

Southern Alberta Vasculitis Patient Registry: Creation And Utility

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Vasculitides encompass a group of rare autoimmune diseases where inflammation of vessels causes multisystem organ damage (i.e. kidneys, lungs, nervous system, Figure 1). The overall incidence is currently estimated to be 10-20/million/year, however there is an increasing trend noted in the pediatric population in Alberta^{1,2}.

As with other rare autoimmune diseases such as lupus, the limited number of patients impairs recruitment into clinical trials, impedes research, and hinders the generation of evidence-based treatment recommendations and protocols⁴. Moreover, the low patient numbers provide a barrier to obtaining reliable rare disease prevalence statistics and to gaining a clear understanding of the natural history of specific vasculitides.

To address some of these limitations, we propose to generate a prospective vasculitis registry for patients in Southern Alberta. This initial registry will serve as a foundation for collaboration with other established or prospective centres at the provincial, national, and international levels (Figure 2). The contribution of Canadian centres is coordinated through the Canadian network for research on vasculitides (CanVasc), representing a network of physicians with expertise in vasculitis. To ensure the seamless amalgamation of information among the centres, the same nomenclature and database platform will be used by each provincial vasculitis centre. The establishment of such a registry

will create a framework for knowledge translation, discovery, and best practice management.

Methods

Recruitment will initially be limited to patients with vasculitis residing in Southern Alberta who have been referred to the Rheumatology division in Calgary through the central triage system to ensure capture of patients with a confirmed diagnosis. Ethics approval for this research project was obtained in May 2012. Patients complete written informed consent at their baseline visit in the Rheumatology vasculitis clinic at the South Health Campus in Calgary, Alberta. Upon consenting, the patients are examined yearly by Dr. Aurore Fifi-Mah, a Rheumatologist at the South Health Campus. The research team will complete yearly case report forms where information on patient symptoms, test results, response to treatment, activity of disease and complications will be longitudinally collected. Each patient will also complete a Patient Quality of Life form (SF-36).

Patient consent also extends to the collection of de-identified sera for storage in the Mitogen Advanced Diagnostics Laboratory in Calgary, Alberta, under the direction of Dr. Fritzler. As new diagnostic and prognostic biomarkers are identified, these sera will constitute a biobank to be analyzed for the presence of new target antibodies and proteins. Such information can enhance understanding of the pathogenesis of these diseases to inform and direct future therapeutic approaches. Biomarkers could also help with early

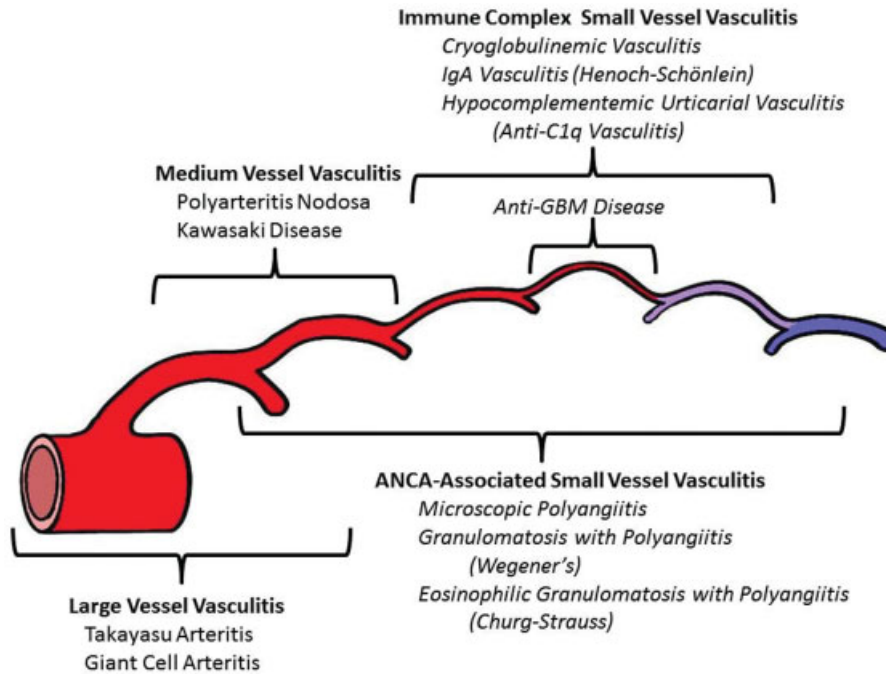


Figure 1:

The 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides³.

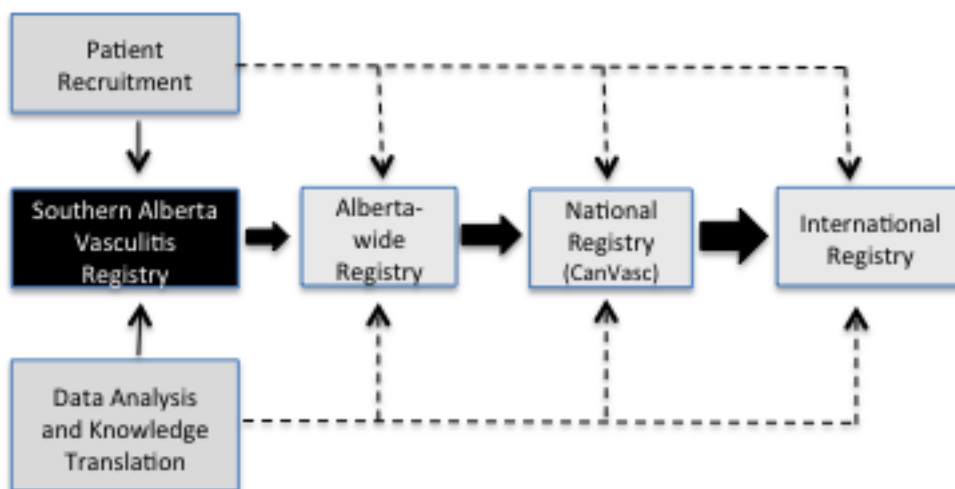


Figure 2:

Schematic of expansion of the Southern Alberta Vasculitis Registry. (CanVasc: Canadian network for research on vasculitides; <http://www.canvasc.ca>)

Table 1: *Current numbers and proportions of patients categorized by specific vasculitides in the Southern Alberta Vasculitis Patient Registry.*

Diagnosis	Number	Percentage
ANCA-associated Vasculitis	12	11.2
Behçets Disease	7	6.5
Connective Tissue Disease Associated Vasculitis	6	5.6
Central Nervous System Vasculitis	4	3.7
Cryoglobulinemic Vasculitis	2	1.8
Giant Cell Arteritis	9	8.4
IgA Vasculitis	5	4.6
Leukocytoclastic Vasculitis	11	10.2
Polyarteritis Nodosa	11	10.2
Polymyalgia Rheumatica	13	12.1
Takayasu Arteritis	6	5.6
Vasculitis Associated with Other Disease	22	20.5

detection of disease relapse to reduce permanent organ damage and improve prognosis.

As this is an observational study, no power calculation is involved. Descriptive analysis will be used to summarize participant characteristics and comorbid conditions.

Results

The initial project infrastructure has been successfully established. To date, 107 patients have been recruited (Table 1).

Discussion and Conclusions

The establishment of a Southern Alberta Vasculitis Patient Registry allows the development of a prospective cohort of patients with vasculitis seen in Calgary, the catchment area for Southern Alberta. The data collected include clinical and serological information that will be captured in a database. The development of a biobank of sera with access to an internationally recognized immunology laboratory in Calgary will provide the basic science and technical support to improve diagnosis and management of patients based on their biomarker profile. This will help address issues of inaccurate rare disease statistics, inadequate understanding of disease natural history, and limited diagnostic and treatment regimes. Through systematic collection and analysis of data, the registry will create a foundation of knowledge on

which to build informed, standardized models of care by identifying gaps in patient care.

Moreover, the additional benefit of serum analysis may permit the discovery of specific biomarkers and prognostic factors to classify and predict the future course of different vasculitis subtypes. This will permit individualized, patient-centred treatment.

The extension of this project to include centres provincially, nationally, and internationally would magnify the utility of the initial project. The previous establishment of provincially-based vasculitis research centres through CanVasc expedites project expansion.

References

1. Ntatsaki E, Watts RA, Scott DGI. *Rheum Dis Clin North Am.* 36:447-61, 2010
2. Grisaru S et al. *J Rheumatol* 37:440-442, 2010.
3. Jennette JC et al. *Arthritis and Rheumatism* 65:1-11, 2013.
4. Barbour S et al. *BMC Nephrology* 14:236, 2013.