New primary renal diagnosis codes for the ERA-EDTA

Gopalakrishnan Venkat-Raman1, Charles R.V. Tomson2, Yongsheng Gao3, Ronald Cornet4, Benedicte Stengel5, Carola Gronhagen-Riska6, Chris Reid7, Christian Jacquelinet8, Elke Schaeffner9, Els Boeschoten10, Francesco Casino11, Frederic Collart12, Johan De Meester13, Oscar Zurriaga14, Reinhard Kramar15, Kitty J. Jager16 and Keith Simpson16, for the ERA-EDTA Registry*

Benedicte Stengel5, Carola Gronhagen-Riska6, Chris Reid7, Christian Jacquelinet8, Elke Schaeffner9, Bruxelles, Belgium,13Department of Nephrology, Dialysis and Hypertension, AZ Nikolaas, Sint-Niklaas, Belgium,14Registro de Universitätsmedizin Charité, Charité Campus Virchow Klinikum, Berlin, Germany,10Hans Mak Institute, Naarden, The Netherlands, NHS Data Standards & Products, NHS Connecting for Health, Princes Exchange, Princes Square, Leeds, UK, 4Academic Medical Center, Department of Medical Informatics, ERA-EDTA Registry, Amsterdam, The Netherlands, 15INSERM CESP, U1018, Épidémiologie et Biostatistiques, Villejuif Cedex, France, 2Department of Medicine, Helsinki University Hospital, Helsinki, Finland, 7Evelina Children’s Hospital, London, UK, 8Coordination Nationale, Réseau Epidémiologie et Information en Néphrologie, Agence de la biomédecine, Saint-Denis La Plaine Cedex, France, 3Medizinische Klinik m.S. Nephrologie und Int. Intensivmedizin, Universitätsmedizin Charité, Charité Campus Virchow Klinikum, Berlin, Germany, 10Hans Mak Institute, Naarden, The Netherlands, 11Nephrology, Ospedale Madonna delle Grazie, Matera, Italy, 12Clinique de Nephrologie et Dialyse, CHU Brugmann, Bruxelles, Belgium, 13Department of Nephrology, Dialysis and Hypertension, AZ Nikolaas, Sint-Niklaas, Belgium, 14Registro de Enfermos Renales de la Comunitat Valenciana, Conselleria de Sanitat y CIBERESP, Spain, 15Austrian Dialysis & Transplant Registry, Scharmuehle, Kematen, Austria and 16Renal Unit, Glasgow Western Infirmary, Glasgow, UK

Correspondence and offprint requests to: Kitty J. Jager; E-mail: k.j.jager@amc.uva.nl
*The Authors are members of the ERA-EDTA Registry Coding and Definitions Working Group.

Abstract

The European Renal Association-European Dialysis and Transplant Association (ERA-EDTA) Registry has produced a new set of primary renal diagnosis (PRD) codes that are intended for use by affiliated registries. It is designed specifically for use in renal centres and registries but is aligned with international coding standards supported by the WHO (International Classification of Diseases) and the International Health Terminology Standards Development Organization (SNOMED Clinical Terms). It is available as supplementary material to this paper and free on the internet for non-commercial, clinical, quality improvement and research use, and by agreement with the ERA-EDTA Registry for use by commercial organizations. Conversion between the old and the new PRD codes is possible. The new codes are very flexible and will be actively managed to keep them up-to-date and to ensure that renal medicine can remain at the forefront of the electronic revolution in medicine, epidemiology research and the use of decision support systems to improve the care of patients.

Keywords: codes; ERA-EDTA; ICD; PRD; SNOMED CT

Introduction

Accurate reporting of basic epidemiology data underpins the practice of medicine, health care planning and social policy. The European Dialysis and Transplant Association (EDTA) has promoted epidemiology research since its formation in 1963. The EDTA Registry was established in 1964. At that time it published a list of diagnoses that led to end-stage renal failure (ESRF) [1]. This was called the primary renal diagnosis (PRD) list. In 1983, the EDTA changed its name to the European Renal Association–European Dialysis and Transplant Association (ERA-EDTA) and the EDTA Registry became the ERA–EDTA Registry.

Patients who started renal replacement therapy (RRT) for ESRF in countries affiliated to the ERA-EDTA (and formerly the EDTA) were registered by their renal centres and were followed up annually. Initially, this was done by returning a paper form to the Registry office where a computer database was updated. The ERA-EDTA thus established a voluntary comprehensive, longitudinal, international registry many years before other medical specialties and often reported more detailed information than was available even to national authorities including incidence, prevalence, diagnosis, treatment modality and survival.

The adoption of a set of standard diagnostic terms improves communication and supports data analysis. Unfortunately, it can also coerce users, impose unreasonable constraints and provide a false sense of accuracy. Code sets must be designed and used carefully with these benefits and limitations in mind. It must be possible to record events in free text in the primary medical record in order to preserve details and context...
and to allow previously unrecognized entities to emerge.

In 2000, the ERA-EDTA Registry office, which at that point was based in London, was reorganized and returned to Amsterdam [1, 2]. The responsibility for patient registration and follow-up was devolved on national and regional renal registries, which collected the data from renal centres, undertook initial validation and sent them in an agreed, secure electronic format to the ERA-EDTA Registry. These data files were sometimes augmented with specially collected data and were used to produce annual reports on incidence, prevalence, treatment methods and survival and scientific papers. They demonstrate the continuing value of good-quality long-term observational studies [3–6]. However, deficiencies imposed by the limited options for recording the PRDs caused frustration [7]. This paper describes the development and publication of a new list of PRD codes that adheres to international standards and will extend their use and reliability. In line with the Strengthening the Reporting of Observational Studies in Epidemiology recommendations for reporting observational studies [8], we provide background on the choices made during the development process.

The problem

The ERA-EDTA Registry Committee recognized that after having served well for nearly 40 years, the old PRD codes were incomplete and inflexible. The terms lacked definitions, used the word ‘other’ without qualification and because there were no guidelines, the PRDs were applied inconsistently between and even within countries. It was not possible to indicate the accuracy of the code used and there was no formal mechanism for adding new codes or retiring redundant ones.

In the 1960s, when the PRDs were introduced, computers were not widely used in clinical practice, and at that time, it was important that the list could be printed on a single sheet of paper.

Initially, there was very limited quality assurance and data validation, and it was clear that while attempting to convey the limited but useful insights they had about patients, some nephrologists were coding with spurious accuracy (e.g. a PRD of Immunoglobulin A (IgA) nephropathy based purely on clinical presentation). The PRD assigned may have reported the clinical diagnosis but it was sometimes inadequate for secondary uses and epidemiological analysis. There was widespread variation in the use of some codes and it was difficult to find the appropriate codes for some patients, particularly those with systemic vasculitis, where understanding and classification have changed greatly since the PRD list was first agreed. Over the decades, other new diseases have been described that did not fit into existing categories. These problems could not be fixed simply by extending the old PRD list and it was therefore decided that a completely new list of PRD terms and a new philosophy for maintaining it would be produced.

Possible solutions

A Registry Coding and Definitions Working Group was established to develop a new list of Primary Renal Diagnoses and to report to the ERA-EDTA Registry Committee as part of the QUIET European Studies (QUality) initiative [9]. The brief was: ‘to improve and standardize the definitions, terminology and coding used by renal registries in Europe to describe primary renal diagnoses’.

The first task was to review the existing codes and to seek the views of the national and regional registries affiliated to the ERA-EDTA. That was done by a questionnaire and discussion at meetings of registries during ERA-EDTA congresses. It was apparent that there were many very different, well-considered and deeply held views. Some countries had also modified the code set by expanding some sections.

A similar problem has been clearly reviewed by Agar et al. [10] in a paper on the terminology of chronic maintenance haemodialysis. They point out that the meaning of terms varies from one geographical region to another and that many publications prompt the reader to ask ‘What exactly did they mean?’ They offer suggestions for improving the situation but realistically warn that changes will not always be welcomed, particularly by those who use terms that have been rejected. In the development of the new PRD codes, we faced the same problem. The key point is that to be of full use in later epidemiology and clinical studies, the contextual information or ‘meta data’ which supports the PRD and other clinical data must be recorded at the same time as the diagnosis and inextricably linked.

In their response to our questionnaire and in subsequent discussions, renal registries expressed a huge range of sometimes irreconcilable views concerning the new PRD terms. It became apparent that many registries and national authorities were already committed to using the International Classification of Diseases (ICD)-10 or the SNOMED Clinical Terms (SNOMED CT). The latter nomenclature offers enormous benefits and is fully maintained by the International Health Terminology Standards Development Organization (IHTSDO), a not-for-profit multinational association.

The main suggestions from Registries are summarized in the following list, which retains the telegraphic style of the questionnaire responses:

(i) simply retain the existing PRD list without modification because it has served well and is understood,
(ii) adopt ICD-10 without modifications,
(iii) adopt SNOMED CT without modifications,
(iv) create a new list to be maintained entirely by the ERA-EDTA Registry,
(v) create a new list to be maintained by the ERA-EDTA Registry and keep it aligned with other international coding schemes,
(vi) include all aspects of a PRD in a single line of text that does not require further qualification,
Elsewhere Classification

It was clear that a ready-made solution was not available anywhere else. This is the common strategy adopted by classification systems that are attempting to achieve complete coverage. The concept of 'other' changes every time the code set is altered. Even with good version control, it is often not clear what alternatives were available when a 'NOS' code was used, and therefore, it was unclear what it might include or exclude. Furthermore, ambiguity is introduced when new codes are added because the domain deferred to 'other' changes every time the code set is altered.

Modern medical terminology systems try to avoid this problem by capturing clinical information at the level of detail appropriate for clinical practice and by allowing new codes to be added in an easy but controlled way. The approach now favoured is to provide high-level terms called 'generic', 'less granular' or 'superconcept'. This allows complex analysis of huge databases and enables appropriate literature to be linked to individual patient records for optimal clinical practice and decision support. Care must be taken when generating detailed (granular) terms from a diagnosis that was initially saved at a generic (less granular) level but the utility of automatically providing all the available attributes of a diagnostic term is obvious.

The new PRD list

The 2012 ERA-EDTA PRD codes are published as supplementary material to this paper. In addition, they are freely available in a spreadsheet and in a searchable web browser on the ERA-EDTA Registry website at www.era-edta-reg.org.

As requested by the national registries, the 273 new PRD terms not only include all the common and many rare nephropathies that result in ESRF, but also many other kidney conditions which do not usually cause advanced chronic kidney disease (CKD).

In the list, each new PRD term has been assigned a unique number, which serves only to identify it and to help electronic communication. We came to the conclusion that a single hierarchy of codes would be impossible. For example, it would be equally logical to include familial IgA nephropathy in a 'familial' category or in a 'glomerulonephritis' category. The many complex ways in which lists of diagnoses can be searched, sorted and examined are already well handled in SNOMED CT and these tools will be available when the new PRD terms are accessed on a modern computer browser. A default sorting order decided by the working group is included so that if required, the list can be viewed in a standard order. This default order has no particular significance and users can rearrange the list and use any convenient software tools to search, sort and manipulate it. If required, printed copies can be used. Suggestions for addition or inactivation of terms should be sent to the ERA-EDTA Registry office at erareg@amc.uva.nl.
Further details about the codes and their attributes are given in ‘Notes for Users’ which are published with the codes. An unusual feature of the PRDs is the inclusion of the term ‘histologically proven’ and ‘no histology’ even for some PRDs, which should not reasonably be used without histological evidence. These terms are used to satisfy the requirement to provide detail, as a measure of the certainty of the diagnosis and to say everything in a single line of text. We provide PRDs that contain the words ‘no histology’ for diagnoses that clearly require histological proof (eg ‘IgA nephropathy - no histology’). It is clear from Registry records that PRDs which describe the histological appearance of a kidney biopsy are frequently used even when histological evidence is not available and an alternative less granular PRD would be more suitable. We do not condone this practice. We hope that nephrologists who wish to record such a diagnosis will use the PRD which contains the words ‘no histology’ so that if required they can be analysed separately from the similar PRD which contains the words ‘histologically proven’. We hope that it will henceforth be possible to distinguish an honest guess from a dishonest guess from a

We know from previous Registry analyses that many patients with Type 2 diabetes are incorrectly coded as Type 1. There is also marked variation in the use of codes for diabetic glomerulosclerosis, suggesting that clinicians may have chosen these codes without considering the possibility of alternative aetiologies for kidney disease in patients with diabetes mellitus.

The new classification gives additional guidance on the choice of the code in this situation. For instance, the code ‘2337 Diabetic nephropathy in Type II diabetes—no histology’ includes the guidance:

(i) A diagnosis of Type II diabetes mellitus must have been made.
(ii) For a diagnosis of diabetic nephropathy, proteinuria must have been documented at some point in the patient’s history.
(iii) A PRD of diabetic nephropathy is not mandatory in the presence of DM with proteinuria and alternative diagnoses can be considered.
(iv) In the absence of renal histology, the differential diagnosis will include ‘Chronic kidney disease (CKD) / chronic renal failure (CRF) - aetiology uncertain/unknown - no histology’, ischaemic nephropathy, renovascular disease and atheroembolic renal disease.
(v) Distinguish from: Inherited/genetic diabetes mellitus Type II.

In addition, newly described nephropathies have been added to the PRD coding system, e.g.

2274 Nephropathy related to HIV—no histology.
2288 Nephropathy related to HIV—histologically proven.

Finally, many rare diagnoses were omitted from the old PRD list but are now included, e.g.

1074 Denys–Drash syndrome,
2929 Dent disease and
2938 Lowe syndrome (oculocerebrorenal syndrome).

When a definitive test has not been used, there will always be uncertainty, but the new PRD codes allow both
a clinical diagnosis to be recorded faithfully. At the same time, by noting the absence of a definitive test, we can also convey the degree of uncertainty. Where appropriate, patient records can be grouped according to the degree of accuracy and certainty required for a particular analysis and research teams can decide whether to examine a small number of patients with accurate diagnoses or larger numbers with phenotypic similarities. Cohorts can be combined if required and nothing is lost by using the more granular coding scheme apart from the slight additional effort of choosing the best diagnostic term from a larger list. With computer aids, the extra effort is trivial although we must recognize that we now have more diagnostic options than we can commit to memory.

No current coding system in widespread use has full definitions. While desirable, this would be a huge task and would require extensive international and cross-specialty collaboration. In our PRD list, we have made some progress by providing partial definitions that indicate what type of diagnostic information should be used to support each PRD but with the exception of histological evidence, these are not mandatory and we must still rely on the good judgement of individual nephrologists.

Fortuitously, while the work on the PRDs was proceeding, the renal community in the UK was establishing a subset of existing renal SNOMED CT codes and they agreed to incorporate the new ERA–EDTA PRD codes in their list without modification. That work introduced one of the authors (Y.G.), who is an expert in clinical terminology, to the PRD codes. He was co-opted onto the PRD working group and undertook the detailed mapping of the new PRDs to SNOMED CT and ICD-10. Not only did that ensure that the terms favoured by the nephrologists were acceptable to professional terminologists, but it also allowed the new PRDs to be aligned to SNOMED CT with all the subtle semantic links that make it such a powerful tool.

Each new PRD has been linked (often called mapping) to the most appropriate PRD in the old code set and vice versa. These translation tables are offered for use where automated conversion of large numbers of records is required. For detailed research work where the historic coding practice is well understood, it may be appropriate to develop alternative mappings or to re-examine individual records to ensure that the correct new PRD has been chosen. They need not be followed slavishly but users are encouraged to publish the mapping tables they use along with their results. Conversion from old to new PRDs may be necessary when historic data are being combined with contemporary data or when historic data with modern data analysis techniques that use SNOMED CT or ICD codes.

In addition to allowing conversion between new and old PRD codes, a powerful feature of the new PRD coding system is that the assignment of an ICD-10 code, a SNOMED CT identifier and where necessary a set of post-coordinated SNOMED CT codes will allow users to access the full power and utility of these modern and internationally supported clinical terminology systems. The most immediate and obvious benefit of a link to SNOMED CT may be the provision of validated translation into other languages and the possibility of participating in further translation work if required. This makes the codes and their extended uses (e.g. links to literature and semantic links) available to non-English speakers and allows codes to be entered in one language and displayed in another. A single example persuaded the coding group of the utility of this approach. We considered the diagnosis of ‘renal vein thrombosis’. Using a SNOMED CT browser and irrespective of the language that is used or whether the fully specified name or a local preferred term is used, the relationship to renal disorders, venous disorders and thrombotic disorders is clear. SNOMED CT uses words with which we are familiar but behind the scenes it preserves the concepts and the true biological meaning via the codes and not simply by looking up the words.

The introduction of SNOMED CT into clinical practice is still at an early stage but its power and potential are obvious and the ERA-EDTA will be able to contribute to it as it develops. The new PRD list also has extensive links to the Online Mendelian Inheritance in Man (OMIM) database, which is a comprehensive medical and scientific resource maintained by US National Library of Medicine and the William H. Welch Medical Library at Johns Hopkins University. The online version was developed by the US National Center for Biotechnology Information.

The ERA-EDTA Registry will accept patient data returns using the new PRDs from its contributing registries from January 2012. It will continue to accept data using the old PRDs for some time and it will announce on its website 2 years before the date on which it will no longer be able to accept the old PRDs.

Further developing the codes and putting them to use

Future development of the codes will be the responsibility of the ERA-EDTA Registry committee and its coding group. The initial task will be to collaborate with other specialties under the general guidance of IHTSDO to develop codes relevant to the disorders on the boundary with another specialty or affecting more than one organ, e.g. extra-renal vasculitis and amyloidosis. We hope that national registries and renal centres will help us to improve the PRD list by notifying the Registry of any errors, omissions, redundancies, clarifications or new terms that are required. Within the structure of SNOMED CT, there are stable mechanisms for undertaking this work, which allows the codes to evolve while retaining all the information in the existing records.

We believe that we have produced a useful new list of PRDs which satisfies most of the requirements set by renal registries affiliated to the ERA-EDTA and which incorporates most of the suggestions from colleagues who reviewed the work.

The new PRD codes will be maintained by the ERA-EDTA Registry and are aligned with major international coding schemes.
Supplementary data

The ‘2012 ERA-EDTA Primary Renal Diagnosis Codes’ are available as supplementary data online at http://ndt.oxfordjournals.org.

Acknowledgements. This work was part of the ERA-EDTA Registry QUEST initiative. We thank the members of the national and regional renal registries affiliated to the ERA-EDTA Registry who provided the impetus and the specification for this work and senior members of the IHTSDO and WHO ICD group.

Conflict of interest statement. None declared.

References

1. Briggs JD. The ERA-EDTA Registry returns to Amsterdam. Nephrol Dial Transplant 2000; 15: 1326–1327
2. Briggs JD, Jager K. The first year of the new ERA-EDTA Registry. Nephrol Dial Transplant 2001; 16: 1130–1131
3. Geddes CC, van Dijk PCW, McArthur S et al. The ERA-EDTA cohort study–comparison of methods to predict survival on renal replacement therapy. Nephrol Dial Transplant 2006; 21: 945–956
4. Jager KJ, Stel VS, Wanner C et al. The valuable contribution of observational studies to nephrology. Kidney Int 2007; 72: 671–675
5. Jager KJ, van Dijk PCW, Dekker FW et al. The epidemic of aging in renal replacement therapy: an update on elderly patients and their outcomes. Clin Nephrol 2003; 60: 352–360
6. Tsakiris DJ, Stel VS, Finne P et al. Incidence and outcome of patients starting renal replacement therapy for end-stage renal disease due to multiple myeloma or light-chain deposit disease: an ERA-EDTA Registry study. Nephrol Dial Transplant 2010; 25: 1200–1206
7. Ronco P. Disease classification: a pitfall of the ERA/EDTA registry? Nephrol Dial Transplant 2010; 25: 1022–1024
8. von Elm E, Altman DG, Egger M et al. Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement: guidelines for reporting observational studies. Br Med J 2007; 335: 806–808
9. Jager KJ, Zoccali C. QUality European STudies (QUEST) a step forward in the quality of RRT care. Nephrol Dial Transplant 2005; 20: 2005–2006
10. Agar JW, MacGregor MS, Blagg CR. Chronic maintenance hemodialysis: making sense of the terminology. Hemodial Int 2007; 11: 252–262

Received for publication: 10.5.2012; Accepted in revised form: 30.8.2012