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Gaps in MPO and PR3 ELISA Performance in ANCA-Associated Vasculitis: A Thai Cohort Analysis

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ABSTRACT

Objective: Early and accurate diagnosis of antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis (AAV) is critical to initiating appropriate treatment and for improving patient outcomes. Delayed diagnosis may result in irreversible organ damage, prolonged disease activity and increased mortality. Although, myeloperoxidase (MPO)–ANCA and proteinase 3 (PR3)–ANCA detection by enzyme–linked immunosorbent assay (ELISA) is widely implemented, diagnostic performance may vary across different AAV subtypes and ethnic populations. This study aimed to assess the diagnostic accuracy of MPO–ANCA and PR3–ANCA ELISA in a hospital–based cohort of Thai patients.

Material and Methods: A retrospective analysis was conducted on clinical and serological data from a hospital–based Thai cohort of patients tested for ANCA, using MPO and PR3 ELISA; from January 2019 until August 2024. Sensitivity and specificity were calculated using confirmed AAV diagnoses as the reference standard.

Results: Among the 55 confirmed AAV cases, ELISA detected MPO–ANCA or PR3–ANCA in 39 cases (70.9% sensitivity), and showed 95.0% specificity among non–AAV controls. Subtype analysis revealed the highest sensitivity for microscopic polyangiitis (MPA, 76.7%) and granulomatosis with polyangiitis (GPA, 66.7%), but low sensitivity for eosinophilic granulomatosis with polyangiitis (EGPA, <30.0%). MPA was the most common subtype (48.4%), with a median age of 67 years. ELISA failed to detect ANCA in 29.1% of the confirmed AAV cases, most of which were EGPA.

Conclusion: MPO–ANCA and PR3–ANCA ELISA are highly specific; however, they have limited sensitivity, particularly for EGPA. Given the regional variations in AAV subtype prevalence, reliance on ELISA alone may cause underdiagnosis. Complementary testing strategies may improve detection of ELISA–negative samples.

Keywords: ANCA–associated vasculitis; ANCA–negative AAV; Asian population; immunoassay validation; indirect immuno luorescence

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INTRODUCTION

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) represents a group of life-threatening autoimmune small-vessel vasculitis that include granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA)[1-3]. AAV is a rare but serious autoimmune condition, with multisystem involvement. The clinical manifestations vary by subtype, with renal and pulmonary involvement being common and potentially life-threatening. The estimated prevalence ranges from: 4.6 to 42.1 cases per 100,000 population worldwide, and the mortality rate can reach 10%-20% within 1 to 5 years, particularly in cases with delayed diagnosis or severe organ involvement. These figures underscore the importance of early and accurate diagnosis to reduce adverse outcomes[2,4]. Given the heterogeneous and sometimes nonspecific clinical presentations, laboratory detection of ANCA plays a pivotal role in the diagnostic workup of suspected AAV[1,5]. The past decade has witnessed a change in how ANCA testing is approached. Historically, indirect immunofluorescence (IIF) has been used as a screening method, followed by antigen-specific immunoassays targeting proteinase 3 (PR3) and myeloperoxidase (MPO)[5,6]. However, the 2017 international consensus now recommends the use of high-quality immunoassays; such as enzyme-linked immunosorbent assay (ELISA), as the primary screening method, with IIF reserved for selected cases[5,7]. This change reflects the improved standardization and specificity of ELISA[8,9]. Despite these advances, several studies have highlighted the limitations of the current ELISA-based methods. Highly specific, MPO-ANCA and PR3-ANCA immunoassays have variable sensitivities; depending on the AAV subtype, with particularly low detection rates in EGPA and atypical cases[4,10,11]. A substantial proportion of patients with clinically diagnosed AAV, especially in non-Western populations, may remain ELISA negative owing to atypical antigen targets or limited expression of PR3 and MPO antibodies[2,7]. However, a recent systematic review revealed suboptimal pooled sensitivities of 62% for PR3-ANCA in GPA and 58% for MPO-ANCA in MPA, despite high specificities approaching 95%-99%[4]. These data suggest that a substantial proportion of AAV cases may be missed by ELISA alone, underscoring the need for validation in real-world settings. This diagnostic gap raises concerns regarding potential underdiagnosis in routine clinical settings; especially when complementary assays are not used[6,11].

In Southeast Asia, where AAV prevalence data and test performance metrics remain underreported, it is essential to validate the diagnostic accuracy of ELISA under real-world conditions. Differences in disease phenotype, ANCA target specificity, and laboratory infrastructure may influence test utility compared with that observed in Western populations[12,13]. Moreover, most existing performance studies were conducted in highly selected cohorts or reference laboratories, limiting their generalizability to routine hospital settings[8,9].

Therefore, this study aimed to evaluate the diagnostic performance of MPO-ANCA and PR3-ANCA ELISA in a large cohort of patients tested for ANCA in a real-world Southeast Asian tertiary-care setting. We assessed the sensitivity, specificity, and diagnostic accuracy across AAV subtypes and analyzed the characteristics of ELISA-negative cases to identify potential gaps in current testing approaches.

MATERIAL AND METHODS

Study design

This retrospective study evaluated the prevalence of AAV, its subtypes along with the diagnostic performance of ELISA for MPO-ANCA and PR3-ANCA in suspected AAV cases. Data were collected from patients at Songklanagarind Hospital, a tertiary-care center in Southern Thailand, having undergone ANCA testing via ELISA from January 2019 to August 2024. Inclusion criteria were patients that presented clinical features suggestive of ANCA-AAV, or were clinically suspected to have AAV based on clinical evaluation, and for those that underwent laboratory testing for MPO-ANCA and PR3-ANCA using the ELISA method.

Data collection

This study included patients that presented clinical features suggestive of AAV, or were clinically suspected as having AAV based on physician evaluation, and whom underwent laboratory testing for MPO-ANCA and PR3-ANCA using the ELISA method. Clinical and laboratory data were retrospectively collected from suspected AAV cases; including GPA, MPA, and EGPA. Eligible cases being selected based on the availability of both clinical records and ELISA results for MPO-ANCA and PR3-ANCA. AAV diagnosis and subtype classification were performed using the International Classification of Diseases, 10th Revision (ICD-10, 2019 update), developed by the World Health Organization and adapted by the Centers for Medicare and Medicaid Services and National Center for Health Statistics.

ELISA-based autoantibody detection

MPO-ANCA and PR3-ANCA were detected using two separate commercial ELISA kits: Anti-MPO ELISA (IgG), Order No. EA 1211-9601 G, and Anti-PR3-hn-hr ELISA (IgG), Order No. EA 1201-9601-2 G (EUROIMMUN Medizinische Labordiagnostika AG, Lübeck, Germany). These kits are designed for quantitative and semi-quantitative detection of IgG-class autoantibodies in human serum or plasma, based on the indirect ELISA principle. Diluted serum samples (1:101) were incubated in antigen-coated microplate wells, followed by detection using a peroxidase-conjugated anti-human IgG antibody. The reaction was developed with 3,3',5,5'-tetramethylbenzidine and hydrogen peroxide (H_2O_2), then stopped with 0.5 M sulfuric acid. Absorbance was measured at 450 nm, with a reference wavelength of 620-650 nm.

Quantitative results were obtained using a calibration curve derived from three calibrators (2, 20, and 200 RU/mL), with ≥20 RU/mL interpreted as positive. For semi-quantitative analysis, the ratio of sample absorbance to that of calibrator 2 (20 RU/mL) was calculated (ratio=OD_sample/OD_calibrator2); ratios ≥1.0 were considered positive and <1.0 negative, in accordance with the manufacturer's instructions.

All procedures were performed using an automated microplate analyzer (EUROIMMUN Analyzer I-2P, Lübeck, Germany) to ensure precision and minimize operator variability. Internal positive and negative controls were included in each run, and assay precision was monitored by calculating intra- and inter-assay coefficients of variation (% CV). Statistical analyses were conducted using RStudio (RStudio, PBC, Boston, MA, USA).

Statistical analysis

Categorical variables were summarized as frequencies and percentages; whereas, continuous variables; such as age, were reported as medians with interquartile ranges (IQR). Group comparisons were performed using the chi-squared test or Fisher's exact test. Statistical significance was set at a p-value<0.05. The diagnostic performances of MPO-ANCA and PR3-ANCA ELISA were assessed based on sensitivity, specificity, likelihood ratios (positive and negative) and overall accuracy. The sensitivity and specificity of MPO-ANCA and PR3-ANCA ELISA kits were calculated using 2×2 contingency tables, with ICD-10–based diagnoses of AAV (GPA, MPA, and EGPA) serving as the reference standard. Diagnostic codes were obtained from clinical records, and the 2019 update of the ICD-10 classification was used to define true disease status. All statistical analyses were conducted using RStudio version 4.4.1 and Microsoft Excel.

Ethical considerations

Patient data were anonymized using coded identifiers to ensure confidentiality. This study was approved by the Ethics Committee of the Faculty of Medicine, Prince of Songkla University (EC approval number: REC.67-387-5-8).

RESULTS

Prevalence and demographic distribution of ANCA-associated vasculitis and its subtypes

A total of 1,458 patients were tested for ANCA during the study period; 62 (4.3%) were diagnosed with AAV and 1,396 (95.7%) were classified as non-AAV. The median

age of patients with AAV was 62 years (IQR: 48–70 years), which was significantly higher than that of patients without AAV (55 years, IQR: 40–67 years: p-value<0.05). The age group with the highest AAV prevalence was 61–70 years (29.0%) followed by 71–79 years (19.4%). Only 12.8% of AAV cases occurred in patients younger than 40 years, indicating a predominance in older adults (Table 1).

The distribution of the AAV subtypes is presented in Table 2. MPA was the most common subtype, accounting for 48.4% of cases, followed by GPA (29.0%), and EGPA (22.6%). A statistically significant difference in gender distribution was observed across the subtypes (p-valu<0.05). MPA was predominantly observed in female patients, accounting for 37.1% of all AAV cases in females. In contrast, males were more evenly distributed across the GPA and EGPA groups.

There was also a significant variation in the median age across subtypes (p-value<0.05), with patients with MPA being the oldest (median 67 years), followed by EGPA (64.5 years) and GPA (57 years). EGPA was absent in patients under 40; whereas, GPA peaked in the 51–60 years age group, and MPA was most prevalent in the 61–70 years age range. These age-related trends are illustrated in Figure 1, which shows the distribution of AAV subtypes across the age groups. MPA dominated among older adults; whereas, GPA was observed primarily in individuals over 50, with some cases occurring in younger patients. In contrast, EGPA was not identified in individuals aged<40 years, suggesting a predilection for older age groups.

ELISA diagnostic performance in AAV: subtypes and overall accuracy

The diagnostic performance of MPO-ANCA and PR3-ANCA ELISA varied across AAV subtypes (Table 3). In GPA, PR3-ANCA demonstrated moderate sensitivity (72.0%; 95% confidence interval [CI], 47.0–90.0%) and high specificity (98.0%); whereas, MPO-ANCA was not detected. For MPA, MPO-ANCA showed a high sensitivity (83.0%, 95% CI: 65.0–94.0%); whereas, PR3-ANCA had a low sensitivity (7.0%, 95% CI: 1.0–24.0%). In EGPA, the MPO-ANCA sensitivity was 31.0% (95% CI: 9.0–61.0%), and PR3-ANCA detected no case. The differing serological profiles across AAV subtypes were demonstrated, particularly the limited detection of EGPA using ELISA.

Table 1 Prevalence and demographic distribution of ANCA-associated vasculitis

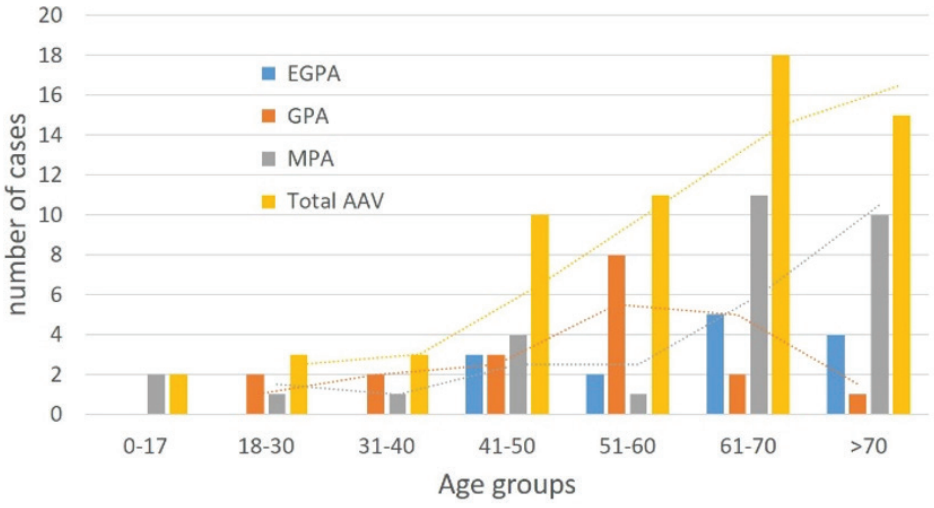
Variable	AAV	non-AAV	Total	p-value
Total n, (%)	62 (4.3)	1,396 (95.7)	1,458 (100.0)	
Gender n, (%)				0.635
Female	36 (58.1)	756 (54.2)	792 (54.3)	
Male	26 (41.9)	640 (45.8)	666 (45.7)	
Age, Median (IQR)	62 (48,70)	55 (40,67)	56 (41,67)	0.013
Age groups n, (%)				0.320
0–17	2 (3.2)	56 (4.0)	58 (4.0)	
18–30	3 (4.8)	135 (9.7)	138 (9.5)	
31–40	3 (4.8)	165 (11.8)	168 (11.5)	
41–50	10 (16.1)	208 (14.9)	218 (14.9)	
51–60	11 (17.7)	285 (20.4)	296 (20.3)	
61–70	18 (29.0)	308 (22.1)	326 (22.4)	
71–79	12 (19.4)	169 (12.1)	181 (12.4)	
>80	3 (4.8)	70 (5.0)	73 (5.0)	

ANCA=antineutrophil cytoplasmic antibody; AAV=ANCA-associated vasculitis; IQR=interquartile range

Table 2 Distribution of AAV subtypes

Variable	GPA	MPA	EGPA	Total	p-value
Total n, (%)	18 (29.0)	30 (48.4)	14 (22.6)	62 (100.0)	
Sex n, (%)					0.0157
Female	7 (11.3)	23 (37.1)	6 (9.7)	36 (58.1)	
Male	11 (17.7)	7 (11.3)	8 (12.9)	26 (41.9)	
Age, Median (IQR)	57 (47,59)	67 (50,72)	64.5 (52,70)	62 (48,70)	0.0325
Age groups n, (%)					0.0142
0–17	0 (0.0)	2 (3.2)	0 (0.0)	2 (3.2)	
18–30	2 (3.2)	1 (1.6)	0 (0.0)	3 (4.8)	
31–40	2 (3.2)	1 (1.6)	0 (0.0)	3 (4.8)	
41–50	3 (4.8)	4 (6.5)	3 (4.8)	10 (16.1)	
51–60	8 (12.9)	1 (1.6)	2 (3.2)	11 (17.7)	
61–70	2 (3.2)	11 (17.7)	5 (8.1)	18 (29.0)	
71–80	1 (1.6)	7 (11.3)	4 (6.5)	12 (19.4)	
>80	0 (0.0)	3 (4.8)	0 (0.0)	3 (4.8)	

AAV=ANCA-associated vasculitis; ANCA=antineutrophil cytoplasmic antibody; GPA=granulomatosis with polyangiitis; MPA=microscopic polyangiitis; EGPA=eosinophilic granulomatosis with polyangiitis; IQR=interquartile range



AAV=ANCA-associated vasculitis; EGPA=eosinophilic granulomatosis with polyangiitis; GPA=granulomatosis with polyangiitis; MPA=microscopic polyangiitis

Figure 1 Age distribution by AAV subtype

When combining the MPO and PR3 results, 55 AAV cases and 1,354 non-AAV cases were analyzed after excluding those with missing data. Nearly one-third of the confirmed patients with AAV (29.1%) tested negative for both markers (Table 4a). A high proportion of ELISA-negative cases was observed in EGPA (72.7%), although

this pattern was also seen in GPA (29.4%) and MPA (11.1%): as shown in Table 4b. Combined marker analysis yielded an overall sensitivity of 70.9%, a specificity of 95.0% and a diagnostic accuracy of 94.0%. The positive likelihood ratio was 14.1, and the negative likelihood ratio was 0.3 (Table 4a). These values indicate strong diagnostic confirmation with positive ELISA results, but limited capacity to exclude AAV when results are negative.

Table 3 Demographic data of the volunteers

AAV subtypes	Analyte	n	Sensitivity, % (95% CI)	Specificity, % (95% CI)
GPA (n=18)	MPO	0	0.0 (0–2)	96.0 (95–97)
	PR3	13	72.0 (47–90)	98.0 (97–99)
MPA (n=30)	MPO	25	83.0 (65–94)	96.0 (95–97)
	PR3	2	7.0 (1–24)	98.0 (97–99)
EGPA (n=14)	MPO	4	31.0 (9–61)	96.0 (95–97)
	PR3	0	0.0 (0–26)	98.0 (97–99)

AAV=ANCA-associated vasculitis; MPO=myeloperoxidase; PR3=proteinase 3; GPA=granulomatosis with polyangiitis; MPA=microscopic polyangiitis; EGPA=eosinophilic granulomatosis with polyangiitis; 95% CI=95% confidence interval

Table 4a Combined MPO/PR3 ELISA results and diagnostic performance for AAV

ELISA result	AAV (n=55, %)	non-AAV (n=1354, %)
MPO neg. with PR3 neg.	16 (29.1%)	1286 (94.9%)
MPO pos. with PR3 neg.	25 (45.5%)	39 (2.9%)
MPO neg. with PR3 pos.	12 (21.8%)	21 (1.6%)
MPO pos. with PR3 pos.	2 (3.6%)	8 (0.6%)
Accuracy	94.0%	
Sensitivity	70.9%	
Specificity	95.0%	
Likelihood ratio+	14.1	
Likelihood ratio-	0.3	

AAV=ANCA-associated vasculitis; ELISA=Enzyme-linked immunosorbent assay; MPO=myeloperoxidase; PR3=proteinase 3; MPA=microscopic polyangiitis; EGPA=eosinophilic granulomatosis with polyangiitis; neg.=negative; pos.=positive; LR+=positive likelihood ratio; LR-=negative likelihood ratio

Table 4b Combined MPO/PR3 ELISA results in confirmed AAV cases

ELISA result	GPA (n=17, %)	MPA (n=27, %)	EGPA (n=11, %)
MPO neg. with PR3 neg.	5 (29.4%)	3 (11.1%)	8 (72.7%)
MPO pos. with PR3 neg.	0 (0.0%)	22 (81.5%)	3 (27.3%)
MPO neg. with PR3 pos.	12 (70.6%)	0 (0.0%)	0 (0.05%)
MPO pos. with PR3 pos.	0 (0.0%)	2 (7.4%)	0 (0.0%)

ELISA=Enzyme-linked immunosorbent assay; MPO=myeloperoxidase; PR3=proteinase 3; GPA=granulomatosis with polyangiitis; MPA=microscopic polyangiitis; EGPA=eosinophilic granulomatosis with polyangiitis; neg.=negative; pos.=positive

DISCUSSION

This study evaluated the diagnostic performance of anti-MPO and anti-PR3 ELISA in a large cohort of patients with suspected AAV. While the ELISA assays demonstrated high specificity, their sensitivity varied across AAV subtypes, particularly showing limited detection in EGPA cases. These findings highlight both the clinical value and limitations of MPO/PR3-targeted immunoassays in the diagnostic workup of AAV.

The subtype-specific performance of ELISA in this cohort aligns with the previously established serological patterns. As expected, PR3-ANCA and MPO-ANCA were useful markers for GPA and MPA,[1,2] respectively; whereas, their sensitivity was markedly lower in EGPA. EGPA demonstrated markedly lower sensitivity for both markers, consistent with its recognized serological heterogeneity and frequent absence of detectable ANCA[10]. Notably, over 70.0% of patients with EGPA in this study were ELISA-negative, reaffirming the limited value of MPO/PR3 immunoassays in this subtype and highlighting the need for broader ANCA-testing approaches.

These findings reinforce the importance of incorporating clinical context and disease subtype into the interpretation of ELISA results. Despite their high specificity, the ELISA-based detection of MPO-ANCA and PR3-ANCA alone might not rule out AAV in seronegative

cases. This limitation is relevant given that international consensus recommendations have shifted toward using antigen-specific immunoassays as the first-line approach for ANCA testing, foregoing indirect IIF in most cases[7,9]. While this strategy enhances specificity and standardization, it may cause underdiagnosis of ELISA-negative AAV, especially in EGPA, and certain atypical presentations; such as ANCA with non-MPO/non-PR3 specificity or isolated extrapulmonary manifestations[4,11].

Recent systematic reviews and multicenter studies have confirmed that antigen-specific immunoassays; including ELISA, outperform IIF in terms of diagnostic accuracy for GPA and MPA[5,8,9]. However, these methods still demonstrate limited sensitivity in detecting AAV cases outside classical serotypes namely, those not associated with PR3-ANCA or MPO-ANCA, especially when atypical ANCA antigens are involved[14]. The potential contribution of IIF in identifying non-MPO/non-PR3 ANCA; such as those targeting lactoferrin, elastase or other neutrophil antigens, remains clinically relevant[6,7].

Demographic trends were also observed in the prevalence of AAV subtypes. MPA was the most common, occurring predominantly in older adults; with a median age of 67 years (IQR, 58-73 years). In contrast, EGPA was absent in patients under 40 years. GPA showed a broader age distribution; including that of younger individuals. These patterns are consistent with those of previous epidemiological studies and underscore the need for age and subtype informed diagnostic interpretations[12,15]. Notably, previous studies have reported regional variations in AAV subtype prevalence, with MPA being more common in Asian populations than in Western cohorts[12,13]. This is consistent with our findings that MPA accounted for 48.4% of AAV cases in the studied population.

Several strengths were identified in this study; including its large and representative cohort, detailed clinical characterization and evaluation of real-world diagnostic performance using a widely available ELISA platform. However, this study had limitations. First, we did not incorporate IIF or alternative ANCA antigens in parallel testing, limiting our ability to characterize ELISA-negative samples. Second, disease controls were not systematically subclassified, which may have affected the specificity estimates. Finally, this study was conducted in a single-center setting, which may have limited the generalizability of our findings.

Given the potentially life-threatening and rapidly progressive nature of AAV, delayed or missed diagnosis can result in irreversible organ damage and increased mortality. Although, MPO-ANCA and PR3-ANCA ELISA remain highly specific and diagnostically valuable for GPA and MPA, their sensitivity is suboptimal in a significant proportion of AAV cases, particularly EGPA. These findings underscore the need for complementary testing strategies; including IIF or expanded antigen panels, especially in clinically suspected ELISA-negative cases. Refinement of diagnostic algorithms through future studies is warranted by incorporating novel ANCA targets and evaluating their utility in diverse clinical settings. Diagnostic approaches should also be adapted to the regional serological landscape to optimize test performance.

This study has certain limitations. The limited number of cases in each AAV subtype, particularly in the EGPA group (n=14), may reduce the statistical precision of performance estimates and affect the generalizability of findings. Future studies with larger and more balanced sample sizes are warranted to validate these results and support subgroup comparisons.

CONCLUSION

High diagnostic specificity but limited sensitivity for detecting AAV, particularly in patients with EGPA, was demonstrated for MPO-ANCA and PR3-ANCA ELISA. It was observed that nearly one-third of confirmed AAV cases were not identified using ELISA alone, emphasizing the importance of clinical correlation and recognition of subtype-specific serological patterns. Incorporating complementary diagnostic strategies may improve early

detection, particularly in ELISA-negative patients. Given the regional differences in AAV subtype prevalence, test selection should be aligned with the serological profile characteristics of the local population. Future studies should investigate extended antigen panels, IIF, and novel biomarkers to enhance diagnostic accuracy in atypical or ELISA-negative cases.

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CONFLICT OF INTEREST

The authors declare no conflicts of interest regarding the contents of this article.

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