Computed tomography of the skeletal musculature

M. de Visser, neurologist (Amsterdam)

Recently computed tomography of skeletal musculature has been introduced as a diagnostic tool in the field of neuromuscular diseases.

By means of a standardized program the muscles of the neck, the shoulder girdle, the lumbar region, the pelvic girdle, the thighs and the lower legs are visualised.

Radiological findings in patients with neuromuscular diseases were not always consistent with the results on clinical examination. On one hand computed tomographic abnormalities, consisting of areas of decreased density, may be observed in non-paretic muscles and on the other hand paretic muscles may have a normal appearance on computed tomographic examination. These findings are not easy to explain. One has to give account to the fact, however, that morphological abnormalities of muscles are not identical to clinical abnormalities, i.e. weakness of the muscles. Also, the function of muscles can not be determined by radiological examination. Moreover, compound muscles like the quadriceps femoris muscle or the adductors, may show areas of decreased density in part of the muscle, whereas on clinical examination of the whole muscle is tested. Finally, technical imperfection of the present apparatus may not reveal small areas of decreased density.

Computed tomographic examination of skeletal musculature can be applied for selecting the site of a muscle biopsy. Moreover, it can be useful in differentiating various neuromuscular diseases.

The practical significance of investigations in muscle diseases with respect to the patient and his family

H.F.M. Busch, neurologist (Rotterdam)

Some recent advances in nosology and diagnosis of myopathies were reviewed, in particular in myotonic dystrophy, the X-linked muscular dystrophies, malignant hyperthermia, mitochondrial myopathies and toxic myopathies. A number of genetic aspects in particular the present methods of carrier detection and antenatal diagnosis in the hereditary myopathies were briefly discussed.

Biochemical studies in biopsy material, taken for diagnostic reasons and properly preserved have revealed important pathogenetic mechanisms in metabolitic and other myopathies.

A close cooperation between diagnostic, genetic and rehabilitation centres, family physicians and the Muscular Diseases Association of the Netherlands increases the availability of the advantages of investigative results to the patients and their families.

The ischemic forearm test revised: more information with less effort.

P.R. Bär, biochemist and F.G.I. Jenneken, neurologist (Utrecht)

The ischemic test is used in neuromuscular diagnosis. People with enzyme deficiencies in glycogen metabolism fail to show a rise in venous lactate levels in this test.

A more recently discovered deficiency (of myoadenylate deaminase, MAD) can be detected in the same test: NH3, formed in muscle during exercise, rises in healthy people, not in MAD-def. people. For both parameters (Lac and NH3) false-positive results occur easily and we therefore standardized the test to obtain unambiguous results. The amount of work was normalized by using a wristmuscle-training device; we tested three levels of work: 60, 90 and 120 contractions in one minute. Three bloodsamples were collected, at 0, 2 and 12 minutes.

Results

1. At 60/min there is already a clear lactate response (6.4× resting value), which is near-maximal, as at 90 and 120/min the rise is only slightly higher (7.1× and 7.5× resp.).
2. At 60/min the NH3 response is small: 1.4× rest. value; at 90 and 120/min it is significant, 3.1× and 5.7× resp.

Conclusion

At 60/min the lactate-response has sufficient discriminating power; however, the NH3-response will give rise to false-positive results, as is reported. We therefore choose 90/min as routine testload: both responses are significantly different from pre-test values in all controls. No one so far has been unable to perform the test. Data of controls and patients will be shown.

Comparison of protein synthesis in red and white muscle

H.A. Boekholt, animal physiologist and V.V.A.M. Schreurs, biochemist (Wageningen)

Protein synthesis was studied in m. longissimus and m. soleus of the rabbit after labeling the proteins by infusion with ^{14}C-tyrosine. Muscle samples were solubilised in SDS and separated by SDS gel filtration. Protein fractions were analysed for total protein and ^{14}C-radioactivity. Protein synthesis was measured as the amount of ^{14}C-tyrosine incorporated per mg protein, corrected for tyrosine content of the protein. Incorporation of ^{14}C was three times higher for proteins in m. soleus than for proteins in m. longissimus. This may be due to real differences in synthesis rate or to differences in specific activity of the precursorpool. Gel filtration provides a separation between main muscle proteins. Although both elution profiles show roughly the same pattern with 5 protein peaks clear characteristics can be distinguished.

The main myofibrillar proteins of both muscles show similar relative synthesis rates (relative to actin = 1): myosine heavy chains: myosine light chains : actin = 1.6 : 3.4 : 1. These results indicate that there exist differences in synthesis rates of polypeptides which constitute a protein, as do the heavy and light chains of myosine. The same holds for polypeptides which constitute a functional unit like the myofibril.

Digital simulation of the mitochondrial energy generating processes in human muscular tissue homogenates

J.C. Fischer, pediatrician, W. Ruitenbeek, biochemist, J.M.F. Trijbels, clinical chemist, R.C.A. Sengers, pediatrician, A.J.M. Janssen, laboratory assistant and A.M. Stadhouders, cell biologist (Nijmegen)

A digital simulation program for the recently developed incubation system to estimate ATP and creatinephosphate generating capacity of skeletal muscle mitochondria[1] has been developed. The BASIC program calculates in an iterative procedure the response of the creatine kinase and adenylate kinase mediated reactions upon the generation of ATP by oxidative phosphorylation. It has shown that the creatine kinase present in the incubation system had all the capacity of maintaining a steady state mass action ratio at equilibrium value, whether the characteristics of the adenine nucleotide pool are controlled by adenylate kinase activity (at the start of the incubation) or by ATP generated by mitochondrial metabolism. Use of the program facilitated the choise of initial ADP and Cr concentrations for the recognitition of possible defects in the creatine kinase enzymes.

References

[1] RUITENBEEK W, SENGERS RCA, TRIJBELS JMF, STADHOUDERS AM, JANSSEN AJM. Estimation of energy metabolism in human skeletal muscle homogenate as a diagnostic aid. J Inher Metab Dis 1981; 4:91.