

Diagnosis and Management of Adult Primary Angiitis of Central Nervous System: an International Survey on Current Practice

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Background

Primary angiitis of the central nervous system (PACNS) is a rare disease, with an estimated prevalence of 2.4 cases per 1 million person-years.¹ Our understanding of the disease is based on a few single-center¹⁻⁴ and multi-center^{5,6} cohorts of limited size. No validated guidelines exist to guide clinicians in the diagnosis and treatment of PACNS, and the level of agreement between clinicians in the management of the disease remains unknown. This international online survey aims at reporting clinical practices of physicians who manage PACNS in adults, by identifying main commonalities but also differences and challenges.

Methods

Two investigators (AN and HDB) developed an online survey (<https://fr.surveymonkey.com/>), which was improved from other investigators' (MB, SL, JPM, CP, MR and ET) reviews and comments. Implementation (completion time) and efficacy (absence of ambiguity or redundancy) was verified by 5 physicians (neurologists and internists) at our institutions (Caen, France and Montreal, Canada) prior to final modifications and dissemination.

The survey will be distributed through the newsletters of Canadian and European vasculitis research groups, internal medicine, rheumatology, and neurology societies. (Table 1). If possible, a reminder will be sent out 6 weeks after the initial newsletter. The survey will remain open for 3 months. Participants will consent to the survey on the Survey Monkey platform. No financial compensation is planned. The project was approved by the ethics committee at the CHU Caen-Normandie.

The current study includes a screening phase, followed by an extensive survey for respondent meeting the eligibility criteria. The screening phase collects data on the training specialty, years in practice, country of practice, type of practice, and estimated number of adult patients with PACNS (proven or presumed) for whom they were directly involved as treating physicians during their entire career. Respondents who were directly involved as treating physicians for 2 or more patients will be enrolled in the extensive survey. Subsequent survey questions examine 3 key areas: diagnostic work-up, pharmacological treatment, and monitoring of disease activity.

Table 1 – Societies

Internal Medicine/Rheumatology	Neurology
Canadian Vasculitis Research Network	Canadian Neurological Sciences Federation
Société Nationale Française de Médecine Interne	Canadian Stroke Consortium
Société Française de Rhumatologie	Société Française de Neurovasculaire
GFEV	European Stroke Organization
EUVAS	Société des Sciences Vasculaires du Québec

Canadian Rheumatology Association	
Société des Sciences Vasculaires du Québec	
Canadian Stroke Consortium	

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