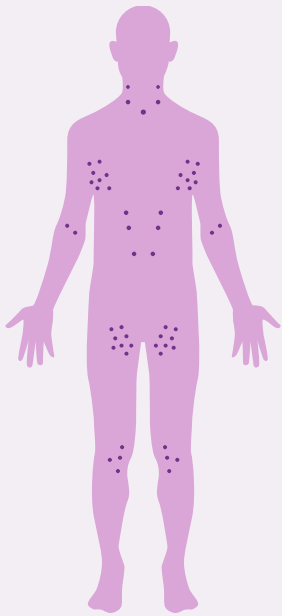


Multicentric Castleman's Disease (MCD)

A rare disorder of the lymph node system

What is Castleman's Disease (CD)?



Castleman's disease (CD) is a rare, serious disease of lymph nodes and related tissues.¹

CD is not officially a cancer, but the multicentric form of this disease acts very much like lymphoma (cancer of lymph nodes).¹

There are two forms of CD, each of which affect people very differently:¹

- **Unicentric Castleman's disease (UCD)** affects only a single area or group of lymph nodes.
- **Multicentric Castleman's disease (MCD)** affects more than one group of lymph nodes.

What is Multicentric Castleman's Disease (MCD)?



LYMPHOCYTES

MCD is a disorder in which **lymphocytes**, a certain type of white blood cells, are **over-produced and lead to enlargement of lymph nodes** or other internal organs containing lymphoid tissue.^{1,2}

- This can cause a variety of symptoms and weaken the immune system, making it hard to fight infection.^{1,2}
- Infections in people with MCD can be very serious and may even be fatal.¹

Multicentric disease is less common and more aggressive than unicentric disease³



IL-6

- While the official cause of CD currently is unknown, overproduction of interleukin-6 (IL-6) is considered a key pathogenic mechanism in MCD.^{1,3}
- People infected with the human immunodeficiency virus (HIV), the virus causing AIDS, may be at increased risk for MCD.¹

Prevalence, Incidence and Patients

CD is so rare that it is difficult to track the number of cases worldwide. Experts place the number at fewer than **1 in 100,000** people.^{1,4}



In Europe, diseases are categorised as rare if they affect **less than five in 10,000** people (0.05%).⁵

RARE:
<0.05%

The average age range of those diagnosed with MCD is between **50 and 60**.⁷



Symptoms



Symptoms of MCD may include enlarged nodes (appear as lumps under the skin), fever, weakness, fatigue, night sweats, weight loss, loss of appetite, nausea, vomiting, nerve damage that leads to numbness and weakness.¹

Some symptoms can be life threatening.^{7,8}

Diagnosis

As with other rare diseases, it can be **difficult to diagnose CD**, especially because the **symptoms are often similar to those of other diseases** and conditions.⁹



CD is formally diagnosed through **lymph node biopsy**. Other diagnostics used to confirm disease include:¹



Physical examination



Blood tests



Imaging tests

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References

- 1 American Cancer Society. Castleman disease. Available from: <http://www.cancer.org/acs/groups/cid/documents/webcontent/003093-pdf.pdf>.
- 2 Memorial Sloan Kettering Cancer Center. Uncommon Lymphoproliferative Disorders. Available from: <http://www.mskcc.org/cancer-care/adult/rare-hematologic-disorders/uncommon-lymphoproliferative-disorders>.
- 3 El-Osta HE, Kurzrock R. Castleman's disease: from basic mechanisms to molecular therapeutics. *Oncologist*. 2011;16(4):497-511.
- 4 Farruggia P et al. Castleman's disease in childhood: report of three cases and review of the literature. *Italian Journal of Pediatrics*. 2011; 37(50).
- 5 European Commission. Rare diseases – what are they? Available at: http://ec.europa.eu/health/rare_diseases/policy/index_en.htm
Last accessed December 2013.
- 6 Dispenzieri A, Gertz MA. Treatment of Castleman's disease. *Current Treatment Options in Oncology*. 2005;6:255-266.
- 7 Greiner T, Armitage JO, Gross TG. Atypical Lymphoproliferative Diseases. *American Society of Hematology Education Program*. 2000:133-146.
- 8 Peterson, B. Multicentric Castleman's disease. *Seminars in Oncology*. 1993 Dec;20(6):636-47.
- 9 Bonekamp D, Horton KM, Hruban RH, Fishman EK. Castleman disease: the great mimic. *Radiographics*. 2011;31:1793-1807.