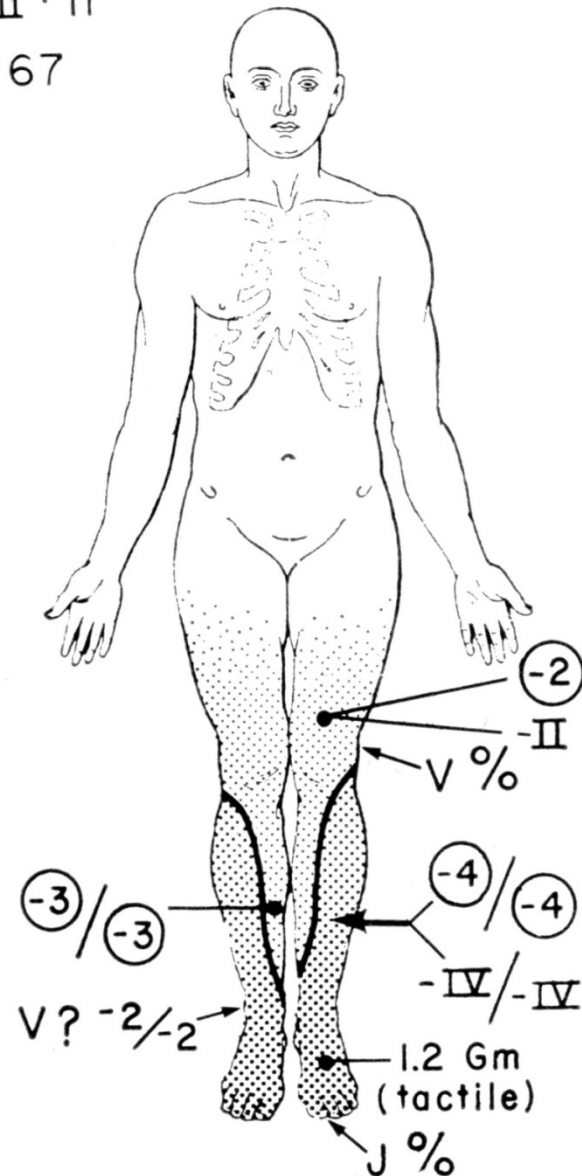
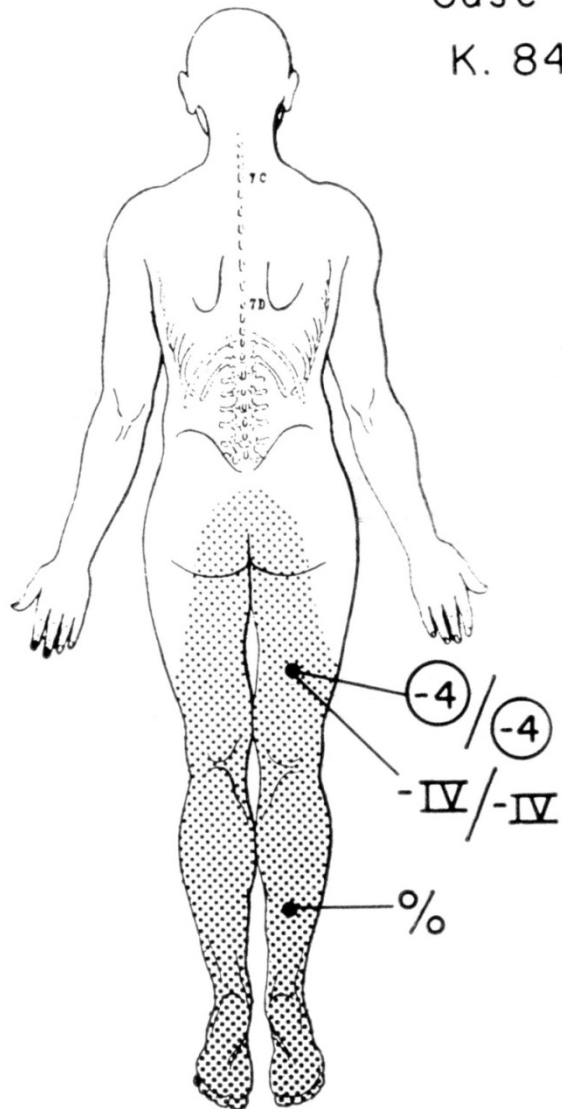


Historic patient I saw approximately 40 years ago with painless injury of the foot and who then developed incontinence of urine and bowel motions.

The patient was of Greek-American descent and the disorder was inherited from a parent to half of the children (on average) typical of autosomal dominant inheritance.

Case III · 11

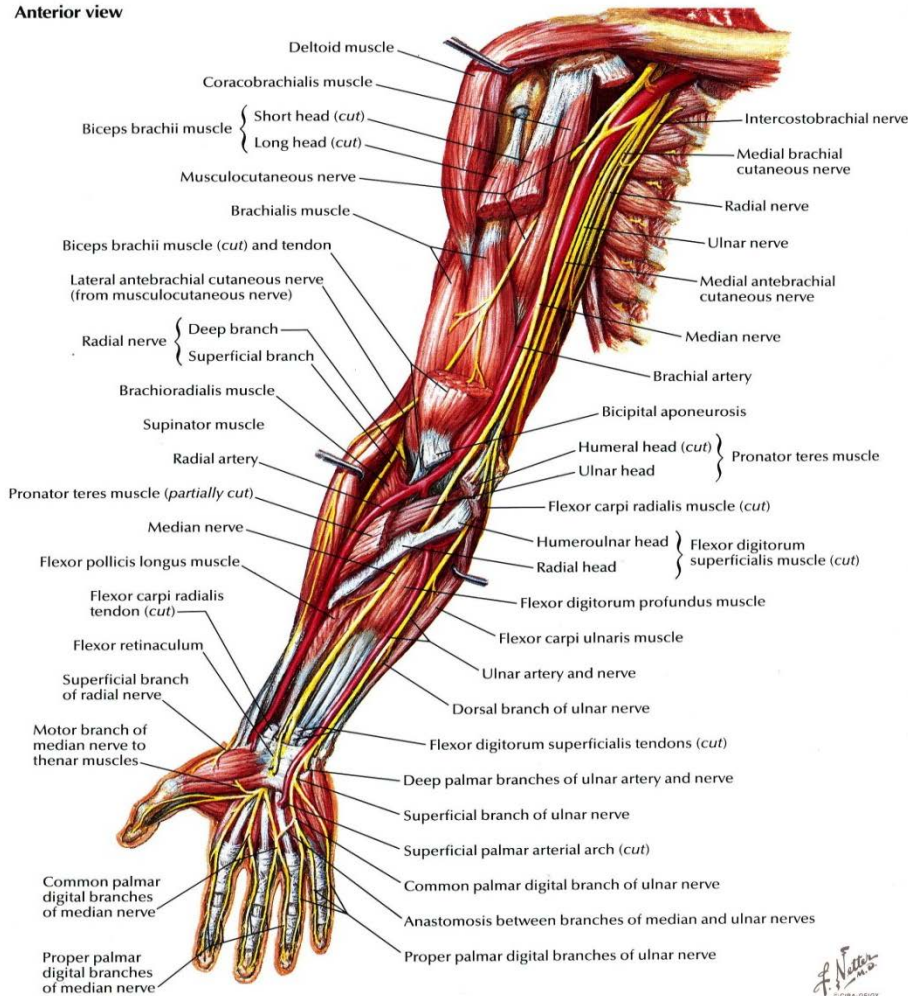
K. 84-67



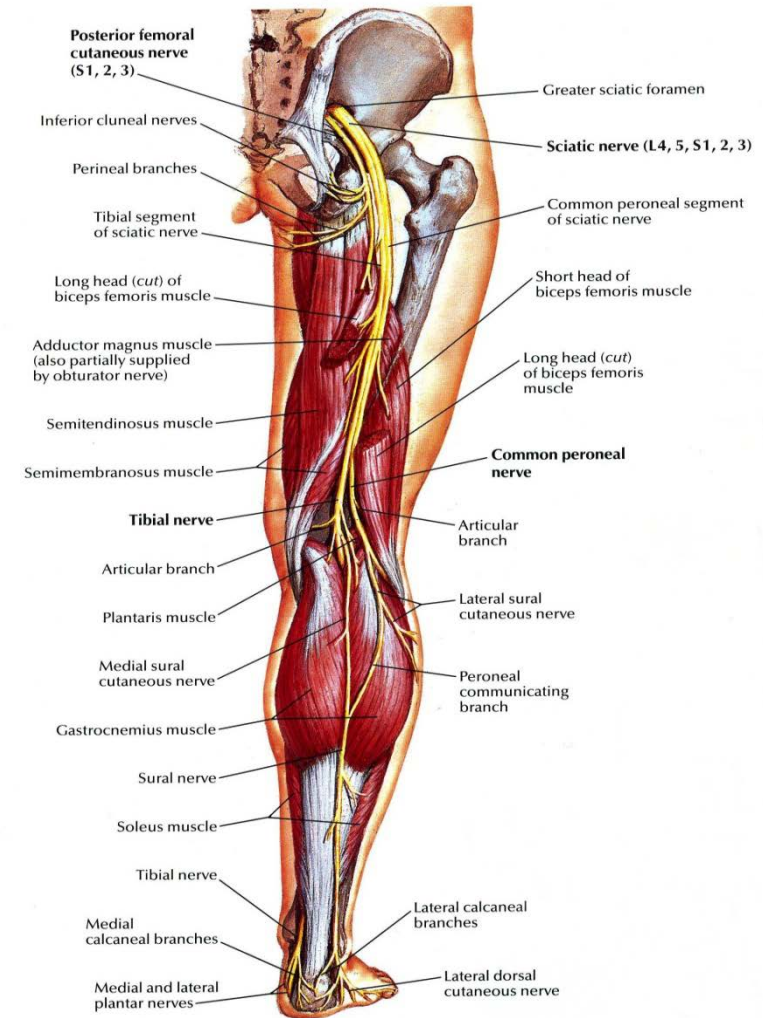
The sensation loss over the feet, legs, thighs and saddle area is shown and it was mainly of small fiber function, i.e., pain and temperature sensation.

Arteries and Nerves of Upper Limb

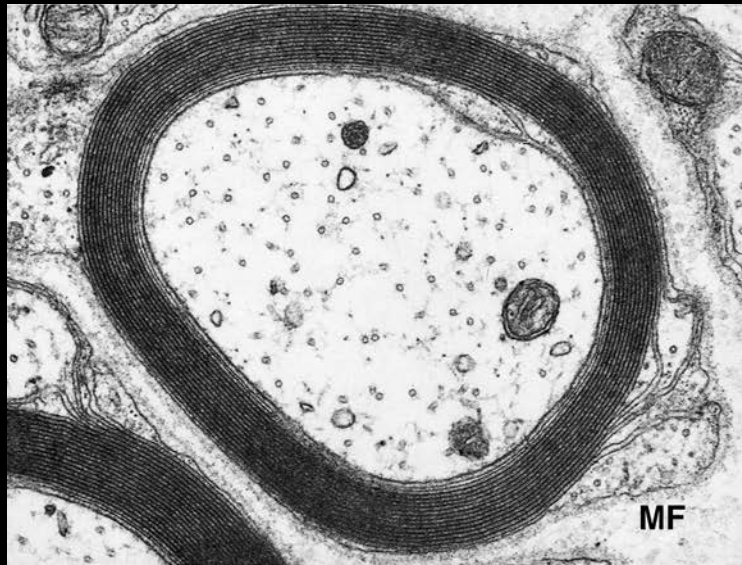
Anterior view



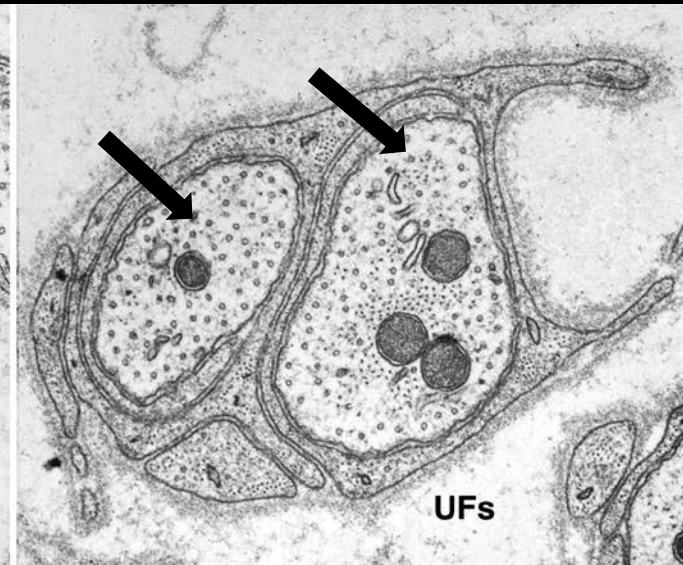
Sciatic Nerve and Posterior Femoral Cutaneous Nerve



This illustration taken from Netter's Atlas shows the nerves (in yellow) of the upper and lower limbs.

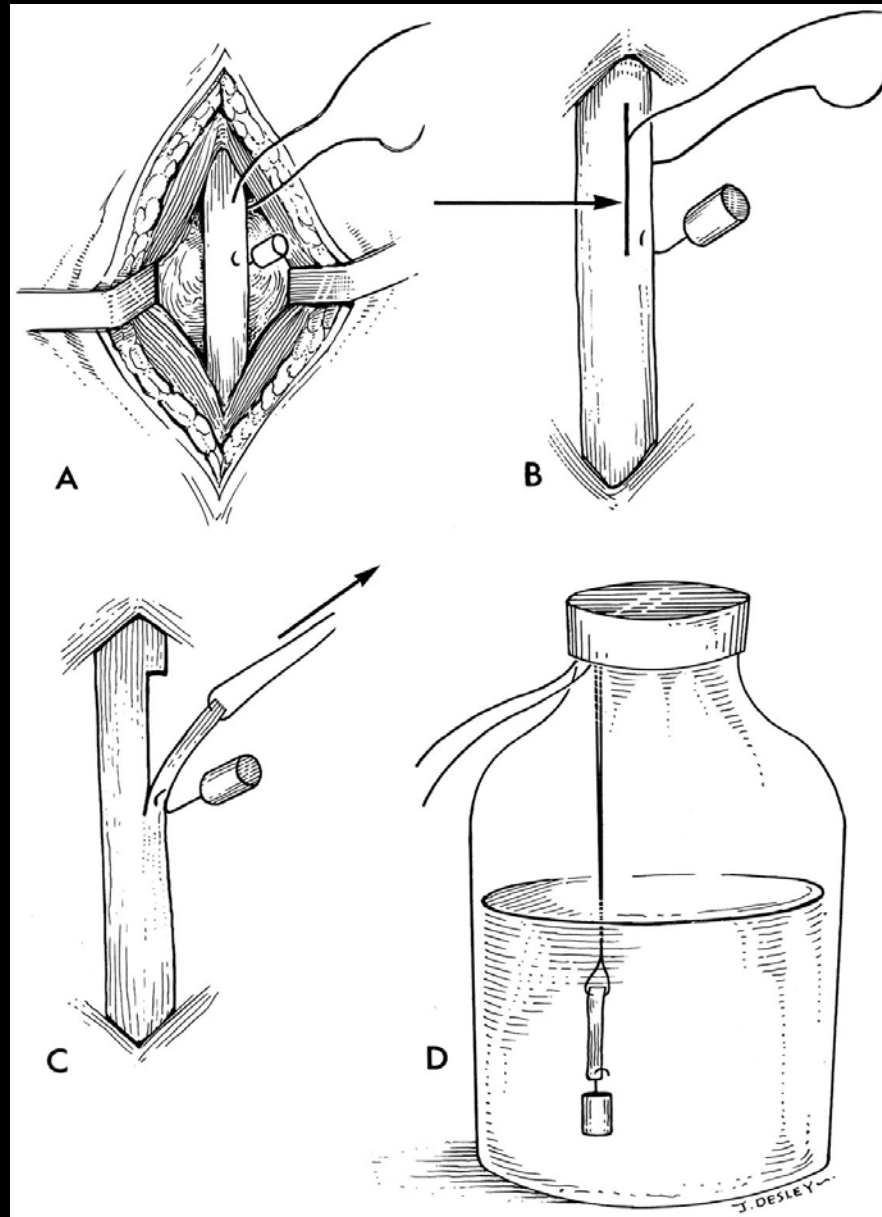


A myelinated nerve fiber



Two unmyelinated nerve fibers

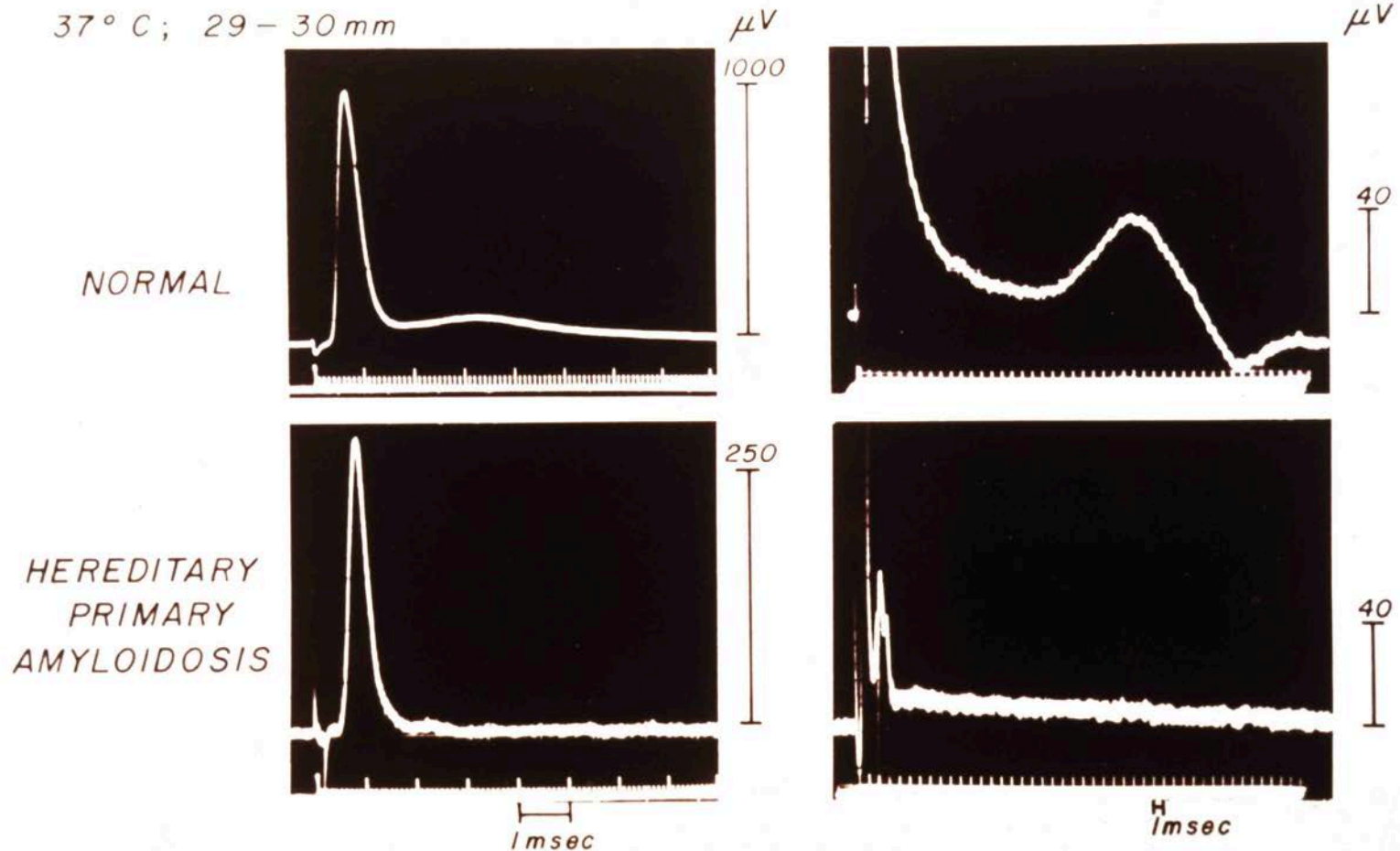
Myelinated fibers are covered by a protein fatty material called myelin (the black rim surrounding the fiber). Note that the two unmyelinated nerve fibers (arrows) are also surrounded by another cell, the Schwann cell.



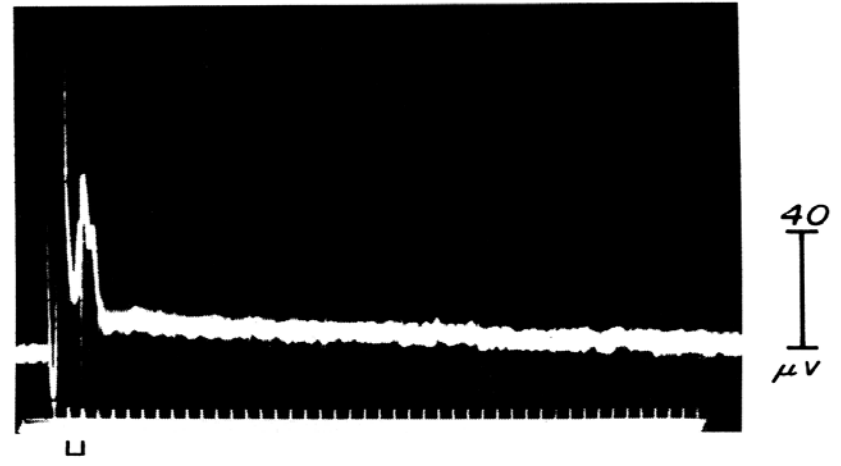
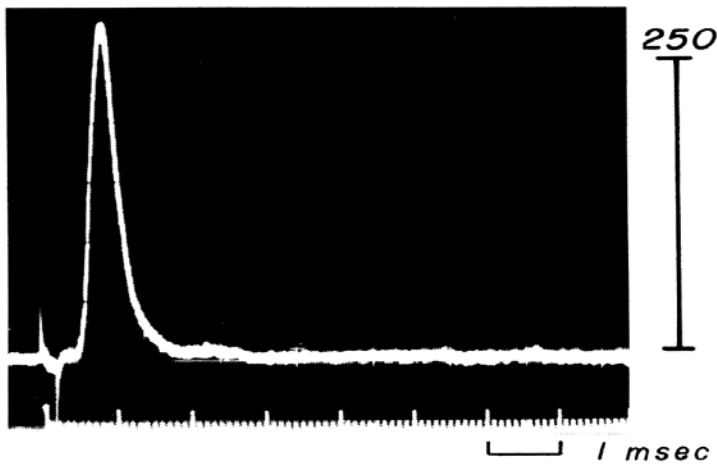
This slide shows the steps in taking a fascicular nerve biopsy (an historic drawing by Dyck and Lofgren about 40 years ago).

ACTION POTENTIALS OF HUMAN SURAL NERVE, IN VITRO

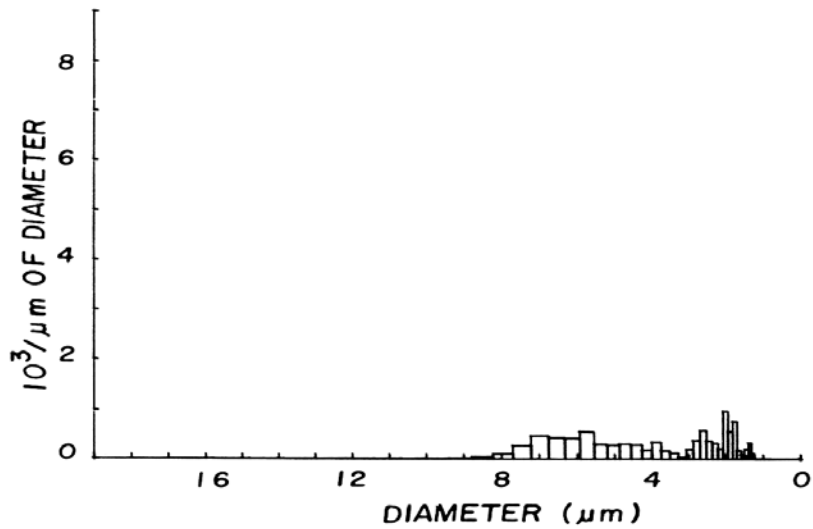
37° C; 29 - 30 mm



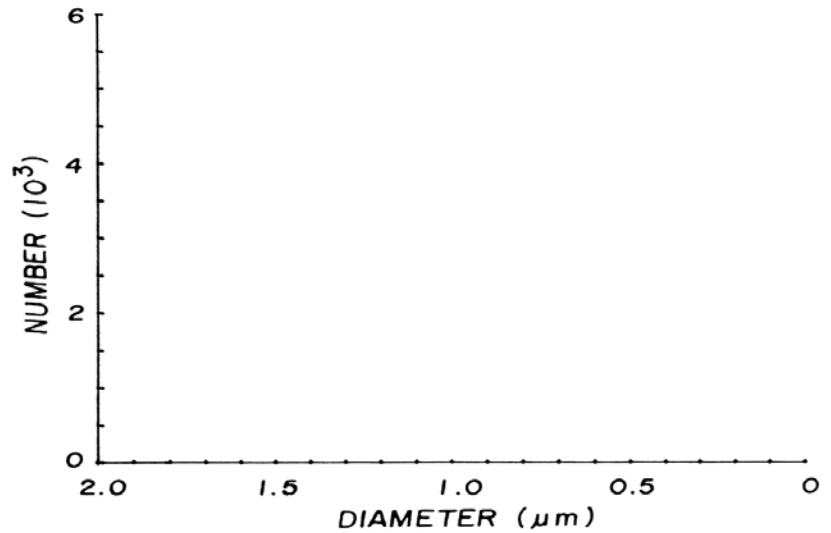
The upper two tracings are from physiological recordings of three peaks of a healthy sural nerve. From left to right they are referred to as alpha beta, delta and C. In the tracings below, the action potentials of our patient's nerve with TTR amyloid neuropathy is shown. Only reduced numbers of alpha-beta fibers were present; no delta fibers or C fibers. This degree of selective involvement of small fibers is actually somewhat atypical for TTR amyloid polyneuropathy. Most patients with this disease have a greater involvement also of large fibers.



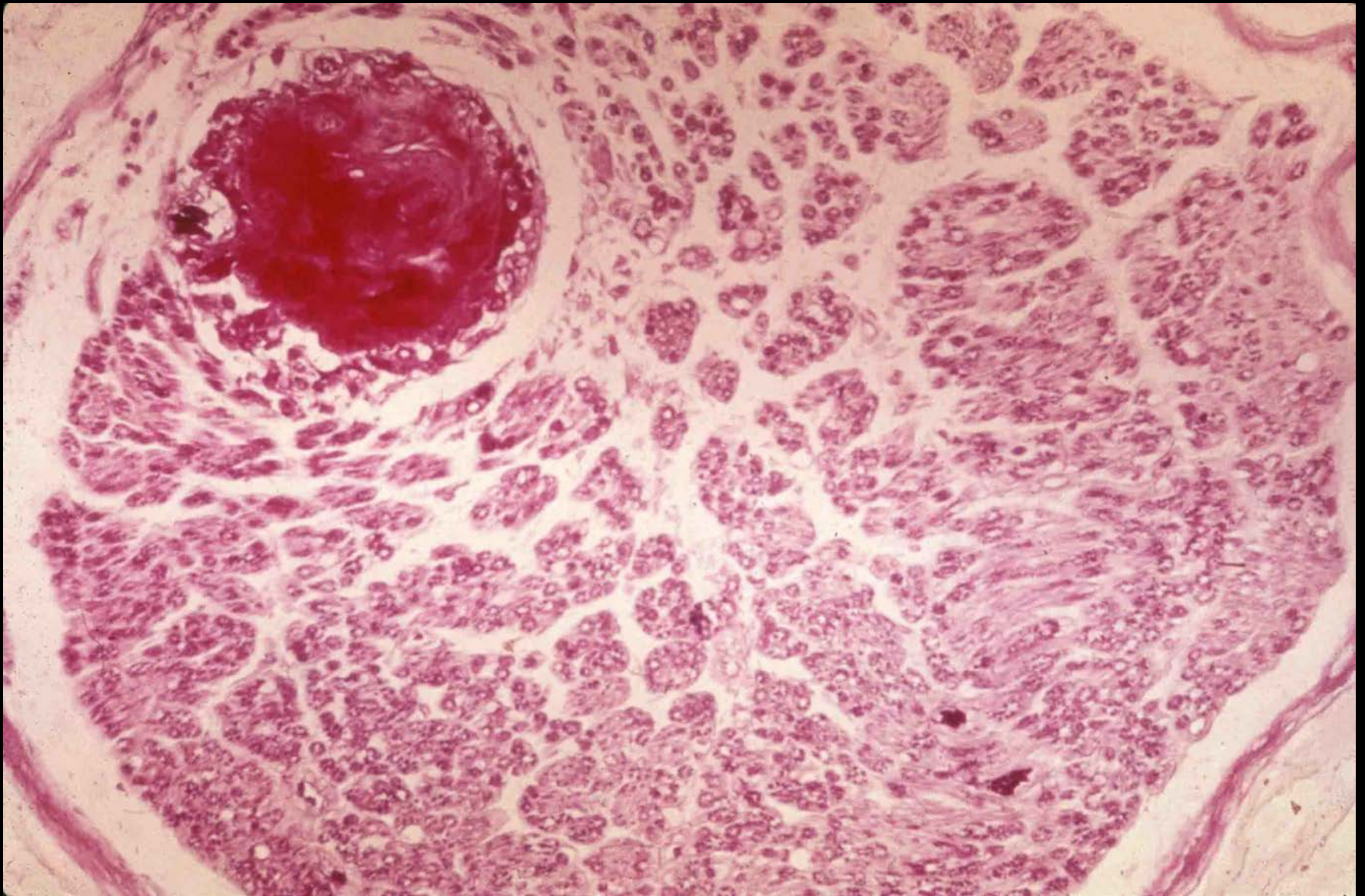
A



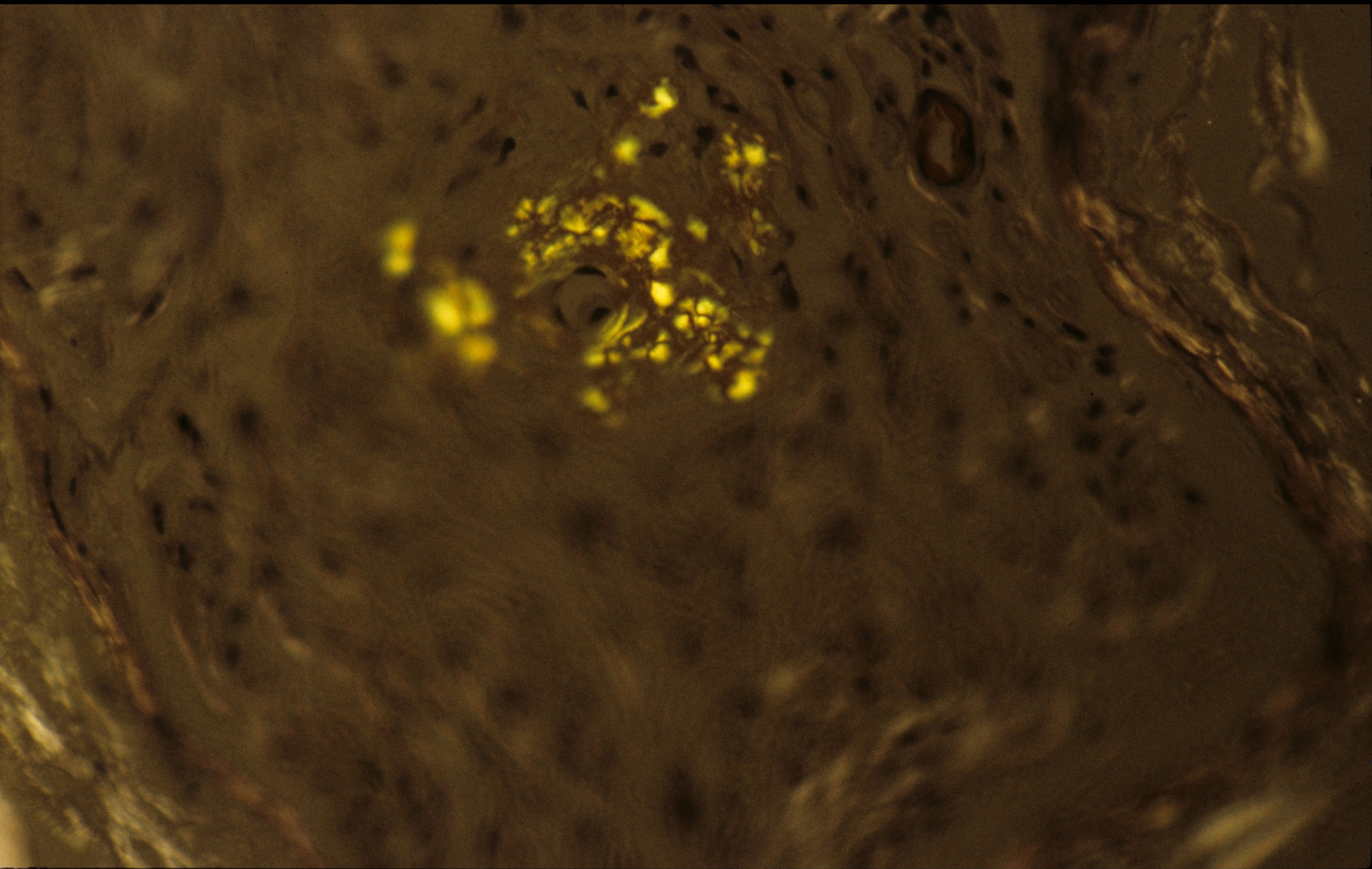
B



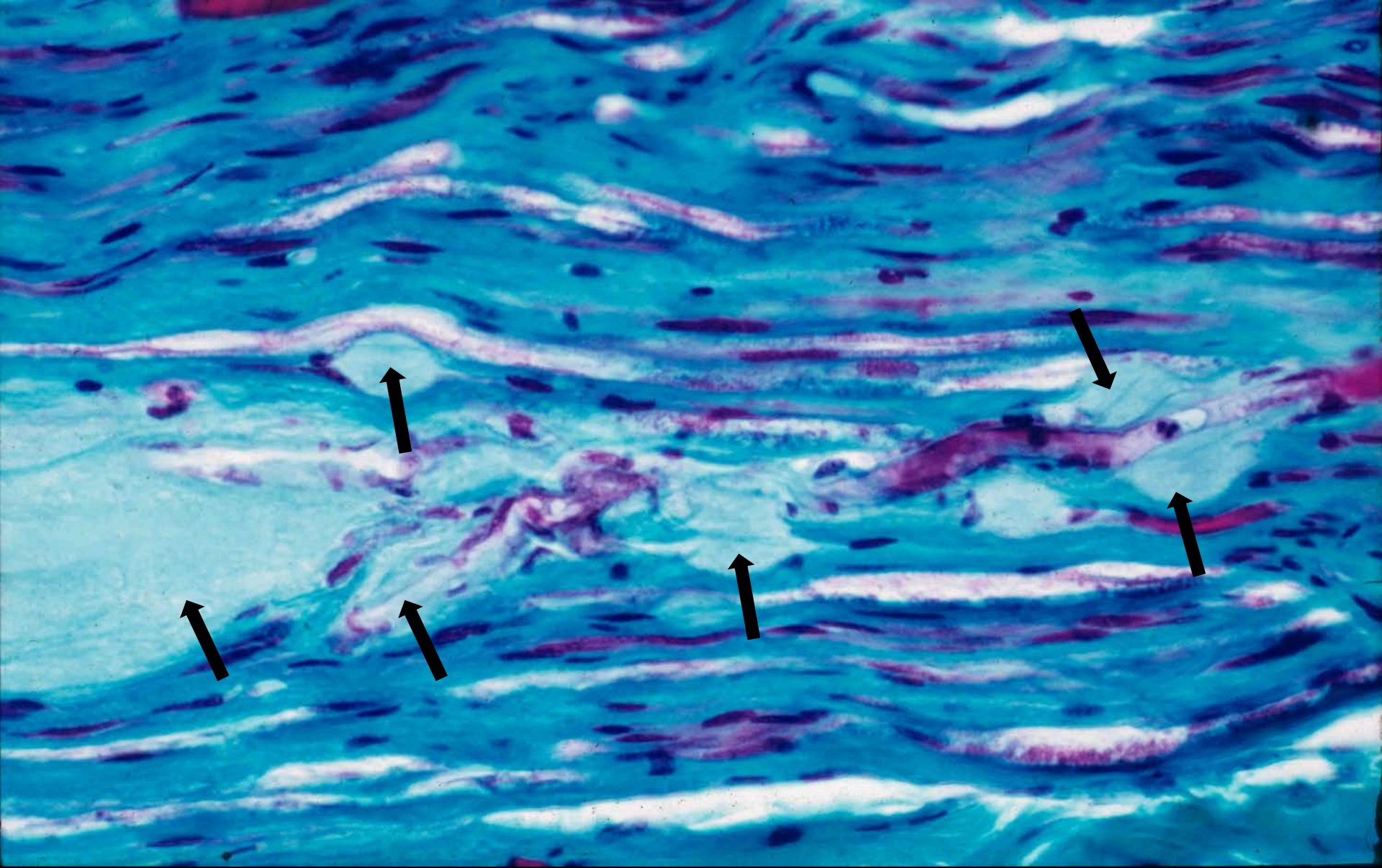
In microscopic measurements, the severe absence of unmyelinated and small myelinated fibers was confirmed. Small myelinated fibers and unmyelinated fibers were essentially absent.



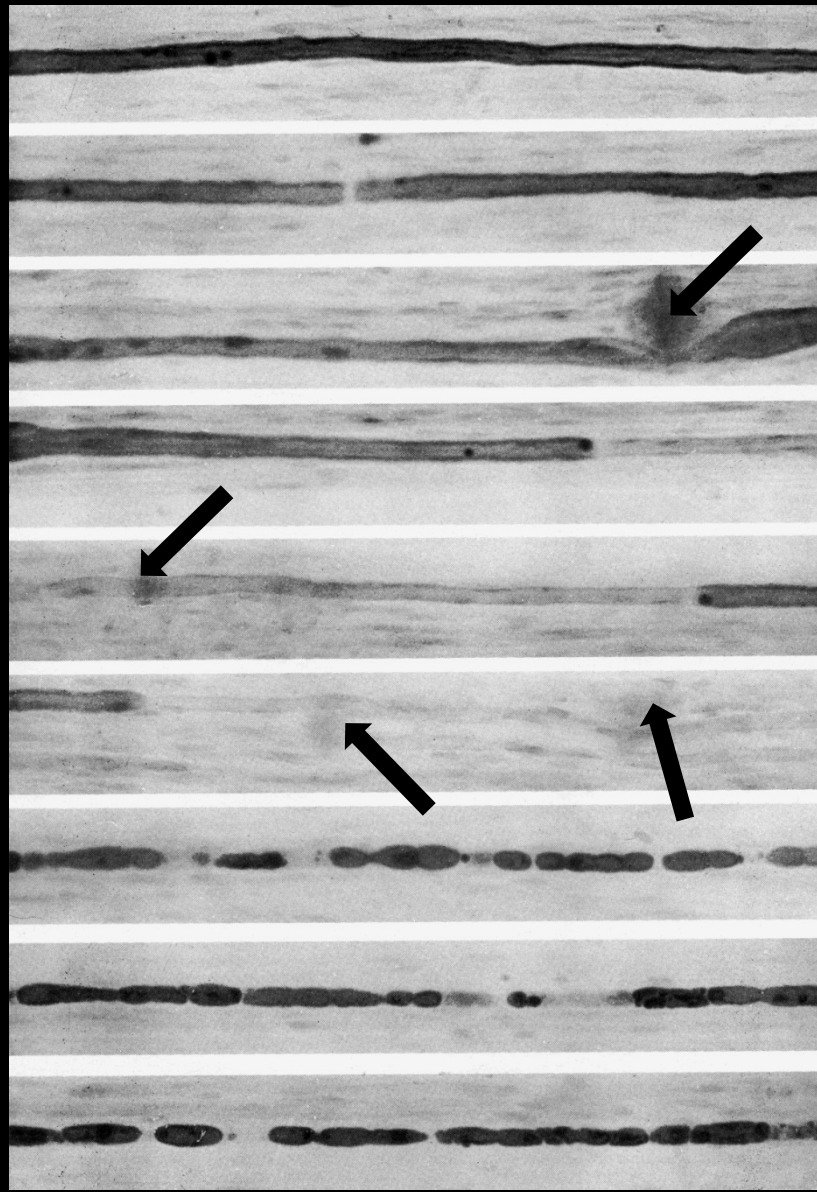
The large red clump (left upper) is methylviolet staining of amyloid in this patient's nerve.



This shows the apple-green birefringence in the Congo-red stain under polarizing filters.



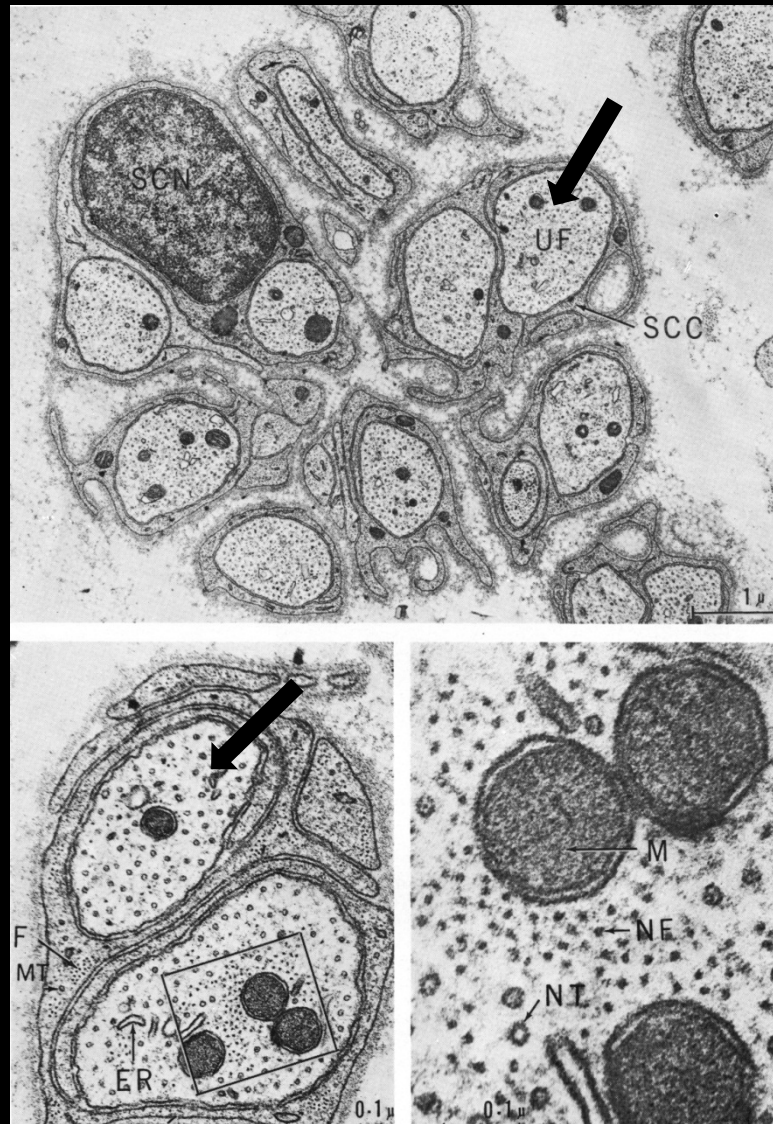
In longitudinal sections of the nerve, large deposits of amyloid (arrows) are shown.



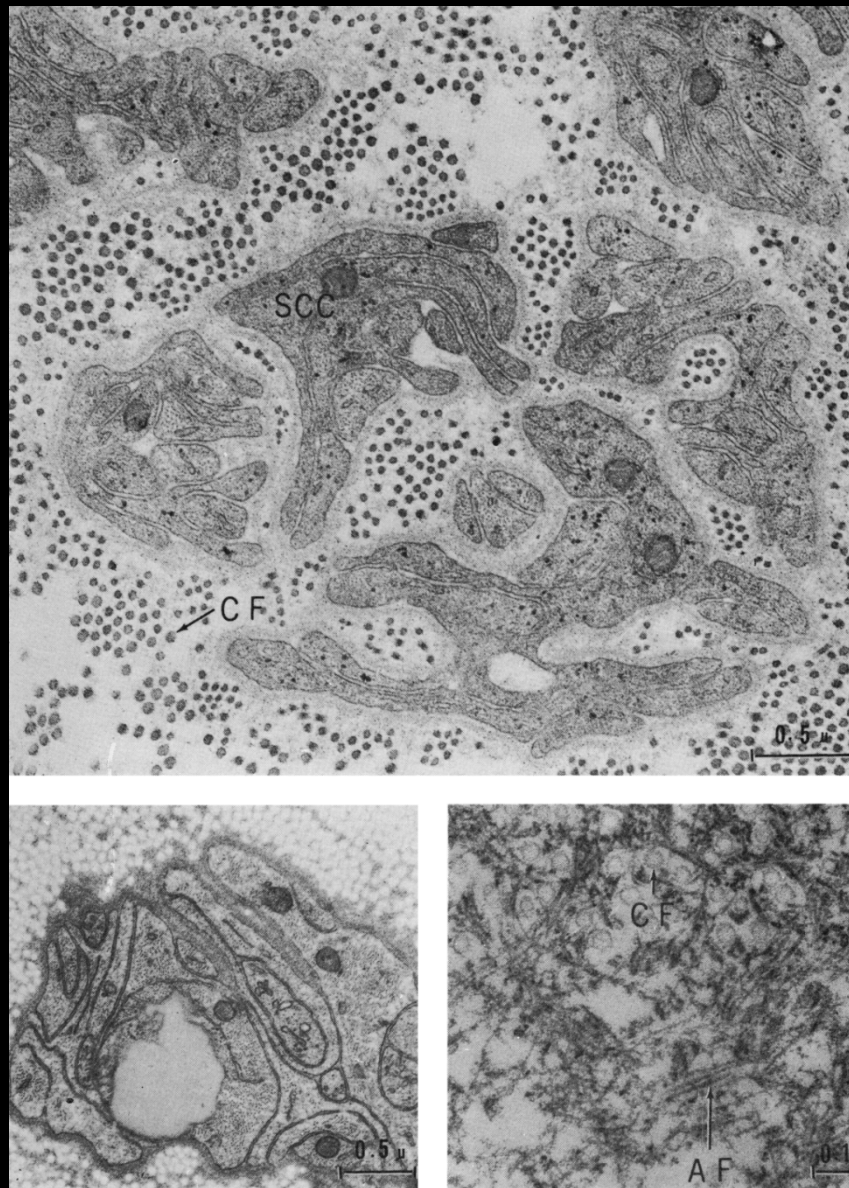
These are consecutive lengths of teased fibers from our patient. The arrows indicate the deposits of amyloid which is indenting a myelinated fiber, and in more distal parts of the fiber, one sees de- and remyelination and even axonal degeneration.



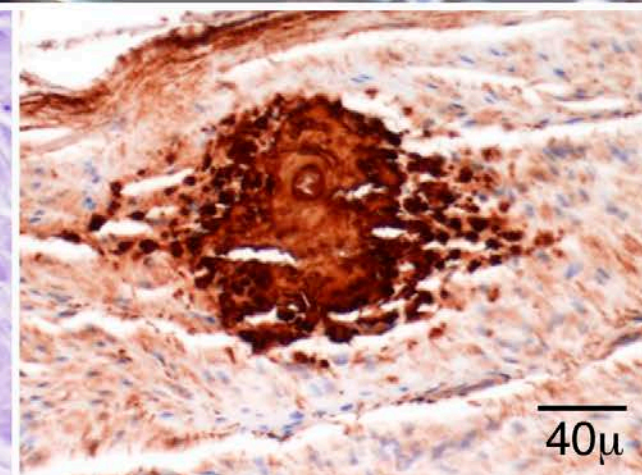
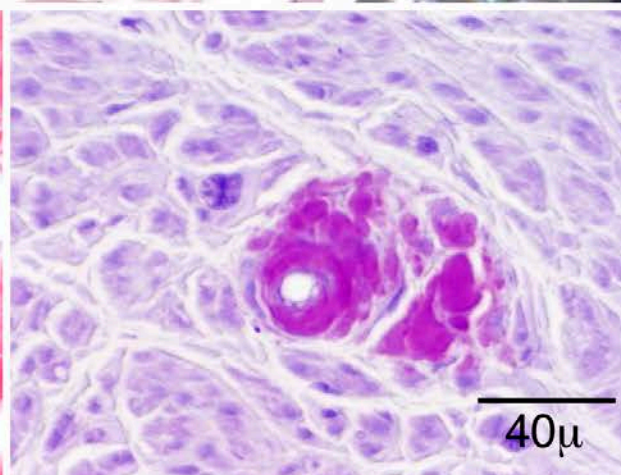
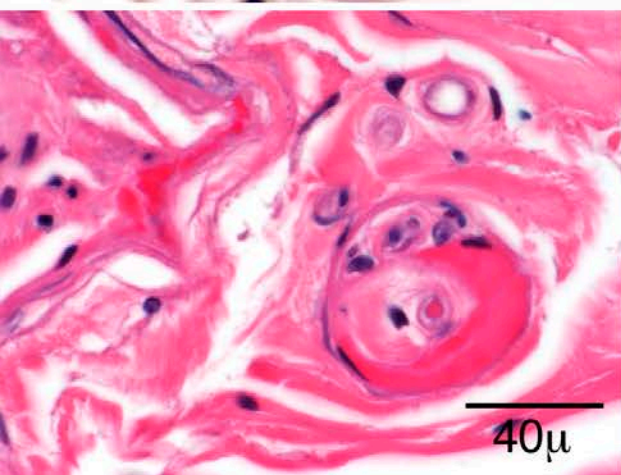
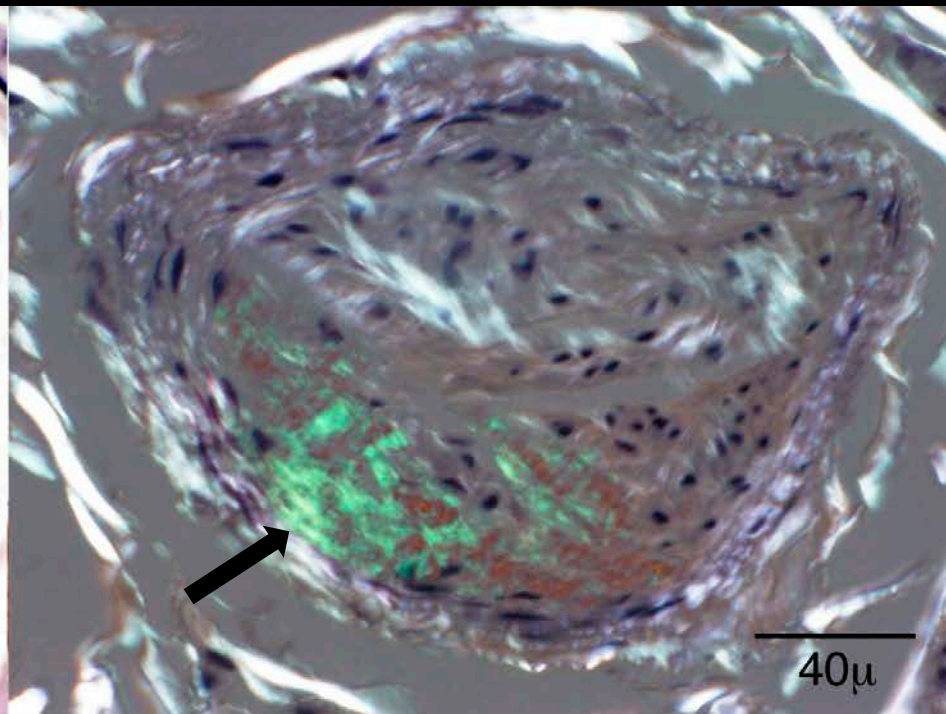
The indentation of a fiber by amyloid.



A cluster of unmyelinated nerve fibers in healthy nerve at various degrees of magnification.



Electron micrograph of a former unmyelinated fiber cluster without the unmyelinated fibers in our patient's nerve.



Deposits of amyloid in nerve from another patient left upper Congo red positivity (arrow) right upper apple-green birefringence (arrow).