

In-Hospital Vital Status and Heart Transplants After Intervention for Congenital Heart Disease in the Pediatric Cardiac Care Consortium: Completeness of Ascertainment Using the National Death Index and United Network for Organ Sharing Datasets

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Background—The long-term outcomes of patients undergoing interventions for congenital heart disease (CHD) remain largely unknown. We linked the Pediatric Cardiac Care Consortium (PCCC) with the National Death Index (NDI) and the United Network for Organ Sharing Dataset (UNOS) registries to study mortality and transplant occurring up to 32 years postintervention. The objective of the current analysis was to determine the sensitivity of this linkage in identifying patients who are known to have died or undergone heart transplant.

Methods and Results—We used direct identifiers from 59 324 subjects registered in the PCCC between 1982 and 2003 to test for completeness of case ascertainment of subjects with known vital and heart transplant status by linkage with the NDI and UNOS registries. Of the 4612 in-hospital deaths, 3873 were identified by the NDI as “true” matches for a sensitivity of 84.0% (95% CI, 82.9–85.0). There was no difference in sensitivity across 25 congenital cardiovascular conditions after adjustment for age, sex, race, presence of first name, death year, and residence at death. Of 455 known heart transplants in the PCCC, there were 408 matches in the UNOS registry, for a sensitivity of 89.7% (95% CI, 86.9–92.3). An additional 4851 deaths and 363 transplants that occurred outside the PCCC were identified through 2014.

Conclusions—The linkage of the PCCC with the NDI and UNOS national registries is feasible with a satisfactory sensitivity. This linkage provides a conservative estimate of the long-term death and heart transplant events in this cohort. (*J Am Heart Assoc.* 2016;5:e003783 doi: 10.1161/JAHA.116.003783)

Key Words: cohort study • congenital heart disease • mortality • sensitivity and specificity • transplant

The first and second natural history studies of congenital heart disease (CHD) reported on the short- and long-term mortality of patients preceding the era of modern surgical correction. Subsequent contemporary reports have been small and/or limited to single surgical centers.^{1–15}

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Hence, the Centers for Disease Control and Prevention has identified linkage of databases to assess the long-term outcomes of persons with CHD as filling a critical gap in knowledge.^{16,17}

As the oldest, and one of the largest, registries of CHD in the United States, the Pediatric Cardiac Care Consortium (PCCC) is uniquely placed to address the long-term outcomes of these patients. The PCCC was established in 1982 as a multicenter registry in support of quality improvement in the care of patients with CHD.¹⁸ Between 1982 and 2007, clinical information, including detailed diagnoses and procedure data, was entered by 57 centers, resulting in 112 030 hospital admissions and 118 084 operations for over 137 000 patients.

As part of our effort to study the long-term mortality of patients who have undergone intervention for CHD, we submitted information sufficient for linkage to the National Death Index (NDI)¹⁹ and the United Network for Organ Sharing (UNOS).²⁰ The objective of this study was to determine the sensitivity of this linkage in identifying patients

in the PCCC who are known to have died in the hospital or undergone a heart transplant. Our estimate of sensitivity may then be applied to subsequent analysis of death and transplant events up to 32 years postdischarge as well as other studies seeking to link pediatric patients to surveillance data using a limited set of identifiers.

Methods

The PCCC collected limited identifiers and details about specific CHD, inpatient interventions (including operations as well as transcatheter and electrophysiology procedures), and vital status at discharge, but not long-term outcomes postdischarge. We created an initial data set of 59 324 subjects registered with the PCCC who had codes consistent with an intervention for CHD between January 1, 1982 and April 21, 2003 (the date the Health Insurance Portability and Accountability Act [HIPAA] privacy rule took effect), regardless of vital status at discharge, and who had sufficient information available for linkage. The data set was restricted to subjects undergoing intervention in US-based PCCC centers and included only subjects with a US residence at the time of the intervention (Figure 1). Because the PCCC allows the submission of information on adult patients with CHD, the initial data set included 4049 patients (6.8%) and 139 (3.0%) in-hospital deaths greater than 18 years of age at last admission to a PCCC center. Direct identifiers included first and middle names (when available), surname, date of birth (DOB), sex, and state of last known residence. These data were submitted to the NDI in November 2014 (and updated in November 2015) and to the UNOS in May 2015 to ascertain death or transplant through the latest years available (December 31, 2014 for both). The UNOS registry began tracking transplant events that occurred on or after January 1, 1988. This analysis focuses on the 4612 known in-hospital deaths and 455 heart transplants in patients with CHD that occurred during the respective eligibility period as recorded in the PCCC. All study conduct was approved by the Human Subjects Research Protection Program of the University of Minnesota (Minneapolis, MN) and the Emory School of Medicine (Atlanta, GA) with a waiver of consent for the linkages. All analyses were conducted using R software (version 3.2.3; R Foundation for Statistical Computing, Vienna, Austria).

Because names were collected in a single field, without the intention of later linkage, these data required cleaning before submission to the NDI. In particular, it was not possible to distinguish between middle names and hyphenated surnames among 440 subjects, where 2 names were concatenated. Therefore, duplicate records were submitted for subjects who encompassed both possibilities. A small number of subjects

($n=60$) had 2-letter entries for first name, which may have represented abbreviations or legitimate names, and were submitted as is. Likewise, 634 subjects had “Baby boy” or “Baby girl” entered rather than a first name, and these records were also submitted as is. At this stage, we did not submit duplicate records encompassing common alternate spellings or nicknames for first names.

All records were assigned a score by the NDI based on its standard algorithm, which is explained in detail in the NDI User’s Guide.²¹ Briefly, the NDI matches²² are based on 7 criteria: (1) Social Security number (SSN); (2) exact month and ± 1 year of birth, first and last name; (3) exact month and ± 1 year of birth, first and middle initials, and last name; (4) exact month and day of birth, first and last name; (5) exact month and day of birth, first and middle initials, and last name; (6) exact month and year of birth, first name, and father’s surname; and (7) if the subject is female: exact month and year of birth, first name, last name (on user’s record), and father’s surname (on NDI record) and computes a probabilistic match score. Based on their matching algorithm, the NDI assigns each possible match to 1 of 5 classes, 1 through 5, that reflect the fact that some of the matching criteria are more important than others, with 1 being the highest quality and 5 the poorest. All of class 1 matches are considered real, class 5 matches are considered false, and classes 2 to 4 are assigned as either true or false based on score cut-off points determined for best performance. Class 1, 2, and 5 matches require an SSN, which was not collected by the PCCC; hence, all of our matches were class 3 or 4. Class 3 matches are those without an SSN, but match on 8 or more of the following items: first name, middle initial, last name, father’s surname (for females), birth day, birth month, birth year, sex, race, marital status, or state of birth. Class 4 matches are those that match on fewer than 8 of the items required for a class 3 match. The NDI provides all possible matches to each submitted record with both a class and probabilistic score so that investigators may devise their own cutoffs for determining deaths.

We conducted a custom multistep matching algorithm to determine deaths from the NDI that systematically selected matches according to match quality. Our first step was to accept any original or duplicate (ie, alternate surname) record that which matched perfectly with the NDI.²⁰ Next, we accepted any class 3 record with an NDI score >37.5 ; third we accepted any class 3 record, regardless of NDI score <37.5 , if there was an exact match on full name, sex, DOB, and residence at death. This was then repeated using residence at birth rather than death. Finally, we accepted class 4 matches with scores >32.5 and names that were phonetic matches of the last name if sex, DOB, and residence at death or birth completely matched.

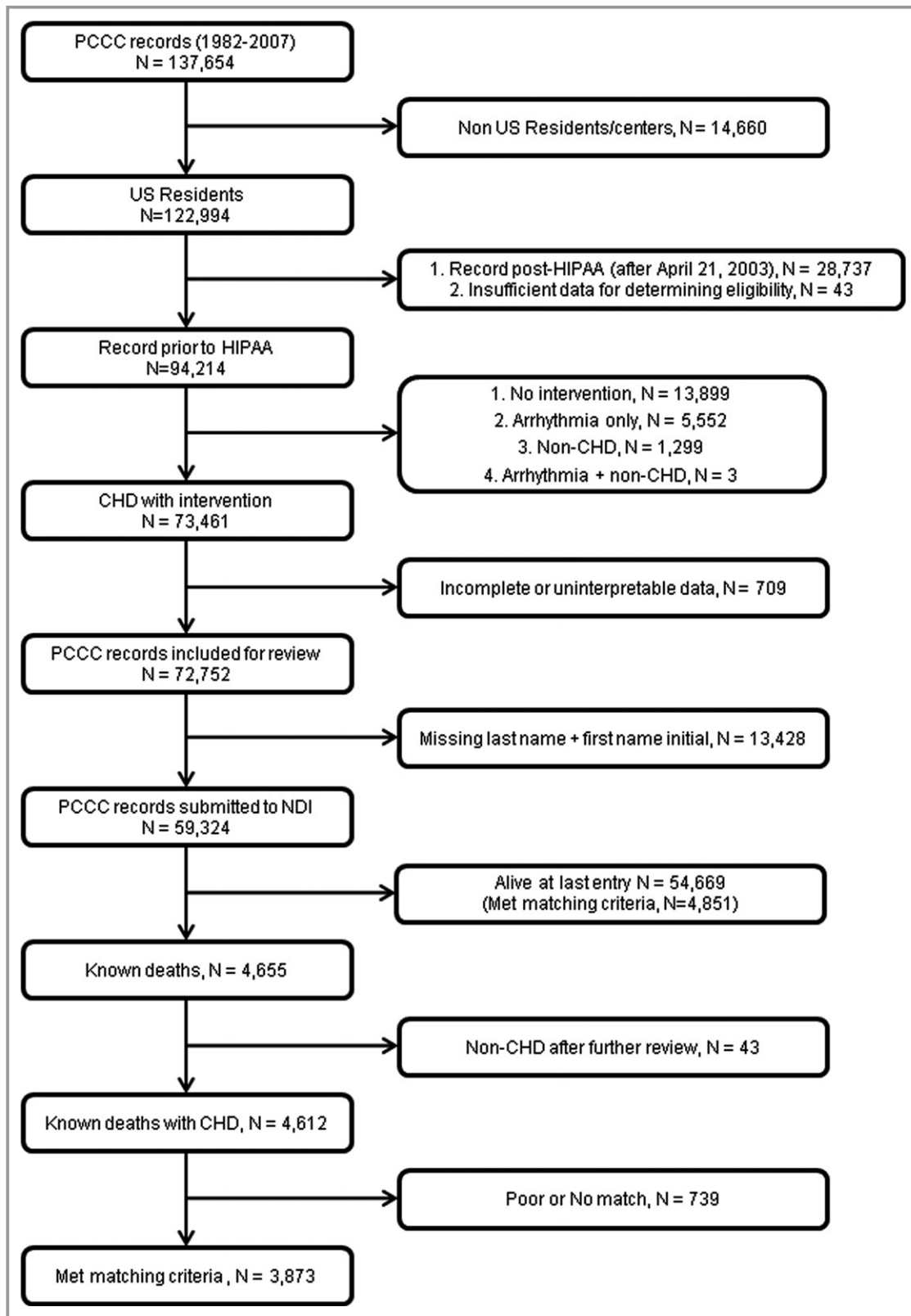


Figure 1. STROBE-style flow diagram displaying selection of the study PCCC cohort for linkage with NDI and UNOS after exclusion of patients because of non-US-resident status, non-US-center where intervention was performed, inadequate direct identifiers, and lack of pediatric/congenital heart diagnosis or intervention for such condition. CHD indicates congenital heart disease; HIPAA, Health Insurance Portability and Accountability Act; NDI, National Death Index; PCCC, Pediatric Cardiac Care Consortium; UNOS, United Network for Organ Sharing.

All 59 324 PCCC subjects included in the NDI search were also sent to UNOS for matching with their Organ Procurement and Transplantation Network (OPTN) database, which began on January 1, 1988. The UNOS used an internal probabilistic matching algorithm that assigned a likelihood of match based on name, sex, and DOB; “ambiguous” matches were then settled by comparing common variations of names when sex and DOB matched. All matches determined by the UNOS were returned to us. Multiple matches for a single individual for those who had more than 1 transplant were created by linking to the OPTN database. Next, we determined which transplant records in the UNOS corresponded to those in the PCCC by matching on date; those UNOS records of transplant that occurred within 1 week before or after the date of transplant in the PCCC were considered true matches. In instances where only the year or year and month of transplant were known (N=12), matching was considered satisfactory when the UNOS year or year and month of transplant matched.

For the purposes of calculating sensitivity and specificity, we treated CHD deaths reported to the PCCC as a gold standard. These were subjects for whom death was confirmed by a discharge summary, autopsy report, or death certificate available in the PCCC. Similarly, subjects with a PCCC procedure code indicating heart transplant were considered true recipients because these codes were assigned on the basis of direct review of operative reports. Sensitivity was calculated as the number of NDI or UNOS matches divided by the number of known deaths or heart transplants in the PCCC. The 95% CIs of match success were calculated by using the binomial exact procedure for the NDI, whereas a clustered bootstrap using 10 000 samples was used for the UNOS because of the potential for within-person correlation when patients had multiple transplants. To estimate the specificity of the linkages, we analyzed individuals with multiple hospital admissions documented in the PCCC, for whom a period of transplant-free survival could be inferred, consisting of 24 818 individuals for the PCCC-NDI linkage and 22 910 individuals for the PCCC-UNOS linkage (because UNOS did not begin operation until 1988). Specificity for each of the linkages was then calculated as the number of NDI or UNOS nonevents, respectively, up to the last known date alive, using the same algorithm as for known deaths, divided by the denominator above. The 95% CI was calculated as above.

In order to better understand factors affecting death ascertainment, we conducted a multivariable analysis of patient characteristics and match success. The independent variables were sex, race, primary diagnosis, year of death, age at death, state of residence at death, and availability of first name. All variables were categorical, with the exception of age, which was transformed using restricted cubic splines with 3 knots at the 10th, 50th, and 90th percentiles to allow for nonlinearity. Race was not explicitly captured by the PCCC

and so was abstracted from records in which it was included incidentally; missing data were addressed by creating a category to indicate missing race. Logistic regression was used to model matches in the NDI. Overall tests for each factor were computed using likelihood ratio tests. We present only descriptive data on heart transplant recipients, because there were too few nonmatches to similarly conduct multivariable analysis of match success. Sex, age, race, ethnicity, and availability of first name are summarized at the person level for UNOS matches. Year of transplant and state of transplant are summarized separately for first and second transplant in the PCCC.

Results

Characteristics of known deaths in the PCCC by success of matching with the NDI are presented in Table 1. Overall, 3873 of 4612 deaths had a matching record in the NDI, for a sensitivity of 84.0% (95% CI, 82.9–85.0). Sensitivity was higher among the 4392 subjects who had a first name available within the PCCC, with 3869 deaths matching for a sensitivity of 88.1% (95% CI, 87.1–89.0).

Among the 24 818 PCCC patients who had 2 or more inpatient visits with a median observation time between visits of 1.57 years (or 88 783 person-years total), 63 had a matching NDI record indicating death on or preceding the second visit, resulting in a specificity of 99.75% (95% CI, 99.68–99.80). Given the high specificity, we did not pursue further analysis.

The results of the multivariable analysis of matches with the NDI are shown graphically in Figure 2 and presented in full in Table 2. Odds Ratios (ORs) represent the ratio of the odds of a match with the NDI. Overall tests of association were significant for year of death ($P=0.010$), age at death ($P<0.001$), residence at death ($P<0.001$), availability of first name ($P<0.001$), and race ($P<0.001$). Sex ($P=0.116$) and diagnosis group ($P=0.294$) were not significantly associated with match success.

Individual year of death from 1982 to 2002 was compared to 2003; whereas all ORs were between 0.10 and 0.60, indicating lower odds of match than in 2003, those for 1983, 1984, 1989, 1991–1994, 1998, 2000, and 2002 were significant. To present estimates of the odds of matching by age at death, 3 ages were compared to 1 year of age at death (18 years, 9 years, and 28 days, respectively) that are commonly used for categorization of age in CHD patients. Higher odds of matching were observed for 18 versus 1 year (OR=1.68; 95% CI, 1.11–2.54) and 9 versus 1 year (OR=1.68; 1.38–20.04), and lower odds of matching were observed for 28 days versus 1 year (OR=0.59; 95% CI, 0.48–0.71). Because the relationship between age at death and matching

Table 1. PCCC Deaths by Characteristic and Match Success*

Characteristic	Match	No Match
N (%)	3873 (84.0)	739 (16.0)
Sex, N (%)		
Female	1714 (82.9)	354 (17.1)
Male	2159 (84.9)	385 (15.1)
Death age, y		
Death age, mean (SD)	2.3 (6.1)	0.9 (3.5)
Death age, median [range]	0.3 [0.0, 77.0]	0.1 [0.0, 42.2]
Race as modeled, N (%)		
Black	419 (77.3)	123 (22.7)
Other	80 (75.5)	26 (24.5)
White	2097 (88.1)	283 (11.9)
Missing	1277 (80.6)	307 (19.4)
First name missing, N (%)		
No	3869 (88.1)	523 (11.9)
Yes	4 (1.8)	216 (98.2)
First initial only, N (%)		
No	3872 (84.0)	739 (16.0)
Yes [†]	1 (100.0)	0 (0.0)
Death year, N (%)		
1982	40 (87.0)	6 (13.0)
1983	37 (82.2)	8 (17.8)
1984	57 (90.5)	6 (9.5)
1985	157 (89.2)	19 (10.8)
1986	161 (89.0)	20 (11.0)
1987	128 (87.7)	18 (12.3)
1988	149 (90.3)	16 (9.7)
1989	200 (81.3)	46 (18.7)
1990	167 (87.0)	25 (13.0)
1991	180 (82.6)	38 (17.4)
1992	240 (84.5)	44 (15.5)
1993	222 (81.9)	49 (18.1)
1994	254 (83.0)	52 (17.0)
1995	279 (86.6)	43 (13.4)
1996	287 (86.2)	46 (13.8)
1997	252 (86.3)	40 (13.7)
1998	226 (83.4)	45 (16.6)
1999	199 (82.6)	42 (17.4)
2000	177 (74.4)	61 (25.6)
2001	167 (79.9)	42 (20.1)

Continued

Table 1. Continued

Characteristic	Match	No Match
2002	203 (79.0)	54 (21.0)
2003 and 2004 (N=1) [‡]	58 (84.1)	11 (15.9)
State, N (%)		
AK	12 (92.3)	1 (7.7)
AL [†]	3 (100.0)	0 (0.0)
AR	202 (93.1)	15 (6.9)
AZ	34 (73.9)	12 (26.1)
CA	51 (85.0)	9 (15.0)
CO [†]	3 (100.0)	0 (0.0)
DC	19 (70.4)	8 (29.6)
FL	242 (65.4)	128 (34.6)
GA	81 (65.9)	42 (34.1)
HI [†]	3 (50.0)	3 (50.0)
IA	333 (93.0)	25 (7.0)
ID	11 (84.6)	2 (15.4)
IL	114 (92.7)	9 (7.3)
IN	26 (89.7)	3 (10.3)
KS	83 (85.6)	14 (14.4)
KY	198 (85.7)	33 (14.3)
LA	121 (84.0)	23 (16.0)
MD	59 (80.8)	14 (19.2)
MI [†]	4 (100.0)	0 (0.0)
MN	441 (89.8)	50 (10.2)
MO	320 (89.9)	36 (10.1)
MS	203 (86.4)	32 (13.6)
NC [†]	2 (66.7)	1 (33.3)
ND	42 (91.3)	4 (8.7)
NE	137 (92.6)	11 (7.4)
NM [†]	6 (100.0)	0 (0.0)
NY [†]	5 (83.3)	1 (16.7)
OH	235 (91.1)	23 (8.9)
OK	34 (81.0)	8 (19.0)
OR	25 (54.3)	21 (45.7)
PA	9 (90.0)	1 (10.0)
PR [†]	0 (0.0)	1 (100.0)
SC	192 (78.0)	54 (22.0)
SD	42 (87.5)	6 (12.5)
TN	74 (81.3)	17 (18.7)
TX	253 (74.4)	87 (25.6)
VA	52 (74.3)	18 (25.7)
WA	36 (85.7)	6 (14.3)

Continued

Table 1. Continued

Characteristic	Match	No Match
WI	61 (93.8)	4 (6.2)
WV	104 (86.0)	17 (14.0)
WY [†]	1 (100.0)	0 (0.0)
Primary diagnosis group, N (%)		
AI [†]	4 (66.7)	2 (33.3)
AS/subAS	98 (85.2)	17 (14.8)
ASD	20 (87.0)	3 (13.0)
BVOTO [†]	1 (100.0)	0 (0.0)
CCAA	22 (95.7)	1 (4.3)
CCAVC	348 (92.3)	29 (7.7)
ccTGA	55 (90.2)	6 (9.8)
coA	193 (81.1)	45 (18.9)
Cor-Triatr	9 (90.0)	1 (10.0)
DORV	10 (83.3)	2 (16.7)
dTGA	340 (81.3)	78 (18.7)
IAA	96 (78.0)	27 (22.0)
MR	12 (85.7)	2 (14.3)
MS/supra MV ring	14 (87.5)	2 (12.5)
PA/IVS	74 (76.3)	23 (23.7)
PAA	16 (84.2)	3 (15.8)
PAPVR	12 (92.3)	1 (7.7)
PAVC/TAVC	29 (80.6)	7 (19.4)
PDA	19 (70.4)	8 (29.6)
PS/subPS	33 (80.5)	8 (19.5)
Supra AS	23 (85.2)	4 (14.8)
TAC	154 (85.1)	27 (14.9)
TAPVR	127 (78.9)	34 (21.1)
TOF	498 (86.8)	76 (13.2)
TVA	28 (90.3)	3 (9.7)
UVH	1479 (83.0)	304 (17.0)
VSD	159 (85.9)	26 (14.1)

AS/subAS indicates aortic stenosis/subaortic stenosis; ASD, atrial septal defect; CCAA, congenital coronary artery anomalies; CCAVC, complete common atrioventricular canal; ccTGA, congenitally corrected transposition of great arteries; CoA, coarctation of aorta; Cor-Triart, cor triatriatum; DORV, double outlet right ventricle; dTGA, dextro-transposition of great arteries; IAA, interrupted aortic arch; MR, mitral regurgitation; MS/supra MV ring, mitral stenosis/supra-mitral valve ring; PA/IVS, pulmonary atresia/intact ventricular septum; PAA, pulmonary artery atresia; PAPVR, partial anomalous pulmonary venous return; PAVC/TAVC, partial atrioventricular canal/transitional atrioventricular canal; PCCC, Pediatric Cardiac Care Consortium; PDA, patent ductus arteriosus; PS/subPS, pulmonary stenosis/subpulmonary stenosis; Supra AS, supra aortic stenosis; TAC, transverse aortic constriction; TAPVR, total anomalous pulmonary venous return; TOF, tetralogy of fallot; TVA, tricuspid valve atresia; UVH, univentricular heart; VSD, ventricular septal defect.

*Percents are within characteristic and stratified by match success.

[†]Excluded from model.

[‡]Because of the small number of deaths that occurred in 2004 (partial year), these were added to the deaths that occurred at 2003.

was nonlinear, we also present these results graphically in Figure 3; matching probability was sharply decreased for deaths under 3 years of age and especially poor for infants. We compared matching odds to that in Minnesota, the most frequent state of residence at death. Significantly lower odds of match were observed for deaths occurring in Southern states (Florida, Georgia, Kentucky, Louisiana, South Carolina, and Virginia), Oregon, and Texas. Compared to white patients, black patients (OR=0.53; 95% CI, 0.39–0.72), patients with other race (OR=0.35; 95% CI, 0.20–0.63), and patients with missing race (OR=0.57; 95% CI, 0.45–0.73), each had lower odds of an NDI match. The odds of matching were very low if first name was unavailable, because only 4 of 220 such patients matched with an NDI record. There was no overall association between diagnostic group and the odds of an NDI match.

Overall sensitivity of known heart transplants matching to the UNOS, as well as that by patient characteristic, is shown in Table 3. Overall, there were matching UNOS records for 408 of 455 known heart transplant events, resulting in a sensitivity of 89.7% (95% CI, 86.9–92.3). Males were more likely to match (247 of 263; 93.9%) than females (156 of 173; 90.2%). Nonmatches were slightly older (mean=8.5 years) than matches (mean=7.3 years). Distribution of matches by race and year of transplant was similar, and there were too few children without first name to evaluate its association with matching. We assessed the identification of transplant among 22 910 individuals in the PCCC who had 2 or more inpatient visits. Among these patients with median observation time of 1.84 years (87 249 person-years), there were 21 heart transplants in the UNOS that were not captured by the PCCC where the heart transplant occurred inside the earliest and latest dates of observation for a person. Thus, specificity was 22 889 of 22 910 or 99.91% (95% CI, 99.86–99.94). An additional 363 heart transplants occurring after the last PCCC record were detected by linkage to the UNOS through 2014.

Discussion

Using a large and long-standing registry of CHD patients and their clinical care, we were able to estimate the accuracy of matching with the NDI and UNOS without the availability of SSN. A large majority of both known deaths and known transplants were detected by probabilistic linkage, a finding that supports the use of the PCCC to evaluate long-term outcomes in CHD patients. Moreover, this analysis is one of few reports of linkage to the NDI in a primarily pediatric population,²³ and, to our knowledge, the first regarding linkage with the UNOS, which may be of interest to other pediatric researchers.

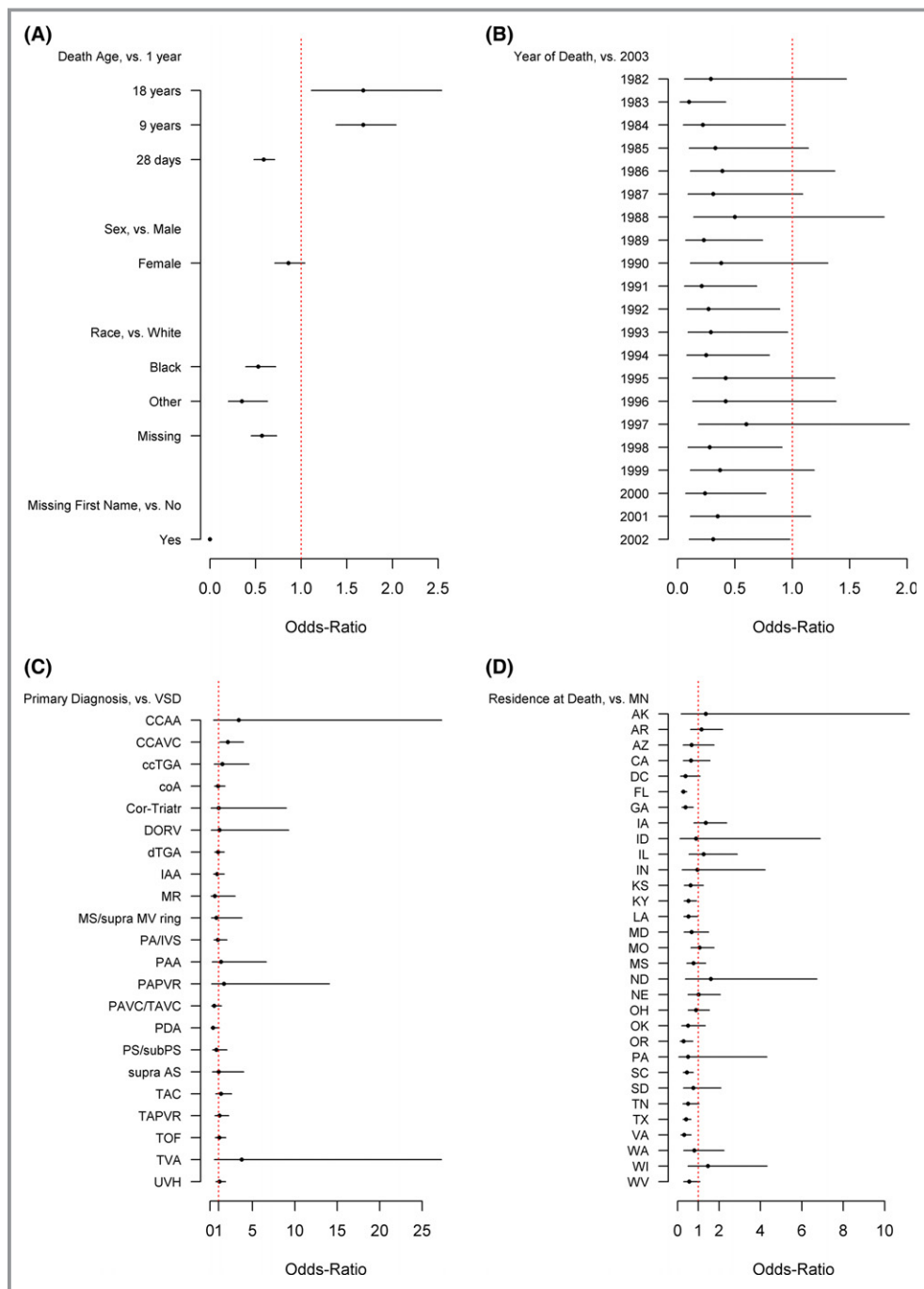


Figure 2. Tree plot of odds ratios for each factor in the multivariable logistic regression model with the outcome, successful match with the NDI (A: Age at death, sex, race, and availability of first name; B: Primary diagnosis; C: Year of death; D: Residence at death). AS/subAS indicates aortic stenosis/subaortic stenosis; ASD, atrial septal defect; CCAA, congenital coronary artery anomalies; CCAVC, complete common atrioventricular canal; ccTGA, congenitally corrected transposition of great arteries; CoA, coarctation of aorta; Cor-Triart, cor triatriatum; DORV, double outlet right ventricle; dTGA, dextro-transposition of great arteries; IAA, interrupted aortic arch; MR, mitral regurgitation; MS/supra MV ring, mitral stenosis/supra-mitral valve ring; NDI, National Death Index; PA/IVS, pulmonary atresia/intact ventricular septum; PAA, pulmonary artery atresia; PAPVR, partial anomalous pulmonary venous return; PAVC/TAVC, partial atrioventricular canal/transitional atrioventricular canal; PDA, patent ductus arteriosus; PS/subPS, pulmonary stenosis/subpulmonary stenosis; Supra AS, supra aortic stenosis; TAC, transverse aortic constriction; TAPVR, total anomalous pulmonary venous return; TOF, tetralogy of fallot; TVA, tricuspid valve atresia; UVH, univentricular heart; VSD, ventricular septal defect.

Table 2. ORs Corresponding to Tree Plots

Comparison	OR	Lower	Upper
Death age			
28 days vs 1 year	0.59	0.48	0.71
9 years vs 1 year	1.68	1.38	2.04
18 years vs 1 year	1.68	1.11	2.54
Sex, vs male			
Female	0.86	0.71	1.04
Death year, vs 2003			
1982	0.29	0.06	1.47
1983	0.10	0.02	0.42
1984	0.22	0.05	0.94
1985	0.33	0.10	1.14
1986	0.39	0.11	1.37
1987	0.31	0.09	1.09
1988	0.50	0.14	1.80
1989	0.23	0.07	0.74
1990	0.38	0.11	1.31
1991	0.21	0.06	0.69
1992	0.27	0.08	0.89
1993	0.29	0.09	0.96
1994	0.25	0.08	0.80
1995	0.42	0.13	1.37
1996	0.42	0.13	1.38
1997	0.60	0.18	2.02
1998	0.28	0.09	0.91
1999	0.37	0.11	1.19
2000	0.24	0.07	0.77
2001	0.35	0.11	1.16
2002	0.31	0.10	0.98
Residence at death, vs MN			
AK	1.36	0.17	11.19
AR	1.16	0.62	2.17
AZ	0.68	0.26	1.76
CA	0.65	0.27	1.56
DC	0.39	0.14	1.08
FL	0.28	0.18	0.44
GA	0.39	0.21	0.75
IA	1.36	0.78	2.37
ID	0.89	0.11	6.89
IL	1.26	0.55	2.88
IN	0.96	0.22	4.22
KS	0.63	0.32	1.24

Continued

Table 2. Continued

Comparison	OR	Lower	Upper
KY	0.53	0.31	0.91
LA	0.53	0.30	0.96
MD	0.68	0.31	1.50
MO	1.06	0.64	1.76
MS	0.77	0.44	1.35
ND	1.61	0.38	6.73
NE	1.02	0.50	2.06
OH	0.89	0.51	1.53
OK	0.51	0.20	1.34
OR	0.29	0.12	0.73
PA	0.51	0.06	4.31
SC	0.45	0.27	0.75
SD	0.76	0.28	2.09
TN	0.51	0.25	1.03
TX	0.41	0.26	0.64
VA	0.32	0.16	0.65
WA	0.81	0.29	2.24
WI	1.47	0.50	4.32
WV	0.56	0.29	1.07
First name missing, vs No			
Yes	0.00	0.00	0.01
Race, vs White			
Black	0.53	0.39	0.72
Other	0.35	0.2	0.63
Missing	0.57	0.45	0.73
Diagnosis group, vs VSD			
AS/subAS	1.06	0.49	2.27
ASD	0.91	0.19	4.30
CCAA	3.39	0.42	27.31
CCAVC	2.12	1.14	3.95
CCTGA	1.47	0.47	4.60
CoA	0.96	0.52	1.76
Cor-Triatr	1.04	0.12	8.99
DORV	1.12	0.14	9.28
D-TGA	0.96	0.55	1.68
IAA	0.84	0.42	1.68
MR	0.57	0.11	2.97
MS/supra MV ring	0.75	0.15	3.77
PA/IVS	0.94	0.45	2.00
PAA	1.31	0.26	6.64
PAPVR	1.66	0.19	14.07

Continued

Table 2. Continued

Comparison	OR	Lower	Upper
PAVC/TAVC	0.50	0.19	1.35
PDA	0.37	0.12	1.08
PS/subPS	0.76	0.29	2.00
Supra AS	1.04	0.27	3.98
TAC	1.30	0.67	2.54
TAPVR	1.13	0.57	2.21
TOF	1.09	0.63	1.87
TVA	3.74	0.51	27.29
UVH	1.13	0.69	1.84

AS/subAS indicates aortic stenosis/subaortic stenosis; ASD, atrial septal defect; CCAA, congenital coronary artery anomalies; CCAVC, complete common atrioventricular canal; ccTGA, congenitally corrected transposition of great arteries; CoA, coarctation of aorta; Cor-Triart, cor triatriatum; DORV, double outlet right ventricle; dTGA, dextro-transposition of great arteries; IAA, interrupted aortic arch; MR, mitral regurgitation; MS/supra MV ring, mitral stenosis/supra-mitral valve ring; ORs, odds ratios; PA/IVS, pulmonary atresia/intact ventricular septum; PAA, pulmonary artery atresia; PAPVR, partial anomalous pulmonary venous return; PAVC/TAVC, partial atrioventricular canal/transitional atrioventricular canal; PDA, patent ductus arteriosus; PS/subPS, pulmonary stenosis/subpulmonary stenosis; Supra AS, supra aortic stenosis; TAC, transverse aortic constriction; TAPVR, total anomalous pulmonary venous return; TOF, tetralogy of fallot; TVA, tricuspid valve atresia; UVH, univentricular heart; VSD, ventricular septal defect.

The sensitivity achieved by our matching to the NDI was similar to that reported by others. In an early test of matching in the absence of a unique identifier, Williams et al. linked data from residents of Veterans Affairs nursing homes to the NDI with and without the use of SSN.²⁴ Using first initial, last

name, and complete DOB, they achieved a sensitivity of 83% compared with 97% if only SSN was used. Similar sensitivity was achieved in other studies of adults in a review by Cowper et al.¹⁹ A report from the National Wilms Tumor Study, a cancer that occurs mainly in children under 5 years of age, is the only relevant assessment of NDI linkage in a pediatric population.²³ A sensitivity of 87.8% was achieved when SSN was not always available; in multivariable analysis, lack of SSN, Hispanic ethnicity, and foreign birth were associated with lower sensitivity. Unlike our study, sensitivity did not differ by age at death.

Our larger goal is to describe the long-term mortality and transplant experience of the PCCC cohort after discharge. Because this cohort includes patients from 50 states plus Puerto Rico and 44 institutions enrolled over 21 years, we believe it to be representative of the population operated on for CHD between 1982 and 2003. In addition, the case mix at the PCCC and distribution of surgical risk categories are fairly comparable with the largest available clinical and administrative data sets and with a very large international referral center for the same period.¹⁸

The understanding of the altered natural history (life-span length, end-stage heart failure, and causes of death) of this population remains a significant knowledge gap for this population.^{16,17} Although the creation of a prospective, longitudinal database is the ideal source to address this gap, it will take a considerable amount of time before collecting similar duration of longitudinal data. Within the realm of currently available resources, the use of the PCCC

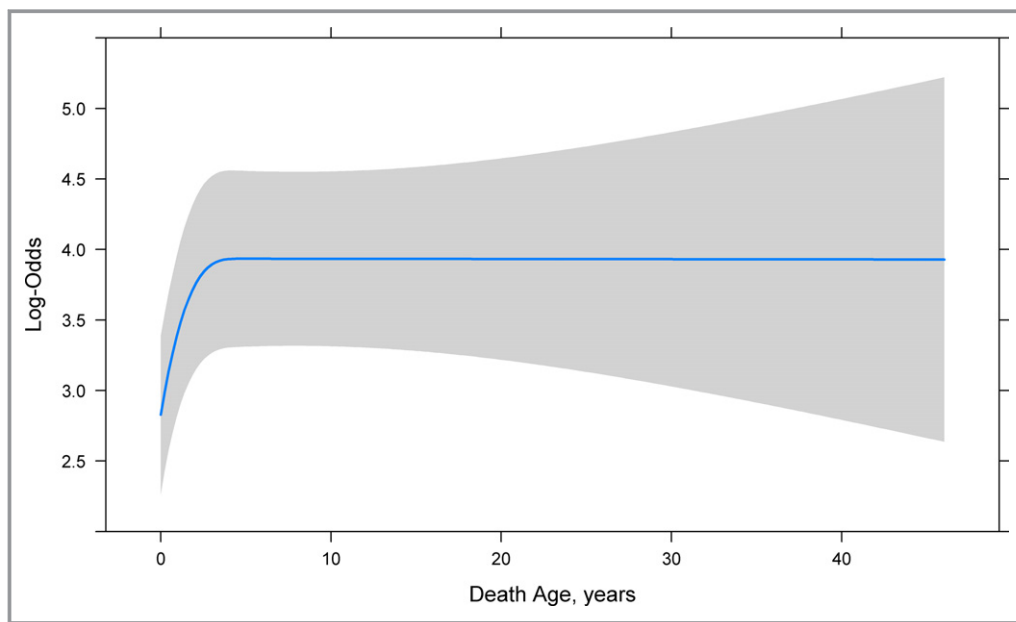


Figure 3. Predicted log odds and 95% CI of NDI match by death age in years from the restricted cubic spline in the multivariable logistic regression model. Other variables in the model were set at their mode. NDI, National Death Index.

Table 3. PCCC Transplants by Characteristic and Match Success

Characteristic	Match	Non-Match
N (%)	403 (92.4)	33 (7.6)
Sex, N (%)		
Female	156 (90.2)	17 (9.8)
Male	247 (93.9)	16 (6.1)
Approximate age at first transplant, y		
Age, mean (SD)	7.3 (8.3)	8.5 (11.7)
Age, median [range]	4.1 [0.0, 36.4]	1.3 [0.0, 44.2]
Race, N (%)		
American Indian/Alaskan Native	0 (0.0)	1 (100.0)
Asian	2 (100.0)	0 (0.0)
Black or African American	33 (94.3)	2 (5.7)
More than 1 race	1 (100.0)	0 (0.0)
White	174 (92.1)	15 (7.9)
Unknown or not reported	193 (92.8)	15 (7.2)
Ethnicity, N (%)		
Hispanic	11 (91.7)	1 (8.3)
Not Hispanic	2 (100.0)	0 (0.0)
Unknown or not reported	390 (92.4)	32 (7.6)
First name missing, N (%)		
No	402 (92.8)	31 (7.2)
Yes	1 (33.3)	2 (66.7)
Year of first transplant, N (%)		
1988	7 (100.0)	0 (0.0)
1989	11 (100.0)	0 (0.0)
1990	14 (100.0)	0 (0.0)
1991	20 (90.9)	2 (9.1)
1992	15 (100.0)	0 (0.0)
1993	39 (95.1)	2 (4.9)
1994	22 (84.6)	4 (15.4)
1995	24 (88.9)	3 (11.1)
1996	33 (94.3)	2 (5.7)
1997	37 (88.1)	5 (11.9)
1998	29 (90.6)	3 (9.4)
1999	19 (86.4)	3 (13.6)
2000	24 (92.3)	2 (7.7)
2001	30 (93.8)	2 (6.3)
2002	24 (96.0)	1 (4.0)
2003	17 (89.5)	2 (10.5)
2004	16 (100.0)	0 (0.0)

Continued

Table 3. Continued

Characteristic	Match	Non-Match
2005	11 (91.7)	1 (8.3)
2006	4 (80.0)	1 (20.0)
2007	7 (100.0)	0 (0.0)
Year of second transplant, N (%)		
1988	0 (0.0)	1 (100.0)
1991	0 (0.0)	2 (100.0)
1992	0 (0.0)	1 (100.0)
1993	0 (0.0)	2 (100.0)
1994	0 (0.0)	1 (100.0)
1995	1 (100.0)	0 (0.0)
1996	0 (0.0)	1 (100.0)
1998	1 (100.0)	0 (0.0)
1999	1 (100.0)	0 (0.0)
2003	1 (50.0)	1 (50.0)
2004	1 (50.0)	1 (50.0)
2005	0 (0.0)	3 (100.0)
2006	0 (0.0)	1 (100.0)
State of first transplant, N (%)		
AR	73 (96.1)	3 (3.9)
AZ	4 (100.0)	0 (0.0)
CA	8 (80.0)	2 (20.0)
FL	69 (87.3)	10 (12.7)
GA	1 (100.0)	0 (0.0)
IA	23 (95.8)	1 (4.2)
IL	1 (100.0)	0 (0.0)
KY	6 (100.0)	0 (0.0)
LA	3 (100.0)	0 (0.0)
MD	6 (85.7)	1 (14.3)
MN	26 (89.7)	3 (10.3)
MO	126 (94.7)	7 (5.3)
MS	3 (100.0)	0 (0.0)
NE	9 (90.0)	1 (10.0)
OH	4 (100.0)	0 (0.0)
OK	1 (100.0)	0 (0.0)
OR	0 (0.0)	1 (100.0)
SC	11 (91.7)	1 (8.3)
SD	1 (100.0)	0 (0.0)
TN	4 (80.0)	1 (20.0)
TX	22 (91.7)	2 (8.3)
VA	2 (100.0)	0 (0.0)

Continued

Table 3. Continued

Characteristic	Match	Non-Match
State of second transplant, N (%)		
AR	2 (50.0)	2 (50.0)
FL	0 (0.0)	3 (100.0)
IA	0 (0.0)	2 (100.0)
LA	0 (0.0)	1 (100.0)
MO	2 (33.3)	4 (66.7)

PCCC indicates Pediatric Cardiac Care Consortium.

registry linkage with existing national registries provides one of the best opportunities to inform the field about long-term outcomes of patients surviving with repaired or palliated CHD. Our ultimate goal is to both compare mortality to that in the general US population adjusting for age, sex, and race (when available) and to compare mortality rates within the cohort. The successful matching of 85% to 90% of known deaths and transplants gives confidence that similar success will be attained for long-term outcomes. We found a number of factors independently associated with match success to the NDI, which will inform the analysis and interpretation. In particular, young age at death, nonwhite race/ethnicity, and Southern state of residence lowered the odds of matching. These associations may be caused by unique features of these subgroups, such as frequent change of names attributed to change in maternal marital status, nonmainstream English names that are prone to differential spelling and typos, and a relatively larger immigrant population in these states. Given that we found different sensitivities by nonclinical characteristics, it is possible that relative risks for these variables may be biased. To correct this in future analyses, sensitivities can be derived from our modeling of match success of known deaths in the PCCC and used to quantitatively adjust for undercounting deaths.²³ Importantly, match success was not associated with the underlying cardiac lesion that necessitated surgery, suggesting that comparison of lesion-specific mortality within the PCCC will not be biased by differential sensitivity.

Conclusions

In summary, these results support the use of PCCC linkage with national registries such as the NDI and UNOS to address a major public health gap of knowledge related to the long-term outcomes of patients with repaired or palliated CHD. The identified death and transplant events provide a conservative estimate that will form the basis for estimating the long-term survival of this cohort, whereas the sensitivities found will be used to adjust for undercounting deaths.

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Disclosures

None.

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