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The 745.5 issue in code-based, adult congenital heart disease population studies: Relevance to current and future ICD-9-CM and ICD-10-CM studies

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Abstract

Objective: Although the ICD-9-CM code 745.5 is widely used to indicate the presence of a secundum atrial septal defect (ASD), it is also used for patent foramen ovale (PFO) which is a normal variant and for "rule-out" congenital heart disease (CHD). The ICD-10-CM code Q21.1 perpetuates this issue. The objective of this study was to assess whether code 745.5 in isolation or in combination with unspecified CHD codes 746.9 or 746.89 miscodes for CHD, and if true CHD positives decrease with age.

Design: Echocardiograms of patients with an ICD-9-CM code of 745.5 in isolation or in combination with unspecified CHD codes 746.9 or 746.89 were reviewed to validate the true incidence of an ASD. This observational, cross-sectional record review included patients between 11 and 64 years of age.

Results: Medical charts and echocardiograms of 190 patients (47.9% males) were reviewed. The number of falsely coded patients with 745.5 (no ASD) was high (76.3%). Forty-five (23.7%) patients had a true ASD. Among the 145 patients without an ASD, 100 (52.6%) were classified as having a PFO, 37 (19.5%) had a normal non-CHD echocardiogram, and 8 (4.2%) had some other CHD anomaly. The likelihood that 745.5 coded for a true ASD was higher in children aged 11-20 (64.3%) than adults aged 21-64 years (20.6%).

Conclusions: This validation study demonstrates that 745.5 performed poorly across all ages. As 745.5 is widely used in population-level investigations and ICD-10-CM perpetuates the problem, future analyses utilizing CHD codes should consider separate analysis of those identified only through code 745.5.

KEYWORDS
adult congenital heart disease, atrial septal defect, ICD-9-CM congenital heart defect coding

1 | INTRODUCTION

With increasing use of administrative data sets to conduct population health research, the validity of International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) and ICD-10-CM codes to identify true cases is of increasing importance. Some ICD-9-CM and ICD-10-CM codes lack specificity and may correspond to 1 or more diagnoses. In addition, a common practice of using "rule-out" codes when ordering a test may further increase the number of false cases associated with a particular ICD-9-CM or ICD-10-CM code. Databases that identify patients with congenital heart disease (CHD) frequently use the ICD-9-CM code 745.5 or ICD-10-CM code Q21.1 to categorize patients with secundum atrial septal defects (ASDs).[1] The publications based on the data from these databases often include

the spectrum of ICD-9-CM codes 745.xx-747.xx as "cases," without validation of individual codes. True ASDs are present in about 0.05% of the population.[2] However, code 745.5 also includes those patients with patent foramen ovale (PFO) which is a normal variant present in >25% of the population. In addition, 745.5 also serves as a "rule out" for PFO or ASD, often in the setting of a transient ischemic attack (TIA) or nonspecific cardiac symptoms which may represent a normal echocardiogram and no diagnosis of CHD, and other CHD for which a more specific code might exist.

Inclusion of 745.5 in the studies of CHD based on the large administrative data sets may overestimate true cases of CHD, and may misrepresent more complex CHDs as simple. Research studies which interrogate large administrative databases for patients with CHD often search for ASDs using the ICD-9-CM code 745.5. More likely than not, these studies end up falsely classifying the patients as CHD cases when they either actually have a PFO, which is technically not a CHD, or the test was performed to "rule-out" CHD-ASD. The previous quality improvement (QI) studies have shown a very low sensitivity (23%)[3] and accuracy (50%)[4] of ICD-9-CM codes, especially the inability to differentiate between ASD and PFO, along with the considerable variability in procedure reporting formats and medication schemes, often with center-specific modifications.[5,6] Also, these studies were performed in tertiary centers with large populations of the patients with CHD, which makes for a "best case" scenario to identify true patients with CHD.

The objectives of this study are to assess the validity of the ICD-9-CM code 745.5 for true secundum ASDs as well as the risk factors for incorrect use of ICD-9-CM code 745.5, including nonspecific cardiac codes. It is hypothesized that the ICD-9-CM code for ASD (745.5, in isolation or with 746.9 or 746.89) frequently does not code for significant CHD, and that true positives decrease with the age of the population being examined.

2 | MATERIALS AND METHODS

This observational, cross-sectional study was conducted at the Emory Healthcare (EH) system (Georgia). It included patients aged 11–64 years of either gender, seen between January 1, 2008 and January 31, 2010 at 1 of the EH sites, with an ICD-9-CM diagnosis of 745.5 with or without nonspecific codes 746.9 (unspecified anomaly of heart) and 746.89 (other congenital anomalies of heart), and an echocardiogram available for review. After deduplication and linkage of data sets, patients who lacked either an echocardiogram or ICD-9-CM code 745.5 as well as those with additional ICD-9-CM codes for more specific CHD (in addition to 745.5, 746.89, or 746.9) were excluded.

2.1 | Definitions

Normal echocardiograms are defined as those which have no structural abnormalities. Trivial to mild pulmonary and tricuspid insufficiency and trivial mitral and aortic valve insufficiency are considered normal variants. A PFO is defined as a persistent connection from fetal life that allows for oxygenated blood from the umbilical vein to pass from the right atrium to the left atrium and out to the systemic circulation. After birth, this connection is covered by the flap of the fossa ovalis, but may persist as a small communication between the two atria in about 26%–27% of adults and as such, is considered a normal structure.[7,8] To diagnose a PFO by echocardiogram, the flap of the fossa ovalis should be visualized adjacent to the defect. A secundum ASD is defined as a connection between the left and the right atria in the area of the fossa ovalis that results from deficiency, perforation, or the absence of the septum primum during embryologic life.[9] Both secundum ASDs and PFOs are coded by 745.5 in the ICD-9-CM coding system and by Q21.1 in the ICD-10-CM coding system. Secundum ASDs have distinctive echocardiographic features, such as a complete absence of the flap of the fossa ovalis. Other suggestive features of a secundum ASD rather than a PFO are the presence of right atrial or right ventricular enlargement, and a large defect in the atrial septum often >6 mm. An "agitated saline bubble study" or "microcavitation study" is sometimes performed to diagnose an ASD or PFO. When the agitated saline is injected into an IV, the right atrium and right ventricle are opacified by the microbubbles of air and the presence of bubbles in the left atrium and left ventricle indicate either an intracardiac or extracardiac shunt. If the bubbles are visualized within 3 beats in the left atrium, a PFO or ASD is detected. If bubbles are detected in the left atrium after more than 3 beats, then a pulmonary AVM may be more likely.

There are other types of ASDs, but they are coded by different ICD-9-CM codes. A primum ASD is coded by 745.61 in the ICD-9-CM coding system and by Q21.2 in the ICD-10-CM system. A sinus venosus ASD is coded by 745.8 in the ICD-9-CM coding system, but is also coded by Q21.1 in the ICD-10-CM system such as secundum ASDs and PFOs. The term "more specific CHD" by echocardiogram indicates that the study does not demonstrate a PFO or ASD, but does include another CHD. The term "rule-out CHD" is commonly used to obtain an echocardiogram in the setting of strokes, migraine headaches, endocarditis, arrhythmias requiring cardioversion, and murmurs to assess for any cardiac defects.

2.2 | Data collection

From an IRB-approved data repository (IRB00064051) created with funds from a Cooperative Agreement with The Centers for Disease Control and Prevention, 995 patients between 11 and 64 years of age were identified as having ICD-9-CM code 745.5 in isolation or in combination with nonspecific CHD codes 746.89 and/or 746.9. Data from the EH system which include Emory University hospitals and clinics, Emory Saint Joseph's Cardiology Clinics, Children's Healthcare of Atlanta, and Sibley Heart Center Cardiology were utilized; CHD data obtained from other data sources were not included owing to the lack of access to echocardiographic images. The 995 cohorts of patients were then assigned to 1 of the 3 age groups (11–20, 21–40, or 41–64 years), and a 20% sample was randomly selected within each age group based on the number of individuals within that group to obtain a total of 200 patients. Ten patients without echocardiogram images for review were excluded. Characteristics of the remaining study sample

including demographics, diagnoses, surgical histories, medications, laboratory data, hospitalizations, pregnancy history when appropriate, electrocardiograms, magnetic resonance imaging studies, echocardiograms, and reason for echocardiogram were obtained through a retrospective review of the electronic medical record. Echocardiograms were reviewed by a board-certified adult congenital and pediatric cardiologist to assess whether the patient had a normal echocardiogram or a true CHD. A second reviewer who was board certified in pediatric cardiology and blind to the interpretations of initial echocardiogram review, reevaluated 29 (15%) of the echocardiograms to ensure accuracy and measure interobserver's reliability.

2.3 | Ethics

The study was approved by the Emory University Institutional Review Board (IRB00083563). The requirement for obtaining informed consent was waived as this was a retrospective review of medical records and echocardiographic images.

2.4 | Statistical analysis

A 20% random sample provided a sample size of 200 which was determined to be large enough to find a small difference (effect size) between true ASDs and those misclassified as having an ASD when a real difference exists for $\alpha = .05$ and power = .80. Data were analyzed using SAS software, version 9.4 TS Level 1M4 for Windows 1.0, 32-bit platform. Frequencies were calculated and chi-square analysis for all categorical variables was conducted. Sensitivity and specificity by age was also assessed.

3 | RESULTS

The study included 190 patients (47.9% males) between 11 and 64 years of age with 92.7% of the sample between 21 and 64 years; the majority of the sample (63.7%) fell into the oldest age group (age, 41–64 years). The majority of patients were white (70.9%), nonsmokers (69.4%) and who had private or commercial health insurance coverage (62.9%). Approximately, 7% were uninsured or self-payers. Demographic characteristics of the sample are summarized in Table 1.

The top 6 reasons for the first echocardiogram are listed in Table 2. The most common reasons for a first echocardiogram were neurologic symptoms of a stroke or TIA followed by chest pain. The vast majority of these patients, 95.7% and 84.0%, respectively, did not have an ASD. The only reason for a first echocardiogram for a majority of those patients with a true ASD was among those patients with known CHD (81.8%).

Table 3 summarizes the validation of true ASDs. Forty-five (23.7%) patients had a true ASD, whereas 145 patients (76.3%) were falsely coded as having a CHD. Among these 145 patients, 100 (52.6%) were classified as having a PFO, 37 (19.5%) had a normal non-CHD echocardiogram, and 8 (4.2%) had some other sort of CHD anomaly. The likelihood that 745.5 coded for a true ASD was higher in children (64.3%) than adults (20.6%). There was no statistical difference based on race, smoking, or insurance type.

TABLE 1 Characteristics of the study sample (n = 190)

Variable	n (%)
Age group (years)	
<21	14 (7.3%)
21–40	55 (29.0%)
41–64	121 (63.7%)
Gender	
Male	91 (47.9%)
Female	99 (52.1%)
Race	
White	95 (70.9%)
Black	36 (26.9%)
Asian/Pacific Islander	3 (2.2%)
Missing	56
Smoking	
Yes	44 (30.1%)
No	102 (69.9%)
Missing	44
Insurance type	
Self/uninsured	11 (6.9%)
Private/commercial	100 (62.9%)
Government	37 (23.3%)
Other	11 (6.9%)
Missing	31
Data source	
Emory/St. Joseph's Hospital	158(83.2%)
CHOA/Sibley	32 (16.8%)

Abbreviations: CHOA, Children's Healthcare of Atlanta.

4 | DISCUSSION

The patients with CHD constitute a real, yet small subset of the general population,[2] of increasing prevalence.[10] As such, conducting research in this field tends to be difficult and fraught with various challenges, especially sample size. Future directions to overcome these obstacles include the advent of registries and the accumulation of data over time. Birth registries such as the Metropolitan Atlanta Congenital Defects Program (MADCP), a population-based system for all babies born with birth defects in the metropolitan five-county Atlanta area, have all diagnoses verified by chart abstraction and hence identifying true cases.[11] However, these birth registries may identify the defects that spontaneously close. The studies utilizing echocardiography in neonates have described up to 92% of ASDs spontaneously closing.[12] The studies utilizing large, available, administrative, and electronic record-based databases use ICD-9-CM codes to identify the cases and constitute an appealing recourse to investigators.[1,13,14] The danger in this strategy, however, emanates from the known lack of granularity in these data sets.[4,5] Although the ICD-9-CM code 745.5 is widely used in published literature as equivalent to the presence of a secundum ASD, this study demonstrated that this code correlated only with actual CHD pathology in 24% of occurrences overall. The only patients who were

TABLE 2 Top 6 reasons for first echocardiogram by validation status

	n (% of Total 190 patients)	ASD	Not ASD	
Stroke/TIA	47 (24.7%)	2 (4.3%)	45 (95.7%)	
Chest pain	25 (13.2%)	4 (16.0%)	21 (84.0%)	
Known CHD	22 (11.6%)	18 (81.8%)	4 (18.2%)	$X^2 = 81.2$, $P < .0001$[a]
Dyspnea	18 (9.5%)	9 (50.0%)	9 (50.0%)	
Noncardiac surgery/transplant	18 (9.5%)	1 (5.6%)	17 (94.44%)	
Migraine headaches	16 (8.4%)	0 (0.0%)	16 (100.0%)	

[a]X^2 and *P* value analysis are for the entire group.
Abbreviations: ASD, atrial septal defect; CHD, congenital heart disease; TIA, transient ischemic attack.

more likely to be correctly identified with 745.5 as having an ASD were those <21 years (Table 3). The code becomes less likely to correctly classify those with a true ASD as patients' age. We suspect this is owing to the use of 745.5 as a "rule-out" code in adults more frequently than children. The patients who present with a stroke are more likely to be adults than children, and when the 745.5 code is used to order an echocardiogram for a patient with a stroke it often does not correctly identify a patient with an ASD. In addition, adult patients may see a broader range of noncardiac adult subspecialists, increasing the chances 745.5 may be used as a "rule-out ASD or PFO" code for a variety of nonspecific symptoms ranging from heart palpitations to shortness of breath to neurologic or TIA symptoms. In brief, 745.5 was, in fact, more likely to code for "not CHD" than for a true CHD. Given the high prevalence of this code in the range of 745.xx-747.xx used to define CHD in database studies, inaccurate conclusions may be drawn about CHD populations when this code is included. Plainly, implications of such findings suggest that patients are included in the studies identifying them as having true CHD when they in fact do not. Conversely, other patients who actually have a true ASD may not be coded accurately, and therefore, may be missed.

This study has ramifications for hundreds of previously published data which likely influenced guidelines, policies, and recommendations for patient care, as well as future studies. Unfortunately, the ICD-10-CM classification system has the same issues. For instance, ICD-10-CM code Q21.1 codes for PFO, secundum ASD, sinus venosus ASD, and coronary sinus ASD, and has the same issues as ICD-9-CM 745.5. In a study on the outcomes of hospitalization in adults with ASD in the United States, ventricular septal defect, and ASDs,[1] defined by an ICD-9-CM code of 745.5, constituted 48% of the study population. As per the findings of this study as well as those of the previous

TABLE 3 Characteristics of sample by validation status

	Total (n = 190)	ASD (n = 45, 23.7%)	Not ASD (n = 145, 76.3%)	*P* value
Age group				
<21	14 (7.4%)	9 (64.3%)	5 (35.7%)	
21–40	55 (28.9%)	13 (23.7%)	42 (76.4%)	$X^2 = 14.2$, $P < .001$
41–64	121 (63.7%)	23 (19.0%)	98 (81.0%)	
Male	91 (47.9%)	17 (18.7%)	74 (81.3%)	ns
Race				
White	95 (70.9%)	22 (23.2%)	73 (76.8%)	ns
Black	36 (26.9%)	10 (27.8%)	26 (72.2%)	
Asian/Pacific Islander	3 (2.2)	0 (0%)	3 (100.0%)	
Missing	56	13	43	
Smoking	44 (30.1%)	8 (18.2%)	36 (81.8%)	ns
Insurance type				
Self/uninsured	11 (6.0%)	5 (45.5%)	6 (54.5%)	ns
Private/commercial	100 (54.0%)	23 (23.0%)	77 (77.0%)	
Government supported	37 (20.0%)	7 (18.9%)	30 (81.1%)	
Other	11 (6.0%)	2 (18.2%)	9 (81.8%)	
Missing	31	8	23	
Validation type				
Normal	37 (19.5%)	0 (0.0%)	37 (100.0%)	$X^2 = 190.5$, $P < .0001$
ASD	45 (23.7%)	45 (100.0%)	0 (0.0%)	
PFO	100 (52.6%)	0 (0.0%)	100 (100.0%)	
Other CHD	8 (4.2%)	0 (0.0%)	8 (100.0%)	

Abbreviations: ASD, atrial septal defect; CHD, congenital heart disease; PFO, patent foramen ovale.

QI projects,[3] this fact would suggest that 76.3% of the patients identified as having ASDs are actually normal; that is, CHD free. This seriously impacts the validity and generalizability of the findings as false-positive cases are likely to dilute the effect size and mitigate the results. Given the large percentage of patients identified in database studies as CHD through code 745.5, future analyses utilizing codes 745.xx-747.xx to define CHD should consider separate analysis of those identified only through code 745.5. In the absence of an algorithm to distinguish true ASD from no CHD, database studies analyzing 745.5 separately may more accurately represent the findings in those with true CHDs.

5 | LIMITATIONS

Several study limitations should be considered in the interpretation of the results. This study is a retrospective analysis and has all the inherent limitations of such a design. For example, 10 subjects from the random sample of 200 taken from the original full 995 cohort did not have an echocardiogram available for review, and hence they were excluded from the analysis. The current investigation utilized the data from a single health system, and hence the coding practice of using 745.5 to classifying patients as CHD cases when they actually do not have an ASD may reflect a local or regional standard of coding practice, and thus limiting external validity. However, the study population was diverse from a racial perspective, and the clinical characteristics of patients admitted to the hospital did not differ from those in other studies suggesting reasonable generalizability. Despite accounting for multiple variables and assessing for potential confounding factors and effect modifiers, residual confounding variables could have led to the observed results.

6 | CONCLUSIONS

This study demonstrated that the use of ICD-9-CM code 745.5 as an equivalent of the presence of secundum ASD, and therefore CHD, was accurate only in 24% of the cohort who was primarily an adult-aged population. As this code is widely used in population-level investigations of the CHD patient population, it is important to remediate to this problem to avoid the continued generation of conclusions marred by up to 76.3% of false positives. Adequate measures should include performing similar analyses on national and international levels to validate the proportion of false-positive CHD patients in the data sets and allow for solutions to correct previously collected information. The creation and adoption of separate diagnosis codes for ASD, PFO, and "rule-out CHD" should be the focus going forward.

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CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

All authors contributed significantly to the design of the study. All authors take responsibility for the content of the manuscript. All authors have approved the manuscript and agree with submission to Congenital Heart Disease.

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