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Title: Burden of Idiopathic Multicentric Castleman Disease (iMCD) in the US: A Population-Level Real World Analysis using a Health-Claims Dataset

Introduction: Idiopathic Multicentric Castleman Disease (iMCD) is a rare cytokine-driven disorder characterized by generalized lymphadenopathy and chronic inflammation. The purpose of the present study was to refine the previously developed administrative-claims diagnostic algorithm to more accurately capture iMCD cases and analyze trends in the use of siltuximab.

Methods: We developed three different claims-based algorithms using a combination of the CD-specific diagnostic code (ICD-10-CM: D4.7Z2), corresponding claim with diagnosis codes for iMCD minor criteria, exclusion of HHV-8 and HIV infection and siltuximab treatment claims using the MarketScan Commercial and Medicare administrative claims databases between October 1, 2016 and July 31, 2023.

Results: Of 868 people with at least one diagnosis of CD and analyzed using our selected algorithm, we identified 186 iMCD patients after exclusion of 52 HIV/HHV-8 patients and 65 patients with CD mimics. In the entire iMCD cohort, 22% had a claim for siltuximab.

Conclusions:
The utility of health-claims based datasets to conduct studies on epidemiology, disease burden and treatment patterns in iMCD is dependent on an accurate patient pool. An algorithm relying on use of CD-specific ICD-10 code in conjunction with international diagnostic criteria and exclusion of CD mimics may provide the most accurate estimates of prevalent cases. Although there has been a doubling in the proportion of iMCD patients receiving siltuximab between the periods 2017-2018 and added analysis extending to 2023, the number of treated patients remain dismal indicating a high unmet treatment need.