

Caplan's syndrome

Synonyms: rheumatoid pneumoconiosis, silicoarthritis and rheumatoid lung silicosis

What is Caplan's syndrome?^[1]

Caplan's syndrome is a condition which occurs due to scarring of the lungs resulting from exposure to coal mine dust, asbestos and silica. It more commonly affects patients suffering with [rheumatoid arthritis](#).^[1]

The syndrome is named after Dr Tony Caplan who was a doctor on the [pneumoconiosis](#) board in Cardiff.^[2] Since it was first described by Caplan in 1953, the exact correlation between rheumatoid arthritis, [pneumoconiosis](#) and autoimmunity has been a topic of debate.

How common is Caplan's syndrome? (Epidemiology)^[3]

It was said to affect 1 in 100,000 people but the incidence is falling as the coal mining industry has been in decline.

Although the syndrome was originally described in coal miners, several cases have since been diagnosed in individuals exposed to free silica or asbestos. However, such cases are much rarer than those occurring in individuals exposed to mineral coal dust.

There is a significantly higher frequency of TB in cases of Caplan's syndrome when compared with cases of pneumoconiosis without rheumatoid nodules.^[3]

Risk factors

Rheumatoid arthritis (RA) is a systemic disease and not just a disease of joints. The combination of RA and exposure to coal dust produces the condition. It develops especially in miners working in anthracite coal mines and in persons exposed to silica and asbestos.^[4]

There is probably also a genetic predisposition and smoking is thought to be an aggravating factor.

Caplan's syndrome symptoms (presentation)^[5]

Symptoms

There is cough and shortness of breath. In addition there are the features of rheumatoid arthritis (RA) with painful joints and morning stiffness.

Signs

There are features of RA, including tender swollen metacarpophalangeal (MCP) joints and rheumatoid nodules. The nodules may pre-date the appearance of RA by several years. Examination of the chest may show diffuse rales that do not disappear on coughing or taking a deep breath.

Differential diagnosis^[1]

The combination of rheumatoid arthritis (RA) and exposure to coal dust are essential for the diagnosis but silicosis and [asbestosis](#) must be considered. The X-ray appearance can resemble [tuberculosis \(TB\)](#), [sarcoidosis](#), lymphoma or metastases.

In RA, lung disease can develop even in the absence of dust exposure. It includes interstitial fibrosis, [pleural effusion](#), pulmonary nodules, pulmonary arteritis and [pulmonary hypertension](#).^[6]

Investigations^[1]

CXR shows multiple, round, well-defined nodules, usually 0.5–2.0 cm in diameter, which may cavitate and resemble tuberculosis (TB). CT scanning gives a better picture of cavitation.^[7]

[Spirometry](#) may reveal a mixed restrictive and obstructive ventilatory defect with a loss of lung volume. There may also be irreversible airflow limitation and a reduced gas transfer factor.

Rheumatoid factor, antinuclear antibodies, and non-organ specific antibodies may be present in the blood. ESR or PV and CRP will be elevated. X-rays of affected joints will show the features of rheumatoid arthritis (RA) with bone erosions.

Caplan's syndrome treatment and management

Nondrug

- Exposure to coal dust must cease.
- Physical treatment should proceed as for rheumatoid arthritis (RA).
- Smoking should cease.

Drugs

After exclusion of TB, steroids are used. Treatment of the RA will include [disease-modifying anti-rheumatic drugs](#) (DMARDs) at an early stage.

One trial found that rituximab does not alter the evolution of the pulmonary nodules in Caplan's syndrome.^[8]

Complications

There can be complications relating to rheumatoid arthritis as well as pneumoconiosis, in addition to side effects from steroids and other forms of treatment, whether [non-steroidal anti-inflammatory drugs](#) (NSAIDs) or DMARDs.

Prognosis

This is as for RA. Severe respiratory disability is uncommon but massive [pulmonary fibrosis](#) can progress at times. Spontaneous remission of the lung disease can occur.

Preventing Caplan's syndrome

People with rheumatoid arthritis (RA) must not be exposed to additional risk factors for lung disease, particularly smoking.

Further reading

- [Nemakayala DR, Surmachevska N, Ramphul K](#); Caplan Syndrome. StatPearls, Jan 2022.

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