Publication of the Month

EliA™ Excellence in Autoimmunity

April 04/11: The outcome of Wegener patients improved significantly

Key messages:

- *Improved diagnosis and treatment reduce mortality and relapse rates of Wegener patients.*
- *Comparing diagnosis in the last four decades, the interval between first symptoms and diagnosis was reduced from 8 to 4 months.*

Holle JU, Gross WL, Latza U, Nolle B, Ambrosch P, Heller M, Fertmann R, Reinhold-Keller E
Improved outcome in 445 patients with Wegener's granulomatosis in a German vasculitis center over four decades
Arthritis Rheum. 2011; 63: 257-266

Background: Wegener's granulomatosis is a severe autoimmune disease leading to massive damage of organs or even death. The importance of an early but correct diagnosis is indispensable to induce counteractions in time.

Summary: Three cohorts of patients diagnosed in 1966 – 1993 (cohort 1), 1994 – 1998 (cohort 2) and 1999 – 2002 (cohort 3) were retrospectively assessed for clinical manifestations, therapy procedures, trends in therapy, mortality, and incidence of malignancies.

Unchanged:
- Organ manifestations are similar.
- Cyclophosphamide (CYC) is still the most prescribed drug.
- No increased rate of malignancies

Changed:
- Interval between first symptoms and diagnosis was reduced by half to 4 month.
- The median cumulative dose of CYC was significantly reduced.
- Declined standardized mortality ratios (SMR) with fewer deaths and decreased relapse rates.

Young males had a considerably higher SMR and more frequent renal manifestations compared to young females.

Conclusions: The decline in mortality is probably due to improved diagnostic and therapeutic procedures and increased awareness of WG, leading to earlier diagnosis, reduction in relapse rates, and lower cumulative CYC dose with fewer deaths related to therapy.
Shorter periods of remission induction and increasing intravenous administration of CYC account for a reduced cumulative CYC dose.
The decline in relapse rates is associated with the implementation of a regular maintenance therapy.

Comment: This study confirms the better outcome of Wegener patients during the last 40 years. And the development of better diagnostic tests, finding more Wegener patients and less controls positive is still going on. Phadia's highly sensitive and specific EliA PR3^S assay yields a good evidence for the determination of the disease.
The panel for detection ANCA-associated Vasculitis is complemented by the sensitive and specific EliA MPO^S assay.