

# IgG4-related hypertrophic pachymeningitis treatment

The diagnosis of [IgG4-related hypertrophic pachymeningitis](#) is challenging but is of great relevance as treatment differs significantly from other forms of pachymeningitis and a specific therapeutic approach may avoid long-term neurological complications <sup>1)</sup>.

There is no specific treatment protocol for IgG4 related pachymeningitis <sup>2)</sup>.

High-dose steroids are the initial treatment of choice; other immunosuppressive agents have shown variable efficacy in reducing meningeal hypertrophy and mostly used in disease relapse setting <sup>3)</sup>

Second-line is chemotherapy ([methotrexate](#), [azathioprine](#), etc.).

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Surgery is necessary when lesions progress and patients start to develop cranial nerve function deficit <sup>4)</sup>.

## Rituximab

see [Rituximab for IgG4-related hypertrophic pachymeningitis treatment](#).

<sup>1)</sup>

Esmailzadeh M, Dadak M, Atallah O, Möhn N, Skripuletz T, Hartmann C, Banan R, Krauss JK. IgG4-related hypertrophic pachymeningitis with tumor-like intracranial and intracerebral lesions. *Acta Neurochir (Wien)*. 2022 Aug 17. doi: 10.1007/s00701-022-05340-5. Epub ahead of print. PMID: 35974231.

<sup>2)</sup>

Gospodarev V, Câmara J, Chakravarthy V, Perry A, Wood M, Dietz R, Wang J, De Los Reyes K, Raghavan R. Treatment of IgG4-related pachymeningitis in a patient with steroid intolerance: The role of early use of rituximab. *J Neuroimmunol*. 2016 Oct 15;299:62-65. doi: 10.1016/j.jneuroim.2016.08.009. Epub 2016 Aug 16. PMID: 27725123.

<sup>3)</sup>

Lu LX, Della-Torre E, Stone JH, Clark SW. IgG4-related hypertrophic pachymeningitis: clinical features, diagnostic criteria, and treatment. *JAMA Neurol*. 2014 Jun;71(6):785-93. doi: 10.1001/jamaneurol.2014.243. PMID: 24733677.

<sup>4)</sup>

Yu Y, Lv L, Yin SL, Chen C, Jiang S, Zhou PZ. Clivus-involved immunoglobulin G4 related hypertrophic pachymeningitis mimicking meningioma: A case report. *World J Clin Cases*. 2022 Jun 26;10(18):6269-6276. doi: 10.12998/wjcc.v10.i18.6269. PMID: 35949844; PMCID: PMC9254204.

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