A SNOMED CT linked diagnosis coding scheme for European Renal Medicine

Keith Simpson

Nephrologist
Chair ERA – EDTA Registry Codes and Definitions Working Group
Glasgow Royal Infirmary
Scotland UK

keith.simpson@nhs.net

Yongsheng Gao

Advanced Clinical Terminology Specialist
NHS Data Standards & Products
NHS Connecting for Health
Leeds
England UK

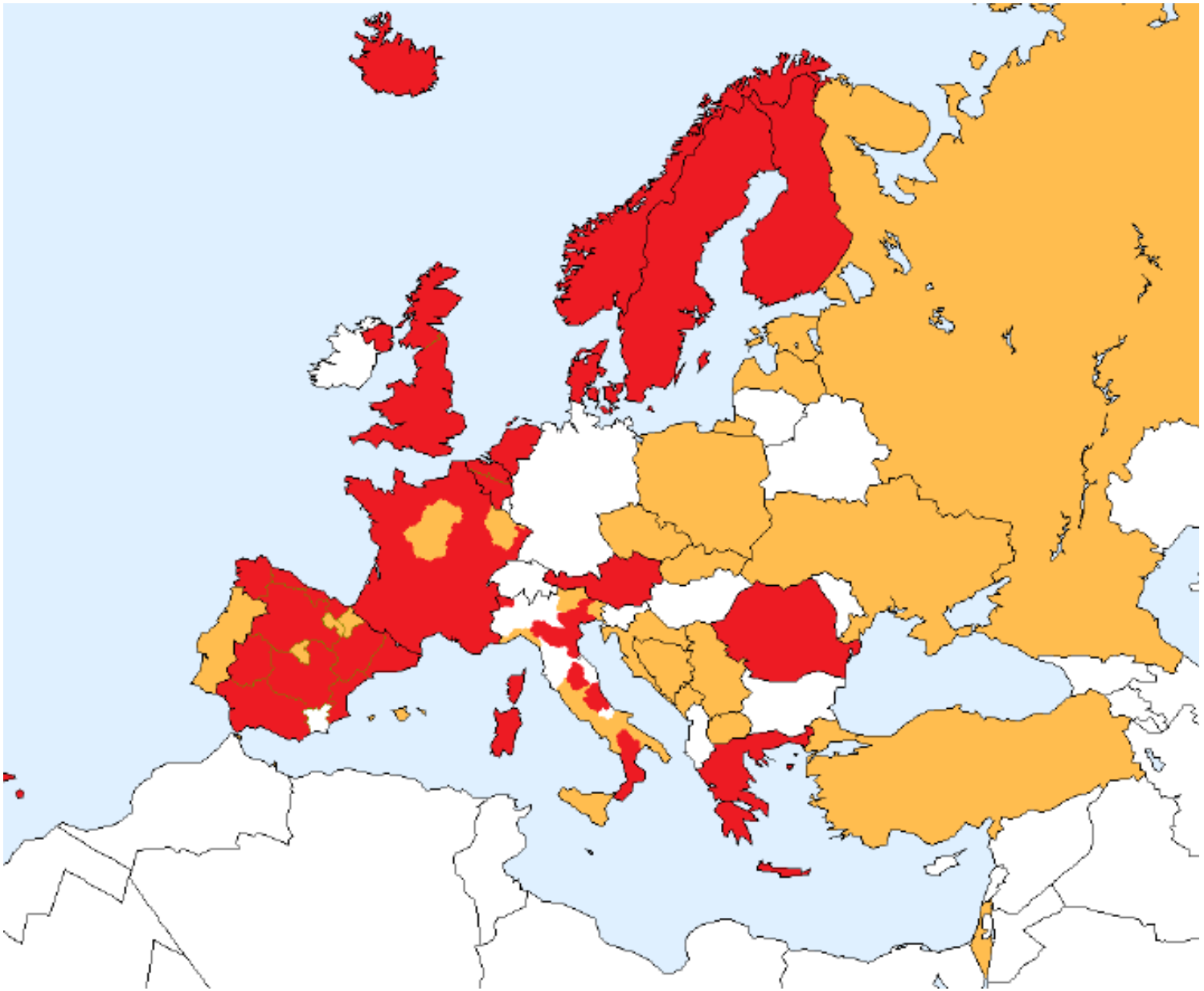
Kitty Jager

ERA-EDTA Registry,
Academic Medical Center,
Dept. of Medical Informatics,
Amsterdam,
The Netherlands.

On behalf of the European Renal Association-European Dialysis and Transplant Association (ERA-EDTA) Registry Coding and Definitions Working Group

Thanks to:
Ian Arowsmith, Denise Downs, Ed Cheetham – NHS Data Standards
Charlie Thomson UK Renal Association
Ronald Cornet ERA-EDTA

National & regional Renal registries affiliated to the ERA EDTA



Individual patient data

Aggregated data

GREATER GLASGOW HEALTH BOARD

GLASGOW ROYAL INFIRMARY

CONFIDENTIAL

This Casesheet must **not** be handled by Patients or Unauthorised Persons


1989

Appendix 1 ERA-EDTA Primary Renal Diagnosis Codes and Groupings

Group 1 Primary Glomerulonephritis

10 Glomerulonephritis; histologically NOT examined
11 Focal segmental glomerulosclerosis with nephrotic syndrome in children
12 IgA nephropathy (proven by immunofluorescence, not 85)
13 Dense deposit disease; membranoproliferative GN; type II (proven by immunofluorescence and/or electron microscopy)
14 Membranous nephropathy
15 Membranoproliferative GN; type I (proven by immunofluorescence and/or electron microscopy-not code 84 or 89)
16 Crescentic (extra-capillary) glomerulonephritis (type I, II, III)
17 Focal segmental glomerulosclerosis with nephrotic syndrome in adults
19 Glomerulonephritis; histologically examined, not given above

Group 2 Interstitial Nephropathies

20 Pyelonephritis cause not specified
21 Pyelonephritis associated with neurogenic bladder
22 Pyelonephritis due to congenital obstructive uropathy with/without vesico-ureteric reflux
23 Pyelonephritis due to acquired obstructive uropathy
24 Pyelonephritis due to vesico-ureteric reflux without obstruction
25 Pyelonephritis due to urolithiasis
29 Pyelonephritis due to other cause
30 Interstitial nephritis (not pyelonephritis) due to other cause, or unspecified (not mentioned below)
31 Interstitial nephropathy due to analgesic drugs
32 Interstitial nephropathy due to cis-platinum
33 Interstitial nephropathy due to cyclosporin A
34 Lead induced interstitial nephropathy
39 Drug induced interstitial nephropathy not mentioned above
40 Cystic kidney disease-type unspecified
41 Polycystic kidneys; adult type (dominant)
42 Polycystic kidneys; infantile (recessive)
43 Medullary cystic disease; including nephronophthisis
49 Cystic kidney– disease-other specified type
50 Hereditary/Familial nephropathy-type unspecified
51 Hereditary nephritis with nerve deafness (Alport's Syndrome)
52 Cystinosis
53 Primary oxalosis
54 Fabry's disease
59 Hereditary nephropathy-other specified type
61 Oligomeganephronic hypoplasia
63 Congenital renal dysplasia with/without urinary tract malformation
66 Syndrome of agenesis of abdominal muscles (Prune Belly)
92 Gout nephropathy (urate)
93 Nephrocalcinosis and hypercalcaemic nephropathy

Group 3 Multisystem Diseases

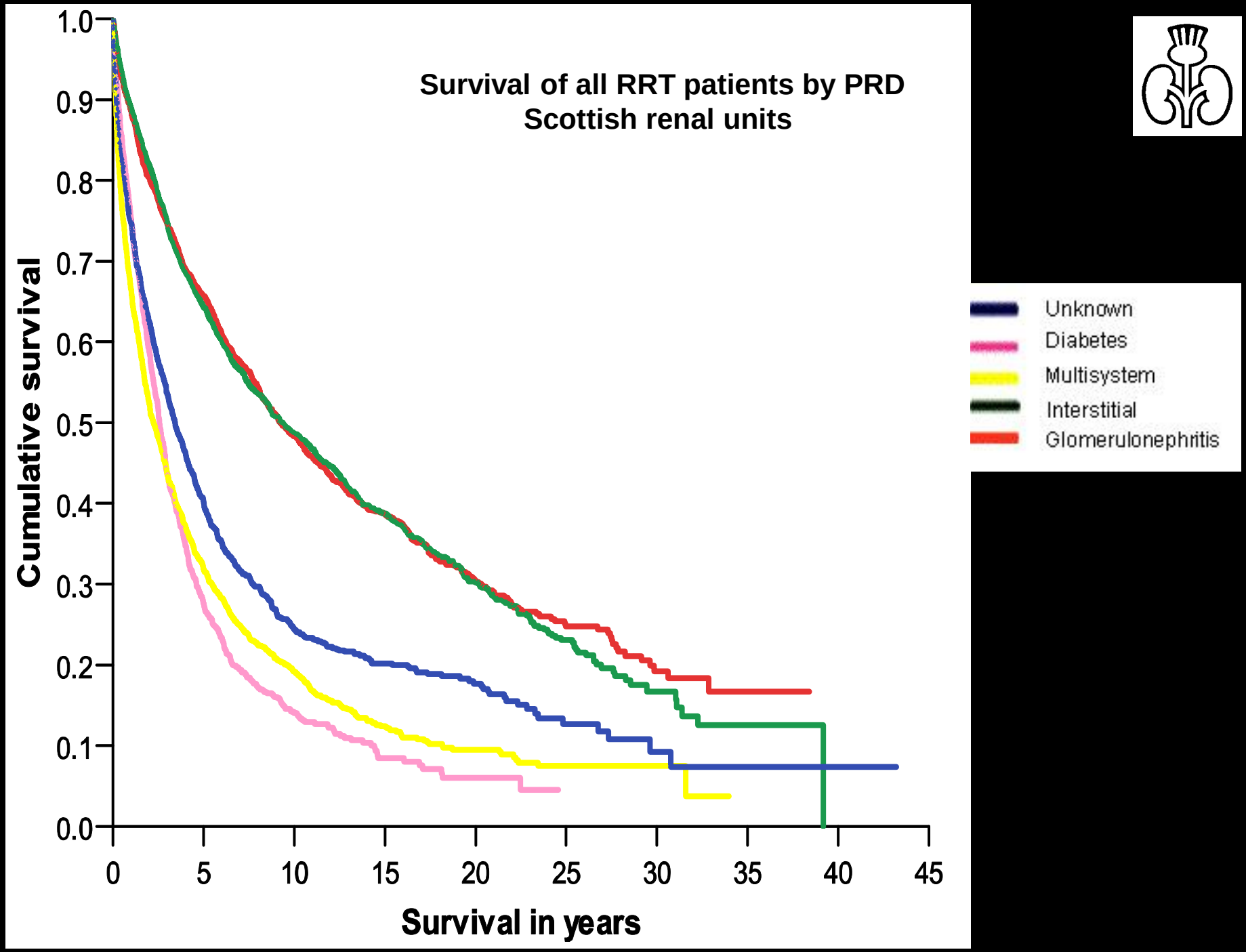
70 Renal vascular disease-type unspecified
71 Renal vascular disease due to malignant hypertension (No PRD)
72 Renal vascular disease due to hypertension (No PRD)
73 Renal vascular disease due to polyarteritis
74 Wegeners Granulomatosis
75 Ischaemic renal disease / cholesterol embolisation
76 Glomerulonephritis related to liver cirrhosis
78 Cryoglobulinaemic glomerulonephritis
79 Renal vascular disease-due to other cause (not given above and not code 84-88)
82 Myelomatosis/light chain deposit disease
83 Amyloid
84 Lupus erythematosus
85 Henoch-Schonlein purpura
86 Goodpasture's Syndrome
87 Systemic sclerosis (scleroderma)
88 Haemolytic uraemic Syndrome (including Moschcowitz Syndrome)
89 Multi-system disease-other (not mentioned above)
90 Tubular necrosis (irreversible) or cortical necrosis (different from 88)
91 Tuberculosis
94 Balkan nephropathy
95 Kidney tumour
96 Traumatic or surgical loss of kidney

Group 4 Diabetes

80 Diabetic glomerulosclerosis or diabetic nephropathy

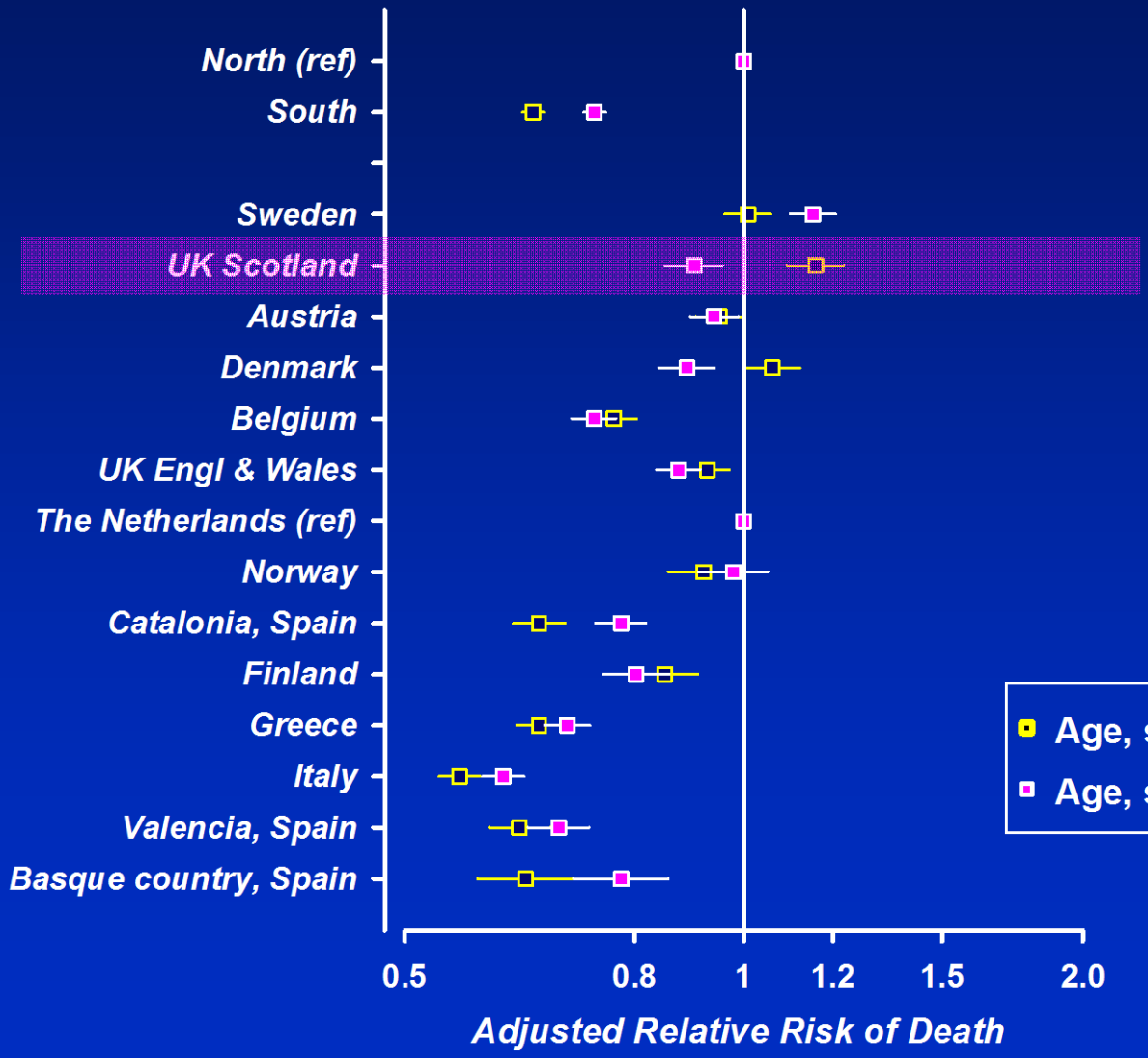
Group 5 Not Known and Other

00 Chronic renal failure; aetiology uncertain/unknown/unavailable
60 Renal hypoplasia (congenital)-type unspecified
99 Other identified renal disorders


Survival of all RRT patients by PRD
Scottish renal units
Cumulative survival
1.0
0.9
0.8
0.7
0.6
0.5
0.4
0.3
0.2
0.1
0.0
0
5
10
15
20
25
30
35
40
45
Survival in years
Unknown
Diabetes
Multisystem
Interstitial
Glomerulonephritis

Results

General population mortality adjusted relative risk of death for incident RRT patients by country and region



RR 0.65 → 0.74

- Age, sex and diab adjusted
- Age, sex, diab and gen pop mort adjusted

Do we need new codes for renal registries?

Yes

if we want

- to pursue old topics properly
- new topics
- International comparisons
  - investigating differences
  - quality improvement (audit)
  - service planning
  - evidence based practice
  - use data derived from electronic records
  - decision support & machine intelligence to improve the care of individual patients and not just for summary statistics.

No

If we plan to continue doing more of the same !

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Aim

To improve and standardise the coding and definitions used by renal registries in Europe.

Some principles

Usable by ordinary clinicians for routine work – care of patients diagnosis, prognosis, treatment

Incorporate the existing ERA-EDTA Codes & definitions PRD, CoD
Provide working definitions
Consistent with emerging medical standards SNOMED CT
Rigorous enough to support research, quality improvement, decision support,
health care planning & management
Compatible with other international registries
Acknowledge the uncertainties in clinical practice
Capable of electronic transmission

Acceptable to the ERA-EDTA registry and affiliated registries

PRD Headings

1) **ERA-EDTA PRD Code** (a meaningless number to identify it)
2) **ERA-EDTA Primary Renal Diagnosis (PRD) terms**
3) **Diagnostic criteria:**
    i) Histology
    ii) Clinical history
    iii) Family history
    iv) Clinical Exam
    v) Biochemistry
    vi) Immunology
    vii) Urine analysis
    viii) Imaging
    ix) Gene test
    x) Other criteria & notes
4) **SNOMED CT concept identifier for focus concept**
5) **SNOMED CT fully specified name**
6) **SNOMED CT expression constraint**
7) **Mapping to old PRD code**
8) **Mapping to old PRD term**
9) **Online Mendelian Inheritance in Man - link**
10) **ICD10 code**
11) **ICD10 rubric**

.

PRD Headings

1) ERA-EDTA PRD Code (a meaningless number to identify it)	1074
2) ERA-EDTA Primary Renal Diagnosis (PRD) terms	Denys-Drash Syndrome
3) Diagnostic criteria:	
i) Histology	x
ii) Clinical history	
iii) Family history	x
iv) Clinical Exam	
v) Biochemistry	
vi) Immunology	
vii) Urine analysis	
viii) Imaging	
ix) Gene test	x
x) Other criteria & notes	WT1 gene mutation. Onset in 1st 3 months of life
4) SNOMED CT concept identifier for focus concept	236385009
5) SNOMED CT fully specified name	Drash syndrome (disorder)
6) SNOMED CT expression constraint	
7) Mapping to old PRD code	99
8) Mapping to old PRD term	Other identified renal disorder
9) Online Mendelian Inheritance in Man - link	http//omim.org/entry/194080
10) ICD10 code	N048
11) ICD10 rubric	Nephrotic syndrome

ERA-EDTA Primary Renal Diagnosis

Renal Diagnoses	Biopsy	Clinical history	Family history	Clinical Exam	Biochemistry	Immunology	Urine dipstick	Urine microscopy	Imaging	Gene test	Other criteria		SNOMED CT focus concept ID	SNOMED CT Fully Specified Name of focus concept	The smantic mapping of SNOMED CT (conform to context model)
Congenital nephrotic syndrome (CNS) - Congenital infection		x			x									Congenital nephrotic syndrome (disorder)	48796009 | Congenital nephrotic syndrome |: 47429007 | Associated with | = ((40733004 | Infectious disease |) AND (! 278929008 | Congenital hepatitis C infection |) AND (! << 52079000 | Congenital human immunodeficiency virus infection |)
Minimal change nephropathy - No histology					x						*A history of heavy proteinuria at some point is required*		44785005	Minimal change disease (disorder)	44785005 | Minimal change disease |: 418775008 | Finding method | = ! <<252416005 | Histopathology test |
Minimal change nephropathy - biopsy proven	x				x						*A history of heavy proteinuria at some point is required*		44785005	Minimal change disease (disorder)	44785005 | Minimal change disease |: 418775008 | Finding method | = <<7246002 | Kidney biopsy |
IgA nephropathy - No histology		x											236407003	IgA nephropathy (disorder)	236407003 | IgA nephropathy |: 418775008 | Finding method | = ! <<252416005 | Histopathology test |
IgA nephropathy - biopsy proven	x										*IgA must be demonstrated in a renal biopsy.*		236407003	IgA nephropathy (disorder)	236407003 | IgA nephropathy |: 418775008 | Finding method | = <<7246002 | Kidney biopsy |
Familial IgA nephropathy - No histology		x	x										zzz	Familial IgA nephropathy	zzz | Familial IgA nephropathy |: 418775008 | Finding method | = ! <<252416005 | Histopathology test |
Familial IgA nephropathy - biopsy proven	x		x								*IgA must be demonstrated in a renal biopsy.*		zzz	Familial IgA nephropathy	zzz | Familial IgA nephropathy |: 418775008 | Finding method | = <<7246002 | Kidney biopsy |
IgA nephropathy secondary to liver cirrhosis - No histology		x											282364005	IgA nephropathy associated with liver disease (disorder)	236407003 | IgA nephropathy |: 47429007 | Associated with | =19943007 | Cirrhosis of liver |, 418775008 | Finding method | = ! <<252416005 | Histopathology test |
IgA nephropathy secondary to liver cirrhosis - biopsy proven	x	x											282364005	IgA nephropathy associated with liver disease (disorder)	282364005 | IgA nephropathy associated with liver disease |: 42752001 | Due to | =19943007 | Cirrhosis of liver |, 418775008 | Finding method | = <<7246002 | Kidney biopsy |
IgM-associated nephropathy	x												236411009	IgM nephropathy (disorder)	236411009 | IgM nephropathy |
Membranous nephropathy - idiopathic	x												197590001	Nephrotic syndrome with membranous glomerulonephritis (disorder)	197590001 | Nephrotic syndrome with membranous glomerulonephritis |
Membranous nephropathy - malignancy associated	x	x											197590001	Nephrotic syndrome with membranous glomerulonephritis (disorder)	197590001 | Nephrotic syndrome with membranous glomerulonephritis |: 47429007 | Associated with | =<< 363346000 | Malignant neoplastic disease |
Membranous nephropathy - drug induced	x	x											197590001	Nephrotic syndrome with membranous glomerulonephritis (disorder)	197590001 | Nephrotic syndrome with membranous glomerulonephritis |: 246075003 | causative agent | =< 410942007 | Drug or medicament |
Membranous nephropathy - infection associated	x	x											197590001	Nephrotic syndrome with membranous glomerulonephritis (disorder)	197590001 | Nephrotic syndrome with membranous glomerulonephritis |: 47429007 | Associated with | =< 40733004 | Infectious disease |
Mesangiocapillary glomerulonephritis type 1	x												75888001	Mesangiocapillary glomerulonephritis, type I (disorder)	75888001 | Mesangiocapillary glomerulonephritis, type I |
Mesangiocapillary glomerulonephritis type 2	x													Mesangiocapillary glomerulonephritis, type II	

Major Heading	Mapping to old P	Mapping to old PRD term	ERA-ED	Online Mendelian Inheritance in Man - link from the National Center for Biotechnology Information http://www.ncbi.nlm.nih.gov/ OMIM Home page, http://www.ncbi.nlm.nih.gov/omim/	ICD10 code	ICD10 Rubric
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	1		N049	Nephrotic syndrome, unspecified
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	2		N049	Nephrotic syndrome, unspecified
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	3	for background information see: http://omim.org/entry/600995 about nephrotic syndrome, type 2; NPHS2 = nephrotic syndrome, steroid-resistant, autosomal recessive; SRN1 http://omim.org/entry/610725 about nephrotic syndrome, type 3; NPHS3 = nephrotic syndrome, early-onset, type 3	N049	Nephrotic syndrome, unspecified
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	4		N049	Nephrotic syndrome, unspecified
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	5	for background information see: http://omim.org/entry/256300 about nephrotic syndrome, type 1; NPHS1 http://omim.org/entry/602716 about nephrin; NPHS1 http://omim.org/entry/600995 about nephrotic syndrome, type 2; NPHS2 = nephrotic syndrome, steroid-resistant, autosomal recessive; SRN1 http://omim.org/entry/604766 about podocin; NPHS2 http://omim.org/entry/610725 about nephrotic syndrome, type 3; NPHS3 = nephrotic syndrome, early-onset, type 3 http://omim.org/entry/256370 about nephrotic syndrome, type 4; NPHS4 http://omim.org/entry/614199 about nephrotic syndrome, type 5, with or without ocular abnormalities; NPHS5	N049	Nephrotic syndrome, unspecified
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	6	http://omim.org/entry/256300 about nephrotic syndrome, type 1; NPHS1 http://omim.org/entry/602716 about nephrin; NPHS1	N049	Nephrotic syndrome, unspecified
Glomerular disease	19	Glomerulonephritis, histologically examined	7	http://omim.org/entry/256300 http://omim.org/entry/602716 about nephrin; NPHS1	N049	Nephrotic syndrome, unspecified
Glomerular disease	19	Glomerulonephritis, histologically examined	8	for background information see: http://omim.org/entry/256370 about nephrotic syndrome, type 4; NPHS4 http://omim.org/entry/607102 about WT1 gene; WT1	N049	Nephrotic syndrome, unspecified
Glomerular disease	11	Severe nephrotic syndrome with focal sclerosis (Paed	9	http://omim.org/entry/600995	N071	Focal and segmental glomerular lesions
Glomerular disease	99	Other identified renal disorder	10	http://omim.org/entry/194080 http://omim.org/entry/607102 about WT1 gene; WT1	N048	Nephrotic syndrome, other
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	11		N049	Nephrotic syndrome, unspecified
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	12		N050	Unspecified nephritic syndrome, minor glomerular abnormality
Glomerular disease	19	Glomerulonephritis, histologically examined	13		N050	Unspecified nephritic syndrome, minor glomerular abnormality
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	14	for background information see: http://omim.org/entry/161950 http://omim.org/entry/613944	N028	Recurrent and persistent haematuria, other
Glomerular disease	12	IgA nephropathy proven by immunofluorescence	15	for background information see: http://omim.org/entry/161950 http://omim.org/entry/613944	N028	Recurrent and persistent haematuria, other
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	16	for background information see: http://omim.org/entry/161950 http://omim.org/entry/613944	N028	Recurrent and persistent haematuria, other
Glomerular disease	12	IgA nephropathy proven by immunofluorescence	17	for background information see: http://omim.org/entry/161950 http://omim.org/entry/613944	N028	Recurrent and persistent haematuria, other
Glomerular disease	10	Glomerulonephritis, histologically NOT examined	18		N028 AND K746	Recurrent and persistent haematuria, other Other and unspecified cirrhosis of liver
Glomerular disease	12	IgA nephropathy proven by immunofluorescence	19		N028 K746	Recurrent and persistent haematuria, other Other and unspecified cirrhosis of liver
Glomerular disease	19	Glomerulonephritis, histologically examined	20		N053	Diffuse mesangial proliferative glomerulonephritis
Glomerular disease	14	Membranous nephropathy	21	for background information see: http://omim.org/entry/604939 about 604939, phospholipase A2 receptor 1; PLA2R1	N042	Nephrotic syndrome, diffuse membranous glomerulonephritis
Glomerular disease	14	Membranous nephropathy	22		N042	Nephrotic syndrome, diffuse membranous glomerulonephritis

The diagnosis
The term we use
= the ‘PRD’

The evidence
<> a definition (yet)

Notes
Because we don’t have a definition

Renal Diagnoses	Biopsy	Clinical history	Family history	Clinical Exam	Biochemistry	Immunology	Urine dipstick	Urine microscopy	Imaging	Gene test	Other criteria
Minimal change nephropathy - No histology					x						*A history of heavy proteinuria at some point is required*
Minimal change nephropathy - biopsy proven	x				x						*A history of heavy proteinuria at some point is required*

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PRD SNOMED id Concept mapping of the meaning

Renal Diagnoses	SNOMED CT focus concept ID	SNOMED CT Fully Specified Name of focus concept	The smantic mapping of SNOMED CT (conform to context model)
Minimal change nephropathy - No histology	44785005	Minimal change disease (disorder)	44785005 | Minimal change disease |: 418775008 | Finding method | = ! <<252416005 | Histopathology test |
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Minimal change nephropathy - biopsy proven	44785005	Minimal change disease (disorder)	44785005 | Minimal change disease |: 418775008 | Finding method | = <<7246002 | Kidney biopsy |

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Why map to SNOMED?

SNOMED CT Provides

Extensive and well supported list of terms current
language translation,
literature links eg BMJ, Map of Medicine
decision support,
queries using multiple domain hierarchies,

secondary functions eg research, hospital management

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literature links eg BMJ, Map of Medicine
decision support,
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secondary functions eg research, hospital management

Will soon be the only terminology supported by the NHS in the UK

Why not stick to ICD10 ?

ICD10 Terms including ‘renal’

one example per letter

A985	Haemorrhagic fever with renal syndrome
C65X	Malignant neoplasm of renal pelvis
D301	Benign neoplasm of renal pelvis
E112	Non-insulin-dependent diabetes mellitus with renal comps
I120	Hypertensive renal disease with renal failure
K767	Hepatorenal syndrome
M8312/3	Renal cell carcinoma (C64)
N170	Acute renal failure with tubular necrosis
O904	Postpartum acute renal failure
P960	Congenital renal failure
Q600	Renal agenesis, unilateral
R392	Extrarenal uraemia
S354	Injury of renal blood vessels
Z992	Dependence on renal dialysis

ICD10 which language ?

N200	Calculus of kidney
N140	Analgesic nephropathy
P960	Congenital renal failure

ICD10 which language ?

N119 Chronic tubulo-interstitial nephritis, unspecified

N159 Renal tubulo-interstitial disease, unspecified

ICD10

No semantic links

ICD10

Renal examples of synonyms

NHS
Connecting for Health

One concept, many names

Some of the descriptions associated with ConceptID 22298006:

- **Fully Specified Name**: *Myocardial infarction (disorder)*
  DescriptionID 751689013
- **Preferred term**: Myocardial infarction
  DescriptionID 37436014
- **Synonym**: Cardiac infarction
  DescriptionID 37442013
- **Synonym**: Heart attack
  DescriptionID 37443015
- **Synonym**: Infarction of heart
  DescriptionID 37441018

Includes foreign language and local terms

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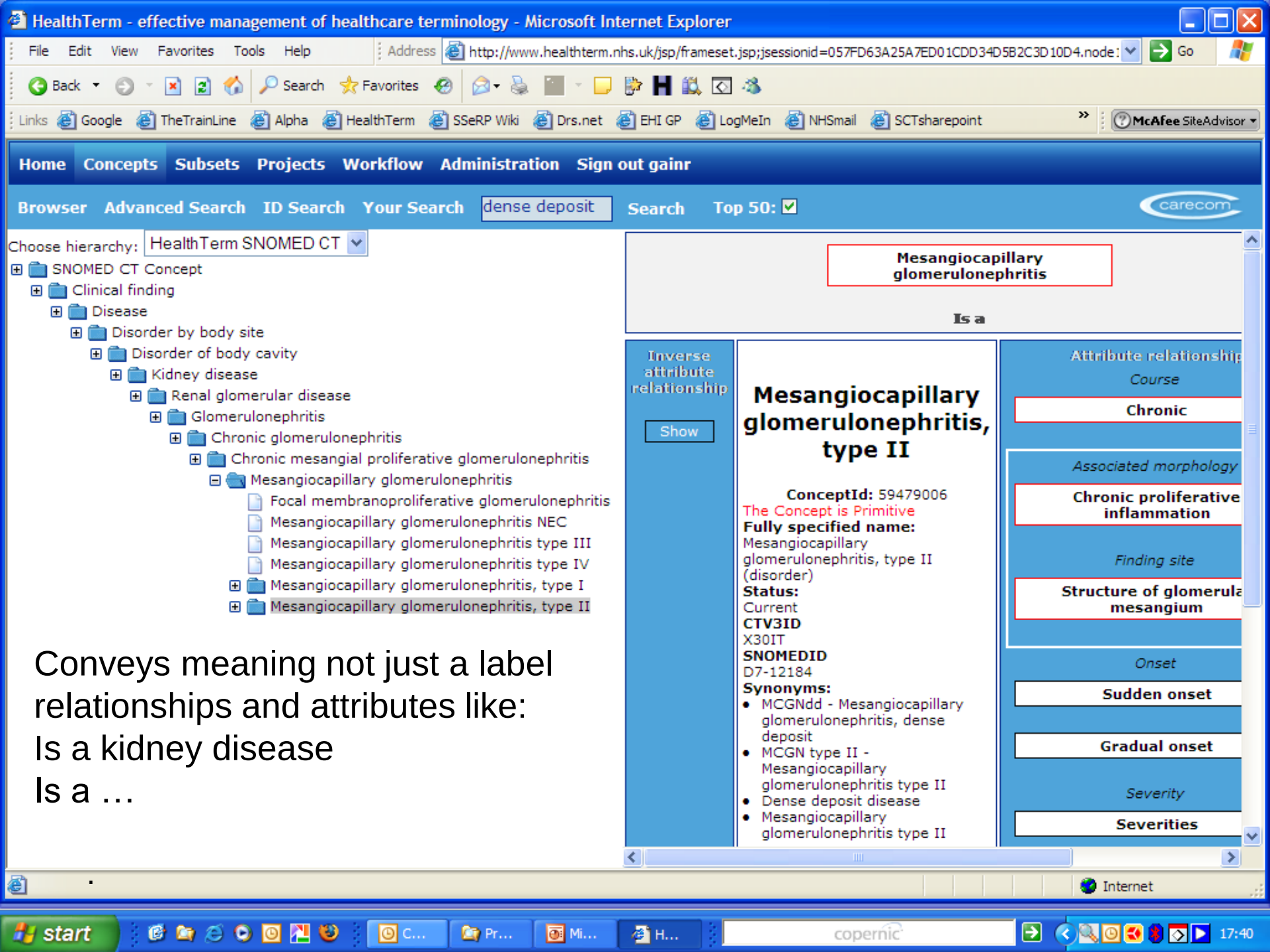
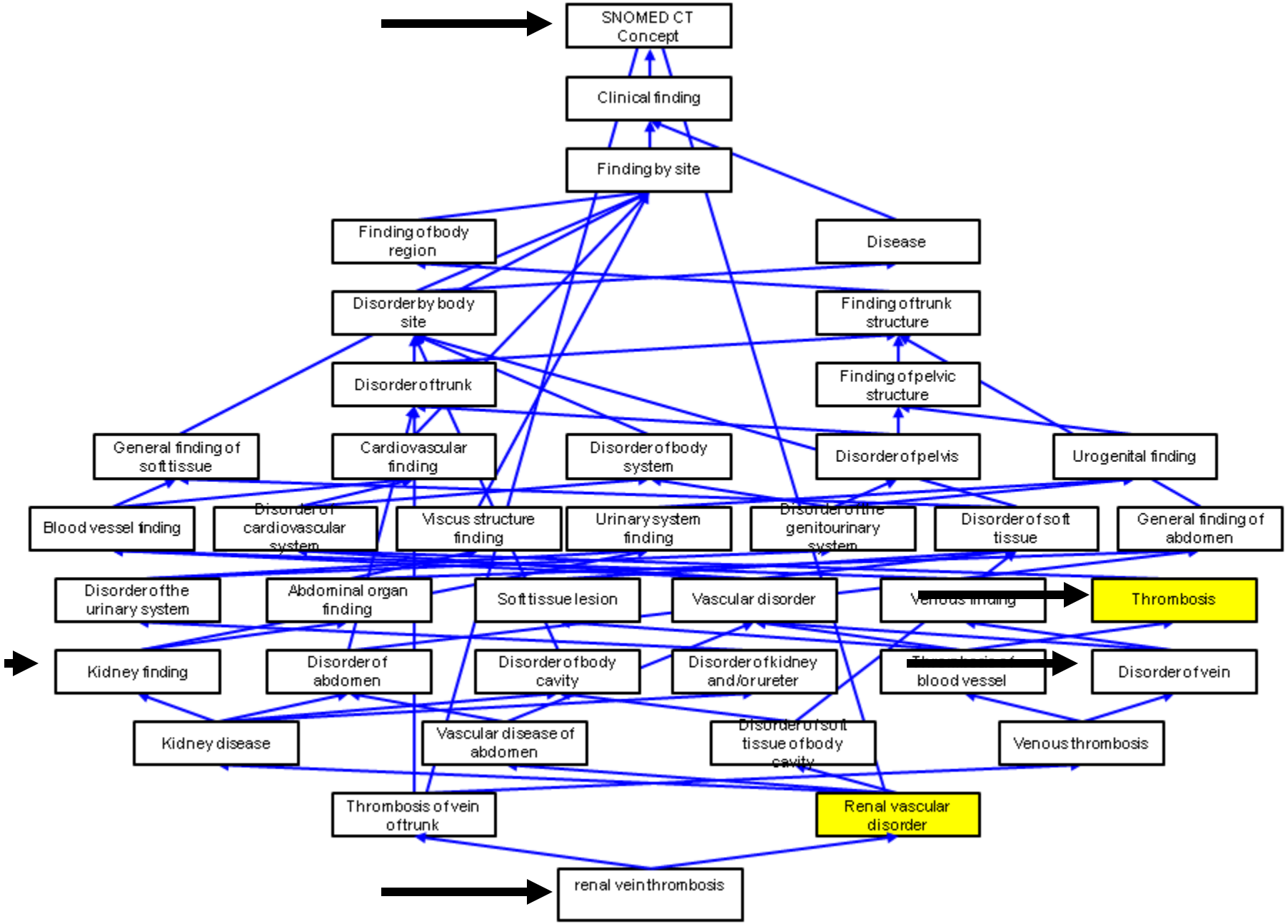


Conveys meaning not just a label relationships and attributes like:
Is a kidney disease
Is a …


SNOMED CT Concept
Clinical finding
Finding by site
Finding of body region
Disease
Disorder by body site
Finding of trunk structure
Disorder of trunk
Finding of pelvic structure
General finding of soft tissue
Cardiovascular finding
Disorder of body system
Disorder of pelvis
Urogenital finding
Blood vessel finding
Disorder of cardiovascular system
Viscus structure finding
Urinary system finding
Disorder of the genitourinary system
Disorder of soft tissue
General finding of abdomen
Disorder of the urinary system
Abdominal organ finding
Soft tissue lesion
Vascular disorder
Venous finding
Thrombosis
Kidney finding
Disorder of abdomen
Disorder of body cavity
Disorder of kidney and/or ureter
blood vessel
Disorder of vein
Kidney disease
Vascular disease of abdomen
Disorder of soft tissue of body cavity
Venous thrombosis
Thrombosis of vein of trunk
Renal vascular disorder
renal vein thrombosis

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UK Renal SNOMED CT subset

Includes:
most of the existing renal terms – about 1100
and the new ERA EDTA PRDs

Managed by:
UK Renal Terminology Committee (RIXG & RA)
working with the NHS Terminology Centre

Purpose:
Patient records, Registries, RADAR, RDGs,
research, TheProject

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Summary

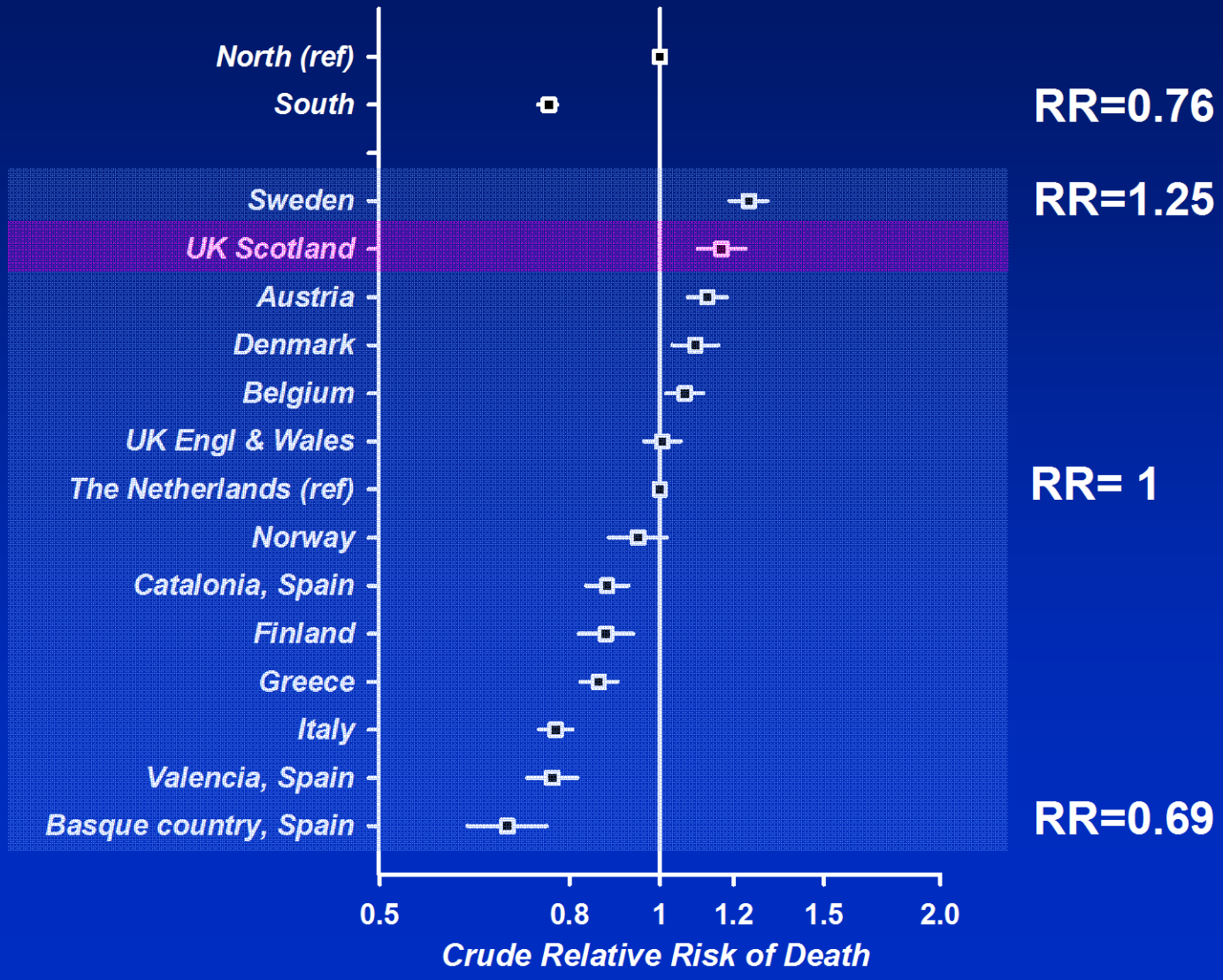
New ERA EDTA PRD terms & codes

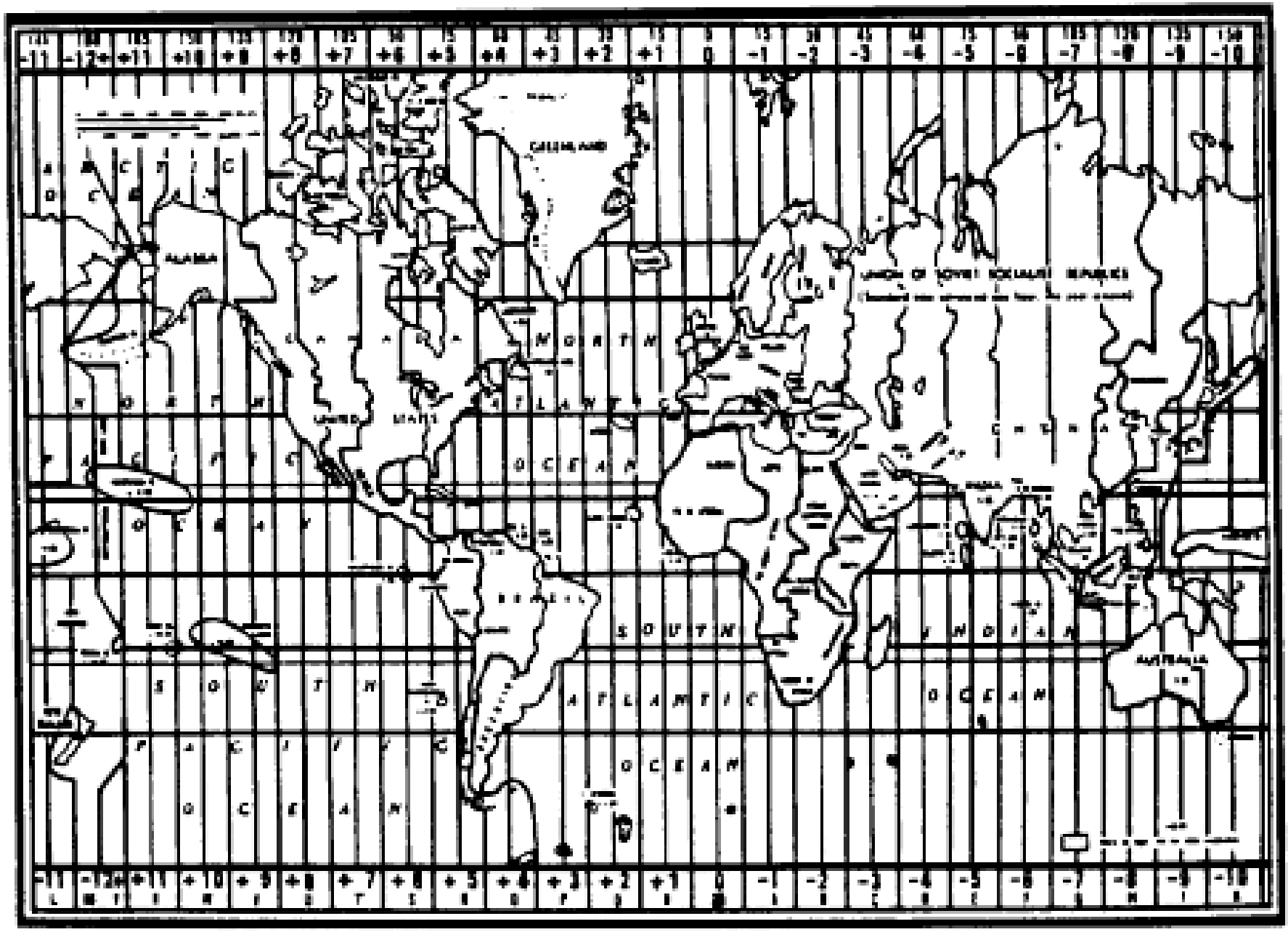
Mapped to SNOMED CT, ICD10 and old EDTA PRD codes

Benefits of SNOMED CT for nephrology
Set will be maintained by
Nephrologists, epidemiologists and terminologists

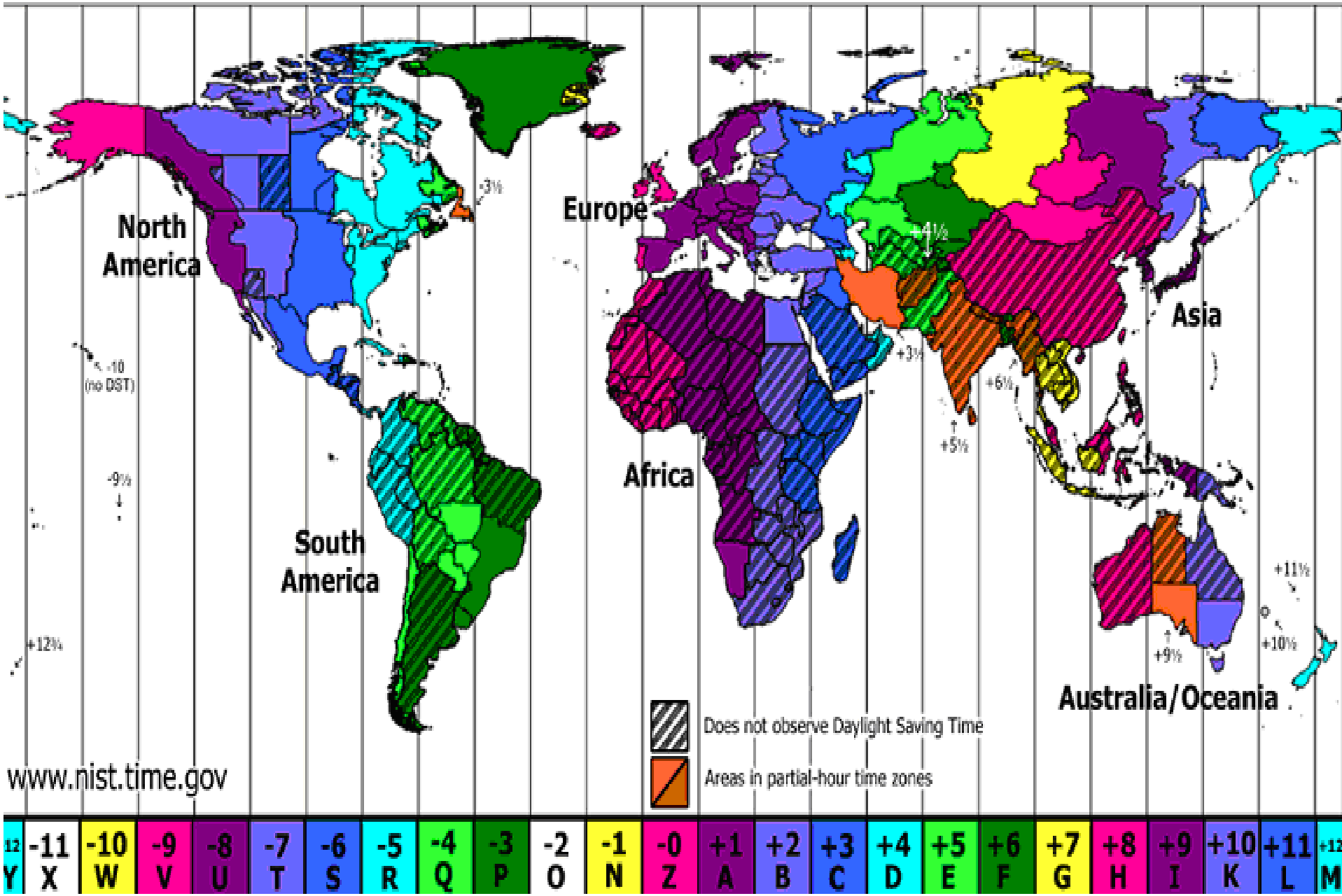
Results

Crude relative risk of death for incident RRT patients by Country and region




GREENLAND
ALASKA
UNITED STATES
UNION OF SOVIET SOCIALIST REPUBLICS
NORTH ATLANTIC OCEAN
SOUTH ATLANTIC OCEAN
INDIAN OCEAN
AUSTRALIA
PACIFIC OCEAN

ISO Time Zones



Publication:
Nephrology Dialysis & Transplantation

Translated to Italian and German

Will be adopted by National Registries affiliated the the ERA-EDTA

Maintained by an ERA-EDTA Registry sub-committee with help from a professional terminologist

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