RHEUMATOLOGY RESEARCH JOURNAL

Vol. 9, No. 2, April 2024,
Webpage: http://rheumres.org
Email: rheumres@gmail.com
ISSN:2476-5856
doi: 10.32592/RR.2024.9.2.103

Original Article **Open Access**

Development and organization of Tabriz University of Medical Sciences Vasculitis Registry: TUOMS-VR

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This manuscript describes the establishment and initial results of a patient-driven registry in Tabriz, Iran to collect demographic, clinical, and paraclinical data, treatment and outcomes of patients with vasculitis. The Tabriz University of Medical Sciences Vasculitis Registry (TUOMS-VR), a patient-driven, prospective and web-based disease registry system, is conducted at the Connective Tissue Diseases Research Center, TUOMS located in Tabriz, Iran. The primary objective of this registry is to comprehensively document prospective data of patients diagnosed with systemic or single-organ vasculitis according to the Chapel Hill Consensus Conference nomenclature and definitions. To date, the registry has successfully enrolled 743 patients. The gender distribution within this existing cohort is balanced, with 50.5% male and 49.5% female participants. The most frequently diagnosed condition was Behcet's disease, representing 56.5% of cases. Polymyalgia rheumatica also emerged as a notable diagnosis, accounting for 13.7% of cases. By providing a comprehensive and prospective documentation of vasculitis cases, the TUOMS-VR aims to enhance vasculitis patients care and outcomes.

Keywords: *vasculitis; registry; outcome; treatment; damage; remission*

Introduction

Vasculitis comprises a spectrum of rare diseases marked by inflammation of blood vessels, leading to organ ischemia and damage. It affects individuals across all age groups and is classified based on the predominant size of the involved vessel. These classifications include large vessel vasculitis (including giant cell arteritis (GCA) and Takayasu arteritis), medium vessel vasculitis (including polyarteritis nodosa (PAN) and Kawasaki disease), small vessel vasculitis (including anti-neutrophil cytoplasmic auto-antibody (ANCA)-associated vasculitis (AAV), IgA vasculitis, and hypocomplementemic urticarial vasculitis), and variable vessel vasculitis (Behcet's disease (BD) and Cogan's Syndrome) [1]. Vasculitis is usually chronic in nature, characterized by significant morbidity and mortality and an increased likelihood of recurrence [2]. It is crucial to emphasize the importance of prevention, early detection, and effective management of vasculitis-related

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Received: 13 April 2024; **Accepted:** 14 April 2024

damage [3]. Damage in vasculitis often occurs early in the disease progression, its severity varies depending on the type of vasculitis, and is closely associated with the disease course and its treatment [3]. Damage significantly influencing prognosis [3]. The literature lacks sufficient data regarding the long-term prognosis, co-morbidities and mortality rates among vasculitis patients in routine clinical settings, beyond controlled trials. Moreover, the intricate nature of vasculitis, which involves challenges in differential diagnosis, the necessity for inter-disciplinary cooperation, treatment administration, outcome assessment, and continuous monitoring for relapses, contributes to the complexity in managing these patients [4]. Hence, there is a continuing need for registries that systematically document the characteristics, progression, treatments, comorbidities, and long-term outcomes of vasculitis patients at multiple centers in a standardized and prospective manner. Establishing such registries will facilitate the prospective recording of these rare diseases, allowing for the standardized documentation of disease outcomes under the guidance of physicians specialized in the care of vasculitis patients [2]. This information will also reveal how treatment instructions and therapy standards are put into practice in local centers that manage vasculitis patients, offering valuable insights into the present state of medical care. Moreover, data from the registry could serve as a foundation for identifying areas in healthcare infrastructure that require enhancement, with a focus on addressing the specific needs of individuals with vasculitis [2]. In recent times, the growing number of clinical trials focusing on vasculitis, along with the necessity to collect long-term data regarding treatment safety and effectiveness in everyday practice, has spurred the establishment of multiple vasculitis registries across Europe and the United States [5-7]. The majority of these registries are web-based, facilitating straightforward access and data input for participating centers. These registries serve various purposes including research, patient care, and healthcare strategy development. In this paper, our aim is to present the protocol, methodology, and current status of a vasculitis registry developed in Tabriz, Iran.

Materials and Methods

Objectives

The primary objective of this registry is to comprehensively document prospective data of patients diagnosed with systemic or single-organ vasculitis according to the Chapel Hill Consensus Conference (CHCC) nomenclature and definitions [8].

Study Design

The registry adopts a prospective approach and functioning as a web-based registry.

Setting

The Tabriz University of Medical Sciences Vasculitis Registry (TUOMS-VR), a patient-driven, prospective and web-based disease registry system, is conducted at the Connective Tissue Diseases Research Center, TUOMS, located in Tabriz, Iran. This registry was launched in Jan 2019. The center is dedicated to providing specialized care for vasculitis patients, with a team of licensed medical specialists, specialized outpatient departments, and wards within secondary and tertiary care facilities. The focus of this study is on capturing a comprehensive representation of the target patient population within this region. Eligible patients receive care within this specialized center, and data collection is facilitated through a web-based registry system. The study involves both prospective data collection of newly diagnosed patients and retrospective data collection of patients' medical histories within this setting.

Ethics

The study protocol (ethical code: IR.TBZMED.REC.1396.271) was approved by the local Ethics Committee of the TUOMS, Tabriz, Iran. Informed consent will be obtained from all study subjects.

Subjects

The registry will include patients diagnosed with vasculitis based on expert opinion and ruling out of vasculitis mimickers. Patients of all ages will be enrolled. Patients will undergo prospective registration and assessment at baseline, followed by evaluations every 1-6 months or upon suspected or confirmed relapse. Data will be entered into an electronic web-based platform. Table 1 displays the vasculitis entities that meet the criteria for inclusion in this registry.

Table 1: Table 1. Vasculitis types eligible for inclusion in the TUOMS-VR

Vasculitis classification		
1. Large vessel vasculitis	Giant cell arteritis (GCA)	
	Takayasu arteritis	
2. Medium size vessel vasculitis	Polyarteritis nodosa (PAN)	
	Kawasaki disease	
	Polymyalgia rheumatic (PMR)	
3. Small vessel vasculitis	ANCA-associated vasculitides	Microscopic Polyangiitis (MPA)
		Granulomatosis with Polyangiitis (GPA)
		Eosinophilic Granulomatosis with Polyangiitis (EGPA)
	Immune complex vasculitis	IgA vasculitis (Henoch-Schönlein purpura)
		Hypocomplementemic urticarial vasculitis (Anti-C1q vasculitis)
		Anti-GBM (glomerular basement membrane) disease
		Cryoglobulinemic vasculitis
		Hypersensitivity vasculitis (HSV)
4. Variable vessel vasculitis	Behcet's disease (BD)	
	Cogan's syndrome	
5. Single organ vasculitis	Cutaneous (leukocytoclastic) small vessel vasculitis	
	Cutaneous arteritis	
	Primary central nervous system vasculitis	
	Others	
6. Vasculitis associated with probable etiology	Infection	
	Malignancy	
	Medication	
	Connective tissue diseases	

TUOMS-VR, Tabriz University of Medical Sciences Vasculitis Registry; ANCA, anti-neutrophil cytoplasmic autoantibody

Interventions

This registry operates solely in an observational capacity, relying on the documentation of routine data. Therefore, no study-specific interventions are administered.

Data collection methods and data management

Our aim with the TUOMS-VR is to comprehensively document and analyze various demographic and clinical factors pertinent to vasculitis patients. From basic demographics such as age, body mass index, and city of residence to detailed medical history including disease duration, age at onset, smoking status, and comorbidities, we aim to build a comprehensive profile of each patient.

Additionally, we meticulously record clinical manifestations and laboratory findings during each visit, including results from bone densitometry and vascular imaging studies, as well as any biopsy results. This information allows us to accurately diagnose and assess disease activity, utilizing standardized disease activity indices and vasculitis damage indices. Furthermore, we monitor treatment regimens and disease activity status, while also documenting any side effects experienced from treatment. All this data is entered into electronic forms directly by expert physicians.

Statistical Analysis

Statistical analysis will be conducted based on the specific type and nature of the data being processed, in accordance with the particular objectives of studies carried out by our network. In all cases, the analysis will encompass fundamental principles of descriptive statistics, correlations among groups, and comparisons between subgroups. More detailed information regarding statistical methodologies will be provided in future publications stemming from data collected in TUOMS-VR.

Results

As an ongoing initiative, the TUOMS-VR continues to evolve, and the data presented here represent the registry's state up until April 2024. To date, the registry has successfully enrolled 743 patients, with plans to expand this cohort as the project progresses. The gender distribution within this existing cohort is balanced, with 50.47% male and 49.53% female participants. Diagnoses within the registry were established through clinical assessments by physicians.

The most frequently diagnosed condition was BD, representing 56.5% of cases (n = 420), highlighting its significant prevalence within the registry cohort. Polymyalgia rheumatica (PMR) also emerged as a notable diagnosis, accounting for 13.7% of cases (n = 102). These two conditions together underscore the diverse clinical spectrum of vasculitis encountered within the registry. Further analysis of the registry data revealed the presence of various other vasculitis subtypes, each contributing to the overall disease landscape. Among these, granulomatosis with polyangiitis (previously known as Wegener's granulomatosis) and Takayasu arteritis were notably present. Other diagnosed conditions included PAN and GCA. Additionally, less common forms of vasculitis, such as urticarial vasculitis, eosinophilic granulomatosis with polyangiitis (formerly Churg-Strauss Syndrome), and Henoch-Schönlein Purpura, contributed to the registry's comprehensive disease landscape, though they occurred with lesser frequency.

It's significant to note that certain forms of vasculitis, such as Kawasaki disease, microscopic polyangiitis, vasculitis affecting the central nervous system, hypersensitivity vasculitis, and vasculitis associated with other underlying conditions, were encountered infrequently in our group of patients. This aspect highlights the registry's ability to encompass a broad spectrum of vasculitis types, thereby providing essential epidemiological insights into these multifaceted disorders. For comprehensive insights into the distribution and prevalence of each vasculitis subtype diagnosed in our cohort, detailed numerical data and percentages are provided in Table 2.

Table 2: Distribution of vasculitis subtypes within the TUOMS-VR

Vasculitis Subtype	Frequency	Percentage
Behcet’s disease	420	56.52
Polymyalgia rheumatic	102	13.72
Polyarteritis nodosa	65	8.74
Granulomatosis with Polyangiitis	56	7.53
Takayasu Arteritis	40	5.38
Eosinophilic Granulomatosis with Polyangiitis	16	2.15
Giant cell arteritis	16	2.15

Hypocomplementemic urticarial vasculitis (anti-C1q vasculitis)	9	1.21
IgA vasculitis (Henoch-Schönlein purpura)	7	0.94
Cutaneous (leukocytoclastic) small vessel vasculitis	4	0.53
Vasculitis associated with connective tissue diseases	3	0.40
Hypersensitivity vasculitis	2	0.26
Primary central nervous system vasculitis	2	0.26
Microscopic polyangiitis	1	0.13
Kawasaki disease	1	0.13

TUOMS-VR, Tabriz University of Medical Sciences Vasculitis Registry

Discussion

The TUOMS-VR was launched in Jan 2019, marking a significant step forward as the first prospective registry dedicated to vasculitis patients within northwest Iran. The TUOMS-VR offers an extensive view of the different types of vasculitis across a substantial number of patients, delivering crucial information for grasping the prevalence, demographic profiles, and clinical features of these conditions. This comprehensive data collection acts as a pivotal foundation for subsequent studies, with the goal of refining the accuracy of diagnoses and improving treatment strategies for individuals with vasculitis. Since its inception, the registry has been on a continuous trajectory of growth, with the inclusion of patients and expansion of participating sites. As of April 2024, the registry has successfully enrolled 743 patients, reflecting a broad and diverse patient population.

Previous research efforts in this field were often constrained by single-center data collection, retrospective designs, and a limited scope in terms of outcomes measured and patient numbers. This situation has created a gap in our understanding, particularly when it comes to representing the full range of patient experiences and disease manifestations. The contrast seen between the controlled conditions of clinical trials and the day-to-day realities observed in broader patient cohorts serves to underline the critical importance of gathering authentic, wide-ranging data that mirrors the actual experiences of individuals living with vasculitis. By providing a comprehensive and prospective documentation of vasculitis cases, the TUOMS-VR aims to fill critical gaps in our understanding of these conditions and ultimately enhance patient care and outcomes.

Acknowledgments

The authors of this manuscript thanks TUOMS for supporting this registry.

Conflict of Interests

The authors declare no conflict of interest regarding the publication of this paper.

Funding

This registry was supported by TUOMS.

References

1- Watts RA, Hatemi G, Burns JC, Mohammad AJ. Global epidemiology of vasculitis. ***Nat Rev Rheumatol*** 2022; 18(1):22-34. doi: 10.1038/s41584-021-00718-8.

2- Iking-Konert C, Wallmeier P, Arnold S, Adler S, de Groot K, Hellmich B, et al. The Joint Vasculitis Registry in German speaking countries (GeVas) – a prospective, multicenter registry for the follow-up of long-term outcomes in vasculitis. ***BMC Rheumatol*** 2021; 5(1):40. doi: 10.1186/s41927-021-00206-2.

3- Robson J, Doll H, Suppiah R, Flossmann O, Harper L, Höglund P, et al. Damage in the anca-associated vasculitides: long-term data from the European vasculitis study group (EUVAS) therapeutic trials. ***Ann Rheum Dis*** 2015; 74(1):177-84. doi: 10.1136/annrheumdis-2013-203927.

4- Thomas K, Panagiotopoulos A, Banos A,

Argyriou E, Boki K, Boumpas D, Vassilopoulos D. Development of an ANCA-Associated Vasculitides Patient Registry in Greece. ***Mediterr J Rheumatol*** 2020;31(1):84-6. doi: 10.31138/mjr.31.1.84.

5- Sánchez Álamo B, Moi L, Bajema I, Faurschou M, Flossmann O, Hauser T, et al. Long-term outcomes and prognostic factors for survival of patients with ANCA-associated vasculitis. ***Nephrol Dial Transplant*** 2023; 38(7):1655-1665. doi: 10.1093/ndt/gfac320.

6- Egan AC, Kronbichler A, Neumann I, Bettiol A, Carlson N, Cid MC, et al. The sound of interconnectivity; the European vasculitis society 2022 report. ***Kidney Int Rep*** 2022; 7:1745–57. doi: 10.1016/j.ekir.2022.05.018.

7- Bajema IM, Bruijn JA, Casian A, Cid MC, Csernok E, van Daalen E, et al. The European Vasculitis Society 2016 Meeting Report. ***Kidney Int Rep*** 2017; 2(6):1018-31. doi: 10.1016/j.ekir.2017.09.008.

8- Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 revised international Chapel Hill consensus conference nomenclature of Vasculitides. ***Arthritis Rheum*** 2013; 65(1):1-11. doi: 10.1002/art.377