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RESEARCH

The effect of acute zonular occult outer retinopathy on retinal vessel oxygen saturation

Shahad Zaid Salim Al-Hamdani[1] · Belal Al Tweish[1] · Annekatrin Rickmann[2] · Berthold Seitz[3] · Hendrik PN Scholl[4,5,6] · Nóra Szentmáry[7,8] · Fabian N Fries[3] · Maria della Volpe Waizel[1]

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Abstract

Purpose To measure the retinal vessel oxygen saturation alteration with retinal oximetry (RO) in patients with acute zonular occult outer retinopathy (AZOOR) and compare these findings with retinitis pigmentosa (RP) and controls.

Methods Prospective observational study examining 189 eyes of 98 patients: 12 eyes of 6 AZOOR patients (1♂5♀; 48.5 ± 19.2 years (mean ± SD) were compared to 77 eyes from 39 patients with RP (15♀ 24♂; 45.6 ± 14.7 years) and 100 eyes from 53 controls (31♀ 22♂; 40.2 ± 13.4 years). Main outcome measures were the mean arterial (A-SO_2; %), venular (V-SO_2; %) oxygen saturation, as well as their difference (A-V SO_2; %) measured with the oxygen saturation tool of the Retinal Vessel Analyzer (IMEDOS Systems UG, Germany).

Results Eyes affected by AZOOR differed significantly from both RP and control eyes in terms of retinal oxygen metabolic parameters. Compared to controls, AZOOR eyes showed significantly higher A-SO_2 and V-SO_2 values ($p = 0.031$ and $p = 0.044$, respectively), while V-SO_2 was significantly lower in AZOOR compared to RP eyes ($p = 0.035$). A–V SO_2, which reflects retinal oxygen consumption, was significantly reduced only in RP eyes, but not in AZOOR eyes, when compared to controls.

Conclusion Assessments of retinal oxygen metabolism might contribute to a correct diagnosis and a better understanding of the pathogenesis of AZOOR and RP.

Key Messages

What is known

- Retinal oximetry (RO) is a useful tool for (differential) diagnosis of retinal diseases.
- Acute zonular occult outer retinopathy (AZOOR) often presents clinically as a phenocopy of retinitis pigmentosa (RP).

What is new

- RO function in AZOOR shows a different pattern when compared to RP, thus offering a fast and efficient way to find the correct clinical diagnosis.
- By offering a deeper understanding of retinal oxygen metabolism, RO may contribute to uncovering the underlying pathophysiological processes in AZOOR.

Keywords acute zonular occult outer retinopathy · retinal oxygen metabolic function · inherited retinal diseases · retinal oximetry · retinitis pigmentosa

Introduction

Retinal oximetry (RO) is a medical Device that provides a non-invasive way to measure oxygen saturation levels in the retinal vessels by capturing the fundus [1]. It is well known that oxygen plays a crucial role in the maintenance of optimal visual function [2–3] and RO facilitates the assessment of in vivo retinal oxygen metabolic alterations [4–5].

Shahad Zaid Salim Al-Hamdani and Belal Al Tweish contributed equally to this work.

Extended author information available on the last page of the article

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Recent studies comparing the retinal vessel oxygen saturation in eyes affected by retinitis pigmentosa (RP) and other inherited retinal diseases (IRDs) with that of healthy controls have demonstrated significantly altered saturation levels in IRD patients [6–16]. These findings suggest that retinal oximetry (RO) could be a valuable tool for (differential) diagnosis and therapy monitoring [6, 7, 16], provided that influencing factors are carefully considered [17]. Consequently, RO offers a range of potential clinical applications, particularly for distinguishing phenocopies of IRDs.

Acute zonal occult outer retinopathy (AZOOR) is an inflammatory retinopathy within the wide range of white dot syndromes and is characterized by photoreceptor functional loss due to cell dysfunction [18–19]. Despite its distinct etiology, the clinical presentation of AZOOR often resembles that of an IRD, making it an important differential diagnosis, particularly in relation to RP [18–19]. AZOOR and RP share key features, including progressive visual field defects and outer retinal dysfunction detectable on electroretinography [18–22]. Both can eventually affect central and peripheral vision, and advanced stages may show similar outer retinal atrophy. AZOOR typically presents with a sudden or subacute onset of paracentral or peripheral field loss, often accompanied by photopsias, while early fundus findings can be subtle. Its course is usually monophasic or limited to a few episodes, with stabilization or partial recovery while RP develops insidiously, beginning with night blindness and slow peripheral field constriction, with characteristic fundus changes and continuous progression [6–16]. Currently neither etiology nor the pathology of the retinal pigment epithelium and photoreceptor epithelium complex malfunction caused by AZOOR is definitively understood [18–22]. Studies have suggested viral infections or an autoimmune etiology due to findings of antiretinal antibodies: Forooghian et al. evaluated 25 AZOOR patients for antiretinal antibodies for which all tested positive on Western blot analysis [21].

While RP and AZOOR are classified as distinct clinical entities, they both share an association with antiretinal antibodies, which are plausibly involved in the pathological changes of the outer retina [22]. Thus, analogous to RP [6–16], an alteration in oxygen metabolic function in patients with AZOOR is a reasonable assumption.

Therefore, this study aims to evaluate retinal vessel oxygen saturation in patients with AZOOR, in comparison to patients with RP and controls, in order to contribute to a better understanding of the underlying pathophysiology in AZOOR.

Methods

This prospective observational double-armed study was performed on a total of 189 eyes of 98 patients. The first report presenting the effect of oximetry on choroideremia in a fourth arm of the study has already been published [16], the present data focusses on the analysis regarding the included patients of the AZOOR arm and includes a shared control group. All study participants were evaluated at a single ophthalmology center (Department of Ophthalmology, University of Basel, Switzerland) and were included between August 2020 and January 2025. The study received approval from the local ethics committee (Ethics Commission of Central and Northern Switzerland, EKNZ, Basel, Switzerland) and was authorized as a prospective observational study (trial number EKNZ BASEC 2020−00122).

Inclusion criteria for all patients and control subjects included Caucasian ethnicity and a refractive error with a spherical equivalent of less than ±6 diopters for both myopia and hyperopia. Participants with ocular (e.g. cataract, diabetic retinopathy, glaucoma, any further retinal disease) or systemic conditions (e.g. hypertension, diabetes mellitus, cardiovascular diseases, respiratory disorders affecting oxygenation, anemia or polycythemia, and smoking) that could potentially affect retinal vessel oximetry measurements were excluded [17]. Additional exclusion criteria included poor-quality fundus oximetry images or a lack of willingness to participate.

All research procedures adhered to institutional guidelines and the principles of the Declaration of Helsinki. Written informed consent was obtained from all participants prior to examination. Each subject underwent a comprehensive ophthalmologic assessment, including refraction, best-corrected visual acuity (BCVA) using the ETDRS chart [23], slit-lamp examination, biomicroscopy, and fundoscopy.

All AZOOR and RP patients received comprehensive diagnostic evaluations tailored to their clinical presentation, performed by experienced, fellowship-trained retina specialists (M.d.V.W. and H.P.N.S.). These assessments included, among others, electrophysiological testing, short-wavelength fundus autofluorescence, optical coherence tomography (OCT), visual field testing using semi-automated kinetic perimetry (background illumination 10 cd/m²; V4e, III4e, I4e, III3e isopters assessed with the Octopus 900®), microperimetry, and genetic testing. Control subjects were healthy individuals with no evidence of ocular or systemic disease.

Prior to retinal oximetry measurements, both pupils were maximally dilated using Tropiphen eye drops (a preparation from our institutional pharmacy containing tropicamide 0.5% and phenylephrine 1%). Three drops were administered in each eye at 10-minute intervals.

Retinal vessel oximetry acquisition

Retinal vessel oximetry was performed using a spectrophotometric oximetry system (IMEDOS Systems UG, Jena, Germany) in combination with a fundus camera (FF450, Carl Zeiss Meditec AG, Jena, Germany). Fundus images were captured using a DCC Digital Camera KY-F75 (JVC Inc., Yokohama, Japan) mounted on the Zeiss fundus camera, with a 50-degree field of view. The system's software (VISUALIS; IMEDOS Systems UG) distinguishes oxygenated from deoxygenated hemoglobin by analyzing differences in light absorption at specific wavelengths, thereby determining the oxygen saturation in the retinal vessels being examined. RO imaging is conducted at two wavelengths: the green channel (548 ± 10 nm), which is relatively insensitive to oxygen, and the red channel (610 ± 10 nm), which is oxygen-sensitive [24–25].

A standardized imaging protocol centered on the optic disc was applied, utilizing two concentric rings in the peripapillary region: an inner ring with a radius of 1.0 optic disc diameters and an outer ring with a radius of 1.5 optic disc diameters. The area between these rings defined the region of interest for all measurements. For each eye, four test–retest fundus images were acquired [26]. Only images with optimal quality—defined as red channel illumination below 160 units and green channel illumination above 60 units on the respective scale—were included in the final analysis [27].

During RO analysis, all major arterioles and venules within the measurement zone were manually selected. The mean oxygen saturation in arterioles (A-SO_2) and venules (V-SO_2) were calculated, and their difference (A–V SO_2), representing an index of retinal oxygen metabolic consumption, was determined.

Statistical analysis

The primary outcomes of the study included mean arterial oxygen saturation (A-SO_2; %), mean venous oxygen saturation (V-SO_2; %), the arteriovenous difference (A–V SO_2; %), as well as the average diameters of retinal arterioles (D-A; µm) and venules (D-V; µm). Oximetry measurements were compared across all three groups: AZOOR patients, RP patients, and controls.

Prior to statistical analysis, the normal distribution of all parameters was assessed using histograms and the Shapiro–Wilk test. To compare the subgroups, ANOVA with pairwise comparisons was conducted, and all reported p-values were adjusted using the Bonferroni correction. To rule out potential age-related differences between male and female participants within each group, unpaired t-tests were performed for all parameters.

ANOVA-based linear mixed-effects models were computed using SPSS® (IBM SPSS Statistics®, Version 30.0.0.0 (172), including the Advanced Statistics package). This approach accounts for the inter-eye correlation in the same subject and is appropriate for analyzing repeated measurements. To assess the influence of subgroup membership on oximetry values, the model included eye (left or right), refractive spherical equivalent, and group as fixed factors, with the subject treated as a random factor. Results are reported as means and standard deviations (± SD) for each group, along with the corresponding p-values. A p-value of <0.05 was considered statistically significant.

Results

A total of 189 eyes of 98 patients were examined: 12 eyes of 6 AZOOR patients (1♂5♀; 48.5 ± 19.2 years (mean ± SD) were compared to 77 eyes from 39 patients with RP (15♀ 24♂; 45.6 ± 14.7 years) and 100 eyes from 53 controls (31♀ 22♂; 40.2 ± 13.4 years). BCVA was logMAR 0.61 ± 1.70 in AZOOR patients, logMAR 0.39 ± 0.67 in RP patients, and logMAR 0.00 ± 0.09 in controls. Both disease groups showed a significant functional deterioration compared to controls ($p<0.001$). The clinical characteristics of all AZOOR patients are shown in Supplementary File 1.

Oximetry results

I. Comparison of oxygen saturation values of diseased eyes to controls

In controls, the mean ± SD for A-SO_2 was 92.69 ± 5.95%, V-SO_2 was 53.96 ± 9.25% and A-V SO_2 was 38.73 ± 7.26%. In RP the corresponding values were: A-SO_2 98.73 ± 9.94%, V-SO_2 was 62.06 ± 10.59% and A-V SO_2 was 36.62 ± 8.85%; in AZOOR these measures were: A-SO_2 97.59 ± 8.01%, V-SO_2 54.30 ± 9.10% and A-V SO_2 43.30 ± 10.64%.

In general, the disease subgroups differed from the controls when all oxygen metabolic parameters (A-SO_2, A–V SO_2, and V-SO_2) were considered: in RP, as previously reported, both A-SO_2 and V-SO_2 showed a statistically significant increase ($p<0.001$), while A–V SO_2 exhibited a significant decrease ($p<0.001$; Fig. 1). In contrast, while A-SO_2 and V-SO_2 were also significantly higher in AZOOR patients ($p=0.031$ and $p=0.044$, respectively), A–V SO_2 showed no significant difference compared to controls ($p=0.407$; Fig. 1). The reported results in this section refer all to the comparison with controls.

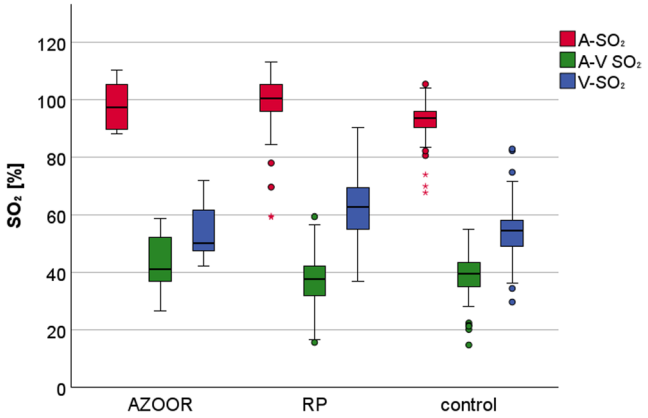


Fig. 1 The box plot displays oxygen saturation levels in the retinal arterioles (red, A-SO_2), venules (blue, V-SO_2), and their difference (green, A–V SO_2). The y-axis represents vessel oxygen saturation percentages across the three groups shown on the x-axis (AZOOR, RP, control). The whiskers indicate the minimum and maximum values, while the box boundaries correspond to the 25th and 75th percentiles. The black horizontal line inside the box denotes the median. Outliers are depicted as individual circles

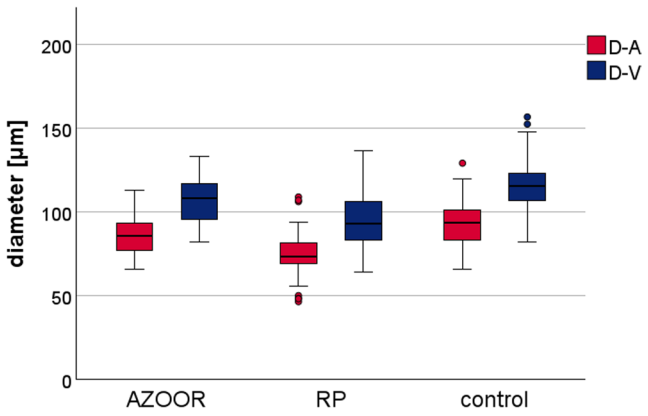


Fig. 2 Box Plot analysis illustrates the retinal vessel diameters of the retinal arterioles (red, D-A) and venules (blue, D-V). The ordinate shows the diameters in micrometers for all three groups shown on the abscissa (AZOOR, RP, control). The upper and lower whiskers of the boxplot present the minimum and maximum value, the upper and lower borders of the box represent the 25th and 75th percentile respectively, and the black horizontal bar within the box represents the median value. Data points classified as outliers are marked as circles

II. Comparison of oxygen saturation values of RP and AZOOR

When comparing the diseased subgroups against each other, the AZOOR subgroup showed similar A-SO_2 ($p=0.236$) and A-V SO_2 ($p=0.115$) values as RP, however, V-SO_2 was still significantly higher in RP than in AZOOR eyes ($p=0.035$). The reported results in this section refer all to the comparison among the diseased eyes groups without comparing to controls.

I. Comparison of retinal vessel diameter values of diseased eyes to controls

In controls, the mean ± SD for D-A was 92.85 ± 12.16 µm and for D-V 116.08 ± 15.40 µm. In RP the respective values were: for D-A 75.03 ± 12.36 µm and for D-V 94.46 ± 15.26 µm; in AZOOR these measures were: for D-A 86.02 ± 13.48 µm and for D-V 106.98 ± 15.57 µm.

In general, the disease subgroups differed from the controls when all retinal vessel diameter parameters (D-A and D-V) were taken into account. In RP, as previously reported, both D-A and D-V showed a statistically significant narrowing of the retinal arterioles and venules ($p<0.001$ for both comparisons; Fig. 2). Similarly, AZOOR eyes showed a significant narrowing of D-A and D-V compared to controls ($p=0.008$ and $p=0.039$, respectively; Fig. 2).

II. Comparison of retinal vessel diameter values of RP and AZOOR

When comparing the disease subgroups with each other, the RP subgroup showed significantly more pronounced narrowing of the retinal arterioles and venules compared to AZOOR, with both D-A and D-V being significantly lower ($p=0.014$ and $p=0.028$, respectively; Fig. 2).

Discussion

Although AZOOR often presents with clinical features similar to those of RP [18–19], our findings reveal significant differences in retinal oxygen metabolism between the two conditions. The exact etiology of AZOOR, as well as the underlying mechanisms affecting the retinal pigment epithelium and photoreceptor complex, remain poorly understood [18–22].

Our study revealed significant alterations in retinal oxygen metabolism in AZOOR patients compared to controls, characterized by elevated arterial and venous hemoglobin oxygen saturation. Despite these changes, oxygen consumption in the AZOOR retina remained within normal limits. In contrast, when compared to the well-characterized IRD RP, different patterns emerged: while arterial oxygen saturation was comparable, RP patients showed significantly higher venous saturation and a marked reduction in oxygen consumption—the latter finding not observed in AZOOR.

Both local retinal oxidative stress and systemic factors appear to contribute to the apoptosis of cones, rods, and inner retinal cells in the pathological progression of RP [28–29]. Multiple studies have highlighted the critical role of oxygen metabolism in the development of RP showing elevated oxygen saturation in both retinal arterioles and venules, along with a decreased arterio-venous oxygen difference which represents the retinal oxygen consumption

and is thought to result from photoreceptor cell loss [6–16]. Additionally, disruption of the blood-retina barrier allows protein-rich fluid and oxygen metabolites from the choriocapillaris to enter the extracellular space, exacerbating its hyperoxygenated state and intensifying oxidative damage [30–31]. These increased extracellular oxygen levels are believed to trigger vasoconstriction, leading to diminished retinal arterial blood flow and disturbed retinal hemodynamics [32]. This mechanism seems to be linked to a neurovascular remodeling [28, 30]: with progression of the dystrophy, the optic disc becomes pale and retinal vessels attenuate [30]. Our study confirms these alterations in retinal vascular oxygenation and the corresponding retinal vessel diameters in RP patients compared to controls.

The pathological changes in AZOOR are thought to be primarily affecting the outer retina, especially involving the photoreceptor layer and the retinal pigment epithelium (RPE). The inflammatory process is thought to target these outer retinal structures, leading to photoreceptor dysfunction and loss. Some studies also suggest involvement of the choroid: Hashimoto et al. assessed choroidal thickness in AZOOR-affected eyes and observed a progressive reduction at multiple locations—subfoveal, nasal, and temporal—over a 3 to 6-month period [33]. Mrejen et al. reported a rather sequential pattern of damage in AZOOR, involving the outer retina first, followed by the RPE and eventually the choroid. Their multimodal imaging approach, including SD-OCT, fundus autofluorescence (FAF), and indocyanine green angiography (ICGA), revealed that RPE and choroidal involvement occurs subsequent to photoreceptor disruption [34]. Our study seems to support the latter hypothesis: higher arterial and venous saturations appear to represent an impaired oxygen consumption in localized areas of the outer retinal photoreceptor layer where the inflammation seems to actually take place, however does not show an extensive pattern as found in RP. If the choroid was a primary focus, analogously to choroideremia [16], we would have found an opposite finding: a lower arterial and venous saturation. The insufficient supply of oxygen from the choroid would have sequentially contributed to the degeneration of photoreceptors and the RPE [16]. However, this seems not to be the case in AZOOR. First, there seem to be outer retinal disturbances that lead to oxygen metabolic dysfunction, followed by the choroidal thinning measured throughout the course of the disease [33].

Compared to controls, we found a significant attenuation of retinal arterioles and venules in AZOOR, however not as pronounced as in RP. The underlying mechanisms are thought to be mainly due to a decreased metabolic demand due to photoreceptor dysfunction or loss [33–35]. As AZOOR primarily affects the outer retina, especially photoreceptors, the retina's oxygen and nutrient demand decreases with progression of the disease and, as a result, retinal blood flow is downregulated [33, 34]. The outer retinal hyperoxia observed in our study, likely resulting from reduced oxygen consumption, appears to contribute to vasoconstriction of both arterioles and venules. This mechanism resembles what has been described in RP. Additionally, in AZOOR, inflammation-related vascular dysregulation may further exacerbate vasoconstriction by impairing endothelial function and disrupting normal autoregulatory processes [33, 34]. Given the ongoing uncertainty surrounding the exact interplay of pathophysiological mechanisms in AZOOR [33–37], RO may become increasingly valuable in detecting early vascular and metabolic alterations in affected patients.

Our study has some limitations with a clinical heterogeneity of RP patients and a small sample size of the AZOOR subgroup. However, our previous work could show that oximetry changes happen early in the course of RP [8]. These significant changes can already be found in children affected by RP, even before neurovascular remodeling takes place [8]. Thus, oximetry values change gradually with more advanced stages, however statistically there is no significant difference between early and late RP stages [15].

Our findings provide the first RO data for AZOOR and demonstrate that RO may be a valuable modality for differentiating IRDs from phenocopies, including AZOOR. In conclusion, our findings provide deeper insight into the pathophysiology of AZOOR and its key differential diagnoses through the use of RO.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s00417-026-07232-1.

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Author contributions MdVW, SSAH and BAT wrote the main manuscript text and MdVW prepared Figs. 1 and 2. All authors reviewed the manuscript and the documents during the revision.

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Data availability The original data are stored in the institutional database and can be provided on request.Trail registry: Ethikkommission Nordwest- und Zentralschweiz (EKNZ) Basel, SwitzerlandTrial registration number: EKNZ BASEC 2020-00122Date of registration: February 11, 2020.

Declarations

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Written informed consent was obtained from all individual participants included in the study.

Competing interests The authors declare no competing interests.

Conflict of interest Maria della Volpe Waizel is a member of the editorial board for Graefes Archive for Clinical and Experimental Ophthalmology, but was not involved in the editorial handling of this manuscript.
Maria della Volpe Waizel is member of the Scientific Advisory Board of backlight therapeutics Switzerland.
Dr. Scholl is chief medical officer of Belite Bio, but this work is performed outside of his activities for Belite Bio and not endorsed by Belite Bio. Dr. Scholl is director of Bioptima AG. He is member of the Scientific Advisory Board of: AIER Hospital Group, Boehringer Ingelheim Pharma GmbH & Co, Janssen Research & Development, LLC (J&J Innovative Medicine), Kerna Ventures, Okuvision GmbH, and Tenpoint Therapeutics. Dr. Scholl is member of the Investment Advisory Board of Droia NV.
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Authors and Affiliations

Shahad Zaid Salim Al-Hamdani[1] · Belal Al Tweish[1] · Annekatrin Rickmann[2] · Berthold Seitz[3] · Hendrik PN Scholl[4,5,6] · Nóra Szentmáry[7,8] · Fabian N Fries[3] · Maria della Volpe Waizel[1]

✉ Maria della Volpe Waizel
maria.waizel@gmail.com

[1] Department of Ophthalmology, University of Basel, Basel, Switzerland

[2] Sulzbach Eye Clinic, Knappschafts Hospital Sulzbach, Sulzbach, Saar, Germany

[3] Department of Ophthalmology, Saarland University Medical Center, Homburg, Saar, Germany

[4] Department of Clinical Pharmacology, Medical University of Vienna, Vienna, Austria

[5] Pallas Kliniken AG, Pallas Klinik Zürich, Zürich, Switzerland

[6] European Vision Institute, Basel, Switzerland

[7] Department of Experimental Ophthalmology, Saarland University, 66424 Homburg, Germany

[8] Department of Ophthalmology, Semmelweis-University, Mária u. 39, Budapest 1085, Hungary