Positive Predictive Values of International Classification of Diseases, 10th Revision Coding Algorithms to Identify Patients With Autosomal Dominant Polycystic Kidney Disease

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Abstract

Background: International Classification of Diseases, 10th Revision codes (ICD-10) for autosomal dominant polycystic kidney disease (ADPKD) is used within several administrative health care databases. It is unknown whether these codes identify patients who meet strict clinical criteria for ADPKD.

Objective: The objective of this study is (1) to determine whether different ICD-10 coding algorithms identify adult patients who meet strict clinical criteria for ADPKD as assessed through medical chart review and (2) to assess the number of patients identified with different ADPKD coding algorithms in Ontario.

Design: Validation study of health care database codes, and prevalence.

Setting: Ontario, Canada.

Patients: For the chart review, 201 adult patients with hospital encounters between April 1, 2002, and March 31, 2014, assigned either ICD-10 codes Q61.2 or Q61.3.

Measurements: This study measured positive predictive value of the ICD-10 coding algorithms and the number of Ontarians identified with different coding algorithms.

Methods: We manually reviewed a random sample of medical charts in London, Ontario, Canada, and determined whether or not ADPKD was present according to strict clinical criteria.

Results: The presence of either ICD-10 code Q61.2 or Q61.3 in a hospital encounter had a positive predictive value of 85% (95% confidence interval [CI], 79%-89%) and identified 2981 Ontarians (0.02% of the Ontario adult population). The presence of ICD-10 code Q61.2 in a hospital encounter had a positive predictive value of 97% (95% CI, 86%-100%) and identified 394 adults in Ontario (0.003% of the Ontario adult population).

Limitations: (1) We could not calculate other measures of validity; (2) the coding algorithms do not identify patients without hospital encounters; and (3) coding practices may differ between hospitals.

Conclusions: Most patients with ICD-10 code Q61.2 or Q61.3 assigned during their hospital encounters have ADPKD according to the clinical criteria. These codes can be used to assemble cohorts of adult patients with ADPKD and hospital encounters.

Abrégé

Mise en contexte: La 10e révision des codes de l’International Classification of Diseases (ICD-10) est utilisée dans plusieurs bases de données administratives des centres de soins pour le classement de la maladie polykystique autosomique dominante (MPR). On ignore toutefois si ces codes permettent d’identifier clairement les patients qui satisfont les critères cliniques stricts de la maladie.

Objectifs de l’étude: 1) Déterminer si les différents algorithmes de codage de la ICD-10 réussissent à identifier de manière efficace les patients adultes satisfaisant les critères cliniques stricts de la MPR tels qu’évalués par la consultation des dossiers médicaux; 2) Évaluer le nombre de patients qui sont identifiés par les différents algorithmes de codage pour la MPR, en Ontario.

Cadre et type d’étude: Il s’agit d’une étude de validation des codes de classification obtenus dans les bases de données des centres de soins de l’Ontario, au Canada, ainsi que de leur prévalence.
Patients: On a révisé les dossiers médicaux de 201 patients adultes ayant reçu une consultation en centre hospitalier entre le 1er avril 2002 et le 31 mars 2014, et à qui les codes ICD-10 Q61.2 ou Q61.3 pour la MPR ont été assignés.

Mesures: Les valeurs prédictives positives des algorithmes de codage ICD-10 ainsi que le nombre d’Ontariens identifiés comme patients atteints de MPR par les différents algorithmes de codage ont été retenus pour l’étude.

Méthodologie: Un échantillon aléatoire de dossiers médicaux en provenance de London, en Ontario (Canada) a été révisé manuellement afin de déterminer lesquels indiquaient la présence d’une MPR selon les critères cliniques stricts pour cette maladie.

Résultats: La présence des codes ICD-10 Q61.2 ou Q61.3 lors d’une consultation à l’hôpital a eu une valeur prédictive positive dans 85% des cas (IC 95%: 79 à 89%), et a permis l’identification d’un total de 2 981 patients ontariens (0,02% de la population adulte en Ontario). Le codage ICD-10 Q61.2 à lui seul a eu une valeur prédictive positive dans 97% des cas (IC 95%: 86 à 100%) et a permis l’identification de 394 patients (0,003% de la population adulte en Ontario).

Limites de l’étude: 1) Nous n’avons pu calculer aucune autre mesure de validité; 2) Les algorithmes de codage n’identifient pas les patients s’ils ne sont pas en consultation en centre hospitalier; 3) Les pratiques de codage peuvent varier d’un hôpital à un autre.

Conclusions: La majorité des patients codés ICD-10 Q61.2 ou Q61.3 à la suite d’une consultation en centre hospitalier était atteinte de maladie polykystique autosomique dominante selon les critères cliniques stricts pour cette maladie. Ainsi, cette codification peut être utilisée pour jumeler des cohortes de patients adultes atteints de MPR avec leurs consultations en hôpital.

Keywords
positive predictive value, administrative data, polycystic kidney disease, validation study, validity

What Was Known Before?
Health care administrative databases may be an attractive way to identify population-based samples of patients with autosomal dominant polycystic kidney disease (ADPKD) as long as the administrative codes used to identify such patients are accurate.

What Does This Add?
Most patients with International Classification of Diseases, 10th Revision codes (ICD-10) have ADPKD according to the strict clinical criteria. These codes can be used to assemble a study cohort of adult patients with ADPKD and hospital encounters.

Background
ADPKD is a genetic condition characterized by focal cyst development leading to bilateral enlargement of both kidneys.1 Approximately, half of these patients will require end-stage kidney disease care by the age of 50.2 ADPKD has an estimated prevalence of 1 in 1000 to 1 in 400 (0.1%-0.25%) persons worldwide.3 As ADPKD is a relatively uncommon disease, using large health care administrative databases may allow a large number of patients with ADPKD to be identified and studied in a time-efficient and cost-effective manner.4 However, this approach requires assurances that ADPKD is coded accurately in these data sources and an appreciation that different administrative databases only apply to patients with certain health care encounters (eg, hospital records only apply to ADPKD patients with at least 1 hospital encounter during a period of interest). Furthermore, information available from administrative databases are collected primarily to monitor health care use and to assess health care needs, without the same rigor used in clinical research studies to assess conditions of interest.5 Physician misdiagnoses, incomplete documentation in medical records, or errors by personnel who assign codes to each hospital...
encounter can all potentially lead to misclassification of a condition.6

We conducted a comprehensive search of bibliographic databases (search last performed December 2015) and found only a single study in the United States that described any aspect of the accuracy of health care administrative database codes for ADPKD. Blanchette and colleagues7 assessed the positive predictive value of a single International Classification of Diseases, Ninth Revision (ICD-9) code for any kind of polycystic kidney disease (PKD) (753.12), where a medical chart review was used to ascertain whether PKD was truly present or not. In this study, the clinical criterion used to define PKD in the medical chart was not defined. In addition, despite knowing that the population comprised of members of commercial health plans, it was not clear whether the charts were from an outpatient and/or hospital-based setting.7 In 132 patients, the positive predictive value of ICD-9 code 753.12 was 95%, indicating that most patients identified with the ICD-9 code 753.12 had ADPKD according to their medical chart review.7

We undertook 2 studies. First, we determined whether different coding algorithms containing ICD-10 codes for ADPKD assigned during hospital encounters (emergency room visits or hospital admissions) can be used to identify adult patients who meet the clinical criteria for ADPKD in the province of Ontario, Canada. This was done to estimate the positive predictive value of various coding algorithms considering the manual chart review and a rigorous definition of ADPKD as the reference standard. Second, we used Ontario-wide health care databases to assess the number of patients identified with different sets of ADPKD codes to determine the proportion of the general public identified with ADPKD with each of the coding algorithms (where the expected prevalence is 0.1%-0.25%).

Methods

Study Design

For our first study, we manually reviewed inpatient and outpatient medical records (including both electronic medical records and paper charts) to assess the positive predictive values of different ICD-10 coding algorithms for ADPKD. For our second study, we conducted analyses of large health care databases housed at the Institute for Clinical Evaluative Sciences (ICES) to understand the frequency of ICD-10 coding algorithm use in the province of Ontario, Canada.8

Data Sources and Database Algorithms

The World Health Organization (WHO) developed the ICD-10 codes collaboratively with 10 international centers to promote comparability in mortality data across countries. In Canada, the National Implementation Advisory Committee (established by the Canadian Institute for Health Information [CIHI]) modified and enhanced some of the ICD-10 codes developed by WHO to better accommodate Canadians’ administrative, epidemiological, and public health research needs prior to implementation. ICD-10-CA is the Canadian modification of the ICD-10 codes. The ICD-10 codes related to ADPKD used in Canada were not modified and are identical to those developed by the WHO.

ICD-10 and ICD-10-CA codes are used in Canadian administrative databases such as the CIHI Discharge Abstract Database (CIHI-DAD) and the CIHI National Ambulatory Care Reporting System (CIHI-NACRS). The CIHI-DAD houses administrative, demographic, and clinical information on hospital discharge and day surgery procedures, and the CIHI-NACRS database contains information on all emergency room visits.9 Neither CIHI-DAD nor CIHI-NACRS houses information on outpatient physician office visits. Trained personnel at each hospital in Ontario reviewed the medical charts of all patients with health care encounters. Based on the rules and guidelines provided by CIHI, these trained personnel coded all diagnoses and procedures using the ICD-10 coding system, and then entered these codes into the CIHI-DAD and CIHI-NACRS databases.8 These trained personnel only consider physician-recorded diagnoses in a patient’s medical chart when assigning the codes, and do not review or interpret diagnostic imaging reports, laboratory values, family history, or signs and symptoms of ADPKD.

In this study, we compiled a list of relevant ICD-10 codes for ADPKD (Table 1) and developed 9 unique algorithms using 2 databases (CIHI-DAD and CIHI-NACRS) and 2 ICD-10 codes, Q61.2 (PKD, autosomal dominant) and Q61.3 (PKD, unspecified).

| Table 1. International Classification for Diseases, 10th Revision Codes Relevant for Autosomal Dominant Polycystic Kidney Disease. |
|---|
| Database | Code | Description |
| CIHI-DAD | Q61.3 | Polycystic kidney disease—unspecified |
| CIHI-DAD | Q61.2 | Polycystic kidney disease—autosomal dominant |
| CIHI-NACRS | Q61.3 | Polycystic kidney disease—unspecified |
| CIHI-NACRS | Q61.2 | Polycystic kidney disease—autosomal dominant |

Patient Selection

For the chart abstraction study, we compiled a list of adult patients (age ≥18 years) with emergency department visits and/or hospital admissions (CIHI-DAD or CIHI-NACRS) assigned the ICD-10 Q61.2 code, Q61.3 code, or both codes between April 1, 2002, and March 31, 2014, at 2 major teaching hospitals (Victoria Hospital and University Hospital) in
London, Ontario. We assigned a unique patient identification number (ID) to each patient and saved a list of all patients’ medical record numbers and patient IDs in a password-protected Microsoft Excel file, which was stored on a secure hospital network. If a patient had more than 1 code or more than 1 hospital and/or ambulatory care encounter, we assigned the unique patient ID to the first hospital or ambulatory care encounter. We included all patients with ICD-10 code Q61.2. We then stratified all patients with ICD-10 code Q61.3 by database (CIHI-DAD or CIHI-NACRS) and by year of hospital encounter, and randomly sampled within strata to review the medical records of a total of 201 patients from a list of 305 patient charts eligible for review.

For the ICES study, we linked CIHI-DAD, and CIHI-NACRS using unique encoded identifiers, which were analyzed at ICES. We identified all adult patients who were assigned either an ICD-10 Q61.2 code or Q61.3 code during an emergency department visit or hospital admission between April 1, 2002, and March 31, 2014. For patients with more than 1 hospital encounter, we only considered the first encounter.

**Data Collection**

For the chart abstraction study, we manually reviewed the medical records of the 201 patients. We abstracted information on physician report of ADPKD, family history of ADPKD, indication of ADPKD on surgical pathology reports or autopsy reports, and information from imaging reports (reason for examination, number of cysts in each kidney, and dimensions of each kidney). Certain imaging reports did not specify the exact number of cysts. In these instances, we interpreted “multiple cysts bilaterally” as at least 3 cysts in each of the 2 kidneys and “innumerable cysts bilaterally” as at least 4 cysts in each of the 2 kidneys. Sensitivity analysis were performed to determine whether interpreting “multiple cysts bilaterally” as at least 4 cysts in each kidney meaningfully changed the results. If information was missing in an electronic medical record, we obtained the paper inpatient chart. If information was still missing after reviewing the paper chart, we reviewed the nephrology outpatient chart when available. Subsequently, a senior radiology resident (M.R.) retrieved and reviewed available diagnostic images for patients with missing or ambiguous information. We recorded all abstracted information onto a detailed data abstraction form.

**Clinical Definition of ADPKD**

In the chart abstraction study, 2 reviewers (V.K. and R.K.M.) independently determined whether each of the 201 patients had ADPKD or not using strict criteria (described in the next paragraph). These criteria were developed in consultation with 2 experienced nephrologists (A.X.G. and Y.P.). To determine the final ADPKD status, any disagreements between the 2 reviewers were resolved by consensus.

First, we assessed whether patients met the internationally accepted diagnostic criteria for ADPKD, which requires the presence of a positive family history of ADPKD and evidence of the following number of cysts on a conventional kidney ultrasound: (1) for patients 15 to 39 years old, at least 3 cysts when one counts the total number of cysts in both kidneys combined; (2) for patients 40 to 59 years old, at least 2 cysts in each kidney; and (3) for patients 60 years of age or older, at least 4 cysts in each kidney. Second, we classified patients with a negative or indeterminate family history of ADPKD as affected if they had innumerable cysts in both kidneys with each kidney greater than 13 cm in length. Third, we classified all patients who had a nephrectomy performed and with a diagnosis of ADPKD in a surgical pathology or autopsy report as affected irrespective of their ADPKD family history status. Finally, we classified patients with missing imaging reports as affected with ADPKD if they had a family history of ADPKD and a clear physician-reported diagnosis of ADPKD. When ADPKD status was still ambiguous, an experienced nephrologist (A.X.G.) reviewed all medical records to make a determination of whether ADPKD was present or not according to clinical criteria. When there was insufficient information to make a determination of whether ADPKD was present or not, patients were excluded from the analysis. Sensitivity analyses were performed to determine whether classifying the excluded patients as having ADPKD, or as not having ADPKD, meaningfully changed the results.

**Analysis**

For the chart abstraction study, we expressed continuous variables as median and interquartile ranges (IQRs) and binary variables as percentages. We calculated the positive predictive value for each of the 9 coding algorithms and calculated their respective 95% confidence intervals (CIs) using the Wilson score method. For the ICES study, we estimated the number of patients with ADPKD in Ontario identified with different sets of codes and calculated the percentage of adult Ontarians with the code sets. We conducted all statistical analyses using SAS 9.3 (SAS Institute, Inc, Cary, North Carolina).

**Results**

**Chart Abstraction Study Sample**

We obtained a list of unique patients with ICD-10 codes Q61.3 and Q61.2 using the CIHI-DAD and CIHI-NACRS databases. We then included all patients with the ICD-10 code Q61.2 and stratified random sampled patients with ICD-10 code Q61.3 to sample a total of 201 patients. We abstracted information using electronic medical records for all 201 patients, inpatient charts for 117 patients, and nephrology outpatient charts for 52 patients. A senior radiology
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resident (M.R.) reviewed the images of 65 patients with ADPKD because imaging reports did not clearly provide all the required information. After excluding 14 patients because of insufficient information to determine ADPKD status, our final cohort consisted of 187 patients.

Chart Abstraction Patient Characteristics

Among the 187 patients identified in our cohort through database codes, median (IQR) patient age was 61 (53-70), and 95 (50%) were men. Family history of ADPKD was positive in 116 (62%) patients, negative in 42 (22%) patients, and was missing or indeterminate in 29 (16%) patients. A total of 158 (85%) patients met the clinical criteria of ADPKD. The number and percentage of patients who satisfied each ADPKD criteria are presented in Table 2.

Coding Algorithm Positive Predictive Value and Frequency

The positive predictive values, their respective 95% CIs (from our chart abstraction study), and the number of Ontarians with the 9 different coding algorithms (from our ICES study) are presented in Table 3. The presence of either ICD-10 code Q61.2 or Q61.3 in either the CIHI-DAD or CIHI-NACRS database had a positive predictive value of 85% (95% CI, 79%-89%) and identified 2981 adults in Ontario (0.02% of the Ontario adult population). The presence of ICD-10 code Q61.2 in either the CIHI-DAD or CIHI-NACRS database had a positive predictive value of 97% (95% CI, 86%-100%) and identified 394 adults in Ontario (0.003% of the Ontario adult population). Sensitivity analyses did not meaningfully change the results.

Discussion

Although past studies have assessed the positive predictive value of different ICD-10 codes or coding algorithms for other diseases or conditions, there is a lack of information on the positive predictive value of ICD-10 coding algorithms for ADPKD. The positive predictive value is reported as a number from 0% to 100%, where a high value indicates that individuals who are identified with the coding algorithm truly have the condition. We manually reviewed a random sample of medical charts from 2 tertiary care hospitals in London, Ontario, where the medical coders in routine care had assigned a code for PKD. Using rigorous clinical criteria, we then determined whether ADPKD was present or not. We found that the presence of the ICD-10 code Q61.2 in hospital admissions or emergency visits had an excellent positive predictive value of 97% (95% CI, 86%-100%). The positive predictive value of the presence of either the ICD-10 code Q61.2 or Q61.3 in either hospital admissions or emergency visit was also good at 85% (95% CI, 79%-89%). Therefore, our study shows that administrative coding algorithms for ADPKD successfully identify patients who truly have ADPKD, which is consistent with the findings from a study conducted by Blanchette and colleagues.7 These values in the ADPKD setting are similar or better than the positive predictive value of ICD-10 codes or ICD-10 coding algorithms for shock (86%; 95% CI, 80%-91%), infant respiratory distress syndrome (81%; 95% CI, 73%-80%), and heart failure (84%; 95% CI, 81%-87%).12-14

Although our study has several strengths, results of this study must be interpreted with caution given the limitations. First, because we only reviewed the medical charts of patients with assigned ICD-10 database codes for ADPKD, we cannot estimate other measures of validity such as negative predictive

| Table 2. Number and Percentage of Patients Who Satisfied Each Criterion for ADPKD. |
|-------------------------------------|--------------------------------------|
| ADPKD criteria | Number of patients (%) |
|-------------------------------------|------------------------|
| Current ultrasonographic diagnostic criteria: family history and age-dependent, | 108 (53.73) |
| a. Ages 15 to 39: at least 3 cysts in 1 or both kidneys | |
| b. Ages 40 to 59: at least 2 cysts in each kidney | |
| c. Ages 60 and above: at least 4 cysts in each kidney | |
| No family history, both kidneys >13 cm and age-dependent minimal number of cysts | 37 (18.41) |
| a. Ages 15 to 39: at least 3 cysts in 1 or both kidneys | |
| b. Ages 40 to 59: at least 2 cysts in each kidney | |
| c. Ages 60 and above: at least 4 cysts in each kidney | |
| Indication of ADPKD in surgical pathology report or autopsy report | 7 (3.48) |
| Physician report of ADPKD and family history of ADPKD or patient has ADPKD | 6 (2.98) |
| based on nephrologist adjudication | |
| Did not meet any criteria | 29 (14.43) |
| Excluded from the study given a lack of information to make a determination of whether ADPKD was present or not | 14 (6.97) |

Note. These data were obtained from chart review. In accordance with privacy regulations, cell sizes less than or equal to 5 cannot be reported. ADPKD = autosomal dominant polycystic kidney disease.
value, sensitivity, and specificity. We expect the sensitivity of the ICD-10 codes for ADPKD to be low. As the prevalence of ADPKD is estimated to be 1 in 1000 to 1 in 400, we would expect 13,000 to 32,500 Ontarians to be affected with ADPKD. However, the expansive coding algorithm (any of the 2 ICD-10 codes in CIHI-DAD or CIHI-NACRS) only identified approximately 3000 patients. Thus, although the 2 ICD-10 codes appear to have a high positive predictive value, it is possible only 9% to 23% of the patients with ADPKD in the province were captured with the algorithm.

Second, by its design, we would expect the ICD-10 coding algorithm would preferentially identify a spectrum of ADPKD patients with moderate to advanced disease requiring hospital encounters, rather than ADPKD patients managed in the community who did not have hospital encounters. The code sets may also identify some mild cases, such as patients with ADPKD admitted for uncomplicated pregnancy. Therefore, these algorithms should only be used to assemble and study cohorts of adult patients with ADPKD and hospital encounters, rather than all patients in the province with ADPKD. Unfortunately, there are no relevant codes that can be used to identify the presence of ADPKD in the Ontario outpatient billing system.

Third, we reviewed medical charts from 2 hospitals at the London Health Sciences Centre. While coding practices are standardized across hospitals, any differences in coding between these 2 hospitals and other hospitals would influence the generalizability of our study results.

Fourth, there were no reports from genetic testing in any of the patient charts, which could have helped further ascertain the presence of ADPKD in cases when a family history is absent or not available.

Fifth, we are not sure that all imaging or other ancillary information for a given patient was found. For example, a patient may have had an ultrasound performed in an outpatient lab, and the nephrologist may not have a record of it. Therefore, the positive predictive value may be underestimated. In addition, this also may explain why the percentage of our cohort is lower than the estimates reported in the published literature.

Finally, our adjudicators were aware that all reviewed records had ICD-10 codes assigned for PKD in the health care database records. Although this may have influenced their adjudication of the records, we minimized the risk of information bias through the use of predefined diagnostic criteria for ADPKD, where 2 reviewers independently adjudicated each case.

Conclusions

In conclusion, the positive predictive value of the various coding algorithms for ADPKD is moderately high. These codes can be used to assemble and study cohorts of adult patients with ADPKD and hospital encounters but are expected to miss many patients with milder forms of ADPKD who are healthy without hospital encounters.

List of Abbreviations

ADPKD, autosomal dominant polycystic kidney disease; CI, confidence interval; CIHI-DAD, Canadian Institute for Health Information Discharge Abstract Database; CIHI-NACRS, Canadian Institute of Information National Ambulatory Care Reporting System; ICD-9, International Classification of Diseases, Ninth revision; ICD-10, International Classification of Diseases, Tenth revision; ID, identification number; IQR, interquartile range.

Ethics Approval and Consent to Participate

The institutional review board at Western University, London, Ontario, Canada, approved the chart abstraction study, and the one at Sunnybrook Health Sciences Centre, Toronto, Ontario, Canada, approved the second study using the health care administrative data housed at the Institute for Clinical Evaluative Sciences (ICES). The institutional review boards waived the need for patient consent. The ICES is a designated prescribed entity under Section 45 of the Personal Health Information Protection Act (PHIPA), and as such, the need for patient consent is waived (as confirmed by the institutional review board that approved this study).
Consent for Publication
Not applicable.

Availability of Data and Materials
The data supporting the findings of this article are not available in the public domain.

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Author Contributions
V.K., Y.P., and A.X.G. conceived and actively participated in the design and coordination of the study. V.K. was the main reviewer, conducted the main analysis, and wrote the first draft of the manuscript. K.C. assisted with the data collection, and R.K.M. was the second reviewer. M.R. reviewed images of patients that required additional information. S.D. conducted analysis and provided analytical support for the Institute for Clinical Evaluative Sciences (ICES) portion of the data. All authors read and approved the final article.

Declaration of Conflicting Interests
The author(s) declared the following potential conflicts of interest with respect to the research, authorship, and/or publication of this article: Dr York Pei served as an expert consultant on drug development (Otsuka, Pfizer, and Genzyme/Sanofi) related to autosomal dominant polycystic kidney disease (ADPKD). The other authors declare no competing interests.

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