Cogan's Syndrome

What is Cogan’s Syndrome?

Cogan's Syndrome is a rare inflammatory disease characterised by inflammation of the inner ears and eyes. It can lead to vision difficulties, hearing loss and dizziness. Commonly there is also inflammation in other organs as well, particularly the heart and large blood vessels, nervous system and bowels.

Who are affected?

Although any age can be affected the syndrome is commonest in young adults (20's and 30's). It affects males and females equally.

What is the aetiology (cause)?

The cause of Cogan's Syndrome is not yet known.

What are the symptoms?

The most common symptoms include red, painful, light-sensitive or blurred vision; hearing loss (which may become profound and permanent); vertigo (dizziness); poor balance; nausea and vomiting; fever, fatigue and weight loss.

Making a diagnosis

There are no specific diagnostic tests. The diagnosis is made on clinical examination and history where a combination of problems in the eyes and inner ears are described. Other infections/diseases, including Wegener's Granulomatosis and Rheumatoid Arthritis need to be excluded.
Treatment

There are no clinical trials of treatment in Cogan's Syndrome. Most people with Cogan's Syndrome will need treatment with moderately high doses of prednisolone or other types of steroids. A few patients with very mild eye disease may be treated with anti-inflammatory drugs including steroids and nonsteroidal anti-inflammatory drugs (NSAIDs) which are applied to the eye. Many patients will also require additional treatment with other immunosuppressive drugs including Methotrexate, Ciclosporin, Azathioprine, Tacrolimus or Cyclophosphamide.

Drugs and Side effects

For information on the main drugs prescribed for Cogan's Syndrome see:

- Azathioprine
- Ciclosporin
- Cyclophosphamide
- Methotrexate
- NSAIDs
- Steroids
- Tacrolimus

For information on other drugs used in the treatment of vasculitis see Glossary of drugs and side effects.

Prognosis

The course of the disease varies significantly from patient to patient. In some patients there is an initial flare, which may last several weeks to months. Following this there may be a slowly progressive course in some patients while others have a course of complete remission with intermittent episodes of disease activity. Fortunately blindness occurs in less than five per cent of patients. Deafness is a frequent and debilitating outcome occurring in up to 54 per cent of patients.

Key Points

- Cogan's Syndrome commonly affects the eyes and ears causing vision, hearing and balance problems
- Treatment will depend on the disease severity

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Further reading