NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health Technology Evaluation

Canakinumab for untreated Schnitzler syndrome

Draft scope

Draft remit/evaluation objective

To appraise the clinical and cost effectiveness of canakinumab within its marketing authorisation for treating Schnitzler syndrome.

Background

Schnitzler syndrome is a rare auto-inflammatory disease. The exact cause of Schnitzler syndrome is unknown but it may be associated with the immune system not functioning properly. It is characterised by long-term urticaria and elevated levels of monoclonal IgM gammopathy, a specific protein in the blood. Symptoms may include repeated episodes of fever, joint inflammation (arthritis), joint pain, bone pain and enlarged lymph nodes. In the longer term, Schnitzler syndrome may also lead to debilitating and life-threatening complications, including severe anaemia, amyloidosis (build-up of abnormal protein deposits) and lymphoproliferative disorders.

The prevalence of Schnitzler syndrome is estimated to be less than 50 patients in England, with an estimated incidence of 5 new cases every year.[1] In most cases, characteristic symptoms develop in people in their 50s, but have also been noted in individuals in their 30s. It affects men more often than women.[2]

Treatment options for Schnitzler syndrome are limited. Therapies such as antihistamines, non-steroidal anti-inflammatory drugs (NSAIDs), steroids, colchicine, hydroxychloroquine and pefloxacin are used as first line treatments for patients with Schnitzler's syndrome. These treatments usually provide only partial or temporary improvement of the symptoms.[1] Anakinra may be used as a first line treatment in patients with a documented diagnosis of Schnitzler syndrome.[1]

The technology

Canakinumab (Illaris, Novartis Pharmaceuticals Ltd) does not currently have a marketing authorisation in the UK for treating Schnitzler syndrome. It has been studied in clinical trials compared with placebo in people with Schnitzler syndrome.

Intervention(s)	Canakinumab
Population(s)	Adults with Schnitzler syndrome.

Comparators	Established clinical management without canakinumab, including but not limited to: • Antihistamines • Non-steroidal anti-inflammatory drugs • Corticosteroids • Colchicine • Hydroxychloroquine • Pefloxacin • Anakinra
Outcomes	The outcome measures to be considered include: • response rates • change in biomarkers of inflammation • adverse effects of treatment • health-related quality of life.
Economic analysis	The reference case stipulates that the cost effectiveness of treatments should be expressed in terms of incremental cost per quality-adjusted life year. The reference case stipulates that the time horizon for estimating clinical and cost effectiveness should be sufficiently long to reflect any differences in costs or outcomes between the technologies being compared. Costs will be considered from an NHS and Personal Social Services perspective. The availability of any commercial arrangements for the intervention, comparator and subsequent treatment technologies will be taken into account. The availability and cost of biosimilar and generic products should be taken into account.
Other considerations	Guidance will only be issued in accordance with the marketing authorisation. Where the wording of the therapeutic indication does not include specific treatment combinations, guidance will be issued only in the context of the evidence that has underpinned the marketing authorisation granted by the regulator.
Related NICE recommendations	**Related Technology Appraisals:** None. **Related Guidelines:** None. **Related Interventional Procedures:**

	None. **Related Public Health Guidance/Guidelines:** None.
Related National Policy	The NHS Long Term Plan, 2019. NHS Long Term Plan NHS England (2018/2019) NHS manual for prescribed specialist services (2018/2019) NHS England (2020) Clinical Commissioning Policy: Canakinumab for treating periodic fever syndrome TRAPS, HIDS/MKD and FMF (ages 2 years and older). Reference 200209P NHS England (2018) Clinical Commissioning Policy: Anakinra to treat periodic fevers and autoinflammatory diseases (all ages). Reference 170062P NHS England (2013) 2013/14 NHS Standard Contract for Cryopyrin Associated Periodic Syndrome (all ages). Reference E13/S(HSS)/b. Department of Health and Social Care, NHS Outcomes Framework 2016-2017: Domains 1, 2 and 3 https://www.gov.uk/government/publications/nhs-outcomes-framework-2016-to-2017

Questions for consultation

Where do you consider canakinumab will fit into the existing care pathway for Schnitzler syndrome?

Have all relevant comparators for canakinumab been included in the scope? Which treatments are considered to be established clinical practice in the NHS for Schnitzler syndrome?

Are the outcomes listed appropriate?

Are there any subgroups of people in whom canakinumab is expected to be more clinically effective and cost effective or other groups that should be examined separately?

Would canakinumab be a candidate for managed access?

Do you consider canakinumab to be innovative in its potential to make a significant and substantial impact on health-related benefits and how it might improve the way that current need is met (is this a 'step-change' in the management of the condition)?

Do you consider that the use of canakinumab can result in any potential substantial health-related benefits that are unlikely to be included in the QALY calculation?

Please identify the nature of the data which you understand to be available to enable the committee to take account of these benefits.

NICE is committed to promoting equality of opportunity, eliminating unlawful discrimination and fostering good relations between people with particular protected characteristics and others. Please let us know if you think that the proposed remit and scope may need changing in order to meet these aims. In particular, please tell us if the proposed remit and scope:

- could exclude from full consideration any people protected by the equality legislation who fall within the patient population for which canakinumab will be licensed;

- could lead to recommendations that have a different impact on people protected by the equality legislation than on the wider population, e.g. by making it more difficult in practice for a specific group to access the technology;

- could have any adverse impact on people with a particular disability or disabilities.

Please tell us what evidence should be obtained to enable the committee to identify and consider such impacts.

NICE intends to evaluate this technology through its Single Technology Appraisal process. We welcome comments on the appropriateness of appraising this topic through this process. (Information on NICE's health technology evaluation processes is available at https://www.nice.org.uk/about/what-we-do/our-programmes/nice-guidance/nice-technology-appraisal-guidance/changes-to-health-technology-evaluation).

References

1. NHS England (2018) Clinical Commissioning Policy: Anakinra to treat periodic fevers and autoinflammatory diseases (all ages). Accessed: May 2022.

2. National organization for rare disorders (NORD) (2018) Schnitzler Syndrome. Accessed May 2022.