Dec. 5, 2025

Innovations in GN: Trials, Registries, and Digital Pathology

Kidney Biopsy Registry in Japan

NAGOYA UNIVERSITY

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COI Disclosure

Presenter: Shoichi Maruyama

☑ I have completed training on research and medical ethics.

Conflicts of Interest to disclose related to companies over the past 3 years:

- **Executive/Advisory Roles:** ZOME Corporation
- **Stock Ownership/Profits:** ZOME Corporation
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- **Travel/Gifts:** None

1. Background

2. What is J-RBR?

3. Prevalence of Kidney Diseases in Japan

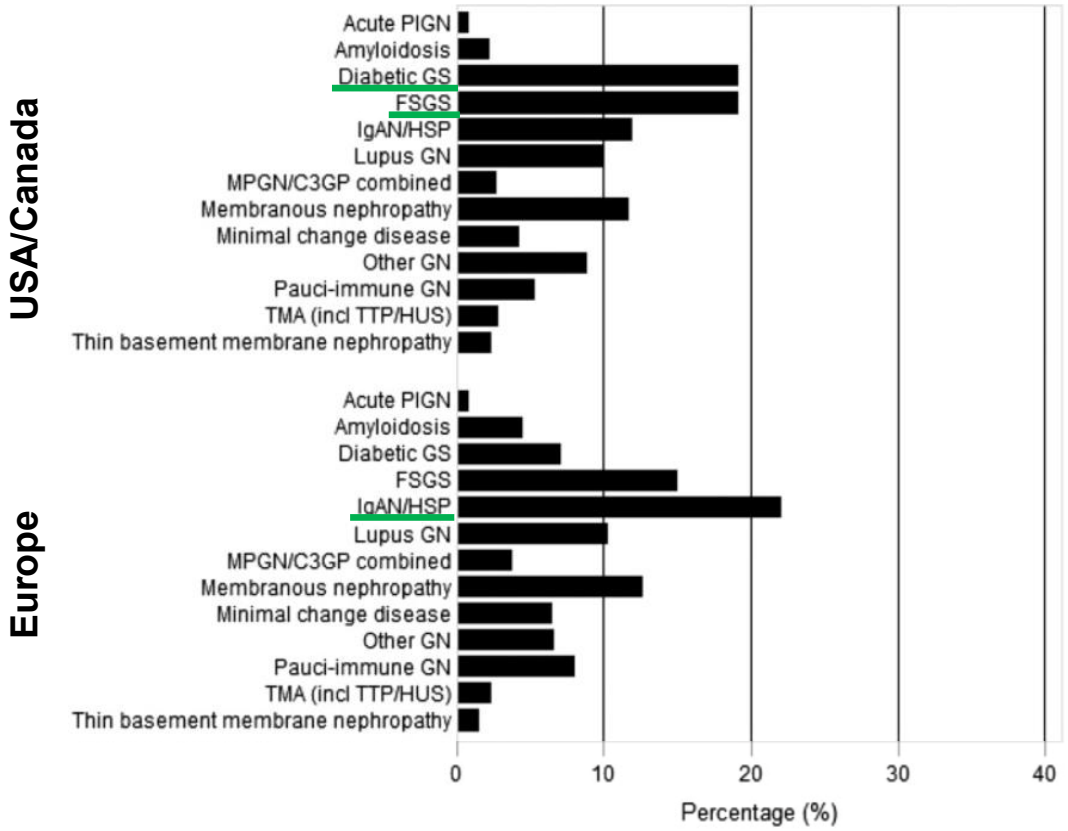
- Analysis of the J-RBR data collected during 2007-2017
- Analysis of the J-RBR data collected during 2018-2024

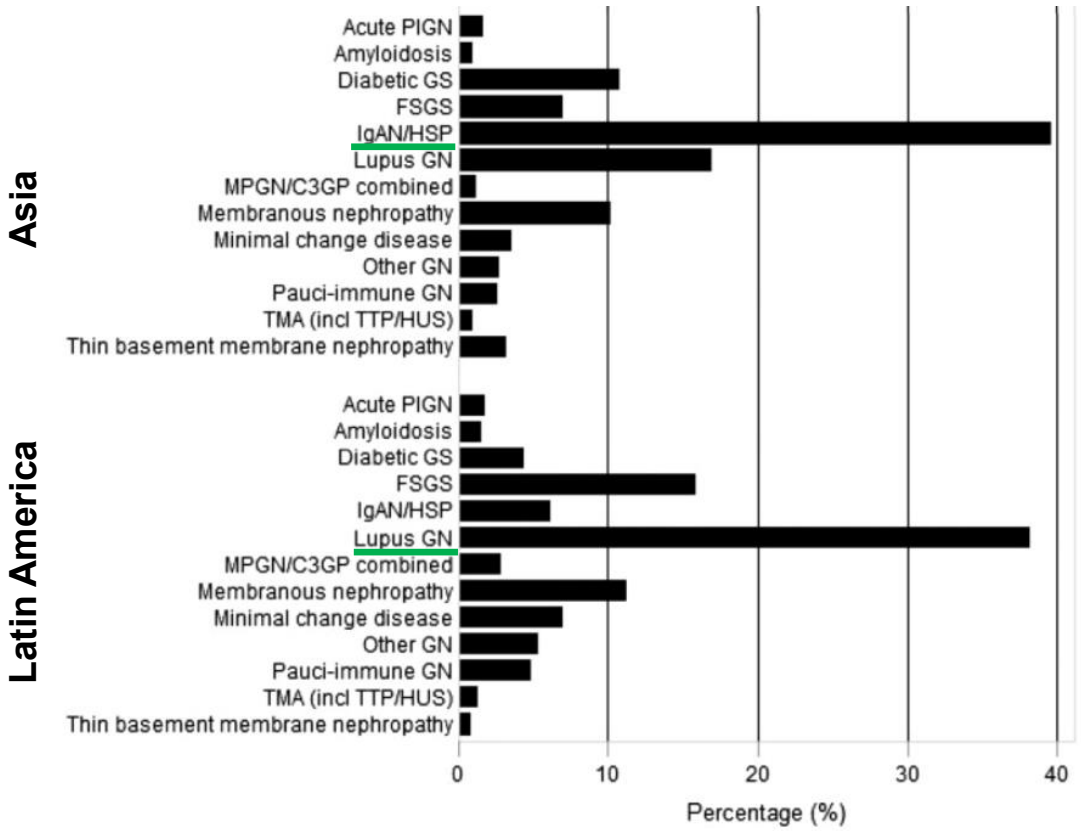
4. Ancillary and Collaborative Studies

Background

The epidemiology of kidney diseases varies widely across countries.
Understanding these demographic patterns—along with differences in healthcare systems and kidney disease management strategies—is essential for advancing our clinical practice and deepening our understanding of renal diseases.

Nephrol Dial Transplant (2017) 1–9





In Europe, IgA nephropathy is the most common, followed by FSGS and membranous nephropathy.
In Latin America, lupus nephritis is the leading diagnosis, whereas in Asia, IgA nephropathy is the most prevalent.

Kidney Biopsy Registry in Japan

ORIGINAL ARTICLE

Clin Exp Nephrol (2011) 15:493–503

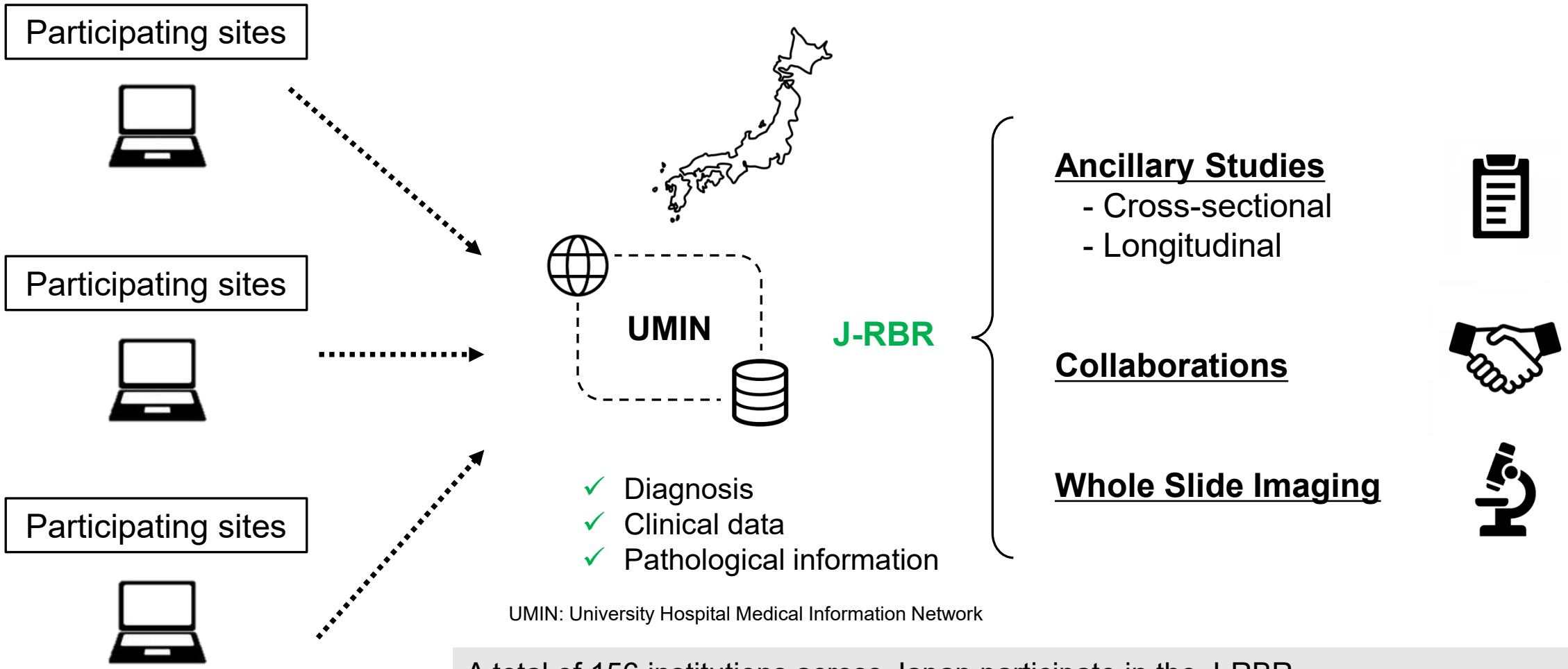
Japan Renal Biopsy Registry: the first nationwide, web-based, and prospective registry system of renal biopsies in Japan

Hitoshi Sugiyama · Hitoshi Yokoyama · Hiroshi Sato · Takao Saito · Yukimasa Kohda · Shinichi Nishi · Kazuhiko Tsuruya · Hideyasu Kiyomoto · Hiroyuki Iida · Tamaki Sasaki · Makoto Higuchi · Motoshi Hattori · Kazumasa Oka · Shoji Kagami · Michio Nagata · Tetsuya Kawamura · Masataka Honda · Yuichiro Fukasawa · Atsushi Fukatsu · Kunio Morozumi · Norishige Yoshikawa · Yukio Yuzawa · Seiichi Matsuo · Yutaka Kiyohara · Kensuke Joh · Takashi Taguchi · Hirofumi Makino · Committee for Standardization of Renal Pathological Diagnosis and Working Group for Renal Biopsy Database, Japanese Society of Nephrology, Tokyo, Japan

- **J-RBR** Japan Renal Biopsy Registry 2007～
  *participants who were performed biopsy
- **J-KDR** Japan Kidney Disease Registry 2009～
  *also includes non-biopsy cases

The Japanese Society of Nephrology (JSN) launched the web-based Renal Biopsy Registry (J-RBR) in 2007. In 2009, JSN established the Japan Kidney Disease Registry (J-KDR) to include cases that do not undergo kidney biopsy, such as polycystic kidney disease.

Kidney Biopsy Registry in Japan



UMIN: University Hospital Medical Information Network

156 sites

A total of 156 institutions across Japan participate in the J-RBR.
It is estimated that this covers about 15% of all kidney biopsy cases in Japan.
Clinical diagnoses, pathological diagnoses, and relevant clinical data are registered.
In principle, only cross-sectional data are entered for each case; however, longitudinal data are additionally collected when needed in ancillary studies.

Overview of the J-RBR (2007-2017, n=32254)

ORIGINAL ARTICLE

Check for updates

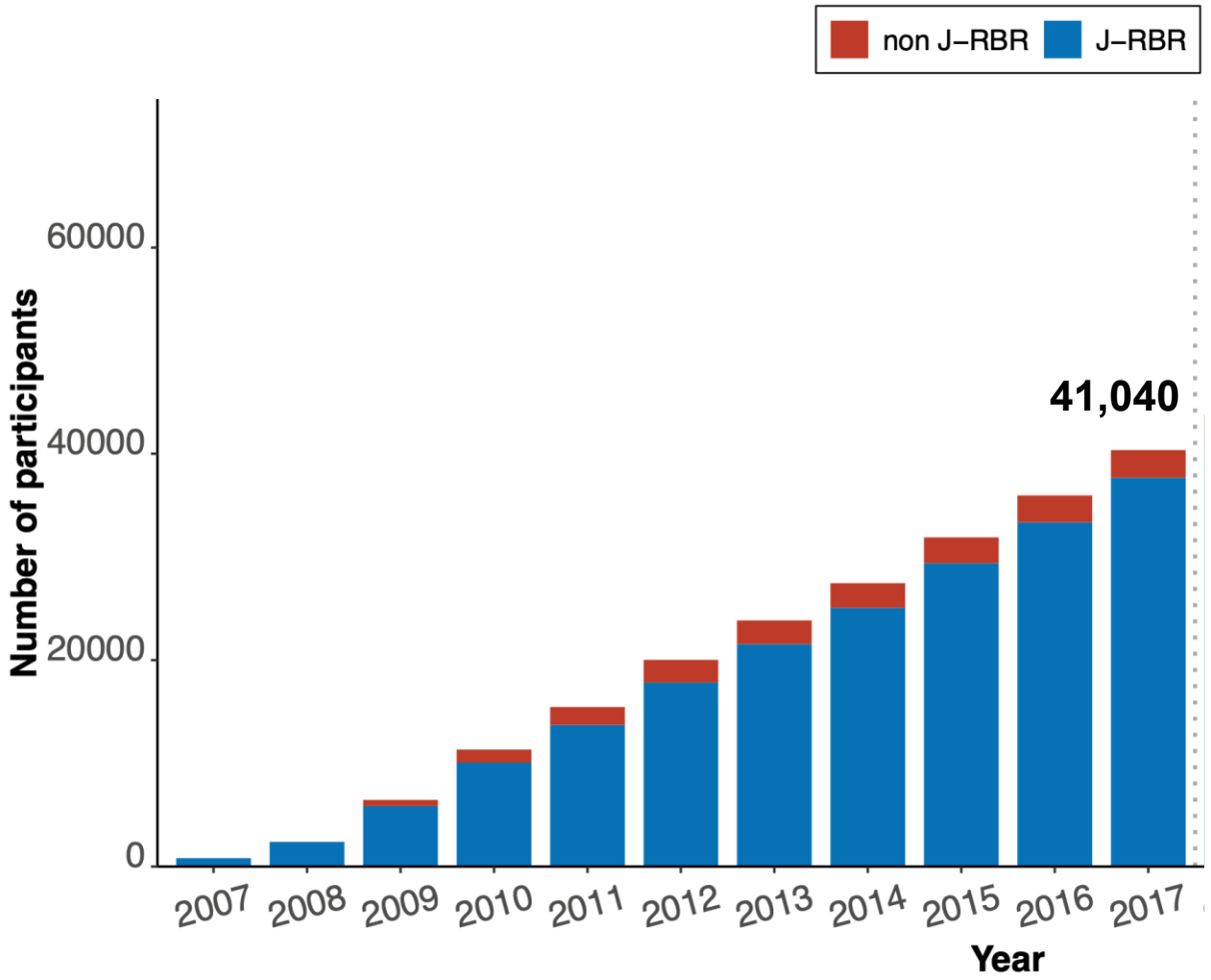
Renal pathology in adult and paediatric population of Japan: review of the Japan renal biopsy registry database from 2007 to 2017

Kazunori Goto[1] · Takahiro Imaizumi[1,2] · Riku Hamada[3] · Kenji Ishikura[4] · Tomoki Kosugi[1] · Ichiei Narita[5] · Hitoshi Sugiyama[6,7] · Akira Shimizu[8] · Hitoshi Yokoyama[9] · Hiroshi Sato[10] · Shoichi Mauryama[1]

Journal of Nephrology (2023) 36:2257–2267

In 2023, we reported a comprehensive analysis of about ten years of registry data, spanning 2007 to 2017.

Trend in the Cumulative Number of Registered Patients in J-RBR/J-KDR (2007–2017)

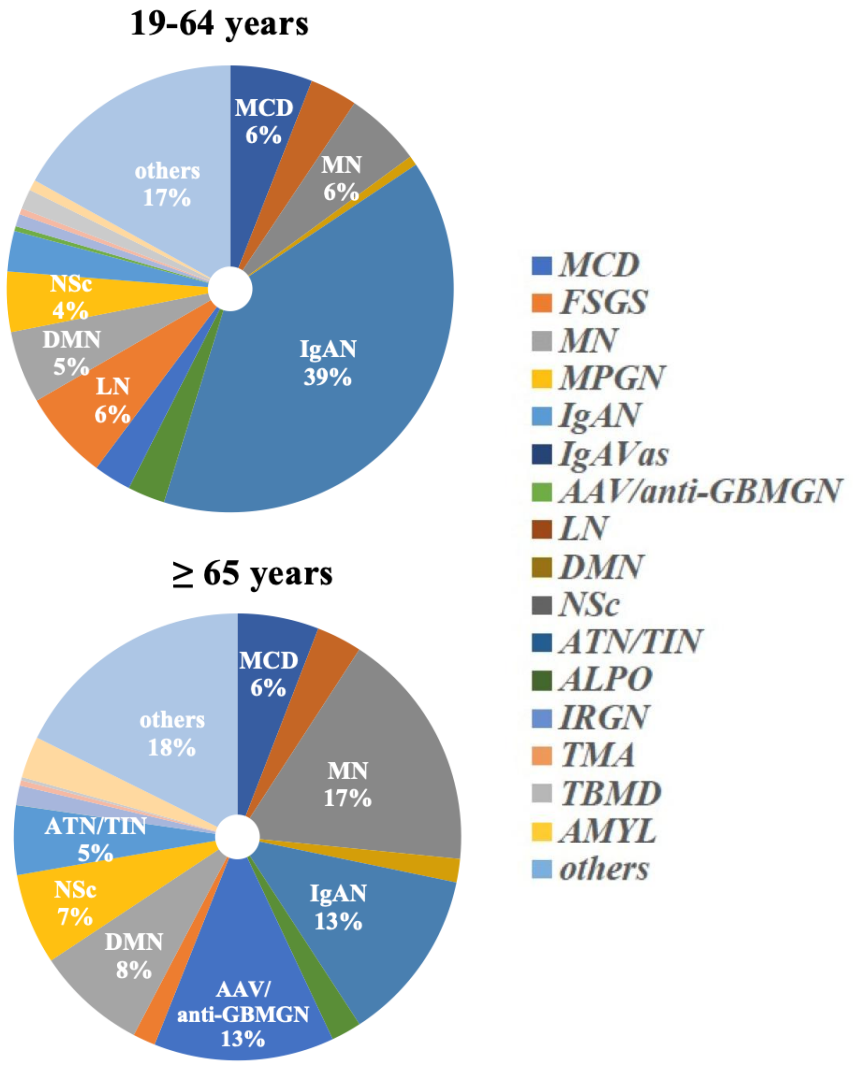


As of the end of 2017: total n = 41,040

Overview of the J-RBR (2007-2017, n=32254)

Main Diagnosis

Diagnosis	Pediatrics (<19 years)	Adults (≥19 years)
MCD	620 (17.6)	1705 (5.9)
FSGS	129 (3.7)	973 (3.4)
Membranous Nephropathy	71 (2.0)	2666 (9.3)
MPGN	55 (1.6)	290 (1.0)
IgA Nephropathy	1273 (36.1)	8838 (30.8)
IgA vasculitis	302 (8.6)	730 (2.5)
AAV/anti-GBMGN	31 (0.9)	1718 (6.0)
Lupus Nephritis	160 (4.5)	1416 (4.9)
Diabetic Nephropathy	2 (0.1)	1754 (6.1)
Nephrosclerosis	7 (0.2)	1453 (5.1)
ATN/TIN	58 (1.6)	1031 (3.6)
Alport syndrome	63 (1.8)	70 (0.2)
Infection-related glomerulonephritis	30 (0.9)	304 (1.1)
TMA	10 (0.3)	120 (0.4)
Thin Basement Membrane Disease	45 (1.3)	305 (1.1)
Renal amyloidosis	2 (0.1)	424 (1.5)
Others	668 (18.9)	4931 (17.2)
Total	3526 (100.0)	28728 (100.0)

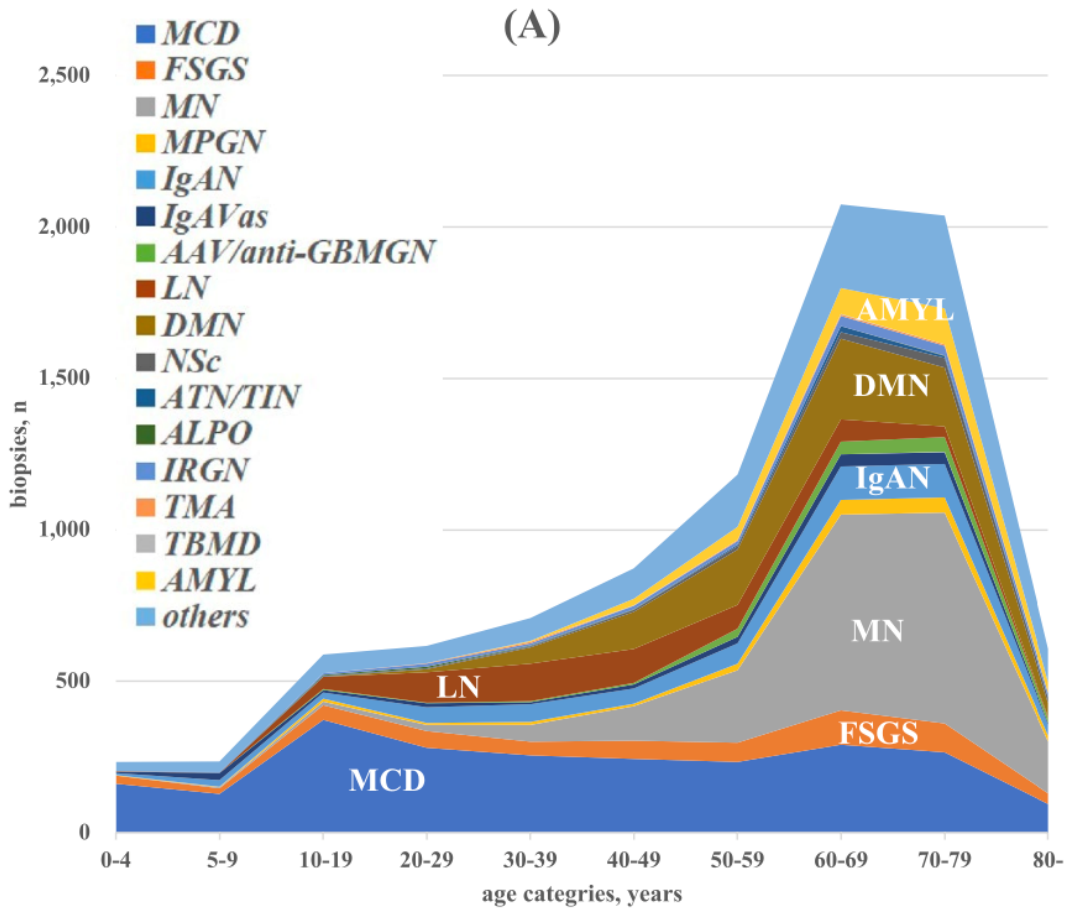


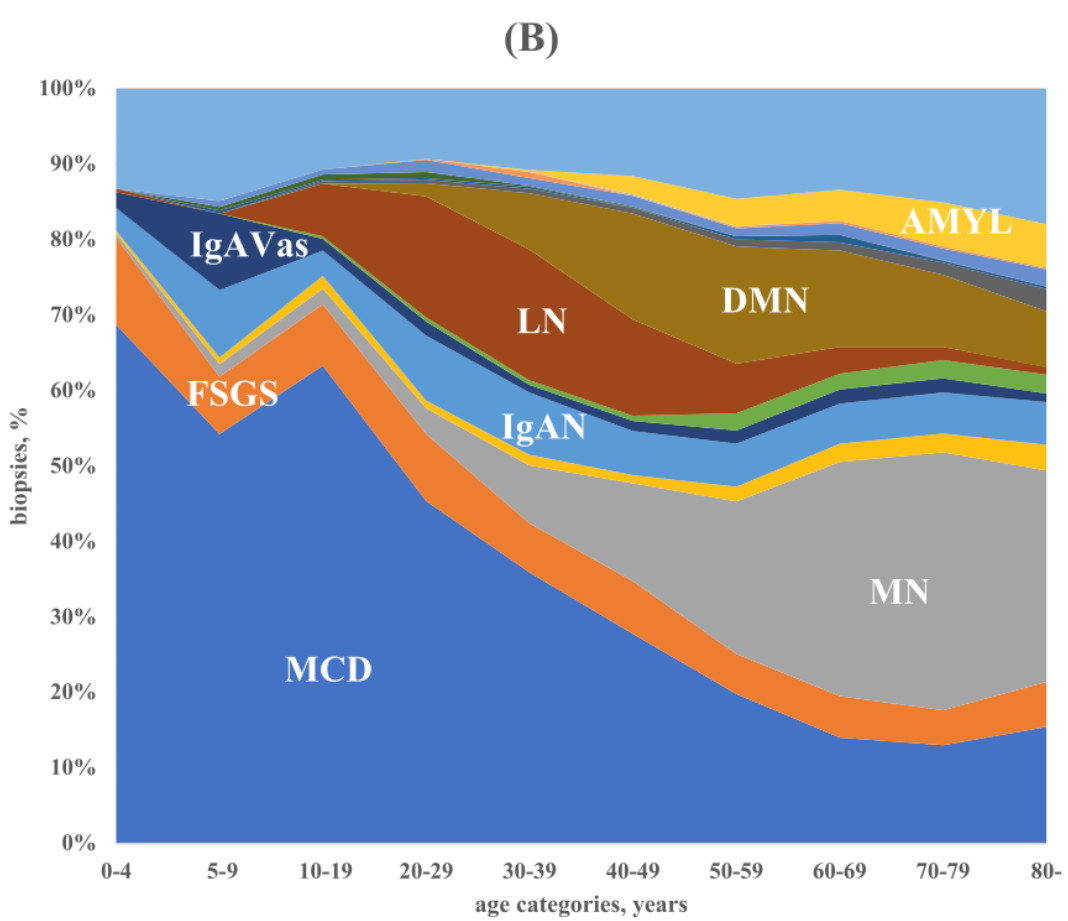
Journal of Nephrology (2023) 36:2257–2267

MCD minimal change disease, *FSGS* focal segmental glomerulosclerosis, *MPGN* membranoproliferative glomerulonephritis, *AAV* antineutrophil cytoplasmic antibody-associated vasculitis, *anti-GBMGN* anti-glomerular basement membrane glomerulonephritis, *ATN* acute tubular necrosis, *TIN* tubulointerstitial nephritis, *TMA* thrombotic microangiopathy

Overview of the J-RBR (2007-2017, n=32254)

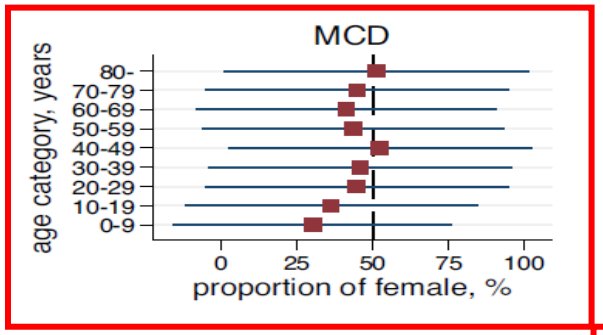
Nephrotic Syndrome in different Age-groups

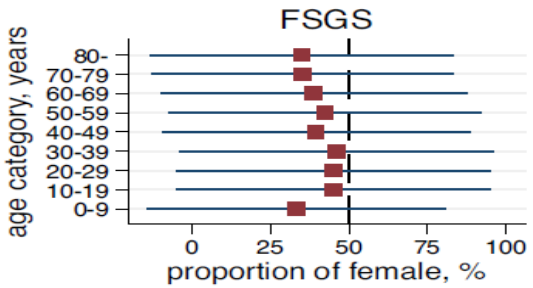


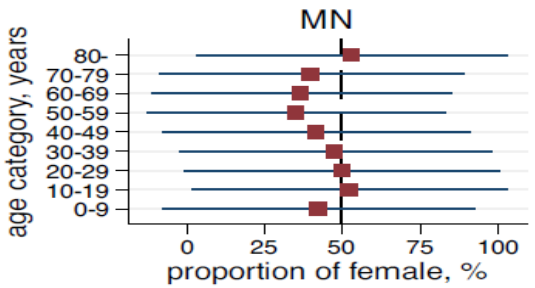


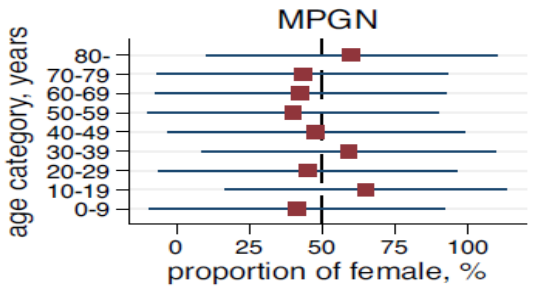
Journal of Nephrology (2023) 36:2257–2267

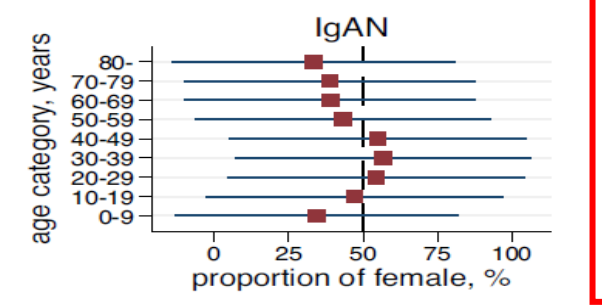
The proportion of females for each diagnosis

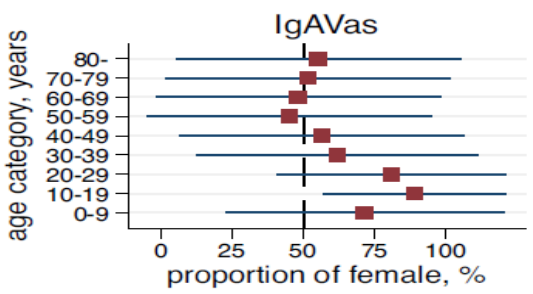


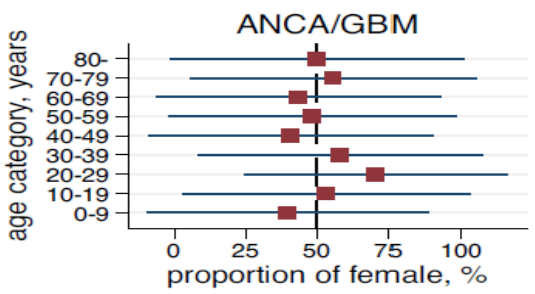


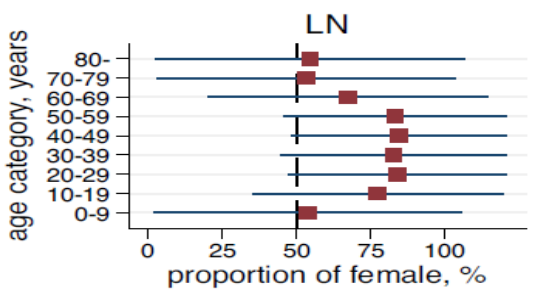


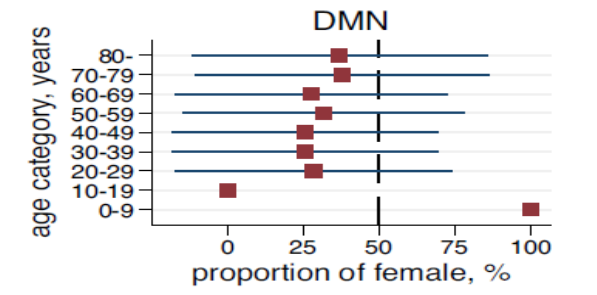


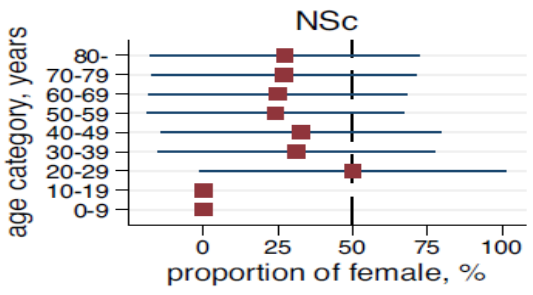


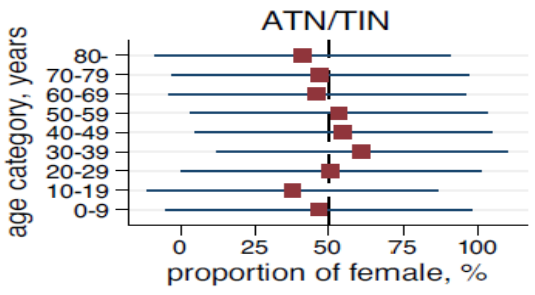


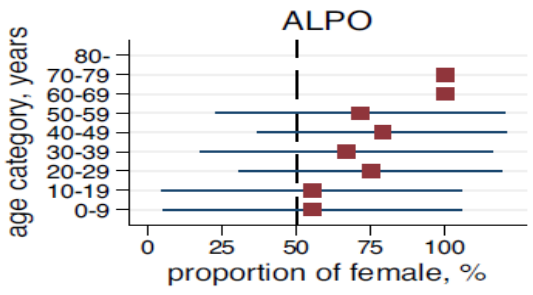


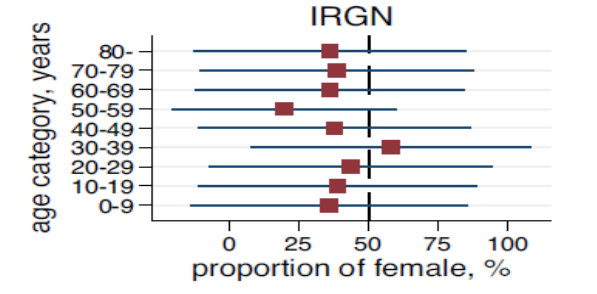


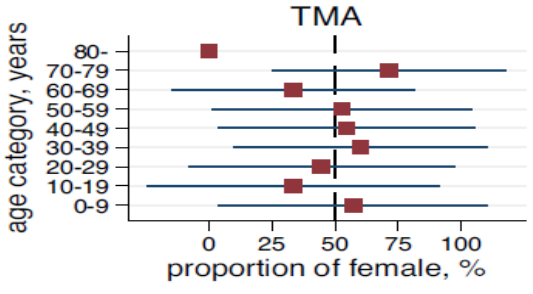


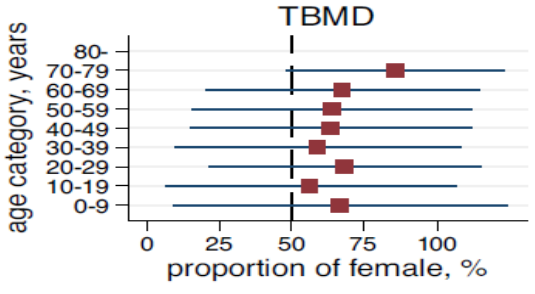


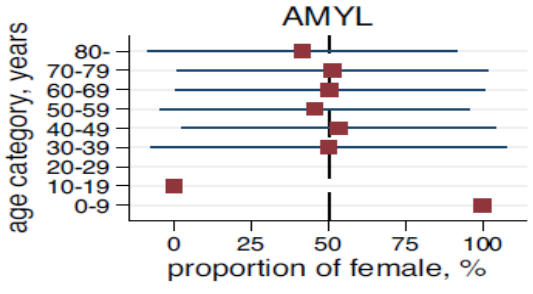












J-RBR Registration Form (2018-)

ORIGINAL ARTICLE

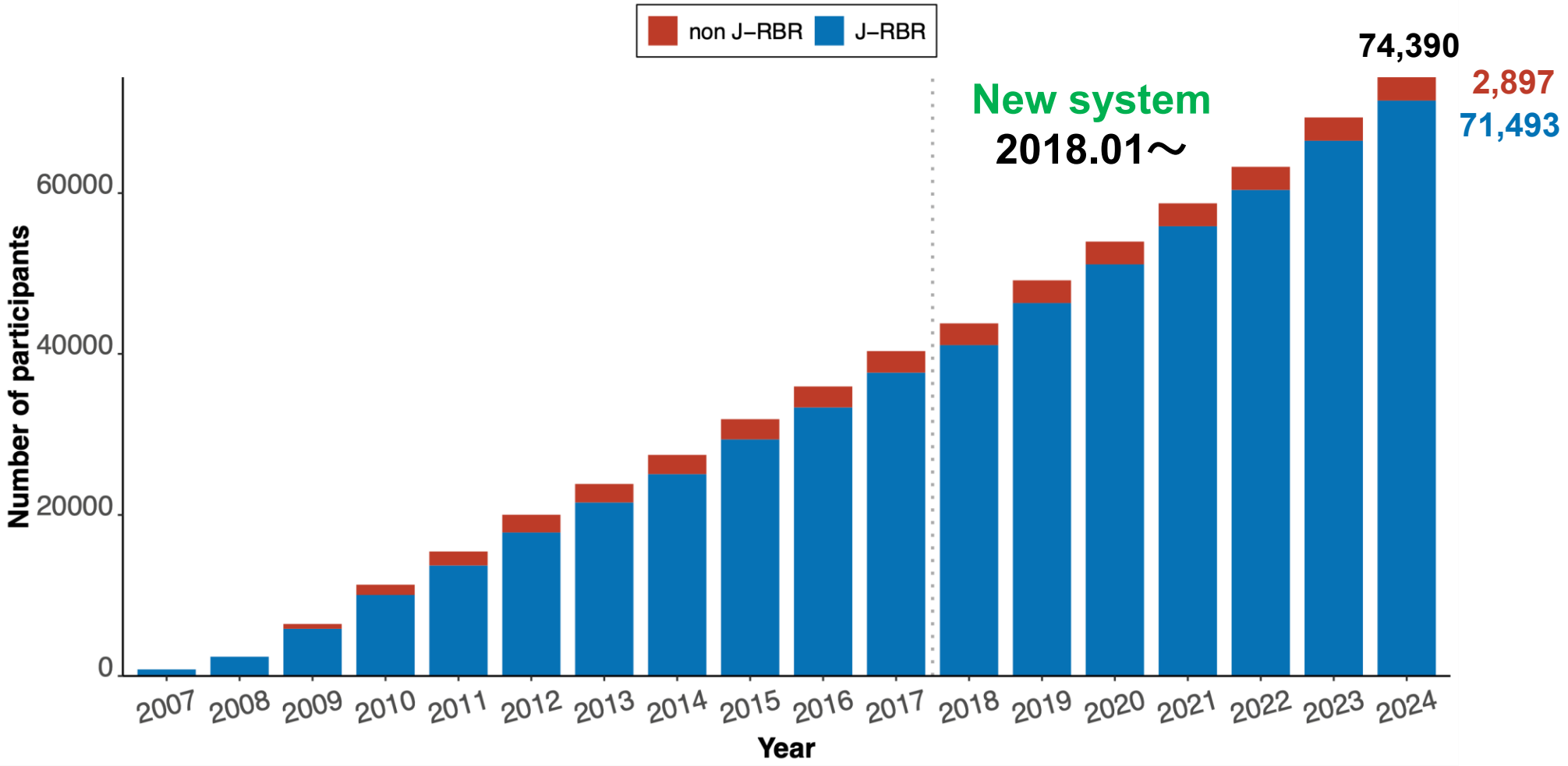
The revised version 2018 of the nationwide web-based registry system for kidney diseases in Japan: Japan Renal Biopsy Registry and Japan Kidney Disease Registry

Takaya Ozeki[1] · Shoichi Maruyama[1] · Michio Nagata[2] · Akira Shimizu[3] · Hitoshi Sugiyama[4] · Hiroshi Sato[5] · Hitoshi Yokoyama[6] on behalf of the Committee for Renal Biopsy and Disease Registry of the Japanese Society of Nephrology

Clin Exp Nephrol (2020) 24:1058–1068

We revised the J-RBR in 2018, updating the system for registering clinical and pathological diagnoses. We also added newly recognized disease categories, including C3 glomerulopathy and PGNMID. These updates improve the accuracy and relevance of nationwide data collection.

Trend in the Cumulative Number of Registered Patients in J-RBR/J-KDR (2007–2024)



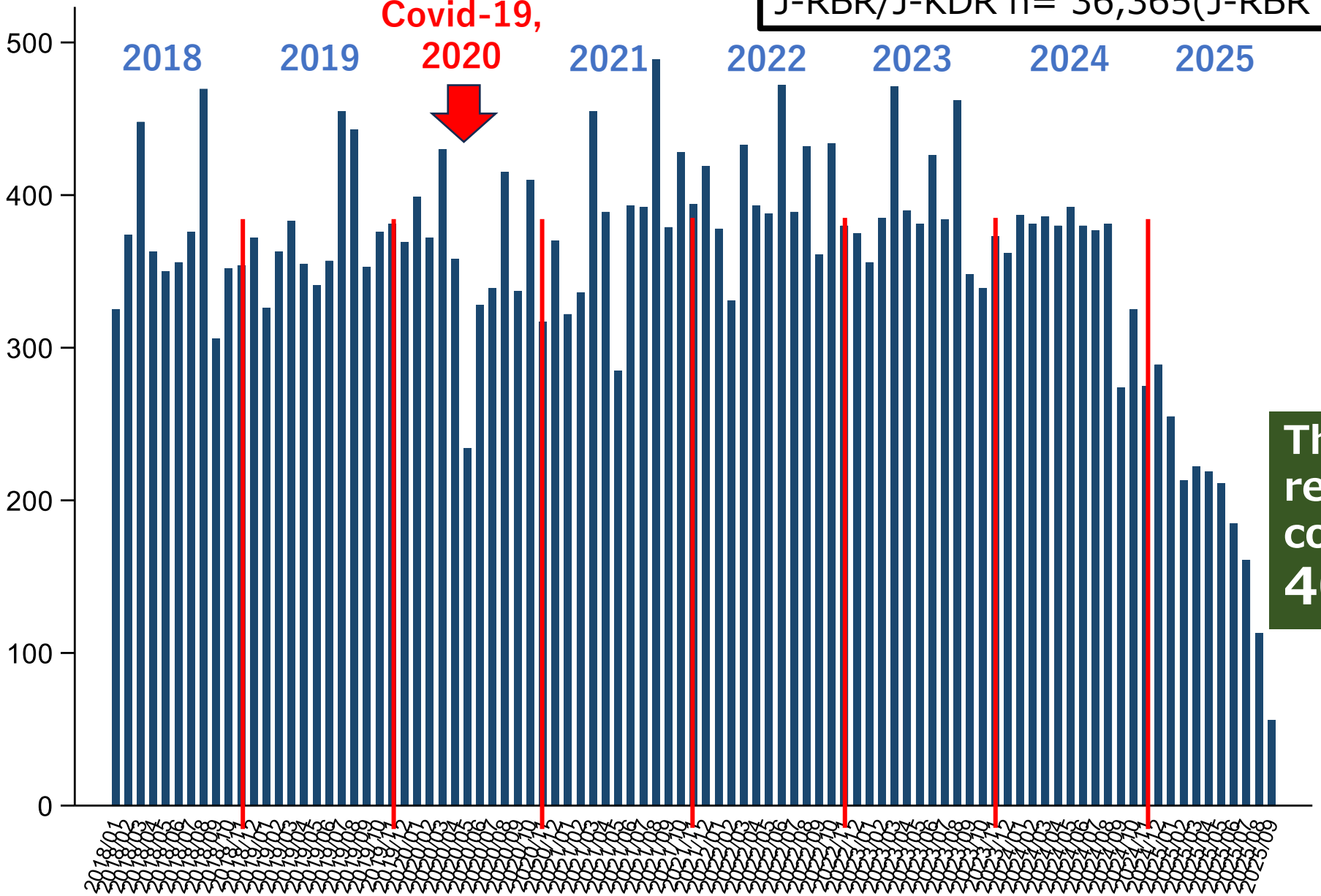
As of the end of 2024: total n=74,390（J-RBR : n=71,493　Non-Bx cases: 2,897 cases)

The JRBR is likely one of the largest kidney disease registries in the world.

Monthly Biopsy Counts

J-RBR/J-KDR n= 36,365(J-RBR n=36,133, non-Bx n=232)

Total number (2018～)
n=33,245

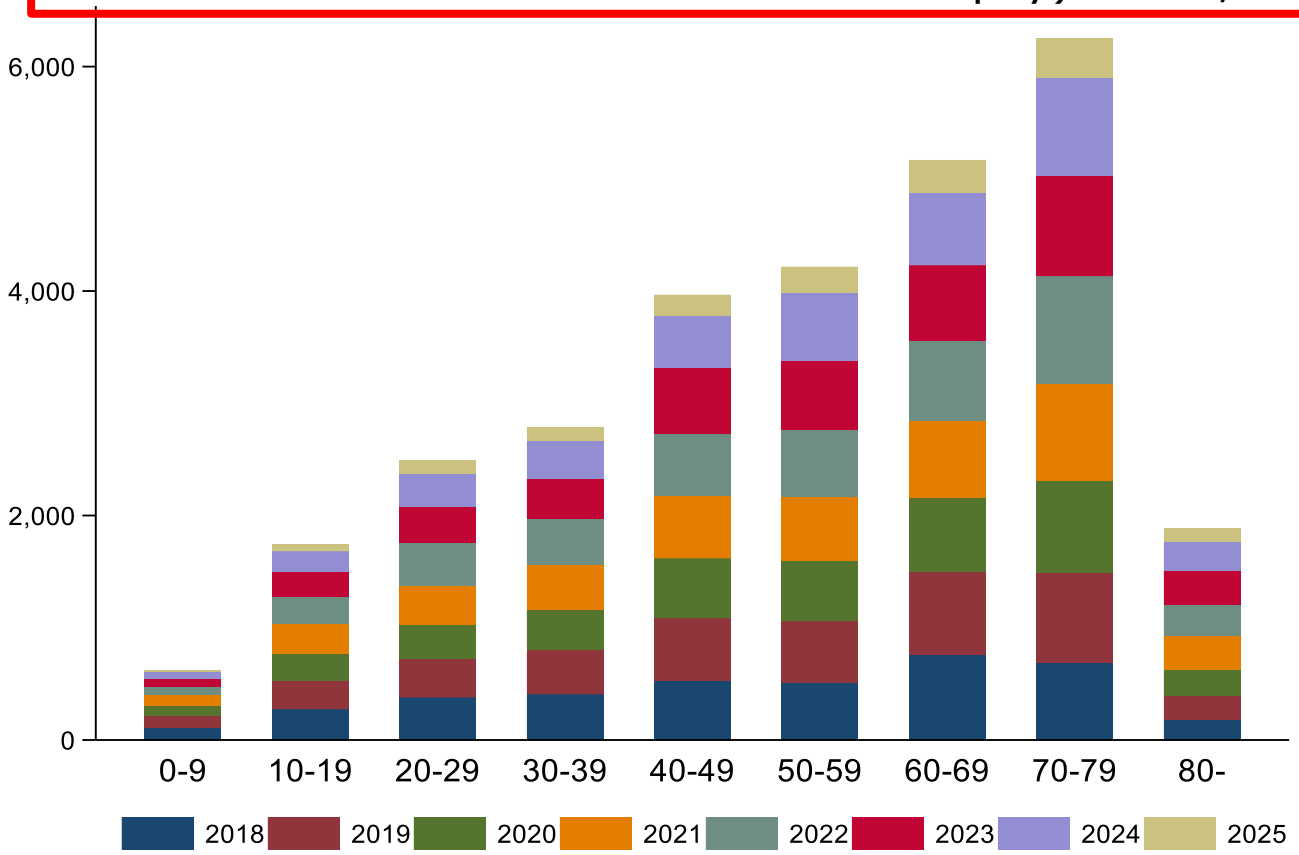


The number of biopsy registry has remained constant at around 4000-4500/Y.

Number of Biopsies by Age Group and Sex

J-RBR/J-KDR　　n= 36,365 (J-RBR n=36,133, Non-Bx n=232)

J-RBR（2018年1月～The first native biopsy) n=29,121



Age	n	%
0-9	619	2.13
10-19	1,740	5.98
20-29	2,491	8.55
30-39	2,787	9.57
40-49	3,967	13.62
50-59	4,217	14.48
60-69	5,168	17.75
70-79	6,248	21.46
80-	1,884	6.47
Total	29,121	100.00

性別	n	%
男性	15,202	52.20
女性	13,919	47.80
Total	29,121	100.00

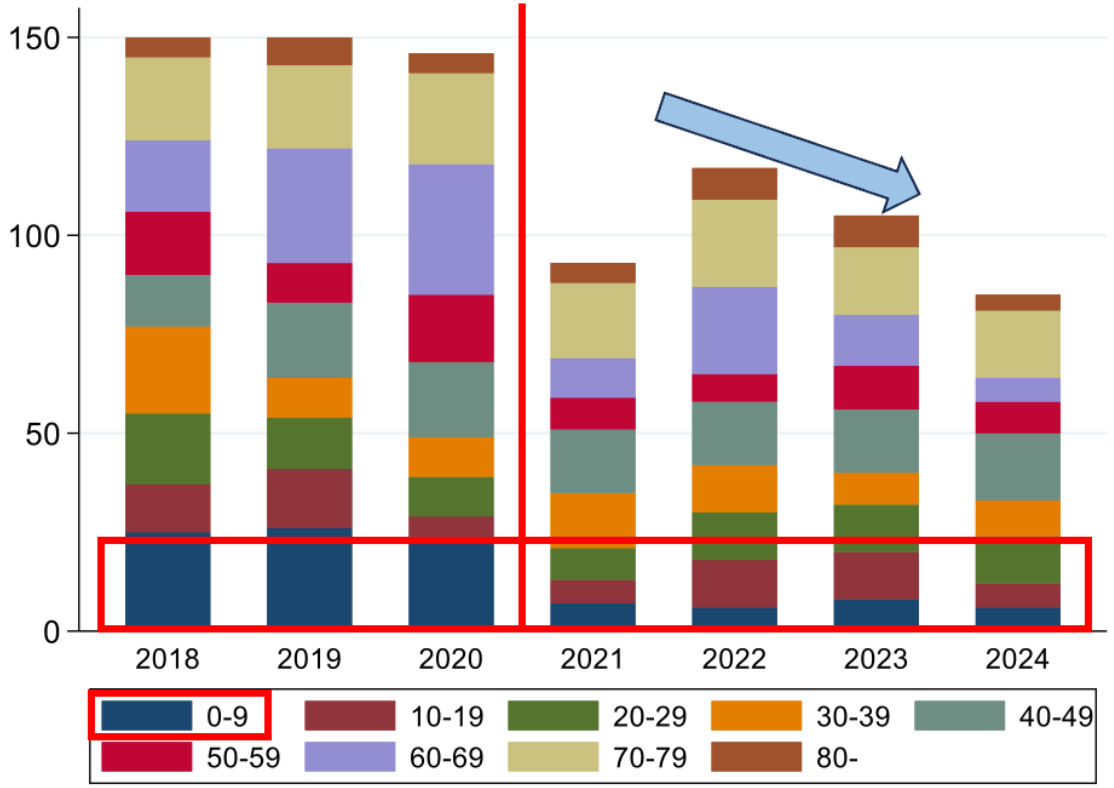
Kidney biopsies are most common in patients in their 70s. This reflects Japan's super-aged society, and this age distribution is one of the most distinctive features of the J-RBR.

Primary Diagnoses by Frequency 2018~2024

Main Diagnosis	n	%
IgA nephropathy	8,266	28.39
Membranous nephropathy	2,308	7.93
Minimal change disease	2,251	7.73
Vasculitis syndrome	2,077	7.13
Hypertensive nephrosclerosis	1,771	6.08
Tubulointerstitial nephritis	1,679	5.77
Diabetic nephropathy	1,357	4.66
FSGS	1,318	4.53
Collagen disease related	1,326	4.55
IgA vasculitis	939	3.22
Congenital/Gentetic	548	1.88
Amyloidosis	397	1.36
TMA	306	1.05
Infection related	296	1.02
MPGN	287	0.99
Paraprotein nephropathy	207	0.71
C3 nephropathy	78	0.27
Cryoglobulin nephropathy	44	0.15
Organized deposits	41	0.14
C1q nephropathy	25	0.09
IgM nephropathy	11	0.04
Altered lipid metabolism	9	0.03
others	3,580	12.29
Total	29,121	100.00

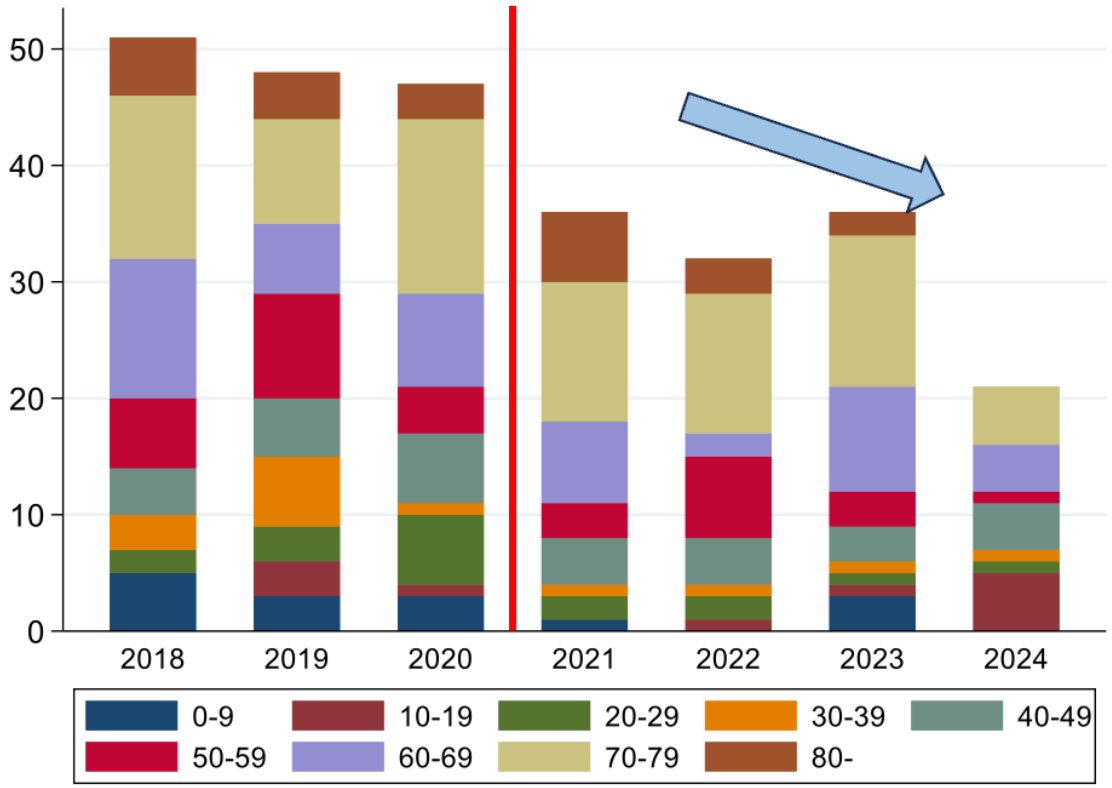
Changes in Age Distribution of Diagnoses of IgA Vasculitis and Infection-Related Glomerulonephritis (2018–2024)

IgA vasculitis



A marked decrease in IgAV among young children, particularly those aged 0–9 years, was observed.

Infection related GN



Infection-related glomerulonephritis declined overall.

Behavioral changes and modifications in clinical practice associated with the COVID-19 pandemic may have influenced these trends.

Japan Renal Biopsy Registry

Participating sites

Participating sites

Participating sites

UMIN

J-RBR

- ✓ Diagnosis
- ✓ Clinical data
- ✓ Pathological information

Ancillary Studies
- Cross-sectional
- Longitudinal

Collaborations

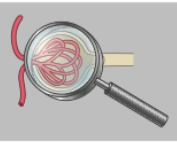
Whole Slide Imaging

J-RBR Ancillary Studies: Longitudinal

Retrospective

Focal segmental glomerulosclerosis histologic variants and renal outcomes

Background



The prognostic significance of the histologic variants of focal segmental glomerulosclerosis (FSGS) is unclear

Methods

Data from J-RBR, Japanese nationwide registry of kidney biopsies, between 2010–2013

Participants: Patients with FSGS
n=304 (173 nephrotic, 131 non-nephrotic)
*Secondary causes of FSGS and transplant biopsies excluded

Median follow-up: 4.8 years

Exposure: FSGS histologic variant

Primary outcome:
30% decline in eGFR or progression to ESKD

Results

Primary outcome
(30% decline in eGFR or ESKD)
n = 87 (29%)

Variant	Distribution	Adjusted HR
NOS	31%	1.00 (ref)
Tip	23%	0.86 p = 0.65
Perihilar	32%	1.11 p = 0.74
Cellular	23%	1.08 p = 0.84
Collapsing	33%	1.20 p = 0.71

Additional analysis

HR of proteinuria remission for the primary outcome, adjusted for FSGS variant and potential confounders:

0.19, p<0.001

Conclusion

FSGS variants alone might not have significant impact on renal outcome after 5 years.
Proteinuria remission was associated with better renal outcome irrespective of FSGS variant.

NEPHROLOGY DIALYSIS TRANSPLANTATION

ERA LEADING EUROPEAN NEPHROLOGY

Kawaguchi et al. NDT (2021)
@NDTSocial

Specific histological variant was not associated with renal outcome in FSGS.

Nephrol Dial Transplant. 2022 Aug 22;37(9):1679-1690

J-RBR Ancillary Studies: Longitudinal

Clinical Kidney Journal

SUrvey of renal Biopsy registry database and Anticancer dRUg therapy in Japan (SUBARU-J study)

The survey using the Japan Renal Biopsy Registry (J-RBR) was conducted to capture a real-world clinical snapshot and outcomes of anticancer drug-associated kidney complications in Japan.

Methods

- 49 Japanese facilities 2018–2021
- 449 cases with 'drug-induced kidney injury' checked in J-RBR database were surveyed
- 135 anticancer drug-related cases were analyzed
- Target:
  - Real-world clinical snapshot
  - Concomitant medications
  - Prognostic factors

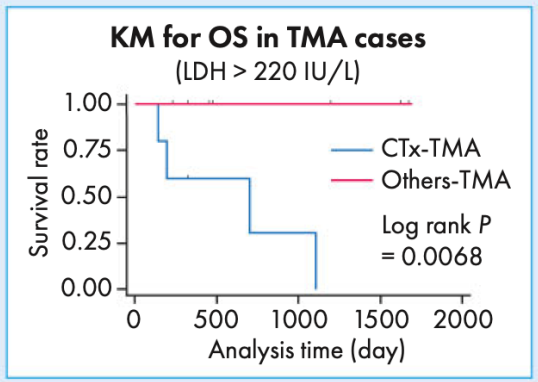
Results

Top 3 primary sites of malignancies	n (%)
Lung	45 (33.3)
Gastrointestinal	22 (16.3)
Gynecological	15 (11.1)

Top 3 histopathological diagnoses	n (%)
TIN	64 (47.4)
TMA	48 (35.6)
FSGS	5 (3.7)

Cox model for death event

	Multivariate analysis		
	OR	95% CI	P
Metastasis	2.95	1.32–6.56	0.008
PPI use	2.49	1.43–4.34	0.001
Serum Alb	0.42	0.28–0.63	0.000
Serum Cr	1.04	0.93–1.16	0.545
CRP	1.01	0.96–1.05	0.773



Conclusion: Our nationwide survey highlighted real-world pictures of biopsy-proven kidney complications associated with anticancer-drug therapy, and reminded us of several pitfalls including pathophysiology of CTx-TMA and prognostic role of PPI use.

Kuwabara, T., Miyasato Y., et al.
Clinical Kidney Journal (2024)
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Clinical Kidney Journal,2024,vol.17,no.12,sfae327

J-RBR Ancillary Studies: Longitudinal

J-NSCS: Japan Nephrotic Syndrome Cohort Study

Primary Nephrotic Syndrome, biopsy performed during 2009-2010
MCD (n=155), MN (n=140), FSGS (n=38), Others (n=33)

	MCD	MN	FSGS	Others
Incidence of infection, *N* (%)	13 (8.4)	9 (6.1)	3 (7.9)	3 (9.1)
Follow-up period (year)	4.9 (2.8, 5.0)	5.0 (2.7, 5.0)	5.0 (3.9, 5.0)	4.1 (2.6, 5.0)
Incidence rate of infection, per 1000 person-years	22.1 (11.8, 37.8)	16.2 (7.4, 30.8)	20.4 (4.2, 59.6)	25.1 (5.2, 73.3)
Hazard ratio (95% CI)[a]				
Unadjusted model	1.33 (0.57, 3.11)	1.00 (reference)	1.27 (0.35, 4.71)	1.48 (0.40, 5.46)
Multivariable-adjusted model 1	2.41 (0.98, 5.94)[†]	1.00 (reference)	1.58 (0.43, 5.88)	1.78 (0.48, 6.58)
Multivariable-adjusted model 2	2.44 (1.00, 5.95)[‡]	1.00 (reference)	1.48 (0.40, 5.50)	1.26 (0.30, 5.29)
Multivariable-adjusted model 3	2.56 (1.04, 6.34)[‡]	1.00 (reference)	1.55 (0.41, 5.83)	1.22 (0.29, 5.19)

CI confidence interval, *FSGS* focal segmental glomerulosclerosis, *MCD* minimal change disease, *MN* membranous nephropathy

[†] $P=0.06$

[‡] $P<0.05$

[a]Model 1 adjusted for baseline age and stratified by sex; Model 2 adjusted for baseline age and serum creatinine and stratified by sex; Model 3 adjusted for baseline age, serum creatinine, and urinary protein and stratified by sex

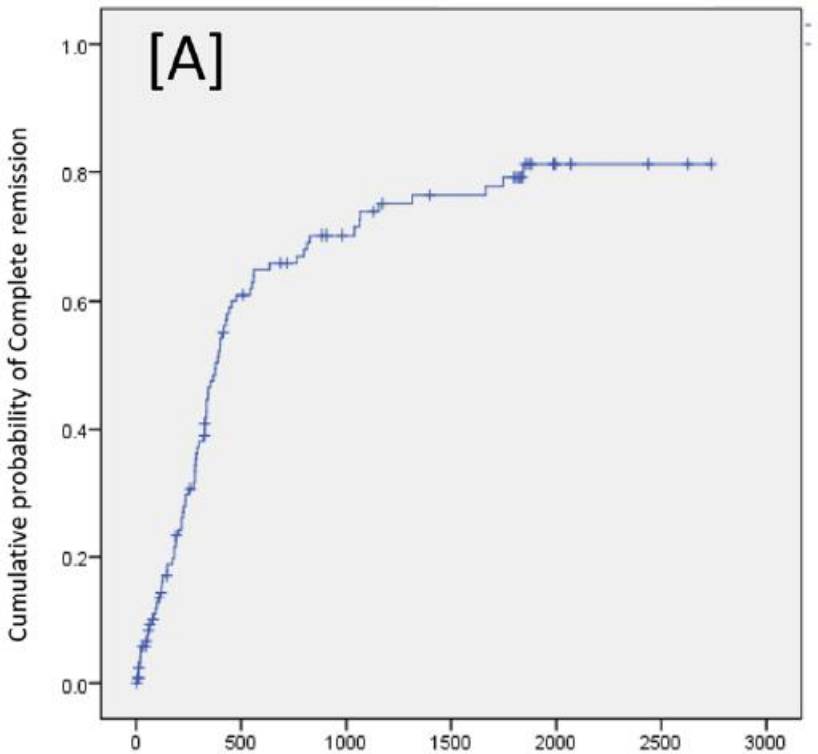
Clin Exp Nephrol. 2020 Jun;24(6):526-540.

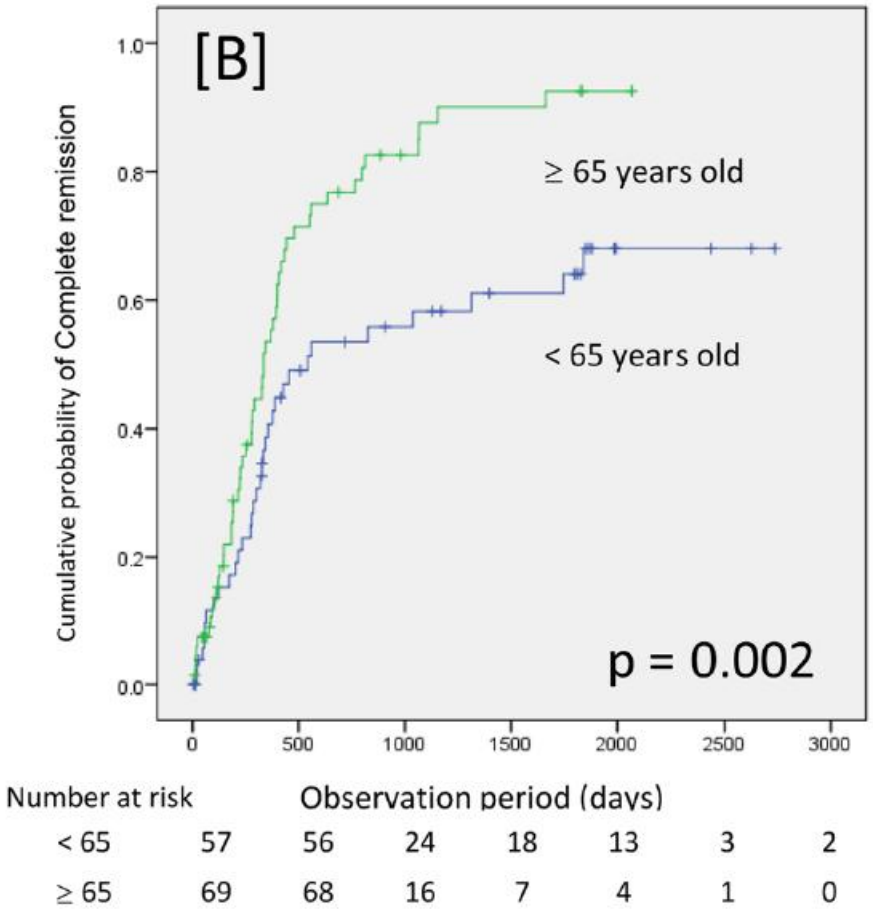
Risk of infection in MCD was higher than other type of NS.

Clinical and Experimental Nephrology (2020) 24:893–909
https://doi.org/10.1007/s10157-020-01913-9

ORIGINAL ARTICLE

Better remission rates in elderly Japanese patients with primary membranous nephropathy in nationwide real-world practice: The Japan Nephrotic Syndrome Cohort Study (JNSCS)

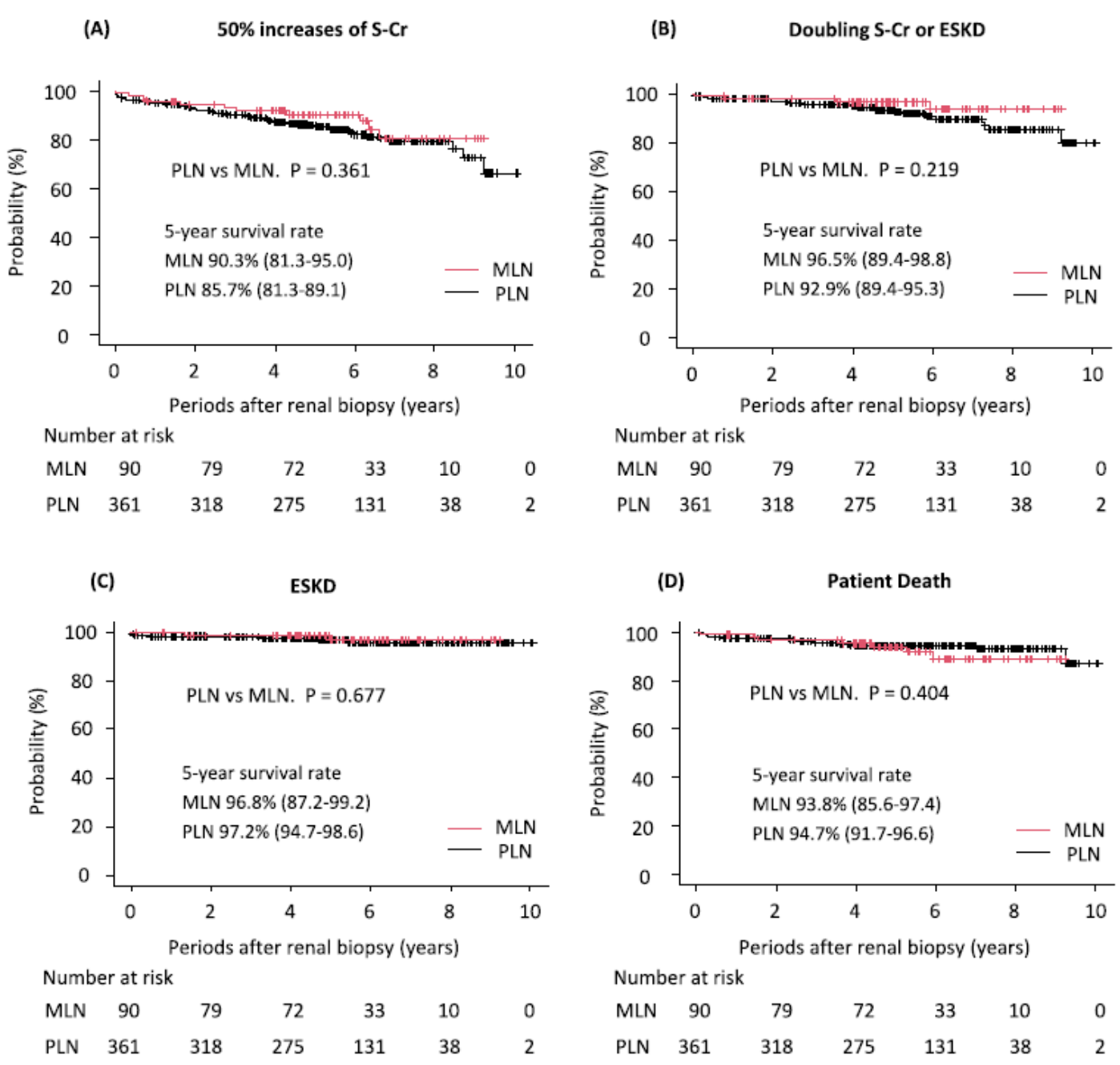




Clinical and Experimental Nephrology (2025) 29:1363–1372
https://doi.org/10.1007/s10157-025-02709-5

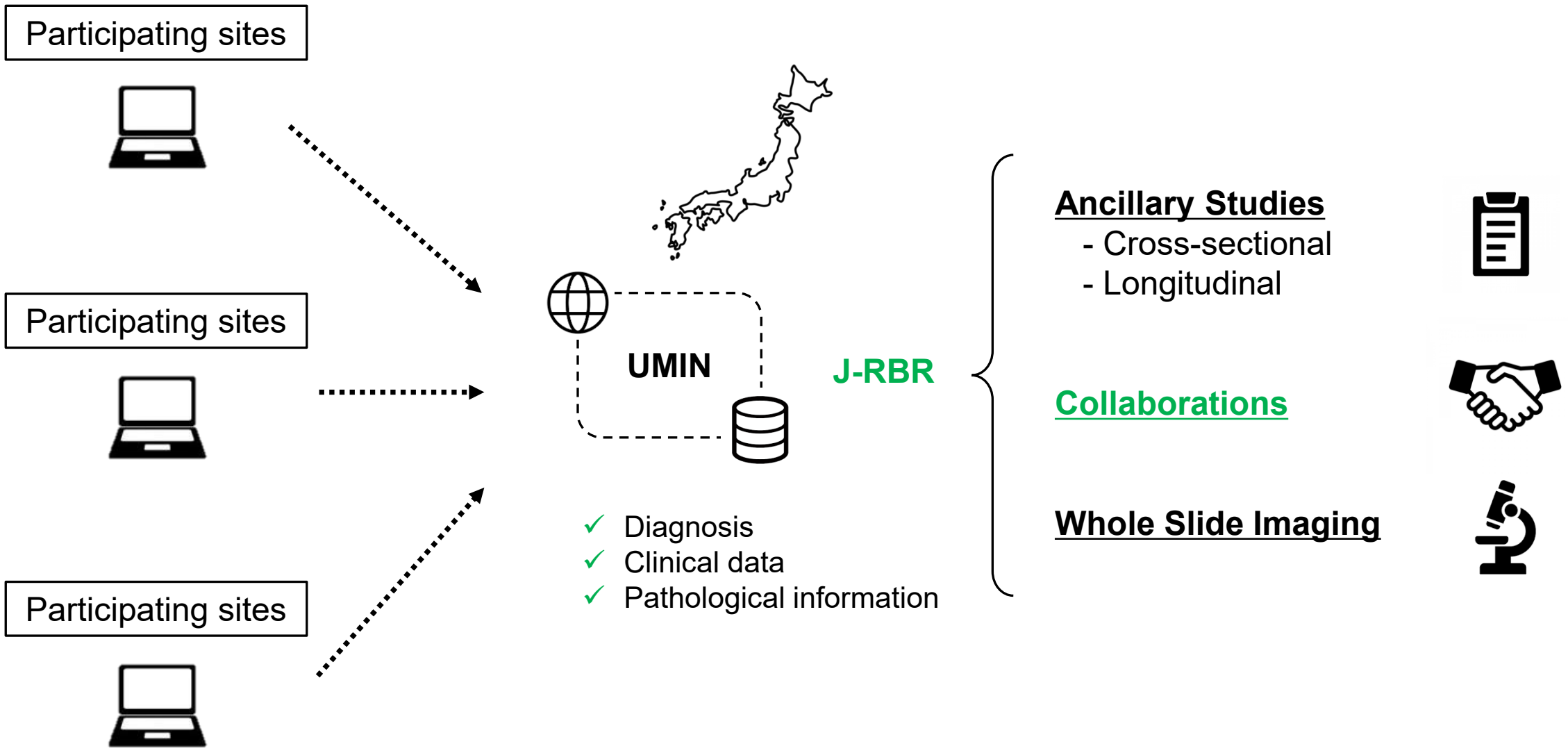
ORIGINAL ARTICLE

Long-term prognosis of pure membranous lupus nephritis: a comparison with proliferative lupus nephritis in Japan



In JRBR, no statistically significant differences in renal outcomes were observed between pure MLN and PLN.

Japan Renal Biopsy Registry



Clinical Course of Adult FSGS and Minimal Change Disease in North American and Japanese Cohorts

Kidney360

Methods

Patients with **FSGS and MCD**

Who recieved **Inmunosuppresive Therapy (IST)**

Neptune Cohort (American) **n=89**

N-KDR Cohort (Japanese) **n=288**

Baseline characteristics and Complete Remission were compared

Results

Baseline characteristics

	Neptune (American)	N-KDR (Japanese)
FSGS	53.9%	17.0%
Family History of Kidney Disease	35.2%	3.2%
Age (Median)	43	56
UPCR	6.65	7.73
Albumin	2.2 mg/dL	1.6 mg/dL

*FSGS= Focal and Segmental Glomerulosclerosis
MCD= Minimal Change Disease
UPCR = Urine Protein creatinine ratio

Complete Remission (CR)

	N-KDR	Neptune
Overall	89.2%	62.9%
FSGS	67.3%	43.7%
MCD	93.7%	85.4%

Multivariable Model

Time to CR

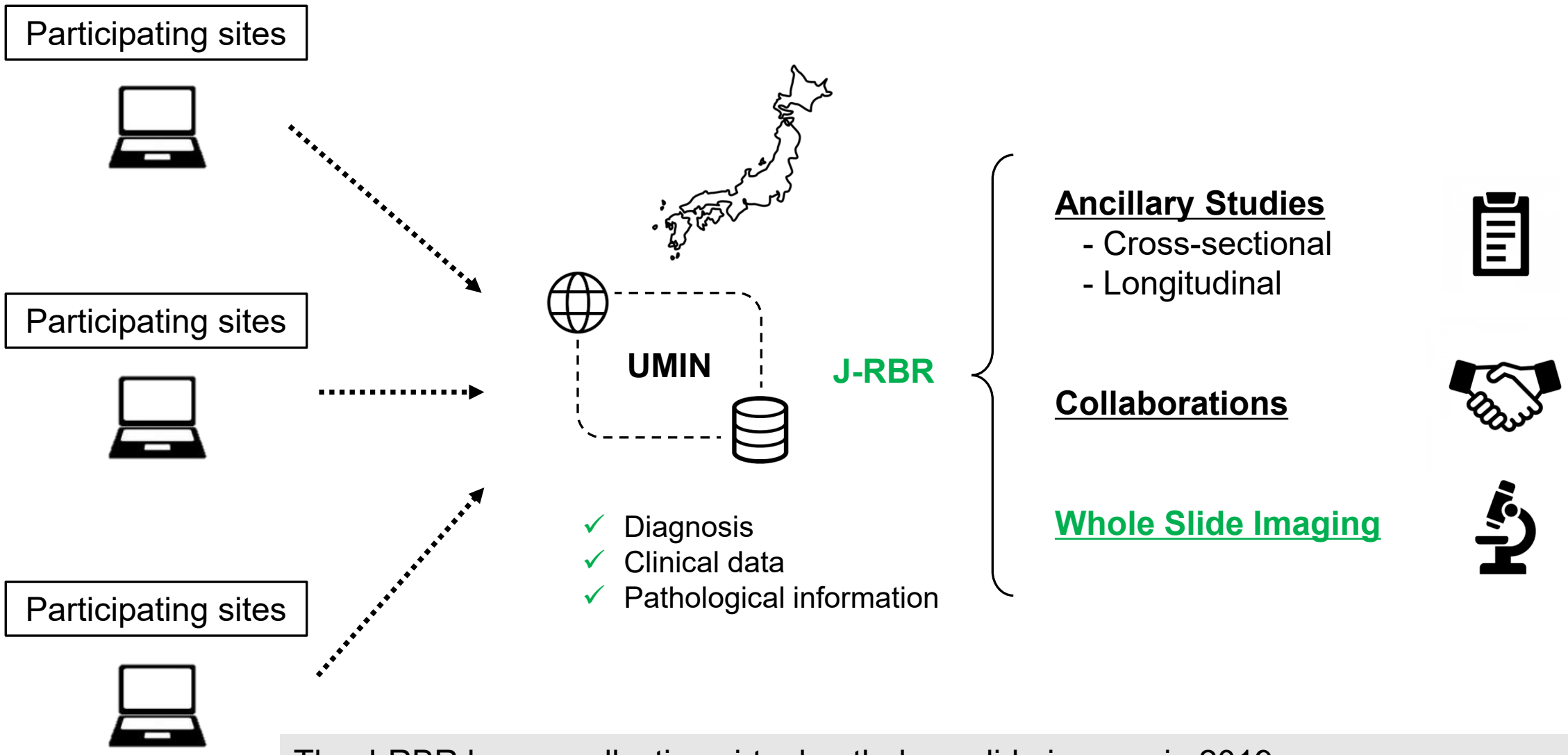
- HR=0.93 SBP per 10mmHg
- HR=1.16 eGFR per 10ml/min
- HR=0.28 FSGS

Conclusions: The North American cohort had more FSGS and more frequent family history. Japanese patients showed more severe NS with better response to IST. FSGS, hypertension and lower eGFR were shared predictors of poor treatment response. Identifying shared and unique features across geographically diverse populations may help uncover biologically relevant subgroups, improve prediction of disease course and better design future multi-national clinical trials.

Takaya Ozeki, Brenda Gillespie, Maria Larkina, *et al.* ***Clinical Course of Adult FSGS and Minimal Change Disease in North American and Japanese Cohorts.*** *Kidney360.* 10.34067/KID.0000000000000133

Visual Abstract by Verner Venegas

Japan Renal Biopsy Registry



The J-RBR began collecting virtual pathology slide images in 2019.
Although the number of uploaded images is still limited, it has been gradually increasing.

Conclusion

- The J-RBR is the first nationwide web-based registry system for the patients who are performed kidney biopsy in Japan, which currently has over 70,000 registration.
- The J-RBR highlighted unique distribution and clinical manifestations of kidney diseases in Japan, and also helped to reveal their diagnostic and prognostic factors.
- The J-RBR will bring novel insights for better understanding of kidney diseases and be able to provide fundamental data for future intra- and international collaborations.

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Innovations in GN: Trials, Registries, and Digital Pathology

Kidney Biopsy Registry in Japan

NAGOYA UNIVERSITY

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