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Vasculitis

A glossary of signs and symptoms of giant cell arteritis

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ABSTRACT

Objectives: This study seeks to create consensus-based definitions of signs and symptoms of giant cell arteritis (GCA) for use by health care professionals, primarily in research settings.
Methods: Core definitions of signs and symptoms of GCA were extracted from 11 randomised controlled trials of GCA previously reviewed in a systematic literature review conducted in the context of the development of response criteria for GCA. This information was supplemented by definitions from other sources, such as rheumatology textbooks. A 2-round Delphi was performed within an international task force (32 members from 11 countries). The first round aimed to obtain consensus on the descriptive terms defining each sign or symptom, and round 2 rated the importance of these terms. Based on the Delphi study method, preliminary definitions were developed. In 4 online meetings, results of the Delphi were reviewed, and a consensus was achieved on final definitions.
Results: Twenty-nine signs and symptoms of GCA were reviewed. Six signs or symptoms of GCA had previously been defined in the literature. A high level of agreement was reached on the definition of 23 signs and symptoms with the following 12 considered characteristic of GCA: headache, temporal artery abnormalities, scalp tenderness, scalp necrosis, jaw claudication, tongue claudication, tongue necrosis, amaurosis fugax, permanent vision loss, fever, limb claudication, and blood pressure inequality.
Conclusions: A glossary of definitions for 23 signs and symptoms of GCA was developed through a consensus process involving international experts.

WHAT IS ALREADY KNOWN ON THIS TOPIC

- The paucity of standardised definitions for clinical manifestations of giant cell arteritis (GCA) leads to variable interpretations in research settings. Thus, there is an unmet need to establish uniform definitions of signs and symptoms of GCA to ensure standardised patient enrolment into clinical trials and characterisation of patients in clinical studies.

WHAT THIS STUDY ADDS

- This study defined 23 signs and symptoms of GCA. Of these, 12 were considered characteristic of GCA and are as follows: headache, temporal artery abnormalities, scalp tenderness, scalp necrosis, jaw claudication, tongue claudication, tongue necrosis, amaurosis fugax, permanent vision loss, fever, limb claudication, and blood pressure inequality.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

- These 23 definitions can complement inclusion criteria in clinical trials and other clinical studies, allow precise application of classification criteria, and lay the groundwork for developing response criteria for GCA.

INTRODUCTION

Giant cell arteritis (GCA) is a large vessel vasculitis and the most common form of vasculitis in adults over the age of 50 [1,2]. There are no standardised definitions of the clinical manifestations of GCA in the literature. Classification criteria related manuscripts usually include definitions of individual parameters (for example, classification criteria for spondyloarthritis), whereas the classification criteria for GCA do not comprehensively include definitions for all signs and symptoms [3–5]. This lack of standardised definitions might lead to inconsistency in application in research settings. Furthermore, the diagnostic and therapeutic landscapes of GCA are rapidly expanding [6,7]. Inconsistent definitions of GCA features hinder the validity of the data and the inclusion of a homogeneous group of patients into clinical trials, thereby affecting the results and limiting the comparability of studies.

An international task force supported by the European Alliance of Associations for Rheumatology (EULAR) and the American College of Rheumatology (ACR) was established to develop response criteria for GCA. For the development of these criteria, a multistep approach is followed that includes but is not limited to a systematic literature review and a Delphi exercise to evaluate candidate descriptors in the response criteria [8]. During the Delphi, features of GCA (for example, jaw claudication) were defined to allow consistent interpretation when doing this exercise. While defining those signs and symptoms of GCA, it was noted that there were no standardised definitions in the literature. Therefore, it was decided to create a glossary of signs and symptoms of GCA. This glossary is intended to be used in research settings by health care professionals as a resource to facilitate the recruitment of patients into clinical trials by standardising the definitions of commonly used features of GCA.

METHODS

Initial development of the definitions of signs and symptoms of GCA

An international task force endorsed by EULAR and ACR to develop new response criteria for GCA conducted the current glossary project [8]. The 32 task force members consisted of 28 rheumatologists, 1 internist, 1 health professional in rheumatology, and 2 patient research partners from 11 countries (Austria, Canada, France, Germany, Italy, Netherlands, Portugal, Spain, Switzerland, United States of America, United Kingdom).

Two patient research partners were involved in each step of this study including but not limited to the Delphi study and consensus meetings.

Seven members (CAL, CD, ZT, SR, MS, MB, and CS-A) formed the Steering Committee, 3 of which are fellows (MS, MB, and CS-A). Using the systematic literature review (SLR) conducted in the context of the response criteria for GCA project as a basis, the fellows extracted definitions of the signs and symptoms of GCA from 11 randomised controlled trials (RCTs) (Supplementary Table S1) [8]. To ensure comprehensiveness, this information was supplemented by definitions from other sources, including rheumatology textbooks [9–11], the 2022 ACR/EULAR classification criteria for GCA [4], the ACR 1990 classification criteria for GCA [12], and a Delphi exercise prepared as part of the GCA response criteria project (which was different from the Delphi described below) (Fig).

For each sign and symptom of GCA, keywords or features (termed 'descriptors' in this paper) were extracted from each definition (for example, 'new onset') by 2 of the fellows (MS and MB). Descriptors that were frequently mentioned in the above sources (for example, 'new onset,' was mentioned in most RCTs) were incorporated into the potential new definition, whereas the other descriptors that were infrequently mentioned were kept as potential options. A model was created with a basic phrase that includes the common descriptors of the signs and symptoms of GCA that cannot be changed and several descriptors that can be added to make the definition comprehensive (Supplementary Fig S1). The information retrieved during this step was used as a basis for the subsequent Delphi exercise.

Delphi

The preliminary descriptors for each sign and symptom of GCA were refined via a 2-round Delphi performed among the task force members.

During round 1, participants were asked to select the descriptors that most appropriately matched each feature of GCA. Participants were also allowed to propose new descriptors.

Any descriptive term with ≥70% consensus was added to the definition of the respective sign/ symptom. Descriptors with <30% agreement were excluded. Descriptors with 30% to 70% agreement were reassessed in a second Delphi survey. The goal of round 2 was to rate the importance of the descriptors that most appropriately matched the features of GCA. The rating was as follows: 0–3 = not important, 4–6 = important, and 7–9 = critically important (Supplementary Fig S2).

Based on the results of the 2 Delphi rounds, a preliminary definition of each feature of GCA was developed by the Steering Committee as follows: those descriptors rated as critically important by ≥70% of participants were included in the preliminary definition. The rest of the descriptors were shown to task force members during subsequent meetings (below). The preliminary definitions were discussed and refined by the Steering Committee via e-mail and in online meetings until a final proposal was achieved, which was then presented to the task force.

Based on the Delphi results, the Steering Committee identified fundamental overarching principles essential for contextualising and interpreting the definitions of the features of GCA. These were also proposed to task force members.

Consensus meetings

The results of the Delphi exercise and preliminary definitions were presented to the task force members during 4 online meetings (Supplementary Fig S3).

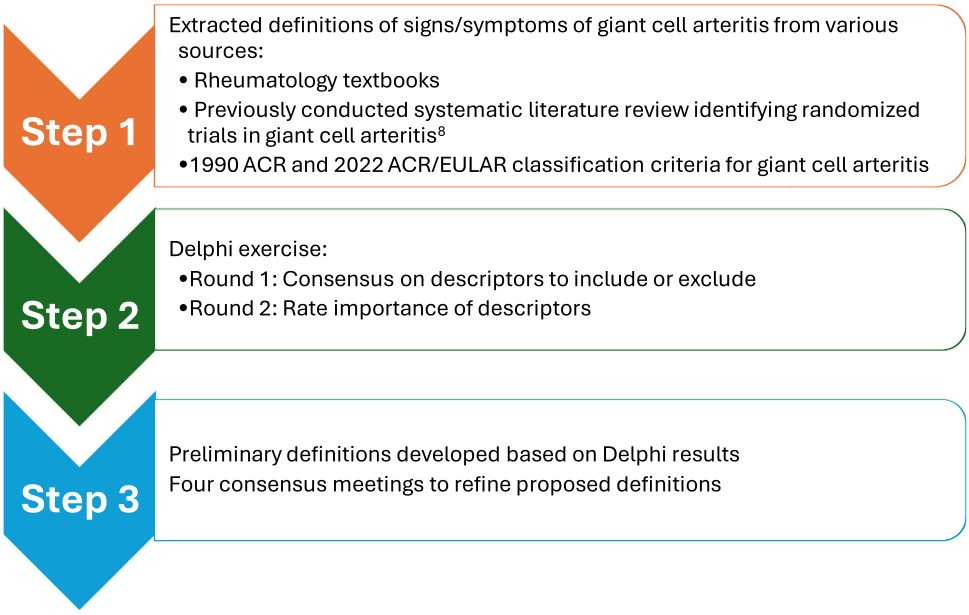

Figure. Flow chart of the study process. The steps taken to develop the glossary of signs and symptoms of giant cell arteritis. ACR, American College of Rheumatology; EULAR, European Alliance of Associations for Rheumatology.

During the meetings, task force members discussed the Delphi results and the proposed definitions. Consensus among task force members was achieved through an online live voting process. Agreement was reached if at least 70% of participants concurred on a definition. If consensus was not achieved, the definition under consideration was rediscussed. In the second round, consensus was accepted if >67% of the members voted in favour of the revised definition, and in the third round, >50% was accepted.

Finally, each task force member anonymously indicated the level of agreement (LoA) via an online survey (LoA, 0–10 numeric rating scale with 0 = do not agree and 10 = fully agree). The mean and SD of the LoA, as well as the percentage of task force members with an agreement ≥8, are presented.

RESULTS

A total of 29 out of 32 (92%) and 27 out of 32 (84%) task force members participated in rounds 1 and 2 of the Delphi exercise, respectively. Attendance by members at online meetings was >75% (meeting 1, 81%; meeting 2, 87%; meeting 3, 87%; meeting 4, 78%).

Overarching principles and definitions of signs/symptoms of GCA

The task force formulated 2 overarching principles forming a framework for all the definitions of signs and symptoms of GCA (Table 1).

Twenty-nine items were reviewed. Four were not defined (polymyalgia rheumatica [13], stroke [14], transient ischaemic attack [15], and myocardial infarction [16]) because they are clinical diagnoses with established definitions in the literature. Similarly, 2 other signs or symptoms (digital ulcers [17] and fatigue [18]) were not defined due to their accepted definitions in the literature. During online meetings, out of the 23 signs and symptoms left to define, 12 were considered characteristic for GCA, whereas 11 were regarded as self-explanatory or not highly specific for GCA. Three online meetings were dedicated to defining those 12 signs and symptoms of GCA, and 1 online meeting was used for the remaining 11 signs and symptoms.

The 12 signs and symptoms of GCA with their definitions are listed in Table 1 (including LoA) and discussed below. All statements obtained a high LoA, ranging from 8.7 to 9.9. The remaining 11 definitions of the symptoms considered self-explanatory or not highly specific for GCA are reported in Table 2 (including LoA), whereas the respective discussion points are included in Supplementary Table S2.

Overarching principles

Overarching principle 1: All the definitions from this glossary refer to signs or symptoms not better explained by other clinical conditions (that is, they are primarily attributed to GCA)

The first principle emphasises that the definitions provided in the glossary are specifically tailored to recognise signs or symptoms of GCA. This principle clarifies that these definitions are most relevant when other clinical conditions cannot better explain the symptoms, ensuring that the focus remains on identifying GCA-related signs and symptoms.

All signs or symptoms described generally represent a new occurrence (ie, not chronic). Additionally, signs and symptoms that self-resolve or resolve with conventional treatments (eg, headache responding to acetaminophen) are generally not attributable to GCA.

Table 1
Overarching principles and definitions of 12 signs and symptoms of giant cell arteritis

Overarching principles		**LoA (0–10) Mean (SD)**	**% with LoA ≥8**
1.	All the definitions from this glossary refer to signs or symptoms not better explained by other clinical conditions (that is, they are primarily attributed to GCA).	9.8 (0.5)	100
2.	The glossary should not serve to prevent patients who do not completely fit these definitions from receiving the necessary diagnostic procedures or therapies.	9.9 (0.4)	100
Signs or symptoms of GCA	**Definitions**		
Headache	New-onset pain localised to the head, not typical of headaches the patient previously experienced. The pain is usually persistent, continuous, and not easily alleviated by analgesics.	9.4 (0.8)	96.4
Temporal artery abnormalities	Any of the following features of a temporal artery: thick, firm, tender, or with a diminished or absent pulse.	9.6 (0.6)	100
Scalp tenderness	Pain/discomfort on touching the scalp, occurring on one or both sides, often elicited by brushing or combing hair.	9.7 (0.5)	100
Scalp necrosis	Ischaemic damage to the scalp marked by altered colour and compromised integrity of the skin.	9.5 (0.8)	96.4
Jaw claudication	Pain, fatigue, or discomfort in jaw muscles occurring when chewing and resolving shortly after chewing stops.	9.6 (0.7)	100
Tongue claudication	Pain, fatigue, or discomfort in the tongue when chewing or talking that resolves after chewing or talking stops.	9.5 (0.8)	100
Tongue necrosis	Ischaemic damage to the tongue marked by altered colour and compromised integrity of the mucosa.	9.3 (1.0)	92.9
Amaurosis fugax	Transient loss of vision in one or both eyes, without associated ocular pain, that is usually sudden and resolves within minutes or rarely hours.	9.3 (1.09)	96.4
Permanent loss of vision	Sudden and irreversible, partial, or complete, loss of sight in one or both eyes.	9.7 (0.6)	100
Fever	Temperature ≥38°C (100.4°F).	8.7 (1.6)	78.6
Limb claudication	Pain, fatigue, or discomfort in limb muscles that occurs with use and is relieved by rest.	9.17 (1.5)	89.3
Blood pressure inequality	A difference of ≥20 mm Hg in systolic blood pressure between contralateral limbs.	9.4 (0.9)	92.9

GCA, giant cell arteritis; LoA, level of agreement.
Numbers in the columns 'LoA' indicate the mean and SD (in parenthesis) of the LoA (assessed on a scale from 0 = no agreement to 10 = full agreement), and the proportion of task force members with a score of at least 8/10.

Table 2
Definitions of 11 additional signs and symptoms of giant cell arteritis

Additional signs or symptoms of GCA	Definitions	LoA (0–10) Mean (SD)	% with LoA ≥8
Abdominal angina	Recurrent pain or discomfort in the abdomen, usually occurring or worsening after eating, considered due to vascular insufficiency.	9.3 (1.1)	89.3
Anorexia	Diminished desire to eat.	9.2 (1.4)	89.3
Blurry vision	A visual disturbance in which objects appear unclear, making it difficult to see things sharply. *Typically sudden in onset.*	9.3 (1.3)	92.9
Carotidynia	Pain or tenderness over one or both carotid arteries.	9.8 (0.5)	100
Diplopia	Transient or persistent visual disturbance in which an object is seen partially or fully in duplicate.	9.6 (0.7)	96.4
Dry cough	A type of cough not accompanied by expectorated phlegm, mucus or blood.	9.6 (0.8)	96.4
Hearing loss	Partial or complete inability to hear sounds in one or both ears. *Typically rapidly progressive.*	9.1 (1.2)	92.9
Odynophagia	Pain or discomfort with swallowing.	9.7 (0.7)	100
Peripheral arthralgia	Pain or discomfort in the joints of the extremities.	9.3 (1.4)	92.9
Pulse abnormalities	Pulse that is difficult to detect or feels faint when palpating arteries in the extremities.	9.0 (1.2)	89.3
Weight loss	Reduction of body weight of at least 5%.	8.4 (2.4)	78.6

GCA, giant cell arteritis; LoA, level of agreement.
Numbers in the columns 'LoA' indicate the mean and SD (in parenthesis) of the LoA (assessed on a scale from 0 = no agreement to 10 = full agreement), and the proportion of task force members with a score of at least 8/10.

Overarching principle 2: The glossary should not serve to prevent patients who do not completely fit these definitions from receiving necessary diagnostic procedures or therapies

The second principle ensures that the glossary definitions do not hinder access to appropriate diagnostic procedures and treatment for patients whose symptoms may not fully align with the defined signs or symptoms. This principle acknowledges the variability in how diseases present and emphasises the importance of clinical flexibility and discretion. It underlines that the glossary serves as a guide, mainly for research, advocating for comprehensive patient care regardless of predefined signs or symptoms.

Definitions of signs and symptoms of GCA

Headache

Definition: New-onset pain localised to the head, not typical of headaches the patient previously experienced. The pain is usually persistent, continuous, and not easily alleviated by analgesics.

The concepts of new-onset (previously not experienced by the patient) and atypical headache were commonly noted for patients included in clinical trials [19–24] as well as in the ACR 1990 classification criteria for GCA [12]. However, aside from a few research papers [21,23], the medical literature offered limited additional information to further characterise a GCA-related headache. These 2 concepts were thus included in the initial potential headache definition and other descriptors extracted from the literature and textbooks were added as options for the task force members to choose from (Supplementary Fig S1).

Task force members agreed that the definition should aim to characterise the type of headache that makes a health care provider concerned about GCA rather than defining any type of headache. The headache definition had to strike a balance between avoiding oversimplification akin to other headache definitions and not being overly restrictive, which can limit inclusion of patients in clinical trials; it should thus provide some flexibility.

In contrast to the International Headache Society definition of GCA-related headache [25], which emphasises the inclusion of other symptoms of GCA (eg, scalp tenderness), it was decided that such an association should not be incorporated in the definition. The task force was reluctant to combine different features of GCA into a single definition, maintaining a distinct concept of GCA-related headache specifically for research purposes and independent of the presence of other symptoms of GCA.

The appropriateness of including a specification regarding headache characteristics such as inflammation (eg, heightened intensity during the night or in the early morning) was also discussed. However, to maintain broad inclusivity, it was decided to omit such specifications due to the heterogeneous nature of headaches across patients and disease courses.

There was a debate about whether to include the lack of response to treatment (ie, no or little improvement of headache by analgesics) as there might be a subgroup of patients who do not, or cannot, take analgesics. The addition of the adverbs 'usually' and 'not easily' in the definition was intended to accommodate such exceptions, whereas the definition also aimed to include descriptors typically associated with a GCA-related headache, such as 'persistent' and 'continuous'. Although new onset is mentioned, the task force agreed that the definition can also be extended to a patient who is relapsing if the character of headache is referencing to the initial presentation of GCA.

Temporal artery abnormalities

Definition: Any of the following features of a temporal artery: thick, firm, tender, or with a diminished or absent pulse.

Distinct features of temporal artery abnormalities that were agreed upon include thickness, firmness, tenderness and a diminished or absent pulse. Tenderness is pain elicited by touching the temporal artery. When discussing the difference between thickness and firmness, thickness was felt to incorporate the concept of being dense and not completely compressible, while firmness would be synonymous to hard or cord-like. 'Any' was intentionally added to have the flexibility of only having 1 abnormality, such as diminished pulse, and therefore increase sensitivity. However, the more abnormalities that are present, the higher the likelihood of being related to GCA. Particular care was taken in the wording of pulse palpation: 'absent pulse' was chosen to convey a lack of prior knowledge regarding the temporal artery's condition in contrast to 'loss of pulse', which implies previous awareness. The interpretation of the temporal artery pulse should be considered in the absence of a prior temporal artery biopsy, as the latter may alter the pulse characteristics. When assessing the temporal artery for signs of GCA, the

examiner should have adequate experience with the anatomy of the temporal artery and with GCA to ensure that the appropriate amount of pressure is applied for evaluating tenderness, firmness, and pulse characteristics. The task force did not include bruits as a possible abnormality of the temporal artery because auscultation of this artery is rarely conducted in clinical practice and has not been mentioned as a possible descriptor in the literature (in contrast to bruits of extracranial arteries to indicate vessel stenosis).

Scalp tenderness

Definition: Pain/discomfort on touching the scalp, occurring on one or both sides, often elicited by brushing or combing hair.

The term 'discomfort' was introduced as an option to encompass nuances related to tenderness when touching the scalp, ranging from mild discomfort to pain. Hyperesthesia was also discussed but finally not added, as the task force believed that pain/discomfort touching the scalp catches the essence of the definition. The definition included the adjective 'often' to allow room for exceptions. Similar to the discussion on temporal artery abnormalities, interpretation should be considered in the absence of a prior temporal artery biopsy.

Scalp necrosis

Definition: Ischaemic damage to the scalp marked by altered colour and compromised integrity of the skin.

There was initial discussion on whether it should be defined given it is such a rare occurrence but on the other hand, it is a very specific feature of GCA and was thus included [26]. Ischaemia means diminished or absent blood flow. Gangrene, a possible consequence of ischaemia was initially included in the definition of scalp necrosis but ultimately omitted, given its definition is restricted to the end-stage result of an ischaemic process. The term 'ischaemic damage' provides greater flexibility in the definition given that it encompasses altered colour and integrity of the skin, as well as gangrene. The task force also emphasised that a change of colour alone is not enough for this process of ischaemic damage to happen. Usually, the subcutaneous tissue also undergoes damage. It would be exceptional that necrosis occurred without also damaging the skin.

Jaw claudication

Definition: Pain, fatigue, or discomfort in jaw muscles occurring when chewing and resolving shortly after chewing stops.

The challenge was formulating a definition that effectively differentiated jaw claudication from other pathologies such as temporomandibular joint disorders. The task force agreed that the essence of the definition is that jaw claudication happens with prolonged (usually minutes) chewing and resolves once chewing stops. It does not typically occur with the first bites or with the initiation of chewing. The task force decided against including the word 'prolonged' into the definition given that its interpretation could be heterogenous with no specific cut-off in the literature. Discussing the meaning of 'prolonged' as part of the definition would introduce substantial complexity. It is important to note that GCA patients can express several jaw-related symptoms, but jaw claudication was chosen to be defined given its high diagnostic specificity [27]. Jaw muscle was added rather than simply jaw to emphasise that this phenomenon happens in the muscles of mastication and not elsewhere. Fatigue and discomfort were added to the definition because jaw claudication is not always perceived as pain by patients.

Tongue claudication

Definition: Pain, fatigue, or discomfort in the tongue when chewing or talking, that resolves after chewing or talking stops.

The task force aimed for this definition to align with the same conceptual framework as that of jaw claudication. Therefore, the essence of the meaning of claudication was kept, where the tongue pain, fatigue, or discomfort happens with activity (chewing or talking) and resolves once the activity stops.

Tongue necrosis

Definition: Ischaemic damage to the tongue marked by altered colour and compromised integrity of the mucosa.

The task force wanted this definition to mirror the same underlying principles as that of scalp necrosis. The fundamental concept of necrosis was maintained by utilising the phrase 'ischaemic damage' characterised by changes in colour and compromised mucosal integrity.

Amaurosis fugax

Definition: Transient loss of vision in one or both eyes, without associated ocular pain, that is usually sudden and resolves within minutes or rarely hours.

In defining amaurosis fugax, the task force advocated for inclusivity by specifying that this GCA feature can involve one or both eyes. Although predominantly unilateral, the less frequent bilateral presentation was retained for the benefit of a larger audience, aiming to be as inclusive as possible when recruiting patients for trials. The central debate revolved around the necessity of explicitly specifying the painlessness of vision loss itself. Cranial GCA is typically characterised by a headache, amaurosis fugax, and painless loss of vision in general (ie, patients do not experience any pain in the eye) [28]. Moreover, the task force wanted to highlight another crucial characteristic of this symptom, which is its reversibility in a short time frame in most cases [28]. However, for the sake of inclusiveness, the rare circumstance where amaurosis resolves within hours is also reported in the definition.

Permanent vision loss

Definition: Sudden and irreversible, partial or complete, loss of sight in one or both eyes.

The task force advocated for the explicit inclusion of the sudden onset and irreversibility of this feature. Vision loss in GCA is an ischaemic complication occurring in 15% to 35% of patients, almost exclusively before the start of treatment with glucocorticoids, and it may be preceded by amaurosis fugax but may also occur as the first manifestation of disease [28–30]. Visual loss encompasses a wide spectrum of visual impairment. Although some individuals may notice specific areas of visual defects, such as blind spots or scotomas, affecting their ability to perceive objects in certain areas of their visual field, others may suffer more severe impairment or blindness, importantly impacting their daily activities and quality of life [31]. Most task force members argued against the necessity of specifying the ischaemic cause in the definition, contending that the assumption of relevance to GCA covers all potential causes, as specified in the overarching principles.

Fever

Definition: Temperature of ≥38°C (100.4°F).

There was extensive discussion about whether this term warranted definition, as a few task force members deemed it self-explanatory. Although fever is defined by the Centers for Disease Control and Prevention (CDC) as measured temperature of

≥38°C [32], some experts expressed concerns that a proportion of patients with GCA may have a temperature that is only slightly higher than their baseline body temperature yet abnormal for that patient and that these low-grade temperatures might differ between individuals.

To capture the concept of low-grade fever, proposals discussed were 2 above-normal measurements or an elevation of at least 0.5°C above the patient's usual baseline temperature. The debate extended to whether patients are even aware of their normal temperature and whether they regularly measure it. However, no agreement on these aspects was reached. The minimum consensus among the task force was, therefore, to adopt the definition of fever used by the CDC as a temperature of ≥38°C and not to formulate a definition for low-grade fever. Nevertheless, all task force members acknowledged that a patient with active GCA may have a body temperature that is slightly higher than their usual baseline but not exceeding 38°C. Finally, fever in GCA is typically not relapsing and remitting, but generally persistent.

Limb claudication

Definition: Pain, fatigue, or discomfort in limb muscles that occurs with use and is relieved by rest.

Limb claudication occurs in patients with extracranial vessel involvement may affect upper and lower limbs, and symptoms may vary from mild to severe based on the degree of vascular involvement and concomitant factors, such as (pre-existent) atherosclerosis, heart failure, or anaemia. The reported definition emphasises the muscular site of the symptoms and notes that limb claudication is typically triggered by prolonged movements (of at least several seconds to minutes) and rapidly reverses with rest. These characteristics help distinguish it from other non-ischaemic conditions such as osteoarthritis, muscle injury, or tendinopathy.

Blood pressure inequality

Definition: A difference of ≥20 mm Hg in systolic blood pressure between contralateral limbs.

Although the task force voted for a ≥20 mm Hg difference in systolic blood pressure between contralateral limbs, respective cut-offs varied markedly in the literature [33–35]. This choice of the task force was influenced by the recently published classification criteria for Takayasu arteritis, which proposed the same cut-off for blood pressure difference [36]. In GCA, such inequality in blood pressure is usually observed in the upper extremities. The task force also stressed the importance of clarifying that the comparison of blood pressures should be between the 2 upper or the 2 lower limbs, not between 1 upper and 1 lower limb. Finally, accurate measurement of blood pressure is key, and guidelines on proper blood pressure measurements (for example, the American College of Cardiology and American Heart Association guidelines on blood pressure measurement [37]) should be followed.

DISCUSSION

To the authors' knowledge, this glossary reflects the first collaborative effort to establish comprehensive definitions for fundamental symptoms and signs of GCA. The task force agreed on 23 definitions for a more precise application of classification criteria and outcome assessments in clinical trials, including remission, relapse, and response to treatment. Despite these definitions being consensus based, there was substantial agreement among both experts and patient representatives.

No definition alone is assumed to possess the requisite sensitivity and specificity to diagnose a patient with GCA in clinical practice. Instead, these definitions are intended to be incorporated in inclusion criteria for research studies, standardise the application of classification criteria (even though this effort was not part of the classification criteria project), and serve as a basis for the development of the new criteria for response to treatment in GCA. Although not primarily intended for use in clinical practice, these definitions and the detailed accompanying qualifying discussions by the task force members provide useful information about the nuances of clinical features commonly associated with GCA to health care providers who may have less experience with this disease.

Obtaining clarity and homogeneity of definitions has been an exercise performed by several groups to facilitate the development of classification criteria in the field of rheumatology. For example, the third phase of the development of the classification criteria for systemic lupus erythematosus was dedicated to an in-depth examination and refinement of definitions [38]. Similarly, a glossary of definitions was incorporated in several classification criteria (eg, spondyloarthritis) to improve the validity and reliability of the final classification system [3,5]. Previous definitions were mostly based on literature review and consensus by the working group, whereas this GCA glossary included multiple rounds of refinement through iterative processes, encompassing 2 Delphi rounds and 4 consensus meetings of an international task force. In contrast to classification criteria, however, this glossary for GCA concentrated solely on clinical signs and symptoms as definitions of imaging and biopsy findings in GCA have already been described.

One potential limitation of this study is the lack of comprehensive definitions in the literature for certain signs and symptoms of GCA (eg, limb claudication), whereas several definitions are available for other features (eg, headache). Consequently, the absence of initial components could have hindered the definition-building process. Therefore, a few definitions were proposed and discussed by the task force members based on clinical expertise (eg, tongue necrosis). Another limitation of this study is that the generation of definitions was based on a SLR focused on RCTs measuring treatment response and disease activity changes in GCA. Although a dedicated SLR would have been appropriate, the task force believed it would be redundant because all relevant GCA trials had already been identified [19–24,39–43]. Because RCTs include descriptions of GCA signs and symptoms as part of the inclusion criteria, they were considered the most important data source. To enhance completeness for the generation of definitions, multiple resources, including GCA classification criteria, were also incorporated.

In conclusion, an international group of specialists in GCA formulated a glossary of definitions for 23 signs and symptoms occurring in GCA using a consensus process. These definitions are designed for research purposes. Applying these definitions should facilitate uniform characterisation of the features of GCA and harmonise the patient populations enrolled into clinical trials in GCA and outcome assessment.

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Competing interests

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REFERENCES

[1] Pugh D, Karabayas M, Basu N, Cid MC, Goel R, Goodyear CS, et al. Large-vessel vasculitis. Nat Rev Dis Primers 2022;7(1):93. doi: 10.1038/s41572-021-00327-5.
[2] Berti A, Dejaco C. Update on the epidemiology, risk factors, and outcomes of systemic vasculitides. Best Pract Res Clin Rheumatol 2018;32(2):271–94. doi: 10.1016/j.berh.2018.09.001.
[3] Rudwaleit M, van der Heijde D, Landewé R, Listing J, Akkoc N, Brandt J, et al. The development of Assessment of SpondyloArthritis International Society classification criteria for axial spondyloarthritis (part II): validation and final selection. Ann Rheum Dis 2009;68(6):777–83. doi: 10.1136/ard.2009.108233.
[4] Ponte C, Grayson PC, Robson JC, Suppiah R, Gribbons KB, Judge A, et al. 2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis. Arthritis Rheumatol 2022;74(12):1881–9. doi: 10.1002/art.42325.
[5] Aringer M, Costenbader K, Daikh D, Brinks R, Mosca M, Ramsey-Goldman R, et al. 2019 European League Against Rheumatism/American College of Rheumatology classification criteria for systemic lupus erythematosus. Ann Rheum Dis 2019;78(9):1151–9. doi: 10.1136/annrheumdis-2018-214819.
[6] Nepal D, Putman M, Unizony S. Giant cell arteritis and polymyalgia rheumatica: treatment approaches and new targets. Rheum Dis Clin North Am 2023;49(3):505–21. doi: 10.1016/j.rdc.2023.03.005.
[7] Prieto-González S, Espígol-Frigolé G, García-Martínez A, Alba MA, Tavera-Bahillo I, Hernández-Rodríguez J, et al. The expanding role of imaging in systemic vasculitis. Rheum Dis Clin North Am 2016;42(4):733–51. doi: 10.1016/j.rdc.2016.07.009.
[8] Sanchez-Alvarez C, Bond M, Soowamber M, Camellino D, Anderson M, Langford CA, et al. Measuring treatment outcomes and change in disease activity in giant cell arteritis: a systematic literature review informing the development of the EULAR-ACR response criteria on behalf of the EULAR-ACR response criteria in giant cell arteritis task force. RMD Open 2023;9(2):e003233. doi: 10.1136/rmdopen-2023-003233.
[9] Firestein GS, Gabriel SE, McInnes IB, O'Dell JR. Kelley & Firestein's Textbook of Rheumatology. 10th ed. Philadelphia, PA: Elsevier; 2017.
[10] Hochberg MC, Silman AJ, Smolen JS, Weinblatt ME, Weisman MH, Gravallese E, editors. Rheumatology. 7th ed. Philadelphia, PA: Elsevier, Inc.; 2019.
[11] Loscalzo J, Fauci A, Kasper D, Hauser S, Longo D, Jameson J. Harrison's Principles of Internal Medicine. 21st ed. New York: McGraw Hill; 2022.
[12] Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 1990;33(8):1122–8. doi: 10.1002/art.1780330810.
[13] Dasgupta B, Cimmino MA, Maradit-Kremers H, Schmidt WA, Schirmer M, Salvarani C, et al. 2012 provisional classification criteria for polymyalgia rheumatica: a European League Against Rheumatism/American College of Rheumatology Collaborative Initiative. Ann Rheum Dis 2012;71(4):484–92. doi: 10.1136/annrheumdis-2011-200329.
[14] Sacco RL, Kasner SE, Broderick JP, Caplan LR, Connors JJ, Culebras A, et al. An updated definition of stroke for the 21st century: a statement for healthcare professionals from the American Heart Association/American Stroke Association. Stroke. 2013;44(7):2064–89. doi: 10.1161/STR.0b013e318296aeca.
[15] Easton JD, Saver JL, Albers GW, Alberts MJ, Chaturvedi S, Feldmann E, et al. Definition and evaluation of transient ischemic attack: a scientific statement for healthcare professionals from the American Heart Association/American Stroke Association Stroke Council; Council on Cardiovascular Surgery and Anesthesia; Council on Cardiovascular Radiology and Intervention; Council on Cardiovascular Nursing; and the Interdisciplinary Council on Peripheral Vascular Disease. The American Academy of Neurology affirms the value of this statement as an educational tool for neurologists. Stroke. 2009;40(6):2276–93. doi: 10.1161/STROKEAHA.108.192218.
[16] Thygesen K, Alpert JS, Jaffe AS, Chaitman BR, Bax JJ, Morrow DA, et al. Fourth universal definition of myocardial infarction (2018). Circulation 2018;138(20):e618–51. doi: 10.1161/CIR.0000000000000617.
[17] Hughes M, Tracey A, Bhushan M, Chakravarty K, Denton CP, Dubey S, et al. Reliability of digital ulcer definitions as proposed by the UK scleroderma study group: a challenge for clinical trial design. J Scleroderma Relat Disord 2018;3(2):170–4. doi: 10.1177/2397198318764796.
[18] Maxwell LJ, Jones C, Bingham CO, Boers M, Boonen A, Choy E, et al. Defining domains: developing consensus-based definitions for foundational domains in OMERACT core outcome sets. Semin Arthritis Rheum 2024;66:152423. doi: 10.1016/j.semarthrit.2024.152423.
[19] Hoffman GS, Cid MC, Hellmann DB, Guillevin L, Stone JH, Schousboe J, et al. A multicenter, randomized, double-blind, placebo-controlled trial of adjuvant methotrexate treatment for giant cell arteritis. Arthritis Rheum 2002;46(5):1309–18. doi: 10.1002/art.10262.
[20] Villiger PM, Adler S, Kuchen S, Wermelinger F, Dan D, Fiege V, et al. Tocilizumab for induction and maintenance of remission in giant cell arteritis: a phase 2, randomised, double-blind, placebo-controlled trial. Lancet 2016;387(10031):1921–7. doi: 10.1016/S0140-6736(16)00560-2.

[21] Stone JH, Tuckwell K, Dimonaco S, Klearman M, Aringer M, Blockmans D, et al. Trial of tocilizumab in giant-cell arteritis. N Engl J Med 2017;377 (4):317–28. doi: 10.1056/NEJMoa1613849.
[22] Schmidt WA, Dasgupta B, Luqmani R, Unizony SH, Blockmans D, Lai Z, et al. A multicentre, randomised, double-blind, placebo-controlled, parallel-group study to evaluate the efficacy and safety of sirukumab in the treatment of giant cell arteritis. Rheumatol Ther 2020;7(4):793–810. doi: 10.1007/s40744-020-00227-2.
[23] Langford CA, Cuthbertson D, Ytterberg SR, Khalidi N, Monach PA, Carette S, et al. A randomized, double-blind trial of abatacept (CTLA-4Ig) for the treatment of giant cell arteritis. Arthritis Rheumatol 2017;69(4):837–45. doi: 10.1002/art.40044.
[24] Cid MC, Unizony SH, Blockmans D, Brouwer E, Dagna L, Dasgupta B, et al. Efficacy and safety of mavrilimumab in giant cell arteritis: a phase 2, randomised, double-blind, placebo-controlled trial. Ann Rheum Dis 2022;81 (5):653–61. doi: 10.1136/annrheumdis-2021-221865.
[25] Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia 2018;38(1):1–211. doi: 10.1177/0333102417738202.
[26] Chehem Daoud Chehem F, de Mornac D, Feuillet F, Liozon E, Samson M, Bonnotte B, et al. Giant cell arteritis associated with scalp, tongue or lip necrosis: a French multicenter case control study. Semin Arthritis Rheum 2024; 64:152348. doi: 10.1016/j.semarthrit.2023.152348.
[27] Lim J, Dures E, Bailey LF, Almeida C, Ruediger C, Hill CL, et al. Jaw claudication and jaw stiffness in giant cell arteritis: secondary analysis of a qualitative research dataset. Rheumatol Adv Pract 2024;8(1) rkad082. doi: 10.1093/rap/rkad082.
[28] Dejaco C, Duftner C, Buttgereit F, Matteson EL, Dasgupta B. The spectrum of giant cell arteritis and polymyalgia rheumatica: revisiting the concept of the disease. Rheumatology (Oxford) 2017;56(4):506–15. doi: 10.1093/rheumatology/kew273.
[29] Patil P, Williams M, Maw WW, Achilleos K, Elsideeg S, Dejaco C, et al. Fast track pathway reduces sight loss in giant cell arteritis: results of a longitudinal observational cohort study. Clin Exp Rheumatol 2015;33(2 Suppl 89) S-103-6.
[30] Diamantopoulos AP, Haugeberg G, Lindland A, Myklebust G. The fast-track ultrasound clinic for early diagnosis of giant cell arteritis significantly reduces permanent visual impairment: towards a more effective strategy to improve clinical outcome in giant cell arteritis? Rheumatology (Oxford) 2016;55(1):66–70. doi: 10.1093/rheumatology/kev289.
[31] Vodopivec I, Rizzo 3rd JF. Ophthalmic manifestations of giant cell arteritis. Rheumatology (Oxford, England) 2018;57(suppl_2):ii63–72. doi: 10.1093/rheumatology/kex428.
[32] Centers for Disease Control and Prevention, National Center for Emerging and Zoonotic Infectious Diseases (NCEZID), Division of Global Migration Health (DGMH). Fever. https://www.cdc.gov/. Accessed January 28, 2024.
[33] Chen Y, Dong H, Li HW, Zou YB, Jiang XJ. Characteristics of four-limb blood pressure and brachial-ankle pulse wave velocity in Chinese patients with Takayasu arteritis. Blood Press 2022;31(1):146–54. doi: 10.1080/08037051.2022.2091513.
[34] Singh S, Sethi A, Singh M, Khosla K, Grewal N, Khosla S. Simultaneously measured inter-arm and inter-leg systolic blood pressure differences and cardiovascular risk stratification: a systemic review and meta-analysis. J Am Soc Hypertens 2015;9(8) 640-50.e12. doi: 10.1016/j.jash.2015.05.013.
[35] Clark CE, Campbell JL, Powell RJ. The interarm blood pressure difference as predictor of cardiovascular events in patients with hypertension in primary care: cohort study. J Hum Hypertens 2007;21(8):633–8. doi: 10.1038/sj.jhh.1002209.
[36] Grayson PC, Ponte C, Suppiah R, Robson JC, Gribbons KB, Judge A, et al. 2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis. Ann Rheum Dis 2022;81(12):1654–60. doi: 10.1136/ard-2022-223482.
[37] Whelton PK, Carey RM, Aronow WS, Casey Jr DE, Collins KJ, Dennison Himmelfarb C, et al. 2017 ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA guideline for the prevention, detection, evaluation, and management of high blood pressure in adults: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. J Am Coll Cardiol 2018;71(19):e127–248. doi: 10.1016/j.jacc.2017.11.006.
[38] Tedeschi SK, Johnson SR, Boumpas D, Daikh D, Dörner T, Jayne D, et al. Developing and refining new candidate criteria for systemic lupus erythematosus classification: an international collaboration. Arthritis Care Res (Hoboken) 2018;70(4):571–81. doi: 10.1002/acr.23317.
[39] Seror R, Baron G, Hachulla E, Debandt M, Larroche C, Puéchal X, et al. Adalimumab for steroid sparing in patients with giant-cell arteritis: results of a multicentre randomised controlled trial. Ann Rheum Dis 2014;73(12):2074–81. doi: 10.1136/annrheumdis-2013-203586.
[40] Hoffman GS, Cid MC, Rendt-Zagar KE, Merkel PA, Weyand CM, Stone JH, et al. Infliximab for maintenance of glucocorticosteroid-induced remission of giant cell arteritis: a randomized trial. Ann Intern Med 2007;146(9):621–30. doi: 10.7326/0003-4819-146-9-200705010-00004.
[41] Mazlumzadeh M, Hunder GG, Easley KA, Calamia KT, Matteson EL, Griffing WL, et al. Treatment of giant cell arteritis using induction therapy with high-dose glucocorticoids: a double-blind, placebo-controlled, randomized prospective clinical trial. Arthritis Rheum 2006;54(10):3310–8. doi: 10.1002/art.22163.
[42] Spiera RF, Mitnick HJ, Kupersmith M, Richmond M, Spiera H, Peterson MG, et al. A prospective, double-blind, randomized, placebo controlled trial of methotrexate in the treatment of giant cell arteritis (GCA). Clin Exp Rheumatol 2001;19(5):495–501.
[43] Jover JA, Hernández-García C, Morado IC, Vargas E, Bañares A, Fernández-Gutiérrez B. Combined treatment of giant-cell arteritis with methotrexate and prednisone. A randomized, double-blind, placebo-controlled trial. Ann Intern Med 2001;134(2):106–14. doi: 10.7326/0003-4819-134-2-200101160-00010.