Sarcoidosis – patient treatment priorities

Introduction

Sarcoidosis is a rare disease with an estimated incidence of between 4.7 and 64 per 100,000. Some 40% of individuals experience sarcoidosis as a chronic condition, while >60% of individuals find that the disease resolves within 2-5 years.

However, it has been shown that patients can still have symptoms that result in a reduced quality of life even after clinical signs of disease have disappeared. These symptoms include fatigue, pain, everyday cognitive failure, small fibre neuropathy, exercise limitation and depressive symptoms.

In 2016, a European Respiratory Society (ERS) Task Force (TF) was set up to develop an international clinical guideline on sarcoidosis treatment to provide evidence-based recommendations for healthcare professionals.

Sarcoidosis outcomes are traditionally governed by clinical measures, such as blood tests, pulmonary function tests and imaging.

The European Lung Foundation (ELF) supported the facilitation of patient input into the TF via its Sarcoidosis Patient Advisory Group (PAG), which comprises of patient organisation representatives from nine countries.

Methods

The anonymous survey asked patients to rate the following outcomes of sarcoidosis treatment: quality of life, functionality, blood tests, pulmonary function tests, imaging, adverse events, and survival.

Purpose of Study

1. Develop a survey designed to gather views about which treatment outcomes matter most to sarcoidosis patients with the results to inform the work of the TF.

Results

Outcome and definition

Quality of life
Definition: The level of well-being a person feels which might include a standard of health (physical and/or mental), comfort and life satisfaction or other factors important to that person.

Functionality
Definition: the level at which the body (or specific parts of the body) is able to work and function as normal.

Pulmonary function tests
Definition: non-invasive tests that show how well a person’s lungs are working. Different tests can measure lung volume, capacity, rates of flow and gas exchange.

Blood tests
Definition: taking a sample of blood to help with diagnosis (including ACE test).

Imaging
Definition: X-rays and scans e.g. chest X-ray, CT scan, PET scan which take photographic or digital pictures of the body. They can be used to help diagnose a lung condition and to monitor how a condition progresses.

Adverse events
Definition: an undesired or harmful effect from a drug or medical intervention such as surgery, also known as side effects.

Survival
Definition: the length of time a person will continue to live.

Questionnaire

Themes noted in survey

1. QoL and Functionality should be included in treatment outcomes

The survey findings strongly show that respondents want QoL and Functionality to be included as outcomes in their treatment and care. “Scans, tests and examinations are of course crucial in the treatment, but the well-being and the quality of life determine how much I suffer from sarcoidosis. And, it is the most important thing.” (Survey respondent)

2. Clinicians should work in a multidisciplinary way to ensure a holistic approach

In sarcoidosis, it is important to identify every symptom and treat them as a whole rather than in isolation

“I think that although mental health is included in the definition of quality of life in this survey it is important enough to have its own category. Often sarcoidosis patients suffer from depression and low mood. Going through tests, missing appointments, taking drugs everyday and seeing health deteriorate definitely takes its toll on mental health and it’s important because it will ultimately underpin recovery and overall health and well-being.” (Survey respondent)

“A multidisciplinary approach with an emphasis on functionality is important. I have never had another doctor see me other than my internist.” (Survey respondent)

3. Sarcoidosis specialists and specialist centres are needed in every country

Sarcoidosis can be difficult to diagnose and treat when there is a lack of expertise, particularly for the rarer manifestations such as neurological sarcoidosis.

“It is important to increase the number of centres that deal with this disease as seen for other more widespread but also incurable diseases.” (Survey respondent)

Conclusions

1. Quality of life and functionality were the highest priority for outcomes of sarcoidosis patients

2. Blood tests and pulmonary function testing were not viewed as nearly important to the patient

3. We encourage the sarcoidosis health and research community to partner with sarcoidosis patient organisations and to utilise this immense source of sarcoidosis lived experience to develop effective methods of measuring and incorporating QoL and Functionality outcomes as a way of improving sarcoidosis outcomes and future treatment.