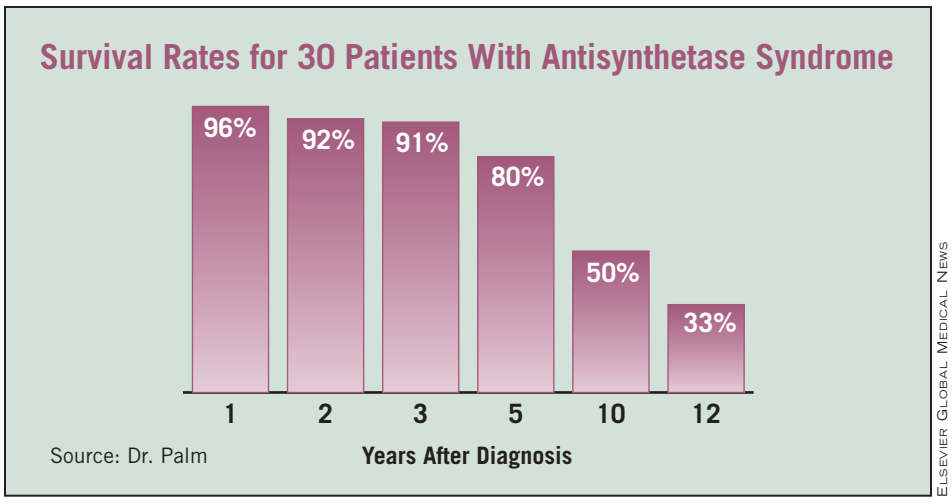


Elsevier Global Medical News

Ten-Year Survival Poor in Antisynthetase Syndrome

BY NANCY WALSH
New York Bureau

BARCELONA — A review of 30 patients with antisynthetase syndrome found that only half survived 10 years after diagnosis, Dr. Oyvind Palm reported at the annual European Congress of Rheumatology.

This idiopathic inflammatory myopathy is characterized by the presence of antibodies directed against tRNA synthetase. The most common antibody is anti-Jo-1, which is found in 80% of cases.

Other antibodies sometimes found include anti-SSA, anti-PL-7, and anti-PL-12.

Clinical manifestations of the disease include interstitial lung disease, which can be severe, arthritis, Raynaud phenomenon, and the hyperkeratotic rash known as mechanic's hands, according to Dr. Palm of the department of rheumatology, Rikshospitalet-Radiumhospitalet Medical Center, Oslo.

With the aim of characterizing the disease's clinical and serologic features, researchers reviewed all hospital records of patients diagnosed with an inflammatory myopathy and analyzed the charts of those who had antisynthetase antibodies and pulmonary disease.

The mean age of these 30 patients was 45.5 years, and in one-third of the group, the disease onset was before age 40. Two-thirds of the patients were women.

'While approximately 90% [of patients] survive the first 3 years of disease, thereafter the mortality increases sharply.'

Most patients had histologic evidence of inflammatory myopathy and elevated serum creatine kinase, but only four had elevations of creatine kinase exceeding 3,000 IU/mL.

Muscular manifestations rarely caused significant patient disability and were present at the onset of disease in only six of the cases.

Anti-Jo-1 antibodies were detected in 90%. Anti-SSA autoantibodies, commonly found in patients with Sjögren syndrome, were detected in 50% but only rarely were they associated with dry eyes and mouth, Dr. Palm wrote in a poster session.

Pulmonary involvement was classified as follows:

► Type I (acute): Found in 24%; rapid onset of dyspnea or cough with development of hypoxemia within 1 month after the onset of disease.

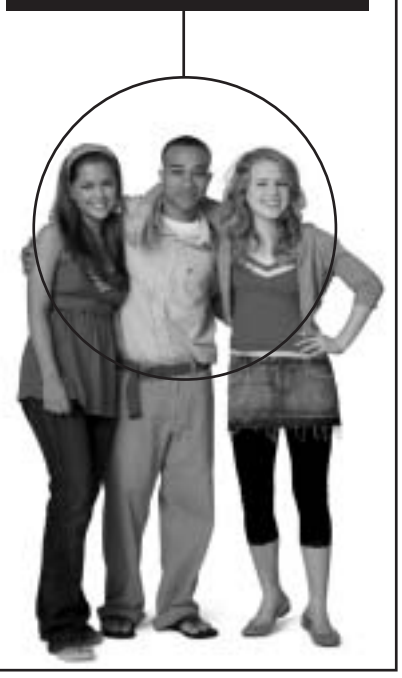
► Type II (subacute): Found in 64%; gradual onset of pulmonary symptoms.

► Type III (asymptomatic): Found in 12%; coincidentally detected pulmonary abnormalities on x-ray or CT scan with subsequent slowly developing pulmonary symptoms.

Honeycombing with end-stage pulmonary disease was found in 30.4%.

All but one patient had received treatment with immunosuppressive drugs including corticosteroids, cyclophosphamide, and rituximab.

Four patients died, two having type I pulmonary involvement."While approximately 90% survive the first 3 years of disease, thereafter the mortality increases sharply, and new treatment strategies are clearly warranted," he concluded. ■



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