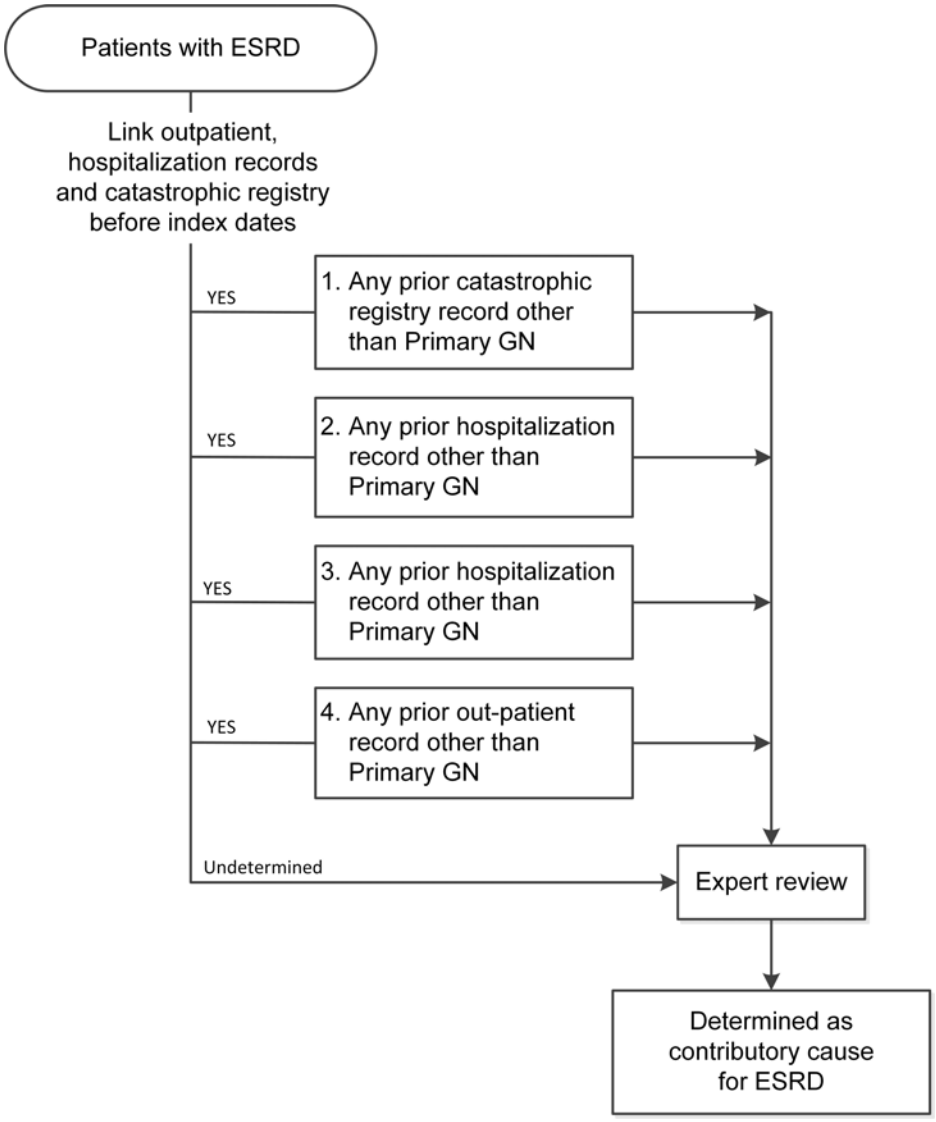
Supplementary Files

Appendix 1. Algorithm to determine the causes of end-stage renal disease (ESRD) in the young (age <30 years old) in the period 1998-2009



The process of Expert review:

Possible causes were identified by diagnosis codes from complete medical claims, out-patient visits, and hospitalization records linked and traced back to the date of birth or one year before the study period. To ensure the validity of the index date, the patients were followed-up for at least one year, those with incomplete observation were not included in renal survival analysis. The differentiation algorithm automatically sorted the possible causes of ESRD for each patient by date and importance. After the automatic aggregation process, a pediatric nephrologist then reviewed the medical claims and determined only one valid contributory cause for ESRD per patient. Followings

are few examples of automatic aggregations, the interpretation and the result of expert reviewing process.

Examples records for expert review:
Each row represents a number of records with different source (field in green: cd for Outpatient visits, hv for catastrophic registry, dd for hospitalization records) and the number of records (field blue).

1. An example of SLE related ESRD

Sex	Age ESRD	src cnt	l1		First Dx	Last Visit	ESRD date
F	28	cd 25	GN	Miscellaneous glomerulonephritis	200904--	200907--	200904--
F	28	hv 3	GN	Miscellaneous glomerulonephritis	200904--	200907--	200904--
F	28	dd 6	GN	Systemic lupus erythematous	200802--	200907--	200904--
F	28	dd 9	GN	Miscellaneous glomerulonephritis	200802--	200907--	200904--
F	28	hv 14	GN	Systemic lupus erythematous	200004--	200511--	200904--
F	28	cd 88	GN	Systemic lupus erythematous	200003--	200907--	200904--

[Expert review]
This case was diagnosed as SLE in 2000 and she received catastrophic illness card of SLE in June of the same year. Then she came to outpatient department and hospitalization for problems about SLE and renal dysfunction (CGN) respectively. She was coded as SLE by rheumatologists and as CGN by nephrologists during these years. It was not until 2009 she got another catastrophic illness card given by the nephrologist with CGN, which should be actually lupus nephritis. According to the behavior of doctors under NHI, the expert reviewed her recordings and concluded her cause and renal survival of ESRD.
[Result]: The case was classified as SLE related ESRD.

2. An example of Wegener's granulomatosis related ESRD

Sex	Age ESRD	src cnt	l1		First Dx	Last Visit	ESRD date
M	20	hv 9	GN	Miscellaneous glomerulonephritis	200810--	201002--	200810--
M	20	dd 10	GN	Miscellaneous glomerulonephritis	200607--	200906--	200810--
M	20	cd 62	GN	Miscellaneous glomerulonephritis	200503--	200910--	200810--
M	20	cd 299	GN	Wegener's granulomatosis	200412--	200910--	200810--
M	20	hv 12	GN	Wegener's granulomatosis	200411--	200411--	200810--
M	20	dd 14	GN	Wegener's granulomatosis	200410--	200904--	200810--

[Expert review]
This patient was hospitalization for 14 times and admitted for 299 times from 2004 to 2009 with diagnosis as Wegener's granulomatous and got identification of catastrophic illness. However, he was referred for renal insufficiency and was diagnosed by nephrologists as CGN. His renal disease came into end-stage in 20081027.
[Result] This patient was classified as Wegener's granulomatous related ESRD.

3. An example of cystic kidney related ESRD

Sex	Age ESRD	src cnt	l1		First Dx	Last Visit	ESRD date
M	10	hv 6	GN	Miscellaneous glomerulonephritis	200712--	200712--	200712--
M	10	cd 55	Congential (CAKUT)	Specified anomalies of kidney	200403--	200512--	200712--
M	10	dd 6	GN	Miscellaneous glomerulonephritis	200402--	200805--	200712--
M	10	cd 207	GN	Miscellaneous glomerulonephritis	200004--	200812--	200712--
M	10	hv 13	Congential (CAKUT)	Cystic kidney	200003--	200303--	200712--
M	10	cd 244	Congential (CAKUT)	Cystic kidney	199911--	200802--	200712--
M	10	dd 8	Congential (CAKUT)	Cystic kidney	199911--	200804--	200712--

[Expert review]

The boy was diagnosed as cystic kidney since 1999 and progressed into ESRD in 2007. The medical claims from catastrophic registry (hv), outpatient visits (cd), and hospitalization (dd) consistently suggest a diagnosis of “cystic kidney”. Although more diagnosis codes were appended to his medical record afterward, the most probable cause of his ESRD should be “cystic kidney”.

[Result] This patient was classified as polycystic kidney related ESRD.

Appendix 2. List of contributory causes of ESRD and ICD-9-CM codes

Contributory causes of ESRD	ICD-9-CM
Glomerulonephropathy	
Unknown glomerulonephritis	580-589, 593.7, 593.70-593.73
Systemic lupus erythematous	710.0
Henoch-Schönlein purpura	287.0
Scleroderma	710.1
Hemolytic uremic syndrome	283.11
Wegener's granulomatosis	446.4
Goodpasture's syndrome	446.21
Genetic and metabolic diseases	
Insulin-dependent diabetes mellitus	250.x except Type II DM (250.00, 250.02, 250.10, 250.20, 250.22, 250.30, 250.32, 250.40, 250.42, 250.50, 250.52, 250.60, 250.62, 250.70, 250.72, 250.80, 250.82, 250.90, 250.92)
Amino-acid transport disorders	270.0
Glycogen storage diseases	271.0
Mucopolysaccharidosis	277.5
Metabolic disorders	277.8-9
Lysosomal storage disorders	271.0, 277.5
Tuberous sclerosis	759.5
Amyloidosis	277.3, 277.30, 277.39
Congenital anomalies (CAKUT)	
Renal hypoplasia, dysplasia, oliognephronia	753.0
Cystic kidney disease	753.1
Congenital anomalies of urinary system	753.20-3, 753.29
Specified anomalies of kidney	753.3
Tumor	
Renal and urinary tract tumor	189.x
Nephrolithiasis	
Nephrolithiasis	592.0
Trauma	
Renal trauma	866.x
ESRD with hypertension	
ESRD with hypertension	403.01, 403.11, 403.91, 404.02, 404.03, 4.12, 404.13, 404.92, 404.93

Appendix 3. List of CAKUT and some metabolic disorders that are existing already at birth (life-long disease)

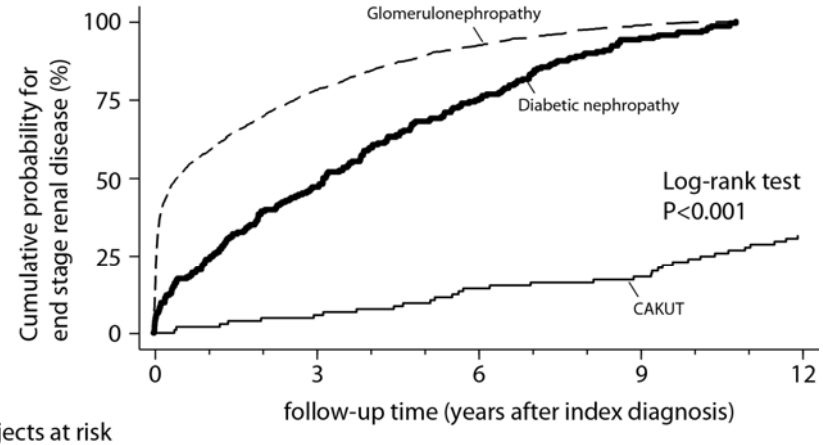
life-long disease
Genetic and metabolic diseases
Amino-acid transport disorders
Glycogen storage diseases
Mucopolysaccharidosis
Metabolic disorders
Lysosomal storage disorders
Tuberous sclerosis
Congenital anomalies (CAKUT)
Renal hypoplasia, dysplasia, oliognephronia
Cystic kidney disease
Congenital anomalies of urinary system
Specified anomalies of kidney

Appendix 4. Characteristics of patients included in the renal survival analysis compared to those of excluded patients

	No. of participants		No. of excluded subjects		
	n=2304	(%)	n=527	(%)	*p* value[a]
Age at onset of ESRD					0.053
Infant	15	(0.7)	0	(0.0)	
Toddler	9	(0.4)	1	(0.2)	
Pre-school	12	(0.5)	1	(0.2)	
School age	94	(4.1)	14	(2.7)	
Adolescent	275	(11.9)	60	(11.4)	
Young adult	1899	(82.4)	451	(85.6)	
Sex					0.070
Female	1049	(45.5)	288	(54.6)	
Male	1255	(54.5)	239	(45.4)	
Contributory causes					0.185
Glomerulonephropathy	1411	(61.2)	323	(61.3)	
Genetic and metabolic diseases	233	(10.1)	117	(22.2)	
Congenital anomalies (CAKUT)	106	(4.6)	36	(6.8)	
Tumor	36	(1.6)	8	(1.5)	
Nephrolithiasis	30	(1.3)	5	(0.9)	
Trauma	5	(0.2)	1	(0.2)	
ESRD with hypertension	483	(21.0)	37	(7.0)	

[a]Chi-square test

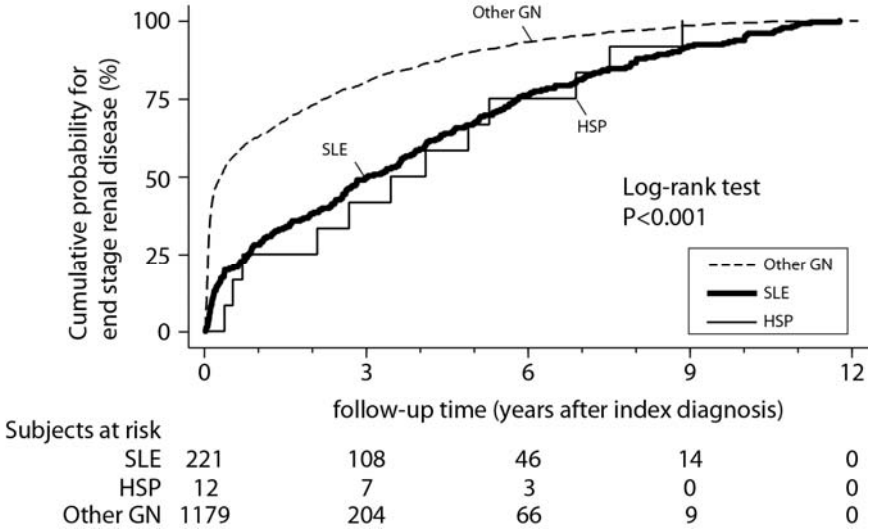
Appendix 5. Renal survivals of ESRD related to glomerulonephritis, CAKUT, and diabetic nephropathy in the young (n=2304) in 1998-2009.



Subjects at risk					
Glomerulonephropathy	1412	319	115	23	0
Diabetic nephropathy	206	110	53	12	0
CAKUT	105	100	90	87	74

[1]Renal survivals of ESRD related to glomerulonephritis, CAKUT, and diabetic nephropathy in the young were plotted for illustration. However, as a retrospective case-only study, the Kaplan–Meier estimator is **not** the **true "disease specific" risk** (probability) for renal failure. The curves here should be interpreted as the **renal survival** (duration of renal deterioration) but the incidence rate in the traditional sense.

Appendix 6. Renal survivals of ESRD related to Systemic lupus erythematous (SLE), Henoch-Schönlein purpura (HSP), and other types of glomerulonephritis (Other GN) in the young (n=1412) in 1998-2009.



Subjects at risk					
SLE	221	108	46	14	0
HSP	12	7	3	0	0
Other GN	1179	204	66	9	0

[1] Renal survivals of ESRD related to SLE, HSP, and other GN in the young were plotted for illustration. However, as a retrospective case-only study, the Kaplan–Meier estimator is **not** the **true "disease specific" risk** (probability) for renal failure. The curves here should be interpreted as the **renal survival** (duration of renal deterioration) but the incidence rate in the traditional sense.