Classification Criteria for Punctate Inner Choroiditis

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

• PURPOSE: The purpose of this study was to determine classification criteria for punctate inner choroiditis (PIC).

• DESIGN: Machine learning of cases with PIC and 8 other posterior uveitides.

• METHODS: Cases of posterior uveitides were collected in an informatics-designed preliminary database, and a final database was constructed of cases achieving supermajority agreement on diagnosis by using formal consensus techniques. Cases were split into a training set and a validation set. Machine learning using multinomial logistic regression was used in the training set to determine a parsimonious set of criteria that minimized the misclassification rate among the posterior uveitides. The resulting criteria were evaluated in the validation set.

• RESULTS: A total of 1,068 cases of posterior uveitides, including 144 cases of PIC, were evaluated by machine learning. Key criteria for PIC included: 1) "punctate"-appearing choroidal spots <250 μm in diameter; 2) absent to minimal anterior chamber and vitreous inflammation; and 3) involvement of the posterior pole with or without mid-periphery. Overall accuracy for posterior uveitides was 93.9% in the training set and 98.0% (95% confidence interval: 94.3-99.3) in the validation set. The misclassification rates for PIC were 15% in the training set and 9% in the validation set.

• CONCLUSIONS: The criteria for PIC had a reasonably low misclassification rate and appeared to perform sufficiently well for use in clinical and translational research. (Am J Ophthalmol 2021;228: 1–6. © 2021 Elsevier Inc. All rights reserved.)

Accepted for publication March 31, 2021.

[1] Supplemental Material available at AJO.com.

[2] WRITING COMMITTEE: Douglas A. Jabs, Antoine P. Brezin, Ralph D. Levinson, Susan L. Lightman, Peter McCluskey, Neal Oden, Alan G. Palestine, Narsing A. Rao, Jennifer E. Thorne, Brett E. Trusko, Albert Vitale, and Susan E. Wittenberg.

[3] AUTHOR AFFILIATIONS: Members of the SUN Working Group are listed online at AJO.com. Department of Epidemiology (D.A.J., J.E.T.), Johns Hopkins University Bloomberg School of Public Health, and Department of Ophthalmology (D.A.J., J.E.T.), Wilmer Eye Institute, Johns Hopkins University School of Medicine, Baltimore, Maryland, USA; Department of Ophthalmology (A.P.B.), University of Paris V-Hôpital Cochin, Paris, France; Department of Ophthalmology (R.D.L.), UCLA Stein Eye Institute, David Geffen School of Medicine, University of Southern California Los Angeles, Los Angeles, California, USA; Moorfields Eye Hospital (S.L.L.), London, UK; Institute of Ophthalmology (S.L.L.), University College London, London, UK; Department of Ophthalmology (P.M.), Save Sight Institute, University of Sydney School of Medicine, Sydney, New South Wales, Australia; Emmes Company (N.O.), LLC, Rockville, Maryland, USA; Department of Ophthalmology (A.G.P.), University of Colorado School of Medicine, Aurora, Colorado, USA; Department of Ophthalmology (N.A.R.), USC Roski Eye Institute, Keck Medicine of USC, University of Southern California, Arcadia,, California, USA; Department of Medicine (B.E.T.), Texas A&M University, College Station, Texas, USA; Department of Ophthalmology (A.V.), University of Utah School of Medicine, Salt Lake City, Utah, USA; and the Houston Eye Associates (S.E.W.), Houston, Texas, USA.

[4] CONFLICT OF INTEREST: Douglas A. Jabs: none. Antoine P. Brezin: none. Ralph D. Levinson: none. Susan L. Lightman: none. Peter McCluskey: none. Neal Oden: none. Alan G. Palestine: none. Narsing A. Rao: none. Jennifer E. Thorne: Dr. Thorne engaged in a portion of this research as a consultant and was compensated for the consulting service. Brett E. Trusko:none. Albert Vitale: none. Susan E. Wittenberg: none.

* **Corresponding author:** Douglas A. Jabs, Department of Epidemiology, Johns Hopkins University Bloomberg School of Public Health, 615 N Wolfe St, Baltimore, Maryland 20215, USA.

PUNCTATE INNER CHOROIDITIS (PIC), ORIGINALLY termed punctate inner choroidopathy, was first described by Watzke and associates[1] in 1984 as a distinct type of multifocal choroiditis occurring typically in young adult myopic women patients and characterized by small "punctate" lesions of the inner choroid or retinal pigment epithelium or both. Lesions often had overlying subretinal fluid, and on fluorescein angiography in the acute phase were hyperfluorescent and leaked fluorescein dye. Anterior chamber and vitreous inflammation typically were absent. The lesions healed into atrophic scars, and choroidal neovascularization developed in 6 of 10 patients. Although often symptomatic with blurred vision, flashing lights, or paracentral scotomata, vision was minimally affected unless choroidal neovascularization developed.

Punctate inner choroiditis is an uncommon disease, accounting for ≤10% of posterior uveitides presenting to a tertiary care referral ophthalmology center,[2] and its incidence has been estimated at 0.4 cases/1,000,000 population/year.[3,4] The cause and pathogenesis of PIC are unknown; PIC is an eye-limited disease unassociated with a systemic disease. It has been speculated that PIC is an autoimmune process,[5] and the association between PIC and polymorphisms in the interleukin-10 and tumor necrosis factor-α genes has been taken as evidence of an autoimmune process.[6] However, it is unclear whether these genetic risk factors are risk factors for the disease itself or for the associated choroidal neovascularization.

Subsequent larger case series[2,7-10] have confirmed the clinical picture described by Watzke and associates.[1] Although there is a wide range of age at presentation, the age ranges from ~32-33 years. More than 90% of reported cases are women, and ~85%-92% are myopic. Active lesions are yellow to yellowish white, estimated at 100-300

0002-9394/$36.00
https://doi.org/10.1016/j.ajo.2021.03.046

μm in size, and largely located in the posterior pole. At presentation, ~55% will have bilateral disease, but bilateral disease has been reported to occur in as many as 88%.[10] Data from 1 study can be used to estimate the rate of bilateral disease as ~0.03/person-years.[7] More than 90% have no anterior chamber or vitreous inflammation, and other signs of inflammation (eg, posterior synechiae) typically are not present.[8] Approximately 50% of cases will present with choroidal neovascularization in at least 1 eye,[2,7-10] but with follow-up, estimates of choroidal neovascularization run as high ~75%-80%.[9,10] One study estimated the incidence of new and recurrent choroidal neovascularization as 0.02/eye-year (EY) and 0.04/EY, respectively.[11] Other structural complications of uveitis (eg, macular edema, disc edema) typically are not present.[8]

The active "punctate" lesions of PIC are distinct, yellow-white or cream-colored, typically round or oval, and <150 μm in size. There may be an overlying serous elevation, and the lesions may resolve or heal with atrophic scarring. Fluorescein angiography demonstrates early hyperfluorescence of active lesions with late staining. Late-stage atrophic scars are seen as window defects on fluorescein angiography.[3] Indocyanine angiography demonstrates hypofluorescent lesions throughout the angiogram.[3,12] More lesions may be evident on imaging than are appreciated on ophthalmoscopy. Optical coherence tomography (OCT) demonstrates focal hyperreflective elevation of the retinal pigment epithelium with corresponding disruption of the inner and outer segment photoreceptor interface.[13] Enhanced depth imaging OCT of acute lesions may demonstrate increased choroidal thickening.[14] Choroidal neovascularization, when present, is seen on fluorescein angiography, OCT, and OCT angiography.[3,13,15] Fundus autofluorescence demonstrates hyperautofluorescence of active lesions and may be useful in following the response to treatment.[15–17]

The course of PIC is variable. Active inflammatory lesions may spontaneously involute to small atrophic scars. Bilateral disease may occur simultaneously or asynchronously, with the disease in the second eye occurring years later. Rarely, choroidal neovascularization may spontaneously involute, but now nearly all choroidal neovascularization is treated with anti-vascular endothelial growth factor (VEGF) agents (eg, bevacizumab, ranibizumab, aflibercept). Patients with recurrent disease, chronic disease, bilateral choroidal neovascularization, or choroidal neovascularization requiring multiple injections of anti-VEGF agents typically are treated with oral corticosteroids and immunosuppression.[11,18,19] Case series suggest that, with appropriate use of immunosuppression, disease control, preservation of vision, and decreased or no need for anti-VEGF injections can be accomplished along with successful corticosteroid-sparing (prednisone, ≤7.5 mg/d) in most patients.[11,19] Rates of visual impairment (worse than 20/40) and blindness (20/200 or worse) have been estimated at 0.06/EY and 0.006/EY, respectively, with preservation of good vision typically observed in at least 1 eye.[11]

The Standardization of Uveitis Nomenclature (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. Among the diseases studied was PIC.[20-26]

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.[22-25]

• **INFORMATICS:** As previously described, the consensus-based informatics phase permitted the development of a standardized vocabulary and the development of a standardized, menu-driven hierarchical case collection instrument.[22]

• **CASE COLLECTION AND CASE SELECTION:** De-identified information was entered into the SUN preliminary database by the 76 contributing investigators for each disease as previously described.[24,25] 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 articles.[24,25] Because the goal was to develop classification criteria,[26] only cases with a supermajority agreement (>75%) that the case was the disease in question were retained in the final database (ie, were "selected").[24,25]

• **MACHINE LEARNING:** The final database then was randomly separated into a learning set (~85% of the cases) and a validation set (~15% of the cases) for each disease, as described in the accompanying article.[25] Machine learning was used on the learning set to determine criteria that minimized misclassification. The criteria then were tested on the validation set; for both the learning 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 PIC, the diseases against which it was evaluated included acute posterior multifocal placoid pigment epitheliopathy (APMPPE); birdshot chorioretinitis (BSCR) multifocal choroiditis with panuveitis (MFCPU); multiple evanescent white dot syndrome (MEWDS); serpiginous choroiditis; sarcoidosis-associated posterior uveitis; syphilitic posterior uveitis: and tubercular (TB) uveitis.

This study adhered to the principles of the Declaration of Helsinki. Institutional Review Boards (IRBs) at each participating center reviewed and approved the study;

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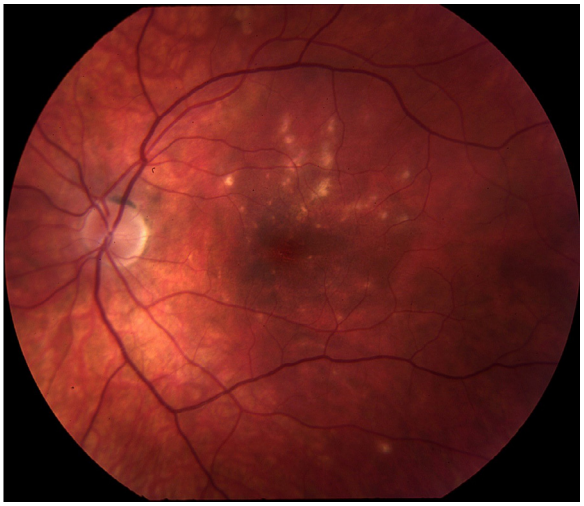

FIGURE 1. Fundus photograph of a case of punctate inner choroiditis, demonstrating "punctate" chorioretinal lesions in the posterior pole.

the study typically was considered either minimal risk or exempt by the individual IRBs.

RESULTS

A total of 250 cases of PIC were collected, and 144 (58%) achieved supermajority agreement on the diagnosis during the "selection" phase and were used in the machine learning phase. Those cases of PIC were compared to cases of posterior uveitides, including 82 cases of APMPPE, 207 cases of BSCR, 51 cases of MEWDS, 138 cases of MFCPU, 122 cases of serpiginous choroiditis, 12 cases of sarcoid posterior uveitis, 35 cases of syphilitic posterior uveitis, and 277 cases of TB posterior or panuveitis (including 96 cases of serpiginous-like TB choroiditis). Details of the machine learning results for those diseases are outlined in the accompanying article.[25] The characteristics of cases with PIC are listed in Table 1, and the classification criteria developed after machine learning are listed in Table 2. Key features of the criteria include: 1) characteristic punctate choroidal lesions (Figure 1); 2) absent or minimal anterior chamber and vitreous inflammation; and 3) posterior pole involvement. The overall accuracies for posterior uveitides were 93.9% in the learning set and 98.0% in the validation set (95% confidence interval: 94.3-99.3). The misclassification rate for PIC in the learning set was 15% and 9% in the validation set. The disease most often confused with PIC was MFCPU.

TABLE 1. Characteristics of Cases with Punctate Inner Choroiditis

Characteristic	Result
Number of cases	144
Demographics	
Median IQR (25th,75th) age, y	32 (25, 39)
Men, %	13
Women, %	87
Race/ethnicity, %	
White, non-Hispanic	81
Black, non-Hispanic	6
Hispanic	1
Asian, Pacific Islander	2
Other	4
Missing	6
Uveitis history	
Uveitis course, %	
Acute, monophasic	26
Acute, recurrent	6
Chronic	65
Indeterminate	3
Laterality, %	
Unilateral	41
Unilateral, alternating	0
Bilateral	59
Ophthalmic examination	
Keratic precipitates, %	
None	100
Anterior chamber cells, %	
Grade 0	99
½+	1
≥1+	0
Anterior chamber flare, %	
Grade 0	100
Iris, %	
Normal	100
IOP, involved eyes	
Median IQR (25th,75th) mm Hg	15 (13, 17)
Proportion of patients with IOP>24 mm Hg either eye, %	2
Vitreous cells, %	
Grade 0	91
½+	9
≥1+	0
Vitreous haze, %	
Grade 0	99
½+	1
≥1+	0
Chorioretinitis characteristics	
Lesion number, %	
Unifocal (1 lesion)	0
Paucifocal (2–4 lesions)	28
Multifocal (≥5 lesions)	72

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TABLE 1. (*continued*)

Characteristic	Result
Lesion shape and character, %	
Ameboid or serpentine	0
Oval or round	18
Placoid	0
Atrophic	38
Punctate	82
Inflammatory lesion/scar location, %[a]	
Posterior pole involved	78
Posterior pole and periphery/mid-periphery involved	21
Mid-periphery and periphery only	1
Typical lesion size, %	
<125 μm	54
125–250 μm	33
250–500 μm	9
>500 μm	3
Missing	1
Other features, %	
Retinal vascular sheathing	3
Retinal vascular leakage	17
Choroidal neovascularization	19

IOP = intraocular pressure.
[a]Based on retinal photographs from 130 cases.

DISCUSSION

The classification criteria developed by the SUN Working Group for PIC had a moderate misclassification rate, indicating reasonable discriminatory performance compared with other posterior uveitides.[25]

Because of the rare occurrence of PIC-like lesions in 1 eye and MFCPU in the other and because of a similar appearance on multimodal imaging (other than lesion size), some investigators have considered PIC and MFCPU to be variants of the same disease.[27,28] Conversely, other investigators, classifying the 2 diseases based solely on chorioretinal morphology, have found clear-cut differences, namely the absence of anterior chamber and vitreous inflammation and the absence of uveitis-related structural complications other than choroidal neovascularization in PIC[8] and differences in course with prognostic import.[29] There also are differences between distributions of lesions of the two diseases; lesions in PIC are most often in the posterior pole, whereas lesions in MFCPU typically involve the mid-periphery and periphery with or without posterior pole involvement.[30,31] A study of cases of MFCPU and PIC using cluster analysis determined that 2 distinct clusters existed, conforming to the diagnoses of PIC and MFCPU and that the 2 distinguishing features were anterior chamber and vitreous inflammation (largely absent in PIC) and lesion location (posterior in PIC and peripheral in MFCPU).[30]

In the series by Shimada and associates,[32] histological evaluation of surgically removed choroidal neovascular membranes demonstrated inflammatory infiltrates in some cases of MFCPU but not in PIC, suggesting that they may be distinct diseases. Conversely, in the case series by Olsen and associates,[33] histological evaluation of surgically removed choroidal neovascular membranes from patients with PIC demonstrated the occasional lymphocyte, suggesting that the pathology may not be completely dissimilar from that of MFCPU. A genetic risk factor association study suggested similar haplotype associations in the *IL-10* and TNF loci for MFCPU and PIC, suggesting possible similarities in pathogenesis but allowing for different inciting events or epigenetic factors to influence phenotype.[29] Finally, if they were a single disease, one might expect the clinical presentation to be a Gaussian distribution with the overlap syndrome to be the most common presentation, which is not the case. The paradigms are the most common presentations, and overlap is uncommon.[8,27,30] Hence, the SUN Working Group has elected to define the diseases separately, recognizing that there will be a small number of cases with the appearance of an overlap. Most such cases behave more like MFCPU

TABLE 2. Classification Criteria for Punctate Inner Choroiditis

Criteria
1. Multifocal choroidal inflammatory lesions
   a. Predominant lesion size of <250 μm and
   b. Punctate lesion appearance
And
2. Lesion involvement of posterior pole with or without mid-periphery
And
3. Absent to minimal anterior chamber and vitreous inflammation
Exclusions
1. Positive serologic test result for syphilis using a treponemal test
2. Evidence of sarcoidosis (either bilateral hilar adenopathy on chest imaging or tissue biopsy results demonstrating non-caseating granulomata)

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than PIC and may be classified as MFCPU but probably should be classified as an overlap syndrome at this time.

The presence of any of the exclusions in Table 2 suggests an alternate diagnosis, and the diagnosis of PIC should not be made in their presence. In prospective studies, many of these tests will be performed routinely and the alternative diagnoses excluded, especially syphilis. However, in retrospective studies based on clinical care, not all of those tests may have been performed. In those studies, the presence of an exclusionary criterion excludes PIC, but the absence of such testing does not always exclude the diagnosis of PIC if the criteria for the diagnosis are met.

Classification criteria are used to diagnose individual diseases for research purposes.[26] Classification criteria differ from clinical diagnostic criteria in that, although both seek to minimize misclassification, when a trade-off is needed diagnostic criteria typically emphasize sensitivity, whereas classification criteria emphasize specificity,[26] 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 might confound the data. The machine learning process used did not explicitly use sensitivity and specificity; instead, it minimized the misclassification rate. Because the study was developing classification criteria and because the typical agreement between 2 uveitis experts as to diagnosis was moderate at best,[24] 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 with PIC may not be so classified by classification criteria.

In conclusion, the criteria for PIC outlined in Table 2 appear to perform sufficiently well for use as classification criteria in clinical research.[25]

CREDIT ROLES

Douglas A. Jabs, MD, MBA: Conceptualization, Methodology, Validation, Investigation, Data curation, Writing–Original draft, Writing–Review and editing, Visualization, Supervision, Project administration, Funding acquisition. **Antoine P. Brezin, MD:** Investigation, Writing–Review and editing. **Ralph D. Levinson, MD:** Investigation, Writing–Review and editing. **Susan L. Lightman, PhD, FRCP, FRCOphth:** Investigation, Writing–Review and editing. **Peter McCluskey, MD:** Investigation, Data curation, Writing–Review and editing. **Neal Oden, PhD:** Methodology, Software, Validation, Formal analysis, Investigation, Resources, Data curation, Writing–Review and editing. **Alan G. Palestine, MD:** Investigation, Writing–Review and editing. **Narsing A. Rao, MD:** Investigation, Writing–Review and editing. **Jennifer E. Thorne, MD, PhD:** Methodology, Software, Validation, Formal analysis, Investigation, Data curation, Writing–Review and editing. **Brett E. Trusko, PhD, MBA:** Methodology, Software, Resources, Data curation, Investigation, Writing–Review and editing. **Albert Vitale, MD:** Investigation, Writing–Review and editing. **Susan E. Wittenberg, MD:** Investigation, Writing–Review and editing.

All authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest and none were reported.

FUNDING/SUPPORT: The Standardization of Uveitis Nomenclature (SUN) Working Group was supported by National Eye Institute/National Institutes of Health grant R01 EY026593; the David Brown Fund; the Jillian M. and Lawrence A. Neubauer Foundation; and the New York Eye and Ear Foundation.

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