

## **The incidence of Castleman disease in The Netherlands: a retrospective study based on Dutch pathology reports.**

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**Background:** Castleman disease (CD) constitutes a group of lymphoproliferative disorders involving one lymph node (unicentric) or multiple lymph nodes (multicentric). Diagnosis is challenging due to its rarity and nonspecific clinical and histologic characteristics. Recognizing idiopathic multicentric CD (iMCD) is especially important, as patients often develop life-threatening multi-organ failure. CD incidence in the US is estimated at ~6 per million person-years (PYs), with iMCD at ~3 per million PYs [1]. The incidence in the Netherlands is currently unknown.

**Methods:** Using the Palga database, we requested reports mentioning CD or related terms between 2000 and 2022 and included additional lymph node reports for each index case. Reports were categorized as definitive, highly probable, unlikely, or definitely not CD based on microscopy and conclusion fields.

**Results:** We identified 810 patients with lymph node biopsies/excisions (1318 reports). Among these, 446 were definitive or highly probable CD cases (52% male, mean age 44.8 years, SD 18.4 years). Subtypes included 43% hyaline vascular, 33% plasmacytic, 4% mixed, and 20% unspecified. Although a diagnosis of iMCD cannot be based on histology alone, we tried to estimate how many cases could potentially fit. After excluding cases with HIV, lymphoma, HHV8, or a clear designation as being unicentric, 159 patients were considered potential iMCD cases.

**Conclusion:** The estimated CD incidence in the Netherlands is 1 per million PYs, ~6 times lower than US rates. Methodological differences, surgical approaches and clinical and diagnostic practices may partly explain this discrepancy, but we consider it likely that a true difference in incidence exists.

1. Mukherjee S, Fajgenbaum D, et al. Epidemiology and treatment patterns of idiopathic multicentric Castleman disease in the era of IL-6 directed therapy. *Blood Adv.* 2022 Jan 25;6(2):359-367.