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The Importance of Nomenclature for Congenital Heart Disease: Implications for Research and Evaluation

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Abstract

Background—Administrative databases are often used for congenital heart disease research and evaluation, with little validation of the accuracy of the diagnostic codes.

Methods—Metropolitan Atlanta Congenital Defects Program surveillance records were reviewed and classified using a version of the International Pediatric and Congenital Cardiac Code. Using this clinical nomenclature as the referent, we report the sensitivity and false positive fraction (1 – positive predictive value) of the International Classification of Diseases, Ninth Revision, Clinical Modification diagnosis codes for tetralogy of Fallot, transposition of the great arteries, and hypoplastic left heart syndrome.

Results—We identified 4918 infants and foetuses with congenital heart disease from the surveillance records. Using only the International Classification of Diseases diagnosis codes, there were 280 records with tetralogy, 317 records with transposition, and 192 records with hypoplastic left heart syndrome. Based on the International Pediatric and Congenital Cardiac Code, 330
records were classified as tetralogy, 163 records as transposition, and 179 records as hypoplastic left heart syndrome. The sensitivity of International Classification of Diseases diagnosis codes was 83% for tetralogy, 100% for transposition, and 95% for hypoplastic left heart syndrome. The false positive fraction was 2% for tetralogy, 49% for transposition, and 11% for hypoplastic left heart syndrome.

Conclusions—Analyses based on International Classification of Diseases diagnosis codes may have substantial misclassification of congenital heart disease. Isolating the major defect is difficult, and certain codes do not differentiate between variants that are clinically and developmentally different.

Keywords
congenital heart disease; nomenclature; surveillance

Introduction
Administrative databases are often the basis for congenital heart disease research and evaluation\textsuperscript{1–9}. In the United States of America, these databases use the International Classification of Diseases, Ninth Revision, Clinical Modification\textsuperscript{10} to describe cardiac lesions. Evidence from two recent investigations suggests that the accuracy of the International Classification of Diseases diagnosis codes for congenital heart defects is likely to be poor\textsuperscript{11,12}. Cronk and colleagues reported that only 52% of the congenital heart defect diagnoses contained in medical records had corresponding diagnosis codes in the hospital discharge database\textsuperscript{11}. Frohnert and colleagues reviewed a series of medical records and were able to confirm only 41% of the diagnosis codes for congenital heart defects that were present in the administrative database\textsuperscript{12}. The investigators offer several possible reasons for the poor diagnostic accuracy of the administrative codes, including accidental miscoding, contradictory or poorly described information in the medical record, and inadequately trained medical coders\textsuperscript{11,12}.

These two studies suggest that administrative databases fail to identify a substantial fraction of true cases of heart defects, identify many false positives, and that the heart defects studied using such databases may be unrepresentative of heart defects in the general population. Furthermore, some members of the paediatric cardiology and cardiac surgery community have argued that the International Classification of Diseases nomenclature used in administrative databases lacks sufficient detail to adequately describe the spectrum of congenital heart defects and have voiced the need for an improved nomenclature\textsuperscript{13,14}.

During the 1990s, both The Society of Thoracic Surgeons and The European Association for Cardio-Thoracic Surgery created databases to assess the outcomes of congenital cardiac surgery\textsuperscript{13}. In 1998 these organizations collaborated to create the International Congenital Heart Surgery Nomenclature and Database Project, and in 2000 a common nomenclature and core minimal dataset was adopted by both The Society of Thoracic Surgeons and The European Association for Cardio-Thoracic Surgery\textsuperscript{13}. By 2005, the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease had crossmapped the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project with the European Paediatric Cardiac Code of the Association for European Paediatric Cardiology, thereby creating the International Pediatric and Congenital Cardiac Code, which is freely available [\text{http://www.IPCCC.NET}]\textsuperscript{15}.

Two commonly used versions of the International Pediatric and Congenital Cardiac Code exist\textsuperscript{16–19}.  

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The version derived from the European Paediatric Cardiac Code of The Association for European Paediatric Cardiology.

The version derived from the International Congenital Heart Surgery Nomenclature and Database Project of The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons.

Recently, the Metropolitan Atlanta Congenital Defects Program used the version of the International Pediatric and Congenital Cardiac Code derived from the International Congenital Heart Surgery Nomenclature and Database Project (hereafter referred to as the “clinical nomenclature”) to classify all of its surveillance records with congenital heart disease. This was the first application of this clinical nomenclature to routinely collected birth defects surveillance data. Our objective was to evaluate the diagnostic accuracy of the administrative nomenclature in the International Classification of Diseases relative to this clinical nomenclature for the cohort of infants and foetuses with congenital heart defects born to mothers residing in metropolitan Atlanta during 1988–2003.

Materials and Methods

The Metropolitan Atlanta Congenital Defects Program is an active, population-based birth defects surveillance system administered by the Centers for Disease Control and Prevention of the United States of America since 1967\(^2\). Cases in the Metropolitan Atlanta Congenital Defects Program include infants and foetuses of at least 20 weeks gestation whose mothers resided in one of five central metropolitan Atlanta counties at delivery. Major structural defects, chromosomal abnormalities, and clinical syndromes diagnosed within six years of delivery are included in Metropolitan Atlanta Congenital Defects Program. Trained abstractors access hospital medical records directly and record information on infant and foetal diagnoses and procedures. A nomenclature developed by the Centers for Disease Control and Prevention\(^2\) based on the International Classification of Diseases, Ninth Revision, Clinical Modification\(^1\) and the British Paediatric Association Classification of Diseases\(^2\), is used to code birth defects, and hereafter is referred to as the “administrative nomenclature.” The codes in the administrative nomenclature, while more detailed, can be mapped directly to the codes in the International Classification of Diseases via a computer algorithm if the extra detail is ignored. In the present study we ignored this extra detail and treated the codes in this administrative nomenclature as if they were codes in the International Classification of Diseases, Ninth Revision, Clinical Modification. Emory University and Centers for Disease Control and Prevention Institutional Review Boards granted waivers of informed consent for this study on July 24, 2004 and February 1, 2005, respectively.

We identified all surveillance records in the Metropolitan Atlanta Congenital Defects Program with congenital heart defects and a delivery date during 1988–2003, inclusive. Each record was manually reviewed by paediatric cardiologists: Mark D Reller, William T Mahle, Lorenzo D Botto, and Tiffany J Riehle-Colarusso. All records were coded using the clinical nomenclature as published by The Society of Thoracic Surgeons Congenital Heart Surgery Database version 2.3\(^2\). This activity was an enrichment of pre-existing surveillance data, based on analysis of the abstracted text and expert opinion, using a standard clinical nomenclature that enables reviewers to accurately describe congenital cardiac lesions. Reviewers determined the anatomical diagnosis based on data from surveillance records in the Metropolitan Atlanta Congenital Defects Program, which included details from echocardiographic reports and the catheterization report, if performed. Comments from the operative note regarding anatomical features were also included in surveillance records. After the review, records with similar clinical nomenclature classifications were grouped to facilitate analysis. For records with several defects,
prioritization was based on presumed developmental mechanisms \cite{24,25}. For example, all records with isomerism of the atrial appendages were grouped into heterotaxy, regardless of other associated defects. Similarly, a constellation of defects might be placed into the "single ventricle/complex group." Although records could be placed into one or more of 35 different aggregation groups, we focus on just three of these groups: tetralogy of Fallot, transposition of the great arteries with concordant atroventricular connections and discordant ventriculo-arterial connections, and hypoplastic left heart syndrome. We focus on these severe, commonly occurring lesions because they are frequently used as benchmarks for surgeon and programmatic performance \cite{26–32}. The administrative nomenclature and clinical nomenclature diagnosis codes that comprise these three groups are presented in Table 1.

In this analysis, the clinical nomenclature-based groups were treated as the referent. The sensitivity and the false positive fraction of the administrative nomenclature codes are reported for each defect group listed in Table 1. Sensitivity is the probability that a case has an appropriate code from the administrative nomenclature, given its membership in a particular group from the clinical nomenclature. The false positive fraction is the probability that a case is not in the group from the clinical nomenclature, given that it has the code from the administrative nomenclature for that diagnosis. The false positive fraction is equivalent to 1 – positive predictive value. If sensitivity = 1.00, this indicates that all records in the group from the clinical nomenclature have an appropriate code from the administrative nomenclature, whereas if sensitivity = 0.00 then no records in the group from the clinical nomenclature have an appropriate code from the administrative nomenclature. A sensitivity = 1.00 does not indicate perfect agreement; excess records not contained in the group from the clinical nomenclature may be present in the group from the administrative nomenclature. The false positive fraction is this proportion of excess, or “false positive,” records.

**Results**

During 1988–2003, there were 691,099 infants born to mothers residing in Atlanta; 4,918 infants and foetuses ascertained by the Metropolitan Atlanta Congenital Defects Program during this period had structural heart defects (0.7%). Using only the codes from the administrative nomenclature, there were 280 records with tetralogy, 317 records with transposition, and 192 records with hypoplastic left heart syndrome. Based on the review using the clinical nomenclature, 330 records were classified as tetralogy, 163 records as transposition, and 179 records as hypoplastic left heart syndrome. The sensitivity and false positive fraction for these three defect groups are presented in Table 2.

**Tetralogy of Fallot**

Of the 330 records classified as tetralogy of Fallot by the review using the clinical nomenclature, 55 did not have a code for tetralogy from the administrative nomenclature (Table 2, sensitivity = 0.83). Many of these hearts had pulmonary atresia and ventricular septal defect (n=36), which is often the extreme end of the anatomical spectrum of tetralogy of Fallot. However, because of limitations in the administrative nomenclature, pulmonary valve atresia cannot be distinguished from congenital absence of the pulmonary valve. Even more problematic is the fact that pulmonary artery atresia, stenosis, agenesis, and hypoplasia are all lumped under one code in the administrative nomenclature. Thus, one cannot reliably identify records with both pulmonary atresia and ventricular septal defect using codes from the administrative nomenclature.

The coding of records with double outlet right ventricle with the administrative nomenclature also decreased the sensitivity for tetralogy. Clinically, double outlet right ventricle has several phenotypes:
• Double outlet right ventricle of the transposition type
• Double outlet right ventricle of the tetralogy type
• Double outlet right ventricle of the ventricular septal defect type
• Double outlet right ventricle with uncommitted ventricular septal defect type
• Double outlet right ventricle with intact ventricular septum

The administrative nomenclature forces all patients with any double outlet right ventricle phenotype into a single code that is a subtype of transposition. The clinical nomenclature, conversely, distinguishes among these phenotypes. The patients with double outlet right ventricle of the tetralogy type can be grouped with tetralogy and the patients with double outlet right ventricle of the transposition type can be grouped with transposition (Table 1). Fourteen of the 55 records that did not have a code from the administrative nomenclature for tetralogy were classified by the review using the clinical nomenclature as double outlet right ventricle of the tetralogy type.

There were five additional records in which the code from the administrative nomenclature did not agree with the classification of tetralogy by the clinical nomenclature. Three records had codes from the administrative nomenclature for both atrioventricular canal defect and pulmonary artery anomaly. One record had a code from the administrative nomenclature for “pulmonary valve anomaly, other,” and the fifth record used a code from the administrative nomenclature for “unspecified anomaly of the heart.”

The false positive fraction for tetralogy was very low (false positive fraction = 0.02); only five of 280 records were false positives. These false positives were classified by the review using the clinical nomenclature as heterotaxy (n = 1), double outlet right ventricle of the transposition type (n = 1), perimembranous ventricular septal defect (n = 1), and perimembranous ventricular septal defect with secundum atrial septal defect (n = 1). The fifth record had insufficient information to confirm a diagnosis of tetralogy.

Transposition of the great arteries

The sensitivity was 1.00 for transposition of the great arteries (Table 2); all records classified as transposition by the review using the clinical nomenclature had an appropriate code from the administrative nomenclature. However, 154 of the 317 records with a code for transposition from the administrative nomenclature were false positives (false positive fraction = 0.49). These records were placed into various groups following the review using the clinical nomenclature, the most frequent being “single ventricle/complex” (n = 38), heterotaxy (n = 38), double outlet right ventricle (n = 32), and tetralogy (n = 27). Other groups include hypoplastic left heart syndrome (n = 6), congenitally corrected transposition of the great arteries (n = 5), ventricular septal defect (n = 3), and atrioventricular septal defect (n = 3). Two surveillance records had insufficient detail to support a diagnosis of transposition.

The majority of false positives in the analysis of transposition were caused by one of two issues. First, complex cardiac lesions frequently include transposed great arteries as part of the anatomical description. The hierarchy used in the clinical aggregation process tended to place these records into the “single ventricle/complex group” or the heterotaxy group rather than the transposition group. Accordingly, nearly half of the false positive records for transposition were classified by the review using the clinical nomenclature as either “single ventricle/complex” (n = 38) or heterotaxy (n = 38).
Second, the single code in the administrative nomenclature used to describe all patients with any of the double outlet right ventricle phenotypes resulted in many false positives. The administrative nomenclature considers all double outlet right ventricle phenotypes to be a subtype of transposition. Our aggregation process grouped double outlet right ventricle of the transposition type with transposition and double outlet right ventricle of the tetralogy type with tetralogy. All other phenotypes were classified as double outlet right ventricle. Records with the code from the administrative nomenclature for double outlet right ventricle classified by the review using the clinical nomenclature as either tetralogy (n = 27) or double outlet right ventricle (n = 32) were therefore counted as false positives.

We conducted a secondary analysis to evaluate whether concordance could be improved by excluding records that had codes from the administrative nomenclature for both transposition and one or more of the following: malposition of the heart and cardiac apex, common ventricle, “situs inversus”, or “spleen anomaly”. Doing so reduced the number of false positives from 154 to 99. The false positive fraction decreased from 0.49 to 0.38, while the sensitivity remained at 1.00.

**Hypoplastic left heart syndrome**

Most records classified as hypoplastic left heart syndrome after the review using the clinical nomenclature had the corresponding code from the administrative nomenclature for hypoplastic left heart syndrome (170 of 179 records, sensitivity = 0.95). Six discrepant records had a code from the administrative nomenclature for hypoplastic left ventricle, two had a code from the administrative nomenclature for “single ventricle”, and one had codes from the administrative nomenclature for mitral valve stenosis and aortic valve stenosis. This finding is not attributable to a limitation in the administrative nomenclature; rather, it is the result of the medical coder or abstractor coding one or more of the component defects of hypoplastic left heart syndrome but failing to recognize the overall syndrome.

Eleven percent of the records with hypoplastic left heart syndrome were false positives (22 of 192 records). The 22 false positives had the code from the administrative nomenclature for hypoplastic left heart syndrome but were classified by the review as “single ventricle/complex” (n = 10), interrupted aortic arch (n = 4), double outlet right ventricle (n = 2), heterotaxy (n = 2), and coarctation of the aorta (n = 2). Two surveillance records had insufficient detail to confirm a diagnosis of hypoplastic left heart syndrome.

Some records were false positives because the code from the administrative nomenclature for hypoplastic left heart syndrome was used to describe hearts with only coarctation of the aorta or interrupted aortic arch. These hearts were miscoded by the medical coder or the abstractor. False positives also occur when records with the component defects for hypoplastic left heart syndrome have additional defects that merit classification as “single ventricle/complex” or heterotaxy. For example, we elected to classify records with “Single ventricle, Unbalanced atroventricular canal (left),” with “single ventricle/complex” defects rather than with hypoplastic left heart syndrome.

**Discussion**

The frequency of misclassification in reporting of tetralogy, transposition, and hypoplastic left heart syndrome suggests caution is needed when administrative diagnosis codes are used to classify congenital heart lesions. Misclassification can occur because of errors on the part of the coder, because of limitations inherent to the administrative nomenclature, or because of failure to distinguish less complicated forms of a lesion from those with heterotaxy or other complex arrangements.
In the tetralogy analysis, coding with the administrative nomenclature missed 17% of tetralogy records. Using codes from the administrative nomenclature, records with pulmonary atresia and ventricular septal defect could not be distinguished from records with pulmonary artery stenosis or hypoplasia. These pulmonary atresia and ventricular septal defect cases represent the extreme end of the anatomic spectrum of tetralogy and bear little or no relationship to simple branch pulmonary artery or valvar stenosis. Estimates of post-surgery mortality for tetralogy based on administrative databases may be overly optimistic if the most severe form of tetralogy, pulmonary atresia with ventricular septal defect, is not included in the evaluation. Additionally, the administrative nomenclature collapses all double outlet right ventricle phenotypes into a single code that is a subtype of transposition. In reality, only a fraction of DORV cases have features fundamentally related to transposition from an anatomic, physiologic, or prognostic standpoint.

Outcomes after surgery for transposition are often used as a benchmark for surgeon and programmatic performance. The transposition analysis revealed that 49% of records classified as transposition by coding with the administrative nomenclature did not actually have transposition as the fundamental problem. Although excluding transposition records with codes from the administrative nomenclature for malposition of the heart and cardiac apex, common ventricle, “situs inversus,” or spleen anomaly reduced the false positive fraction from 0.49 to 0.38, this level of misclassification remains high. Patients with heterotaxy syndrome and/or functionally univentricular hearts tend to have poor survival. If these records are included in the transposition subgroup, then this will lead to a severely flawed and misleading analysis, effectively penalizing surgeons who routinely treat patients with very complex congenital heart disease.

In the hypoplastic left heart syndrome analysis, one in every nine records coded with the administrative nomenclature as hypoplastic left heart syndrome was a false positive. Coding errors were the cause of many false positives, highlighting the value of systematic record review. Hypoplastic left heart syndrome is another benchmark lesion where surgeon and programmatic performance is often measured. Use of administrative databases that might include several false positive hypoplastic left heart syndrome records could result in inaccurate estimates of surgical outcomes.

The generalization of our findings to studies based on hospital billing databases, and other administrative databases based on the International Classification of Diseases, may be limited because of differences between these databases and the Metropolitan Atlanta Congenital Defects Program. Abstractors from the Metropolitan Atlanta Congenital Defects Program receive specialized training in birth defects coding and travel to hospitals and access medical records directly. Nine of the 11 abstractors have nursing degrees and can draw from their clinical background when reviewing medical records. Defects are coded using an enhanced International Classification of Diseases–based nomenclature, and additional information, such as echocardiography report details, is often recorded on the surveillance records. After abstraction, surveillance records are further reviewed by in-house staff to reduce the frequency of coding errors. The common hospital administrative database does not have this level of quality control, and unlike Metropolitan Atlanta Congenital Defects Program abstractors, many hospital-based medical coders do not have a clinical background nor do they receive specialized training in birth defects coding. Consequently, the validity of diagnosis codes from the International Classification of Diseases for congenital heart lesions in administrative databases may be significantly worse than reported in this manuscript, and our results may represent a “best case scenario” with respect to the quality of the administrative diagnosis codes. The extremely poor agreement between the administrative codes and the medical records documented in previous studies further supports this notion.
Unlike hospital billing databases, however, the Metropolitan Atlanta Congenital Defects Program does not abstract International Classification of Diseases procedural codes or other similar codes such as the American Medical Association Current Procedure Terminology codes. These codes describe medical, surgical, and diagnostic services. Many hospital billing departments record both diagnosis and procedural codes. Although many large, widely used administrative datasets, such as those of the Agency for Healthcare Research and Quality of the U.S. Department of Health and Human Services, do not have access to Current Procedure Terminology codes, analyses of these datasets typically incorporate both International Classification of Diseases diagnosis and procedural codes.

Unlike the United States, much of the world has already transitioned to the International Classification of Diseases, Tenth Revision, Clinical Modification. Many problems in the Ninth Revision of the nomenclature persist in the Tenth Revision. The nomenclature of the Tenth Revision continues to collapse all double outlet right ventricle phenotypes into a single code and cannot distinguish less complicated forms of lesions from those with heterotaxy or other complex arrangements. Some problems with the Ninth Revision have been addressed in the Tenth Revision; for example, pulmonary artery stenosis and pulmonary artery atresia have unique codes in the Tenth Revision. Although we cannot quantify the impact that the transition from the Ninth Revision to the Tenth Revision has had on the accuracy of coding patients with congenital heart disease, we would speculate that misclassification continues to be a concern.

Our use of the International Pediatric and Congenital Cardiac Code classifications as a referent requires qualification. A true “gold standard” would require complete echocardiography reports for each infant and foetus included in the analysis, whereas this project relies on information contained in the surveillance records of the Metropolitan Atlanta Congenital Defects Program. As such, some diagnoses could not be confirmed because important details were missing from the surveillance records. We are unable to evaluate the extent to which our findings would differ if complete echocardiography reports were available for every surveillance record. Nevertheless, even without a true gold standard, our results reveal limitations in several codes within the nomenclature system of the International Classification of Diseases.

Our demonstration of the weaknesses of the administrative nomenclature in comparison to the clinical nomenclature, based on surveillance data from Atlanta, Georgia, United States of America, is relevant to the relationship between administrative and clinical databases worldwide. The international scope of this challenge is the driving force behind the ongoing international collaborative efforts to create and maintain the International Pediatric and Congenital Cardiac Code. Reconciling differences between administrative and clinical databases is truly a challenge with global impact in our field, and this challenge will only be met with continued global collaboration.

Ultimately, the optimal classification and coding system will be based on clear, precise definitions of the cardiac phenotype. Our current study documents how an improved classification scheme based on the clinical coding of the International Pediatric and Congenital Cardiac Code is more precise and accurate relative to coding based on administrative nomenclature. Applying standardized definitions of cardiac phenotypes should lead to further improvement. To this end, the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease has provided unified nomenclature and definitions for several complex congenital cardiac malformations, including the functionally univentricular heart, hypoplastic left heart syndrome, congenitally corrected transposition, and heterotaxy. Recently, the...
International Society for Nomenclature of Paediatric and Congenital Heart Disease created two new committees to further the definitions of cardiac phenotypes:

- The International Working Group for Defining the Nomenclatures for Paediatric and Congenital Heart Disease, which will write definitions for the terms used in the International Pediatric and Congenital Cardiac Code.
- The International Working Group for Archiving and Cataloguing the Images and Videos of the Nomenclatures for Paediatric and Congenital Heart Disease, which will link images and videos to the International Pediatric and Congenital Cardiac Code, and create an archive of these images which will be linked to The Cardiothoracic Surgery Network.

Conclusions

The Metropolitan Atlanta Congenital Defects Program is an active birth defects surveillance system committed to excellence in diagnostic accuracy. Numerous quality control procedures have been implemented to enhance data quality. Many common sources of error in administrative databases, including accidental miscodes, poorly trained medical coders, and other similar errors, have been greatly reduced in the Metropolitan Atlanta Congenital Defects Program. Even so, the diagnostic accuracy of certain codes for cardiac defects in the International Classification of Diseases was found to be poor.

Analyses of outcomes from paediatric cardiac surgery based on diagnosis codes from the International Classification of Diseases in administrative databases are likely to be limited by substantial misclassification of cases of congenital heart defects. Although evaluation of surgical outcomes for children with congenital heart disease is critically important for healthcare quality assessment, evaluations that base lesion classification on codes from the International Classification of Diseases risk generating inaccurate results that are potentially misleading. We encourage the use of a more accurate and current nomenclature, such as the International Pediatric and Congenital Cardiac Code, for classification of congenital heart disease prior to evaluation of surgical outcomes.

Acknowledgments

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References


Table 1

Composition of the aggregate cardiac defect groups according to the clinical nomenclature and administrative nomenclature.

<table>
<thead>
<tr>
<th>Aggregate cardiac defect group</th>
<th>Clinical nomenclature‡</th>
<th>Administrative nomenclature‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>Tetralogy of Fallot</td>
<td>745.2 – Tetralogy of Fallot</td>
</tr>
<tr>
<td></td>
<td>Tetralogy of Fallot with absent pulmonary valve</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tetralogy of Fallot with atrioventricular septal defect</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Double outlet right ventricle, tetralogy of Fallot type</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pulmonary atresia with ventricular septal defect§</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral artery(ies)</td>
<td></td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>Transposition with intact ventricular septum</td>
<td>745.10 – Transposition of the great arteries</td>
</tr>
<tr>
<td></td>
<td>Transposition with intact ventricular septum and left ventricular outflow tract obstruction</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Transposition with ventricular septal defect</td>
<td>745.11 – Double outlet right ventricle</td>
</tr>
<tr>
<td></td>
<td>Transposition with ventricular septal defect and left ventricular outflow tract obstruction</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Transposition, not otherwise specified</td>
<td>745.19 – Other transposition</td>
</tr>
<tr>
<td></td>
<td>Double outlet right ventricle, transposition-type</td>
<td></td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>Hypoplastic left heart syndrome</td>
<td>746.7 – Hypoplastic left heart syndrome</td>
</tr>
<tr>
<td></td>
<td>Hypoplastic left heart syndrome with ventricular septal defect</td>
<td></td>
</tr>
</tbody>
</table>

‡“The Clinical nomenclature” is derived from the International Congenital Heart Surgery Nomenclature and Database Project of The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons.

§The “Administrative nomenclature” is derived from the International Classification of Diseases, Ninth Revision, Clinical Modification diagnosis codes.

§For the clinical nomenclature, hearts with discordant ventriculo-arterial connections, pulmonary atresia, and ventricular septal defect are grouped with Transposition of the great arteries, not with Tetralogy of Fallot.
Table 2

Sensitivity and false positive fraction of the administrative nomenclature codes for tetralogy of Fallot, transposition of the great arteries, and hypoplastic left heart syndrome, using the clinical nomenclature codes as the referent.

<table>
<thead>
<tr>
<th>Aggregate cardiac defect group</th>
<th>Sensitivity‡</th>
<th>False Positive Fraction‡</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Tetralogy of Fallot</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinical code +</td>
<td>275/330 (0.83)</td>
<td>5/280 (0.02)</td>
</tr>
<tr>
<td>Administrative code +</td>
<td>275</td>
<td>5</td>
</tr>
<tr>
<td>Administrative code −</td>
<td>55</td>
<td>---</td>
</tr>
<tr>
<td><strong>Transposition of the great arteries</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinical code +</td>
<td>163/163 (1.00)</td>
<td>154/317 (0.49)</td>
</tr>
<tr>
<td>Administrative code +</td>
<td>163</td>
<td>154</td>
</tr>
<tr>
<td>Administrative code −</td>
<td>0</td>
<td>---</td>
</tr>
<tr>
<td><strong>Hypoplastic left heart syndrome</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinical code +</td>
<td>170/179 (0.95)</td>
<td>22/192 (0.11)</td>
</tr>
<tr>
<td>Administrative code +</td>
<td>170</td>
<td>22</td>
</tr>
<tr>
<td>Administrative code −</td>
<td>9</td>
<td>---</td>
</tr>
</tbody>
</table>

‡Sensitivity is the probability that a case has an appropriate administrative nomenclature code given the presence of the clinical nomenclature code for that diagnosis.

‡False Positive Fraction is the probability that a case does not have the clinical nomenclature code given the presence of the administrative nomenclature code ICD for that diagnosis.