A cohort evaluation of surgical pathways reported to a national audit of children undergoing treatment for congenital heart disease in England and Wales

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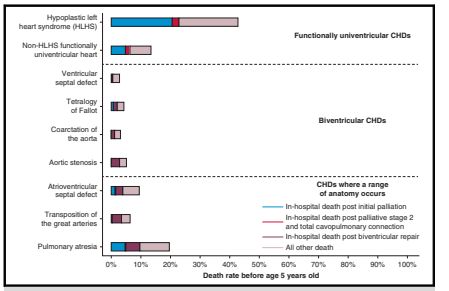
ABSTRACT

Objectives: To ascertain rates of completion of essential cardiac procedures and their overall contribution to longer-term mortality in children with congenital heart disease (CHD).

Methods: In this cohort study using the United Kingdom National CHD Audit, we described the pathway operations required for treatment for 9 sentinel CHDs—hypoplastic left heart syndrome (HLHS), non-HLHS functionally univentricular heart, ventricular septal defect, tetralogy of Fallot, coarctation, aortic stenosis, atrioventricular septal defect, transposition of the great arteries, and pulmonary atresia—that were undertaken at the population level and report the mortality associated with these pathway operations by age 5 years.

Results: Among 28,806 patients, over a median follow-up of 9.8 years (interquartile range, 4.2-15.4 years), 839 (2.9%) had undergone pre-pathway procedures only, 1135 (3.9%) had undergone initial palliation only (inclusive of functionally univentricular and biventricular reparative pathways), 2001 (6.9%) had undergone stage 2 palliation and/or total cavopulmonary connection (TCPC), 2254 (7.8%) had undergone staged biventricular repair after palliation, and 22,572 (78.3%) had undergone primary biventricular repair. Of the 23,239 children with 5 years of follow-up, 1794 (7.7%) had died by age 5 years, consisting of 409 (1.8%) who died following an initial palliative procedure, 43 (0.2%) who died following a stage 2 palliation or TCPC, 379 (1.6%) who died following a biventricular reparative procedure, and 963 (4.1%) who died in other circumstances, such as interstage or following reinterventions.

Conclusions: The outcome metrics of surgical pathway completion (biventricular repair or TCPC) and overall mortality at age 5 years can be evaluated using registry data and could contribute to future assessment of overall CHD service quality. (J Thorac Cardiovasc Surg 2025;170:316-24)



Mortality rates at age 5-years for sentinel CHDs by stage in the surgical pathway.

CENTRAL MESSAGE

Of 28,806 children starting treatment, 24,906 (86.5%) completed repair and 1384 (4.8%) underwent total cavopulmonary connection. Mortality at age 5 years was 7.7%, with 3.6% dying following a pathway procedure.

PERSPECTIVE

In this population-based study of children treated for 9 sentinel congenital heart diseases (CHDs), we captured all stages of planned intervention. The outcome metrics of surgical pathway completion (repair or total cavopulmonary connection) and overall mortality at age 5 years (linked and unlinked to pathway procedures) can be evaluated using registry data and are potentially useful for judging CHD service quality and inform users.

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Abbreviations and Acronyms

AOS = aortic stenosis
AVSD = atrioventricular septal defect
COA = coarctation of the aorta
CHD = congenital heart disease
FUH = functionally univentricular heart
HLHS = hypoplastic left heart syndrome
IQR = interquartile range
IVS = intact ventricular septum
NCHDA = National Congenital Heart Diseases Audit
NHS = National Health Service
PA = pulmonary atresia
PS = pulmonary stenosis
TGA = transposition of the great arteries
TOF = tetralogy of Fallot
TCPC = total cavopulmonary connection
VSD = ventricular septal defect

Scanning this QR code will take you to the table of contents to access supplementary information.

Although postoperative mortality rates for pediatric cardiac surgery are key metrics in quality assurance, these are currently very low.[1-3] Based on the National Congenital Heart Diseases Audit (NCHDA) in the United Kingdom, mortality rates at 30 days improved significantly from 4.3% in 2000 to 2.6% in 2010[4] and 1.6% in 2019 to 2021.[5] Contemporaneously, repairs occurred at younger ages and included a greater proportion of children with complex heart defects and comorbidities.[4] Given these trends in practice and outcomes, clinicians and families have aspired to evaluate a wider range of outcome metrics to better judge the performance of services.[6,7]

Barriers to measuring longer-term outcomes of consistently defined CHD types from procedure-based patient registries include the heterogeneity and complexity of the case mix and the lack of suitable analysis methods. In a prior study,[3] we selected and defined 9 "sentinel CHDs" (including subgroups) suitable for the national audit to use in long-term outcome monitoring. The selection and definition process (covered in detail elsewhere[3]) involved the identification of reasonably prevalent CHDs that frequently require intervention in early life. These sentinel CHDs are hypoplastic left heart syndrome (HLHS); non-HLHS functionally univentricular heart (FUH) conditions (tricuspid atresia and double-inlet ventricle); significant ventricular septal defect (VSD), including single or multiple VSD; tetralogy of Fallot (TOF), including standard TOF, TOF with double-outlet right ventricle, and TOF absent pulmonary valve; coarctation of the aorta (COA), isolated or with VSD; congenital aortic stenosis (AOS), isolated or with multilevel left heart obstruction; atrioventricular septal defect (AVSD), including complete, partial, unbalanced, and TOF with AVSD; transposition of the great arteries (TGA), including TGA with intact ventricle septum (IVS), complex TGA without pulmonary stenosis (PS), and complex TGA with PS; and pulmonary atresia (PA) with VSD or PA IVS. We then created a patient-level cohort of children who were treated for these CHDs in England and Wales between 2000 and 2021 and reported survival rates at age 1, 5, and 10 years.[3] Each child with sentinel CHD is treated with at least one and for many a series of planned cardiac procedures over time.[3,8-10] In the present study, for these sentinel CHDs and their subgroups, we aimed to (1) describe the treatment pathways undertaken and report the proportions of children who completed treatment pathway, defined as either biventricular repair or total cavopulmonary connection (TCPC), and (2) ascertain the contribution of post-treatment pathway procedure mortality to the overall mortality at age 5 years.

METHODS

Study Design

We undertook this retrospective cohort study based on cardiac surgery and interventional catheterization records from the NCHDA for children born between April 2000 and March 2022, with survival status from the Office of National Statistics in August 2023. Submission to the NCHDA is mandatory and subject to external data validation.

Each procedure record contains several diagnostic and procedure codes from the European Paediatric Cardiac Code (itself a derived short list of the International Paediatric and Congenital Cardiac Code, www.ipccc.net).[11] The procedure-based records were linked using pseudonymized patient identifiers to create a patient-based rather than a procedure-based dataset.

Approvals

The study was approved by the North of Scotland NHS Research Ethics Committee on February 14, 2020 (20/NS/0022) and the Health Research Authority Confidentiality Advisory Group on July 12, 2020 (20/CAG/0027), permitting the use of registry data for specific research purposes without consent.

Inclusion Criteria

This cohort study included all children age <18 years who had a cardiac surgery or interventional catheterization recorded in the NCHDA between April 2000 and March 2022 for 1 of the 9 sentinel CHDs.[3] Of note, because the NCHDA is procedure-driven, only patients who underwent cardiac procedures were included in this study.

Exclusion Criteria

We excluded patients born before April 2000 to ensure that a complete procedure history was available for all patients. We excluded patients from overseas, Scotland, and Northern Ireland because we did not have their life status. A very small number of children (n = 651; 2.2%) with major missing procedure records (eg, no cardiac procedure codes) were excluded.

We identified and excluded those patients with miscoded procedure records, informed by clinical expertise (see the flowchart in Figure E1). In future routine monitoring, these patients will be flagged with the treating centers for correction. There was no evidence of worse survival among these patients, as was shown in our prior study.[3]

Risk Factors

We considered 3 patient-level risk factors present at birth: (1) congenital anomalies and genetic syndromes as defined by the NCHDA, which given the high prevalence of Down syndrome, is reported as a standalone category; (2) preterm birth (defined as birth at gestational age <37 weeks),[12] and (3) recent era (born from April 2010 onward).

Outcomes

The outcomes for each sentinel CHD were (1) the proportions of children who completed a pathway, defined as biventricular repair or TCPC, and (2) the cumulative postoperative mortality for all pathway procedures, defined as death during hospitalization. To contextualize the pathway procedure-related mortality, we also report overall mortality at age 5 years.

Interventional Treatment Pathways

We defined the "expected treatment pathways" for each sentinel CHD based on diagnosis and procedure codes as reported previously[3] and present these in Table E1. As depicted in Figure E2, there are 2 broad groups of cardiac procedures that contribute to an overall expected interventional treatment pathway: reparative and palliative (cardiac surgery, interventional catheters, and hybrid types). In our data management, we first identified cardiac procedures on the pathway for FUH; this was most feasible given the narrower range of applicable procedures, and then procedures on the biventricular reparative pathway. Thus, we report outcomes ordered as FUH CHD by prevalence, biventricular CHD by prevalence, then CHD with a range of anatomy by prevalence.

For the FUH sentinel CHDs (HLHS and non-HLHS FUH), we identified the exclusively palliative procedures, which were initial palliation procedures inclusive of surgery and interventional catheterizations, stage 2 Glenn-type surgeries, and TCPC completion.[3,10] For the sentinel biventricular CHDs (VSD, TOF, COA, and AOS), we identified the expected reparative surgery and any prior palliation (eg, systemic-to-pulmonary arterial shunt in TOF, followed by later TOF repair).[9]

For the sentinel CHDs in which a range of anatomy is seen (AVSD, TGA, and PA), the treatment pathway may involve a reparative procedure with or without a prior palliation or, alternatively, staged palliation for FUH. The FUH pathway was identified by the occurrence of stage 1 Norwood type, stage 2 palliation, or TCPC. Patients who had a stage 2 operation and a reparative surgery but no TCPC were assigned to the 1.5-ventricle pathway, reported as a subtype of FUH pathway.

Each child could be allocated only 1 of each defined pathway procedure, and subsequent occurrence of a pathway procedure was classified as a re-intervention.[13] We also captured "pre-pathway procedures," short-term interventions provided after birth and before the pathway surgery (eg, balloon atrial septostomy in TGA).[10]

Statistical Analysis

We report descriptive statistics as number and percentage or as median and interquartile range (IQR). We applied data disclosure control rules that suppress all counts from 1 to 5 (marked as *), allowing 0 to be shown. We calculated postprocedure mortality for all pathway procedures. We considered only those patients who had completed follow-up to age 5 years (born between April 2000 and March 2017) in the analyses of procedure-related and overall mortality at age 5 years. Data management and analyses were performed with Stata version 15 (StataCorp) and R version 4.3.0 (R Foundation for Statistical Computing).

RESULTS

Cohort Population

Figure E1 depicts the inclusion flow chart. Of 67,406 children who underwent a CHD procedure in the study period in England and Wales, there were 28,806 with the 9 sentinel CHDs, excluding those with major data errors or missing data (n = 651; 2.2%); details in Table E2 and Figure E3. The case breakdown was 1228 (4.3%) HLHS, 992 (3.4%) non-HLHS FUH, 6570 (22.8%) VSD, 4568 (15.9%) TOF, 4338 (15.1%) COA, 1624 (5.7%) AOS, 4085 (14.2%) AVSD, 3759 (13.1%) TGA, and 1642 (5.7%) PA. The cohort included 16,557 (57.5%) boys, 2218 (7.7%) preterm births, 3066 (10.6%) patients with Down syndrome, and 3892 (13.5%) patients with another major congenital anomaly or genetic condition.

We depict the median and interquartile range of age at pathway procedures and the number of patients who completed each planned intervention for FUH CHDs in Figure 1 (data provided in Table E3) and for biventricular CHDs when an initial palliation occurred and for biventricular CHDs when no initial palliation occurred in Figure 2 (data provided in Table E4).

Among children with FUH CHDs (HLHS and non-HLHS FUH), and children with complex CHDs (AVSD, TGA, and PA) who had a FUH pathway, the youngest ages at pathway procedures were in children with HLHS. Among children with biventricular sentinel CHDs (VSD, TOF, COA, and AOS), and complex CHDs (AVSD, TGA, and PA) who were on a biventricular reparative pathway, in all subtypes, the age at reparative procedure after prior palliation was older than the age at single-stage biventricular repair. For each sentinel CHD, children who had a prior palliation were less likely to undergo repair than those who did not have prior palliation.

Pathway Types and the Rate of Planned Treatment Pathway Completion

We present the frequency of pathway procedure types by sentinel CHD subtypes in Figure 3 (data provided in Table E5). Among the cohort of 28,806 patients, the median (IQR) follow-up time was 9.8 years (IQR, 4.2-15.4 years), and 839 (2.9%) had undergone a pre-pathway procedure only (eg, balloon atrial septostomy), 1135 (3.9%) had undergone initial palliation only (inclusive of FUH and reparative pathways); 2001 (6.9%) had undergone stage 2 palliation and/or TCPC (including 37 children with 1.5-ventricle circulation), 2254 (7.8%) had a reparative procedure with prior palliation, and 22,572 (78.4%) had a primary biventricular repair. Table 1 describes the number of children who had a pre-pathway procedure and each type of pathway procedure.

In patients with FUH CHDs (HLHS, n = 1228; non-HLHS FUH, n = 992), completion of TCPC occurred in 536 (43.6%) children with HLHS, and 167 (13.6%)

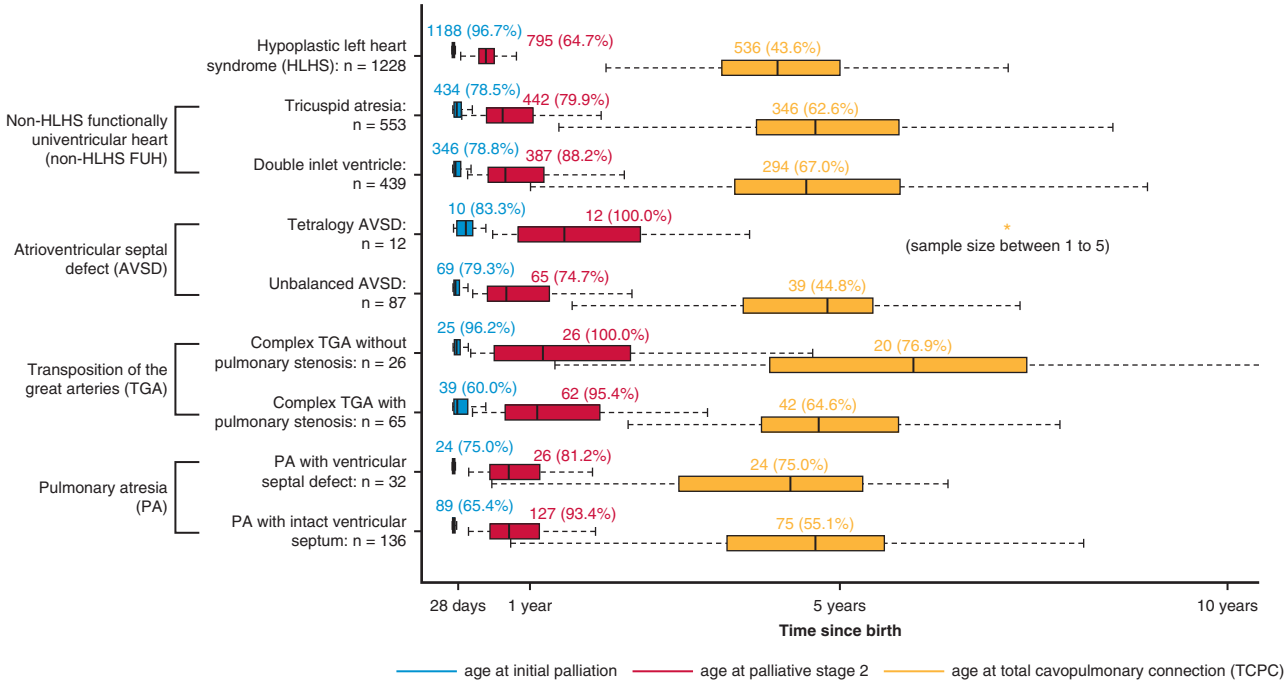


FIGURE 1. Boxplots depicting age at the time of procedure for patients on the functionally univentricular pathway, and the proportion who completed each stage.

children were under follow-up, 22 (0.2%) underwent transplantation, and 503 (41.0%) died before TCPC completion. Of the children with non-HLHS FUH, 640 (64.5%) had completed TCPC and 225 (22.7%) were under follow-up, 1-5 transplanted (number suppressed), and 123 (12.4%) died before TCPC completion.

For sentinel biventricular CHDs (VSD, n = 6570; TOF, n = 4568; COA, n = 4338; AOS, n = 1624), palliative procedures were performed in 469 (7.1%) VSDs, 819 (17.9%) TOFs, 14 (0.3%) COAs, and 137 (8.4%) AOSs. The number of patients with a reparative procedure (with or without prior palliation) was 5860 (89.2%) for VSD, 4481 (98.1%) for TOF, 4282 (98.7%) for COA, and 1514 (93.2%) for AOS. Of those children not completing repair, 71 (1.1%) with VSD died and 639 (9.7%) were under follow-up (including 482 who had a PDA ligation as their only procedure, presumably with small VSD only). For children with TOF, COA, and AOS, 69 (1.5%), 1-5 (number suppressed) and 10 (0.6%), respectively, died without repair; 8 (0.4%), 53 (1.2%), and 105 (6.4%), respectively, remained under follow-up; and <0.1% underwent heart transplantation.

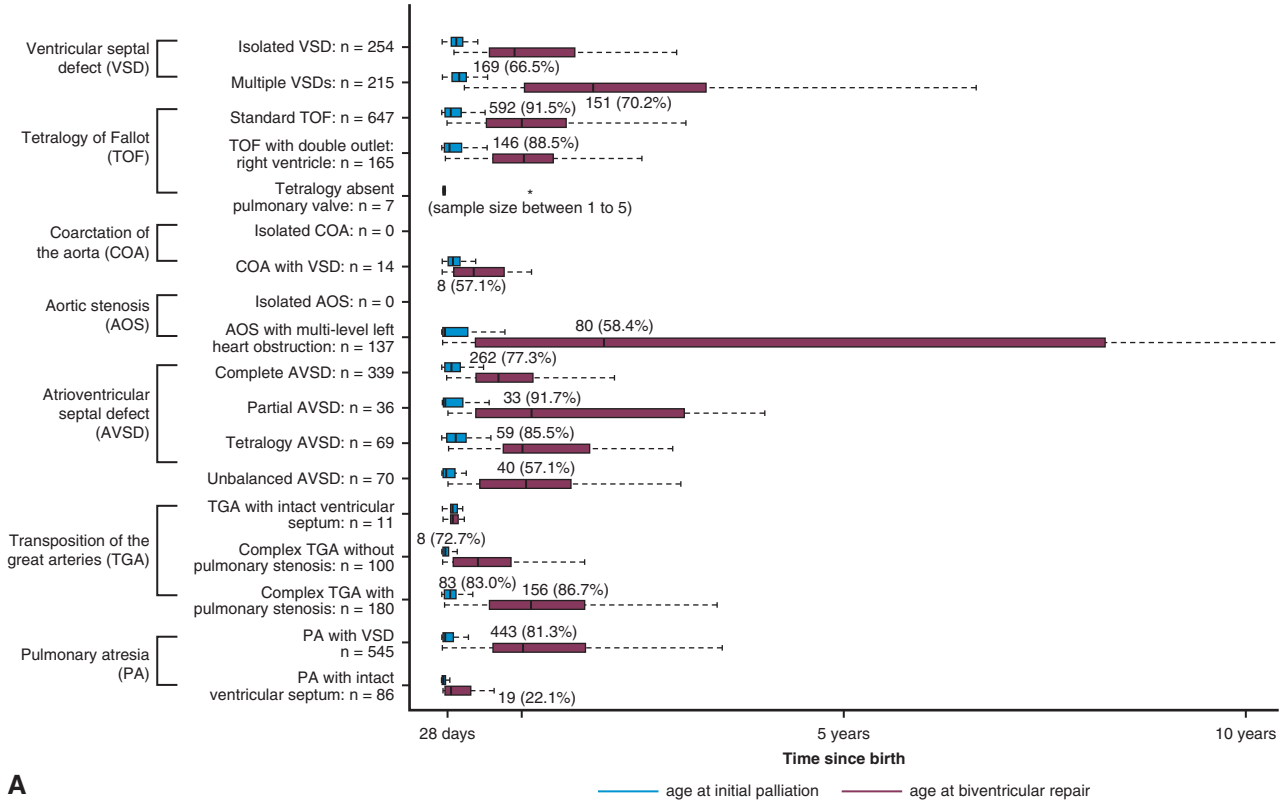
For children with complex sentinel CHDs with variable anatomy (AVSD, n = 4085; TGA, n = 3759; PA, n = 1642), FUH pathway procedures were performed in 99 (2.4%) of those with ASVD, 91 (2.4%) of those with TGA, and 168 (10.2%) of those with PA and TCPC was completed in 44 (44.4%), 62 (68.1%), and 99 (58.9%), respectively. Thirty-five patients (2.1%) with PA had a 1.5-ventricle pathway.

On a biventricular reparative pathway were AVSD (n = 3986; 97.6%), TGA (n = 3668; 97.6%), and PA (n = 1474; 89.8%). Of these, initial palliation occurred in 514 (12.9%), 291 (7.9%), and 631 (42.8%), respectively, and a reparative procedure (with or without a prior palliation) occurred in 3847 (96.4%), 3581 (97.6%), and 1261 (85.5%), respectively. One hundred twenty-five (3.1%) children with AVSD, 68 (1.9%) with TGA, and 155 (10.5%) with PA died without repair; 14 (0.4%) with AVSD, 19 (0.5%) with TGA, and 57 (3.9%) with PA were under follow-up; and <0.1% underwent heart transplantation.

Tables E6 and E7 show the number of children with the considered risk factors. In the whole cohort, initial palliation was more likely with preterm birth (n = 571; 25.7%) and congenital comorbidities (n = 1102; 28.3%), but not with Down syndrome (n = 277; 10.9%). Biventricular repair was less likely with preterm birth (n = 1791; 87.1%) and with congenital comorbidities (n = 3201; 91.4%), but not with Down syndrome (n = 2438; 96.4%).

Postoperative and Overall Mortality at Age 5 Years

In-hospital mortality rates after pathway procedures for all children in the cohort are summarized by sentinel CHD subgroup in Table E8. Procedure-related and overall mortality at age 5 years by sentinel CHD subgroup are depicted in Figure E4, and data are presented in Table E9. Of 23,239 children born between April 2000 and March 2016, 1794 (7.7%) died by age 5 years, consisting of 409 (1.8%) who died following an initial palliative procedure, 43 (0.2%)



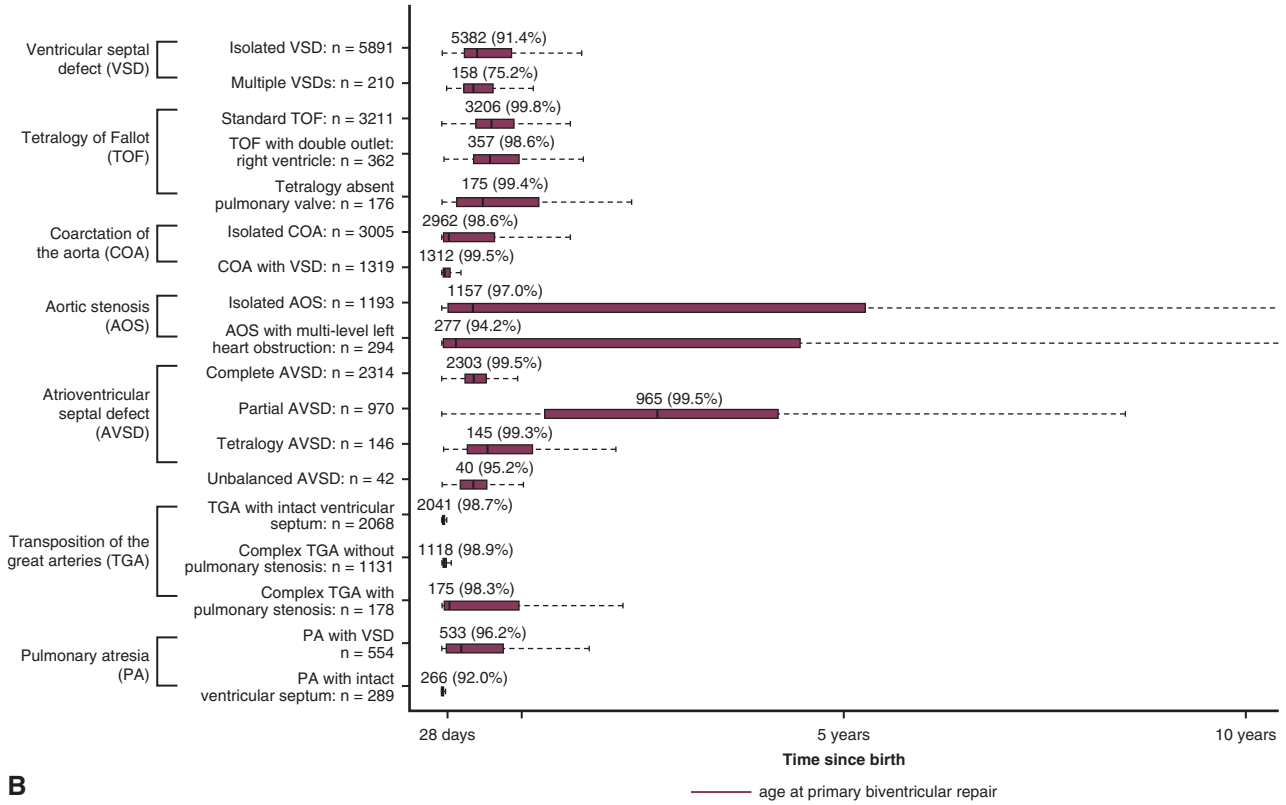


FIGURE 2. Boxplots depicting pathway procedure age and the proportion completing repair for patients with biventricular CHDs who underwent repair with prior palliation (A) or primary biventricular repair (B). *CHD*, Congenital heart disease.

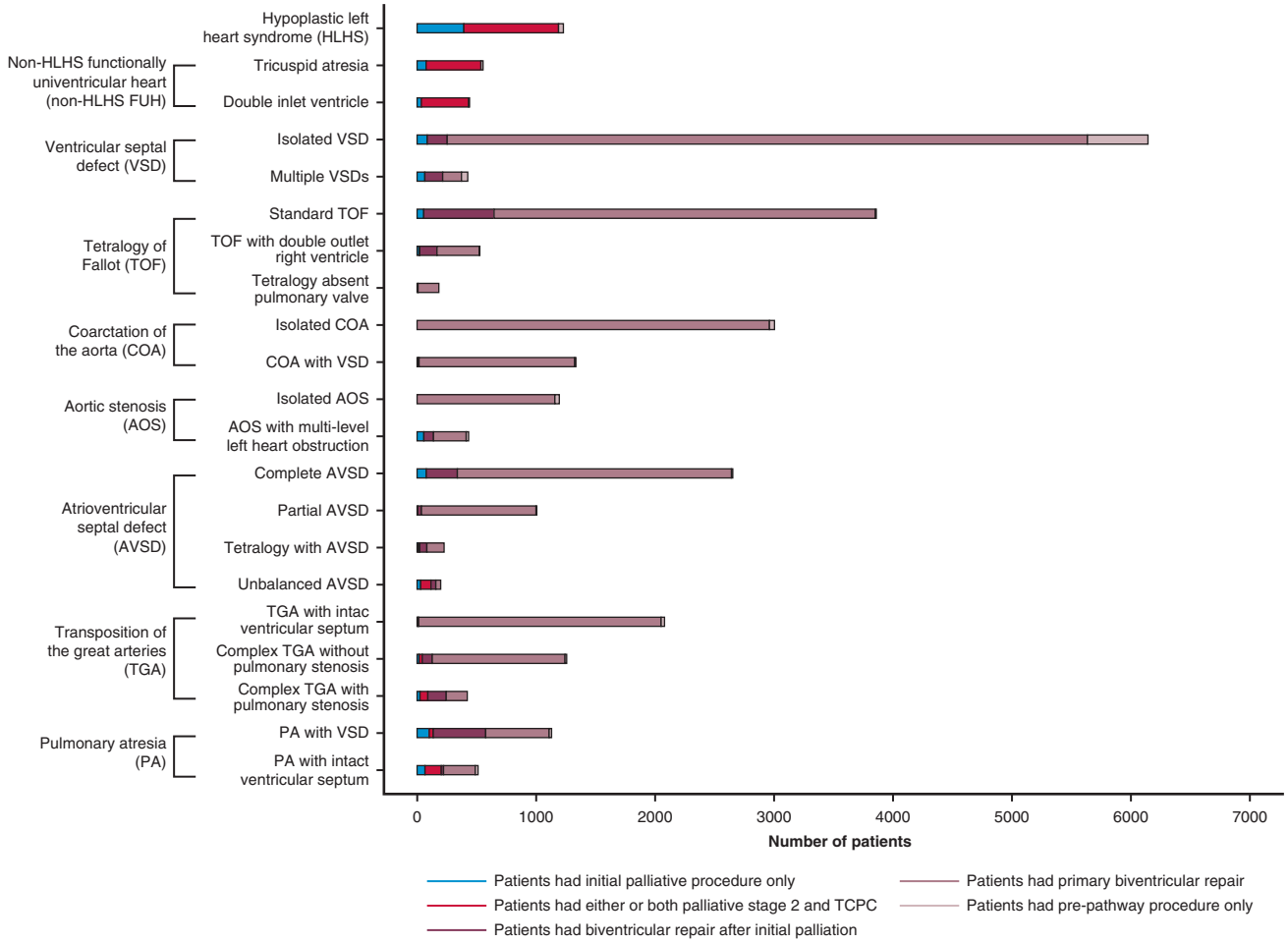


FIGURE 3. Proportion of patients by the type of pathway procedures undertaken for each sentinel CHD subtype. *CHD*, Congenital heart disease; *TCPC*, total cavopulmonary connection.

who died following stage 2 palliation or TCPC, 379 (1.6%) who died following a biventricular repair, and 963 (4.1%) who died in other circumstances, such as interstage or with reinterventions.

Among children with functionally univentricular CHDs (HLHS and non-HLHS FUH), the total overall mortality by age 5 years was 436 (42.8%) for HLHS and 106 (13.4%) for non-HLHS FUH. Mortality related to pathway procedures was 22.9% (n = 233) for HLHS and 6.3% (n = 50) for non-HLHS FUH, and the number who died unrelated to a pathway operation was 203 (19.9%) for HLHS and 56 (7.1%) for non-HLHS FUH.

Among children with biventricular CHD (VSD, TOF, COA, and AOS), the total overall mortality by age 5 years was 2.8% (n = 147) for VSD, 4.3% (n = 158) for TOF, 3.1% (n = 108) for COA, and 5.2% (n = 71) for AOS. Mortality related to pathway procedures was 0.5% (n = 29) for VSD, 2.1% (n = 76) for TOF, 1.1% (n = 40) for COA, and 2.9% (n = 39) for AOS. The number of patients who died unrelated to a pathway operation was 118 (2.3%), 82 (2.2%), 68 (2.0%), and 32 (2.3%), respectively.

By age 5 years, among children with complex CHDs with variable anatomy (AVSD, TGA, and PA), the total overall mortality by age 5 years was 9.6% (n = 321) for AVSD, 6.4% (n = 190) for TGA, and 19.6% (n = 257) for PA. Mortality related to pathway procedures was 3.9% (n = 133) for AVSD, 3.5% (n = 104) for TGA, and 9.7% (n = 127) for PA. The number of patients who died unrelated to a pathway operation was 188 (5.5%) for AVSD, 86 (2.9%) for TGA, and 130 (9.9%) for PA.

Table E10 presents overall and pathway procedure mortality rates by the presence of risk factors. At age 5 years, compared to the mortality of 7.7% in the whole cohort, mortality was 13.4% (n = 193) in children with preterm birth, 11.1% (n = 316) in those with a congenital comorbidity, and 7.9% (n = 184) in those with Down syndrome.

DISCUSSION

Summary of Findings

This novel population-level analysis presents the essential palliative and reparative pathway procedures

TABLE 1. Frequency and percentage of each type of pathway procedure for sentinel CHDs and subgroups

Sentinel CHDs and subgroups	Functionally univentricular pathway					Biventricular reparative pathway			
	Number of patients	Pre-pathway procedures, n (%)	Initial palliation, n (%)	Palliative stage 2, n (%)	TCPC, n (%)	Number of patients	Pre-pathway procedures, n (%)	Prior palliation, n (%)	Reparative procedures, n (%)
Total cohort (n = 28,806)	2583	314 (12.2)	2227 (86.2)	1947 (75.4)	1384 (53.6)	26,223	2943 (11.2)	2872 (11.0)	24,826 (94.7)
Functionally univentricular CHDs									
HLHS (n = 1228)	1228	122 (9.9)	1188 (96.7)	795 (64.7)	536 (43.6)	NA	NA	NA	NA
Non-HLHS functionally univentricular heart (n = 992)	992	134 (13.5)	780 (78.6)	829 (83.6)	640 (64.5)	NA	NA	NA	NA
Tricuspid atresia (n = 553)	553	84 (15.2)	434 (78.5)	442 (79.9)	346 (62.6)	NA	NA	NA	NA
Double-inlet ventricle (n = 439)	439	50 (11.4)	346 (78.8)	387 (88.2)	294 (67.0)	NA	NA	NA	NA
Biventricular CHDs									
VSD (n = 6570)	NA	NA	NA	NA	NA	6570	615 (9.4)	469 (7.1)	5860 (89.2)
Single VSD (n = 6145)	NA	NA	NA	NA	NA	6145	554 (9.0)	254 (4.1)	5551 (90.3)
Multiple VSDs (n = 425)	NA	NA	NA	NA	NA	425	61 (14.4)	215 (50.6)	309 (72.7)
TOF (n = 4568)	*	0 (0)	0 (0)	*	*	4568	115 (2.5)	819 (17.9)	4481 (98.1)
Standard TOF (n = 3858)	NA	NA	NA	NA	NA	3858	76 (2.0)	647 (16.8)	3798 (98.4)
TOF with double-outlet right ventricle (n = 527)	*	0 (0)	0 (0)	*	*	527	25 (4.7)	165 (31.3)	503 (95.4)
TOF absent pulmonary valve (n = 183)	NA	NA	NA	NA	NA	183	14 (7.7)	7 (3.8)	180 (98.4)
COA (n = 4338)	NA	NA	NA	NA	NA	4338	85 (2.0)	14 (0.3)	4282 (98.7)
Isolated COA (n = 3005)	NA	NA	NA	NA	NA	3005	73 (2.4)	0 (0)	2962 (98.6)
COA with VSD (n = 1333)	NA	NA	NA	NA	NA	1333	12 (0.9)	14 (1.1)	1320 (99.0)
AOS (n = 1624)	NA	NA	NA	NA	NA	1624	103 (6.3)	137 (8.4)	1514 (93.2)
Isolated AOS (n = 1193)	NA	NA	NA	NA	NA	1193	51 (4.3)	0 (0)	1157 (97.0)
AOS with multilevel left heart obstruction (n = 431)	NA	NA	NA	NA	NA	431	52 (12.1)	137 (31.8)	357 (82.8)
CHDs with variable anatomy									
AVSD (n = 4085)	99	*	79 (79.8)	77 (77.8)	44 (44.4)	3986	48 (1.2)	514 (12.9)	3847 (96.5)
Complete AVSD (n = 2653)	NA	NA	NA	NA	NA	2653	30 (1.1)	339 (12.8)	2565 (96.7)
Partial AVSD (n = 1006)	NA	NA	NA	NA	NA	1006	11 (1.1)	36 (3.6)	998 (99.2)
Tetralogy AVSD (n = 227)	12	0 (0)	10 (83.3)	12 (100)	*	215	*	69 (32.1)	204 (94.9)
Unbalanced AVSD (n = 199)	87	0 (0)	69 (79.3)	65 (74.7)	39 (44.8)	112	*	70 (62.5)	80 (71.4)
TGA (n = 3759)	91	*	64 (70.3)	88 (96.7)	62 (68.1)	3668	1882 (51.3)	291 (7.9)	3581 (97.6)
TGA with IVS (n = 2079)	NA	NA	NA	NA	NA	2079	1199 (57.7)	11 (0.5)	2049 (98.6)
Complex TGA without PS (n = 1257)	26	*	25 (96.2)	26 (100)	20 (76.9)	1231	493 (40.0)	100 (8.1)	1201 (97.6)
Complex TGA with PS (n = 423)	65	36 (55.4)	39 (60.0)	62 (95.4)	42 (64.6)	358	190 (53.1)	180 (50.3)	331 (92.5)
PA (n = 1642)	168	16 (9.5)	113 (67.3)	153 (91.1)	99 (58.9)	1474	97 (6.6)	631 (42.8)	1261 (85.5)
PA with VSD (n = 1131)	32	*	24 (75.0)	26 (81.2)	24 (75.0)	1099	57 (5.2)	545 (49.6)	976 (88.8)
PA IVS (n = 511)	136	14 (10.3)	89 (65.4)	127 (93.4)	75 (55.1)	375	40 (10.7)	86 (22.9)	285 (76.0)

CHD, Congenital heart disease; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *NA*, not applicable; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *IVS*, intact ventricular septum; *PS*, pulmonary stenosis; *PA*, pulmonary atresia. *Sample size 1-5 (number suppressed).

undertaken for children with 9 sentinel CHDs and their subgroups. While most children starting CHD treatment completed their "pathway" with either a biventricular repair (86.2%) or TCPC (4.8%), the important minority who did not do so were more likely to have FUH CHD, particularly HLHS, or to be more complex children who required palliation before biventricular repair. Children with more complex CHDs requiring FUH palliation and those needing palliation before a repair can experience poorer outcomes, emphasizing the potential value in reporting these pathways. Nearly one-half of the deaths (963 of 1794) that occurred in children who had follow-up to age 5 years were unrelated to pathway operations, and for each sentinel CHD, there were deaths in children who did not complete their pathway, reinforcing the importance of evaluating longer-term mortality to judge service performance alongside short-term surgical metrics.

Context

Most multi-institutional registries report frequency and outcomes for individual operations,[1,2] which, although very important, cannot capture the complete picture. A systematic review and meta-analysis of population-based studies of long-term survival with CHD found only 16 studies worldwide, mainly from an earlier era, with a pooled 10-year survival rate of 81.4%.[14] Patients in our study benefited from improvements in care pathways given their later years of birth when short term-outcomes had improved.[4,15,16] Recognizing the importance of longer-term outcomes, selected population-based (CH STRONG[17]) and registry-based (Australia, New Zealand Fontan Registry[18] and German Registry[19]) analyses have begun to address this important topic, but to date none have adopted our detailed pathway analysis approach.

Strengths and Limitations

This was a registry-based study reflecting practice and outcomes in England and Wales. The study is limited by data quality; however, NCHDA data are of a very high standard. Children with CHD who had no procedures were not captured. There is no perfect method for categorizing CHD types, given the heterogeneity of CHD. Because our focus was on metric development for routine monitoring, we excluded CHD types with small numbers of patients, including some high-impact CHDs. The involvement of experienced clinicians in defining CHD types and developing data quality thresholds was a strength. Because this was a patient-level analysis spanning a series of procedures that vary widely, we did not consider procedure-level risk factors. We divided our data into 2 eras, finding small improvements in outcomes in the later era, and note that this complex topic could not be covered in detail. We performed descriptive analyses and did not undertake the vital next steps of evaluating care.

Future Directions

Procedure-based registry data can be used to evaluate the outcome metrics of pathway completion in terms of TCPC in children on an FUH pathway and repair in children on a biventricular pathway, as well as longer-term mortality at age 5 years. These important outcomes are more complex than procedure-level metrics. Their development for future use in quality assurance will require further work, but they could provide an overall view of service performance and be of interest to patients' families.

Conflict of Interest Statement

Drs Tsang and Brown reported support from the National Institute for Health and Care Research's Biomedical Research Centre at Great Ormond Street Hospital. All other authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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Key Words: congenital heart disease, outcomes, cardiac surgery

APPENDIX E1

Supplemental materials for the manuscript: Evaluating long-term surgical pathways reported to national audit for children undergoing treatment for congenital heart disease in England and Wales.

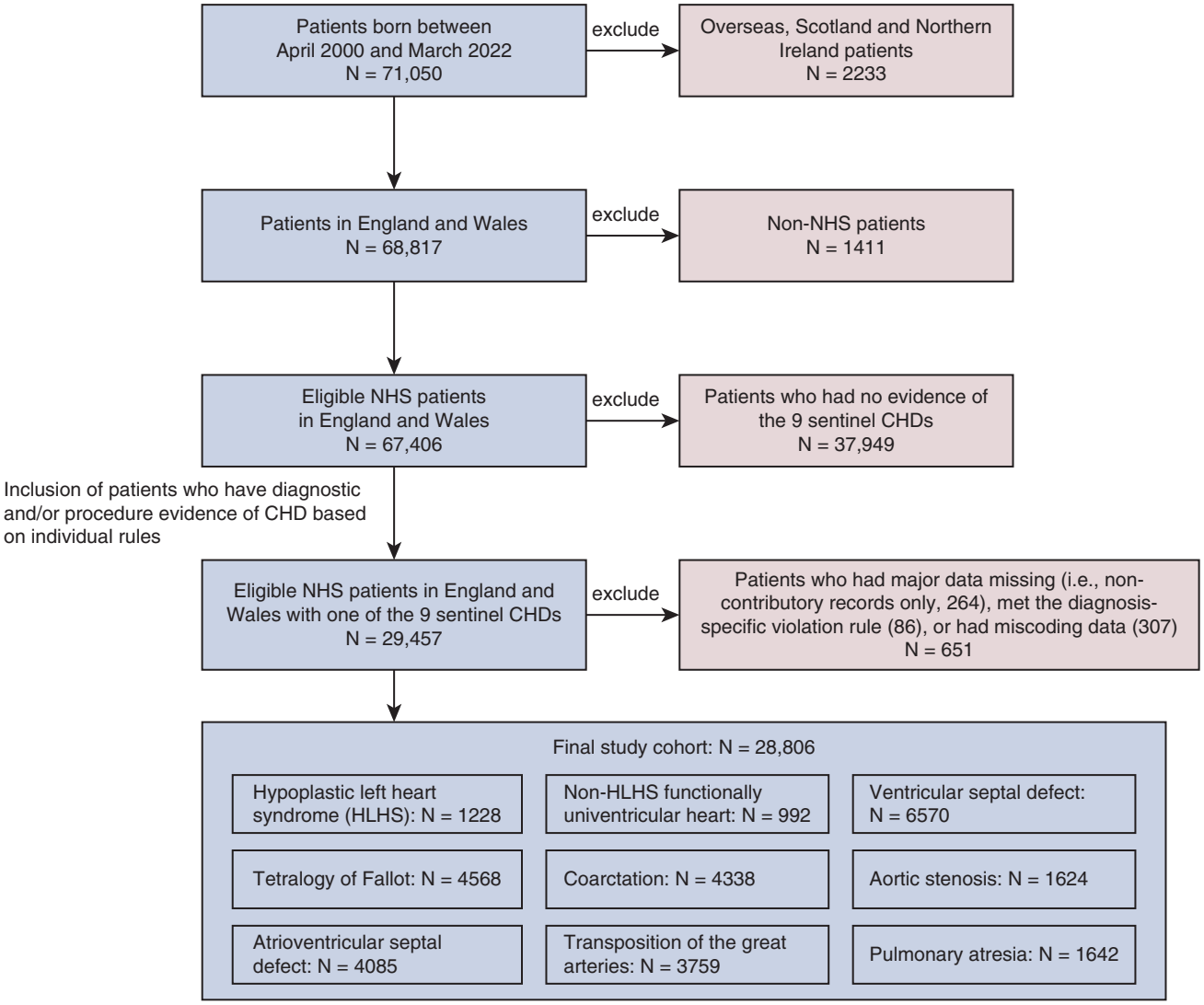

FIGURE E1. Patient inclusion and exclusion flow chart. *NHS*, National Health Service; *CHD*, congenital heart disease.

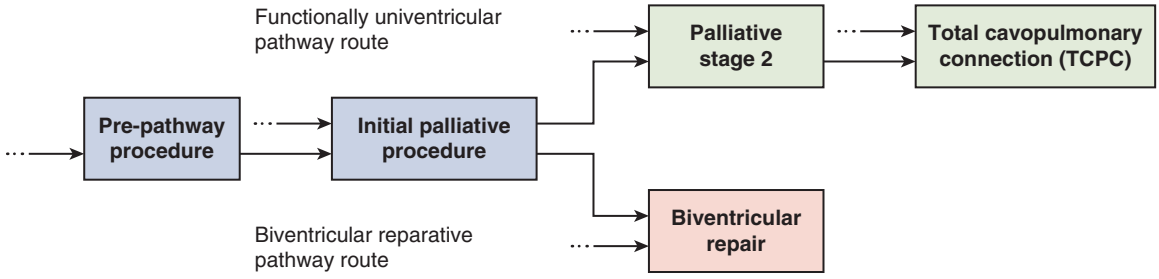

FIGURE E2. Flowchart of typical treatment pathways. Patients with functionally univentricular congenital heart defects (*CHD*) were expected to undergo a palliative pathway. Patients with selected biventricular CHDs were expected to undergo a reparative pathway. For CHDs with a range of anatomy, the treatment pathway may be either biventricular repair or a functionally univentricular pathway. For certain sentinel CHDs, patients may skip some pathway procedures.

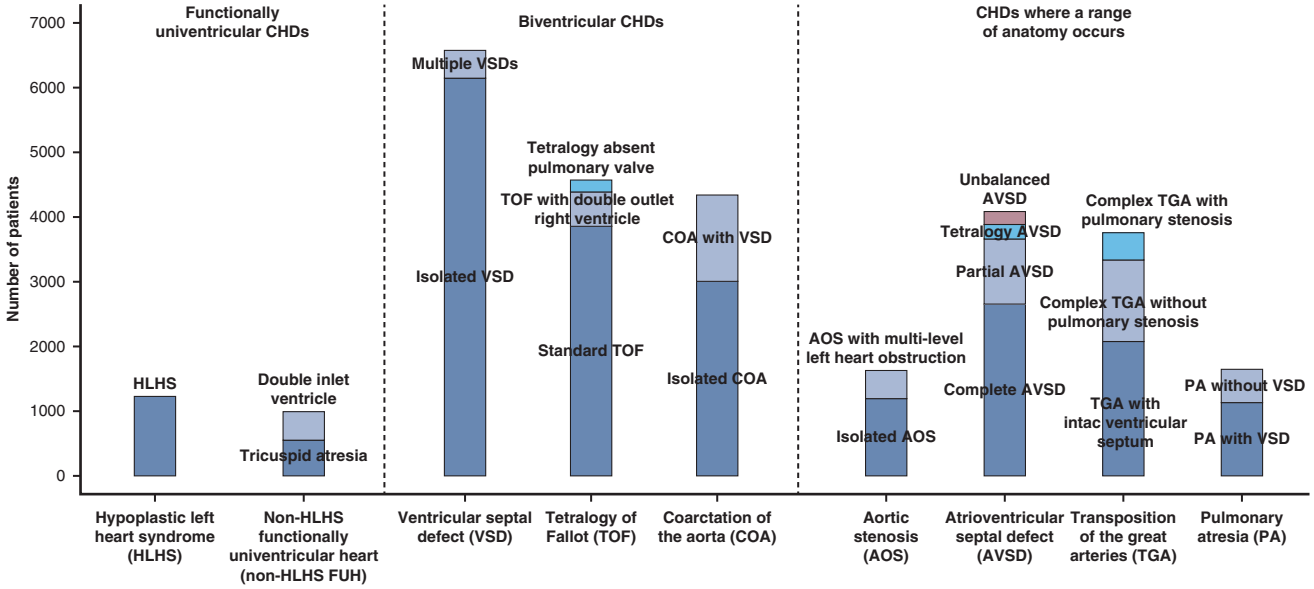

FIGURE E3. Breakdown of sentinel congenital heart defects by subgroups. *CHD*, Congenital heart disease.

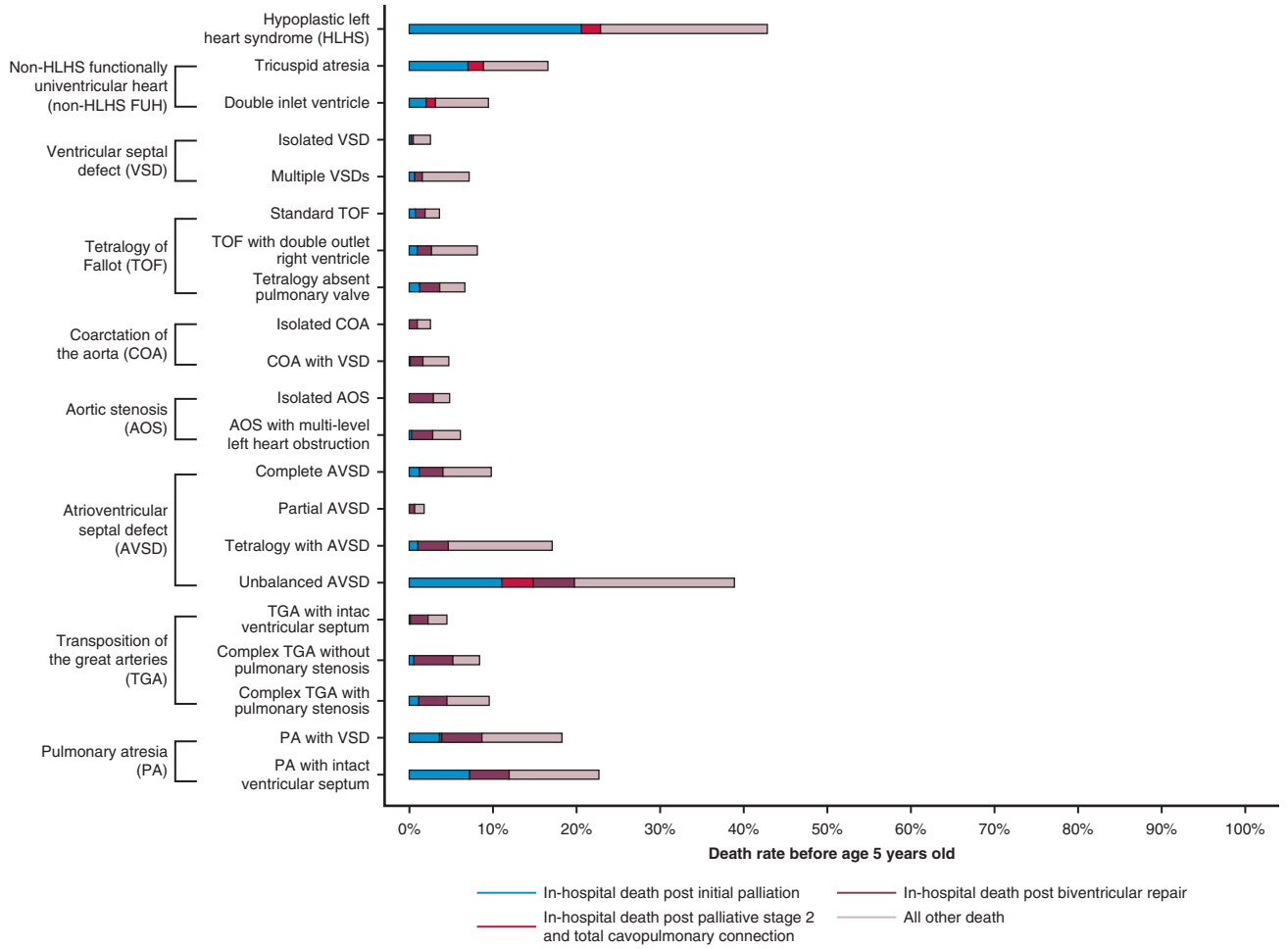

FIGURE E4. Death before age 5 years depicted separately for post-pathway procedure mortalities and other mortalities (patients who completed 5 years of follow-up in terms of procedure history only, born between April 2000 and March 2017).

TABLE E1. Pathway procedure types (and subtypes) for each sentinel CHD

Sentinel CHD	Pre-pathway procedures	Initial palliation procedures	Palliative stage 2	TCPC	Reparative procedure for biventricular pathway
Functionally univentricular CHDs					
HLHS	Application of right and left pulmonary arterial bands Pulmonary trunk band Atrial septectomy	Norwood type operation Hybrid procedure	Glenn Comprehensive stage two	TCPC	NA
Non-HLHS FUH	Balloon atrial septostomy by pull-back (Rashkind) Transluminal occlusion of systemic-to-pulmonary collateral artery(ies) with coil device Atrial septectomy	Procedures to secure pulmonary blood flow, including systemic-to-pulmonary arterial shunts and ductal stenting Pulmonary arterial trunk banding Isolated aortic arch repairs Norwood type operation Hybrid procedure	Glenn Comprehensive stage two	TCPC	NA
Biventricular CHDs					
VSD	Patent ductus arteriosus closure by surgery or transluminal device Atrial septal defect closure with transluminal device Vascular ring procedure	Pulmonary arterial trunk banding	NA	NA	VSD closure and multiple VSD closure
TOF	Balloon dilation of right ventricular outflow tract Transluminal occlusion of systemic-to-pulmonary collateral artery(ies) with coil device Patent arterial duct closure by surgery	Procedures to secure pulmonary blood flow, including systemic to pulmonary arterial shunts, right ventricular outflow tract stenting and ductal stenting.	NA	NA	TOF repair Other reparative procedures for TOF, eg, primary right ventricle-to-pulmonary artery conduit with VSD closure, Fallot and absent pulmonary valve repair, and primary isolated VSD repair ("pink TOF").
COA, isolated with or without VSD	Patent ductus arteriosus closure by surgery or transluminal device	Pulmonary arterial banding	NA	NA	Coarctation surgical repair Coarctation repair by catheterization Coarctation repair with VSD repair Coarctation repair with VSD repair and debanding pulmonary artery Coarctation repair with pulmonary trunk band
AOS with or without multiple-level left heart obstructions	Subaortic fibromuscular shelf resection in the presence of AOS Patent arterial duct closure by surgery or transluminal device.	Isolated arch repairs (coarctation only) in presence of AOS	NA	NA	Balloon dilation of aortic valve by catheterization Surgical relief of aortic stenosis Complex and other types of reparative procedure in aortic stenosis, eg, Ross operation Aortic valve replacement

(Continued)

TABLE E1. Continued

Sentinel CHD	Pre-pathway procedures	Initial palliation procedures	Palliative stage 2	TCPC	Reparative procedure for biventricular pathway
CHDs in which a range of anatomy occurs					
AVSD	Patent arterial duct closure by surgery or transluminal device	Pulmonary arterial banding Isolated arch repairs Procedures to secure pulmonary blood flow, including systemic-to-pulmonary arterial shuntsand ductal stenting Norwood type operation Hybrid procedure	Glenn Comprehensive stage two	Fontan	Complete AVSD repair Partial AVSD repair Other reparative procedure linked to AVSD, eg, common atrioventricular valve repair Tetralogy AVSD repair Atrial septal defect or VSD repair in AVSD
TGA	Balloon atrial septostomy by pull-back (Rashkind) Interatrial communication creation/enlargement Patent ductus arteriosus closure by surgery	Procedures to secure pulmonary blood flow, including systemic-to-pulmonary arterial shunts and ductal stenting Pulmonary arterial trunk banding Isolated arch repairs Norwood type operation Hybrid procedure	Glenn	Fontan	TGA with IVS repair, ie, arterial switch operation Complex TGA without PS repairs Complex TGA with PS repair, eg, Rastelli Senning or Mustard operation (very rare for complex TGA)
PA	Balloon atrial septostomy by pull-back (Rashkind) Balloon dilation of pulmonary artery	Procedures to secure pulmonary blood flow, including systemic-to-pulmonary arterial shunt and ductal stenting	Glenn (in isolation) Glenn with reparative procedure, ie, 1.5- ventricle repair	Fontan	Surgery involving only the pulmonary valve and/or pulmonary arteries PA or PA with VSD full surgical repair Catheter interventions to open the pulmonary valve and arteries Other reparative procedures for PA, eg, unifocalization of aortopulmonary collateral arteries Fallot type repair in setting of PA Reparative procedure for PA with incomplete coding Atrial septal defect or VSD repair in setting of PA

Detailed rules and R code for assigning the treatment pathways to individual patients are available at https://github.com/UCL-CORU/CHD-research-code/tree/main (in the "06 Pathways of care for specific diagnoses" dropdown list). *CHD*, Congenital heart disease; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *NA*, not applicable; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *PA*, pulmonary atresia.

TABLE E2. Patient characteristics

Sentinel CHDs and subgroups	Number of patients	Male sex, n (%)	Preterm birth (before 37 wk), n (%)	Congenital noncardiac comorbidity, n (%)	Down syndrome, n (%)	Recent era (born from April 2010 onward), n (%)
Total cohort	28,806	16,553 (57.5)	2218 (7.7)	3890 (13.5)	3066 (10.6)	15,583 (54.1)
Functionally univentricular CHDs						
HLHS	1228	791 (64.4)	52 (4.2)	170 (13.8)	*	630 (51.3)
Non-HLHS FUH	992	589 (59.4)	78 (7.9)	144 (14.5)	*	554 (55.8)
Tricuspid atresia	553	324 (58.6)	40 (7.2)	87 (15.7)	*	309 (55.9)
Double-inlet ventricle	439	265 (60.4)	38 (8.7)	57 (13.0)	0 (0)	245 (55.8)
Biventricular CHDs						
VSD	6570	3385 (51.5)	656 (10.0)	820 (12.5)	870 (13.2)	3691 (56.2)
Single VSD	6145	3204 (52.1)	591 (9.6)	729 (11.9)	852 (13.9)	3456 (56.2)
Multiple VSDs	425	181 (42.6)	65 (15.3)	91 (21.4)	18 (4.2)	235 (55.3)
TOF	4568	2637 (57.7)	396 (8.7)	876 (19.2)	168 (3.7)	2523 (55.2)
Standard TOF	3858	2239 (58.0)	300 (7.8)	643 (16.7)	144 (3.7)	2092 (54.2)
TOF with DORV	527	306 (58.1)	82 (15.6)	159 (30.2)	24 (4.6)	345 (65.5)
TOF absent pulmonary valve	183	92 (50.3)	14 (7.7)	74 (40.4)	0 (0)	86 (47.0)
COA	4338	2656 (61.2)	315 (7.3)	490 (11.3)	37 (0.9)	2349 (54.1)
Isolated COA	3005	1917 (63.8)	203 (6.8)	304 (10.1)	21 (0.7)	1518 (50.5)
COA with VSD	1333	739 (55.4)	112 (8.4)	186 (14.0)	16 (1.2)	831 (62.3)
AOS	1624	1158 (71.3)	92 (5.7)	139 (8.6)	*	776 (47.8)
Isolated AOS	1193	872 (73.1)	59 (4.9)	76 (6.4)	*	562 (47.1)
AOS with multilevel left heart obstruction	431	286 (66.4)	33 (7.7)	63 (14.6)	0 (0)	214 (49.7)
CHDs with variable anatomy						
AVSD	4093	1869 (45.7)	298 (7.3)	605 (14.8)	1973 (48.2)	2142 (52.4)
Complete AVSD	2653	1193 (45.0)	220 (8.3)	388 (14.6)	1565 (58.9)	1473 (55.5)
Partial AVSD	1006	478 (47.5)	35 (3.5)	116 (11.5)	226 (22.4)	447 (44.4)
Tetralogy AVSD	227	109 (48.0)	20 (8.8)	52 (22.9)	129 (56.8)	113 (49.8)
Unbalanced AVSD	199	85 (42.7)	23 (11.6)	47 (23.6)	53 (26.6)	109 (54.8)
TGA	3759	2583 (68.7)	156 (4.2)	198 (5.3)	*	2074 (55.2)
TGA with IVS	2079	1473 (70.9)	80 (3.8)	59 (2.8)	0 (0)	1103 (53.1)
Complex TGA without PS	1257	834 (66.3)	44 (3.5)	88 (7.0)	*	739 (58.8)
Complex TGA with PS	423	276 (65.2)	32 (7.6)	51 (12.1)	0 (0)	232 (54.8)
PA	1642	889 (54.1)	175 (10.7)	450 (27.4)	13 (0.8)	844 (51.4)
PA with VSD	1131	608 (53.8)	140 (12.4)	394 (34.8)	13 (1.1)	572 (50.6)
PA with IVS	511	281 (55.0)	35 (6.8)	56 (11.0)	0 (0)	272 (53.2)

CHD, Congenital heart disease; *HLHS*, hypoplastic left heart syndrome; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *DORV*, double-outlet right ventricle; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *IVS*, intact ventricular septum; *PS*, pulmonary stenosis; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E3. Age at procedure for patients managed with the functionally univentricular pathway

Sentinel CHDs and subgroups	Number of patients	Initial palliative procedure		Palliative stage two		TCPC	
		n (%)	Age, d, median (IQR)	n (%)	Age, d, median (IQR)	n (%)	Age, y, median (IQR)
Functionally univentricular CHDs							
HLHS	1228	1188 (96.7)	4 (3-7)	795 (64.7)	157 (124-196)	536 (43.6)	4.2 (3.5-5.0)
Non-HLHS FUH	992	780 (78.6)	13 (6-40)	829 (83.6)	241 (165-411)	640 (64.5)	4.6 (3.8-5.8)
Tricuspid atresia	553	434 (78.5)	17 (6-42)	442 (79.9)	236 (161-380)	346 (62.6)	4.7 (3.9-5.8)
Double-inlet ventricle	439	346 (78.8)	10 (5-39)	387 (88.2)	250 (168-430)	294 (67.0)	4.6 (3.6-5.8)
CHDs in which a range of anatomy occurs							
AVSD	99	79 (79.0)	12 (6-48)	77 (78.0)	280 (180-552)	44 (44.0)	5.1 (4.0-5.6)
Tetralogy with AVSD	12	10 (83.3)	63 (27-93)	12 (100)	528 (312-844)	*	*
Unbalanced AVSD	87	69 (78.4)	10 (6-34)	65 (75.0)	250 (165-454)	39 (44.3)	4.8 (3.7-5.4)
TGA	91	64 (70.3)	18 (7-62)	88 (96.7)	420 (241-788)	62 (68.1)	5.0 (4.0-6.4)
Complex TGA without PS	26	25 (96.2)	14 (7-37)	26 (100)	426 (204-836)	20 (76.9)	5.9 (4.1-7.4)
Complex TGA with PS	65	39 (60.0)	23 (8-72)	62 (95.4)	400 (252-689)	42 (64.6)	4.7 (4.0-5.7)
PA	168	112 (66.7)	6 (4-10)	153 (91.1)	267 (176-411)	99 (58.9)	4.5 (3.4-5.5)
PA with VSD	32	24 (75.0)	6 (4-8)	26 (81.2)	264 (178-401)	24 (75.0)	4.4 (3.0-5.3)
PA without VSD	136	88 (64.7)	6 (4-10)	127 (93.4)	267 (176-408)	75 (55.1)	4.7 (3.5-5.6)

This table presents the data shown in Figure 1. *CHD*, Congenital heart disease; *TCPC*, total cavopulmonary connection; *IQR*, interquartile range; *HLHS*, hypoplastic left heart syndrome; *FUH*, functionally univentricular heart; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *PS*, pulmonary stenosis; *PA*, pulmonary atresia.
*Results not shown (sample size 1 to 5).

TABLE E4. Age at procedure for patients managed with a biventricular reparative pathway

Sentinel CHDs and subgroups	Patients with initial palliative procedure				Patients without initial palliative procedure		
			Reparative procedure			Reparative procedure	
	Number of patients	Age at initial palliative procedure, d, median (IQR)	n (%)	Age, d, median (IQR)	Number of patients	n (%)	Age, d, median (IQR)
Biventricular CHDs							
VSD	469	73 (46-105)	320 (68.2)	459 (273-960)	6101	5540 (90.8)	160 (104-313)
Single VSD	254	66 (44-96)	169 (66.5)	332 (217-604)	5891	5382 (91.4)	161 (104-318)
Multiple VSDs	215	80 (48-113)	151 (70.2)	687 (376-1200)	210	158 (75.2)	144 (101-233)
TOF	819	42 (15-89)	743 (90.7)	367 (210-550)	3749	3738 (99.7)	225 (153-332)
Standard TOF	647	42 (16-89)	592 (91.5)	364 (204-565)	3211	3206 (99.8)	226 (157-328)
TOF with DORV	165	36 (13-91)	146 (88.5)	374 (234-506)	362	357 (98.6)	220 (145-352)
TOF absent pulmonary valve	7	12 (8-13)	*	*	176	175 (99.4)	188 (70-442)
COA	14	52 (33-82)	8 (57.1)	147 (56-223)	4324	4274 (98.8)	20 (8-106)
Isolated COA	0	NA	0	NA	3005	2962 (98.6)	33 (9-240)
COA with VSD	14	52 (33-82)	8 (57.1)	147 (56-223)	1319	1312 (99.5)	12 (7-40)
AOS	137	13 (6-119)	80 (58.4)	738 (154-2899)	1487	1434 (96.4)	131 (23-1867)
Isolated AOS	0	NA	0	NA	1193	1157 (97.0)	143 (30-1923)
AOS with multilevel left heart obstruction	137	13 (6-119)	80 (58.4)	738 (154-2899)	294	277 (94.2)	66 (8-1628)
CHDs in which a range of anatomy occurs							
AVSD	514	42 (12-86)	394 (76.7)	296 (173-518)	3472	3453 (99.5)	180 (119-583)
Complete AVSD	339	45 (15-85)	262 (77.3)	258 (158-415)	2317	2304 (99.4)	145 (106-202)
Partial AVSD	36	15 (7-90)	33 (91.7)	408 (156-1102)	971	966 (99.5)	981 (470-1528)
Tetralogy AVSD	69	64 (24-112)	59 (85.5)	367 (280-672)	146	145 (99.3)	209 (116-412)
Unbalanced AVSD	70	21 (7-60)	40 (57.1)	384 (174-584)	42	40 (95.2)	144 (86-195)
TGA	291	24 (10-56)	247 (84.9)	302 (106-569)	3377	3334 (98.7)	10 (7-16)
TGA with IVS	11	48 (42-71)	8 (72.7)	52 (45-72)	2068	2041 (98.7)	9 (6-13)
Complex TGA without PS	100	14 (7-32)	83 (83.0)	165 (52-315)	1131	1118 (98.9)	11 (7-22)
Complex TGA with PS	180	40 (13-66)	156 (86.7)	405 (219-647)	178	175 (98.3)	36 (12-352)
PA	628	12 (5-48)	461 (73.4)	364 (213-641)	846	802 (94.8)	31 (6-203)
PA with VSD	543	15 (6-54)	441 (81.2)	370 (234-651)	556	535 (96.2)	90 (21-280)
PA with IVS	85	7 (4-18)	20 (23.5)	42 (16-132)	290	267 (92.1)	4 (2-9)

This table presents the data shown in Figure 2. *CHD*, Congenital heart disease; *IQR*, interquartile range; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *DORV*, double-outlet right ventricle; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *IVS*, intact ventricular septum; *PS*, pulmonary stenosis; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E5. Proportion of patients by the type of pathway procedures undertaken for each sentinel CHD subtype

Sentinel CHDs and subgroups	Patients with pre-pathway procedure only, n (%)	Patients with initial palliative procedure only, n (%)	Patients with palliative stage 2 and/or TCPC, n (%)	Patients with initial palliation and reparative procedure, n (%)	Patients with single-stage definitive repair, n (%)
Whole cohort (n = 28,806)	1794 (7.7)	409 (1.8)	43 (0.2)	379 (1.6)	963 (4.1)
Functionally univentricular CHDs					
HLHS (n = 1228)	436 (42.8)	210 (20.6)	23 (2.3)	NA	203 (19.9)
Non-HLHS FUH (n = 992)	106 (13.4)	38 (4.8)	12 (1.5)	NA	56 (7.1)
Tricuspid atresia (n = 553)	73 (16.6)	31 (7.0)	8 (1.8)	NA	34 (7.7)
Double-inlet ventricle (n = 439)	33 (9.5)	7 (2.0)	4 (1.1)	NA	22 (6.3)
Biventricular CHDs					
VSD (n = 6570)	147 (2.8)	13 (0.2)	NA	16 (0.3)	118 (2.3)
Single VSD (n = 6145)	124 (2.5)	11 (0.2)	NA	13 (0.3)	100 (2.0)
Multiple VSDs (n = 425)	23 (7.1)	2 (0.6)	NA	3 (0.9)	18 (5.6)
TOF (n = 4568)	158 (4.3)	28 (0.8)	NA	48 (1.3)	82 (2.2)
Standard TOF (n = 3858)	113 (3.6)	22 (0.7)	NA	37 (1.2)	54 (1.7)
TOF with DORV (n = 527)	34 (8.2)	4 (1.0)	NA	7 (1.7)	23 (5.5)
TOF absent pulmonary valve (n = 183)	11 (6.7)	2 (1.2)	NA	4 (2.4)	5 (3.0)
COA (n = 4338)	108 (3.1)	1 (0.0)	NA	39 (1.1)	68 (2.0)
Isolated COA (n = 3005)	62 (2.5)	0 (0)	NA	24 (1.0)	38 (1.5)
COA with VSD (n = 1333)	46 (4.7)	1 (0.1)	NA	15 (1.5)	30 (3.1)
AOS (n = 1624)	71 (5.2)	1 (0.1)	NA	38 (2.8)	32 (2.3)
Isolated AOS (n = 1193)	49 (4.8)	0 (0)	NA	29 (2.9)	20 (2.0)
AOS with multilevel left heart obstruction (n = 431)	22 (6.1)	1 (0.3)	NA	9 (2.5)	12 (3.3)
CHDs in which a range of anatomy occurs					
AVSD (n = 4085)	321 (9.6)	45 (1.3)	6 (0.2)	82 (2.4)	188 (5.6)
Complete AVSD (n = 2653)	209 (9.9)	25 (1.2)	NA	61 (2.9)	123 (5.8)
Partial AVSD (n = 1006)	16 (1.8)	0 (0)	NA	6 (0.7)	10 (1.1)
Tetralogy AVSD (n = 227)	33 (17.1)	2 (1.0)	0 (0)	7 (3.6)	24 (12.4)
Unbalanced AVSD (n = 199)	63 (39.5)	18 (11.1)	6 (3.7)	8 (4.9)	31 (19.1)
TGA (n = 3759)	190 (6.4)	11 (0.4)	0 (0)	93 (3.1)	86 (2.9)
TGA with IVS (n = 2079)	74 (4.5)	2 (0.1)	0 (0)	35 (2.1)	37 (2.3)
Complex TGA without PS (n = 1257)	82 (8.4)	5 (0.5)	0 (0)	46 (4.7)	31 (3.2)
Complex TGA with PS (n = 423)	34 (9.6)	4 (1.1)	0 (0)	12 (3.4)	18 (5.1)
PA (n = 1642)	257 (19.6)	62 (4.7)	*	63 (4.8)	130 (9.9)
PA with VSD (n = 1131)	166 (18.3)	33 (3.6)	*	44 (4.8)	87 (9.6)
PA with IVS (n = 511)	91 (22.7)	29 (7.2)	0 (0)	19 (4.7)	43 (10.7)

This table presents the data shown in Figure 3. *CHD*, Congenital heart disease; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *NA*, not applicable; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *DORV*, double-outlet right ventricle; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *IVS*, intact ventricular septum; *PS*, pulmonary stenosis; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E6. The undertaking of initial palliative procedures and postoperative death by risk factor

	Patients with an initial palliative procedure, n (%)					In-hospital deaths after initial palliative procedure, n (%)				
	All patients	Patients born preterm (before 37 wk)	Patients with congenital noncardiac comorbidity	Patients with Down syndrome	Patients born from April 2010 onward	All patients	Patients born preterm (before 37 wk)	Patients with congenital noncardiac comorbidity	Patients with Down syndrome	Patients born from April 2010 onward
Total cohort	**5099 (17.7)**	**571 (25.7)**	**1102 (28.3)**	**277 (10.9)**	**2706 (17.4)**	**477 (9.4)**	**56 (9.8)**	**81 (7.4)**	**18 (6.5)**	**229 (8.5)**
By sentinel CHDs										
Functionally univentricular CHDs										
HLHS	1188 (96.7)	46 (88.5)	166 (97.6)	*	610 (96.8)	244 (20.5)	15 (32.6)	26 (15.7)	*	110 (18.0)
Non-HLHS FUH	780 (78.6)	67 (85.9)	120 (83.3)	*	451 (81.4)	42 (5.4)	6 (9.0)	9 (7.5)	*	25 (5.5)
Biventricular CHDs										
VSD	469 (7.1)	120 (18.3)	137 (16.7)	23 (3.1)	261 (7.1)	14 (3.0)	6 (5.0)	6 (4.4)	*	8 (3.1)
TOF	819 (17.9)	120 (30.3)	233 (26.6)	27 (19.3)	427 (16.9)	33 (4.0)	9 (7.5)	6 (2.6)	*	15 (3.5)
COA	14 (0.3)	*	*	0 (0)	7 (0.3)	*	*	*	0 (0)	*
AOS	137 (8.4)	*	22 (15.8)	0 (0)	69 (8.9)	*	*	0 (0)	0 (0)	0 (0)
CHDs in which a range of anatomy occurs										
AVSD	593 (14.5)	96 (32.2)	151 (25.0)	216 (13.4)	348 (16.2)	59 (9.9)	8 (8.3)	18 (11.9)	13 (6.0)	35 (10.1)
TGA	355 (9.4)	33 (21.2)	52 (26.3)	*	174 (8.4)	16 (4.5)	*	*	0 (0)	7 (4.0)
PA	744 (45.3)	81 (46.3)	217 (48.2)	9 (75.0)	359 (42.5)	66 (8.9)	7 (8.6)	13 (6.0)	*	28 (7.8)

Only broader CHD diagnoses are presented owing to the limited sample size for breakdown by diagnosis subgroup. *CHD*, Congenital heart disease; *HLHS*, hypoplastic left heart syndrome; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E7. The undertaking of reparative procedures in biventricular CHDs and postoperative death by risk factor group

	Patients undergoing a reparative procedure, n (%)					In-hospital deaths after reparative procedures, n (%)				
Cohort managed under biventricular reparative pathway	**All patients**	**Patients born preterm (before 37 wk)**	**Patients with congenital noncardiac comorbidity**	**Patients with Down syndrome**	**Patients born from April 2010 onward**	**All patients**	**Patients born preterm (before 37 wk)**	**Patients with congenital noncardiac comorbidity**	**Patients with Down syndrome**	**Patients born from April 2010 onward**
	24,826 (94.7)	**1791 (87.1)**	**3200 (91.4)**	**2438 (96.4)**	**13,428 (94.5)**	**457 (1.8)**	**63 (3.5)**	**81 (2.5)**	**49 (2.0)**	**220 (1.6)**
By sentinel CHDs										
Biventricular CHDs										
VSD	5860 (89.2)	493 (75.2)	662 (80.7)	703 (95.0)	3273 (88.7)	20 (0.3)	*	*	*	7 (0.2)
TOF	4481 (98.2)	377 (95.2)	847 (96.9)	135 (96.4)	2477 (98.3)	59 (1.3)	19 (5.0)	24 (2.8)	*	29 (1.2)
COA	4282 (98.7)	306 (97.1)	480 (98.0)	30 (96.8)	2319 (98.7)	45 (1.1)	6 (2.0)	13 (2.7)	*	18 (0.8)
AOS	1514 (93.2)	80 (87.0)	123 (88.5)	0 (0)	717 (92.4)	43 (2.8)	*	*	0 (0)	15 (2.1)
CHDs in which a range of anatomy occurs										
AVSD	3847 (96.5)	261 (90.6)	537 (93.2)	1559 (97.3)	2013 (96.3)	97 (2.5)	7 (2.7)	16 (3.0)	41 (2.6)	45 (2.2)
TGA	3581 (97.6)	137 (92.6)	172 (95.6)	*	1976 (97.6)	122 (3.4)	9 (6.6)	6 (3.5)	*	70 (3.5)
PA	1261 (85.5)	137 (85.1)	379 (89.6)	10 (83.3)	653 (86.1)	71 (5.6)	13 (9.5)	15 (4.0)	*	36 (5.5)

Only broader CHD diagnoses are presented owing to the limited sample size for breakdown by diagnosis subgroup. *CHD*, Congenital heart disease; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E8. Postoperative in-hospital deaths for the pathway procedures by sentinel CHDs and subgroups

Sentinel CHDs and subgroups	Initial palliative procedure		Functionally univentricular pathway: palliative stage 2		Functionally univentricular pathway: TCPC		Biventricular reparative procedure	
	Number of patients	In-hospital deaths, n (%)	Number of patients	In-hospital deaths, n (%)	Number of patients	In-hospital deaths, n (%)	Number of patients	In-hospital deaths, n (%)
Total cohort	5102	479 (9.4)	1948	46 (2.4)	1384	16 (1.2)	24,909	460 (1.8)
Functionally univentricular CHDs								
HLHS	1188	244 (20.5)	795	26 (3.3)	536	6 (1.1)	NA	NA
Non-HLHS FUH	780	42 (5.4)	829	14 (1.7)	640	*	NA	NA
Tricuspid atresia	434	32 (7.4)	442	10 (2.3)	346	*	NA	NA
Double-inlet ventricle	346	10 (2.9)	387	*	294	*	NA	NA
Biventricular CHDs								
VSD	469	14 (3.0)	NA	NA	NA	NA	5860	20 (0.3)
Single VSD	254	12 (4.7)	NA	NA	NA	NA	5551	15 (0.3)
Multiple VSDs	215	*	NA	NA	NA	NA	309	*
TOF	819	33 (4.0)	*	0 (0)	*	0 (0)	4481	59 (1.3)
Standard TOF	647	25 (3.9)	NA	NA	NA	NA	3798	44 (1.2)
TOF with DORV	165	6 (3.6)	*	0 (0)	*	0 (0)	503	9 (1.8)
TOF absent pulmonary valve	7	*	NA	NA	NA	NA	180	6 (3.3)
COA	14	*	NA	NA	NA	NA	4282	45 (1.1)
Isolated COA	0	NA	NA	NA	NA	NA	2962	26 (0.9)
COA with VSD	14	*	NA	NA	NA	NA	1320	19 (1.4)
AOS	137	*	NA	NA	NA	NA	1514	43 (2.8)
Isolated AOS	0	NA	NA	NA	NA	NA	1157	32 (2.8)
AOS with multilevel left heart obstruction	137	*	NA	NA	NA	NA	357	11 (3.1)
CHDs with variable anatomy								
AVSD	593	59 (9.9)	77	*	44	*	3859	99 (2.6)
Complete AVSD	339	32 (9.4)	NA	NA	NA	NA	2565	74 (2.9)
Partial AVSD	36	0 (0)	NA	NA	NA	NA	998	6 (0.6)
Tetralogy AVSD	79	*	12	0 (0)	*	*	204	8 (3.9)
Unbalanced AVSD	139	25 (18.1)	65	*	39	*	92	11 (12.0)
TGA	355	16 (4.5)	88	0 (0)	62	0 (0)	3583	122 (3.4)
TGA with IVS	11	*	0	NA	0	NA	2049	42 (2.0)
Complex TGA without PS	125	7 (5.6)	26	0 (0)	20	0 (0)	1201	67 (5.6)
Complex TGA with PS	219	6 (2.7)	62	0 (0)	42	0 (0)	333	13 (3.9)
PA	744	66 (8.9)	153	*	99	*	1327	71 (5.4)
PA with VSD	569	34 (6.0)	26	*	24	*	989	50 (5.1)
PA with IVS	175	32 (18.3)	127	0 (0)	75	*	338	21 (6.2)

CHD, Congenital heart disease; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *NA*, not applicable; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *DORV*, double-outlet right ventricle; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *IVS*, intact ventricular septum; *PS*, pulmonary stenosis; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E9. Post-pathway procedure mortality percentage and overall mortality percentage at age 5 years in each sentinel CHD (patients who completed 5 years of follow-up in terms of procedure history, born between April 2000 and March 2017)

Sentinel CHDs and subgroups	Total deaths before age 5 y, n (%)	In-hospital deaths after initial palliative procedure, n (%)	In-hospital deaths post-stage 2 or TCPC, n (%)	In-hospital deaths post- biventricular reparative procedure, n (%)	All other deaths, n (%)
Total cohort (born April 2000-March 2017) (n = 23,239)	1794 (7.7)	409 (1.8)	43 (0.2)	379 (1.6)	963 (4.1)
Functionally univentricular CHDs (born before March 2017)					
HLHS (n = 1018)	436 (42.8)	210 (20.6)	23 (2.3)	NA	203 (19.9)
Non-HLHS FUH (n = 789)	106 (13.4)	38 (4.8)	12 (1.5)	NA	56 (7.1)
Tricuspid atresia (n = 440)	73 (16.6)	31 (7.0)	8 (1.8)	NA	34 (7.7)
Double-inlet ventricle (n = 349)	33 (9.5)	7 (2.0)	4 (1.1)	NA	22 (6.3)
Biventricular CHDs (born before March 2017)					
VSD (n = 5241)	147 (2.8)	13 (0.2)	NA	16 (0.3)	118 (2.3)
Single VSD (n = 4919)	124 (2.5)	11 (0.2)	NA	13 (0.3)	100 (2.0)
Multiple VSDs (n = 322)	23 (7.1)	2 (0.6)	NA	3 (0.9)	18 (5.6)
TOF (n = 3701)	158 (4.3)	28 (0.8)	NA	48 (1.3)	82 (2.2)
Standard TOF (n = 3120)	113 (3.6)	22 (0.7)	NA	37 (1.2)	54 (1.7)
TOF with DORV (n = 416)	34 (8.2)	4 (1.0)	NA	7 (1.7)	23 (5.5)
TOF absent pulmonary valve (n = 165)	11 (6.7)	2 (1.2)	NA	4 (2.4)	5 (3.0)
COA (n = 3445)	108 (3.1)	1 (0.0)	NA	39 (1.1)	68 (2.0)
Isolated COA (n = 2446)	62 (2.5)	0 (0)	NA	24 (1.0)	38 (1.5)
COA with VSD (n = 979)	46 (4.7)	1 (0.1)	NA	15 (1.5)	30 (3.1)
AOS (n = 1375)	71 (5.2)	1 (0.1)	NA	38 (2.8)	32 (2.3)
Isolated AOS (n = 1016)	49 (4.8)	0 (0)	NA	29 (2.9)	20 (2.0)
AOS with multilevel left heart obstruction (n = 359)	22 (6.1)	1 (0.3)	NA	9 (2.5)	12 (3.3)
CHDs in which a range of anatomies occurs (born before March 2017)					
AVSD (n = 3388)	321 (9.6)	45 (1.3)	6 (0.2)	82 (2.4)	188 (5.6)
Complete AVSD (n = 2131)	209 (9.9)	25 (1.2)	NA	61 (2.9)	123 (5.8)
Partial AVSD (n = 902)	16 (1.8)	0 (0)	NA	6 (0.7)	10 (1.1)
Tetralogy AVSD (n = 193)	33 (17.1)	2 (1.0)	0 (0)	7 (3.6)	24 (12.4)
Unbalanced AVSD (n = 162)	63 (39.5)	18 (11.1)	6 (3.7)	8 (4.9)	31 (19.1)
TGA (n = 2973)	190 (6.4)	11 (0.4)	0 (0)	93 (3.1)	86 (2.9)
TGA with IVS (n = 1640)	74 (4.5)	2 (0.1)	0 (0)	35 (2.1)	37 (2.3)

(Continued)

TABLE E9. Continued

Sentinel CHDs and subgroups	Total deaths before age 5 y, n (%)	In-hospital deaths after initial palliative procedure, n (%)	In-hospital deaths post-stage 2 or TCPC, n (%)	In-hospital deaths post- biventricular reparative procedure, n (%)	All other deaths, n (%)
Complex TGA without PS (n = 977)	82 (8.4)	5 (0.5)	0 (0)	46 (4.7)	31 (3.2)
Complex TGA with PS (n = 356)	34 (9.6)	4 (1.1)	0 (0)	12 (3.4)	18 (5.1)
PA (n = 1309)	257 (19.6)	62 (4.7)	*	63 (4.8)	130 (9.9)
PA with VSD (n = 908)	166 (18.3)	33 (3.6)	*	44 (4.8)	87 (9.6)
PA with IVS (n = 401)	91 (22.7)	29 (7.2)	0 (0)	19 (4.7)	43 (10.7)

CHD, Congenital heart disease; *TCPC*, total cavopulmonary connection; *HLHS*, hypoplastic left heart syndrome; *NA*, not applicable; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *DORV*, double-outlet right ventricle; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *IVS*, intact ventricular septum; *PS*, pulmonary stenosis; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).

TABLE E10. Overall mortality at age 5 years and post-pathway mortality for each sentinel CHD by risk factors (patients who completed 5 years of follow-up in terms of procedure history, born April 2000-March 2017)

	Patients who died before age 5 y, n (%)					Post-pathway in-hospital deaths before age 5 y, n (%)				
Total cohort (born April 2000-March 2017)	All patients, n (%)	Patients born preterm (before 37 wk), n (%)	Patients with congenital noncardiac comorbidity, n (%)	Patients with Down syndrome, n (%)	Patients born from April 2010 onward, n (%)	All patients, n (%)	Patients born preterm (before 37 wk), n (%)	Patients with congenital noncardiac comorbidity, n (%)	Patients with Down syndrome, n (%)	Patients born from April 2010 onward, n (%)
	1794 (7.7)	**193 (13.4)**	**316 (11.1)**	**184 (7.9)**	**687 (6.9)**	**831 (3.6)**	**92 (6.4)**	**130 (4.6)**	**67 (2.9)**	**321 (3.2)**
By sentinel CHDs										
Functionally univentricular CHDs										
HLHS	436 (42.8)	19 (57.6)	43 (29.9)	*	162 (38.6)	233 (22.9)	12 (36.4)	23 (16.0)	*	84 (20.0)
Non-HLHS FUH	106 (13.4)	14 (23.3)	19 (16.7)	0 (0)	49 (14.0)	50 (6.3)	6 (10.0)	10 (8.8)	0 (0)	26 (7.4)
Biventricular CHDs										
VSD	147 (2.8)	35 (8.6)	41 (7.3)	22 (3.4)	54 (2.3)	29 (0.6)	10 (2.5)	6 (1.1)	*	11 (0.5)
TOF	158 (4.3)	30 (11.5)	53 (7.9)	12 (9.8)	62 (3.7)	76 (2.1)	18 (6.9)	21 (3.1)	*	28 (1.7)
COA	108 (3.1)	13 (6.3)	26 (7.7)	*	32 (2.2)	40 (1.2)	6 (2.9)	9 (2.7)	0 (0)	12 (0.8)
AOS	71 (5.2)	9 (13.0)	7 (6.2)	0 (0)	21 (4.0)	39 (2.8)	*	*	0 (0)	10 (1.9)
CHDs in which a range of anatomy occurs										
AVSD	321 (9.5)	33 (17.5)	58 (14.1)	143 (9.5)	131 (9.1)	133 (3.9)	10 (5.3)	27 (6.6)	57 (3.8)	54 (3.7)
TGA	190 (6.4)	14 (14.7)	12 (8.5)	0 (0)	77 (6.0)	104 (3.5)	8 (8.4)	*	0 (0)	43 (3.3)
PA	257 (19.6)	26 (22.2)	57 (16.4)	*	99 (19.4)	127 (9.7)	20 (17.1)	26 (7.5)	*	53 (10.4)

CHD, Congenital heart disease; *HLHS*, hypoplastic left heart syndrome; *FUH*, functionally univentricular heart; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *COA*, coarctation of the aorta; *AOS*, aortic stenosis; *AVSD*, atrioventricular septal defect; *TGA*, transposition of the great arteries; *PA*, pulmonary atresia. *Results not shown (sample size 1 to 5).