4CPS-161 ATC: L04 - Immunosuppressive agents

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1. OBJECTIVES

The deficiency of interleukin (IL)-1 receptor (IL-R) antagonist (DIRA) is a rare genetic auto-inflammatory disease related to alterations in antagonists of the IL-1 pathway. We aimed to summarize the efficacy and safety of IL-1 antagonists drugs in DIRA following the scoping review methodology.

2. MATERIAL AND METHODS

The study was conducted and reported using the methodology described in the Joanna Briggs Institute Reviewer’s Manual1 and the PRISMA Extension for Scoping Reviews2. Data extraction was carried out by two researchers independently.

3. RESULTS

15 case reports found between 2009 and 2017

19 Patients

19 anakinra

2 canakinumab

9 different mutations in IL1RN (all homozygous)

17 anakinra

Response

Previous treatment

100%

19 patients

78.94%

15 patients

52.63%

10 patients

68.42%

13 patients

63.15%

12 patients

15.78%

4 patients

15.78%

3 patients

Antibiotics

Corticosteroids

NSAIDs

Acitretin

Medium-long term (> 24weeks)

Short term(<12 weeks)

Immediate (day-hours)

The length of therapy) varied between 2 weeks and 4.5 years.

Discontinuation of anakinra in 9 patients was reported following a flare-up of their disease.

Anakinra was associated with transient injection site reactions (n=3) and anaphylactic reactions (n=2).

1 episode of vomiting and diarrhoea was reported in a patient treated with high doses of canakinumab.

5. DISCUSSION

Anakinra is the most commonly used drug in patients with DIRA. The observed efficacy was high in patients with DIRA at all times, both with anakinra and canakinumab.

6. CONCLUSIONS

The use of IL-1 antagonists may prove to be a valid therapeutic alternative, but there is a lack of secondary studies that summarise the use of IL-1-targeting agents in this disease. Larger studies with better methodological quality are needed to increase confidence in the use of these drugs in patients with DIRA.

REFERENCES AND/OR ACKNOWLEDGEMENTS
