SUMMARY POINTS

- Sarcoidosis is a multisystemic inflammatory disorder characterised by the presence of epithelioid non-caseating granulomata in affected organs.
- It has a worldwide distribution and is slightly more common in women than in men. Prevalence peaks between 20 and 40 years of age and in women over 50. Studies on sarcoidosis show significant heterogeneity in incidence, prevalence, disease presentation and severity among different ethnic racial groups.
- The aetiology of sarcoidosis remains unknown, but several infectious agents and genetic factors have been implicated.
- Pulmonary involvement is present in 90% of cases with sarcoidosis, and up to 30% of patients present with extra pulmonary disease, including musculoskeletal, dermatological, ocular, cardiovascular, neurological and renal manifestations.
- Pulmonary involvement in sarcoidosis may range from radiographic stage I disease with bilateral hilar adenopathy without parenchymal involvement, to stage III or IV disease with pulmonary fibrosis, lung insufficiency and/or cor pulmonale.
- Musculoskeletal manifestations in sarcoidosis include acute arthritis, Lofgren's syndrome, chronic arthritis, tenosynovitis, myopathy and bone involvement.
- Lofgren's syndrome is characterised by hilar adenopathy, acute arthritis/arthralgias (ankle involvement is typical), and erythema nodosum. Lofgren's syndrome is usually self-limiting and has a good prognosis, usually requiring treatment with non-steroidal anti-inflammatory drugs only.
- A variety of non-specific laboratory abnormalities may be seen in patients with sarcoidosis. Sarcoidosis has been associated with hypercalciuria, hypercalcaemia and elevations in serum angiotensin-converting enzyme levels.
- Pulmonary imaging may reveal different stages of pulmonary sarcoidosis. Joint and bone radiographs may show non-specific radiographic changes.
- There are no specific diagnostic tests for sarcoidosis. The diagnosis rests on a combination of compatible clinical and radiographic manifestations, histological proof of non-caseating granulomata and exclusion of other diseases with similar presentation.
- Histological proof is not required in patients presenting with classic Lofgren's syndrome, but for all other patients with suspected sarcoidosis, biopsy is usually needed to confirm the diagnosis.
- Sarcoidosis is often a benign disease with good prognosis, unless there is cardiac and neurological involvement. Spontaneous remission occurs in 60–70% of cases, even more frequently in Lofgren's syndrome, and relapses are uncommon. Mortality is <5% and mainly results from respiratory failure related to pulmonary fibrosis, and refractory neurological or cardiac involvement.
- Glucocorticoids have been the most commonly used agents for the treatment of pulmonary and extra pulmonary sarcoidosis. Experience with other drugs is limited.