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Standardization of Uveitis Nomenclature for Reporting Clinical Data. Results of the First International Workshop

THE STANDARDIZATION OF UVEITIS NOMENCLATURE (SUN) WORKING GROUP

Abstract

● **PURPOSE:** To begin a process of standardizing the methods for reporting clinical data in the field of uveitis.

● **DESIGN:** Consensus workshop.

● **METHODS:** Members of an international working group were surveyed about diagnostic terminology, inflammation grading schema, and outcome measures, and the results used to develop a series of proposals to better standardize the use of these entities. Small groups employed nominal group techniques to achieve consensus on several of these issues.

● **RESULTS:** The group affirmed that an anatomic classification of uveitis should be used as a framework for subsequent work on diagnostic criteria for specific uveitic syndromes and that the classification of uveitis entities should be on the basis of the location of the inflammation and not on the presence of structural complications. Issues regarding the use of the terms “intermediate uveitis,” “pars planitis,” “panuveitis,” and descriptors of the onset and course of the uveitis were addressed. The following were adopted: standardized grading schema for anterior chamber cells, anterior chamber flare, and for vitreous haze; standardized methods of recording structural complications of uveitis; standardized definitions of outcomes, including “inactive” inflammation, “improvement” and “worsening” of the inflammation, and “corticosteroid sparing,” and standardized guidelines for reporting visual acuity outcomes.

● **CONCLUSIONS:** A process of standardizing the approach to reporting clinical data in uveitis research has begun, and several terms have been standardized.

The field of uveitis deals with multiple disease entities, some of which are caused directly by infectious agents and others of which appear to be immune-mediated. Many uveitic entities are associated with systemic immune-mediated diseases, such as sarcoidosis, the HLA-B27-associated spondyloarthropathies, and Behçet’s disease, whereas others are limited to the eye. Although attempts have been made to standardize some aspects of uveitis,[1] in general there is limited standardization of classification criteria, inflammation grading schema, and outcomes.[2] Standardization would enhance greatly the comparability of clinical research from different centers, permit meta-analyses, and assist in the development of a more complete and meaningful picture of the clinical course of these diseases and their response to treatment.

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A listing of members of The Standardization of Uveitis Nomenclature (SUN) Working Group appears in the Appendix.

The American College of Rheumatology (ACR) has developed classification criteria for many of the rheumatic diseases such as rheumatoid arthritis and systemic lupus erythematosus.[3,4] These criteria have been developed through a standard process and validated against large databases, in an effort to maximize sensitivity and specificity. In the field of uveitis, provisional criteria have been developed for a limited number of disorders (acute retinal necrosis, progressive outer retinal necrosis, Vogt-Koyanagi-Harada disease, and tubulointerstitial nephritis with uveitis),[5–8] and they still await validation. Additionally, there are criteria for the systemic portion of three diseases in which uveitis is an important feature (ankylosing spondylitis, juvenile idiopathic arthritis, and Behçet's disease),[9–11] but not for the uveitis in these diseases. Like the ACR classifications of arthritis and vasculitis, which are on the basis of the anatomic pattern of the disease, the most widely used classification of uveitis is the one devised by the International Uveitis Study Group (IUSG),[1] and is based on the anatomic location of the inflammation. Nevertheless, there are ambiguities in its use, and it does not provide criteria for the diagnosis of specific uveitic entities. Although the establishment of criteria for specific uveitic entities is a major undertaking, resolution of some of the ambiguities can be addressed more easily.

Grading schema for intraocular inflammation typically uses an ordinal scale ranging from 0 to 4+. However, there are at least four systems for anterior chamber cells,[12–15] three for anterior chamber flare,[12–15] two for vitreous cells,[14,15] and three for vitreous haze or debris.[15–17] Although these systems generally are similar, there are differences, and the number of ordinal grades ranges from six to nine. Therefore, data from different groups are difficult to compare, and concepts, such as a two-step increase in the inflammation, are difficult to apply. A standardized set of criteria for grading the four aspects of intraocular inflammation (anterior chamber cells, anterior chamber flare, vitreous cells, and vitreous haze or debris) would enable the data from different groups and different studies to be compared directly. Although disease-specific scoring systems may require more complicated grading schema (for example, for the multifocal choroidopathies), these four grading schema form the building blocks of more complicated systems, and for some types of uveitis (for example, anterior uveitis) they may suffice.

The choice of outcomes for a clinical study depends on the goals of the study, and for many studies, multiple outcomes are appropriate. Nevertheless, for many clinical studies, particularly therapeutic studies, a primary outcome is needed, and for randomized clinical trials, one outcome typically is chosen as the basis for the sample size calculation. In rheumatology, composite scoring systems, such as the ACR scoring systems for improvement in rheumatoid arthritis (ACR 20, 50, and 70), are used.[18] However, in ophthalmology the ability to observe directly the amount of inflammation and measure directly the eye's ability to function (visual acuity and visual field) may allow simpler systems to be used. For long-term studies of visual impairment in uveitis, loss of visual function, such as visual acuity or visual field,[19] may be appropriate outcomes. For short-term studies of the effect of a new treatment on active uveitis, control of the inflammation is an appropriate outcome. For studies of corticosteroid-sparing agents, such as immunosuppressive drugs or biologic agents, in patients on chronic corticosteroid treatment with quiet disease, the ability to taper the prednisone dose below a clinically meaningful threshold[20] while maintaining inactive disease is an appropriate outcome.

Although the outcome of choice will vary with the type of study, standardized terminology and standardized definitions are needed.

To begin the process of addressing these issues, the First International Workshop on Standardization of Uveitis Nomenclature was held on November 8 to 9, 2004 in Baltimore, Maryland, USA. Attendees included individuals invited to ensure a diverse group from leading centers around the world. Additionally, the meeting was announced and opened to all interested parties, although attendance was capped at 50 participants in order for the breakout sessions to be a manageable size. Fifty individuals from 35 centers in 13 countries participated in some part of the process, and 45 individuals from 33 of the 35 centers in these countries attended the meeting. The leaderships of American Uveitis Society (AUS) and of the International Uveitis Study Group (IUSG) endorsed the workshop, the process involved, and its conclusions, and members of both organizations participated in the workshop.

METHODS

Before the workshop, the meeting organizers discussed possible areas to be addressed by the working group. A survey was developed to determine where there were areas of agreement and where there was a diversity of opinions. The survey instrument was pilot-tested on a small group of participants and revised. It then was sent to the members of the working group, and the responses compiled.

The working group then met for the workshop. At the meeting, the first one-half day was devoted to prepared presentations detailing the issues involved, and the results of the survey were presented. The 45 attendees were assigned to one of three groups for the afternoon sessions, each of which addressed one of the three following subjects: (1) terminology; (2) grading inflammation and documenting complications; and (3) outcomes and results reporting. The small group sessions used nominal group techniques to achieve consensus when possible.[21] Items for which consensus could not be reached were tabled for future work. On the second day, the results of the small group sessions were presented to the entire group for review and acceptance (or tabling if there was substantial disagreement from the other groups). Issues related to the definitions of glaucoma and elevated intraocular pressure were tabled at the workshop and were addressed further by Delphi techniques[21] after consultation with glaucoma experts outside the group.

RESULTS

TERMINOLOGY:

There was consensus that an anatomic classification of uveitis should be used and should serve as a framework for subsequent work on diagnostic criteria for specific uveitic diagnoses. The IUSG anatomic classification scheme[1] (Table 1) was endorsed. Furthermore, it was agreed that the classification of the anatomic location of the uveitis should be on the basis of the site(s) of inflammation and not on the presence of structural complications.

Ambiguities in the IUSG system were addressed. There was consensus that the term *intermediate uveitis* should be used for that subset of uveitis where the vitreous is the major site of the inflammation, and that the presence of peripheral vascular sheathing and macular edema should not change the classification. The diagnostic term *pars planitis* should be used only for that subset of intermediate uveitis where there is snowbank or snowball formation occurring in the absence of an associated infection or systemic disease (that is, "idiopathic"). If there is an associated infection (for example, Lyme disease) or systemic disease (for example, sarcoidosis), then the term intermediate uveitis should be used. The term *panuveitis* should be reserved for those situations in which there is no predominant site of inflammation, but inflammation is observed in the anterior chamber, vitreous, and retina and/or choroid (that is, retinitis, choroiditis, or retinal vasculitis). For the definition of panuveitis, structural complications such as macular edema or neovascularization should not be considered in classifying the anatomic location of the uveitis. Inflammation in the anterior chamber and vitreous (that is, more vitritis than in an iridocyclitis and more anterior chamber inflammation than in intermediate uveitis) should be referred to as anterior and intermediate uveitis and not as panuveitis.

The term *retinal vasculitis* was addressed. There was consensus that it is a descriptive term for those situations in which there is evidence of ocular inflammation and retinal vascular changes. The presence of occlusive retinal vasculopathy, in the absence of visible inflammation such as in the antiphospholipid antibody syndrome, should not be considered retinal vasculitis. Achieving consensus on which retinal vascular changes constituted retinal vasculitis was more problematic. Although the group provisionally agreed to consider perivascular sheathing and vascular leakage or occlusion on fluorescein angiogram as evidence of retinal vascular disease for the classification of retinal vasculitis, there was consensus that the definition of retinal vasculitis required more work. For example, it was unresolved as to how to distinguish between retinal vasculitis and the peripheral vascular sheathing sometimes seen in intermediate uveitis.

The terms "acute" and "chronic" have been used inconsistently in the literature and have been used variably to refer to the onset of the uveitis, the duration of an attack of uveitis, or to the course of uveitis. Consensus was obtained that the use of these terms should be reserved for the description of the clinical course of the uveitis, and that other terms should be used to describe the onset of the uveitis and the duration of an attack of uveitis (Table 2). The onset of uveitis should be described either as *sudden* or *insidious*. The duration of an attack of uveitis should be described as either *limited*, if it is 3 months or less in duration or as *persistent*, if it is greater than 3 months in duration. The term *acute* should be used to describe the course of specific uveitic syndromes characterized by sudden onset and limited duration, such as HLA-B27-associated "acute anterior uveitis."[22] The term *recurrent* should be used to describe repeated episodes of uveitis separated by periods of inactivity without treatment, in which these periods of inactivity without treatment are at least 3 months in duration. The term *chronic* should be used to describe persistent uveitis characterized by prompt relapse (in less than 3 months) after discontinuation of therapy.

It was agreed that the appearance of the keratic precipitates potentially conveys useful clinical information and may have diagnostic implications. However, keratic precipitates are

not described in a universally standardized fashion. Although no consensus could be reached either on how to describe keratic precipitates or on the use of the term “granulomatous” as a descriptor for keratic precipitates, there was consensus that a series of standardized photographs should be used by a panel of experts to develop appropriate descriptive terms and then published as a standard reference.

GRADING INFLAMMATION AND DOCUMENTING COMPLICATIONS:

Consensus was achieved regarding standard methods for grading anterior chamber cells (Table 3) and anterior chamber flare (Table 4). Additionally, there was consensus that a set of standardized photographs should be developed to assist in grading anterior chamber flare. Although the level 0.5+ was selected over the term “trace,” the system is an ordinal one, in which the levels represent a nonlinear hierarchy of increasing magnitude, but do not have a numerical relationship to the amount of inflammation. For the grading of anterior chamber cells, the presence or absence of a hypopyon should be recorded separately. Although it was agreed that the presence of vitreous cells was an important clinical feature, no consensus could be reached on a standard grading system for vitreous cells. The National Eye Institute system for grading vitreous haze was adopted with the proviso that the designation “trace” be recorded as 0.5+.[17]

The level of evidence required for reporting structural complications of uveitis depended on the complication being reported. It was agreed that macular edema could be reported as present or absent as determined clinically. However, ancillary testing can provide a greater level of sensitivity and specificity, and macular edema may be confirmed or excluded by either fluorescein angiography or optical coherence tomography. Similarly it was agreed that epiretinal membrane formation could be reported as present or absent as determined clinically and may be confirmed or excluded by either fundus photography or optical coherence tomography. It was expected that retrospective studies might report both levels of evidence, particularly if there was a variable use of ancillary studies, but that for clinical trials and epidemiologic studies in which there is prospective data collection, reporting results on the basis ancillary studies is preferable.

For reporting purposes, it was agreed that subretinal neovascularization should be reported only if confirmed by either fluorescein angiography or fluorescein angiography and indocyanine green angiography. Disk and retinal neovascularization should be reported if confirmed by fundus photography and fluorescein angiography. Although clinical grading of these outcomes may be acceptable in the setting of other retinal diseases, for uveitis, in which these complications are less common, the group decided that reporting was appropriate only if based on the appropriate ancillary testing. Although there was consensus that ancillary tests that document retinal dysfunction, such as perimetry or electroretinography, should be used for those diseases that cause diffuse retinal dysfunction (for example, birdshot chorioretinitis), there was no consensus on which tests are most appropriate for which disease.

There was consensus that the term *glaucoma* should not be considered synonymous with elevated intraocular pressure in a patient with uveitis, but that it should be reserved for those situations where there is either observed glaucomatous disk damage or demonstrated visual

field loss. The term *elevated intraocular pressure* should be used for those situations where there is an intraocular pressure above a defined normal range or when there is an increase in intraocular pressure from baseline during a study with longitudinal data. The threshold for considering a rise in intraocular pressure substantial (for example, as in a rise in intraocular pressure attributable to corticosteroid use) was 10 mm Hg or greater. Although consensus was not achieved on the threshold for considering an intraocular pressure as elevated, the choices were narrowed to two. The first was to report at two levels: above 21 mm Hg (the traditional "upper limit of normal") and above 30 mm Hg (a level above which many practitioners would initiate treatment even without evidence of glaucomatous damage). The second option was to report intraocular pressure above the 24 mm Hg as elevated, as the risk of glaucoma appears to increase substantially as the intraocular pressure increases beyond this level.[23–25] The use of antiglaucoma treatment can be reported, but because of the variability among practitioners in indications for treatment, it should not be used as the only criterion for reporting elevated intraocular pressure.

OUTCOMES AND RESULTS REPORTING:

The activity of anterior chamber inflammation should be on the basis of the cells in the anterior chamber. High-speed optical coherence tomography of the anterior chamber has demonstrated that a rare cell (but less than 1 per field on standard slit-lamp examination) may be present in the anterior chamber of normal individuals.[26] Therefore, it was agreed that for reporting purposes, *inactive* anterior uveitis should be defined as rare cells or less. The presence of one cell in every field is indicative of 0.5+ cells (or in some systems "trace cells") and, for reporting purposes, should not be considered inactive uveitis. As with the inability to reach consensus on a grading system for vitreous cells, no consensus could be reached on a definition of inactive vitritis on the basis of vitreous cells.

Although the goal of treatment of uveitis is to suppress the inflammation completely ("inactive" disease), for the short-term evaluation of new therapies, it may be appropriate to determine whether the inflammation has improved or worsened (Table 5). Given the semiquantitative nature of the grading systems (for example, for anterior chamber cells, vitreous haze), it was agreed that at least a two-step decrease in the level of inflammation for improvement and at least a two-step increase in the level of inflammation for worsening were better criteria than one-step changes. However, because of floor and ceiling effects (that is, 3+ only can increase by one step and 0.5+ only can decrease by one step), the definition of improvement should include a decrease in inflammation from 0.5+ to inactive, and the definition of worsening should include an increase from 3+ to the maximum grade. Hence, *improvement* in the inflammation will be defined as either a two-step decrease in the level of inflammation or a decrease to "inactive," and *worsening* of the inflammation will be defined as either a two-step increase in the level of inflammation or an increase to the maximum grade. The group considered several definitions of the term *remission*, including inactive disease on treatment, inactive disease after discontinuing treatment, and inactive disease for a specified duration after discontinuing treatment. Because chronic uveitis may be a life-long problem, which can be controlled by treatment but relapses promptly after discontinuing treatment, there was consensus that the term *remission* should be reserved for inactive disease for at least 3 months after discontinuing all treatments for eye disease.

Clinical studies of immunosuppressive drugs and biologic agents for severe uveitis can evaluate either the response of active uveitis to the drug being evaluated or the ability of the drug being evaluated to maintain inactive disease in the face of tapering other drugs, such as systemic corticosteroids. Although global scoring systems for a reduction in the total corticosteroid and immunosuppressive drug regimen may be a desirable goal for research studies, in clinical practice a reduction in the prednisone dose for adults to 10 mg per day or less is a primary goal of immunosuppressive drug therapy.[20] Therefore, there was consensus that for reporting purposes in studies of adult patients, reduction in the dose of prednisone to 10 mg per day or less (or its equivalent for other corticosteroids) while maintaining inactive uveitis be considered the primary outcome for successful *corticosteroid sparing*. Although other outcomes also may be reported (for example, discontinuation of prednisone), in studies where corticosteroid sparing is an outcome, reduction in the dose of prednisone to 10 mg per day or less should be reported and should be the primary measure of this outcome.

Data from clinical series should use accepted statistical methods and should not report events or outcomes as the proportion of a population when there is variable follow-up.[27] Instead, the proportion with the outcome at presentation (or study entry) should be reported and the event rate during follow-up should be reported for longitudinal studies. In those series with complete or nearly complete follow-up, the distribution of outcomes at defined time point(s) after presentation (or study entry) can be reported. “Final visual acuity,” defined as the last measured acuity in a series of patients with variable follow-up, should not be reported, because it is a flawed concept that may introduce uncontrolled bias into the study.[27,28] Instead, rates of visual acuity loss or gain either below or above specified thresholds or the rate of acuity change (for example, doubling of the visual angle), should be reported. Alternatively, when there is complete or nearly complete follow-up, the distribution of visual acuities at a specified time after presentation (or study entry), also known as “interval visual acuity results,”[28] may be reported. There was consensus that key visual acuity thresholds that should form the basis for reporting results of uveitis studies include 6/15 or worse (20/50 or worse) and 6/60 or worse (20/200 or worse), and that key acuity changes include a doubling of the visual angle (or for improvement, a halving of the angle). The latter measurement is the basis for the widely used “three lines on an ETDRS visual acuity chart.”[29] In situations where logarithmic visual acuity charts, such as the ETDRS charts, are not available, other acuity measurements (for example, Snellen acuities) should be converted to logMAR format to evaluate doubling of the visual angle. The formula for logMAR is:

$$\text{logMAR} = -\log_{10}(\text{visual acuity fraction})$$

Although logMAR reporting is superior to the number of lines on a Snellen chart approach, there remain problems introduced by the limitations of Snellen Charts, particularly in the poorer ranges of visual acuity (that is, 20/100 or worse), where a line may be represented by one or two letters. Therefore, the use of logarithmic charts, especially in prospective studies, is encouraged.

CONCLUSIONS

The first international workshop on standardization of Uveitis Nomenclature (SUN) has produced consensus on several items, beginning the process of developing international standards for reporting clinical data in the field of uveitis. Standardization should provide greater precision and enhance comparability among reports from different groups. Long term, a set of classification criteria for specific uveitic entities, completion of the process of standardizing the grading of inflammation, and the development of disease-specific outcomes are needed. The SUN Working Group advocates that the standards herein be applied in all studies of uveitis begun after publication of this manuscript.

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APPENDIX

Standardization of uveitis nomenclature working Group.

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TABLE 1.

The SUN* Working Group Anatomic Classification of Uveitis

Type	Primary Site of Inflammation†	Includes
Anterior uveitis	Anterior chamber	Iritis
		Iridocyclitis
		Anterior cyclitis
Intermediate uveitis	Vitreous	Pars planitis
		Posterior cyclitis
		Hyalitis
Posterior uveitis	Retina or choroid	Focal, multifocal, or diffuse choroiditis
		Chorioretinitis
		Retinochoroiditis
		Retinitis
		Neuroretinitis
Panuveitis	Anterior chamber, vitreous, and retina or choroid	

*SUN = Standardization of uveitis nomenclature.

†As determined clinically. Adapted from the International Uveitis Study Group anatomic classification in reference 1.

TABLE 2.

The SUN* Working Group Descriptors of Uveitis

Category	Descriptor	Comment
Onset	Sudden	
	Insidious	
Duration	Limited	≤3 months duration
	Persistent	>3 months duration
Course	Acute	Episode characterized by sudden onset and limited duration
	Recurrent	Repeated episodes separated by periods of inactivity without treatment ≥3 months in duration
	Chronic	Persistent uveitis with relapse in <3 months after discontinuing treatment

*SUN = Standardization of uveitis nomenclature.

TABLE 3.

The SUN* Working Group Grading Scheme for Anterior Chamber Cells

Grade	Cells in Field†
0	<1
0.5+	1–5
1+	6–15
2+	16–25
3+	26–50
4+	>50

*SUN = Standardization of uveitis nomenclature.

†Field size is a 1 mm by 1 mm slit beam.

TABLE 4.

The SUN* Working Group Grading Scheme for Anterior Chamber Flare

Grade	Description
0	None
1+	Faint
2+	Moderate (iris and lens details clear)
3+	Marked (iris and lens details hazy)
4+	Intense (fibrin or plastic aqueous)

Adapted from reference 12.

*SUN = Standardization of uveitis nomenclature.

TABLE 5.

The SUN* Working Group Activity of Uveitis Terminology

Term	Definition
Inactive	Grade 0 cells†
Worsening activity	Two step increase in level of inflammation (e.g. anterior chamber cells, vitreous haze) or increase from grade 3+ to 4 +
Improved activity	Two step decrease in level of inflammation (e.g. anterior chamber cells, vitreous haze) or decrease to grade 0
Remission	Inactive disease for ≥3 months after discontinuing all treatments for eye disease

*SUN = Standardization of uveitis nomenclature.

†Applies to anterior chamber inflammation.