

Perspectives in refractory diabetic and amyloid small-fiber neuropathy

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Small fiber neuropathies (SFN) are characterized by alteration of unmyelinated and small myelinated nerve fibers with impairment of pain and temperature sensations, often associated with autonomic disturbances. Pains and numbness in distal lower limbs are the most common symptoms. Loss of temperature and pain sensations in distal lower limbs contrasts with relative preservation of light touch, position and vibratory sense, tendon reflexes and muscle strength. Nerve conduction and sensory action potentials may remain unaltered for a long time. Diabetes is responsible for most cases of SFN in the world. In severe cases progression of sensory deficit follows a length dependent pattern with gradual involvement of proximal lower limbs; upper limbs and anterior trunk. In this setting, nerve biopsy specimens show predominant involvement of unmyelinated and small myelinated fibers, and a dying back process followed by spontaneous ax-

onal regeneration by sprouting. Ideally, only perfect control of blood glucose, as achieved by β -cell replacement by Langherans islets or pancreas transplantation can prevent further alteration of the PNS and allow efficient regeneration.

Familial amyloid polyneuropathy (FAP) is very similar to severe forms of diabetic polyneuropathy with a devastating course leading to death within 10 years on average. Fiber degeneration is linked to the presence of transthyretin-derived amyloid deposits in the endoneurium. Liver transplantation dramatically reduces the amount of circulating mutated transthyretin and, a drug recently introduced Tafamidis[®] seems to favorably influence the outcome. However genetic counseling remains highly recommended for prevention of FAP.