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1. OBJECTIVES

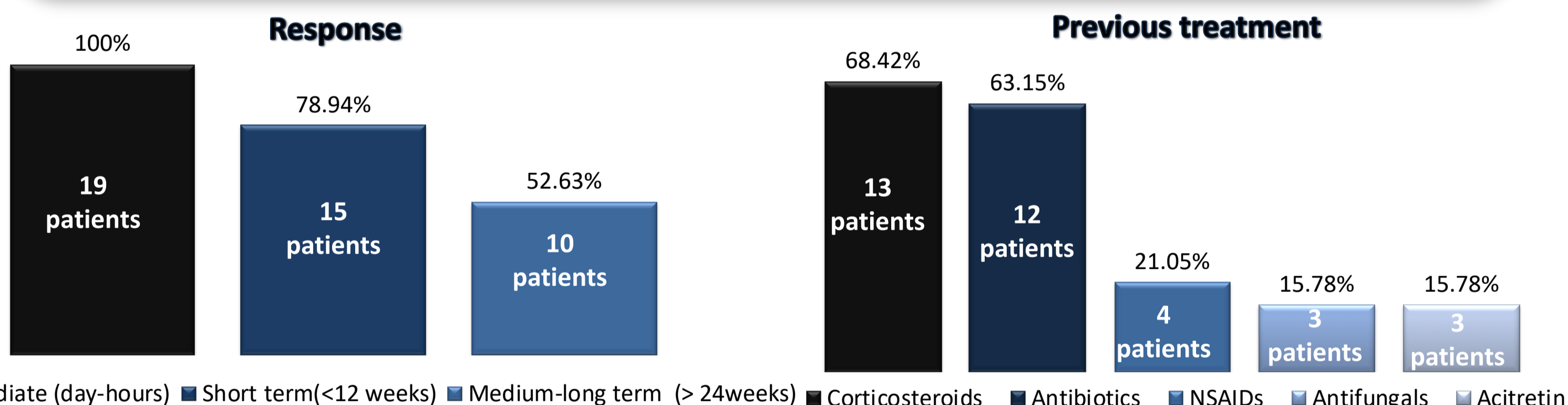
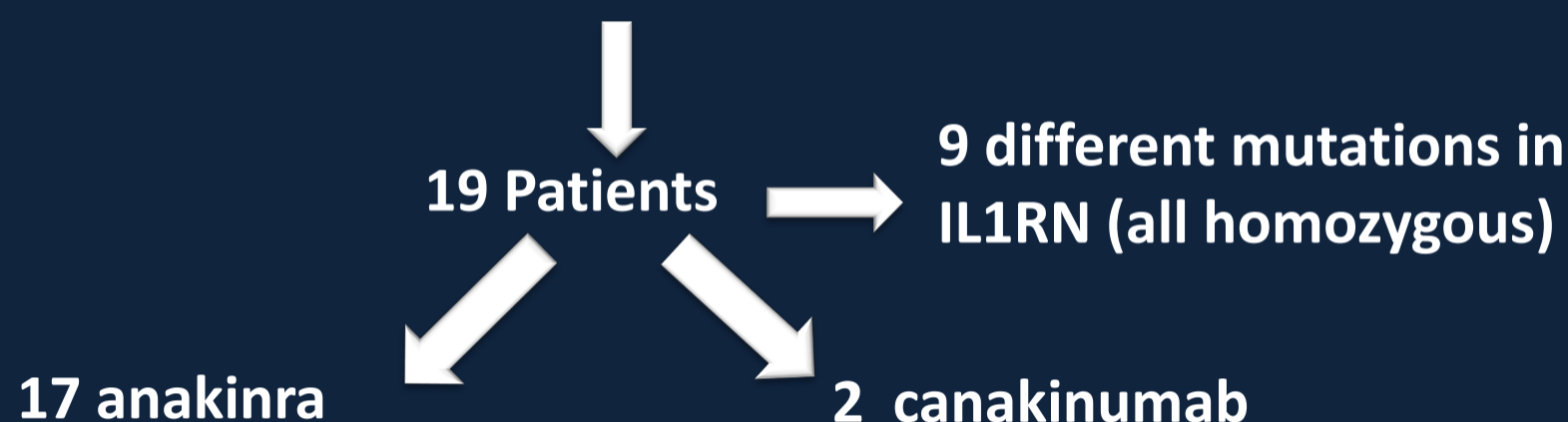
The deficiency of interleukin (IL)-1 receptor (IL-R) antagonist (DIRA) is a rare genetic auto-inflammatory disease related to alterations in antagonists of the IL-1 pathway. We aimed to summarize the efficacy and safety of IL-1 antagonists drugs in DIRA following the scoping review methodology.

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



- The length of therapy) varied between 2 weeks and 4.5 years.
- Anakinra was associated with transient injection site reactions (n=3) and anaphylactic reactions (n=2).
- Discontinuation of anakinra in 9 patients was reported following a flare-up of their disease.
- 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

1. The Joanna Briggs Institute. The Joanna Briggs Institute Reviewer's Manual 2015. http://joannabriggs.org/assets/docs/sumari/Reviewers-Manual_Methodology-for-JBIScoping-Reviews_2015_v2.pdf (accessed 29 Sept 2018.)
2. PRISMA Extension for Scoping Reviews (PRISMA-ScR): Checklist and explanation. <http://annals.org/aim/fullarticle/2700389/prisma-extension-scoping-reviews-prismascr-checklist-explanation> (accessed 29 Sept 2018.)

