**Information for patients**

**Sarcoidosis**

Sarcoidosis is an inflammatory disease that can affect any organ and involves the lung in 90% of cases. In tissue of biopsies from patients with sarcoidosis characteristic clumps of white blood cells called granulomas, can be identified. Granulomas are an immune response to foreign particles within the body and this is why sarcoidosis is thought to have an environmental cause. In many patients sarcoidosis exists without causing symptoms and is, therefore, often discovered during routine checkup. Usually it is detected by chest radiographs or a chest computer tomography scan showing enlarged mediastinal lymph nodes and reticular-nodular patterns over the lungs. The disease can last one or two years and require minimal or no treatment or it can span decades and require interventions. The organ involvement determines the symptoms and chronic progressive forms of the disease involving the lungs result in shortness of breath. Patients with neurological or cardiac manifestations have the poorest outcome.

**Whom does it affect?**

Sarcoidosis affects people of all ages throughout the world with the highest incidence in those between the age of 20 and 40 with a second peak for females in the age of 60. There are significant racial and gender differences in disease severity, incidence and prevalence. Women are more often affected than men and tend to suffer from more severe disease. The highest prevalence has been observed in northern Europe with 65 per 100.000 population in Sweden. In Central Europe the prevalence is about 40 per 100.000 population and prevalence numbers about 10 per 100.000 population are reported from Southern Europe.

Exposure to inorganic particles, insecticides and moldy environments has been reported to be associated with sarcoidosis but eliciting agents have not been identified. A number of occupations are associated with sarcoidosis e.g. metal working, firefighters, teachers, nurses.

Susceptibility to sarcoidosis depends on an interaction between variants of inherited genes and environmental exposures. Relatives of sarcoidosis patients are at higher risk of developing the disease compared to the general population. About five percent of the sarcoidosis patients are from families with several affected individuals. Although remnants of bacteria can be found in some granulomas of sarcoidosis, it is not a contagious disease.

**Clinical course and pathophysiology**

Sarcoid granulomas may spontaneously resolve or may accumulate and persist. Persistent granulomas may lead to organ dysfunction and cause scarring. The extent and location of the granulomata determine the severity of the disease. For example, large accumulation of granulomas in lymph nodes is in most situations of no relevance for the patient. However, few granulomas in the ventricular conduction system of the heart may lead to arrhythmias with sudden death. There are studies which indicate that a considerable number of sudden death of young “healthy” people is caused by unrecognized sarcoidosis.

Pulmonary fibrosis occurs in about 25 % of patients and can lead to respiratory failure. In cases with pulmonary insufficiency lung transplantation is the last resort. Why fibrosis occurs in some individuals and not in others is not known.

Several studies have shown that animated and non-animated agents may cause sarcoidosis because sarcoidosis most commonly involves the lungs followed by the eyes and skin. Therefore, exposure to air-born agents in susceptible individuals has been suspected as the cause. This concept is supported by the observation of a higher incidence in the spring and the summer months when different infections peak. However, no infectious agent has been unequivocally identified to cause the disease. Sarcoidosis occurs worldwide in all climates and, therefore, it is likely that it is the immunological response to various environmental agents, rather than to a single agent. Those who develop sarcoidosis likely inherit a tendency to respond more intensely to the unknown eliciting agents.

**Prevention, treatment, prognosis**

Since the course of sarcoidosis is unknown, prevention does not exist. Although familiar sarcoidosis takes place in about 5 % of cases routine screening of family members is ineffective and should not be done.

Not all patients with sarcoidosis need treatment. In at least two thirds of patients sarcoidosis resolves without treatment and causes no further problems. The decision to commence treatment is based on weighing the risks of treatment against its potential benefits. In general, treatment is begun for the following reasons: Symptoms interfere with activities of daily living; or organ function is threatened. When treatment is started with a higher dose of prednisolone, it should be tapered-off over six months. Corticosteroids are the most effective treatment and improve symptoms and lung function and probably prevent complications. In some patients sarcoidosis does only respond to high dose of corticosteroids which leads to the necessity of combining corticosteroids with immunosuppressive drugs. In desperate cases the off-label use of biological drugs interfering with inflammatory processes may stabilize pulmonary function and suppresses symptoms.

Prognosis is variable and depends on gender, race, age and the pattern of organ manifestation. In the majority of patients spontaneous resolution takes place. With 5% the risk of relapses is low. Progressive sarcoidosis may lead to respiratory insufficiency requiring oxygen supplementation or even lung transplantation. Sarcoid manifestations in heart or the nervous system frequently cause organ damage.

**Research**

Large clinical studies have demonstrated the broad spectrum of clinical manifestations, occupational associations, symptoms, courses and therapeutic responses; however, the eliciting agents are still elusive. Genetic studies demonstrated that the susceptibility to develop sarcoidosis depends on the inheritance of gene variants which promote an exaggerated immune response against the unknown sarcoid triggers.