## **SUPPLEMENTARY APPENDIX**

# Mortality in IgA Nephropathy – a nationwide population-based study

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## Description of the national registers used in the paper

#### Personal identity number (PIN)

All Swedish citizens are assigned a unique ten-digit personal identity number (PIN), consisting birth date in YYMMDD format, a three-digit serial number and a one-digit checksum. The PIN is used for identification in a wide range of contexts, including all healthcare contacts, and it is also the identifier in national census and health registers.<sup>1</sup>

#### The Total Population Register (TPR)

The Swedish Total Population Register (TPR) contains demographical data on all Swedish residents, including date of birth, death, emigration or immigration, area of birth and residence, and level of education. The Multi-Generation Register (MGR) is part of the TPR and links legal spouses and first-degree relatives to all individuals born in Sweden since 1932 who were still alive in 1961. <sup>2</sup>

### The National Patient Register (NPR)

The Swedish National Patient Register (NPR) contains information on inpatient discharge records from 1964 onward, with national coverage since 1987. Hospital-based outpatient visits are included since 2001, with a coverage of more than 99%. The register includes data on admission and discharge, main and additional diagnoses and surgical procedures, with corresponding International Classification of Diseases (ICD) codes and procedure codes. The register has been validated for several diagnoses, with positive predictive values of 85-95 % for most diagnoses, with a tendency to higher accuracy with more severe diagnoses. <sup>4</sup>

#### The Prescribed Drug Register (PDR)

The Swedish Prescribed Drug Register was started on July 1 2005 and contains information on drugs dispensed in Swedish Pharmacies, including substance with corresponding ATC code, brand name, formulation, amount and dosage, age sex, place of residence and PIN of the patient. The register has a high sensitivity. Patient identity is available for 99,7% of the drug disposals. <sup>5</sup>

#### The Cause of Death Register (CDR)

The Swedish Cause of Death Register (CDR) was launched in 1952 and became complete in 1961. Registration of a cause of death is mandatory and should be completed within three weeks from the death certificate. The register includes a primary and, if applicable, contributing cases of death. A cause of death is registered in about 99.5% of the deceased (the remaining 0.5% are assigned the ICD code R99.9. For this study, we used only the primary death cause.

<sup>&</sup>lt;sup>1</sup> Ludvigsson, JF, Otterblad-Olausson, P, Pettersson, BU, Ekbom, A: The Swedish personal identity number: possibilities and pitfalls in healthcare and medical research. *Eur J Epidemiol*, 24: 659-667, 2009.

<sup>&</sup>lt;sup>2</sup> Ludvigsson, JF, Almqvist, C, Bonamy, AK, Ljung, R, Michaelsson, K, Neovius, M, Stephansson, O, Ye, W: Registers of the Swedish total population and their use in medical research. *Eur J Epidemiol*, 31: 125-136, 2016.

<sup>&</sup>lt;sup>3</sup> Ludvigsson JF, Andersson E, Ekbom A, et al. External review and validation of the Swedish national inpatient register. *BMC Public Health* 2011; **11:** 450).

<sup>&</sup>lt;sup>4</sup> Wettermark, B, Hammar, N, Fored, CM, Leimanis, A, Otterblad Olausson, P, Bergman, U, Persson, I, Sundstrom, A, Westerholm, B, Rosen, M: The new Swedish Prescribed Drug Register--opportunities for pharmacoepidemiological research and experience from the first six months. *Pharmacoepidemiology and drug safety*, 16: 726-735, 2007.

<sup>&</sup>lt;sup>5</sup> Brooke, HL, Talback, M, Hornblad, J, Johansson, LA, Ludvigsson, JF, Druid, H, Feychting, M, Ljung, R: The Swedish cause of death register. *Eur J Epidemiol*, 32: 765-773, 2017.

**eTable S1.** International classification of disease (ICD) codes for renal disease, cardiovascular disease, diabetes, cancer, autoimmune disease, Henoch-Schönlein purpura (HSP) and other systemic inflammatory diseases.

Diagnostic group	ICD-8	ICD-9	ICD-10
Heredity for renal disease at study entry (adjus	ted for)		
End-stage renal disease		see eTable S2	
IgA Nephropathy	defined acco	ording to renal bio	psy records
IgA Nephropathy phenotype			
Concurrent extrarenal IgA vasculitis (Henoch-	287,00	278A	D69.0
Schönlein purpura) before or <1 year after			
IgAN diagnosis			
Comorbidity at study entry (adjust for 1-4)			
1. Cardiovascular disease/death	390-458	390-459	100-99
2. Cancer	140-209	140-208	C00-D48
3. Diabetes mellitus (type 1 or 2)	250	250	E10-14
4. Other systemic inflammatory diseases (any			
of those below but other than diabetes)			
-Psoriasis	696 (969,30 excluded)	696A-C + E-W	L40
-SLE	734,1	710A	M32.1; M32.8;
			M32.9
-Rheumatoid arthritis	712,3; 714,93	714	M05; M06; M08;
Call to the same	572.00 572.00 562.00		M09; M12.3
-Crohn's disease	572,00; 572,09; 563,00	555	K50
-Ulcerative colitis	572,20; 572,21; 563,10	556	K51
-Thyroiditis	245	245A-X	E06
-Hyperthyroidism	242	242	E05
-Sarcoidosis	135	135	D86
-Primary biliary cirrhosis		571G	K74.3
-ANCA vasculitis and other vasculitis	446,	446	M31
-Celiac disease	269,0	579A	K90.0
-Pelvospondylitis			M45.9
-Autoimmune hepatitis	573,08; 573,09	573D	K75.4; K75.5; K75.9
-Primary sclerosing cholangitis			K83.0A
	1	i	i e

**eTable S2.** International classification of disease (ICD) codes for renal endpoints using hospital-based in- or outpatient diagnosis.

Diagnostic group	ICD-8	ICD-9	ICD-10	Procedure codes
Medical diagnosis of end-stage renal	-	585	N18.0; N18.5	
disease (ESRD) <sup>a</sup>				
Renal dialysis <sup>b</sup>	Y29,01	V45B; V56	Z49; Z99.2	9200; V9200; 9212; V9212; 9314;
				V9531; DR012; DR013; DR016;
				DR024; QF006
				9211; V9211; 9213; V9213;
				V9532; DR015; DR023; DR055;
				DV056
				9219; V9219; 9223; V9223;
				DR017; DR020; DR055; DR056
Renal transplantation		V42A	Z94.0	6070; KAS10; KAS20

<sup>&</sup>lt;sup>a</sup> In the Patient Register or the Cause of Death Register (underlying or contributory cause).

eTable S3. Anatomical therapeutic chemical (ATC) codes for medications

Drug group	ATC code
Steroids	H02AB, except H02AB08 (triamcinolone)
	H02B
Immunosuppressants, other	Cyclophosphamide (L01AA01);
	Rituximab (L01XC02)
	Mycophenolic acid (L04AA06)
	Cyclosporine (L04AD01
	Tacrolimus (L04AD02)
	Azathioprine (L04AX01),
Inhibitors of the renin-angiotensin-aldosterone system	C09
(RAAS inhibitors).	
Anti-hypertensives, other	C02, C03 C07 and C08
Statins	C10AA

eTable S4. International classification of disease (ICD) codes for cause-specific death.

Cause of death <sup>a</sup>	ICD8	ICD9	ICD10
Cardiovascular	390-458	390-459	100-99
Cancer	140-209	140-208	C00-D48
Other		All other cause	S

<sup>&</sup>lt;sup>a</sup> According to *underlying* cause but not contributory cause.

<sup>&</sup>lt;sup>b</sup> Regarded as ESRD if occurring ≥3 times in a patient with ≥4 months between the first and last dialysis.

**eTable S5.** Death hazard in patients with IgA nephropathy (IgAN) diagnosed in Sweden from 1974-2011 compared with the general reference population (corresponds to figure 2, column 1).

Total         577/54058         2066/277463         10.7 vs. 7.4         1.53 (1.37, 1.72)           Length of follow-up         Vol 1 years         43/3590         71/17957         12 vs. 4         2.47 (1.50, 4.06)           10 5 years         411/17286         355/86664         6.4 vs. 4.1         1.21 (0.93, 1.58)           > 5 years³         423/52782         1640/271683         8 vs. 6         1.58 (1.39, 1.80)           ≥ 10 years²         293/463349         1204/240801         6.3 vs. 5         1.56 (1.34, 1.82)           ≥ 20 years²         99/26236         438/14027         3.8 vs. 3.1         1.59 (1.22, 2.08)           Nome         134/16422         502/83079         8.2 vs. 6         1.65 (1.32, 2.08)           Men         443/37636         156/419438         12 vs. 8.1         1.50 (1.32, 1.71)           Age         4/5754         9/29152         0.70 vs. 0.31         4           1-3-39 years         37/26519         211/131622         2.8 vs. 1.6         1.73 (1.28, 2.34)           40-59 years         221/17491         781/90355         13 vs. 8.6         1.25 (1.04, 1.50)           260 years         279/4294         1065/26324         65 vs. 40         1.78 (1.51, 2.11)           219-41 years         121/1788 <t< th=""><th>Variable</th><th>IgAN: Events/py</th><th>Ref: Events/py</th><th>Events/1000 py: IgAN vs. ref</th><th>Adj HR (95% CI)</th></t<>	Variable	IgAN: Events/py	Ref: Events/py	Events/1000 py: IgAN vs. ref	Adj HR (95% CI)
0 to 1 years         43/3590         71/17957         12 vs. 4         2.47 (1.50,4.06)           1 to 5 years         111/17286         355/86664         6.4 vs. 4.1         1.21 (0.93,158)           5 years²         423/52782         1640/271683         8 vs. 6         1.58 (1.39,1.80)           >20 years²         99/26236         438/140427         3.8 vs. 3.1         1.59 (1.22, 2.08)           3.0 years²         13/7754         56/42980         1.7 vs. 1.3         2.03 (0.92, 4.48)           Sex           Women         143/16422         502/83079         8.2 vs. 6         1.65 (1.32, 2.08)           Men         443/37636         156/194384         1 vs. 8.1         1.50 (1.32, 1.08)           Age         7         17/26519         211/131622         2.8 vs. 1.6         1.53 (1.82, 2.48)           40.59 years         73/26519         211/131622         2.8 vs. 1.6         1.73 (1.82, 3.44)           40.59 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           202-201	Total	577/54058	2066/277463	10.7 vs. 7.4	1.53 (1.37, 1.72)
110 5 years	Length of follow-up				
>5 years³         423/52782         1640/271683         8 vs. 6         1.58 (1.39, 1.80)           >10 years³         293/46349         1204/240801         6.3 vs. 5         1.56 (1.34, 1.82)           >20 years³         99/26236         438/140427         3.8 vs. 3.1         1.59 (1.22, 2.08)           30 years³         13/7754         56/42980         1.7 vs. 1.3         2.03 (0.92, 448)           Sex           Women         134/16422         502/83079         8.2 vs. 6         1.65 (1.32, 2.08)           Men         443/37636         1564/194384         12 vs. 8.1         1.50 (1.32, 1.71)           Age           -17 years         4/5754         9/29152         0.70 vs. 0.31         a 1.31 (1.8, 2.34)           40-59 years         73/26519         211/131622         2.8 vs. 1.6         1.25 (1.04, 1.50)           ≥60 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           ≥60 years         127/14791         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           ≥96 years         127/14791         1781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           1974-1988         121/11788         459/61222         10 vs. 7.5 <t< td=""><td>0 to 1 years</td><td>43/3590</td><td>71/17957</td><td>12 vs. 4</td><td>2.47 (1.50, 4.06)</td></t<>	0 to 1 years	43/3590	71/17957	12 vs. 4	2.47 (1.50, 4.06)
>10 years	1 to 5 years	111/17286	355/86664	6.4 vs. 4.1	1.21 (0.93, 1.58)
>20 years of solution of soluti	>5 years <sup>a</sup>	423/52782	1640/271683	8 vs. 6	1.58 (1.39, 1.80)
Say of the say of th	>10 years <sup>a</sup>	293/46349	1204/240801	6.3 vs. 5	1.56 (1.34, 1.82)
Sex           Women         134/16422         502/83079         8.2 vs. 6         1.65 (1.32, 2.08)           Men         43/37636         1564/194384         12 vs. 8.1         1.50 (1.32, 1.71)           Age           -17 years         4/5754         9/29152         0.70 vs. 0.31         4           18-39 years         73/26519         211/11362         2.8 vs. 1.6         1.73 (1.28, 2.34)           40-59 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           60 years         279/4294         1065/26324         50 vs. 40         1.78 (1.51, 2.11)           Calendar year           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           1989-2001         313/26891         4871/7943         83 vs. 5.7         1.48 (1.18, 1.85)           1981-201         313/26891         4871/29491         83 vs. 5.5         1.52 (1.26, 1.83)           1991-201 <td< td=""><td>&gt;20 years <sup>a</sup></td><td>99/26236</td><td>438/140427</td><td>3.8 vs. 3.1</td><td>1.59 (1.22, 2.08)</td></td<>	>20 years <sup>a</sup>	99/26236	438/140427	3.8 vs. 3.1	1.59 (1.22, 2.08)
Women         134/16422         502/83079         8.2 vs. 6         1.65 (1.32, 2.08)           Men         443/37636         1564/194384         12 vs. 8.1         1.50 (1.32, 1.71)           Age           -17 years         4/5754         9/29152         0.70 vs. 0.31         4           18-39 years         73/26519         211/131622         2.8 vs. 1.6         1.73 (1.28, 2.34)           40-59 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           260 years         279/4294         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           26-19 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           26-10 years         279/4294         160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           1999-2015         143/15379         477793         23 vs. 15         1.52 (1.26, 1.83)           19per sec. school (1-9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.26, 1.83)           University	>30 years <sup>a</sup>	13/7754	56/42980	1.7 vs. 1.3	2.03 (0.92, 4.48)
Men         443/37636         1564/194384         12 vs. 8.1         1.50 (1.32, 1.71)           Age           -17 years         4/5754         9/29152         2.70 vs. 0.31         4           8-89 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           260 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           Cologous Septions         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           Cologous Septions         221/17789         1065/26324         65 vs. 40         1.78 (1.51, 2.11)           Cologous Septions         221/17188         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           Cologous Septions         131/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           Cologous Jaly 15379         447/77943         93 vs. 5.7         1.48 (1.18, 1.85)           Cologous Jaly 15379         891/59247         23 vs. 15         1.52 (1.26, 1.83)           Cologous Jaly 15379         891/59247         83 vs. 5.5         1.47 (1.19, 1.81)           Cologous Jaly 15379         891/59247         11 vs. 7.6         1.52 (1.36, 1.71)	Sex				
Age         4/5754         9/29152         0.70 vs. 0.31         d           18-39 years         73/26519         211/131622         2.8 vs. 1.6         1.73 (1.28, 2.34)           40-59 years         221/17491         781/93656         13 vs. 8.6         1.25 (1.04, 1.50)           ≥60 years         279/4294         1065/26324         65 vs. 40         1.78 (1.51, 2.11)           Calledar year           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           2989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           2002-2015         143/15379         447/77943         9.3 vs. 5.7         1.48 (1.18, 1.85)           Colovasion (0.9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.26, 1.83)           Liper sec. school (1.3 years)         206/24829         687/124915         3.3 vs. 5.5         1.47 (1.19, 1.81)           Univer sib full         76/17636         344/87645         4.3 vs. 3.9         1.21 (0.84, 1.79)           Nordic         545/50257         1961/257647         11 vs. 7.6         1.52 (1.36, 1.71)           Nordic         545/50257         1961/257647         11 vs. 7.6         1.52 (1.36, 1	Women	134/16422	502/83079	8.2 vs. 6	1.65 (1.32, 2.08)
-17 years         4/5754         9/29152         0.70 vs. 0.31         delication           18-39 years         73/26519         211/131622         2.8 vs. 1.6         1.73 (1.28, 2.34)           40-59 years         221/17491         781/90365         13 vs. 8.6         1.25 (1.04, 1.50)           260 years         279/4294         1065/6324         65 vs. 40         1.78 (1.51, 2.10)           Calledar year           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           2002-2015         13/31339         44777943         9.3 vs. 5.7         1.88 (1.47, 2.41)           2002-2015         13/415379         44777943         23 vs. 15         1.52 (1.26, 1.83)           Upper sec. school (0-9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.36, 1.71)           Upper sec. school (1-3 years)         26/24829         687/124915         8.3 vs. 5.5         1.47 (1.19, 1.81)           Upiver sity         76/17636         1961/257647         11 vs. 7.6         1.52 (1.36, 1.73)           Nortic         \$45/50257         1961/257647         11 vs. 7.6         1.52 (1.36, 1.73)	Men	443/37636	1564/194384	12 vs. 8.1	1.50 (1.32, 1.71)
18-39 years   73/26519   211/131622   2.8 vs. 1.6   1.73 (1.28, 2.34)	Age				
40-59 years       221/17491       781/90365       13 vs. 8.6       1.25 (1.04, 1.50)         ≥60 years       279/4294       1065/26324       65 vs. 40       1.78 (1.51, 2.11)         Calendar year         1974-1988       121/11788       459/61222       10 vs. 7.5       1.88 (1.47, 2.41)         1989-2001       313/26891       1160/138298       12 vs. 8.4       1.46 (1.25, 1.71)         2002-2015       143/15379       447/77943       9.3 vs. 5.7       1.48 (1.18, 1.85)         Education         Compulsory school (0-9 years)       257/10974       891/59247       23 vs. 15       1.52 (1.26, 1.83)         Upper sec. school (1-3 years)       206/24829       687/124915       8.3 vs. 5.5       1.47 (1.19, 1.81)         Upper sec. school (1-3 years)       206/24829       687/124915       8.3 vs. 5.5       1.47 (1.19, 1.81)         Upper sec. school (1-3 years)       266/17636       344/87645       8.3 vs. 5.5       1.47 (1.19, 1.81)         Upper sec. school (1-3 years)       26/17636       348/87645       1.8 vs. 5.3       1.52 (1.36, 1.71)         Nordic       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Nordic       5	-17 years	4/5754	9/29152	0.70 vs. 0.31	d
≥60 years         279/4294         1065/26324         65 vs. 40         1.78 (1.51, 2.11)           Calendar year           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           2002-2015         43/15379         447/77943         93 vs. 5.7         1.84 (1.18, 1.85)           Education           Compulsory school (0-9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.26, 1.83)           Upper sec. school (1-3 years)         206/24829         687/124915         8.3 vs. 5.5         1.47 (1.19, 1.81)           University         76/17636         344/87645         4.3 vs. 3.9         1.21 (0.84, 1.74)           Computy of birth           Nor-Nordic         32/3796         1961/257647         11 vs. 7.6         1.52 (1.36, 1.71)           Non-Nordic         32/3796         185/1912         7.7 vs. 5.2         1.54 (0.76, 3.13)           Presentation purpura*           Yes         20/2607         8/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Yes         1053/1992 </td <td>18-39 years</td> <td>73/26519</td> <td>211/131622</td> <td>2.8 vs. 1.6</td> <td>1.73 (1.28, 2.34)</td>	18-39 years	73/26519	211/131622	2.8 vs. 1.6	1.73 (1.28, 2.34)
Calendar year           1974-1988         121/11788         459/61222         10 vs. 7.5         1.88 (1.47, 2.41)           1989-2001         313/26891         1160/138298         12 vs. 8.4         1.46 (1.25, 1.71)           2002-2015         143/15379         447/77943         9.3 vs. 5.7         1.46 (1.25, 1.71)           2002-2015         143/15379         447/77943         9.3 vs. 5.7         1.52 (1.26, 1.83)           Chaction           Compulsory school (0-9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.26, 1.83)           Upper sec. school (1-3 years)         206/24829         687/124915         8.3 vs. 5.5         1.47 (1.19, 1.81)           University         76/17636         344/87645         4.3 vs. 3.9         1.21 (0.84, 1.74)           Chaction           Wolf-17636         1961/257647         11 vs. 7.6         1.52 (1.36, 1.71)           Non-Nordic         545/50257         1961/257647         11 vs. 7.6         1.54 (0.76, 3.13)           Webook-Schönlein purpura*           Yes         20/2607         68/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Chaction         1.52 (0.33, 1.95	40-59 years	221/17491	781/90365	13 vs. 8.6	1.25 (1.04, 1.50)
1974-1988       121/11788       459/61222       10 vs. 7.5       1.88 (1.47, 2.41)         1989-2001       313/26891       1160/138298       12 vs. 8.4       1.46 (1.25, 1.71)         2002-2015       143/15379       447/77943       9.3 vs. 5.7       1.48 (1.18, 1.85)         Education         Compulsory school (0-9 years)       257/10974       891/59247       23 vs. 15       1.52 (1.26, 1.83)         Upper sec. school (1-3 years)       206/24829       687/124915       8.3 vs. 5.5       1.47 (1.19, 1.81)         University       76/17636       344/87645       8.3 vs. 5.5       1.47 (1.19, 1.81)         University       76/17636       344/87645       8.3 vs. 5.5       1.47 (1.19, 1.81)         Country of birth         World       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpura*         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Yes       307/14633       1013/77927       21 vs. 13       1.62 (1.38, 1.89)         Yes	≥60 years	279/4294	1065/26324	65 vs. 40	1.78 (1.51, 2.11)
1989-2001       313/26891       1160/138298       12 vs. 8.4       1.46 (1.25, 1.71)         2002-2015       143/15379       447/77943       9.3 vs. 5.7       1.48 (1.18, 1.85)         Education         Compulsory school (0-9 years)       257/10974       891/59247       23 vs. 15       1.52 (1.26, 1.83)         Upper sec. school (1-3 years)       206/24829       687/124915       8.3 vs. 5.5       1.47 (1.19, 1.81)         University       761/7636       344/87645       4.3 vs. 3.9       1.21 (0.84, 1.74)         Country of birth         Nordic       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpurab         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease*         No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       30/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.54 (1.3	Calendar year				
2002-2015         143/15379         447/77943         9.3 vs. 5.7         1.48 (1.18, 1.85)           Education           Compulsory school (0-9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.26, 1.83)           Upper sec. school (1-3 years)         206/24829         687/124915         8.3 vs. 5.5         1.47 (1.19, 1.81)           University         76/17636         344/87645         4.3 vs. 3.9         1.21 (0.84, 1.74)           Country of birth           Nordic         545/50257         1961/257647         11 vs. 7.6         1.52 (1.36, 1.71)           Non-Nordic         32/3796         195/19712         8.4 vs. 5.3         1.86 (1.14, 3.05)           Henock-Schönlein purpursie         20/2607         68/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Cardiovascular disease*           Yes         20/2607         68/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Posson (1.54 (0.74) (0.76) (0.74)         1.0 (0.77)         1.52 (1.38, 1.89)           Yes         307/14633         1013/77927         21 vs. 13         1.52 (1.38, 1.89)           Yes         47/1997         168/1076         24 vs. 15         1.56 (1.38, 1.75)           Yes         51/25602 <td>1974-1988</td> <td>121/11788</td> <td>459/61222</td> <td>10 vs. 7.5</td> <td>1.88 (1.47, 2.41)</td>	1974-1988	121/11788	459/61222	10 vs. 7.5	1.88 (1.47, 2.41)
Education           Compulsory school (0-9 years)         257/10974         891/59247         23 vs. 15         1.52 (1.26, 1.83)           Upper sec. school (1-3 years)         206/24829         687/124915         8.3 vs. 5.5         1.47 (1.19, 1.81)           University         76/17636         344/87645         4.3 vs. 3.9         1.21 (0.84, 1.74)           Country of birth           Nordic         545/50257         1961/257647         11 vs. 7.6         1.52 (1.36, 1.71)           Non-Nordic         32/3796         105/19712         8.4 vs. 5.3         1.86 (1.14, 3.05)           Henoch-Schönlein purpurab           Yes         20/2607         68/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Cardiovascular disease <sup>c</sup> No         270/39425         1053/199536         6.8 vs. 5.3         1.62 (1.38, 1.89)           Yes         307/14633         1013/77927         21 vs. 13         1.56 (1.38, 1.75)           Yes         47/1997         168/11076         24 vs. 15         1.56 (1.38, 1.75)           Yes         47/1997         168/11076         24 vs. 15         1.54 (1.37, 1.73)           Yes         60/1456         183/8895         9.8 vs. 7         1.54 (1.37, 1.73)<	1989-2001	313/26891	1160/138298	12 vs. 8.4	1.46 (1.25, 1.71)
Compulsory school (0-9 years)       257/10974       891/59247       23 vs. 15       1.52 (1.26, 1.83)         Upper sec. school (1-3 years)       206/24829       687/124915       8.3 vs. 5.5       1.47 (1.19, 1.81)         University       76/17636       344/87645       4.3 vs. 3.9       1.21 (0.84, 1.74)         Country of birth         Nordic       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpurab         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease <sup>c</sup> No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.56 (1.38, 1.75)         Cancer <sup>c</sup> No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.54 (1.37, 1.73)         Yes       78/3186       215/18001	2002-2015	143/15379	447/77943	9.3 vs. 5.7	1.48 (1.18, 1.85)
Upper sec. school (1-3 years)       206/24829       687/124915       8.3 vs. 5.5       1.47 (1.19, 1.81)         University       76/7636       344/87645       4.3 vs. 3.9       1.21 (0.84, 1.74)         Country of birth         Nordic       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpurab         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease'         No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       30/714633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer*         No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)*         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41	Education				
University       76/17636       344/87645       4.3 vs. 3.9       1.21 (0.84, 1.74)         Country of birth         Nordic       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpurab         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease'         No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer'         No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)*         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory discovered by a colspan="6">Diabetes mellitus (type 1 or 2)*         Yes       78/3186       215/18001	Compulsory school (0-9 years)	257/10974	891/59247	23 vs. 15	1.52 (1.26, 1.83)
Country of birth           Nordic         545/50257         1961/257647         11 vs. 7.6         1.52 (1.36, 1.71)           Non-Nordic         32/3796         105/19712         8.4 vs. 5.3         1.86 (1.14, 3.05)           Henoch-Schönlein purpurab           Yes         20/2607         68/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Cardiovascular disease*           No         270/39425         1053/199536         6.8 vs. 5.3         1.62 (1.38, 1.89)           Yes         307/14633         1013/77927         21 vs. 13         1.33 (1.10, 1.62)           Cancer*           No         530/52061         1898/266387         10 vs. 7.1         1.56 (1.38, 1.75)           Yes         47/1997         168/11076         24 vs. 15         1.51 (0.83, 2.78)           Diabetes mellitus (type 1 or 2)*           No         517/52602         1883/268568         9.8 vs. 7         1.54 (1.37, 1.73)           Yes         60/1456         183/8895         41 vs. 21         1.30 (0.61, 2.77)           Other systemic inflammatory           Yes         78/3186         215/18001         24 vs. 12         1.79 (0.85, 3.79)           First-grade rel	Upper sec. school (1-3 years)	206/24829	687/124915	8.3 vs. 5.5	1.47 (1.19, 1.81)
Nordic       545/50257       1961/257647       11 vs. 7.6       1.52 (1.36, 1.71)         Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpurab         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease <sup>c</sup> No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer <sup>c</sup> No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)*         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.50 (1.33, 1.68)         Other systemic inflammatory         Ves       78/3186       215/18001       24 vs. 12       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal di	University	76/17636	344/87645	4.3 vs. 3.9	1.21 (0.84, 1.74)
Non-Nordic       32/3796       105/19712       8.4 vs. 5.3       1.86 (1.14, 3.05)         Henoch-Schönlein purpurab         Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease <sup>c</sup> No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer <sup>c</sup> No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2) <sup>c</sup> No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)       1.59 (1.33, 1.68)       1.50 (0.31, 1.79)       1.50 (0.31, 1.69)       1.50 (0.31, 1.69)       1.50 (0.33, 1.68)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.69)       1.50 (0.35, 1.6	Country of birth				
Henoch-Schönlein purpurab           Yes         20/2607         68/13101         7.7 vs. 5.2         1.54 (0.76, 3.13)           Cardiovascular disease*           No         270/39425         1053/199536         6.8 vs. 5.3         1.62 (1.38, 1.89)           Yes         307/14633         1013/77927         21 vs. 13         1.33 (1.10, 1.62)           Cancer*           No         530/52061         1898/266387         10 vs. 7.1         1.56 (1.38, 1.75)           Yes         47/1997         168/11076         24 vs. 15         1.51 (0.83, 2.78)           Diabetes mellitus (type 1 or 2)*           Yes         60/1456         183/8895         9.8 vs. 7         1.54 (1.37, 1.73)           Yes         60/1456         183/8895         41 vs. 21         1.30 (0.61, 2.77)           Other systemic inflammatory disease*           Yes         78/3186         215/18001         24 vs. 12         1.50 (1.33, 1.68)           Yes         78/3186         215/18001         24 vs. 12         1.79 (0.85, 3.79)           First-grade relative with renal disease*           IgAN         2/501         11/2474         4 vs. 4.4         1.80 (0.30, 10.8)	Nordic	545/50257	1961/257647	11 vs. 7.6	1.52 (1.36, 1.71)
Yes       20/2607       68/13101       7.7 vs. 5.2       1.54 (0.76, 3.13)         Cardiovascular disease*         No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer*         No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)**         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease*         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Non-Nordic	32/3796	105/19712	8.4 vs. 5.3	1.86 (1.14, 3.05)
Cardiovascular disease*         No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer*         No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)*         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease*         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Henoch-Schönlein purpurab				
No       270/39425       1053/199536       6.8 vs. 5.3       1.62 (1.38, 1.89)         Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer <sup>c</sup> No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2) <sup>c</sup> No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease <sup>c</sup> No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Yes	20/2607	68/13101	7.7 vs. 5.2	1.54 (0.76, 3.13)
Yes       307/14633       1013/77927       21 vs. 13       1.33 (1.10, 1.62)         Cancer <sup>c</sup> No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2) <sup>c</sup> No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Cardiovascular disease <sup>c</sup>				
Cancer <sup>c</sup> No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2) <sup>c</sup> No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease <sup>c</sup> No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	No	270/39425	1053/199536	6.8 vs. 5.3	1.62 (1.38, 1.89)
No       530/52061       1898/266387       10 vs. 7.1       1.56 (1.38, 1.75)         Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)**         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory discrete         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Yes	307/14633	1013/77927	21 vs. 13	1.33 (1.10, 1.62)
Yes       47/1997       168/11076       24 vs. 15       1.51 (0.83, 2.78)         Diabetes mellitus (type 1 or 2)**         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease*         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Cancer <sup>c</sup>				
Diabetes mellitus (type 1 or 2)°         No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease°         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	No	530/52061	1898/266387	10 vs. 7.1	1.56 (1.38, 1.75)
No       517/52602       1883/268568       9.8 vs. 7       1.54 (1.37, 1.73)         Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease <sup>c</sup> No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Yes	47/1997	168/11076	24 vs. 15	1.51 (0.83, 2.78)
Yes       60/1456       183/8895       41 vs. 21       1.30 (0.61, 2.77)         Other systemic inflammatory disease         No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Diabetes mellitus (type 1 or 2) <sup>c</sup>				
Other systemic inflammatory disease <sup>c</sup> No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	No	517/52602	1883/268568	9.8 vs. 7	1.54 (1.37, 1.73)
No       499/50873       1851/259462       9.8 vs. 7.1       1.50 (1.33, 1.68)         Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Yes	60/1456	183/8895	41 vs. 21	1.30 (0.61, 2.77)
Yes       78/3186       215/18001       24 vs. 12       1.79 (0.85, 3.79)         First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	Other systemic inflammatory di	sease <sup>c</sup>			
First-grade relative with renal disease         IgAN       2/501       11/2474       4 vs. 4.4       1.80 (0.30, 10.8)	No	499/50873	1851/259462	9.8 vs. 7.1	1.50 (1.33, 1.68)
IgAN 2/501 11/2474 4 vs. 4.4 1.80 (0.30, 10.8)	Yes	78/3186	215/18001	24 vs. 12	1.79 (0.85, 3.79)
	First-grade relative with renal disease				
ESRD 7/942 26/4717 7.4 vs. 5.5 1.21 (0.38, 3.82)	IgAN	2/501	11/2474	4 vs. 4.4	1.80 (0.30, 10.8)
		7/942	26/4717	7.4 vs. 5.5	1.21 (0.38, 3.82)

<sup>&</sup>lt;sup>a</sup> Not mutually exclusive. For instance a patient with a follow-up of 11 years would be included in both the >5-year-and the >10-year-categories.

<sup>&</sup>lt;sup>b</sup> Before or within 1 year after first renal biopsy indicating IgAN.

<sup>&</sup>lt;sup>c</sup> Before study entry (date of IgAN and corresponding date in reference individuals).

<sup>&</sup>lt;sup>d</sup> Adjusted HR not possible to estimate due to lack of variation within strata. py = person-years, HR = hazard ratio.

**eTable S6**: Death hazard in patients with IgA nephropathy (IgAN) diagnosed in Sweden in 1974-2011 compared with siblings (corresponds to Figure 3, column 2).

Variable	IgAN: Events/py	Ref: Events/py	Events/1000 py: IgAN vs. Ref	Adj HR (95% CI)
Total	257/43106	362/99467	6 vs. 3.6	1.62 (1.29, 2.03)
Length of follow-up				
0 to 1 years	10/2762	8/6191	3.6 vs. 1.3	0.90 (0.14, 5.88)
1 to 5 years	43/13425	57/30190	3.2 vs. 1.9	1.63 (0.93, 2.86)
>5 years <sup>a</sup>	204/42272	297/97803	4.8 vs. 3	1.67 (1.30, 2.15)
>10 years <sup>a</sup>	143/37403	220/87394	3.8 vs. 2.5	1.72 (1.29, 2.30)
>20 years <sup>a</sup>	59/21748	79/51663	2.7 vs. 1.5	2.60 (1.61, 4.19)
>30 years <sup>a</sup>	7/6666	8/16333	1 vs. 0.49	0.38 (0.02, 9.46)
Sex				
Women	45/12606	99/28357	3.6 vs. 3.5	1.29 (0.75, 2.23)
Men	212/30499	263/71110	6.9 vs. 3.7	1.41 (1.07, 1.86)
Age				
-17 years	4/5511	4/11968	0.73 vs. 0.33	d
18-39 years	54/23196	85/51255	2.3 vs. 1.7	1.65 (1.01, 2.68)
40-59 years	136/12885	220/32085	11 vs. 6.9	1.39 (1.03, 1.87)
≥60 years	63/1514	53/4159	42 vs. 13	3.01 (1.69, 5.37)
Calendar year				
1974-1988	52/9263	70/21761	5.6 vs. 3.2	1.92 (1.14, 3.25)
1989-2001	126/21492	193/49506	5.9 vs. 3.9	1.42 (1.03, 1.95)
2002-2015	79/12351	99/28201	6.4 vs. 3.5	1.80 (1.17, 2.76)
Education				
Compulsory school (0-9 years)	95/7088	103/18086	13 vs. 5.7	2.33 (1.41, 3.82)
Upper sec. school (1-3 years)	112/20694	186/48868	5.4 vs. 3.8	1.61 (1.10, 2.35)
University	47/14974	71/31257	3.1 vs. 2.3	1.60 (0.85, 3.01)
Country of birth				
Nordic	254/42312	359/96941	6 vs. 3.7	1.63 (1.30, 2.04)
Non-Nordic	3/794	3/2526	3.8 vs. 1.2	d
Henoch-Schönlein purpurab				
Yes	8/2254	12/4857	3.5 vs. 2.5	3.98 (0.36, 44.1)
Cardiovascular disease <sup>c</sup>				
No	126/32727	213/73359	3.8 vs. 2.9	1.65 (1.24, 2.21)
Yes	131/10379	149/26108	13 vs. 5.7	1.68 (1.01, 2.80)
Cancer <sup>c</sup>				
No	244/41671	341/96030	5.9 vs. 3.5	1.63 (1.30, 2.06)
Yes	13/1435	21/3436	9.1 vs. 6.1	1.71 (0.22, 13.1)
Diabetes mellitus (type 1 or 2) <sup>c</sup>				
No	237/42169	344/97275	5.6 vs. 3.5	1.74 (1.38, 2.20)
Yes	20/936	18/2192	21 vs. 8.2	d
Other systemic inflammatory dise				
No	222/40789	332/93905	5.4 vs. 3.5	1.67 (1.32, 2.12)
Yes	35/2316	30/5562	15 vs. 5.4	0.53 (0.10, 2.85)

<sup>&</sup>lt;sup>a</sup> Not mutually exclusive. For instance a patient with a follow-up of 11 years would be included in both the >5-year-and the >10-year-categories.

<sup>&</sup>lt;sup>b</sup> Before or within 1 year after first renal biopsy indicating IgAN.

<sup>&</sup>lt;sup>c</sup> Before study entry (date of first IgAN biopsy and corresponding date in reference individuals).

<sup>&</sup>lt;sup>d</sup> Adjusted HR not possible to estimate due to lack of variation within strata. py = person-years, HR = hazard ratio.

**eTable S7.** Death hazard in patients with IgA nephropathy (IgAN) diagnosed in Sweden in 1974-2011 compared with spouses (corresponds to Figure 2, column 3).

Total	Variable	IgAN: Events/py	Ref: Events/py	Events/1000 py: IgAN vs. Ref	Adj HR (95% CI)
0 to 1 years         27/2213         11/2225         12 vs. 4.9         4.01 (0.65, 24.7)           1 to 5 years         76/10705         31/10861         7.1 vs. 2.8         1.20 (0.55, 261)           5 years²         297/35096         203/37055         8.5 vs. 5.5         1.41 (1.07, 1.86)           >10 years²         220/31711         140/33794         6.9 vs. 4.1         1.55 (1.12, 2.17)           >20 years²         77/18911         59/21031         4.1 vs. 2.8         1.48 (0.82, 2.68)           >30 years²         12/5971         8/6791         2.0 vs. 1.2         1.17 (0.19, 7.06)           Sex           Women         77/11053         108/10543         7.0 vs. 10         0.68²           Men         323/24721         137/27023         13 vs. 5.1         2.46 (1.78, 3.41)           Age         -17 years         0/1463         1/1477         0.0 vs. 0.68         *           -17 years         167/14273         99/15007         12 vs. 6.6         1.19 (0.82, 1.73)           ≥60 years         19/3331         122/4312         59 vs. 28         1.71 (1.15, 252)           Caledaryear           1974-1988         91/8930         60/9514         10 vs. 6.3         2.06 (1.24, 3.40)	Total	400/35774	245/37566	11 vs. 6.5	1.42 (1.11, 1.83)
1 to 5 years	Length of follow-up				
S   S   S   S   S   S   S   S   S   S	0 to 1 years	27/2213	11/2225	12 vs. 4.9	4.01 (0.65, 24.7)
Name	1 to 5 years	76/10705	31/10861	7.1 vs. 2.8	1.20 (0.55, 2.61)
\$20 years   \$77/18911   \$59/21031   \$4.1 vs. 2.8   \$1.48 (0.82, 2.68)   \$20 years   \$12/5971   \$8/6791   \$2.0 vs. 1.2   \$1.17 (0.19, 7.06)   \$Cex	>5 years <sup>a</sup>	297/35096	203/37055	8.5 vs. 5.5	1.41 (1.07, 1.86)
\$30 years³         12/5971         8/6791         2.0 vs. 1.2         1.17 (0.19, 7.06)           Sex           Women         77/11053         108/10543         7.0 vs. 10         0.68d           Men         232/24721         137/27023         13 vs. 5.1         0.68d           Age	>10 years <sup>a</sup>	220/31711	140/33794	6.9 vs. 4.1	1.55 (1.12, 2.17)
Sex           Women         77/11053         108/10543         7.0 vs. 10         0.68d           Men         323/24721         137/27023         13 vs. 5.1         2.46 (1.78, 3.41)           Age           -17 years         0/1463         1/1477         0.0 vs. 0.68         *           18-39 years         35/16707         23/16769         2.1 vs. 1.4         1.34 (0.57, 3.16)           40-59 years         167/14273         99/15007         12 vs. 6.6         1.19 (0.82, 1.73)           560 years         198/331         122/4312         59 vs. 28         1.19 (0.82, 1.73)           560 years         198/331         120/4312         59 vs. 28         1.19 (0.82, 1.73)           560 years         198/3331         105/14         10 vs. 6.3         2.06 (1.24, 3.40)           1989-2001         214/18358         130/19258         12 vs. 6.8         1.26 (0.86, 1.84)           2002-2015         95/8486         53/94         11 vs. 6.3         2.06 (1.24, 3.40)           1989-2001         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-9 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years) <td>&gt;20 years<sup>a</sup></td> <td>77/18911</td> <td>59/21031</td> <td>4.1 vs. 2.8</td> <td>1.48 (0.82, 2.68)</td>	>20 years <sup>a</sup>	77/18911	59/21031	4.1 vs. 2.8	1.48 (0.82, 2.68)
Women         77/11053         108/10543         7.0 vs. 10         0.68 <sup>d</sup> Men         323/24721         137/27023         13 vs. 5.1         2.46 (1.78, 3.41)           Age           -17 years         0/1463         1/1477         0.0 vs. 0.68         e           18-39 years         35/16707         23/16769         2.1 vs. 1.4         1.34 (0.57, 3.16)           40-59 years         167/14273         99/15007         12 vs. 6.6         1.19 (0.82, 1.73)           260 years         198/331         122/4312         59 vs. 28         1.71 (1.15, 2.52)           Calendar year         1974-1988         91/8930         60/9514         10 vs. 6.3         2.06 (1.24, 3.40)           1988-2001         214/18358         130/19258         12 vs. 6.8         1.26 (0.86, 1.84)           2002-2015         95/8486         55/8794         11 vs. 6.3         1.44 (0.84, 2.49)           Education         Compulsory school (0-9 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         143/15594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/12170         4.5 vs. 3.1         0.81 (0.34, 1.90)	>30 years <sup>a</sup>	12/5971	8/6791	2.0 vs. 1.2	1.17 (0.19, 7.06)
Men         323/24721         137/27023         13 vs. 5.1         2.46 (1.78, 3.41)           Age           -17 years         0/1463         1/1477         0.0 vs. 0.68         °           18-39 years         35/16707         23/16769         2.1 vs. 1.4         1.34 (0.57, 3.16)           40-59 years         166/14273         99/15007         12 vs. 6.6         1.19 (0.82, 1.73)           60 years         198/3331         122/4312         59 vs. 28         1.71 (1.15, 2.52)           Calendar year           1974-1988         91/8930         60/9514         10 vs. 6.3         2.06 (1.24, 3.40)           1989-2001         214/18358         130/19258         12 vs. 6.8         1.26 (0.86, 1.84)           2002-2015         95/8486         58/794         11 vs. 6.3         1.40 (0.84, 1.84)           2002-2015         95/8486         58/794         11 vs. 6.3         1.44 (0.84, 2.84)           2002-2015         95/8486         38/16093         9.2 vs. 5.4         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         143/15594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/2170         1.5 vs. 3.1         0.81 (0.34, 1.93)	Sex				
Age           -17 years         0/1463         1/1477         0.0 vs. 0.68         °           18-39 years         35/16707         23/16769         2.1 vs. 1.4         1.34 (0.57, 3.16)           40-59 years         167/14273         99/15007         12 vs. 6.6         1.19 (0.82, 1.73)           £60 years         198/3331         122/4312         59 vs. 28         1.71 (1.15, 2.52)           Calendar year           1974-1988         91/8930         60/9514         10 vs. 6.3         2.06 (1.24, 3.40)           1989-2001         214/18358         130/19258         12 vs. 6.8         1.26 (0.86, 1.84)           2002-2015         95/8486         55/8794         11 vs. 6.3         1.44 (0.84, 2.49)           Education         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         183/7594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/12170         4.5 vs. 3.1         0.81 (0.34, 1.90)           Non-Nordic         377/32924         229/34566         11 vs. 6.6         1.40 (1.08, 1	Women	77/11053	108/10543	7.0 vs. 10	0.68 <sup>d</sup>
-17 years 0/1463 1/1477 0.0 vs 0.68 ° 18-39 years 35/16707 23/16769 2.1 vs 1.4 1.34 (0.57, 3.16) 40-59 years 167/14273 99/15007 12 vs 6.6 1.19 (0.82, 1.73) ≥60 years 198/3331 122/4312 59 vs .28 1.71 (1.15, 2.52)  Calendar year  1974-1988 91/8930 60/9514 10 vs 6.3 2.06 (1.24, 3.40) 1989-2001 214/18358 130/19258 12 vs 6.8 1.26 (0.86, 1.84) 2002-2015 95/8486 55/8794 11 vs 6.3 1.44 (0.84, 2.49)  Education  Compulsory school (0-9 years) 183/7963 106/8821 23 vs .12 1.53 (0.98, 2.40) Upper sec. school (1-3 years) 143/15594 87/16093 9.2 vs .5.4 1.32 (0.80, 2.19) University 84/12016 38/12170 4.5 vs .3.1 0.81 (0.34, 1.90)  Country of birth  Nordic 377/32924 229/34566 11 vs .6.6 1.40 (1.08, 1.82) Non-Nordic 23/2850 16/3000 8.1 vs .5.3 3.86 (1.04, 14.3)  Henoch-Schönlein purpura*  Yes 13/1247 8/1308 10 vs .6.1 °  Cardiovascular disease 124/10382 125/11375 21 vs .11 0.83 (0.44, 1.59)  Cancer 124/10382 125/11375 21 vs .11 0.83 (0.44, 1.59)  Cancer 24/10382 125/11375 21 vs .11 0.83 (0.44, 1.59)  Cancer 5 No 367/34493 224/36075 11 vs .6.2 1.45 (1.12, 1.89) Yes 33/1281 21/1491 26 vs .14 0.52 (0.02, 11,0)  Diabetes mellitus (type 1 or 2)* No 361/34805 219/36350 10 vs .6.0 1.52 (1.17, 1.98) Yes 39/968 26/1216 40 vs .21 °  Cheter systemic inflammatory discusser 143 (1.98, 1.87)	Men	323/24721	137/27023	13 vs. 5.1	2.46 (1.78, 3.41)
18-39 years 35/16707 23/16769 2.1 vs. 1.4 1.34 (0.57, 3.16) 40-59 years 167/14273 99/15007 12 vs. 6.6 1.19 (0.82, 1.73) ≥60 years 198/3331 122/4312 59 vs. 28 1.71 (1.15, 2.52) Calendar year  1974-1988 91/8930 60/9514 10 vs. 6.3 2.06 (1.24, 3.40) 1989-2001 214/18358 130/19258 12 vs. 6.8 1.26 (0.86, 1.84) 2002-2015 95/8486 55/8794 11 vs. 6.3 1.44 (0.84, 2.49) Education  Compulsory school (0-9 years) 183/7963 106/8821 23 vs. 12 1.53 (0.98, 2.40) Upper sec. school (1-3 years) 143/15594 87/16093 9.2 vs. 5.4 1.32 (0.80, 2.19) University 54/12016 38/12170 4.5 vs. 3.1 0.81 (0.34, 1.90) Country of birth  Nordic 377/32924 229/34566 11 vs. 6.6 1.40 (1.08, 1.82) Non-Nordic 23/2850 16/3000 8.1 vs. 5.3 3.86 (1.04, 14.3) Henoch-Schönlein purpurab  Yes 13/1247 8/1308 10 vs. 6.1 °  Cardiovascular disease '  No 186/25391 120/26191 7.3 vs. 4.6 1.74 (1.23, 2.48) Yes 214/10382 125/11375 21 vs. 11 0.83 (0.44, 1.59) Cancer '  No 367/34493 224/36075 11 vs. 6.2 1.45 (1.12, 1.89) Yes 33/1281 21/1491 26 vs. 14 0.52 (0.02, 11,0) Diabetes mellitus (type 1 or 2)*  No 361/34805 219/36350 10 vs. 6.0 1.52 (1.17, 1.98) Yes 39/968 26/1216 40 vs. 21 °  Other systemic inflammatory disease '  No 353/33579 208/35177 11 vs. 5.9 1.43 (1.09, 1.87)	Age				
40-59 years       167/14273       99/15007       12 vs. 6.6       1.19 (0.82, 1.73)         ≥60 years       198/3331       122/4312       59 vs. 28       1.71 (1.15, 2.52)         Calendar year         1974-1988       91/8930       60/9514       10 vs. 6.3       2.06 (1.24, 3.40)         1989-2001       214/18358       130/19258       12 vs. 6.8       1.26 (0.86, 1.84)         2002-2015       95/8486       55/8794       11 vs. 6.3       1.44 (0.84, 2.49)         Education       Upper sec. school (0-9 years)       183/7963       106/8821       23 vs. 12       1.53 (0.98, 2.40)         Upper sec. school (1-3 years)       143/15594       87/16093       9.2 vs. 5.4       1.32 (0.80, 2.19)         University       54/12016       38/12170       4.5 vs. 3.1       0.81 (0.34, 1.90)         Country of birth         Non-Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       31/1247       8/1308       10 vs. 6.1       e         Cardiovascular disease*         Yes       13/1247       8/1308       10 vs. 6.1       e         Cardiovascular disease*         Ye	-17 years	0/1463	1/1477	0.0 vs. 0.68	e
40-59 years       167/14273       99/15007       12 vs. 6.6       1.19 (0.82, 1.73)         ≥60 years       198/3331       122/4312       59 vs. 28       1.71 (1.15, 2.52)         Calendar year         1974-1988       91/8930       60/9514       10 vs. 6.3       2.06 (1.24, 3.40)         1989-2001       214/18358       130/19258       12 vs. 6.8       1.26 (0.86, 1.84)         2002-2015       95/8486       55/8794       11 vs. 6.3       1.44 (0.84, 2.49)         Education       Use of Marketian         Compulsory school (0-9 years)       183/7963       106/8821       23 vs. 12       1.53 (0.98, 2.40)         Upper sec. school (1-3 years)       143/15594       87/16093       9.2 vs. 5.4       1.32 (0.80, 2.19)         University       54/12016       38/12170       4.5 vs. 3.1       0.81 (0.34, 1.90)         Country of birth         Non-Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       31/1247       8/1308       10 vs. 6.1       °         Cardiovascular disease*         Yes       13/625391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48) <td< td=""><td>18-39 years</td><td>35/16707</td><td>23/16769</td><td>2.1 vs. 1.4</td><td>1.34 (0.57, 3.16)</td></td<>	18-39 years	35/16707	23/16769	2.1 vs. 1.4	1.34 (0.57, 3.16)
E60 years         198/3331         122/4312         59 vs. 28         1.71 (1.15, 2.52)           Calendar year           1974-1988         91/8930         60/9514         10 vs. 6.3         2.06 (1.24, 3.40)           1989-2001         214/18358         130/19258         12 vs. 6.8         1.26 (0.86, 1.84)           2002-2015         95/8486         55/8794         11 vs. 6.3         1.44 (0.84, 2.49)           Education           Compulsory school (0-9 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         143/15594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/12170         4.5 vs. 3.1         0.81 (0.34, 1.90)           Country of birth           Nordic         377/32924         229/34566         11 vs. 6.6         1.40 (1.08, 1.82)           Nor-Nordic         23/2850         16/3000         8.1 vs. 5.3         3.86 (1.04, 14.3)           Yes         13/1247         8/1308         10 vs. 6.1         c           Cardiovascular disease*           Yes         12/4/10382         125/11375         21 vs. 11         0.83 (0.44, 1.59)				12 vs. 6.6	
Calendar year   1974-1988	·	198/3331	122/4312	59 vs. 28	
1989-2001       214/18358       130/19258       12 vs. 6.8       1.26 (0.86, 1.84)         2002-2015       95/8486       55/8794       11 vs. 6.3       1.44 (0.84, 2.49)         Education         Compulsory school (0-9 years)       183/7963       106/8821       23 vs. 12       1.53 (0.98, 2.40)         Upper sec. school (1-3 years)       143/15594       87/16093       9.2 vs. 5.4       1.32 (0.80, 2.19)         University       54/12016       38/12170       4.5 vs. 3.1       0.81 (0.34, 1.90)         Country of birth         Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       31/1247       8/1308       10 vs. 6.1       **         Cardiovascular disease*         No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Yes       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       39/968	Calendar year				
2002-2015         95/8486         55/8794         11 vs. 6.3         1.44 (0.84, 2.49)           Education           Compulsory school (0-9 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         143/15594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/12170         4.5 vs. 3.1         0.81 (0.34, 1.90)           Country of birth           Nordic         377/32924         229/34566         11 vs. 6.6         1.40 (1.08, 1.82)           Non-Nordic         23/2850         16/3000         8.1 vs. 5.3         3.86 (1.04, 14.3)           Henoch-Schönlein purpurab           Yes         13/1247         8/1308         10 vs. 6.1         e           Cardiovascular disease c           No         186/25391         120/26191         7.3 vs. 4.6         1.74 (1.23, 2.48)           Yes         214/10382         125/11375         21 vs. 11         0.83 (0.44, 1.59)           Cancer c           No         367/34493         224/36075         11 vs. 6.2         1.45 (1.12, 1.89)           Yes         33/1281         21/1491         26 vs. 14 <td< td=""><td>·</td><td>91/8930</td><td>60/9514</td><td>10 vs. 6.3</td><td>2.06 (1.24, 3.40)</td></td<>	·	91/8930	60/9514	10 vs. 6.3	2.06 (1.24, 3.40)
2002-2015         95/8486         55/8794         11 vs. 6.3         1.44 (0.84, 2.49)           Education         Compulsory school (0-9 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         143/15594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/12170         4.5 vs. 3.1         0.81 (0.34, 1.90)           Country of birth           Nordic         377/32924         229/34566         11 vs. 6.6         1.40 (1.08, 1.82)           Non-Nordic         23/2850         16/3000         8.1 vs. 5.3         3.86 (1.04, 14.3)           Henoch-Schönlein purpurab           Yes         13/1247         8/1308         10 vs. 6.1         e           Cardiovascular disease <sup>c</sup> No         186/25391         120/26191         7.3 vs. 4.6         1.74 (1.23, 2.48)           Yes         214/10382         125/11375         21 vs. 11         0.83 (0.44, 1.59)           Cancer <sup>c</sup> No         367/34493         224/36075         11 vs. 6.2         1.45 (1.12, 1.89)           Yes         33/1281         21/1491         26 vs. 14         0.52	1989-2001	214/18358	130/19258	12 vs. 6.8	1.26 (0.86, 1.84)
Education           Compulsory school (0-9 years)         183/7963         106/8821         23 vs. 12         1.53 (0.98, 2.40)           Upper sec. school (1-3 years)         143/15594         87/16093         9.2 vs. 5.4         1.32 (0.80, 2.19)           University         54/12016         38/12170         4.5 vs. 3.1         0.81 (0.34, 1.90)           Country of birth           Nordic         377/32924         229/34566         11 vs. 6.6         1.40 (1.08, 1.82)           Non-Nordic         23/2850         16/3000         8.1 vs. 5.3         3.86 (1.04, 14.3)           Henoch-Schönlein purpurab           Yes         13/1247         8/1308         10 vs. 6.1         e           Cardiovascular disease <sup>c</sup> No         186/25391         120/26191         7.3 vs. 4.6         1.74 (1.23, 2.48)           Yes         214/10382         125/11375         21 vs. 11         0.83 (0.44, 1.59)           Cancer <sup>c</sup> No         367/34493         224/36075         11 vs. 6.2         1.45 (1.12, 1.89)           Yes         33/1281         21/1491         26 vs. 14         0.52 (0.02, 11,0)           Diabetes mellitus (type 1 or 2) <sup>c</sup> No	2002-2015	95/8486	55/8794	11 vs. 6.3	
Upper sec. school (1-3 years)       143/15594       87/16093       9.2 vs. 5.4       1.32 (0.80, 2.19)         University       54/12016       38/12170       4.5 vs. 3.1       0.81 (0.34, 1.90)         Country of birth         Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       23/2850       16/3000       8.1 vs. 5.3       3.86 (1.04, 14.3)         Henoch-Schönlein purpurab         Yes       13/1247       8/1308       10 vs. 6.1       *         Cardiovascular disease*         No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer*         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2)*         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       *         Other systemic inflammatory disease*	Education				
University       54/12016       38/12170       4.5 vs. 3.1       0.81 (0.34, 1.90)         Country of birth         Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       23/2850       16/3000       8.1 vs. 5.3       3.86 (1.04, 14.3)         Henoch-Schönlein purpurab         Yes       13/1247       8/1308       10 vs. 6.1       °         Cardiovascular disease °         No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer °         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2)°         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       °         Other systemic inflammatory disease °         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87) <td>Compulsory school (0-9 years)</td> <td>183/7963</td> <td>106/8821</td> <td>23 vs. 12</td> <td>1.53 (0.98, 2.40)</td>	Compulsory school (0-9 years)	183/7963	106/8821	23 vs. 12	1.53 (0.98, 2.40)
Country of birth         Nordic       377/32924       229/34566       11 vs. 6.6       1.40 (1.08, 1.82)         Non-Nordic       23/2850       16/3000       8.1 vs. 5.3       3.86 (1.04, 14.3)         Henoch-Schönlein purpurab         Yes       13/1247       8/1308       10 vs. 6.1       e         Cardiovascular disease c         No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer c         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) <sup>c</sup> No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease c         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Upper sec. school (1-3 years)	143/15594	87/16093	9.2 vs. 5.4	1.32 (0.80, 2.19)
Nordic 377/32924 229/34566 11 vs. 6.6 1.40 (1.08, 1.82)  Non-Nordic 23/2850 16/3000 8.1 vs. 5.3 3.86 (1.04, 14.3)  Henoch-Schönlein purpurab  Yes 13/1247 8/1308 10 vs. 6.1 6  Cardiovascular disease c  No 186/25391 120/26191 7.3 vs. 4.6 1.74 (1.23, 2.48)  Yes 214/10382 125/11375 21 vs. 11 0.83 (0.44, 1.59)  Cancer c  No 367/34493 224/36075 11 vs. 6.2 1.45 (1.12, 1.89)  Yes 33/1281 21/1491 26 vs. 14 0.52 (0.02, 11,0)  Diabetes mellitus (type 1 or 2) c  No 361/34805 219/36350 10 vs. 6.0 1.52 (1.17, 1.98)  Yes 39/968 26/1216 40 vs. 21 6  Other systemic inflammatory disease c  No 353/33579 208/35177 11 vs. 5.9 1.43 (1.09, 1.87)	University	54/12016	38/12170	4.5 vs. 3.1	0.81 (0.34, 1.90)
Non-Nordic       23/2850       16/3000       8.1 vs. 5.3       3.86 (1.04, 14.3)         Henoch-Schönlein purpurab         Yes       13/1247       8/1308       10 vs. 6.1       e         Cardiovascular disease <sup>c</sup> No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer <sup>c</sup> No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) <sup>c</sup> No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease c         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Country of birth				
Henoch-Schönlein purpurab         Yes       13/1247       8/1308       10 vs. 6.1       e         Cardiovascular disease c         No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer c         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) c         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease c         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Nordic	377/32924	229/34566	11 vs. 6.6	1.40 (1.08, 1.82)
Yes       13/1247       8/1308       10 vs. 6.1       e         Cardiovascular disease c         No       186/25391       120/26191       7.3 vs. 4.6       1.74 (1.23, 2.48)         Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer c         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) c         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease c         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Non-Nordic	23/2850	16/3000	8.1 vs. 5.3	3.86 (1.04, 14.3)
Cardiovascular disease °  No 186/25391 120/26191 7.3 vs. 4.6 1.74 (1.23, 2.48) Yes 214/10382 125/11375 21 vs. 11 0.83 (0.44, 1.59)  Cancer °  No 367/34493 224/36075 11 vs. 6.2 1.45 (1.12, 1.89) Yes 33/1281 21/1491 26 vs. 14 0.52 (0.02, 11,0)  Diabetes mellitus (type 1 or 2) °  No 361/34805 219/36350 10 vs. 6.0 1.52 (1.17, 1.98) Yes 39/968 26/1216 40 vs. 21 °  Other systemic inflammatory disease °  No 353/33579 208/35177 11 vs. 5.9 1.43 (1.09, 1.87)	Henoch-Schönlein purpurab				
No 186/25391 120/26191 7.3 vs. 4.6 1.74 (1.23, 2.48) Yes 214/10382 125/11375 21 vs. 11 0.83 (0.44, 1.59)  Cancer c  No 367/34493 224/36075 11 vs. 6.2 1.45 (1.12, 1.89) Yes 33/1281 21/1491 26 vs. 14 0.52 (0.02, 11,0)  Diabetes mellitus (type 1 or 2) c  No 361/34805 219/36350 10 vs. 6.0 1.52 (1.17, 1.98) Yes 39/968 26/1216 40 vs. 21 c  Other systemic inflammatory disease c  No 353/33579 208/35177 11 vs. 5.9 1.43 (1.09, 1.87)	Yes	13/1247	8/1308	10 vs. 6.1	е
Yes       214/10382       125/11375       21 vs. 11       0.83 (0.44, 1.59)         Cancer °         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) °         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease °         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Cardiovascular disease <sup>c</sup>				
Cancer °         No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) °         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease °         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	No	186/25391	120/26191	7.3 vs. 4.6	1.74 (1.23, 2.48)
No       367/34493       224/36075       11 vs. 6.2       1.45 (1.12, 1.89)         Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) c         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease c         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Yes	214/10382	125/11375	21 vs. 11	0.83 (0.44, 1.59)
Yes       33/1281       21/1491       26 vs. 14       0.52 (0.02, 11,0)         Diabetes mellitus (type 1 or 2) c         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease c         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Cancer <sup>c</sup>				
Diabetes mellitus (type 1 or 2) °         No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease °         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	No	367/34493	224/36075	11 vs. 6.2	1.45 (1.12, 1.89)
No       361/34805       219/36350       10 vs. 6.0       1.52 (1.17, 1.98)         Yes       39/968       26/1216       40 vs. 21       e         Other systemic inflammatory disease °         No       353/33579       208/35177       11 vs. 5.9       1.43 (1.09, 1.87)	Yes	33/1281	21/1491	26 vs. 14	0.52 (0.02, 11,0)
Yes         39/968         26/1216         40 vs. 21         e           Other systemic inflammatory disease c           No         353/33579         208/35177         11 vs. 5.9         1.43 (1.09, 1.87)	Diabetes mellitus (type 1 or 2) <sup>c</sup>				
Other systemic inflammatory disease °  No 353/33579 208/35177 11 vs. 5.9 1.43 (1.09, 1.87)	No	361/34805	219/36350	10 vs. 6.0	1.52 (1.17, 1.98)
No 353/33579 208/35177 11 vs. 5.9 1.43 (1.09, 1.87)	Yes	39/968	26/1216	40 vs. 21	e
	Other systemic inflammatory dis	sease <sup>c</sup>			
Yes 47/2194 37/2390 21 vs. 15 0.71 (0.08, 6.24)	No	353/33579	208/35177	11 vs. 5.9	1.43 (1.09, 1.87)
	Yes	47/2194	37/2390	21 vs. 15	0.71 (0.08, 6.24)

<sup>&</sup>lt;sup>a</sup> Not mutually exclusive. For instance a patient with a follow-up of 11 years would be included in both the >5-year-and the >10-year-categories.

<sup>&</sup>lt;sup>b</sup> Before or within 1 year after first renal biopsy indicating IgAN.

<sup>&</sup>lt;sup>c</sup> Before study entry (date of first IgAN biopsy and corresponding date in reference individuals).

<sup>&</sup>lt;sup>d</sup> CI:s not available since the estimation of the variance-covariance matrix did not converge.

 $<sup>^{\</sup>rm e}$  Adjusted HR not possible to estimate due to lack of variation within strata. py = person-years, HR = hazard ratio

**eTable S8.** Death in patients with IgA nephropathy (IgAN) diagnosed in Sweden in 1974-2011 compared with the general reference population, with different medications entered as time-dependent covariates in a regular Cox model, with left truncation on July 1, 2005 (this is when the Prescribed Drug Register (PDR) started).

Use of medications <sup>a</sup>	Number of incident users	Adjusted hazard ratio
	(IgAN patients) since July 2005	
Steroids	1745	1.95 (1.53, 2.50)
Immunosuppressants, other	742	1.33 (1.02, 1.74)
Renin angiotensin aldosterone system (RAAS) inhibitor	2694	0.80 (0.61, 1.04)
Anti-hypertensive, other	2562	1.84 (1.19, 2.85)
Statins	1529	0.83 (0.67, 1.04)

<sup>&</sup>lt;sup>a</sup> See codes in table S4 for corresponding anatomical therapeutic chemical (ATC) codes.

**eTable S9.** Hazard of ESRD in patients with IgA nephropathy (IgAN) diagnosed in Sweden 1974-2011 compared with the general reference population (corresponds to figure 2, column 4).

Variable	IgAN: Events/py	Ref: Events/py	Events/1000 py: IgAN vs. Ref	Adj HR (95% CI)
Total	803/42569	67/255365	19 vs. 0.26	100 (67.7, 148)
Length of follow-up				
0 to 1 years	105/3534	2/17955	30 vs. 0.11	1109 (27.1, 45400)
1 to 5 years	294/15853	12/84658	19 vs. 0.14	159 (68.7, 369)
>5 years <sup>a</sup>	404/40359	53/248514	10 vs. 0.21	73.3 (46.2, 116)
>10 years <sup>a</sup>	196/33501	39/217000	5.8 vs. 0.18	69.2 (36.9, 130)
>20 years <sup>a</sup>	33/17118	14/120296	1.9 vs. 0.12	87.4 (17.9, 426)
>30 years <sup>a</sup>	2/4325	4/30956	0.46 vs. 0.13	d
Sex				
Women	147/13765	11/76324	11 vs. 0.14	123 (42.1, 358)
Men	656/28804	56/179041	23 vs. 0.31	106 (68.1, 165)
Age				
-17 years	17/5122	0/26684	3.3 vs. 0	d
18-39 years	311/20861	18/121507	15 vs. 0.15	279 (103, 753)
40-59 years	346/13184	36/82898	26 vs. 0.43	59.5 (36.9, 96.1)
≥60 years	129/3403	13/24277	38 vs. 0.54	201 (49.5, 813)
Calendar year				
1974-1988	117/9667	14/58885	12 vs. 0.24	341 (68.5, 1700)
1989-2001	429/21037	38/129920	20 vs. 0.29	87.7 (53, 145)
2002-2015	257/11866	15/66560	22 vs. 0.23	225 (74.9, 676)
Education				, , ,
Compulsory school (0-9 years)	228/8354	16/55078	27 vs. 0.29	59.4 (23.8, 148)
Upper sec. school (1-3 years)	373/19491	34/114805	19 vs. 0.3	125 (58.6, 268)
University	194/14203	14/80276	14 vs. 0.17	203 (45.2, 908)
Country of birth				
Nordic	718/39857	62/237484	18 vs. 0.26	96.9 (64.4, 146)
Non-Nordic	85/2708	5/17783	31 vs. 0.28	d
Henoch-Schönlein purpurab				
Yes	20/2115	2/11653	9.5 vs. 0.17	d
Cardiovascular disease <sup>c</sup>				
No	419/32331	46/184905	13 vs. 0.25	116 (70.4, 192)
Yes	384/10239	21/70461	38 vs. 0.3	112 (37.1, 338)
Cancer <sup>c</sup>				
No	763/41023	59/245636	19 vs. 0.24	96.3 (64.5, 144)
Yes	40/1546	8/9729	26 vs. 0.82	62.9 (4.43, 894)
Diabetes mellitus (type 1 or 2) <sup>c</sup>				
No	762/41438	60/247360	18 vs. 0.24	115 (75.3, 175)
Yes	41/1132	7/8005	36 vs. 0.87	3.02 (0.13, 68)
Other systemic inflammatory di	•			
No	744/40047	62/239096	19 vs. 0.26	99.1 (66, 149)
Yes	59/2523	5/16269	23 vs. 0.31	20.4 (0.22, 1916)
First-degree relative with renal	•	, -		, , /
IgAN	5/405	1/2239	12 vs. 0.45	5.37 (0.50, 57.6)
ESRD	15/710	1/4242	21 vs. 0.24	824 (51.6, 13179)
	=5, . =5	-,	,0. 0.2 .	(02.0, 101.0)

<sup>&</sup>lt;sup>a</sup> Not mutually exclusive. For instance a patient with a follow-up of 11 years would be included in both the >5-year-and the >10-year-categories.

<sup>&</sup>lt;sup>b</sup> Before or within 1 year after first biopsy indicating IgAN.

<sup>&</sup>lt;sup>c</sup> Before study entry (date of first IgAN biopsy and corresponding date in reference individuals).

 $<sup>^{\</sup>rm d}$  Adjusted HR not possible to estimate due to lack of variation within strata. py = person-years, HR = hazard ratio.

# **STROBE Statement** — Checklist of items:

ltem No	Recommendation
1√	(a) Indicate the study's design with a commonly used term in the title [p. 1]
	(b) Provide in the abstract an informative and balanced summary of what was done and what was found [p. 5]
	done and what was round [p. 5]
2√	Explain the scientific background and rationale for the investigation being reported [p. 6]
3✔	State specific objectives, including any prespecified hypotheses [p. 6] <b>Comment</b> : We have expressed our hypothesis (and thereby our objective) in the last paragraph of our introduction.
4√	Present key elements of study design early in the paper [p. 6]
	Describe the setting, locations, and relevant dates, including periods of
3.	recruitment, exposure, follow-up, and data collection [p. 6-7]
6✔	(a) Give the eligibility criteria, and the sources and methods of selection of
	participants. Describe methods of follow-up [p. 7-8]
	(b) For matched studies, give matching criteria and number of exposed and
	unexposed [p. 7-8]
7✔	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable [p. 7-8]
/0*	For each variable of interest, give sources of data and details of methods of
<b>V</b> 0	assessment (measurement). Describe comparability of assessment methods if
	there is more than one group [p. 7-8 + Supplementary tables]
9✔	Describe any efforts to address potential sources of bias [p. 15]
	Explain how the study size was arrived at
	<b>Comment</b> : This is a nationwide observational study in which we included all
	available patients with a biopsy report of IgA nephropathy. We did not perform
	any a priori power analysis; however, if the editor feels one is needed, we can
	carry out a post-hoc power analysis.
11√	Explain how quantitative variables were handled in the analyses. If applicable,
	describe which groupings were chosen and why [p. 8]
12 <b>√</b>	(a) Describe all statistical methods, including those used to control for confounding
	[p. 9-10]
	(b) Describe any methods used to examine subgroups and interactions [p. 9-10]
	(c) Explain how missing data were addressed n/a (d) If applicable, explain how loss to follow-up was addressed n/a
	(e) Describe any sensitivity analyses [p. 10]
	(E) Describe any sensitivity analyses [p. 10]
<b>√</b> 13*	(a) Report numbers of individuals at each stage of study—e.g., numbers potentially
	eligible, examined for eligibility, confirmed eligible, included in the study,
	completing follow-up, and analysed [p. 11]
	(b) Give reasons for non-participation at each stage
	<b>Comment</b> : Because this was a strict registry-based study, study participants were not contacted (all data were analyzed without knowledge of the identity of the
	study participants). Hence, we had no "non-participation" at different stages of the
	study.
	(c) Consider use of a flow diagram
	Comment: Dropouts occurred because of largely one reason, which is described in
	the text.
	(a) Civa characteristics of study participants (a.g. demographic clinical social) and
<b>√</b> 14*	(a) Give characteristics of study participants (e.g., demographic, clinical, social) and information on exposures and potential confounders [p. 11, + table 1]
	No 1√ 2√ 3√ 4√ 5√ 6√ 7√ √8*

		[table 1]
		(c) Summarize follow-up time (e.g., average and total amount) [p. 11, 13 + table 1]
Outcome data	<b>√</b> 15*	Report numbers of outcome events or summary measures over time [p. 11-13 + supplementary tables S5-S8]
Main results	16✔	(a) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (e.g., 95% confidence interval). Make clear which confounders were adjusted for and why they were included [p. 8, p. 11-13 + supplementary table S5-S9]
		(b) Report category boundaries when continuous variables were categorized <b>table</b> 1, supplementary tables S5-S8
		(c) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful period <b>p. 11-13</b>
Other analyses	17✔	Report other analyses done—e.g., analyses of subgroups and interactions, and sensitivity analyses [p.14-15 + Table 3 & 4 + Suppl. Table 1]
Discussion		
Key results	18 <b>√</b>	Summarize key results with reference to study objectives [p. 13]
Limitations	19 <b>√</b>	Discuss limitations of the study considering sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias [p. 15]
Interpretation	20✔	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence [p. 14-15]
Generalizability	21 <b>√</b>	Discuss the generalizability (external validity) of the study results [p. 14-15]
Other information		
Funding	22✔	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based [p. 2]

<sup>\*</sup>Give information separately for exposed and unexposed groups.

**Note:** An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at http://www.plosmedicine.org/, Annals of Internal Medicine at http://www.annals.org/, and Epidemiology at http://www.epidem.com/). Information on the STROBE Initiative is available at http://www.strobe-statement.org.