

Evidence and Consensus Based Guidelines for Imaging in Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE) - Multimodal imaging in Uveitis (MUV) Taskforce: Report 7

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Evidence and Consensus Based Guidelines for Imaging in Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE) - Multimodal imaging in Uveitis (MUV) Taskforce: Report 7

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Short Title: Imaging Guidelines for APMPPE

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Abstract

Purpose: To develop consensus-based guidelines on use and interpretation of multimodal imaging in acute posterior multifocal placoid pigment epitheliopathy (APMPPE).

Design: Consensus agreement led by literature, and an expert committee using a nominal group technique (NGT).

Methods: The expert committee for APMPPE performed a thorough review of representative cases of APMPPE. The cases were used to develop guidelines for the diagnosis and follow-up of APMPPE using color fundus photography (CFP), optical coherence tomography (OCT), fundus fluorescein angiography (FFA), indocyanine green angiography (ICGA), fundus autofluorescence (FAF), and OCT angiography (OCTA). Structured NGT-based discussions were used to achieve consensus-based recommendations on imaging characteristics, disease activity, and complications, and subsequently were adopted by a vote of the entire taskforce.

Results: Diagnosis of active APMPPE is characterized by distinctive imaging findings on CFP, and hyper-reflectivity of the ellipsoid zone (EZ), external limiting membrane (ELM), and outer nuclear layer (ONL) on OCT. Choriocapillaris non-perfusion, detectable via early-phase FFA, ICGA or OCTA, is crucial. In the early stages of APMPPE, OCT findings may be unremarkable, making FFA, ICGA, and/or OCTA relevant for the diagnosis. Based on the imaging findings, APMPPE can be classified into four stages of activity: choroidal, chorioretinal, transitional, and resolved. Following diagnosis, OCT and OCTA can be used to monitor lesion activity and identify potential complications.

Conclusions: MUV imaging criteria enable the identification of key diagnostic features for APMPPE, extending the Standardization of Uveitis Nomenclature (SUN) classification. These consensus-based guidelines provide a framework for evaluating disease activity and complications, enhancing diagnostic accuracy and guiding clinical management.

Introduction

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) was first described in 1968 by Sir J. Donald M. Gass who initially attributed the condition to an acute cellular response of the retinal pigment epithelium (RPE) to a local harmful agent.¹ However, in 1972, based on early fundus fluorescein angiography (FFA) frames showing choriocapillaris nonperfusion, Deutman and colleagues suggested that the choriocapillaris, rather than the RPE, was primarily involved in the disease.² They proposed renaming the disease to "acute multifocal ischemic choroidopathy" (AMIC).³ Despite this, the original term 'APMPPE' has prevailed in the literature.

In 2021, the Standardization for Uveitis Nomenclature (SUN) Working Group published the classification criteria for APMPPE.⁴ The key criteria for diagnosing the acute phase of the disease were the presence of paucifocal or multifocal choroidal lesions with a plaque-like or placoid appearance, and characteristic FFA features, defined by early hypofluorescence and late diffuse hyperfluorescence. These criteria apply only when both a positive syphilis treponemal test and evidence of sarcoidosis are absent.⁴

While the SUN working group provided a clinical and imaging-based diagnostic criteria for APMPPE, given the availability of multimodal imaging, it is proposed to expand consensus-driven guidelines on the use of these modalities for the diagnosis and management of APMPPE. The advances in ophthalmic imaging modalities, such as fundus autofluorescence (FAF), optical coherence tomography (OCT), OCT angiography (OCTA), FFA, and indocyanine green angiography (ICGA), provide valuable insights that should be incorporated into the evolving classification criteria.

The Multimodal Imaging in Uveitis (MUV) task force, an international collaboration, has developed imaging-based guidelines to enhance the existing SUN classification for five of the most common multifocal choroidopathies. This manuscript specifically focuses on APMPPE, outlining not only proposed diagnostic imaging criteria but also recommendations for the use of imaging in evaluating disease activity and guiding management.

Methods

The MUV taskforce was established as an international research consortium by the International Uveitis Study Group (IUSG) with a goal of bringing together a committee of experts to formulate imaging-based guidelines for diagnosing APMPPE based on nominal group technique (NGT). The MUV utilized the SUN criteria⁴ to diagnose and identify cases of APMPPE to be included in imaging analysis. The study was conducted under the tenets of the Declaration of Helsinki. The study was granted Institute Review Board (IRB) exemption by the Vanderbilt University Medical Center (IRB #240146).

Subcommittee Selection

The MUV task force established a dedicated subcommittee for APMPPE to develop expert consensus-based imaging criteria and guidelines for the use of

multimodal imaging. The expert group oversaw the process to ensure methodological rigor and consistency. The subcommittee comprised of seven members who are leading experts in uveitis and medical retina, selected to represent diverse geographical regions and account for global variations in disease presentation. Before conducting the study, the members of the APMPE subcommittee performed a thorough review of the published literature. The experts agreed to utilize at least 15 illustrative cases of APMPE to support the development of imaging criteria. We followed the principles of Standards for Reporting Qualitative Research: A Synthesis of Recommendations (SRQR)⁵ for reporting the results of our study.

Case Selection

All the members of the subcommittee submitted images of APMPE cases with different stages of activity that met the SUN Working Group criteria.⁴ Cases with multimodal imaging, namely color fundus photographs (CFP) as a surrogate for clinical examination, OCT, FFA, FAF, ICGA and OCTA, were included. Only de-identified images were shared amongst the subcommittee members. The members analyzed the cases of APMPE, and only those categorized as definitive APMPE (where all the experts agreed on the diagnosis of APMPE) were included. Cases with infectious etiologies, such as syphilis and tuberculosis, and other known diseases such as sarcoidosis-associated uveitis, were excluded.

Nominal Group Technique

The subcommittee employed a structured nominal group technique (NGT) to systematically review the critical imaging features of APMPE. The subcommittee consisted of a rigid design with one neutral moderator (EC). A series of online meetings were conducted to include all the members of the expert committee. The subcommittee members first discussed all the available literature specifically related to imaging in APMPE, and the primary research question of the study. Thereafter, the members presented their ideas without any interruption, discussion or criticism. Subsequently, the moderator conducted anonymous voting to identify the features of active disease and distinguishing characteristics of APMPE across various imaging modalities. The NGT discussions were continued for identifying the most effective imaging modalities for assessing complications of the disease, including choroidal neovascularization (CNV). The NGT discussions conducted by the APMPE subcommittee led to the development of consensus statements, each approved by a supermajority (over 75% of votes) within the subcommittee. These consensus statements formed the basis for drafting the recommendations subsequently voted on by the MUV Task Force (see below).

Establishment of Consensus

The subcommittee members drafted recommendations for using imaging to diagnose, monitor and manage complications of APMPE. These preliminary recommendations were developed following online discussions held by the subcommittee members. Subsequently, the MUV taskforce (comprising 49 members; Appendix I) from diverse geographical regions and subspecialty training in uveitis and medical retina reviewed the draft. Using an anonymous online survey system,

the taskforce members assessed the recommendations, and any requested modifications were discussed collaboratively. Finally, a consensus was achieved by all members of the MUV taskforce defined as follows:

Unanimous consensus: 100% participants agree

Strong consensus: > 95% vote

Consensus: > 75-95% vote

Majority agreement: > 50-75% vote

No consensus: < 50% vote (lack of agreement or divided votes)

The percentage thresholds for consensus derived by voting were reported as per the guidelines of various international associations. These included the Guidelines International Network (GIN),⁶ European League Against Rheumatism (EULAR),⁷ and Association of Scientific Medical Associations of Germany (AWMF).⁸ When there was no consensus achieved (<50% vote), the guidelines were rejected. Study data were collected and managed using REDCap electronic data capture tools hosted at Vanderbilt University Medical Center.^{9,10} REDCap (Research Electronic Data Capture) is a secure, web-based software platform designed to support data capture for research studies, providing 1) an intuitive interface for validated data capture; 2) audit trails for tracking data manipulation and export procedures; 3) automated export procedures for seamless data downloads to common statistical packages; and 4) procedures for data integration and interoperability with external sources.

Results

A. Imaging Features of Active APMPE

1. Color Fundus Photography

On CFP, the lesions of APMPE are paucifocal (3 to 5) or multifocal, typically bilateral, though the lesions may appear only in one eye initially.^{11,12} These lesions are whitish or yellow in color, deep, and have poorly defined borders, often appearing slightly elevated or with a blurred contour. They are mainly located in the posterior pole (within the temporal arcades), but significant involvement of the peripheral retina has also been reported.^{13,14} On CFP, the lesions are seen as deep (subretinal) with visible overlying retinal vessels (Figure 1). The size of the lesions usually ranges from 100 to 2000 μm ; however, lesions can occasionally become confluent, forming larger lesions (Figure 2).

2. Optical Coherence Tomography (OCT)

On OCT, the typical active lesion of APMPE is characterized by hyper-reflective areas at the level of the outer retina, involving the ellipsoid zone (EZ), external limiting membrane (ELM) and outer nuclear layer (ONL) and thickened, featureless choriocapillaris zone just underlying the area of hyper-reflectivity and extending slightly beyond (Figure 3). The subcommittee agreed that involvement of the ONL is a typical manifestation of acute APMPE (note that ONL involvement may be absent in very early/choroidal lesions), and absence of ONL involvement should prompt suspicion for an alternative diagnosis.

The non-foveal lesions often exhibit areas of hyperreflectivity in the outer nuclear layer, which align with the angular contours of the Henle fiber layer (HFL) neurons. This pattern is referred to as the ASHH sign (“angular Sign of HFL hyper-reflectivity”) (Figure 4).¹⁵ Although the ASHH sign is commonly associated with APMPE, it is not pathognomonic to this condition and has also been observed in other retinal disorders, including acute macular retinopathy and whiplash maculopathy.¹⁵ The presence of this sign suggests an acute disruption of the photoreceptors involving the HFL. In addition, the presence of dome-shaped elevation of the EZ or bacillary layer detachment mimicking Vogt-Koyanagi-Harada disease can be associated with acute APMPE (Figure 5).^{16–18}

3. Fundus Autofluorescence (FAF)

In FAF imaging of APMPE lesions, the acute phase is characterized by initial hypo-autofluorescence, which results from the masking of the RPE pigments by outer retinal edema. As the disease progresses, there is a gradual increase in hyper-autofluorescence, likely due to lipofuscin accumulation in the damaged RPE. This is followed by a decrease in hyper-autofluorescence with occasional areas of focal hypo-autofluorescence in the later stages, reflecting partial loss of the RPE. Moreover, lesions at different stages of activity may coexist within the same patient (Figure 6).

4. Fundus Fluorescein Angiography (FFA)

The characteristic FFA findings in acute APMPE lesions include hypofluorescence in the early frames, followed by diffuse hyperfluorescence in the late frames of FFA. In addition, the choroidal phase of FFA can identify lesions that may not be visible in the late frames (Figure 7).^{14,19} FFA plays a crucial role in differentiating APMPE from serpiginous choroiditis. In patients with serpiginous choroiditis, FFA can highlight different activity stages within the same lesion. These findings highlight the importance of FFA in distinguishing between these two conditions, as they may have overlapping clinical features but distinct patterns on angiography.

5. Indocyanine Green Angiography (ICGA)

The ICGA in active APMPE typically shows hypofluorescent lesions that persist through the late frames of the angiogram (Figure 8).^{14,20–22} Similar to the early frames of FFA, ICGA can reveal lesions that are not visible in the late frames of fluorescein angiography. However, the subcommittee agreed that a significant discrepancy between FFA and ICGA findings is not typical of APMPE. Lack of choriocapillaris hypoperfusion on ICGA is an exclusion criterion for active APMPE.

6. Optical Coherence Tomography Angiography (OCTA)

OCTA in active APMPE is characterized by areas of flow deficit in the choriocapillaris slab, corresponding to the same areas seen in ICGA (Figure 9).^{19,23–26} Absence of flow deficit on OCTA excludes active APMPE. Similar to ICGA and the early frames of FFA, OCTA can detect lesions that do not show any corresponding reflection in OCT, FAF, or clinical fundus examination (Figure

10).^{19,23,27} The area of flow deficit in the choriocapillaris is larger than the corresponding area of hyper-reflectivity seen in the outer retina on structural OCT. This suggests that the primary site of inflammation in APMPE is at the level of the choriocapillaris, ruling out shadowing as a confounding factor for the observed flow deficit (Figure 11).

The consensus statements for active APMPE are described in Table 1.

B. Imaging Features of Healing APMPE

The healing process of APMPE lesions can be tracked through interval changes observed in various imaging modalities. The subcommittee outlined four phases of activity based on the progression of lesion healing and emphasized the significant role of OCT and OCTA in assessing these changes. Active lesions on structural OCT are characterized by hyper-reflectivity in the outer retina, including the EZ, ELM, and ONL. In later stages, these lesions may show relative hypo-reflectivity or disruption of the EZ, often accompanied by subtle RPE alterations. On OCTA, areas of flow deficit gradually diminish over time, with near-complete resolution of the deficit marking advanced stages of healing.

The consensus statements regarding the four phases of healing in APMPE on multimodal imaging are described in Table 2.

C. Consensus-based Recommendations

The details of the recommendations drafted by the APMPE subcommittee and reviewed by the MUV taskforce is presented in Table 3. The table also provides the strength of the consensus.

Discussion

The MUV task force focused on developing comprehensive imaging guidelines for the diagnosis and management of APMPE and identifying multimodal imaging features to assess disease activity and potential complications. Although clinical information remains essential for a conclusive APMPE diagnosis, advanced imaging techniques provide crucial insights that aid in diagnosis and differentiation from similar conditions, such as serpiginous choroiditis. Consequently, establishing consensus-based imaging diagnostic criteria for APMPE is of paramount importance. This approach helps to move away from the potentially misleading term "white dot syndromes," fostering more precise and standardized diagnostic terminology.

According to the current SUN criteria, a diagnosis of APMPE requires a negative syphilis test and the exclusion of other diseases, such as sarcoidosis or tuberculosis.⁴ These criteria remain essential; however, the integration of multimodal imaging aims to further enhance the SUN criteria, provided that infectious and other identifiable uveitic disease have been excluded. In general, APMPE is a self-limited disease with a favorable outcome.^{1,2,28} Recurrences, however, should prompt reevaluation of the initial diagnosis,²⁸ considering conditions such as relentless placoid chorioretinitis (RPC)²⁹ or persistent placoid maculopathy (PPM)³⁰ in these

cases. It is relevant to distinguish APMPE from other conditions. Conditions such as RPC and PPM can mimic APMPE. However, our expert committee did not reach any consensus on differentiating between APMPE and these conditions using multimodal imaging. Future efforts with imaging research in this direction are needed.

Distinguishing APMPE from serpiginous choroiditis is essential. The expert committee emphasized that disease progression is a key differentiating factor: APMPE lesions typically resolve, with recovery of choriocapillaris perfusion, whereas serpiginous choroiditis is characterized by persistent choriocapillaris ischemia and progressive tissue damage. The committee noted that early-stage serpiginous choroiditis often appears unifocal on CFP, which can aid in differentiating it from APMPE, which typically presents as pauci- or multifocal. Serpiginous choroiditis also shows a characteristic active edge of inflammation. Additionally, the persistent absence of choriocapillaris flow recovery is indicative of serpiginous choroiditis. The committee further observed that serpiginous choroiditis is more prone to recurrence, typically at the edges of existing lesions, whereas APMPE rarely recurs, and if it does, new lesions form rather than reactivating at the edges of prior ones. However, these observations require further validation through future studies and analyses. Lesions indicative of serpiginous choroiditis can coexist with APMPE lesions in the same patient, a rare condition known as ampiginous choroiditis.³¹ However, our expert committee did not specifically analyze such cases with potential overlap. While complications like CNV are uncommon in APMPE, they have been reported in other related conditions, such as PPM.^{30,32–34} When CNV occurs in APMPE, OCTA, alongside FFA and ICGA may be used for the detection of the neovascularization.

The MUV task force emphasized the importance of OCT, ICGA and OCTA for diagnosing and monitoring APMPE lesions. Key elements include detecting outer retinal hyper-reflectivity on OCT and assessing choriocapillaris involvement through early-phase FFA, ICGA, or OCTA. The findings of the committee underscore the critical role of multimodal imaging in diagnosing APMPE, serving as an essential complement to established criteria such as those outlined by the SUN.⁴ This proposed minimal imaging set is slated for a validation exercise to confirm its efficacy and reliability.

The strength of the MUV consensus-based recommendations for APMPE include robust NGT-led discussions by experts in uveitis and medical retina from diverse geographical backgrounds, voted upon by a larger group of taskforce members. Despite the strengths of such an effort, there are several limitations that must be acknowledged. As discussed previously, the MUV expert committee admitted that imaging alone was insufficient to provide differentiating clues between early APMPE and serpiginous choroiditis, and further research is needed to distinguish between APMPE and other placoid chorioretinopathies such as RPC and PPM. The imaging-based consensus guidelines need prospective validation on a larger cohort of patients. Atypical and recurrent cases of APMPE may be difficult to diagnose on multimodal imaging and may have unusual or novel imaging findings. Certain imaging modalities such as OCTA have a learning curve for interpretation of specific features of active disease and identification of various stages of healing.

In conclusion, the consensus guidelines for APMPE by the MUV task force aim to standardize uveitis diagnostics by providing uniform definitions for diagnosis and disease activity. The consistent application and interpretation of multimodal imaging in the management of APMPE are expected to enhance clinical practice and research, leading to more accurate diagnoses and improved treatment outcomes. As imaging technology continues to evolve, ongoing efforts to update and refine these recommendations will be essential to ensure that clinicians use consistent terminology in their reports and practice.

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VG: None

Appendix I

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References

1. Gass JD. Acute posterior multifocal placoid pigment epitheliopathy. *Arch Ophthalmol*. 1968;80(2):177-185. doi:10.1001/archopht.1968.00980050179005
2. Deutman AF, Oosterhuis JA, Boen-Tan TN, Aan de Kerk AL. Acute posterior multifocal placoid pigment epitheliopathy. Pigment epitheliopathy of choriocapillaritis? *Br J Ophthalmol*. 1972;56(12):863-874. doi:10.1136/bjo.56.12.863
3. Deutman AF. Acute multifocal ischaemic choroidopathy and the choriocapillaris. *Int Ophthalmol*. 1983;6(2):155-160. doi:10.1007/BF00127644
4. Standardization of Uveitis Nomenclature (SUN) Working Group. Classification Criteria for Acute Posterior Multifocal Placoid Pigment Epitheliopathy. *Am J Ophthalmol*. 2021;228:174-181. doi:10.1016/j.ajo.2021.03.056
5. O'Brien BC, Harris IB, Beckman TJ, Reed DA, Cook DA. Standards for reporting qualitative research: a synthesis of recommendations. *Acad Med*. 2014;89(9):1245-1251. doi:10.1097/ACM.0000000000000388
6. Qaseem A, Forland F, Macbeth F, et al. Guidelines International Network: toward international standards for clinical practice guidelines. *Ann Intern Med*. 2012;156(7):525-531. doi:10.7326/0003-4819-156-7-201204030-00009
7. EULAR Recommendations: Recommendations for Management. Accessed December 4, 2024. <https://www.eular.org/recommendations-management>
8. Association of the Scientific Medical Societies in Germany (AWMF) – Standing Guidelines Commission. AWMF Guidance Manual and Rules for Guideline Development. Version 2.1, 2023. Accessed December 4, 2024. https://www.awmf.org/fileadmin/user_upload/dateien/downloads_regelwerk/en_20230905_AWMF-Regelwerk_2023_V2.1.pdf
9. Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap)--a metadata-driven methodology and workflow process for providing translational research informatics support. *J Biomed Inform*. 2009;42(2):377-381. doi:10.1016/j.jbi.2008.08.010
10. Harris PA, Taylor R, Minor BL, et al. The REDCap consortium: Building an international community of software platform partners. *J Biomed Inform*. 2019;95:103208. doi:10.1016/j.jbi.2019.103208
11. Mordechaev E, Shakarov G, Parikh D. Unilateral acute posterior multifocal placoid pigment epitheliopathy (APMPPE) with delayed contralateral eye involvement. *BMC Ophthalmol*. 2024;24(1):17. doi:10.1186/s12886-023-03221-8
12. Kutluturk I, Agarwal A, Shulman S, et al. The Clinical Characteristics of Unilateral Placoid Pigment Epitheliopathies. *Ocul Immunol Inflamm*. 2021;29(6):1072-1079. doi:10.1080/09273948.2019.1705498

13. Mrejen S, Sarraf D, Chexal S, Wald K, Freund KB. Choroidal Involvement in Acute Posterior Multifocal Placoid Pigment Epitheliopathy. *Ophthalmic Surg Lasers Imaging Retina*. 2016;47(1):20-26. doi:10.3928/23258160-20151214-03
14. Chan X, Amoroso JR, Hoang QV, Jung JJ. The Spectrum of Relentless Placoid and Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE): Multimodal Imaging Analysis. *Retin Cases Brief Rep*. Published online March 1, 2024. doi:10.1097/ICB.0000000000001555
15. Ramtohul P, Cabral D, Satta S, Freund KB, Sarraf D. The OCT angular sign of Henle fiber layer (HFL) hyperreflectivity (ASHH) and the pathoanatomy of the HFL in macular disease. *Prog Retin Eye Res*. 2023;95:101135. doi:10.1016/j.preteyeres.2022.101135
16. Kohli GM, Bhatia P, Shenoy P, Sen A, Gupta A. Bacillary Layer Detachment in Hyper-acute Stage of Acute Posterior Multifocal Placoid Pigment Epitheliopathy: A Case Series. *Ocul Immunol Inflamm*. Published online September 23, 2020:1-4. doi:10.1080/09273948.2020.1823423
17. Kuroiwa DAK, Ribeiro LZ, Regatieri CVS. Acute posterior multifocal placoid pigment epitheliopathy with bacillary layer detachment. *Am J Ophthalmol*. 2022;235:e345-e346. doi:10.1016/j.ajo.2021.10.026
18. Ramtohul P, Denis D, Gascon P. Bacillary Layer Detachment in Acute Posterior Multifocal Placoid Pigment Epitheliopathy: A Multimodal Imaging Analysis. *Retina*. 2021;41(2):e12-e14. doi:10.1097/IAE.0000000000003014
19. Heiferman MJ, Rahmani S, Jampol LM, et al. ACUTE POSTERIOR MULTIFOCAL PLACOID PIGMENT EPITHELIOPATHY ON OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY. *Retina*. 2017;37(11):2084-2094. doi:10.1097/IAE.0000000000001487
20. Dhaliwal RS, Maguire AM, Flower RW, Arribas NP. Acute posterior multifocal placoid pigment epitheliopathy. An indocyanine green angiographic study. *Retina*. 1993;13(4):317-325. doi:10.1097/00006982-199313040-00009
21. Howe LJ, Woon H, Graham EM, Fitzke F, Bhandari A, Marshall J. Choroidal hypoperfusion in acute posterior multifocal placoid pigment epitheliopathy. An indocyanine green angiography study. *Ophthalmology*. 1995;102(5):790-798. doi:10.1016/s0161-6420(95)30955-4
22. Yuzawa M, Kawamura A, Matsui M. Indocyanine green video angiographic findings in acute posterior multifocal placoid pigment epitheliopathy. *Acta Ophthalmol (Copenh)*. 1994;72(1):128-133. doi:10.1111/j.1755-3768.1994.tb02753.x
23. Burke TR, Chu CJ, Salvatore S, et al. Application of OCT-angiography to characterise the evolution of chorioretinal lesions in acute posterior multifocal placoid pigment epitheliopathy. *Eye (Lond)*. 2017;31(10):1399-1408. doi:10.1038/eye.2017.180

24. Mebsout-Pallado C, Orès R, Terrada C, et al. Review of the Current Literature and Our Experience on the Value of OCT-angiography in White Dot Syndromes. *Ocul Immunol Inflamm*. 2022;30(2):364-378. doi:10.1080/09273948.2020.1837185
25. Klufas MA, Phasukkijwatana N, Iafe NA, et al. Optical Coherence Tomography Angiography Reveals Choriocapillaris Flow Reduction in Placoid Chorioretinitis. *Ophthalmol Retina*. 2017;1(1):77-91. doi:10.1016/j.oret.2016.08.008
26. Furino C, Shalchi Z, Grassi MO, et al. OCT Angiography in Acute Posterior Multifocal Placoid Pigment Epitheliopathy. *Ophthalmic Surg Lasers Imaging Retina*. 2019;50(7):428-436. doi:10.3928/23258160-20190703-04
27. Dolz-Marco R, Sarraf D, Giovinazzo V, Freund KB. OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY SHOWS INNER CHOROIDAL ISCHEMIA IN ACUTE POSTERIOR MULTIFOCAL PLACOID PIGMENT EPITHELIOPATHY. *Retin Cases Brief Rep*. 2017;11 Suppl 1:S136-S143. doi:10.1097/ICB.0000000000000473
28. Papisavvas I, Mantovani A, Herbort CP. Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE): A Comprehensive Approach and Case Series: Systemic Corticosteroid Therapy Is Necessary in a Large Proportion of Cases. *Medicina (Kaunas)*. 2022;58(8):1070. doi:10.3390/medicina58081070
29. Jones BE, Jampol LM, Yannuzzi LA, et al. Relentless placoid chorioretinitis: A new entity or an unusual variant of serpiginous chorioretinitis? *Arch Ophthalmol*. 2000;118(7):931-938.
30. Golchet PR, Jampol LM, Wilson D, Yannuzzi LA, Ober M, Stroh E. Persistent placoid maculopathy: a new clinical entity. *Ophthalmology*. 2007;114(8):1530-1540. doi:10.1016/j.ophtha.2006.10.050
31. Jyotirmay B, Jafferji SS, Sudharshan S, Kalpana B. Clinical profile, treatment, and visual outcome of ampiginous choroiditis. *Ocul Immunol Inflamm*. 2010;18(1):46-51. doi:10.3109/09273940903402637
32. Parodi MB, Iacono P, Bandello F. Juxtafoveal choroidal neovascularization secondary to persistent placoid maculopathy treated with intravitreal bevacizumab. *Ocul Immunol Inflamm*. 2010;18(5):399-401. doi:10.3109/09273948.2010.483316
33. Gendy MG, Fawzi AA, Wendel RT, Pieramici DJ, Miller JA, Jampol LM. Multimodal imaging in persistent placoid maculopathy. *JAMA Ophthalmol*. 2014;132(1):38-49. doi:10.1001/jamaophthalmol.2013.6310
34. Nika M, Kalyani PS, Jayasundera KT. PATHOGENESIS OF PERSISTENT PLACOID MACULOPATHY: A Multimodal Imaging Analysis. *Retina*. 2015;35(8):1531-1539. doi:10.1097/IAE.0000000000000496

References

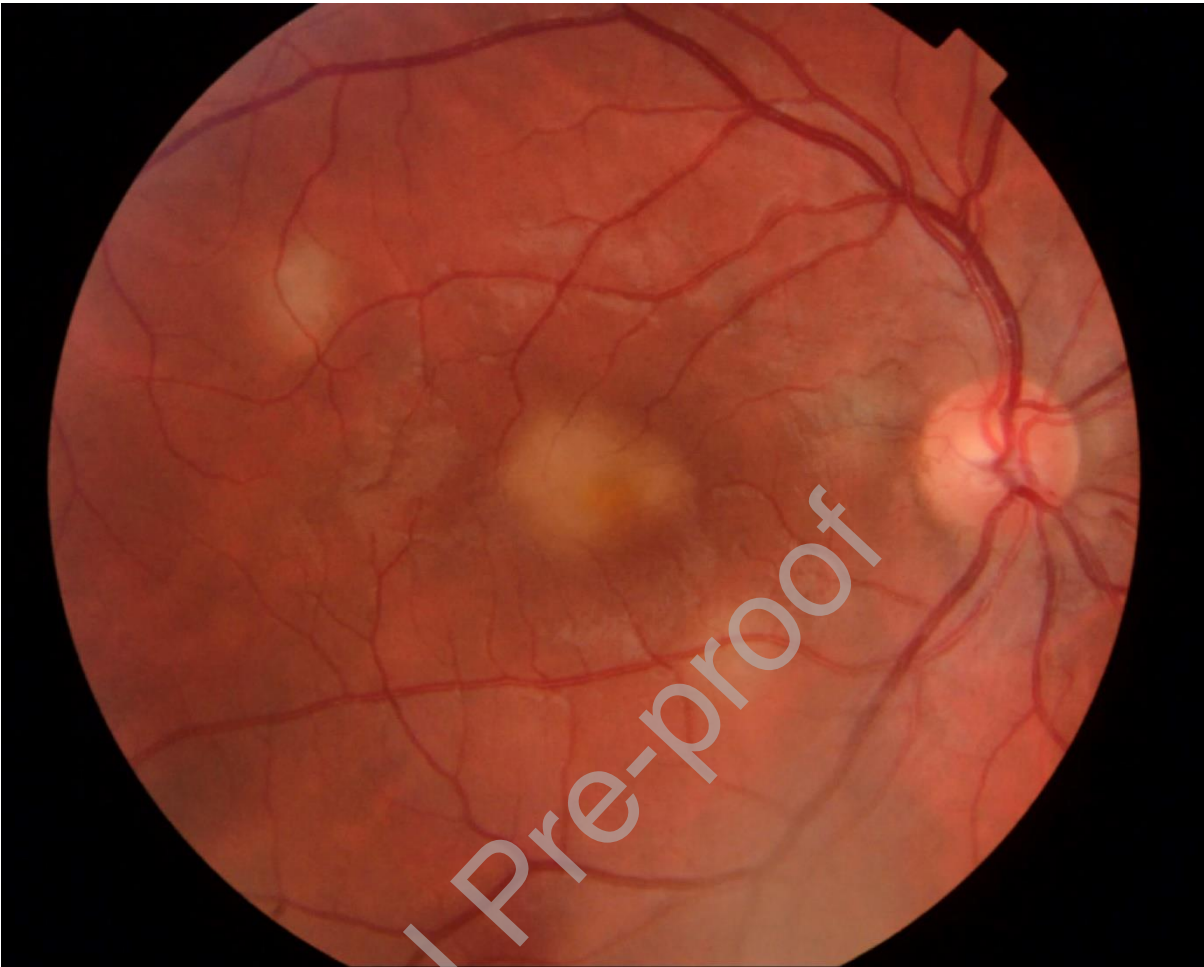


Figure 1: Color fundus photography of the right eye (OD) of a patient with APMPPE showing the typical appearance of the acute outer retinal lesions.

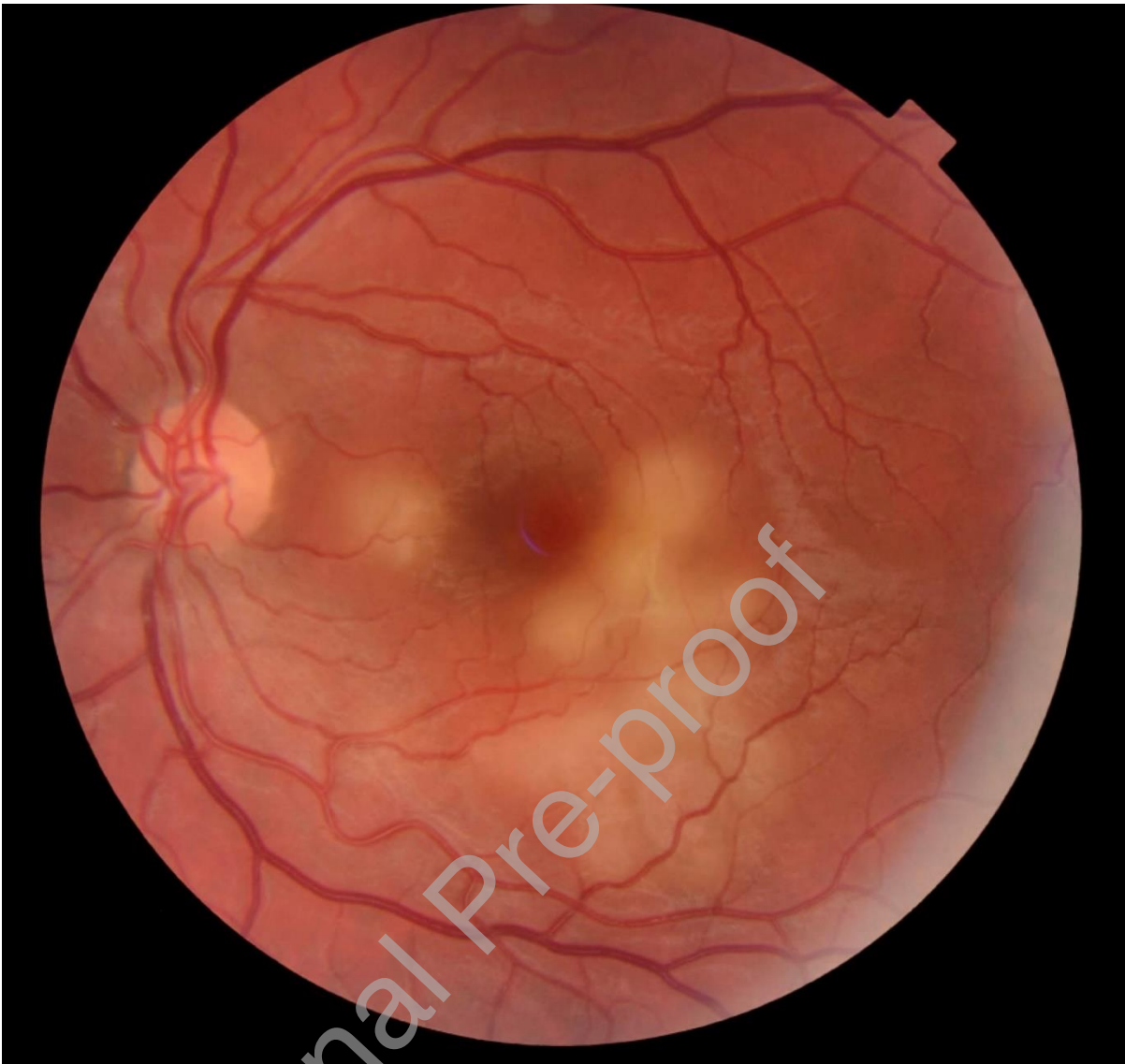


Figure 2: Color fundus photography of the left eye (OS) of a patient with APMPE showing confluent lesions temporal to the fovea.

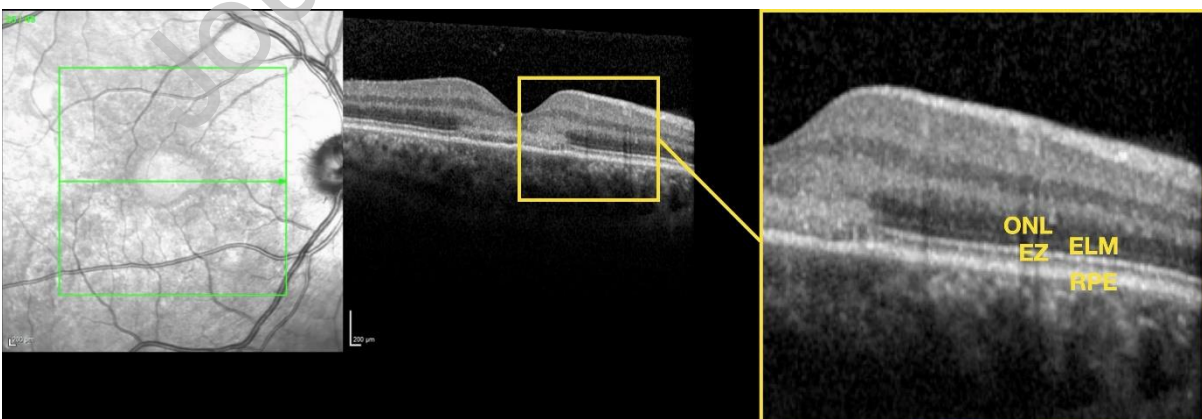


Figure 3: Optical coherence tomography of the right eye (OD) of a patient with APMPE showing a hyperreflective lesion involving the ellipsoid zone (EZ), external limiting membrane (ELM) and outer nuclear layer (ONL). RPE: Retinal Pigment Epithelium.

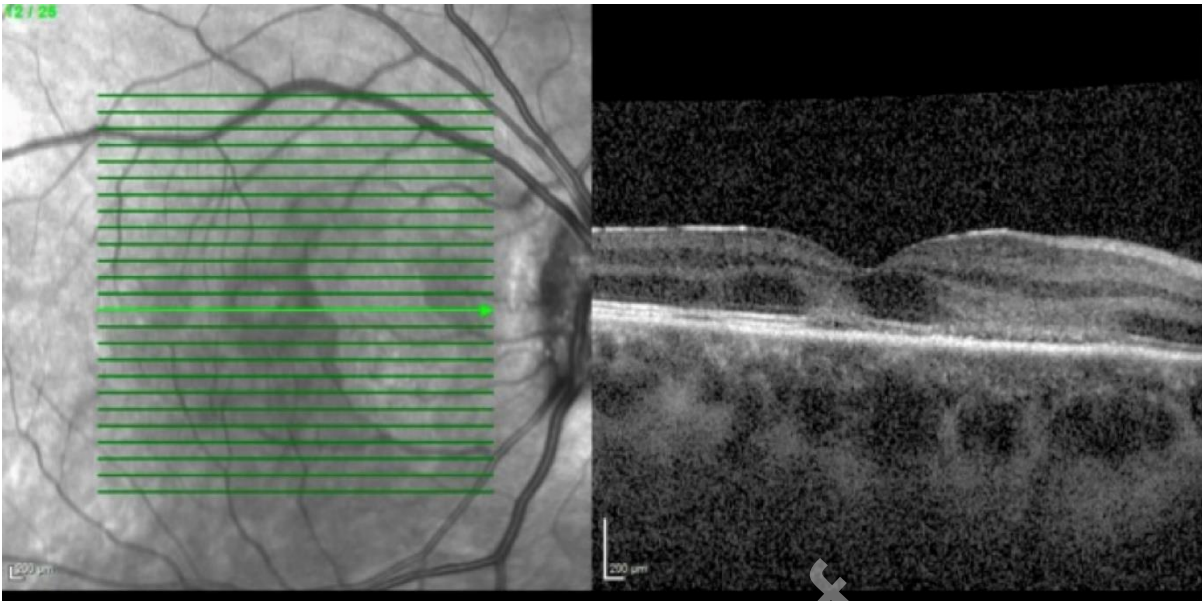


Figure 4: Optical coherence tomography of the right eye (OD) of a patient with APMPE showing the characteristic angular sign of Henle fiber layer (HFL) hyperreflectivity (ASHH sign).

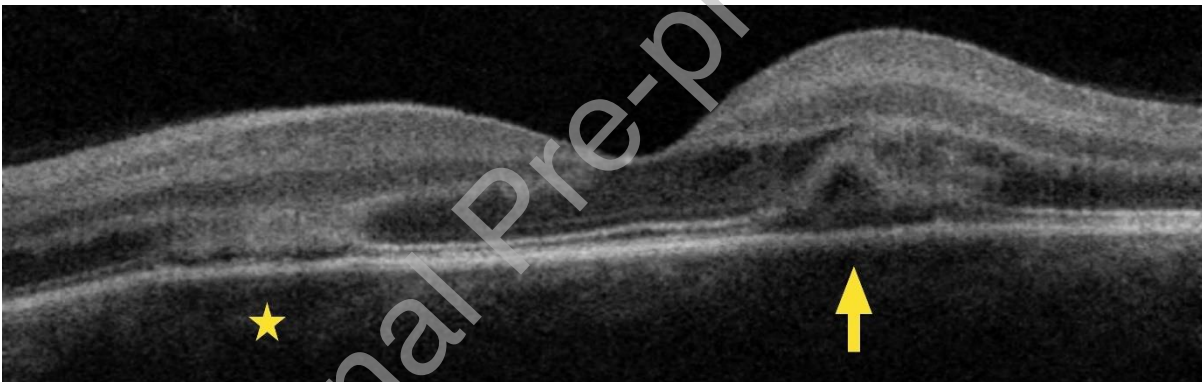


Figure 5: Optical coherence tomography of the left eye (OS) of a patient with APMPE showing a bacillary detachment, especially evident in the more temporal lesion (yellow arrow). The nasal lesion (yellow star) shows a separation between the ellipsoid zone (EZ) and the retinal pigment epithelium (RPE) with mild subretinal fluid consistent with the “double-layer sign” described as well in a case of serpiginous-like choroiditis.²⁷

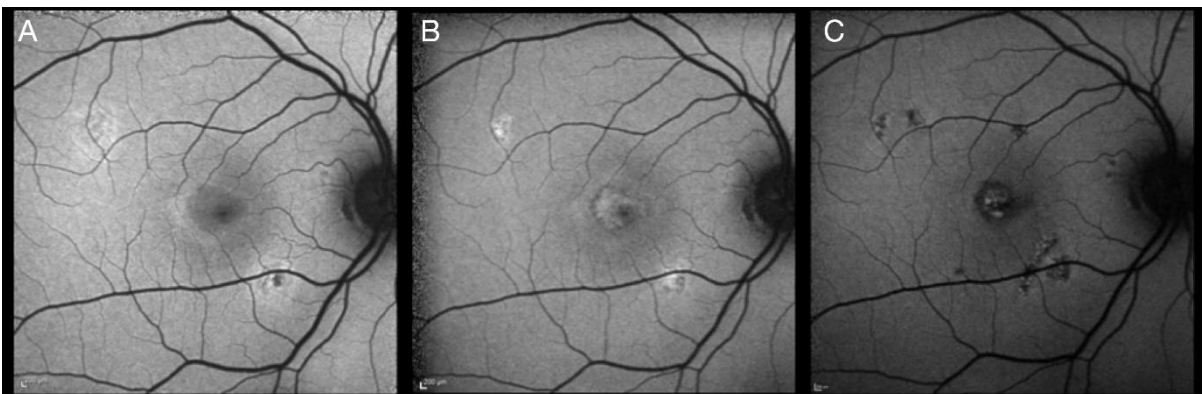


Figure 6: Fundus autofluorescence (FAF) of the right eye (OD) of a patient with APMPE showing the evolution of foveal lesion from early hypo-autofluorescence

(A) to progressive hyper-autofluorescence (B), followed by later hypo-autofluorescence (C). Also note lesions at different stages of activity in the same eye.

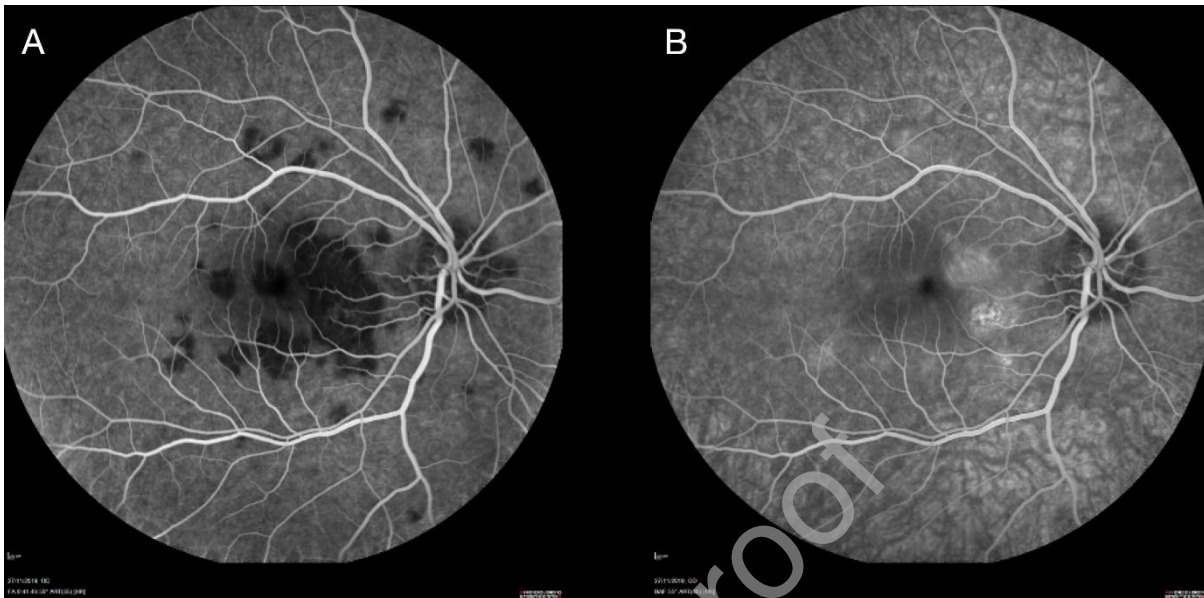


Figure 7: Fluorescein angiography of the right eye (OD) of a patient with APMPPE showing the characteristic pattern of early hypofluorescence (A) and late hyperfluorescence (B). Additionally, it is evident that some lesions are visible in the choroidal phase of FA but no longer visible in the late frames. This underscores the utility of the choroidal phase in detecting APMPPE lesions that may not be apparent in the late stages of angiography alone.

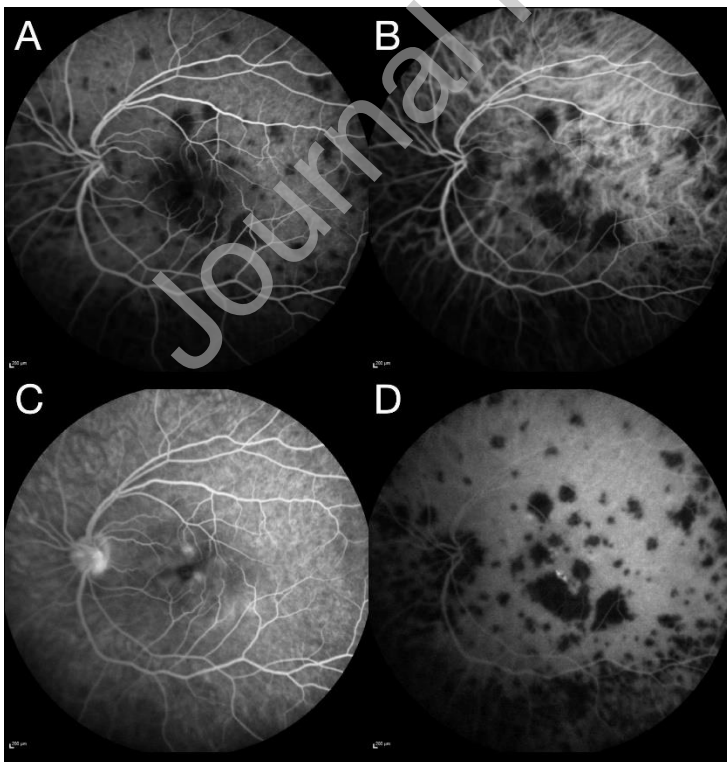


Figure 8: Fluorescein angiography (FA) (A early, C late) and indocyanine green angiography (ICGA) (B early, D late) showing the characteristic early

hypofluorescence and late hyperfluorescence in the FA (A, C) and constant hypofluorescence in the ICGA (B, D). The early frames of FA and ICGA are able to highlight lesions that are not visible in the late frames of the FA (C).

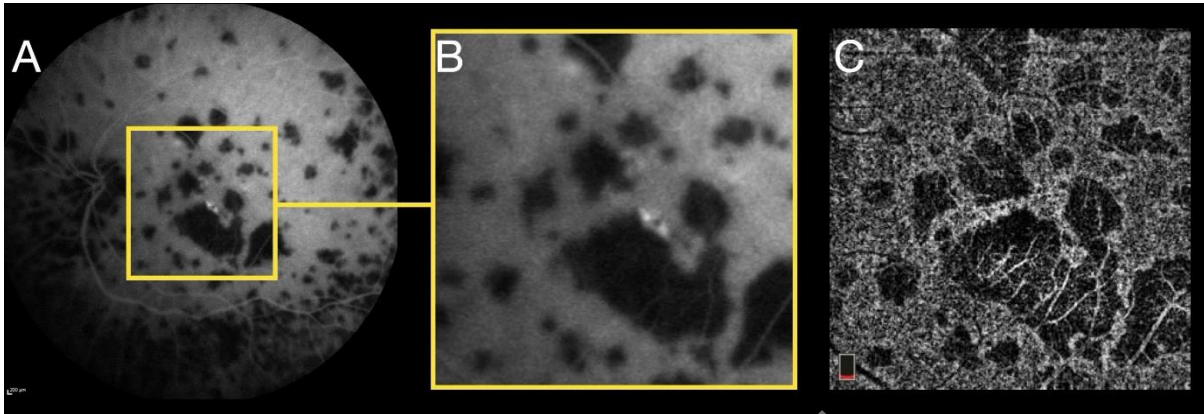


Figure 9: A: Indocyanine green angiography (ICGA) of the left eye (OS) of a patient with APMPE showing the characteristic hypofluorescent lesions. B: Corresponding magnification of the area in the yellow square. C: Optical coherence tomography (OCT) angiography (OCTA) at the level of the choriocapillaris of the same area shows flow deficit in the equivalent areas to the ICGA.

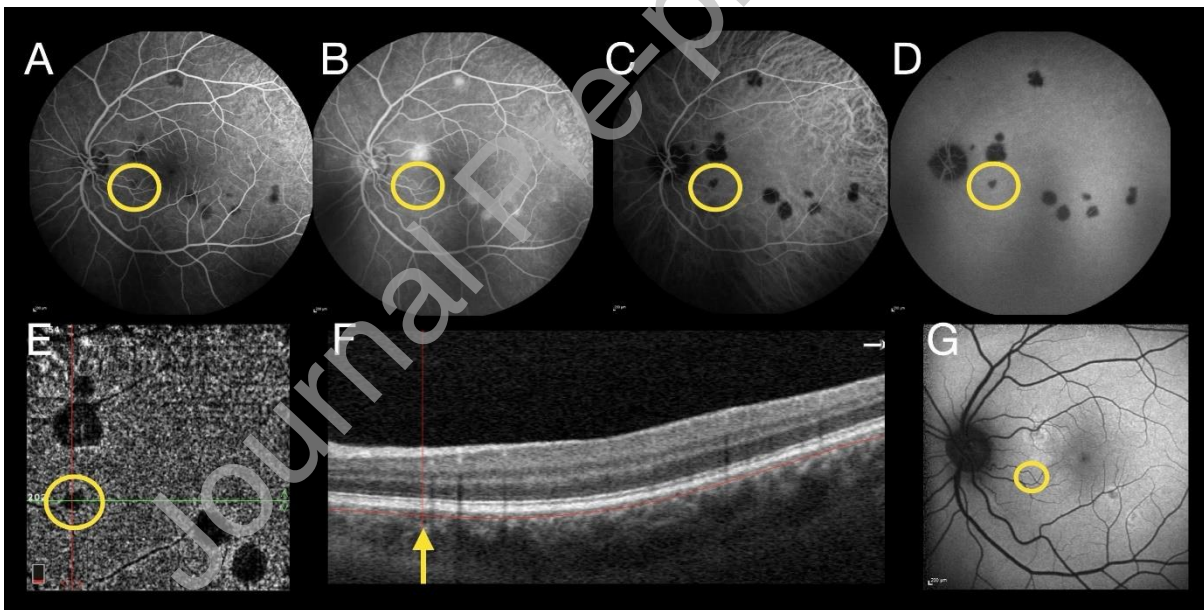


Figure 10: Left eye of a patient with APMPE. The yellow circles highlight the same lesion only visible in the early frames of fluorescein angiography (FA) (A), indocyanine green angiography (ICGA) (C early, D late), and optical coherence tomography (OCT) angiography (OCTA) (E). Whereas it is not seen in the late frames of FA (B), OCT (F, the yellow arrow points to the location of the lesion), or fundus autofluorescence (G).

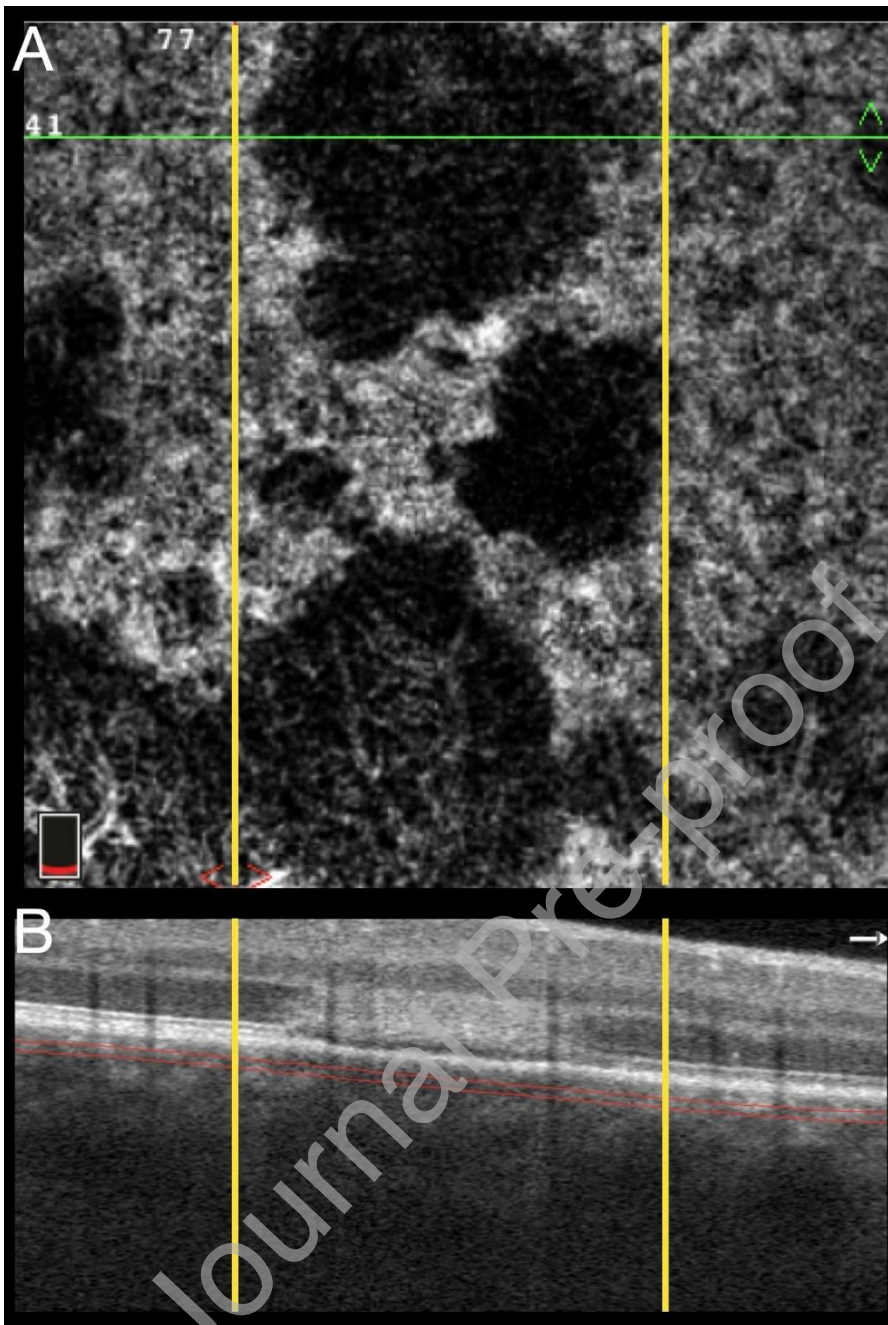


Figure 11: Optical coherence tomography (OCT) angiography (OCTA) at the level of the choriocapillaris disclosing areas of flow deficit (A), the yellow lines mark the borders of the area of flow deficit at the point of the green line that points the location of the structural OCT (B), showing that the area of flow deficit is greater than the area of underlying hyperreflectivity.

Tables

Table 1: Imaging-based Consensus Criteria for Acute Posterior Multifocal Placoid Pigment Epitheliopathy (active stage)

Imaging Modality	Consensus Criteria
Color fundus photography	Paucifocal/multifocal deep yellowish subretinal lesions with ill-defined borders (100 to 2000 μm) Lesions mainly located in the posterior pole
Optical coherence tomography	Active lesions have hyper-reflectivity of the EZ, ELM and ONL Subretinal fluid or bacillary layer detachment can be associated with active lesions
Fundus autofluorescence	Active lesions have initial hypo-autofluorescence with a gradual increase in hyper-autofluorescence
Fluorescein angiography	Hypofluorescence in the early phase followed by late diffuse hyperfluorescence
Indocyanine green angiography	Early frames show hypofluorescence of the lesions, which persists in the late frames
Optical coherence tomography angiography	Flow deficit areas in the choriocapillaris slab at active lesion location

Table 2. Stages of Healing in Acute Posterior Multifocal Placoid Pigment Epitheliopathy based on Multimodal Imaging

Stage	Imaging Findings
1: Choroidal/ Pre-acute	Normal OCT and FAF Choriocapillaris hypoperfusion on FFA (early) and ICGA Choriocapillaris flow deficit on OCTA Lesions can resolve without changes on OCT, OCTA and FAF
2: Chorioretinal/ Acute	Disruption and hyper-reflectivity of EZ, ELM and ONL on OCT Early classic hypofluorescence and late diffuse hyperfluorescence on FFA Persistent hypofluorescence on ICGA Predominantly hypo-autofluorescent lesions on FAF Choriocapillaris flow deficit on OCTA
3: Transitional/ Post-Acute	Disruption and thinning of EZ, ELM and ONL with RPE changes (thickening and/or thinning) Progressive central hyper-autofluorescence on FAF Persistent choriocapillaris flow deficit on OCTA
4: Resolution	Thinning of the outer retina and hypo-reflectivity of the RPE on OCT Predominantly hypo-autofluorescent lesions on FAF Normalized choriocapillaris on OCTA

Table 3. Consensus-based Guidelines for Imaging in Acute Posterior Multifocal Placoid Pigment Epitheliopathy ^a

No.	Guidelines	Strength of Consensus
1.	CFP to document the lesion morphology, size, number and laterality in APMPE (<i>strong consensus</i>)	97.4%
2.	OCT through the lesions of APMPE to determine the disease activity (<i>unanimous consensus</i>)	100%
3.	Serial OCT through the lesions to determine the different phases of healing in APMPE (<i>unanimous consensus</i>)	100%
4.	FAF to assess lesion activity and demonstrate lesions in different stages within the same eye (<i>strong consensus</i>)	97.4%
5.	FFA to assess the lesions and help differentiate APMPE from SC (<i>consensus</i>)	82.1%
6.	ICGA or OCTA to assess the choriocapillaris perfusion and determine the extent of the lesions (<i>strong consensus</i>)	97.4%
7.	Ultra-wide field FFA and/or ICGA for evaluating the peripheral lesions (<i>consensus</i>)	92.3%
8.	OCTA to assess the choriocapillaris perfusion, extent of the lesions and detecting complications such as CNV (<i>consensus</i>)	84.6%

^a The seven members of the expert subcommittee did not cast their votes.

APMPPE: Acute posterior multifocal placoid pigment epitheliopathy; CFP: color fundus photography; CNV: choroidal neovascularization; FFA: fluorescein angiography; FAF: fundus autofluorescence; ICGA: indocyanine green angiography; OCT: optical coherence tomography; OCTA: optical coherence tomography angiography; SC: serpiginous choroiditis

Table of Contents Statement

This report provides expert consensus guidelines for the diagnosis, classification, and monitoring of acute posterior multifocal placoid pigment epitheliopathy. The guidelines emphasize the importance of advanced retinal imaging techniques, including color fundus photography, fluorescein angiography, indocyanine green angiography, fundus autofluorescence, and optical coherence tomography. These imaging modalities enhance diagnostic accuracy and enable the identification of disease activity and complications. The recommendations aim to standardize clinical practice and improve patient management by distinguishing this condition from other similar chorioretinal disorders.

Declaration of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests:

Ester Carreno reports a relationship with Roche that includes: speaking and lecture fees. Rupesh Agrawal reports a relationship with National Medical Research Center that includes: funding grants. Rupesh Agrawal reports a relationship with Council for Scientific and Industrial Research that includes: funding grants. Douglas A. Jabs reports a relationship with AbbVie Inc that includes: funding grants. If there are other authors, they declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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