Classification Criteria For Pars Planitis

THE STANDARDIZATION OF UVEITIS NOMENCLATURE (SUN) WORKING GROUP[1,2,3,4,*]

• PURPOSE: To determine classification criteria for pars planitis.
• DESIGN: Machine learning of cases with pars planitis and 4 other intermediate uveitides.
• METHODS: Cases of intermediate uveitides were collected in an informatics-designed preliminary database, and a final database was constructed of cases achieving supermajority agreement on the diagnosis, using formal consensus techniques. Cases were split into a training set and a validation set. Machine learning using multinomial logistic regression was used on the training set to determine a parsimonious set of criteria that minimized the misclassification rate among the intermediate uveitides. The resulting criteria were evaluated on the validation set.
• RESULTS: Five hundred eighty-nine cases of intermediate uveitides, including 226 cases of pars planitis, were evaluated by machine learning. The overall accuracy for intermediate uveitides was 99.8% in the training set and 99.3% in the validation set (95% confidence interval 96.1, 99.9). Key criteria for pars planitis included unilateral or bilateral intermediate uveitis with either 1) snowballs in the vitreous or 2) snowbanks on the pars plana. Key exclusions included: 1) multiple sclerosis, 2) sarcoidosis, and 3) syphilis. The misclassification rates for pars planitis were 0% in the training set and 1.7% in the validation set, respectively.
• CONCLUSIONS: The criteria for pars planitis had a low misclassification rate and appeared to perform sufficiently well for use in clinical and translational research. (Am J Ophthalmol 2021;228: 1–7. © 2021 Elsevier Inc. All rights reserved.)

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Intermediate uveitis refers to a class of uveitic diseases characterized by inflammation predominantly in the vitreous and an absence of retinitis and choroiditis. Intermediate uveitides may be caused by infections such as Lyme disease or syphilis; or they may be associated with systemic diseases, particularly sarcoidosis and multiple sclerosis (MS); or they may occur as an isolated, presumably immunity-mediated ocular disorder of unknown origin. Pars planitis represents a subset of intermediate uveitis characterized by fibroinflammatory material overlying the pars plana and peripheral retina ("snowbanks").[1,2] Initially noted by Schepens[3] in 1950 and termed "peripheral uveitis," the features of what is now termed pars planitis were described nearly simultaneously in 1960 by Welch and associates[4] and Brockhurst and associates.[5] Also termed cyclitis by Hogan and Kimura,[6] the name "pars planitis" was coined by Welch and associates,[4] and pars planitis has remained the term most commonly used for this intermediate uveitic disease. Although snowbanks have been considered the conventional hallmark of pars planitis, a similar uveitic disorder occurs as an intermediate uveitis without snowbanks or "snowballs" (fibroinflammatory debris typically in the inferior vitreous), which now is termed intermediate uveitis, non-pars planitis type,[2] and which also could be considered an "undifferentiated intermediate uveitis." Case series which have included both pars planitis and non-pars planitis types of intermediate uveitis have made

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interpretations of the published studies more difficult.[7] In 2005, the Standardization of Uveitis Nomenclature (SUN) Working Group at a consensus meeting agreed that the term pars planitis should apply to cases of non-infectious intermediate uveitis with vitritis and either inferior vitreous inflammatory condensates ("snowballs") or pars plana "snowbanks," unassociated with a systemic disease and that it should be distinguished from intermediate uveitis, non-pars planitis type.[2] Furthermore, the group recognized that pars planitis may have peripheral retinal vascular sheathing and non-perfusion (more easily seen on wide-field fluorescein angiography) but should not have posterior pole or mid-peripheral occlusive retinal vasculitis.[2]

Given the definitional variations in the disease, its frequency in referral center case series has been reported to vary from 2.4%-15.4% of uveitis cases,[8,9] and its incidence has been estimated at 2.08/100,000 population/year.[10] Structural complications of intermediate uveitides include macular edema, epiretinal membrane formation, and, uncommonly, retinal neovascularization of either the disc or the snowbank. Anterior chamber inflammation typically is mild, and the eye is not acutely inflamed. Typical presenting symptoms are either floaters or blurred vision, most often due to macular edema.[10–12]

The SUN Working Group is an international collaboration, which has developed classification criteria for 25 of the most common uveitides by using a formal approach to development and classification.[2,13–17] Among the intermediate uveitides studied was pars planitis.

METHODS

The SUN Developing Classification Criteria for the Uveitides project proceeded in 4 phases, as previously described: 1) informatics; 2) case collection; 3) case selection; and 4) machine learning.[13–15,17]

• **INFORMATICS:** As previously described, the consensus-based informatics phase permitted development of a standardized vocabulary and development of a standardized, menu-driven hierarchical case collection instrument.[13]

• **CASE COLLECTION AND CASE SELECTION:** Information was entered into the SUN preliminary database by the 76 contributing investigators for each disease as previously described.[15,17] Cases in the preliminary database were reviewed by committees of 9 investigators for selection into the final database, using the formal consensus techniques described in the accompanying article.[15,17] Because the goal was to develop classification criteria,[16] only cases with a supermajority agreement (>75%) that the case was the disease in question were retained in the final database (ie, were "selected").[17]

• **MACHINE LEARNING:** The final database then was randomly separated into a training set (~85% of cases) and a validation set (~15% cases) for each disease, as described in the accompanying article.[17] Machine learning was used in the training set to determine criteria that minimized misclassification. The criteria then were tested in the validation set; for both the training set and the validation set, the misclassification rate was calculated for each disease. The misclassification rate was the proportion of cases classified incorrectly by the machine learning algorithm compared to the consensus diagnosis. For pars planitis, the diseases against which it was evaluated were MS-associated intermediate uveitis; intermediate uveitis, non-pars planitis type (undifferentiated intermediate uveitis); sarcoidosis-associated intermediate uveitis; and syphilitic intermediate uveitis. There were too few cases of Lyme disease-associated uveitis[14] collected in the data base for analysis by machine learning.

• **COMPARISON OF CASES WITH AND WITHOUT SNOWBANKS:** Comparison of the characteristics of cases with and without snowbanks was performed using the χ^2 test for categorical variables or the Fisher exact test when the count of a variable was less than 5. Continuous variables were summarized as medians and compared using the Wilcoxon rank sum test. For characteristics with multiple categorical grades, values above and below the median were compared. *P* values were nominal and 2-sided.

The study adhered to the principles of the Declaration of Helsinki. Institutional Review Boards (IRBs) at each participating center reviewed and approved the study; the study typically was considered either a minimal risk or exempt by the individual IRBs.

RESULTS

A total of 308 cases of pars planitis were collected, and 226 cases (73%) achieved supermajority agreement on the diagnosis during the "selection" phase and were used in the machine learning phase. Those cases of pars planitis were compared to 363 cases of other intermediate uveitides including 112 cases of MS-associated intermediate uveitis; 114 cases of intermediate uveitis, non-pars planitis type; 52 cases of sarcoidosis-associated intermediate uveitis; and 85 cases of syphilitic intermediate uveitis. Details of the machine learning results for these diseases are outlined in the accompanying article.[17] The characteristics at presentation to a SUN Working Group investigator of cases with pars planitis are listed in Table 1. A comparison between cases with and without snowbanks is listed in Table 2. The only significant differences between those with snowbanks and those without snowbanks were that those with snowbanks were younger. The criteria developed after machine learning are listed in Table 3. The overall accuracy for

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TABLE 1. Characteristics of Cases with Pars Planitis

Characteristics	Result
Number of cases	226
Demographics	
Median IQR age [25th, 75th percentile)	22 [11, 36]
Sex (%)	
Men	48
Women	52
Race/ethnicity, %	
White, non-Hispanic	72
Black, non-Hispanic	5
Hispanic	6
Asian, Pacific Islander	3
Other	6
Missing	8
Uveitis history	
Uveitis course, %	
Acute, monophasic	2
Acute, recurrent	2
Chronic	87
Indeterminate	9
Laterality, %	
Unilateral	15
Unilateral, alternating	0
Bilateral	85
Ophthalmic examination	
Keratic precipitates, %	
None	83
Fine	15
Round	2
Stellate	0
Mutton fat	0
Other	0
Anterior chamber cells, %	
Grade 0	44
½+	27
1+	19
2+	9
3+	1
4+	0
Hypopyon, %	0
Anterior chamber flare, %	
Grade 0	75
1+	21
2+	3
3+	1
4+	0
Iris (%)	
Normal	88
Posterior synechiae	12
Sectoral iris atrophy	0
Patchy iris atrophy	0
Diffuse iris atrophy	0
Heterochromia	0
IOP of the involved eyes	
Median IQR, mm Hg [25th, 75th percentile]	14 [12, 17]
Proportion of patients with IOP >24 mm Hg in either eye, %	4
Vitreous cells (%)*	
Grade 0	4
½+	8
1+	35
2+	39
3+	13
4+	1
Vitreous haze (%)*	
Grade 0	31
½+	15
1+	27
2+	23
3+	3
4+	1
Vitreous snowballs†	83
Pars plana snowbanks†	44
Peripheral retinal vascular sheathing or leakage	25
Macular edema	43

IOP = intraocular pressure; IQR = interquartile range.
*All cases had either vitreous cells or haze; only one case had haze without evident cells.
†All cases snowballs or snowbanks; 124 cases had snowballs without snowbanks.

(*continued on next page*)

intermediate uveitides was 99.8% in the training set and 99.3% in the validation set (95% confidence interval [CI]: 96.1-99.2).[17] The misclassification rate for pars planitis in the training set was 0% and 1.7% in the validation set.

DISCUSSION

The classification criteria developed by the SUN Working Group for pars planitis had a low misclassification rate, indicating good discriminatory performance relative to other intermediate uveitides.

The distinctive feature of pars planitis classically has been the presence of inferior snowbanks (Figure 1). Histopathologic examination has demonstrated fibroglial or fibrovascular proliferation with non-granulomatous inflammation composed of mononuclear inflammatory cells; lymphocytic cuffing and mural infiltration of retinal venules; and hyperplastic non-pigmented epithelium of the pars plana.[18,19] Because the SUN definition of pars planitis[2] allowed inclusion of cases with snowballs but not snowbanks, cases with and without snowbanks were

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TABLE 2. Characteristics of Cases with Pars Planitis with and without Snowbanks

Characteristic	Patients with Snowbanks	Patients without Snowbanks	*P* Value
Number cases	104	124	
Demographics			
Median IQR age [25th, 75th percentile]	19 [10, 30]	39 [27, 52]	<.001
Sex %			.68
Men	50	47	
Women	50	53	
Race/ethnicity, %			.25
White, non-Hispanic	70	73	
Black, non-Hispanic	4	6	
Hispanic	6	6	
Asian, Pacific Islander	2	3	
Other	4	5	
Missing	14	7	
Uveitis History			
Uveitis course, %			.54
Acute, monophasic	1	3	
Acute, recurrent	1	3	
Chronic	87	86	
Indeterminate	11	8	
Laterality, %			.92
Unilateral	16	15	
Bilateral	84	85	
Ophthalmic examination			
Keratic precipitates, %			.31
None	85	80	
Fine	14	18	
Round	1	2	
Anterior chamber cells, %*			.17
Grade 0	50	40	
Grade ½+ or greater	50	60	
Anterior chamber flare, %*			.14
Grade 0	80	72	
Grade 1+ or greater	20	28	
Iris, %			.07
Normal	92	84	
Posterior synechiae	8	16	
IOP of the involved eyes			
Median IQR, mm Hg [25th, 75th percentile)	14 [12, 17]	14 [12, 17]	1.00
Vitreous cells, %*			.17
Grades 0–1+	42	51	
Grades 2+ or greater	58	49	
Vitreous haze, %*			.12
Grades 0–½+	52	41	
Grades 1+ or greater	48	59	
Vitreous snowballs†	71	100	
Pars plana snowbanks†	100	0	-
Peripheral retinal vascular sheathing or leakage	23	27	.51
Macular edema	38	48	.15

IOP = interocular pressure; IQR = interquartile range.

*Analyses compare values above and below the median value.

†Presence or absence of snowbanks = defining characteristic of the 2 subsets; cases without snowbanks required to have snowballs to be classified as having pars planitis.

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TABLE 3. Classification Criteria for Pars Planitis

Criteria
1. Evidence of intermediate uveitis
a. Vitreous cells AND/OR vitreous haze
b. If anterior chamber cells are present, anterior chamber inflammation severity less than vitreous severity
c. No evidence of retinitis or choroiditis
d. No retinal vascular occlusion in posterior pole & mid-periphery*
AND
2. Evidence of pars planitis
a. Vitreous snowballs OR
b. Pars plana snowbanks
Exclusions
1. Multiple sclerosis, defined by the McDonald criteria[28]
2. Positive serology test result for syphilis using a treponemal test
3. Evidence of sarcoidosis (either bilateral hilar adenopathy on chest imaging or tissue biopsy demonstrating non-caseating granulomata)
4. Positive serology for Lyme disease, either IgG or IgM (e.g. positive ELISA AND Western blot with requisite number of bands for assay used)

ELISA = enzyme-linked immunosorbent assay; IgG = immunoglobulin G;
*Peripheral retinal non-perfusion on wide-field angiography is compatible with pars planitis diagnosis.

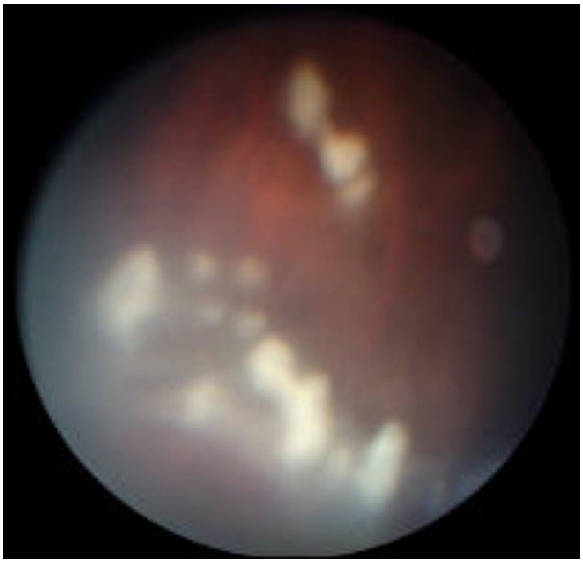

FIGURE 1. Pars plana snowbank in a patient with pars planitis.

compared. The only significant differences detected were the younger age at presentation of those with snowbanks. Whether that difference represents a more exuberant response to the same disease among younger patients or a different pathogenetic mechanism cannot be determined at this time. One study suggested that the course of pars planitis in childhood may be different from that in adults, with a higher rate of sustained, drug-free remissions,[20] but that impression needs to be confirmed. Long-term follow-up studies of patients with and without snowbanks are needed and may help determine if those 2 subsets should continue to be considered within the spectrum of the same disorder or separate ones. However, at this time, the criteria include both subsets in the term "pars planitis"[2]; it would seem prudent that studies of patients with pars planitis report and evaluate the 2 subsets "with and without snowbanks," in order to evaluate any differences.

Ultra-wide-field angiography has demonstrated the presence of peripheral vascular cuffing, leakage, and non-perfusion in patients with pars planitis.[21–23] These findings are distinct from the posterior pole and mid-peripheral occlusive retinal vasculitides, such as those seen in Behçet disease, and pars planitis should be diagnosed separately and not be lumped with the more severe occlusive retinal vasculitides.

Pars planitis has been associated with the HLA type HLA-DR2 and with its split antigen type HLA-DR15 with relative odds in the range of 3-5.[12,24,25] Although there is an association, the positive predictive value[25] of those antigens is poor owing to the high population prevalence of the genes. Furthermore, HLA-DR2 and DR15 are risk factors for MS,[12] rendering them unhelpful for distinguishing between pars planitis and MS-associated uveitis.

Multiple sclerosis has been associated with intermediate uveitis,[11,12] but at this time, it is considered distinct from pars planitis without MS.[2] Nevertheless, the 2 disorders may have overlapping features, including snowballs or snowbanks, or both, in some patients with MS-associated uveitis.[26] Furthermore, patients presenting with pars planitis without MS have been estimated to have a risk of developing MS of ~2%-4%/year,[11,12] so that neuroimaging to exclude MS is likely to have a low yield and is not routinely recommended.[27] Multiple sclerosis should be excluded on clinical grounds, beginning with the absence of relevant neurological lesions or a history of such lesions, and using the McDonald criteria.[28] As such, some cases initially diagnosed as pars planitis may have had their diagnosis changed with follow-up and the development of

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MS. Peripheral vascular changes have been reported as a risk factor for subsequent development of MS,[11] and the prevalence of peripheral vascular sheathing or leakage, or both, was greater in cases with MS-associated uveitis[17,26] but not sufficiently to be of diagnostic utility.[17]

The presence of any of the exclusions in Table 3 suggests an alternate diagnosis, and the diagnosis of pars planitis should not be made in their presence. In prospective studies many of these tests will be performed routinely, and the alternative diagnoses excluded. However, in retrospective studies based on clinical care, not all of these tests may have been performed. Hence the presence of an exclusionary criterion excludes pars planitis, but the absence of such testing does not always exclude the diagnosis of pars planitis if the criteria for the diagnosis are met. Nevertheless, because of the overlapping features of sarcoidosis-associated intermediate uveitis, including snowballs, a reasonable attempt should be made to exclude sarcoidosis, including, at a minimum, chest imaging, for all cases of pars planitis.[29]

The type of uveitis most often seen with Lyme disease is an atypical intermediate or anterior and intermediate uveitis, but disease indistinguishable from pars planitis has been described.[30,31] Lyme uveitis is sufficiently uncommon that the cases collected were too few for analysis. In regions where Lyme disease is not endemic, there appeared to be little value to screening for Lyme disease, as nearly all positive test results would be false positives.[32] Even among patients from Lyme disease-endemic areas undergoing routine testing, the frequency of Lyme disease uveitis has been estimated as no more than 0.35% of uveitis cases, and it has been proposed by some uveitis experts that testing for Lyme disease should be reserved for persons exposed to Lyme disease and those with symptoms suggesting Lyme disease.[33] Nevertheless, in prospective studies from Lyme disease-endemic regions (or in Lyme disease-exposed individuals) testing patients with intermediate uveitis for Lyme disease would appear to be appropriate. The presence of a positive Lyme serology (with appropriate confirmatory testing) excludes the diagnosis of pars planitis.

Classification criteria are used to diagnose individual diseases for research purposes.[16] Classification criteria differ from clinical diagnostic criteria in that, although both seek to minimize misclassification, when a tradeoff is needed, diagnostic criteria typically emphasize sensitivity, whereas classification criteria emphasize specificity,[16] in order to define a homogeneous group of patients for inclusion in research studies and limit the inclusion of patients without the disease in question that may confound the data. The machine learning process used did not explicitly use sensitivity and specificity; instead it minimized the misclassification rate. Because this study focused on developing classification criteria and because the typical agreement between 2 uveitis experts on diagnosis was moderate at best,[15] the selection of cases for the final database ("case selection") included only cases which achieved supermajority agreement on the diagnosis. As such, some cases which clinicians would diagnose as pars planitis will not be so classified by classification criteria. The selection of cases which achieved supermajority agreement on the diagnosis for inclusion in the final data base was used because the study was developing classification criteria.

The criteria for pars planitis outlined in Table 3 appear to perform sufficiently well for use as classification criteria in clinical research.[16,17]

CREDIT ROLES

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