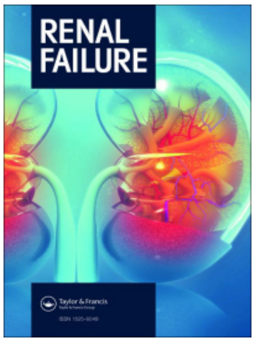

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Validation study of the antineutrophil cytoplasmic antibody (ANCA) kidney risk score in Japanese patients with ANCA-associated glomerulonephritis

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CLINICAL STUDY

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Validation study of the antineutrophil cytoplasmic antibody (ANCA) kidney risk score in Japanese patients with ANCA-associated glomerulonephritis

Ayuko Yamashita[a], Mineaki Kitamura[a], Kumiko Muta[a], Mayu Iwata[a], Emiko Otsuka[a], Kenta Torigoe[a], Tomohisa Uchida[b], Kunihiro Ichinose[c], Takahiro Takazono[d], Noriho Sakamoto[d#], Atsushi Kawakami[b], Hiroshi Mukae[d†] and Tomoya Nishino[a]

[a]Department of Nephrology, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan; [b]Department of Immunology and Rheumatology, Division of Advanced Preventive Medical Sciences, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan; [c]Department of Rheumatology, Shimane University Faculty of Medicine, Izumo, Japan; [d]Department of Respiratory Medicine, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan

ABSTRACT

In 2018, the Antineutrophil Cytoplasmic Antibody (ANCA) Renal Risk Score (ARRS) was proposed as a prognostic tool for renal outcomes in patients with ANCA-associated glomerulonephritis (AAGN). Subsequently, a revised version of the ANCA Kidney Risk Score (AKRiS) was reported in 2024. Nonetheless, its usefulness in Japanese patients has not yet been evaluated. We retrospectively analyzed 109 patients with biopsy-proven AAGN (mean age, 69 years; 50% male) from 13 institutions in Nagasaki Prefecture between January 1995 and December 2019. The prognostic performance of both the ARRS and AKRiS for end-stage kidney disease was assessed using Kaplan–Meier analysis, log-rank test, and receiver operating characteristic (ROC) curve analysis. Myeloperoxidase-ANCA positivity was observed in 90.8% of patients. Based on the AKRiS, 47, 39, 15, and 8 patients were classified as low-, medium-, high-, and very high-risk, respectively. The median observation period was 863 days (interquartile range 254.5–2034). Both the ARRS and AKRiS groups demonstrated progressively worse renal survival with increasing risk categories (both $p < 0.001$). In the ROC analysis, the area under the curve was 0.76 and 0.82 for ARRS and AKRiS, respectively. AKRiS may be a useful prognostic tool for renal outcomes in Japanese patients with AAGN, potentially providing better predictive accuracy compared to the ARRS.

PLAIN LANGUAGE SUMMARY

1. What is known

ARRS and the newer AKRiS are prognostic tools for ANCA-associated glomerulonephritis.

2. What this study adds

This study validates AKRiS in a Japanese cohort and showed good predictive performance.

3. Potential impact

AKRiS may support individualized risk prediction and clinical decision-making in Japanese patients with AAGN.

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ANCA associated glomerulonephritis; ANCA kidney risk score; validation study; end stage kidney disease; risk

Introduction

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a systemic autoimmune disease characterized by necrotizing inflammation of small- to medium-sized blood vessels. The main AAV subtypes include microscopic polyangiitis (MPA), granulomatosis with polyangiitis, and eosinophilic granulomatosis with polyangiitis. Renal involvement is common in AAV, and ANCA-associated glomerulonephritis

CONTACT Mineaki Kitamura minekitamura@nagasaki-u.ac.jp Department of Nephrology, Nagasaki University Graduate School of Biomedical Sciences, 1-7-1 Sakamoto, Nagasaki 852–8501, Japan.
[#]Additional affiliation: Department of Pulmonary Medicine, Graduate School of Medical & Dental Sciences, Kagoshima University, Kagoshima, Japan
[†]Additional affiliation: Nagasaki Harbor Medical Center, Nagasaki, Japan

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(AAGN) frequently leads to rapidly progressive glomerulonephritis. Despite AAV treatment advances, some patients develop end-stage kidney disease (ESKD) [1]. Therefore, predicting the renal prognosis in patients with AAGN is important, and several risk scores have been discussed. Berden et al. [2] proposed the most widely used classification system, which divides patients into four groups based on glomerular lesions as follows: focal, crescentic, mixed, and sclerotic, with renal prognosis reported to worsen in that order. Nevertheless, several meta-analyses have found that only the focal and sclerotic classes are significantly associated with prognosis [3–5]. Sethi et al. [6] reported the Mayo Clinic Chronicity Score, which evaluates factors such as glomerulosclerosis, interstitial fibrosis, tubular atrophy, and arteriosclerosis. Brix et al. [7] developed the ANCA Renal Risk Score (ARRS), which incorporates parameters, including the proportion of normal glomeruli, extent of interstitial fibrosis and tubular atrophy (IF/TA), and the estimated glomerular filtration rate (eGFR) measured at diagnosis. Notably, several validation studies have been conducted on the ARRS, and its usefulness has been suggested in Japanese populations [8–10]. However, Bate et al. [11] proposed the ANCA Kidney Risk Score (AKRiS), an improved ARRS risk score. The ARRS and AKRiS both assess three components: normal glomeruli, IF/TA, and renal function, sharing the approach of evaluating risk by assigning scores to each of these parameters and summing them. The differences are in AKRiS; whereas the quantitative cutoff for IF/TA was retained, a semiquantitative reporting option was also allowed; moreover, renal function was assessed using serum creatinine instead of eGFR. Furthermore, while the ARRS classifies risk into three categories (low, medium, and high), the AKRiS uses a four-tier classification (low, moderate, high, and very high).

The AKRiS has been validated in cohorts from Germany and Mexico, and in both studies, it exhibited comparable or superior utility to that of the ARRS [12,13]. Nonetheless, reports validating this finding in Japanese patients are lacking, and validation in Asian populations has not been reported to date, either. In fact, compared with Western populations, Japanese patients have a higher prevalence of myeloperoxidase (MPO)-ANCA-positive AAV and different patient backgrounds [14]. Because of clinical background variations, whether the AKRiS is useful in the Japanese population remains unclear. We aimed to evaluate the effectiveness of AKRiS in Japanese patients with AAGN.

Methods

Patients

This multicenter retrospective cohort study included patients with AAV diagnosed *via* renal biopsy at Nagasaki University Hospital and its affiliated hospitals between January 1995 and December 2019. The inclusion criteria were as follows: (1) AAV diagnosis based on the Chapel Hill consensus conference definition for AAV [15], (2) renal biopsy consistent with AAGN, and (3) positive ANCA serology. Patients with secondary vasculitis were excluded. Patient characteristics (age, sex, AAV diagnosis, blood tests, urine tests, renal pathological findings, and treatment content) were collected from the medical records. Disease activity was assessed using the Birmingham Vasculitis Activity Score [16].

Ethics

This study was approved by the Institutional Review Board of Nagasaki University Hospital (registration number: 20021012) and adhered to the ethical standards of the 1964 Declaration of Helsinki and its subsequent amendments. Because this study was retrospective, the ethics committees waived informed consent, and patients were informed that they could opt out through their respective centers.

Histological evaluation

Renal biopsy specimens were independently reviewed by two nephrologists with more than 10 years of experience in renal pathology, in accordance with standard pathological criteria. In cases of disagreement, the slides were reviewed jointly, and a consensus was reached through discussion. Renal biopsy specimens were evaluated using multiple serial sections stained with hematoxylin and eosin, periodic acid–Schiff, periodic acid–methenamine silver, and Masson's trichrome. Immunofluorescence studies were performed on frozen sections. Normal glomeruli were defined as those without evidence of sclerosis, crescents, or fibrinoid necrosis within the glomerular tufts. Cellular crescents were identified according to the Renal Pathology Society definition, which included >75% cells and fibrin, with <25% fibrous matrix. The ARRS was determined by combining weighted scores for three pathological and clinical parameters: eGFR at presentation (G0: >15 mL/min/1.73 m^2; G1: ≤15 mL/min/1.73 m^2), the percentage of normal glomeruli (N0: >25%; N1:10–25%; N2: <10%), and the degree of IF/TA (T0: <25%; T1: ≥25%). Each category was assigned a specific point value (G0 = 0, G1 = 3; N0 = 0, N1 = 4, N2 = 6; T0 = 0, T1 = 2), and the total score (range: 0–11) was used to classify patients into the following risk groups: low (0 points), medium (2–7 points), and high (8–11 points) [7]. The AKRiS (ANCA kidney risk score) was determined using the following parameters: creatinine at presentation (K0: <250 µmol/L; K1:250–450 µmol/L; K2: >450 µmol/L), the percentage of normal glomeruli (N0: >25%; N1:10–25%; N2: <10%), and IF/TA (T0: none/mild or <25%; T1: ≥mild to moderate or ≥25%). Each category was assigned a specific point value (K0 = 0, K1 = 4, K2 = 11; N0 = 0, N1 = 4, N2 = 7; T0 = 0, T1 = 3), and the total score (range: 0–21) was used to classify patients into the following risk groups: low (0–4 points), moderate (5–11 points), high (12–18 points), and very high (21 points) [11].

Outcomes

The primary outcome was ESKD development during the observation period, with data censored by death. ESKD was defined as renal replacement therapy for at least 12 weeks. Patients who required dialysis at presentation but discontinued renal replacement therapy within 12 weeks were not classified as having ESKD and were included in the cohort. For each patient, renal survival was calculated from the initial assessment at biopsy to the last follow-up or onset of ESKD.

Statistical analysis

Continuous variables were presented as mean ± standard deviation or median with interquartile range (IQR), depending on their distribution. Categorical variables are presented as counts and percentages. The Wilcoxon rank-sum and Fisher's exact tests compared continuous and categorical variables, respectively. Interobserver agreement for IFTA grading (scores 0–3) between the two nephrologists was assessed using the quadratic weighted kappa coefficient. Renal survival was analyzed using the Kaplan–Meier method, and survival curves were compared using the log-rank test. Kaplan–Meier curves were generated with death treated as censoring events. Competing-risk regression was performed using the Fine–Gray subdistribution hazard model, with death before ESKD treated as a competing event. Subdistribution hazard ratios (sHRs) with 95% confidence intervals were reported. The discriminative ability of the ARRS and AKRiS in predicting ESKD was evaluated using the area under the receiver operating characteristic (ROC) curve, with individual patient-level risk scores analyzed as continuous variables. There were no missing data for any of the variables included in the analyses; therefore, all patients were included without the need for imputation. Calibration was assessed using bootstrap calibration curves with 200 resamples. Sensitivity analyses stratified by treatment era were performed. The interaction between treatment era and AKRiS score was examined using a Cox proportional hazards model. All analyses used a significance threshold of $p < 0.05$. Statistical analyses were performed using JMP® Pro 16 software (SAS Institute, Cary, NC) and EZR (Saitama Medical Center, Jichi Medical University, Japan), a graphical user interface for R.

Results

Baseline clinical feature

The baseline clinical features of the 109 patients are presented in Table 1. The mean age of the patients was 69 years. MPA was diagnosed in 83.5% of cases. As for ANCA type, MPO-ANCA was present in 90.8% of patients. The median eGFR and serum creatinine levels were 24.3 mL/min/1.73 m^2 and 1.9 mg/dL, respectively. At the time of renal biopsy, 9.7% of patients had already initiated dialysis.

Table 1. Patient characteristics.

characteristics	Value ($n = 109$)
Age (year), median (IQR)	69 (62–75)
Male, *n* (%)	55 (50.5)
Hypertension, *n* (%)	45 (41.2)
Diabetes mellitus, *n* (%)	9 (8.3)
Diagnosis	
MPA, *n* (%)	91 (83.5)
GPA, *n* (%)	12 (11.0)
EGPA, *n* (%)	3 (2.8)
unclassifiable, *n* (%)	3 (2.8)
ANCA type	
MPO-ANCA, *n* (%)	99 (90.8)
PR3-ANCA, *n* (%)	6 (5.5)
Double positive, *n* (%)	3 (2.8)
BVAS, median (IQR)	14 (12–18)
eGFR at diagnosis (mL/min/1.73m^2), median (IQR)	24.6 (14.8–38.6)
Cre (mg/dL), median (IQR)	1.9 (1.2–3.0)
CRP (mg/dL), median (IQR)	2.6 (0.3–8.4)
Proteinuria (g/gCr or g/day), median (IQR)	1.1 (0.6–2.6)
RRT at presentation, *n* (%)	9 (8.0)
Treatment	
Steroid pulse, *n* (%)	74 (67.9)
Cyclophosphamide, *n* (%)	22 (20.2)
Rituximab, *n* (%)	1 (0.9)
Plasma pheresis, *n* (%)	15 (13.8)

Variables are shown with median (interquartile range) or number (percentage).
IQR; interquartile range, MPA; microscopic polyangiitis, GPA; granulomatosis with polyangiitis, EGPA; eosinophilic granulomatosis with polyangiitis, ANCA; antineutrophil cytoplasmic antibodies, MPO; myeloperoxidase, PR3; proteinase 3, BVAS; Birmingham Vasculitis Activity Score, eGFR; estimated glomerular filtration rate, Cre; Creatinine, RRT; renal replacement therapy.

Table 2. Histological features and ARRS/AKRiS scores.

Parameters	Value ($n = 109$)
Normal glomeruli (%)	
N0 > 25	37 (33.9)
N1 10–25	24 (22.0)
N2 < 10	48 (44.0)
IFTA (%)	
T0 (none, mild) ≤25	42 (38.5)
T1 (mild–moderate) >25	67 (61.5)
eGFR (mL/min/1.73m^2)	
G0 > 15	82 (75.2)
G1 ≤ 15	27 (24.8)
Creatinine, μmol/L	
K0 < 250	79 (72.5)
K1 250–450	21 (19.3)
K2 > 450	9 (8.3)
ARRS	
ARRS, median (IQR)	6 (2–8)
Low	17 (15.6)
Medium	53 (48.6)
High	39 (35.8)
AKRiS	
AKRiS, median (IQR)	7 (3–10)
Low	47 (43.1)
Moderate	39 (35.8)
High	15 (13.8)
Very high	8 (7.3)

Variables are shown with median (interquartile range) or number (percentage).
IQR; interquartile range, ARRS; ANCA renal risk score, AKRiS; ANCA kidney risk score.

Risk classification based on ARRS and AKRiS

Table 2 presents the pathological features required for evaluating the ARRS and AKRiS, along with each score. The median percentage of normal glomeruli was 12.5%, and moderate-to-severe IF/TA was observed in 61.9% of the patients. The interobserver agreement for IFTA grading between the two nephrologists was moderate (quadratic weighted $\kappa = 0.47$). The ARRS classified more patients into the moderate-to-high risk categories, whereas the AKRiS categorized a greater proportion into the low-to-moderate risk groups. The distribution of AKRiS risk categories did not differ significantly according to clinical phenotype (MPA vs GPA; $p = 0.25$) or ANCA serotype (MPO-ANCA vs PR3-ANCA; $p = 0.77$; Fisher's exact tests). In contrast, clinical diagnosis was significantly associated with ANCA serotype (Fisher's exact test; $p = 0.004$).

ARRS and AKRiS validation

The median follow-up time was 863 days (IQR 254.5–2034). During follow-up, 20 (18.4%) patients progressed to ESKD. The Kaplan–Meier curves for renal outcomes based on the ARRS and AKRiS are displayed in Figure 1. For ARRS (Figure 1A), renal prognosis among the three groups was significantly different ($p < 0.001$). The 3-year renal survival rates were 100% (95% CI, not estimable), 94.0% (95% CI, 77.9–98.6%), and 74.2% (95% CI, 56.2–85.7%) in the low-, moderate-, and high-risk groups, respectively. *Post-hoc* analysis with the Bonferroni correction demonstrated that the severe group exhibited significantly worse renal outcomes compared to the other groups (vs. the mild group, $p < 0.001$; vs. the medium group, $p < 0.001$). No significant differences were noted in renal outcomes between the mild and medium groups ($p = 0.18$). For AKRiS (Figure 1B), the renal prognosis among the four groups was significantly different ($p < 0.001$). The 3-year renal survival rates were 100% (95% CI, not estimable), 88.4% (95% CI, 67.7–96.2%), 76.9% (95% CI, 44.2–91.9%), and 37.5% (95% CI, 8.7–67.4%) in the low-, moderate-, high-, and very high–risk groups, respectively. In the Fine–Gray competing-risk model treating death before ESKD as a competing event, the AKRiS score (continuous) remained significantly associated with ESKD (sHR 1.23 per 1-point increase, 95% CI 1.10–1.37, $p < 0.001$).

ARRS vs. AKRiS: prognostic value comparison

The AUC values were 0.764 (95%CI 0.671–0.858) and 0.822 (95%CI 0.717–0.928) for ARRS and AKRiS, respectively. The ROC curves are presented in Figure 2. Although the difference was not statistically significant, AKRiS demonstrated a tendency toward greater predictive accuracy.

The calibration plot showed reasonable agreement between the predicted and observed 3-year renal survival probabilities (i.e., survival without ESKD) (Supplementary Figure 1). To address potential temporal changes in treatment strategies over the long enrollment period, sensitivity analyses stratified by enrollment era (1995–2007 vs 2008–2019) were performed. Supplementary Table 1 summarizes the induction treatment patterns across eras. The AKRiS discrimination remained similar between the early and late eras (AUC 0.838, 95% CI 0.641–1.000 vs 0.794, 95% CI 0.673–0.916). No significant interaction between AKRiS and treatment era was observed ($p = 0.68$), suggesting that the prognostic performance of AKRiS was broadly consistent across treatment eras.

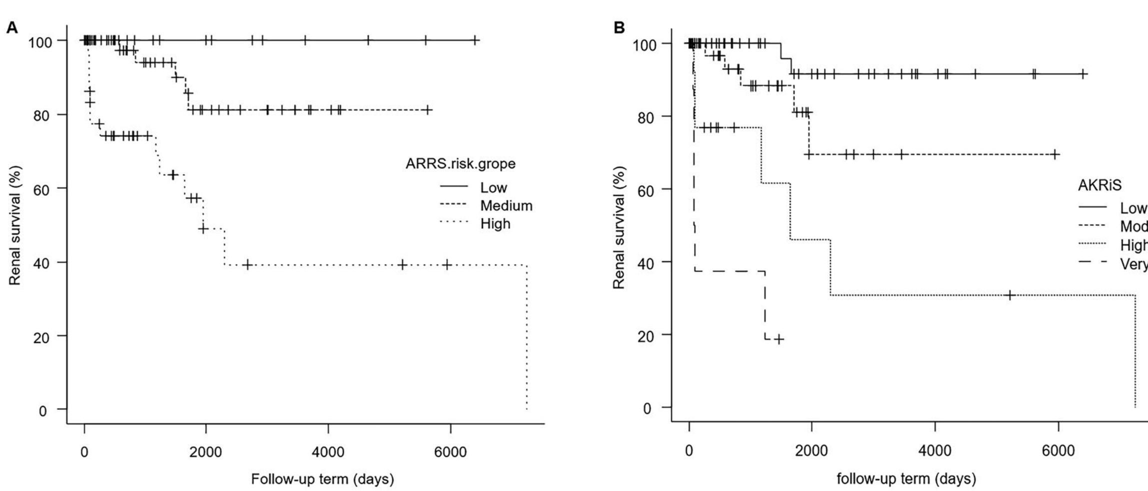

Figure 1. Kaplan–Meier analysis of kidney failure-free survival based on ARRS (A), AKRiS (B).

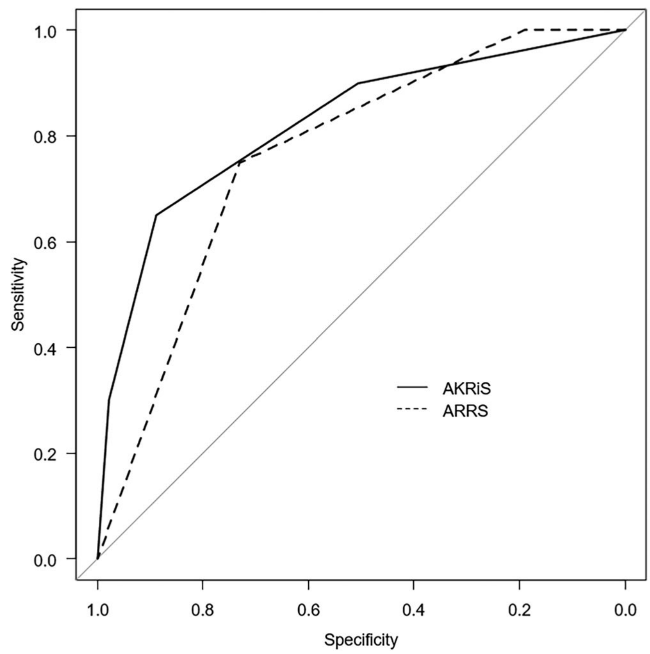

Figure 2. Receiver operating characteristic curves for predicting ESKD using ARRS and AKRiS.

Discussion

This study demonstrated the utility of AKRiS in a Japanese AAV population. Japanese individuals have been reported to exhibit a higher prevalence of MPO-ANCA positivity compared to Western populations [14]. Additionally, research has shown that AAV differs in renal histopathology and prognosis depending on the ANCA subtype. Specifically, MPO-ANCA-positive AAGN tends to demonstrate more chronic lesions, such as interstitial fibrosis, compared to PR3-ANCA–positive AAGN; moreover, it is associated with a poorer renal prognosis [17,18]. Therefore, the applicability of renal prognosis prediction tools developed for Western populations of Japanese individuals must be carefully evaluated.

In a Japanese validation study of the classification proposed by Berden et al. the focal class was consistently associated with renal outcomes. Conversely, the associations with other classes were inconsistent [19–21]. Importantly, the usefulness of the Berden classification in Japanese patients remains controversial: While it assesses risk based solely on glomerular lesions, the ARRS proposed by Brix et al. in 2017 incorporates both the percentage of normal glomeruli and the interstitial changes and renal function into its evaluation [7]. Numerous studies have suggested an association between the number of normal glomeruli and AAV prognosis [22–24]. Notably, the proportion of normal glomeruli is included as an evaluation parameter in the Berden classification. Moreover, the IF/TA ratio has been identified as a factor associated with renal prognosis in AAV [25]. As previously described, IF/TA is more prevalent in MPO-ANCA-positive AAGN, and its assessment may enable evaluation of prognostic differences attributable to ANCA subtypes. Renal function at diagnosis has been reported to correlate with renal prognosis in AAV [26,27]. Furthermore, renal function at diagnosis was poorer in patients with MPO-ANCA-positive AAV than in those with PR3-ANCA-positive AAV [18]. A validation study of the ARRS in a Japanese cohort reported that, although the scoring system was not consistently associated with significant differences in renal prognosis across all risk groups, it demonstrated potential utility [8–10].

Notably, AKRiS is an improved version of the ARRS, with one important modification in risk assessment: the evaluation of renal function using serum creatinine rather than that of eGFR. AAGN often causes rapid declines in renal function, leading to acute kidney injury. It has been reported that eGFR is not a valid measure when serum creatinine is unstable; therefore, in AKRiS, kidney function is assessed using creatinine. Here, the ARRS categorized a greater number of cases as medium to high, whereas the AKRiS classified more cases as low to moderate. This difference may be attributed to the evaluation of serum creatinine levels. Although there was no significant difference in the AUC values, AKRiS was considered as useful as, or possibly more useful than, ARRS in predicting renal prognosis in Japanese patients with AAGN. To date, two validation studies of the AKRiS have been reported. One study conducted in a German cohort of 164 patients with histologically confirmed AAGN demonstrated that AKRiS demonstrated a prognostic performance comparable to that of ARRS in predicting progression to ESKD over 36 months [12]. The second study, published in Rheumatology, evaluated multiple prognostic models, including AKRiS, ARRS, and Berden's histological classification, in a large cohort of patients with ANCA-associated vasculitis involving the kidneys. AKRiS exhibited favorable performance in predicting renal survival, comparable to that of other existing scores. Therefore, these findings support the potential utility of the AKRiS in different patient populations. In both studies, PR3-ANCA-positive patients were more prevalent than MPO-ANCA-positive patients. Conversely, our research population had a higher proportion of MPO-ANCA-positive patients; nonetheless, it showed similar results. Thus, the AKRiS may be applicable to the Japanese population.

This study has some limitations. First, the sample size was relatively small, warranting validation using larger cohorts in future studies. Second, the long observation period from 1995 to 2019 encompassed treatment strategy changes, which may have affected the outcomes. Third, as this was a retrospective observational study, prospective studies are warranted to confirm these findings.

In conclusion, this is the first study to validate the utility of AKRiS for predicting renal prognosis in Japanese patients with AAV. AKRiS may be useful for predicting renal outcomes and potentially offers greater accuracy than the ARRS. Future large-scale validation studies are warranted.

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Author contributions

CRediT: **Ayuko Yamashita**: Conceptualization, Data curation, Formal analysis, Funding acquisition, Investigation, Methodology, Software, Visualization, Writing – original draft; **Mineaki Kitamura**: Conceptualization, Data curation, Formal analysis, Writing – original draft, Writing – review & editing; **Kumiko Muta**: Conceptualization, Data curation, Writing – review & editing; **Mayu Iwata**: Conceptualization, Data curation, Writing – review & editing; **Emiko Otsuka**: Data curation, Writing – review & editing; **Kenta Torigoe**: Data curation, Supervision, Visualization, Writing – review & editing; **Tomohisa Uchida**: Conceptualization, Data curation, Writing – review & editing; **Kunihiro Ichinose**: Conceptualization, Data curation, Supervision, Writing – review & editing; **Takahiro Takazono**: Supervision, Writing – review & editing; **Noriho Sakamoto**: Supervision, Writing – review & editing; **Atsushi Kawakami**: Conceptualization, Data curation, Supervision, Writing – review & editing; **Hiroshi Mukae**: Supervision, Writing – review & editing; **Tomoya Nishino**: Validation, Writing – review & editing.

Disclosure statement

The authors report there are no competing interests to declare.

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ORCID

Mineaki Kitamura http://orcid.org/0000-0002-0003-8645

Kenta Torigoe http://orcid.org/0000-0003-3085-0391

Data availability statement

Due to the need to protect patient privacy, the datasets generated and/or used in this study are not publicly accessible; however, they can be obtained from the corresponding author upon reasonable request.

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