Table 1. Tocilizumab therapy in ten cases of refractory arterio-BD

Patient	Age, year	Disease duration, months	Clinical features ^a	Arterial lesions	Previous therapy	Response at week 24
1	34	228	O, G, P, S, I	Stenosis of CA/ SMA/RA/ SCA, aortic valve prolapse	Pred/CYC/ MMF	CR
2	20	24	O, S	Dissecting aneurysm of AA (Debaque I)	Pred/MMF	PR
3	67	276	O, G, P, S, A	thoracoabdominal aortic aneurysm	Pred/CYC/ TAC	CR
4	67	75	O, S, U	Stenosis of LAD/LCX/RCA	Pred /CYC	CR
5	50	80	O, G, A	Abdominal and coronary aortic aneurysms	Pred /CYC	Relapse
6	48	26	O, G	Aortic insufficiency	Pred /CYC	PR
7	26	147	O, P, S, V	iliac artery aneurysm	Pred /MMF	CR
8	49	466	O, S, V	Thoracoabdominal and coronary aortic aneurysms	Pred /CYC/ AZA	CR
9	27	181	O, P, S	Pseudoaneurysm of CCA	Pred /CYC	PR
10	Male/55	354	O, P, S	Abdominal aneurysm, stenosis of LAD/LCX/RCA	Pred /CYC/ AZA	CR

^aO: oral ulcer; G: genital ulcer; P: pathology test; S: skin lesions; I: intestinal ulcer; A: arthritis; U: uveitis; V: venous thrombosis

Figure 1. Changes from baseline in BSAS, BDCAF, ESR, CRP and steroid daily dose at 24 weeks

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AB0373

PSYCHOMETRIC PROPERTIES OF OUTCOME MEASUREMENT INSTRUMENTS FOR LARGE VESSEL VASCULITIS: A SYSTEMATIC LITERATURE REVIEW

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Background: Giant cell arteritis (GCA) and Takayasu arteritis (TAK) are two forms of chronic progressive large-vessel vasculitis (LVV) of unknown etiology. In 2016, the OMERACT Vasculitis Working Group proposed the development of a Core Domain Set of outcome measures for LVV including organ and arterial function, fatigue, pain, biomarkers, and death (1). Understanding the psychometric properties of outcome measurement instruments is essential when selecting among instruments to use in research; a summary of such properties for measures of LVV has not been developed.

Objectives: To systematically review and summarize the psychometric properties of outcome measurement instruments used to measure the domains of the OMERACT core domain set for LVV.

Methods: A comprehensive search of several databases (Medline, EMBASE, Cochrane, among others) from inception to July 14, 2020 was conducted. Articles were included if they covered psychometric properties of instruments used in LVV. Following the COSMIN and OMERACT frameworks, different psychometric properties (validity, inter- and intra-observer reliability, sensitivity to change, and feasibility) of outcome measurement instruments used in LVV (GCA and TAK) were assessed. Risk of bias was assessed according to the COSMIN checklist.

Results: Among the 3534 articles identified, 15 studies focusing on the development or validation of psychometric properties on LVV met the predefined criteria. Two were development studies and 13 were validation studies. These studies provided information on 13 instruments: 5 instruments specific to TAK, 2 specific to systemic vasculitides, and 6 general, non-disease-specific instruments. No instruments specific to GCA were identified.

Of the main psychometric properties assessed in the included studies, 40% had a low, 47% had moderate, and 13% had high risk of bias. Construct validity was the property most frequently assessed (in 93% of the tools) (Figure 1).

In TAK, the Indian Takayasu Clinical Activity Score 2010 (ITAS2010) showed good consistency ($r=0.97$), reliability (intra-observer, ICC=0.60; inter-observer, ICC=0.92) and validity (correlation with Physician Global Assessment (PGA) ($r=0.73$)) for disease activity. Regarding disease damage, the Disease Extent Index-Takayasu (DEI-Tak) showed good validity (correlation with NIH score 94%, $k=0.85$). Non-specific vasculitis instruments such as the Vasculitis Damage Index (VDI) and the Birmingham Vasculitis Activity Score (BVAS) showed moderate validity in the assessment of disease damage in GCA (cumulative glucocorticoid dose and disease duration, $r=0.30$ and $r=0.29$) and TAK (cumulative glucocorticoid dose and disease duration, $r=0.29$ and $r=0.25$) in the former and disease activity in GCA in the later (PGA, $r=0.50$).

Six non-vasculitis-specific patient-reported outcomes (PROs) instruments were identified, all showing low to moderate validity in GCA/TAK.

Psychometric properties among 13 outcomes instruments developed and/or validated for target-vessel vasculitis									
DOMAIN	INSTRUMENTS	VALIDITY				RELIABILITY			RESPONSENESS
		FACE	CONTENT	CONSTRUCT	CRITERION	INTER-OBSERVER	INTRA-OBSERVER	FEASIBILITY	
Disease activity	ITAS2010								
	ITAS-A								
	BVAS								
Disease activity and damage	DEI-Tak								
	Kerr et al NIH								
Disease damage	VDI								
	TAK-DI								
Patient-reported outcomes	HRPQ								
	ADVICE								
	SG-36								
	VCMI								
	PGA								
	MTI								

Abbreviations: ITAS2010-Indian Takayasu Activity Score 2010; ITAS-A-Indian Takayasu Activity Score A; DEI-Tak-Disease Extent Index-Takayasu; VDI-Vasculitis Damage Index; BVAS-Birmingham Vasculitis Activity Score; TAK-DI-Takayasu Disease Index; HRPQ-Health-Related Quality of Life; ADVICE-Activities of Daily Living Index; SG-36-Short-Form 36; VCMI-Vision Core Measurement; PGA-Physician Global Assessment; MTI-Multidimensional Fatigue Inventory.

Conclusion: The psychometric properties of 13 outcome measures to study LVV covering the OMERACT domains of disease activity, damage, and patient-reported outcomes were assessed. ITAS2010, DEI-Tak, VDI, and BVAS were the instruments with better psychometric properties for disease activity and/or damage. Disease activity and/or damage instruments specific for GCA, and validated PROs for both GCA and TAK are needed.

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AB0374

CHARACTERISTICS OF A GIANT CELL ARTERITIS COHORT IN LATINOAMERICA

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