

193rd ENMC International Workshop: Pathology diagnosis of idiopathic inflammatory myopathies

November 30th – December 2nd 2012

Naarden – The Netherlands

When people experience progressive muscle weakness, muscle pains and fatigue with onset less than a year ago, sometimes accompanied by non-muscle symptoms such as skin rash, they could be suffering from an idiopathic inflammatory myopathy (IM), also known as myositis. There are various subgroups of IIM.

In cases without a characteristic skin rash, it is necessary to perform a muscle biopsy to diagnose IIM. The muscle biopsy serves to distinguish IIM from other myopathies and to identify subgroups. This is very important as the treatment options and prognosis may vary depending on diagnosis. Classification criteria for the neuropathological features of the different myositis subgroups are based on consensus but have not been validated.

World experts in the field of idiopathic inflammatory myopathy gathered in Naarden to address this issue. The workshop brought together neurologists, rheumatologists, immunologists and pathologists active in patient care and research. Knowledge was exchanged regarding classification, testing the classical views with new scientific insights, also making room for discussing the disease subtypes. The focus lay also on the comparison with inflammatory changes that occur in other muscle diseases, to avoid that patients that have diseases of hereditary origin are falsely diagnosed with idiopathic inflammatory myopathy.

A recommendation was developed stating which histopathological stainings should be part of a minimum diagnostic set, and how best to define and score disease signs. The views that were developed, were tested in a practical session at the Amsterdam Medical Center, in which typical and atypical muscle biopsy slides were discussed. At the end of the workshop, a consensus text was drafted. Participants agreed unanimously to continue work on a standardized diagnostic work up and to reconvene in the near future to fine-tune the proposed strategies.