nephritic syndrome (55.4 % and 50.0 % in 2009 and 2010, respectively), followed by nephrotic syndrome (22.4 % and 27.0 %); the most frequent pathological diagnosis as classified by the pathogenesis was IgA nephropathy (31.6 % and 30.4 %), followed by primary glomerular diseases (except IgA nephropathy) (27.2 % and 28.1 %). Among the primary glomerular diseases (except IgA nephropathy) in the patients with nephrotic syndrome, membranous nephropathy was the most common histopathology in 2009 (40.3 %) and minor glomerular abnormalities (50.0 %) were the most common in 2010 in native kidneys in the J-RBR. Five new secondary and longitudinal research studies by the J-KDR were started in 2009 and one was started in 2010.

Keywords Native kidney biopsy · Primary glomerulonephritis · IgA nephropathy · Membranous nephropathy · Renal grafts · National registry

Introduction

The Japanese Society of Nephrology (JSN) established the Japan Renal Biopsy Registry (J-RBR) in 2007, and it conducted analyses for 2007 and 2008 [1]. In 2009, the JSN started the Japan Kidney Disease Registry (J-KDR) to record clinically-diagnosed cases in addition to the J-RBR. Participation in the J-KDR, including the J-RBR, was requested from appointed clinical training hospitals of the JSN and the Japanese Society for Dialysis Therapy in an attempt to extend the registry nationwide. In this report, the detailed data of the J-RBR and the frequencies of the different clinical diagnoses in the J-KDR registered from January to December of 2009 and 2010 are summarized.

Subjects and methods

Registry system and patients

This report includes the data from patients included in the J-RBR and J-KDR (J-RBR/J-KDR), registered prospectively from January 2009 to December 2010. The patients' data, including age, gender, laboratory data, and the clinical and pathological diagnoses, were recorded at each institution and registered on the web page of the J-RBR/J-KDR utilizing the Internet Data and Information Center for Medical Research (INDICE) system of the University Hospital Medical Information Network (UMIN), as described previously [1]. The ethics committee of the JSN and that of Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences comprehensively approved the study, and a local committee of participating centers and their affiliate hospitals individually approved the study. Written informed consent was

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obtained from the patients at the time of biopsy or at the time they were registered to participate in the study. The J-RBR/J-KDR is registered in the Clinical Trial Registry of UMIN (Registered Number UMIN000000618).

Clinical or renal histopathological diagnosis and laboratory data

Three classifications, including the clinical diagnosis, histological diagnosis based on the pathogenesis, and histological diagnosis based on a histopathological examination, were made for each case included in the J-RBR, as described previously [1]. Of these classifications, the clinical diagnosis alone was selected for the J-KDR. The definition of each diagnosis was based on the clinical syndromes and renal histopathology, as described previously [2]. IgA nephropathy (IgAN) (Berger disease) was separated from primary glomerular diseases on the basis of basic glomerular alterations in the classification of glomerular diseases by the World Health Organization [2]. In 2010, hemolytic uremic syndrome and thrombotic thrombocytopenic purpura (HUS/TTP), congenital anomalies of the kidney and urinary tract (CAKUT) and polycystic kidney disease (PKD) were added to the classification of the clinical diagnosis on the case record (Table S1). The clinical data, including the results of the urinalysis, daily proteinuria, serum creatinine concentrations, total protein, albumin, and the total cholesterol values, were always recorded, while the systolic and diastolic blood pressure, prescription use of anti-hypertensive agents, hemoglobin A1c, and presence of diabetes mellitus were optionally recorded. The estimated glomerular filtration rate was calculated as described previously [3]. The frequency of the diseases are here described in general, but the clinical data were also analyzed separately for cases of IgAN, which is the most common renal disease in Japan [1, 4, 5].

Statistical analyses

Data are expressed as the mean ± SD for continuous parametric data, medians and interquartile ranges for continuous non-parametric data, and frequencies for categorical data. The statistical analyses were performed using the JMP software program, version 8 (SAS Institute Inc., Cary, NC, USA).

Results

Baseline characteristics of the J-RBR/J-KDR participants in 2009 and 2010

The numbers of participating facilities and registered renal biopsies or cases without renal biopsies in the registry in 2009 and 2010 are shown in Table 1. The J-KDR was started in 2009 and the number of participating facilities increased by 34 compared to 2008, reaching a total of 57 facilities in the J-RBR and 59 facilities in the J-KDR. The number of total renal biopsies increased to 3,336 in 2009, which was 1,754 more biopsies than in the previous year [1], and in 2010 it further increased to 4,106 in the J-RBR. The number of other cases (not in the J-RBR), which corresponds to the cases without renal biopsies but diagnosed by clinical findings, was 680 and 575 in 2009 and 2010, respectively. The average age of this cohort was more than 10 years higher than that of the J-RBR in each year (Table 1).

The number of native kidney biopsies increased; however, that of renal graft biopsies registered in 2009 slightly decreased compared to 2008 (Table 2). The distribution of age ranges showed a peak distribution in the seventh decade in both genders for native kidneys (Table 3). Patients younger than 20 years of age comprised 12.1 % and 10.3 % of the cases, and those 65 years of age and over comprised 24.5 % and 4.7 % of the native kidney and renal grafts, respectively, during the 2-year period (2009 and 2010). In the patients who underwent renal grafts, both the average age and the peak distribution of age ranges were younger than those of patients who underwent native kidney biopsies (Tables 2, 3).

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Table 1 The number of participated renal centers and registered renal biopsies or other cases without renal biopsies in J-RBR/J-KDR 2009 and 2010

	2009 J-KDR			2010 J-KDR		
	J-RBR	Other cases[a]	Total	J-RBR	Other cases[a]	Total
Renal centers (*n*)[b]	57[c]	–	59	83	–	94
Total biopsies or cases (*n*)	3,336[d] (83.1 %)	680 (16.9 %)	4,016 (100.0 %)	4,106 (87.7 %)	575 (12.3 %)	4,681 (100.0 %)
Average age (years)	46.7 ± 19.9	58.1 ± 17.8	48.7 ± 20.0	46.7 ± 20.6	56.8 ± 21.1	47.9 ± 20.9
Male (*n*)	1,787 (53.6 %)	418 (61.5 %)	2,205 (54.9 %)	2,183 (53.2 %)	335[e] (58.3 %)	2,518[e] (53.8 %)
Female (*n*)	1,549 (46.4 %)	262 (38.5 %)	1,811 (45.1 %)	1,923 (46.8 %)	238[e] (41.4 %)	2,161[e] (46.2 %)

J-RBR Japan Renal Biopsy Registry, *J-KDR* Japan Kidney Disease Registry

Note that J-RBR started in 2007 and J-KDR started in 2009

[a] Other cases include patients diagnosed by clinical findings without renal biopsies

[b] The number represents principal institutions having affiliate hospitals. All of the participated institutions and hospitals in the J-RBR and J-KDR in 2009 and 2010 are shown in the "Appendix". The number of renal centers in total is based on the registration of cases without renal biopsies but diagnosed by clinical findings in addition to that of data with renal biopsy in J-RBR

[c] Increase of 34 when compared to the number in J-RBR 2008

[d] Increase of 1,754 when compared to the number in J-RBR 2008

[e] No registered data for gender in 2 cases

The frequency of clinical diagnoses in the J-RBR

Three classifications, the clinical diagnosis, histological diagnosis based on the pathogenesis, and the histological diagnosis based on a histopathological examination, were included in the J-RBR database, while the clinical diagnosis alone was registered for the other cases. In the J-RBR, a clinical diagnosis of chronic nephritic syndrome was the most common, followed by nephrotic syndrome, in both total biopsies and native kidneys in 2009 and 2010, which was similar to the findings in 2007 and 2008 (Table 4) [1]. In native kidneys, more than half of the cases that were registered had chronic nephritic syndrome. The age distribution according to the classification of clinical diagnoses in native kidneys in the J-RBR in 2009 and 2010 was analyzed, and cases with rapidly progressive nephritic syndrome exhibited the highest mean age while cases with inherited renal diseases showed the youngest mean age (Table 5).

The frequency of pathological diagnoses in the J-RBR

The pathological diagnoses were classified based on the pathogenesis (Table 6) and histopathology (Table 7). In the classification of the pathogenesis, IgAN was diagnosed most frequently (31.6 %), followed by primary glomerular disease other than IgAN (27.2 %) in native kidneys in both 2009 and 2010 (Table 6). Similar frequencies of IgAN, primary glomerular disease other than IgAN and diabetic nephropathy were observed in the combined data for 2007 and 2008 [1]. In the pathological diagnosis classified based on the histopathology in native kidney biopsies, mesangial proliferative glomerulonephritis was the most frequently observed disease, representing 42.5 % and 35.8 % of the cases in 2009 and 2010 (Table 7).

Table 2 The number of registered renal biopsies in J-RBR 2009 and 2010

Years	2009	2010	Total
Native kidneys, *n* (%)	3,165[a] (94.9)	3,869 (94.2)	7,034 (94.5)
Average age (years)	47.0 ± 20.1	47.1 ± 20.8	47.1 ± 20.5
Median age (years)	50 (30–64)	49 (31–65)	49 (30–64)
Male, *n* (%)	1,671 (52.8)	2,035 (52.6)	3,706 (52.7)
Female, *n* (%)	1,494 (47.2)	1,834 (47.4)	3,328 (47.3)
Renal grafts, *n* (%)	171[b] (5.1)	237 (5.8)	408 (5.5)
Average age (years)	40.9 ± 15.0	41.3 ± 15.4	41.1 ± 15.2
Median age (years)	43 (31–52)	41 (33–54)	42 (32–53)
Male, *n* (%)	116 (67.8)	148 (62.4)	264 (64.7)
Female, *n* (%)	55 (32.2)	89 (37.6)	144 (35.3)

[a] Increase of 1,765 when compared to the number in J-RBR 2008

[b] Decrease of 11 when compared to the number in J-RBR 2008

Primary glomerular disease (except IgAN) and nephrotic syndrome in the J-RBR

In the cohort of primary glomerular diseases (except IgA nephropathy) as classified based on the pathogenesis, membranous nephropathy (MN) was predominant in 2009, followed by minor glomerular abnormalities, while minor glomerular abnormalities were the most common diagnosis in 2010, followed by MN (Table 8).

Table 3 Distribution of age ranges and gender in J-RBR 2009 and 2010

	2009									2010								
	Total biopsies (n = 3,336)			Native kidneys (n = 3,165)			Renal grafts (n = 171)			Total biopsies (n = 4,106)			Native kidneys (n = 3,869)			Renal grafts (n = 237)		
Age (years)	Total	Male	Female	Total	Male	Female	Total	Male	Female	Total	Male	Female	Total	Male	Female	Total	Male	Female
0–9	60	33	27	57	32	25	3	1	2	121	94	27	136	87	49	7	7	0
10–19	318	169	149	304	160	144	14	9	5	352	203	149	354	193	161	18	10	8
20–29	413	194	219	392	180	212	21	14	7	406	187	219	429	167	262	22	20	2
30–39	476	221	255	438	193	245	38	28	10	533	278	255	549	248	301	62	30	32
40–49	434	222	212	391	197	194	43	25	18	489	277	212	489	251	238	50	26	24
50–59	545	317	228	509	291	218	36	26	10	575	347	228	541	311	230	49	36	13
60–69	645	382	263	631	371	260	14	11	3	733	470	263	756	452	304	28	18	10
70–79	372	213	159	370	211	159	2	2	0	437	278	159	515	277	238	1	1	0
80+	73	36	37	73	36	37	0	0	0	86	49	37	100	49	51	0	0	0
Total	3,336	1,787	1,549	3,165	1,671	1,494	171	116	55	3,732	2,183	1,549	3,869	2,035	1,834	237	148	89
Under 20 (%)	11.3	11.3	11.4	11.4	11.5	11.3	9.9	8.6	12.7	12.5	13.6	11.4	12.7	13.8	11.5	10.5	11.5	9.0
65 and over (%)	22.4	23.9	20.1	23.4	25.3	21.3	4.7	4.3	5.5	24.2	25.4	20.7	25.4	26.9	23.7	4.6	5.4	3.4

Age (years)	Total								
	Total biopsies (n = 7,442)			Native kidneys (n = 7,034)			Renal grafts (n = 408)		
	Total	Male	Female	Total	Male	Female	Total	Male	Female
0–9	203	127	76	193	119	74	10	8	2
10–19	690	372	318	658	353	305	32	19	13
20–29	864	381	483	821	347	474	43	34	9
30–39	1,087	499	588	987	441	546	100	58	42
40–49	973	499	474	880	448	432	93	51	42
50–59	1,135	664	471	1,050	602	448	85	62	23
60–69	1,429	852	577	1,387	823	564	42	29	13
70–79	888	491	397	885	488	397	3	3	0
80+	173	85	88	173	85	88	0	0	0
Total	7,442	3,970	3,472	7,034	3,706	3,328	408	264	144
Under 20 (%)	12.0	12.6	11.3	12.1	12.7	11.4	10.3	10.2	10.4
65 and over (%)	23.4	24.7	21.8	24.5	26.1	22.6	4.7	4.9	4.2

Table 4 The frequency of classification of clinical diagnoses in J-RBR 2009 and 2010

Classification	2009			2010			Total		
	Total biopsies (n = 3,336)		Native kidneys (n = 3,165)	Total biopsies (n = 4,106)		Native kidneys (n = 3,869)	Total biopsies (n = 7,442)		Native kidneys (n = 7,034)
	n	%	%[a]	n	%	%[a]	n	%	%[a]
Chronic nephritic syndrome	1,759	52.7	55.4	1,944	47.3	50.0	3,703	49.8	52.5
Nephrotic syndrome	711	21.3	22.4	1,044	25.4	27.0	1,755	23.6	24.9
Rapidly progressive nephritic syndrome	200	6.0	6.3	292	7.1	7.5	492	6.6	7.0
Renal transplantation	160	4.8	–	227	5.5	–	387	5.2	–
Renal disorder with collagen disease or vasculitis	116	3.5	3.7	144	3.5	3.7	260	3.5	3.7
Recurrent or persistent hematuria	97	2.9	3.0	111	2.7	2.9	208	2.8	2.9
Renal disorder with metabolic disease	63	1.9	2.0	61	1.5	1.6	124	1.7	1.8
Acute nephritic syndrome	54	1.6	1.6	58	1.4	1.5	112	1.5	1.6
Hypertensive nephropathy	39	1.2	1.2	54	1.3	1.4	93	1.3	1.3
Acute renal failure	36	1.1	1.1	35	0.9	0.9	71	1.0	1.0
Drug-induced nephropathy	13	0.4	0.4	26	0.6	0.6	39	0.5	0.5
Inherited renal disease	6	0.2	0.2	15	0.4	0.4	21	0.3	0.3
HUS/TTP	–	–	–	3	0.1	0.1	3	0.0	0.0
Others	82	2.5	2.6	92	2.2	2.4	174	2.4	2.5
Total	3,336	100.0	100.0	4,106	100.0	100.0	7,442	100.0	100.0

[a] Patients classified as either "Renal graft" or "Renal transplantation" in other categories were also excluded

In the patients with nephrotic syndrome as classified by the clinical diagnosis, primary glomerular disease other than IgAN was the predominant diagnosis in both 2009 and 2010, followed by diabetic nephropathy, which was the same order as in 2007 and 2008 (Table 9). Among the patients with primary glomerular diseases (except IgA nephropathy) who had nephrotic syndrome, MN was dominant, followed by minor glomerular abnormalities, viz., minimal change nephrotic syndrome (MCNS), focal segmental glomerulosclerosis (FSGS), and membranoproliferative glomerulonephritis (MPGN) (types I and III) in 2009. In 2010, minor glomerular abnormalities were the leading diagnosis, followed by MN, FSGS, and MPGN (types I and III) (Table 10).

Clinical diagnosis of membranous nephropathy, minor glomerular abnormalities, and focal segmental glomerulosclerosis in patients with primary glomerular diseases (except IgA nephropathy) in the J-RBR

A subanalysis of the subjects with a clinical diagnosis of MN, minor glomerular abnormalities, and FSGS who had primary glomerular diseases (except IgA nephropathy) was also performed, since these were the most common forms of such diseases. Nephrotic syndrome was the most common clinical diagnosis in cases with primary MN and primary minor glomerular abnormalities (MCNS) (Tables 11, 12), whereas chronic nephritic syndrome and nephrotic syndrome were the most common in cases with primary FSGS in 2009 and 2010, respectively (Table 13).

Subanalysis of cases of IgA nephropathy in the J-RBR

The profile, distribution of age ranges, classification of the clinical diagnosis, and the pathological diagnosis of IgAN, the most common glomerulonephritis reported in the J-RBR, were further analyzed (Tables 14, 15, 16, 17, 18, S2, S3). The average age of the overall subjects was in the fourth decade. There were no differences in the proportion of patients based on gender, but the age was significantly higher in males than in females in 2009 (Table 14). In terms of the distribution of age ranges, the peak distribution was in the twenties individually in both genders and in the overall cases in 2009, while it was in the thirties in both genders and overall in 2010, as well as in the combined data from 2009 and 2010 (Table 15). Patients younger than 20 years of age comprised 14.4 % of the cases and those 65 years and over comprised 7.9 % of the cases in the combined data from 2009 and 2010 (Table 15). The majority of the clinical and pathological diagnoses were chronic nephritic syndrome (Table 16) and mesangial proliferative glomerulonephritis (Table 17), respectively,

Table 5 The age distribution of classification of clinical diagnoses in native kidneys in J-RBR 2009 and 2010

Classification	2009			2010			Total		
	Male	Female	Total	Male	Female	Total	Male	Female	Total
Chronic nephritic syndrome	44.4 ± 18.8	41.2 ± 17.8	42.8 ± 18.4	43.5 ± 19.3	41.0 ± 18.2	42.2 ± 18.8	43.9 ± 19.1	41.0 ± 18.0	42.5 ± 18.6
Nephrotic syndrome	52.6 ± 21.6	54.7 ± 21.1	53.5 ± 21.4	49.5 ± 23.4	50.9 ± 22.6	50.1 ± 23.0	50.8 ± 22.7	52.5 ± 22.0	51.5 ± 22.4
Rapidly progressive nephritic syndrome	64.5 ± 13.0	61.2 ± 17.4	63.0 ± 15.1	65.4 ± 11.5	65.3 ± 15.3	65.4 ± 13.3	65.1 ± 12.1	63.6 ± 16.3	64.4 ± 14.1
Renal disorder with collagen disease or vasculitis	48.0 ± 21.5	46.2 ± 20.1	46.7 ± 20.4	54.3 ± 19.5	46.3 ± 19.6	48.7 ± 19.9	51.6 ± 20.5	46.2 ± 19.8	47.8 ± 20.1
Recurrent or persistent hematuria	33.4 ± 17.4	33.8 ± 16.9	33.6 ± 17.0	49.5 ± 19.0	38.0 ± 17.1	42.6 ± 18.6	41.8 ± 19.9	36.1 ± 17.0	38.4 ± 18.4
Renal disorder with metabolic disease	56.9 ± 12.3	57.9 ± 8.9	57.2 ± 11.5	56.8 ± 14.8	54.8 ± 14.1	56.2 ± 14.5	56.9 ± 13.5	56.2 ± 11.9	56.7 ± 13.0
Acute nephritic syndrome	42.8 ± 19.2	36.0 ± 22.5	39.9 ± 20.7	49.6 ± 17.5	46.6 ± 21.1	48.1 ± 19.3	46.1 ± 18.5	42.0 ± 22.1	44.2 ± 20.3
Hypertensive nephropathy	56.2 ± 13.5	51.0 ± 15.3	55.2 ± 13.8	54.5 ± 15.9	54.7 ± 17.0	54.6 ± 16.0	55.3 ± 14.8	53.3 ± 16.1	54.8 ± 15.1
Acute renal failure	56.0 ± 19.3	56.4 ± 26.2	56.1 ± 21.2	55.2 ± 17.6	58.0 ± 20.6	56.0 ± 18.2	55.6 ± 18.3	57.1 ± 23.1	56.0 ± 19.7
Drug-induced nephropathy	53.6 ± 11.9	35.2 ± 21.6	45.1 ± 18.9	47.3 ± 20.0	60.4 ± 17.6	51.5 ± 19.9	49.1 ± 18.0	49.6 ± 22.7	49.3 ± 19.5
Inherited renal disease	25.0 ± 23.8	40.7 ± 24.1	32.8 ± 23.1	15.0 ± 17.1	24.3 ± 25.3	19.3 ± 21.1	17.7 ± 18.5	29.2 ± 24.9	23.2 ± 22.0
HUS/TTP	–	–	–	10, 69	49	42.6 ± 30.0	10, 69	49	42.6 ± 30.0
Others	50.6 ± 18.2	48.4 ± 19.5	49.6 ± 18.7	48.6 ± 20.9	53.3 ± 18.1	50.5 ± 19.8	49.4 ± 19.6	50.9 ± 18.9	50.0 ± 19.2
Total	48.4 ± 20.0	45.5 ± 20.0	47.0 ± 20.1	48.2 ± 21.0	46.0 ± 20.5	47.1 ± 20.8	48.3 ± 20.6	45.8 ± 20.3	47.1 ± 20.5

in 2009 and 2010. The distribution of chronic kidney disease (CKD) stages, degree of proteinuria and clinical parameters in IgAN were analyzed in the combined data from 2009 and 2010 (Tables 18, S2, S3).

With regard to the stages of CKD in patients with IgAN, stage 2 was predominant in the combined data from 2009 and 2010 (Table 18) and in both genders (Tables S2 and S3). The degree of proteinuria in the 24-h urine or spot urine samples increased with the progression of CKD stages in the combined data from 2009 and 2010 (Table 18) and in both genders (Tables S2 and S3). The systolic and diastolic blood pressure also increased with the progression of the CKD stage (Tables 18, S2, S3). Overall, 37.0 % of IgAN cases were being treated with anti-hypertensive agents and 4.6 % had diabetes mellitus (Table 18).

Cases in the J-KDR not reported in the J-RBR

In cases in the J-KDR not reported in the J-RBR, a clinical diagnosis of chronic nephritic syndrome was predominant in 2009, followed by hypertensive nephropathy, and a clinical diagnosis of renal disorder with metabolic disease (diabetic nephropathy) was predominant in 2010, followed by nephrotic syndrome (Table 19). Polycystic kidney disease was detected in 2010 as a result of the secondary research studies performed on the basis of the J-KDR as described in the "Subjects and methods" section.

Secondary and longitudinal research by the J-RBR/J-KDR

Five of the secondary and longitudinal research studies, viz., the JNSCS, J-IDCS, J-IGACS, JRPGN-CS, and JDNCS, were started in 2009, and the J-PKD was started in 2010 in association with the J-RBR/J-KDR.

Discussion and comments

In 2009, the J-KDR started to register clinically-diagnosed cases without renal biopsies, in addition to cases with renal biopsies included in the J-RBR, which had been started in 2007. More than 80 % of the registered cases were in the J-RBR in 2009 and 2010, and thus the detailed data from the J-RBR and the clinical diagnosis alone for the J-KDR are described in this report.

Table 6 The frequency of pathological diagnoses as classified by pathogenesis in J-RBR 2009 and 2010

Classification	2009			2010			Total		
	Total biopsies (n = 3,336)		Native kidneys (n = 3,165)	Total biopsies (n = 4,106)		Native kidneys (n = 3,869)	Total biopsies (n = 7,442)		Native kidneys (n = 7,034)
	n	%	%[a]	n	%	%[a]	n	%	%[a]
IgA nephropathy	1,003	30.1	31.6	1,177	28.7	30.4	2,180	29.3	31.0
Primary glomerular disease (except IgA nephropathy)	862	25.8	27.2	1,090	26.5	28.1	1,952	26.2	27.7
Diabetic nephropathy	184	5.5	5.8	192	4.7	5.0	376	5.1	5.3
Renal graft	161	4.8	–	235	5.7	–	396	5.3	–
Lupus nephritis	137	4.1	4.3	220	5.4	5.7	357	4.8	5.1
MPO-ANCA positive nephritis	129	3.9	4.1	191	4.7	4.9	320	4.3	4.5
Hypertensive nephrosclerosis	123	3.7	3.9	157	3.8	4.1	280	3.8	4.0
Purpura nephritis	64	1.9	2.0	108	2.6	2.8	172	2.3	2.4
Amyloid nephropathy	45	1.3	1.4	58	1.4	1.5	103	1.4	1.5
Infection-related nephropathy	27	0.8	0.9	31	0.8	0.8	58	0.8	0.8
Thin basement membrane disease	26	0.8	0.8	39	1.0	1.0	65	0.9	0.9
PR3-ANCA positive nephritis	13	0.4	0.4	11	0.3	0.3	24	0.3	0.3
Alport syndrome	10	0.3	0.3	16	0.4	0.4	26	0.3	0.4
Thrombotic microangiopathy	9	0.3	0.3	8	0.2	0.2	17	0.2	0.2
Anti-GBM antibody-type nephritis	8	0.2	0.3	16	0.4	0.4	24	0.3	0.3
Others	535	16.0	16.7	557	13.6	13.6	1,092	14.7	15.4
Total	3,336	100.0	100.0	4,106	100.0	100.0	7,442	100.0	100.0

MPO myeloperoxidase, *ANCA* anti-neutrophil cytoplasmic antibody, *PR3* proteinase 3, *GBM* glomerular basement membrane

[a] Patients classified as either "Renal graft" or "Renal transplantation" in other categories were also excluded

The rates of primary glomerular disease (except IgAN) combined with that of IgAN in native renal biopsies were 59.3 %, 56.5 %, 58.8 %, and 58.5 % in 2007, 2008, 2009, and 2010 in the J-RBR. A recent report from a single center in Japan gave the rates as 77.8 % and 75.9 % between 1979 and 2008 and between 2004 and 2008, respectively [5]. In the present report for the J-RBR, the peak distribution of age was in the sixties in the combined data for 2009 and 2010. The difference in the rates of primary glomerular disease including IgAN may have been due to the higher mean ages of native biopsy cases in the J-RBR compared to the single center in this period (mean age, 46.7 vs. 40.8 years; age of the peak number, sixties vs. twenties), because the incidence of secondary glomerular disease increases in elderly patients, as reported previously [5].

IgAN is still the most frequently diagnosed disease in native kidney biopsies in Japan (33.0 %, 30.2 %, 31.6 %, and 30.4 % of cases in 2007, 2008, 2009, and 2010 in the J-RBR) [1, 4–6] similar to other Asian countries [7, 8] and some European countries [9, 10]. The peak distribution of age ranges was the twenties in 2009 and thirties in 2010. In patients with IgAN, the majority (68.1 %) of renal biopsies were performed in CKD stages G1 and G2, with median proteinuria less than 1 g per day (Table 18), suggesting that there was a relatively early diagnosis of this biopsy-proven disease.

In the present clinical data, the degree of proteinuria increased with the progression of the CKD stage, and was more than 1 g per day for the median value in patients with CKD stages G4 and G5 (Tables 18, S1, S2). Previously, the best single predictor for renal deterioration was severe proteinuria on urine dipstick testing (≥100 mg/dL), followed by hypoalbuminemia, mild hematuria, serum total protein levels, diastolic blood pressure, and histological grade, in a cohort study with 10 years follow-up from 1995 in Japan, the cohort of which exhibited a younger median age (27.7 years) and a peak distribution of age ranges in the teens [11, 12]. A recent report suggested that IgAN with nephrotic syndrome had a worse renal outcome compared to IgAN with non-nephrotic syndrome unless partial or complete remission was achieved [13]. Further studies are necessary to elucidate the risk factors or predictors for renal deterioration in IgAN in the present era utilizing the J-RBR, possibly as part of a new secondary clinical study.

MN was the most common histopathology in terms of primary glomerular disease other than IgAN in 2007

Table 7 The frequency of pathological diagnoses as classified by histopathology in J-RBR 2009 and 2010

Classification	2009			2010			Total		
	Total biopsies (n = 3,336)		Native kidneys (n = 3,165)	Total biopsies (n = 4,106)		Native kidneys (n = 3,869)	Total biopsies (n = 7,442)		Native kidneys (n = 7,034)
	n	%	%[a]	n	%	%[a]	n	%	%[a]
Mesangial proliferative glomerulonephritis	1,346	40.3	42.5	1,388	33.8	35.8	2,734	36.7	38.8
Membranous nephropathy	333	10.0	10.5	418	10.2	10.8	751	10.1	10.7
Minor glomerular abnormality	293	8.8	9.2	559	13.6	14.4	852	11.4	12.1
Crescentic and necrotizing glomerulonephritis	180	5.4	5.7	262	6.4	6.8	442	5.9	6.3
Focal segmental glomerulosclerosis	167	5.0	5.2	211	5.1	5.4	378	5.1	5.3
Nephrosclerosis	163	4.9	5.2	208	5.1	5.4	371	5.0	5.3
Renal graft	151	4.5	–	227	5.5	–	378	5.1	–
Membranoproliferative glomerulonephritis (types I and III)	85	2.5	2.7	97	2.4	2.5	182	2.4	2.6
Chronic interstitial nephritis	71	2.1	2.1	61	1.5	1.6	132	1.7	1.8
Sclerosing glomerulonephritis	63	1.9	2.0	44	1.1	1.1	107	1.4	1.5
Endocapillary proliferative glomerulonephritis	61	1.8	1.9	67	1.6	1.7	128	1.7	1.8
Acute interstitial nephritis	45	1.3	1.4	62	1.5	1.6	107	1.4	1.5
Acute tubular necrosis	9	0.3	0.3	10	0.2	0.2	19	0.3	0.2
Dense deposit disease	3	0.1	0.1	5	0.1	0.1	8	0.1	0.1
Others	366	11.0	11.3	487	11.9	12.5	853	11.5	12.0
Total	3,336	100.0	100.0	4,106	100.0	100.0	7,442	100.0	100.0

[a] Patients classified as either "Renal graft" or "Renal transplantation" in other categories were also excluded

Table 8 The frequency of pathological diagnoses as classified by histopathology in primary glomerular disease except IgA nephropathy in native kidneys in J-RBR 2009 and 2010

Classification	2009		2010		Total	
	n	%	n	%	n	%
Membranous nephropathy	259	30.1	330	30.3	589	30.2
Minor glomerular abnormalities	216	25.1	408	37.5	624	32.0
Mesangial proliferative glomerulonephritis	167	19.4	86	7.9	253	13.0
Focal segmental glomerulosclerosis	113	13.1	149	13.7	262	13.4
Membranoproliferative glomerulonephritis (types I and III)	48	5.6	51	4.7	99	5.1
Crescentic and necrotizing glomerulonephritis	19	2.2	18	1.7	37	1.9
Endocapillary proliferative glomerulonephritis	8	0.9	24	2.2	32	1.6
Chronic interstitial nephritis	7	0.8	3	0.3	10	0.5
Sclerosing glomerulonephritis	7	0.8	3	0.3	10	0.5
Nephrosclerosis	5	0.6	7	0.6	12	0.6
Acute interstitial nephritis	1	0.1	0	–	1	0.1
Acute tubular necrosis	0	–	1	0.1	1	0.1
Others	11	1.3	9	0.8	20	1.0
Total	861	100.0	1,089	100.0	1,950	100.0

Table 9 The frequency of pathological diagnoses as classified by pathogenesis in nephrotic syndrome in native kidneys in J-RBR 2009 and 2010

Classification	2009		2010		Total	
	n	%	*n*	%	*n*	%
Primary glomerular disease (except IgA nephropathy)	442	62.3	696	66.7	1,138	64.9
Diabetic nephropathy	85	12.0	78	7.5	163	9.3
IgA nephropathy	30	4.2	36	3.5	66	3.8
Lupus nephritis	30	4.2	58	5.6	88	5.0
Amyloid nephropathy	27	3.8	41	3.9	68	3.9
Infection-related nephropathy	6	0.8	7	0.7	13	0.7
Hypertensive nephrosclerosis	6	0.8	10	0.9	16	0.9
Purpura nephritis	4	0.6	8	0.8	12	0.7
Alport syndrome	3	0.4	0	–	3	0.2
Thrombotic microangiopathy	1	0.1	1	0.1	2	0.1
PR3-ANCA positive nephritis	1	0.1	0	–	1	0.1
MPO-ANCA positive nephritis	1	0.1	2	0.2	3	0.2
Others	74	10.4	106	10.2	180	10.3
Total	710	100.0	1,043	100.0	1,753	100.0

MPO myeloperoxidase, *ANCA* anti-neutrophil cytoplasmic antibody, *PR3* proteinase 3

Table 10 The frequency of pathological diagnoses as classified by histopathology in primary glomerular disease except IgA nephropathy in nephrotic syndrome in native kidneys in J-RBR 2009 and 2010

Classification	2009		2010		Total	
	n	%	*n*	%	*n*	%
Membranous nephropathy	178	40.3	227	32.6	405	35.6
Minor glomerular abnormalities	172	38.9	348	50.0	520	45.7
Focal segmental glomerulosclerosis	47	10.6	82	11.8	129	11.3
Membranoproliferative glomerulonephritis (types I and III)	25	5.7	18	2.6	43	3.8
Mesangial proliferative glomerulonephritis	11	2.5	13	1.9	24	2.1
Crescentic and necrotizing glomerulonephritis	2	0.5	2	0.3	4	0.4
Sclerosing glomerulonephritis	2	0.5	0	–	2	0.2
Endocapillary proliferative glomerulonephritis	1	0.2	5	0.7	6	0.5
Others	4	0.9	1	0.1	5	0.4
Total	442	100.0	696	100.0	1,138	100.0

(31.4 %), 2008 (25.7 %), and 2009 (30.1 %) in the J-RBR and was also the most common type in primary nephrotic syndrome in 2007 (44.0 %) and 2009 (40.3 %) in the J-RBR. MN was also the most common primary cause of nephrotic syndrome in a northern European Caucasian population, with a biopsy rate of 4.5 per million population per year [14]. A total of 68.7 % and 68.8 % of primary MN cases exhibited nephrotic syndrome as the clinical diagnosis at the time of renal biopsy in 2009 and 2010 in the J-RBR. Yokoyama et al. recently reported in their clinical data analysis of 501 cases collected from the combined data of the J-RBR from 2007 to 2010 that nearly half of primary MN (49.1 %) cases showed a daily proteinuria of 3.5 g or higher [15]. The renal survival rate was 60 % at 20 years after diagnosis in patients with primary MN, and the renal survival rate in patients on steroid therapy was significantly higher in patients on supportive therapy alone in Japan [16], while spontaneous remission was reported to be common (32 %) in patients with primary MN with nephrotic syndrome in Spain [17], even in patients exhibiting chronic renal impairment [18]. Whether treatment with renin–angiotensin blockers or immunoglobulins other than steroids has a favorable effect on the renal prognosis of primary MN should be elucidated in future clinical studies.

The minor glomerular abnormalities in primary nephrotic syndrome, which correspond to MCNS, was the most common histopathology reported in 2008 (44.1 %) and 2010 (50.0 %) in the J-RBR. Since MCNS develops in patients at younger ages [5, 15] while primary MN develops in a relatively elderly population [15, 16], the frequency of these diseases may depend on the distribution of

Table 11 The frequency of clinical diagnoses in membranous nephropathy in primary glomerular disease except IgA nephropathy in native kidneys in J-RBR 2009 and 2010

Classification	2009		2010		Total	
	n	%	*n*	%	*n*	%
Nephrotic syndrome	178	68.7	227	68.8	405	68.8
Chronic nephritic syndrome	74	28.6	93	28.2	167	28.4
Recurrent or persistent hematuria	3	1.2	3	0.9	6	1.0
Renal disorder with collagen disease or vasculitis	1	0.4	1	0.3	2	0.3
Hypertensive nephropathy	1	0.4	0	–	1	0.2
Rapidly progressive nephritic syndrome	0	–	1	0.3	1	0.2
Renal disorder with metabolic disease	0	–	1	0.3	1	0.2
Acute nephritic syndrome	0	–	1	0.3	1	0.2
Acute renal failure	0	–	1	0.3	1	0.2
Others	2	0.8	2	0.6	4	0.7
Total	259	100.0	330	100.0	589	100.0

Table 12 The frequency of clinical diagnoses in minor glomerular abnormalities in primary glomerular disease except IgA nephropathy in native kidneys in J-RBR 2009 and 2010

Classification	2009		2010		Total	
	n	%	*n*	%	*n*	%
Nephrotic syndrome	172	79.6	348	85.3	520	83.3
Chronic nephritic syndrome	35	16.2	50	12.3	85	13.6
Recurrent or persistent hematuria	5	2.3	5	1.2	10	1.6
Acute renal failure	1	0.5	0	–	1	0.2
Rapidly progressive nephritic syndrome	1	0.5	1	0.2	2	0.3
Acute nephritic syndrome	1	0.5	1	0.2	2	0.3
Hypertensive nephropathy	0	–	1	0.2	1	0.2
Others	1	0.5	2	0.5	3	0.5
Total	216	100.0	408	100.0	624	100.0

the age ranges of patients registered in each year. Indeed, the rate of native biopsies of subjects younger than 20 years of age slightly increased from 11.4 % in 2009 to 12.7 % in 2010 (Table 3) and the mean age of patients with nephrotic syndrome slightly decreased from 53.5 years in 2009 to 50.1 years in 2010 (Table 5) in the J-RBR.

The average age of rapidly progressive nephritic syndrome was the highest (64.4 years) in the age distribution in the classification of clinical diagnosis in the J-RBR (Table 5). Elderly subjects (65 years and over) comprised nearly 25 % of cases, and very elderly subjects (80 years and over) comprised 2.5 % of the cases in the combined data for 2009 and 2010 in the J-RBR. It has been reported that there were statistically significant differences in the renal disease spectrum between elderly and younger subjects [19, 20]. The frequency of rapidly progressive nephritic syndrome in the clinical diagnosis dramatically increased from 4.0 % in the younger group (20–64 years) to 19.6 % in the very elderly in the combined data from 2007 to November 2011 in the J-RBR [20]. A nationwide survey of rapidly progressive glomerulonephritis (RPGN) was conducted between 1989 and 2007 in Japan, and showed that 64.0 % of patients had pauci-immune-type RPGN, including 42.0 % renal-limited vasculitis, 19.4 % microscopic polyangiitis, and 2.6 % Wegener's granulomatosis (currently granulomatosis with polyangiitis) [21]. Since the frequency of myeloperoxidase–anti-neutrophil cytoplasmic antibody (MPO-ANCA)-positive nephritis has increased recently [22], a further subanalysis of rapidly progressive nephritic syndrome in the J-RBR should be performed to validate the recently published Japanese guidelines for RPGN [23].

Five new secondary research studies of the J-KDR were started in 2009, viz., the J-NSCS, J-IDCS, J-IGACS, J-RPGNCS, and J-DNCS, and the J-PKD was started in 2010. The J-RBR and J-KDR initiated two more clinical

Table 13 The frequency of clinical diagnoses in focal segmental glomerulosclerosis in primary glomerular disease except IgA nephropathy in native kidneys in J-RBR 2009 and 2010

Classification	2009		2010		Total	
	n	%	*n*	%	*n*	%
Chronic nephritic syndrome	62	54.9	55	36.9	117	44.7
Nephrotic syndrome	47	41.6	82	55.0	129	49.2
Rapidly progressive nephritic syndrome	1	0.9	1	0.7	2	0.8
Renal disorder with metabolic disease	1	0.9	3	2.0	4	1.5
Recurrent or persistent hematuria	1	0.9	1	0.7	2	0.8
Hypertensive nephropathy	0	–	2	1.3	2	0.8
Acute nephritic syndrome	0	–	1	0.7	1	0.4
Inherited renal disease	0	–	1	0.7	1	0.4
Others	1	0.9	3	2.0	4	1.5
Total	113	100.0	149	100.0	262	100.0

Table 14 The profile of IgA nephropathy in native kidneys in J-RBR 2009 and 2010

IgA nephropathy	2009	2010	Total
Total native kidney biopsies (*n*)	1,001	1,176	2,177
Average age (years)	38.1 ± 17.2	39.3 ± 17.0	38.7 ± 17.1
Median age (years)	35 (24–52)	38 (26–53)	37 (25–52)
Male, *n* (%)	498 (49.8 %)[a]	585 (49.7 %)	1,083 (49.7 %)
Average age (years)	39.5 ± 18.2[b]	40.5 ± 18.4[b]	40.0 ± 18.3[b]
Median age (years)	38 (24–55)[b]	39 (25–56)	38 (24–56)[b]
Female, *n* (%)	503 (50.2 %)[a]	591 (50.3 %)	1,094 (50.3 %)
Average age	36.6 ± 15.9[b]	38.1 ± 15.4[b]	37.5 ± 15.7[b]
Median age	34 (24–49)[b]	37 (26–49)	36 (25–49)[b]

[a] Ratio indicates percentage of each gender in each biopsy category

[b] $P < 0.05$ compared to other gender

Table 15 Distribution of age ranges and gender in IgA nephropathy in J-RBR in 2009 and 2010

Age (years)	2009			2010			Total		
	Male	Female	Total	Male	Female	Total	Male	Female	Total
0–9	11	5	16	12	9	21	23	14	37
10–19	73	68	141	80	55	135	153	123	276
20–29	91	116	207	91	127	218	182	243	425
30–39	87	115	202	113	153	266	200	268	468
40–49	65	81	146	94	106	200	159	187	346
50–59	87	62	149	84	75	159	171	137	308
60–69	62	45	107	82	48	130	144	93	237
70–79	19	9	28	20	18	38	39	27	66
80+	3	2	5	9	0	9	12	2	14
Total	498	503	1,001	585	591	1,176	1,083	1,094	2,177
Under 20 (%)	16.9	14.5	15.7	15.7	10.8	13.3	16.3	12.5	14.4
65 and over (%)	9.4	5.2	7.3	11.5	5.4	8.4	10.5	5.3	7.9

Table 16 The frequency of classification of clinical diagnoses in IgA nephropathy in native kidneys in J-RBR 2009 and 2010

Clinical diagnosis	2009		2010		Total	
	n	%	*n*	%	*n*	%
Chronic nephritic syndrome	886	88.5	1,064	90.5	1,950	89.6
Recurrent or persistent hematuria	49	4.9	40	3.4	89	4.1
Nephrotic syndrome	30	3.0	36	3.1	66	3.0
Rapidly progressive nephritic syndrome	14	1.4	20	1.7	34	1.6
Acute nephritic syndrome	8	0.8	9	0.8	17	0.8
Renal disorder with collagen disease or vasculitis	4	0.4	1	0.1	5	0.2
Acute renal failure	2	0.2	2	0.2	4	0.2
Drug-induced nephropathy	2	0.2	1	0.1	3	0.1
Renal disorder with metabolic disease	1	0.1	0	–	1	0.0
Hypertensive nephropathy	0	–	1	0.1	1	0.0
Others	5	0.5	2	0.2	7	0.3
Total	1,001	100.0	1,176	100.0	2,177	100.0

Table 17 The frequency of pathological diagnoses as classified by histopathology in IgAN in native kidneys in J-RBR 2009 and 2010

Pathological diagnosis by histopathology	2009		2010		Total	
	n	%	*n*	%	*n*	%
Mesangial proliferative glomerulonephritis	937	93.6	1,111	94.5	2,048	94.1
Endocapillary proliferative glomerulonephritis	12	1.2	2	0.2	14	0.6
Minor glomerular abnormalities	12	1.2	15	1.3	27	1.2
Focal segmental glomerulosclerosis	9	0.9	6	0.5	15	0.7
Crescentic and necrotizing glomerulonephritis	8	0.8	10	0.9	18	0.8
Nephrosclerosis	6	0.6	4	0.3	10	0.5
Membranous nephropathy	4	0.4	2	0.2	6	0.3
Membranoproliferative glomerulonephritis (types I and III)	4	0.4	5	0.4	9	0.4
Sclerosing glomerulonephritis	3	0.3	2	0.2	5	0.2
Chronic interstitial nephritis	1	0.1	2	0.2	3	0.1
Acute interstitial nephritis	0	–	1	0.1	1	0.0
Others	5	0.5	16	1.4	21	1.0
Total	1,001	100.0	1,176	100.0	2,177	100.0

research studies (J-RBR201001 and J-KDR201001) being performed by members of the JSN who had already participated in the registry and who registered cases under the precise regulations presented on the website of the JSN in 2011.

With regard to estimating the number of yearly native renal biopsies in Japan, the Research Group on Progressive Renal Disease from the Ministry of Health, Labor and Welfare of Japan recently reported by a questionnaire method that it was between 18,000 and 21,000 in 2010. The J-RBR may cover nearly one fourth to one fifth of the number of yearly native renal biopsies in Japan in 2010. Since 128,057,352 people resided in Japan in 2010, the estimated rate of renal biopsy was 140.6 to 164.0 per million population. This rate was higher than that in Romania [24], Spain [25], the Czech Republic [10], Denmark [26], and Scotland [27], was similar to that in France [28], and was lower than that in USA, Finland [29], and Australia [30].

There are some limitations in the J-RBR and J-KDR. The J-RBR records three diagnoses for each case, viz., the clinical diagnosis, diagnosis based on the pathogenesis, and the diagnosis based on a histopathological examination, so there may be still some inconsistency in the case records. The terms hypertensive nephropathy, hypertensive nephrosclerosis, nephrosclerosis, and diabetic nephropathy may need to be defined more precisely to improve the accuracy of the report by the J-RBR. The incidence of renal biopsy

Table 18 Distribution of CKD stages and clinical parameters in total in IgA nephropathy in J-RBR: Combined data of 2009 and 2010

	CKD stage					Total	*P* value*
	G1	G2	G3a/b	G4	G5		
Total	663	814	551	111	30	2,169	–
n (%)	30.6	37.5	25.4	5.1	1.4	100.0	–
Age (years), average	23.5 ± 10.9	40.3 ± 13.5	50.9 ± 13.0	55.7 ± 16.2	46.3 ± 20.4	38.7 ± 17.1	<0.0001
Median	22 (17–29)	38 (30–50)	52 (42–61)	59 (44–68)	46 (29–62)	37 (25–52)	<0.0001
Body mass index	21.0 ± 4.0	22.9 ± 3.8	23.6 ± 3.7	23.0 ± 4.5	23.4 ± 5.9	22.5 ± 4.0	<0.0001
Estimated GFR (mL/min/1.73 m^2)	108.2 (96.9–128.0)	75.2 (67.8–82.7)	49.1 (42.0–54.6)	23.6 (20.9–27.6)	8.5 (6.1–12.0)	74.6 (53.8–95.0)	<0.0001
Proteinuria (g/day)	0.30 (0.10–0.81)	0.50 (0.21–1.00)	0.92 (0.40–2.00)	1.60 (0.71–2.84)	2.81 (1.17–4.58)	0.59 (0.22–1.29)	<0.0001
Proteinuria (g/gCr)	0.39 (0.14–0.91)	0.63 (0.28–1.23)	1.03 (0.51–2.01)	1.69 (0.77–4.21)	2.91 (1.30–4.58)	0.70 (0.27–1.47)	<0.0001
Sediment RBC ≥*5/hpf (%)*	82.4	81.3	74.6	82.0	86.7	80.0	0.0075
Serum creatinine (mg/dL)	0.60 (0.53–0.70)	0.79 (0.70–0.91)	1.16 (1.00–1.36)	2.10 (1.86–2.47)	5.34 (4.06–7.66)	0.81 (0.65–1.07)	<0.0001
Serum albumin (g/dL)	4.15 ± 0.46	4.02 ± 0.49	3.79 ± 0.59	3.45 ± 0.63	3.22 ± 0.59	3.96 ± 0.56	<0.0001
Serum total cholesterol (mg/dL)	184.6 ± 37.4	204.3 ± 46.2	209.9 ± 51.1	211.6 ± 52.3	221.0 ± 58.6	200.2 ± 46.8	<0.0001
Systolic BP (mmHg)	113.9 ± 14.0	123.3 ± 16.2	130.3 ± 17.5	137.6 ± 22.5	147.5 ± 27.9	123.2 ± 18.1	<0.0001
Diastolic BP (mmHg)	67.6 ± 11.4	75.1 ± 12.3	78.9 ± 12.5	81.0 ± 15.6	87.8 ± 18.0	74.2 ± 13.3	<0.0001
Anti-hypertensive agents (%)	13.8	33.3	59.6	75.8	71.4	37.0	<0.0001
Diabetes mellitus (%)	1.5	3.1	7.7	21.1	0.0	4.6	<0.0001

Data are presented as the mean ± SD or the medians (interquartile ranges)

CKD chronic kidney disease, *GFR* glomerular filtration rate, *RBC* red blood cell count, *BP* blood pressure

* ANOVA, Kruskal–Wallis or χ^2-test as appropriate. There are eight (0.4 %) missing values of CKD stage because of inappropriate data for serum creatinine

Table 19 The frequency of classification of clinical diagnoses in other 680 cases than J-RBR in J-KDR 2009 and 2010

Classification	Other cases 2009 (n = 680)		Other cases 2010 (n = 575)		Total (n = 1,255)	
	n	%	n	%	n	%
Chronic nephritic syndrome	165	24.3	72	12.5	237	18.9
Hypertensive nephropathy	142	20.9	43	7.5	185	14.7
Renal disorder with metabolic disease	106	15.6	177	30.8	283	22.5
Nephrotic syndrome	86	12.6	118	20.5	204	16.3
Renal disorder with collagen disease or vasculitis	24	3.5	7	1.2	31	2.5
Rapidly progressive nephritic syndrome	21	3.1	18	3.1	39	3.1
Inherited renal disease	18	2.6	3	0.5	21	1.7
Acute renal failure	9	1.3	10	1.7	19	1.5
Recurrent or persistent hematuria	8	1.2	0	–	8	0.6
Acute nephritic syndrome	5	0.7	4	0.7	9	0.7
Drug-induced nephropathy	5	0.7	0	–	5	0.4
Renal transplantation	2	0.3	9	1.6	11	0.9
Polycystic kidney disease	–	–	82	14.3	82	6.5
Others	89	13.1	32	5.6	121	9.6
Total	680	100.0	575	100.0	1,255	100.0

and the incidence of biopsy-proven renal diseases such as IgAN and primary glomerular disease (except IgAN) could be surveyed in major renal centers in Japan in terms of the epidemiological aspects to work out appropriate countermeasures. In this aspect, the incidence of pediatric IgAN was reported to be 4.5 cases/year per 100,000 children under 15 years of age from 1983 to 1999 in Yonago City, Japan [31], although center variations in the country in terms of the incidence, indications and diagnosis of adult native renal biopsy have been reported [27].

Finally, a committee report of J-KDR including J-RBR in 2009, 2010 and their total was conducted. The J-RBR exhibited the majority of the registry system to elucidate yearly demographic data of renal biopsies in Japan, and J-KDR was utilized to promote advanced clinical research in the field of nephrology in our country.

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ORIGINAL ARTICLE

Renal disease in the elderly and the very elderly Japanese: analysis of the Japan Renal Biopsy Registry (J-RBR)

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Abstract

Background and objectives Data regarding renal disease in the elderly (age ≥65 years old) and very elderly (age ≥80 years old) Japanese are extremely limited. The aim of this study was to examine the causes of renal disease and their clinical presentations in elderly patients who underwent renal biopsy.

Design, setting, participants, and measurements From July 2007 to November 2011, all of the elderly native renal biopsy patients who had been registered in the Japan Renal Biopsy Registry (J-RBR; 2802 including 1596 males and 1206 females) were identified. Their data were compared with a control group of 7416 patients who ranged in age from 20 to 64 years old and were registered on the J-RBR

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over the same period. In addition, the clinical and pathological classifications of 276 very elderly patients were also analyzed.

Results The indications for biopsy were nephrotic syndrome (NS) in 36.2 and 50.7 % of the elderly and the very elderly patients, chronic nephritic syndrome in 31.8 and 17.4 %, and acute kidney injury including rapidly progressive glomerulonephritis in 18.6 and 22.5 %, respectively. Primary glomerular disease was the most frequent diagnosis, followed by MPO-ANCA-positive nephritis, IgA nephropathy (IgAN), and diabetic nephropathy. In primary GN including IgAN, membranous nephropathy (MN) was the most frequent histological type, followed by IgAN and minor glomerular abnormalities. A comparison with the control group showed that MN, MPO-ANCA-positive nephritis, and amyloid nephropathy were more common in the elderly ($P < 0.001$), and IgAN was less common ($P < 0.001$). As for nephrotic syndrome in the elderly, MN was the most common histological type, followed by minimal change NS, diabetic nephropathy, amyloid nephropathy, and focal segmental glomerulosclerosis. There was a significant discrepancy between the urinary protein/creatinine ratio and daily proteinuria after the 7th decade of life.

Conclusions Renal biopsy is a valuable diagnostic tool, even in elderly and very elderly Japanese patients. In the future, modified clinical guidelines for elderly renal disease should be developed.

Keywords Elderly · Very elderly · Japanese · Renal biopsy · Registry · Nephrotic syndrome · IgA nephropathy · Rapidly progressive glomerulonephritis · Proteinuria

Introduction

In Japan, the elderly population; i.e., those aged 65 and over, accounted for 25.8 % of the total population in October 2010, and this will increase to 30.5 % by 2025 [1]. As life expectancy increases, more elderly patients with acute and chronic renal diseases are surviving longer. In particular, the progressive decline in GFR that occurs with age; the decline in GFR brought about by cardiovascular and other systemic diseases, such as anti-neutrophil cytoplasmic antibody (ANCA)-positive vasculitis; and reductions in GFR due to the nephrotoxic effects of medical/surgical treatment are expected to contribute to an increased incidence of renal disease in the elderly population. Several studies involving limited numbers of elderly Japanese patients have suggested that renal biopsy can provide significant diagnostic and prognostic information

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