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ORIGINAL RESEARCH

Timing of Cardiac Surgical Interventions and Postoperative Mortality in Children With Severe Congenital Heart Defects Across Europe: Data From the EUROlinkCAT Study

Mads Damkjær, MD, PhD; Ester Garne, MD; Maria Loane, PhD, MSc, BA (Hons); Stine K. Urhoj, PhD; Elisa Ballardini, MD; Clara Cavero-Carbonell, BSPharm, MPH, PhD; Alessio Coi, PhD; Laura García-Villodre, RN; Joanne Given, PhD; Mika Gissler, PhD; Anna Heino, M.Soc.Sc; Sue Jordan, PhD; Elizabeth Limb, MSc; Amanda J Neville, BSc; Anna Pierini, BSc; Anke Rissmann, MD, PhD; Joachim Tan, PhD; Ieuan Scanlon, MSc; Joan K Morris, PhD, MSc, MA

BACKGROUND: The purpose of this study was to evaluate the timing of the first cardiac surgery, the number of cardiac surgeries performed, and 30-day postoperative mortality rate for children with severe congenital heart defects (sCHDs) in their first 5 years of life.

METHODS AND RESULTS: This was a population-based data linkage cohort study linking information from 9 European congenital anomaly registries to vital statistics and hospital databases. Data were extracted for 5693 children with sCHDs born from 1995 to 2004. Subgroup analyses were performed for specific types of sCHD. Children with sCHDs underwent their first surgical intervention at a median age of 3.6 (95% CI, 2.6–4.5) weeks. The timing of the first surgery for most subtypes of sCHD was consistent across Europe. In the first 5 years of life, children with hypoplastic left heart underwent the most cardiac surgeries, with a median of 4.4 (95% CI, 3.1–5.6). The 30-day postoperative mortality rate in children aged <1 year ranged from 1.1% (95% CI, 0.5%–2.1%) for tetralogy of Fallot to 23% (95% CI, 12%–37%) for Ebstein anomaly. The 30-day postoperative mortality rate was highest for children undergoing surgery in the first month of life. Overall 5-year survival for sCHD was <90% for all sCHDs, except transposition of the great arteries, tetralogy of Fallot, and coarctation of the aorta.

CONCLUSIONS: There were no major differences among the 9 regions in the timing, 30-day postoperative mortality rate, and number of operations performed for sCHD. Despite an overall good prognosis for most congenital heart defects, some lesions were still associated with substantial postoperative death.

Key Words: cardiac surgery ■ congenital heart defects ■ pediatric cardiology

Congenital heart defects (CHDs) are a considerable cause of both morbidity and death in infants and children. For instance, we showed in an earlier EUROlinkCAT (Establishing a Linked European Cohort of Children With Congenital Anomalies) study that, 40% of all surgical interventions in children aged <1 year are performed in those with a CHD. The most prevalent severe CHDs (sCHDs) include atrioventricular septal defect (AVSD), tetralogy of Fallot (TOF), and transposition of the great arteries (TGA).[1] Although the surgical strategy differs among lesions (ie, complete operative correction in 1 procedure for AVSD versus

Correspondence to: Mads Damkjær, MD, PhD, Department of Paediatrics, Lillebaelt Hospital, University Hospital of Southern Denmark, Sygehusvej 24, DK-6000, Kolding, Denmark. Email: mads.damkjaer2@rsyd.dk

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CLINICAL PERSPECTIVE

What Is New?

- This study offers a comprehensive analysis of surgical interventions in severe congenital heart disease across European regions, revealing consistent timing for conditions like tetralogy of Fallot, transposition of the great arteries, and atrioventricular septal defect. Notably, hypoplastic left heart and hypoplastic right heart cases involve early, frequent surgeries.
- While overall survival is positive, critical conditions like hypoplastic left heart show 5-year survival of ≈50%.

What Are the Clinical Implications?

- The study informs optimal surgical timing for congenital heart disease, aligning with recommendations for tetralogy of Fallot and atrioventricular septal defect.
- Vigilance is crucial for managing high-frequency surgeries and mortality risks in patients with hypoplastic left heart and hypoplastic right heart.
- Population-based data are vital for accurate mortality assessment, emphasizing the risk of delayed surgical repair, especially for atrioventricular septal defect.

Nonstandard abbreviations and acronyms

AVSD	atrioventricular septal defect
CoA	coarctation of the aorta
EUROCAT	European Concerted Action on Congenital Anomalies and Twins
EUROlinkCAT	Establishing a Linked European Cohort of Children With Congenital Anomalies
HLH	hypoplastic left heart
HRH	hypoplastic right heart
sCHD	severe congenital heart defect
TGA	transposition of the great arteries
TOF	tetralogy of Fallot

staged repair in hypoplastic left heart [HLH]), complete surgical correction or stage II palliation is recommended in all children with sCHDs within the first year of life.[2–6] The optimal timing with regard to age or weight of the child remains to be determined. As pointed out by Holst et al,[7] "outcomes and practices in CHD continue to be relatively heterogeneous; this is largely because of surgeon-specific practices, institutional preferences, and heterogeneity inherent to CHD." Differences in surgical timing between institutions may further be affected by differences in case mix, and, as such, it can be difficult to directly compare tertiary sCHD surgical centers. Additionally, not all children with sCHDs reach the tertiary surgical centers, or they may not be offered surgery. Therefore, population-based studies including all liveborn children with sCHDs are important for evaluating their morbidity and death.

As part of the EUROlinkCAT project,[8] we sought to evaluate the timing of the first cardiac surgical intervention, the number of cardiac surgical interventions, postoperative death, and survival in children born with sCHDs across 9 European regions in 6 countries for the first 5 years of life.

METHODS

Aggregated data from the local registries were uploaded to a secure central data repository at Ulster University. A condition for local approval for linking databases was that the linked data cannot be shared.

This is a European, population-based linkage cohort study arising from the EUROlinkCAT project.[8] This project includes data on morbidity and death for children born with congenital anomalies. As described in detail in Loane et al,[9] 21 registers originally agreed to participate. Due to challenges in obtaining local ethics permits or quality of data linkage, 9 registers were able to participate in the present study, all of which are from Western Europe.

Study Population

All children reported in the European Concerted Action on Congenital Anomalies and Twins (EUROCAT) congenital anomaly registries with the *International Classification of Diseases, Tenth Revision* (*ICD-10*) diagnoses listed below. Finland and Funen (Denmark) used the equivalent *International Classification of Diseases, Ninth Revision* (*ICD-9*) codes for part of the study period, and these were translated and mapped to corresponding *ICD-10* codes. For comparison of *ICD-9* and *ICD-10* codes, please refer to Table S1. The following EUROCAT-defined cardiac subgroups were included in the study.

Severe CHDs

All diagnoses of common arterial truncus, double-outlet right ventricle, TGA, single ventricle, AVSD, TOF, pulmonary atresia, tricuspid atresia or stenosis, Ebstein anomaly, hypoplastic right heart (HRH) syndrome, aortic valve atresia or stenosis, mitral valve anomalies, HLH, coarctation of the aorta (CoA), interrupted aortic arch, and total anomalous pulmonary venous return.

The main results presented in this study are for children with isolated sCHDs only (although a child could have >1 type of cardiac anomaly). Children with associated major anomalies in other organ systems or a genetic diagnosis were excluded.[10] The consequence of excluding genetic diagnoses is that, for instance, for AVSD, only children with AVSD who do not have Down syndrome were included in the study population. Data on all children with sCHDs, including those with associated anomalies/genetic syndromes, are presented for comparison in Table S2.

Data on survival and on surgical procedures performed during in-patient hospital stays for all children up to the child's 10th birthday or the end of 2015, whichever came earlier, were obtained by electronic linkage to mortality statistics or vital statistics databases and hospital databases. Details about the linkage methods have been published elsewhere.[8,9]

The hospital databases in Finland; Funen, Denmark; Tuscany; and England (East Midlands and South Yorkshire, Thames Valley, and Wessex) covered hospitalizations in the whole country. For Wales, this included procedures carried out in England. For the Valencian Region and Emilia Romagna, the hospital databases covered the same region as the EUROCAT registry.

Surgical procedures were coded according to the coding systems used in the national health systems. Italy and Spain used *International Classification of Diseases, Ninth Revision, Clinical Modification* (*ICD-9-CM*) for the study period; Wales and England used Classifications of Interventions and Procedures; and Finland and Denmark used national adaptions of the Nordic Medico-Statistical Committee Classification of Surgical Procedures. Surgical procedures included catheter procedures but did not include diagnostic procedures (eg, right heart catheterization); for an overview of what was included as a surgery, please refer to Table S3.

Specific Surgical Procedures

For some anomalies, it was possible to identify specific codes for surgical correction of a given CHD; this was the case for CoA, AVSD, modified Blalock–Thomas–Taussig-shunt (which included the original subclavian artery to pulmonary artery shunt), and hemi-Fontan (which included all codes for superior cavopulmonary anastomosis and therefore also the bidirectional Glenn procedure). Fontan codes could be reliably identified only in the Classifications of Interventions and Procedures/Nordic Medico-Statistical Committee Classification of Surgical Procedures/*ICD-9-CM* codes from Denmark, Italy, and Wales. Data from these registries were also used to compare surgical timing for the specific condition versus having any cardiac surgery. For those lesions without lesion-specific codes, we identified having any cardiac surgery or any surgery in general as a proxy for cardiac surgery.

Statistical Analysis

The proportions of children having surgery were calculated using Kaplan–Meier survival estimates to allow for the censoring of children occurring on December 31, 2015, date of death, or date of emigration from the study region or country, as previously described in detail.[11] The numbers of surgical procedures the children had and the age at the time of the first surgery were non-Gaussian with a few extreme outliers, and therefore they were reported as medians and interquartile ranges; meta-analytic methods to combine all results across registries have been previously described elsewhere.[12] Briefly, quantile estimation methods were used to obtain pooled estimates of the median age at first operation and the 95% CIs using the metamedian package in R, version 4.0.3 (R Foundation for Statistical Computing, Vienna, Austria).[13]

Release of Small Numbers

The release of small numbers (<5) was not allowed for several of the registries; therefore, for most of the rare anomalies, survival/mortality rates are pooled estimates from all 9 European regions. Data on survival/death could only be reported for individual registries on AVSD, TGA, and TOF.

Postoperative Death

For analysis, results were subdivided into 5 different age categories for each anomaly: 0 to 27 days, 28 days to <1 year, 0 to <1 year, 1 to 4 years, and 5 to 9 years. Please note that for postoperative death, the denominator is only children with a specific diagnosis and a surgical code; this is in contrast to the 1- and 5-year mortality and survival rates where the denominator is all children with the specific diagnosis. Due to the smaller numbers of children in the postoperative mortality rate, the combined mortality rate was not estimated using a meta-analytic approach; rather, all the deaths after surgery from each registry were combined and divided by the total number of children with a specific diagnosis and surgical code.

The 1- and 5-year mortality rates were obtained from data that were aggregated from 13 registries (both regional and national), as previously described in detail in Glinianaia et al and Coi et al.[14–16] The aggregated mortality data from these papers are presented separately in Table S4.

Ethical Clearance

All registries affiliated with the EUROCAT network possess the essential ethical authorizations and protocols

to facilitate standard surveillance, data compilation, and secure transmission of deidentified information to the central EUROCAT database. Adhering to pertinent national directives, the registries provided substantiation of these clearances to the EUROlinkCAT ethical dossier. Ulster University, the central data repository, also secured ethical endorsement (approval reference: FCNUR-21-060). Notably, as per the nature of the study, no institutional review board approval was requisite. The implementation of parental consent for the registration of infants with anomalies by local registries aligns with respective national legislations. For comprehensive insights, kindly consult the dedicated protocol paper.

RESULTS

Population Characteristics

We obtained data from 9 EUROCAT registries from 6 countries for children born from 1995 to 2014. Data were available on 5693 children born with an sCHD (Table 1).

Timing of First Surgical Intervention

For all children born with an sCHD, the median age at first surgical intervention was 3.6 (95% CI, 2.6–4.5) weeks (aged up to 5 years) (Table 2). For children with an sCHD, 78% (95% CI, 70%–85%) had a surgery in the first year of life and 36% (95% CI, 31%–42%) between ages 1 and 4 years. The same pattern was observed for all the subtypes of sCHD with a higher percentage of children undergoing surgery in the first year of life, than at age 1 to 4 years. The timing of the first surgical intervention varied greatly among different sCHDs, ranging from 0.3 (95% CI, 0.0–0.7) week for HRH to 38.6 (95% CI, 33.0–44.1) weeks for AVSD (Table 2). Measures of heterogeneity (I^2 values) for all variables can be found in Table S2.

Number of Surgical Interventions

Children with sCHDs were operated on a median of 3.2 (95% CI, 2.6–3.9) times in their first 5 years of life. This includes both cardiac and noncardiac operations. Those with the highest median number of cardiac operations in the first 5 years of life were children with HRH and HLH who underwent 5.0 (95% CI, 3.4–6.5) and 4.4 (95% CI, 3.1–5.6) cardiac surgical procedures, respectively (Table 2). Measures of heterogeneity (I^2 values) for all variables can be found in Table S2.

Difference in Timing of Cardiac Surgery Between European Regions

The timing of the first cardiac surgical intervention for each of the 9 European registries are summarized as the median age and interquartile ranges in Figure 1A and 1B. The timing of the first surgery for specific sCHDs was fairly consistent in the first couple of weeks of life for TGA, total anomalous pulmonary venous return, and CoA. Children with AVSD were operated on around age 5 to 6 months and TOF at age 4 to 5 months (Figure 1A). For both the Blalock shunt and hemi-Fontan procedures, we observed consistent timing across Europe (Figure 1B).

Postoperative Death

We evaluated the 30-day postoperative mortality rate (based on the variable *any surgery*) for children with

Table 1. Overview of the Participating Registries From the 9 Different European Regions

Region	Birth years	Number of live births	Percentage of children with sCHD having any surgery (95% CI)			
			Age <1 y	Age 1–4 y	Age 5–9 y	Surgeries at both <1 y and 1–4 y
Denmark, Funen	1995–2014	159	72.6 (65.2–79.6)	41.0 (32.8–50.4)	44.2 (34.5–55.2)	29.1 (20.0–40.0)
Finland	1997–2014	2051	57.1 (54.9–59.3)	35.9 (33.7–38.3)	23.6 (21.2–26.2)	23.4 (21.1–25.9)
Italy, Tuscany	2005–2014	287	83.0 (78.4–87.1)	24.1 (18.8–30.5)	35.7 (23.9–51.2)	17.9 (12.3–25.0)
Italy, Emilia Romagna	2008–2014	358	83.2 (79.1–86.9)	24.3 (19.2–30.6)		21.3 (15.9–27.8)
Spain, Valencian Region	2010–2014	265	67.2 (61.5–72.9)	32.8 (25.6–41.4)		25.4 (18.2–34.3)
United Kingdom, Wales	1998–2014	895	72.5 (69.5–75.5)	36.1 (32.6–39.8)	29.8 (25.8–34.3)	26.6 (22.9–30.7)
United Kingdom, Thames Valley	2005–2013	314	85.2 (80.8–89.1)	43.2 (36.8–50.3)	34.8 (23.9–48.8)	37.1 (30.1–45.0)
United Kingdom, Wessex	2004–2014	426	88.5 (85.1–91.4)	38.5 (33.1–44.4)	44.4 (36.1–53.6)	31.9 (25.7–38.8)
United Kingdom, East Midlands and South Yorkshire	2003–2012	938	83.7 (81.2–86.1)	51.0 (47.4–54.8)	38.0 (33.0–43.5)	38.8 (34.2–43.7)
Total		5693				

Shown are the births years available from each registry, the number of live births with severe congenital heart disease (sCHD), and the percentage of children having any surgery for 4 different age categories.

Table 2. Median Age at First Cardiac Surgery and Median Number of Cardiac Surgical Procedures in the First 10 Years of Life for All Participating Registries

Anomaly	Surgery coding	Number of children with anomaly	Percentage with any surgery			Median age first operation in weeks up to age 5 y (95% CI)	Median number of operations up to age 5 y (95% CI)
			<1 y	1–4 y	0–4 y		
sCHD	Any surgery	5693	78 (70 to 85) I^2=97.7	36 (31 to 42) I^2=91.2	88 (81 to 92) I^2=97.2	3.6 (2.6 to 4.5) I^2=87.5	3.2 (2.6 to 3.9) I^2=97.7
AVSD	Any surgery	453	58 (48 to 66)	47 (36 to 57)	82 (70 to 89)	21.6 (17.7 to 25.4)	3.0 (2.4 to 3.6)
AVSD	AVSD surgery	453	29 (22 to 36)	21 (14 to 28)	51 (41 to 60)	38.6 (33.0 to 44.1)	1.0 (1.0 to 1.0)
TGA	Any surgery	980	90 (86 to 94)	31 (24 to 38)	94 (89 to 96)	1.1 (0.7 to 1.5)	3.9 (3.2 to 4.6)
TOF	Any surgery	805	90 (83 to 94)	41 (33 to 49)	97 (95 to 98)	24.7 (21.1 to 28.3)	3.0 (2.4 to 3.6)
Pulmonary valve atresia	Cardiac surgery	245	84 (69 to 92)	56 (45 to 66)	92 (78 to 98)	0.9 (0.5 to 1.3)	3.8 (3.3 to 4.2)
TAPVR	Cardiac surgery	193	87 (75 to 93)	13 (8 to 20)	93 (81 to 97)	2.1 (0.9 to 3.2)	2.0 (1.7 to 2.4)
CoA	Any surgery	1566	81 (73 to 87)	33 (27 to 38)	89 (82 to 93)	2.4 (1.8 to 3.1)	2.4 (2.0 to 2.9)
CoA	Surgery for coarctation	1566	66 (54 to 76)	7 (5 to 10)	73 (61 to 83)	2.7 (1.9 to 3.6)	1.0 (1.0 to 1.0)
HLH	Any surgery	476	90 (81 to 95)	71 (48 to 85)	97 (88 to 99)	0.9 (0.7 to 1.0)	5.3 (3.8 to 6.7)
HLH	Cardiac surgery	476	88 (76 to 95)	68 (44 to 83)	97 (86 to 99)	0.9 (0.7 to 1.0)	4.4 (3.1 to 5.6)
HLH	Blalock shunt	476	25 (8 to 47)	2 (0 to 9)	26 (8 to 49)	0.9 (0.5 to 1.4)	1.2 (0.8 to 1.6)
HLH	Hemi to Fontan	476	60 (41 to 75)	19 (4 to 42)	67 (46 to 82)	25.4 (19.8 to 30.9)	1.1 (0.9 to 1.4)
HLH	Complete Fontan	94	39 (27 to 52)	19 (8 to 33)	39 (13 to 64)	26.5 (0.4 to 52.7)	1.0 (1.0 to 1.0)
HRH	Cardiac surgery	62	95 (83 to 99)	52 (24 to 74)	98 (88 to 100)	0.3 (0.0 to 0.7)	5.0 (3.4 to 6.5)
HRH	Blalock shunt	62	48 (26 to 68)	2 (0 to 12)	51 (27 to 70)	0.4 (0.0 to 0.9)	1.0 (1.0 to 1.0)
HRH	Hemi to Fontan	62	58 (41 to 72)	24 (10 to 42)	81 (65 to 90)	26.1 (19.3 to 32.8)	1.0 (1.0 to 1.0)
HRH	Complete Fontan	20	31 (0 to 96)	23 (5 to 47)	28 (6 to 57)	213.2 (90.4 to 336.0)	1.0 (1.0 to 1.0)
Aortic valve atresia/stenosis	Any surgery	661	63 (49 to 74)	38 (26 to 49)	69 (57 to 78)	3.1 (1.4 to 4.8)	2.0 (1.3 to 2.8)
Double outlet right ventricle	Any surgery	226	89 (84 to 93)	52 (36 to 67)	97 (93 to 99)	5.1 (2.4 to 7.7)	4.1 (3.4 to 4.9)
Common arterial truncus	Any surgery	106	76 (63 to 84)	36 (23 to 48)	94 (73 to 99)	2.6 (1.5 to 3.8)	3.1 (2.3 to 3.8)
Single ventricle	Any surgery	222	89 (75 to 96)	77 (40 to 93)	96 (88 to 99)	1.8 (1.3 to 2.3)	5.8 (4.3 to 7.4)
Triscuspid atresia and stenosis	Any surgery	318	80 (67 to 88	55 (40 to 67)	86 (75 to 92)	2.7 (1.0 to 4.5)	5.0 (4.4 to 5.6)
Ebstein anomaly	Any surgery	194	35 (19 to 52)	29 (21 to 37)	45 (31 to 58)	13.3 (2.8 to 29.5)	2.3 (1.4 to 3.2)

Numbers in parentheses indicate 95% CIs. For those anomalies where it was possible to identify the specific surgical codes for anatomic correction of the anomaly, rather than just the overarching category of cardiac surgery, we have indicated the median age for both. For the anomaly-specific surgical procedures, the median number of surgical procedures refers to that specific surgery alone, such that this will be >1 only if the surgery is undertaken more than once in the first 5 years of life. The number of children undergoing surgery can be calculated directly from the table. Please note that surgical codes for the Fontan procedure could be reliably identified only in Denmark, Italy, and Wales. I^2 values are shown only for sCHD. All these values can be found in Table S2. AVSD indicates atrioventricular septal defect; CoA, coarctation of the aorta; HLH, hypoplastic left heart; HRH, hypoplastic right heart; sCHD, severe congenital heart defect; TAPVR, total anomalous pulmonary venous return; TGA, transposition of the great arteries; and TOF, tetralogy of Fallot.

sCHDs across all 9 European regions divided into 5 different age categories (Table 3). The highest 30-day postoperative mortality rates occurred when the surgery was performed in the first month of life (0–28 days), which for all children with an sCHD was 7.0% (95% CI, 6.1%–8.0%). The postoperative mortality rate was 3.8% (95% CI, 3.2%–4.5%) for children aged 29 days to 1 year, 1.7% (95% CI, 1.2%–2.5%), for children aged 1 to 4 years, and 1.9% (95% CI, 1.0%–3.5%) for children aged 5 to 9 years. The highest postoperative mortality rates were found in children with Ebstein anomaly, HLH, HRH, and common arterial truncus.

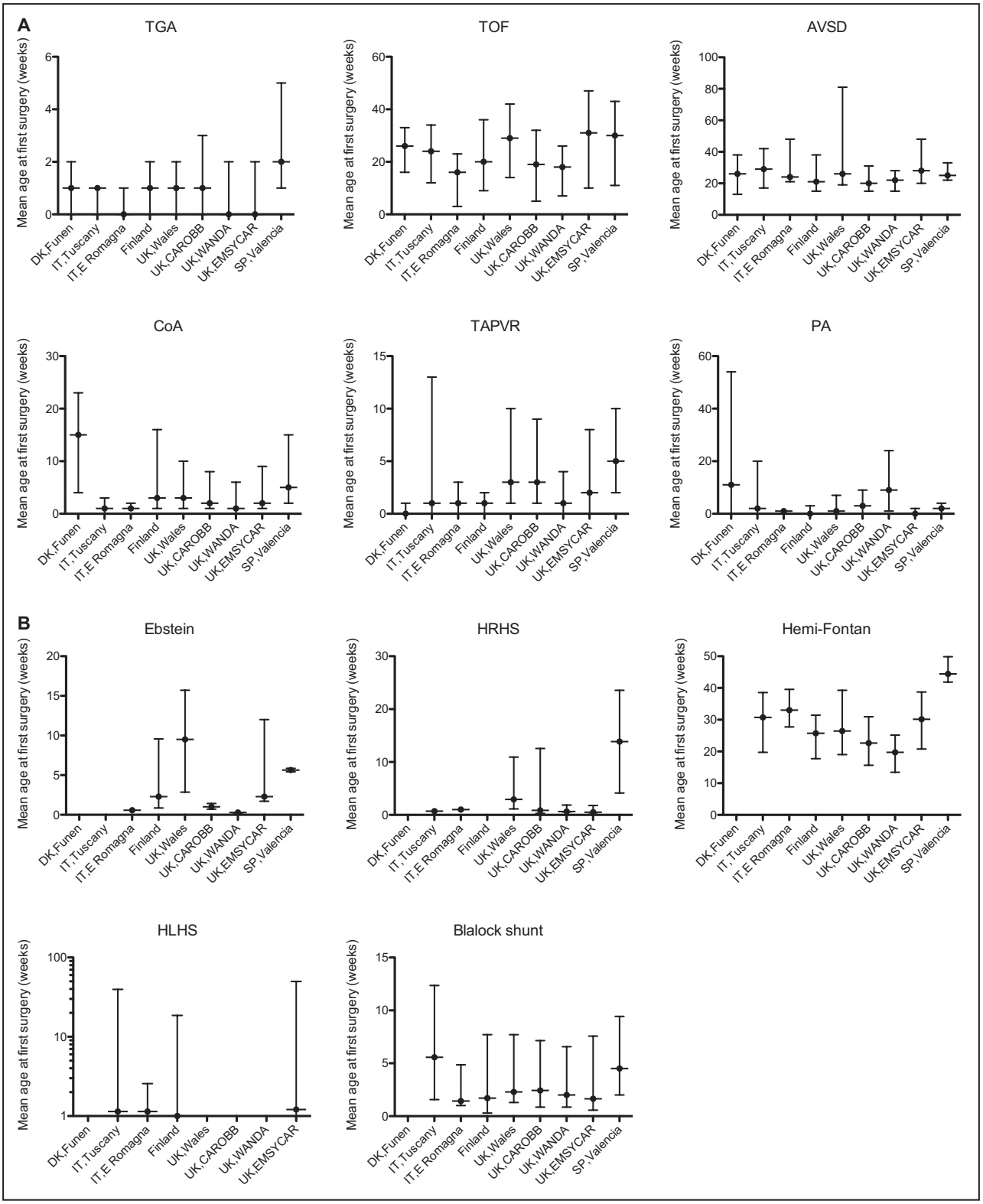

Figure 1. Median age and interquartile range (in weeks) at first cardiac surgery for children with congenital heart defects from the participating European regions.

A, Timing of first surgical interventions for specific conditions and procedures. **B**, Timing of first surgical interventions for common CHDs. Please note that for the Blalock shunt and hemi-Fontan, this is not restricted to any specific cardiac diagnosis, only the surgical codes, and as such will include any anomaly in which these surgical procedures have been undertaken. AVSD indicates atrioventricular septal defect; CoA, coarctation of the aorta; CAROBB, Congenital Anomaly Register of Oxfordshire, Berkshire, and Buckinghamshire; EMSYCAR, The East Midlands & South Yorkshire Congenital Anomalies Register; HLHS, hypoplastic left heart syndrome; HRHS, hypoplastic right heart syndrome; PA, pulmonary atresia; TAPVR, total anomalous pulmonary vein return; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; and WANDA, Wessex Registry of Antenatally Detected Anomolies.

Table 3. The 30-Day Postoperative Mortality Rate for All Included Severe Congenital Heart Defects, Divided Into 4 Age Categories

Anomaly	0–28 d		28 d to <1 y		0 to <1 y		1–4 y		5–9 y	
	Number of surgeries	Postoperative mortality rate per 100 surgeries (95% CI)	Number of surgeries	Postoperative mortality rate per 100 surgeries (95% CI)	Number of surgeries	Postoperative mortality rate per 100 surgeries (95% CI)	Number of surgeries	Postoperative mortality rate per 100 surgeries (95% CI)	Number of surgeries	Postoperative mortality rate per 100 surgeries (95% CI)
sCHD	2707	7.0 (6.1–8.0)	3608	3.8 (3.2–4.5)	6315	5.2 (4.6–5.8)	1682	1.7 (1.2–2.5)	565	1.9 (1.0–3.5)
TGA	726	4.5 (3.1–6.3)	296	3.4 (1.6–6.1)	1022	4.2 (3.1–5.6)	184	1.1 (0.1–3.9)	67	3.0 (0.4–10.4)
TOF	99	5.1 (1.7–11.4)	634	0.5 (0.1–1.4)	733	1.1 (0.5–2.1)	231	1.7 (0.5–4.4)	62	0.0 (0.0–5.8)
Pulmonary atresia	201	7.0 (3.9–11.4)	193	6.7 (3.6–11.2)	394	6.9 (4.6–9.8)	153	3.3 (1.1–7.5)	56	3.6 (0.4–12.3)
TAPVR	141	9.9 (5.5–16.1)	104	8.7 (4.0–15.8)	245	9.4 (6.0–13.8)	41	7.3 (1.5–19.9)	9	11.1 (0.3–48.2)
CoA	742	3.2 (2.1–4.8)	647	2.0 (1.1–3.4)	1389	2.7 (1.9–3.7)	277	2.5 (1.0–5.1)	91	0.0 (0.0–4.0)
HLH	43	11.6 (3.9–25.1)	44	9.1 (2.5–21.7)	87	10.3 (4.8–18.7)	25	0.0 (0.0–13.7)	17	0.0 (0.0–19.5)
HRH	316	15.5 (11.7–20.0)	259	5.8 (3.3–9.4)	575	10.8 (8.4–13.6)	186	1.6 (0.3–4.6)	62	0.0 (0.0–5.8)
Aortic atresia/ interrupted aortic arch	102	11.8 (6.2–19.6)	72	6.9 (2.3–15.5)	174	9.8 (5.8–15.2)	57	0.0 (0.0–6.3)	11	0.0 (0.0–28.5)
Double-outlet right ventricle	121	9.9 (5.2–16.7)	206	6.3 (3.4–10.5)	327	7.6 (5.0–11.1)	125	1.6 (0.2–5.7)	38	5.3 (0.6–17.7)
Common arterial truncus	71	14.1 (7.0–24.4)	90	5.6 (1.8–12.5)	161	9.3 (5.3–14.9)	53	1.9 (0.0–10.1)	25	0.0 (0.0–13.7)
Single ventricle	112	7.1 (3.1–13.6)	149	2.7 (0.7–6.7)	261	4.6 (2.4–7.9)	124	1.6 (0.2–5.7)	38	5.3 (0.6–17.7)
Triscuspid atresia and stenosis	113	11.5 (6.3–18.9)	168	6.0 (2.9–10.7)	281	8.2 (5.3–12.0)	105	1.9 (0.2–6.7)	47	4.3 (0.5–14.5)
Ebstein anomaly	28	32.1 (15.9–52.4)	24	16.7 (4.7–37.4)	52	23.1 (12.5–36.8)	20	0.0 (0.0–16.8)	15	0.0 (0.0–21.8)
Aortic valve atresia/stenosis	194	7.2 (4.0–11.8)	215	5.6 (2.9–9.5)	409	6.4 (4.2–9.2)	135	1.5 (0.2–5.2)	63	0.0 (0.0–5.7)

For each age category, the number of children having surgery, the number who died within 30 d, and the calculated postoperative mortality rate are shown. AVSD indicates atrioventricular septal defect; CoA, coarctation of the aorta; HLH, hypoplastic left heart; HRH, hypoplastic right heart; sCHD, severe congenital heart defects; TAPVR, total anomalous pulmonary vein return; TGA, transposition of the great arteries; and TOF, tetralogy of Fallot.

Differences in Surgical Outcomes Across Europe

We examined the 30-day postoperative mortality rate in the first year of life for all cases of sCHD. Due to problems with small numbers, data on most of the specific sCHD diagnoses had to be excluded from the analysis. It was possible to compare only the 30-day postoperative mortality rate across the 9 regions for cases of AVSD, TGA, and TOF. With this restriction, data from Funen, Denmark, had to be excluded due to small numbers for all 3 anomalies, and data from the Valencia Region had to be excluded for AVSD. Generally, 30-day postoperative mortality rates for TGA and TOF are similar across the 9 regions. The only notable exception appears to be that Wales has a higher AVSD mortality rate than the other regions (Figure 2). Data on all-cause mortality in the first year of life for all 9 European regions are included in Table S5.

DISCUSSION

In this cohort from Western Europe, we show that children with sCHDs are operated on at a median age of 3.6 weeks. For most of the specific sCHDs (TOF, TGA, total anomalous pulmonary venous return, AVSD, and CoA) timing was broadly similar across the included regions. Children born with HLH and HRH underwent the most cardiac surgical procedures, with half of them having at least 5 surgical procedures on average in the first 5 years of life. The 30-day postoperative mortality rate was highest for children undergoing surgery in the first month of life. The 30-day postoperative mortality rate was comparable across the European regions. Although overall survival is good (close to 90%), the 5-year survival for critical conditions such as HLH is still only around 50% (Table S4).

The suggested optimal timing for surgical correction of TOF in infants with no or mild symptoms is around 3 to 6 months[17] and for AVSD around 4 months.[2] This

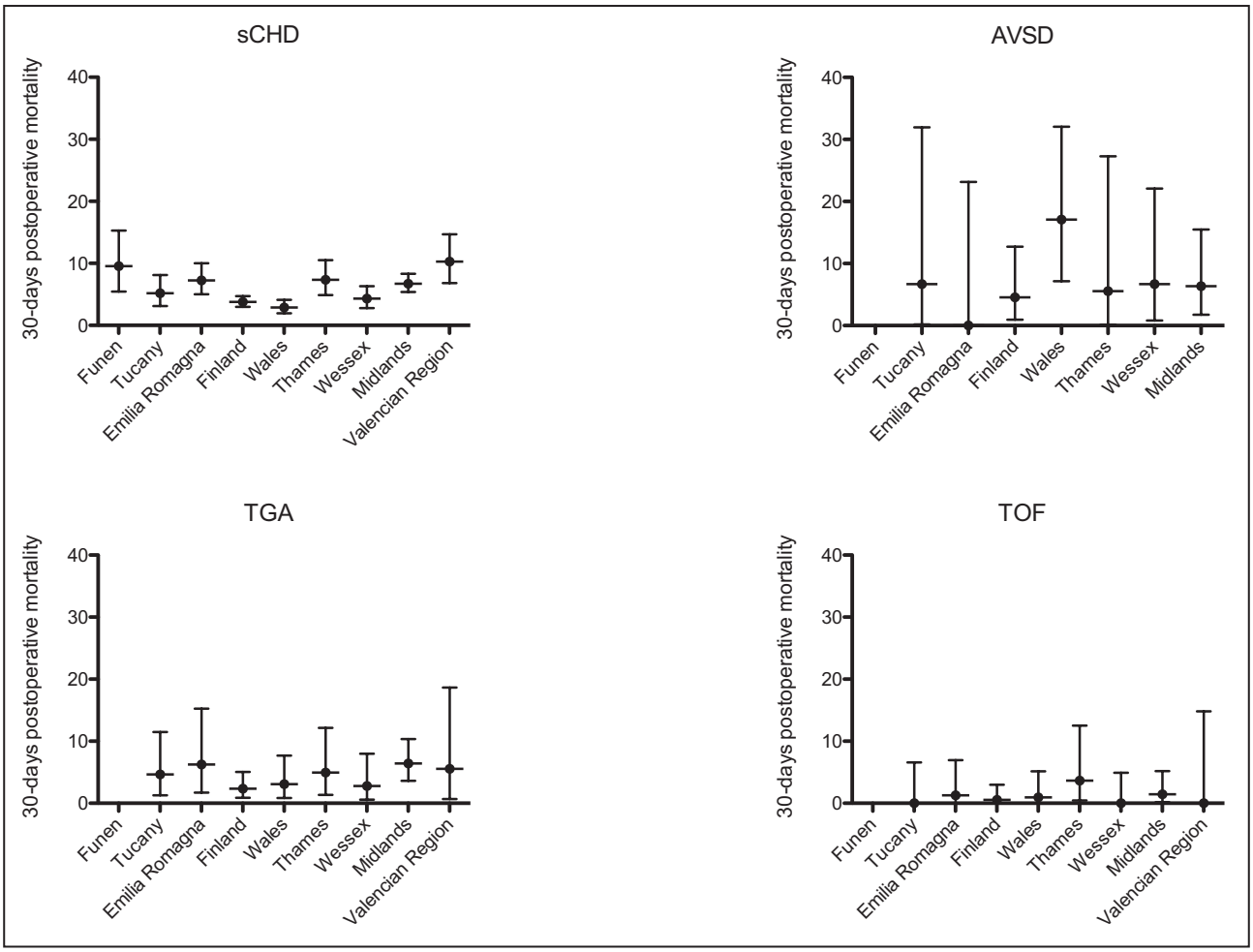

Figure 2. Plot of 30-day postoperative mortality rate in the first year of life for 9 different European regions.
Data from Funen, Denmark had to be excluded due to small numbers for all 3 anomalies, and data from the Valencia Region had to be excluded for AVSD. Also please note that AVSD includes only nonchromosomal cases. AVSD indicates atrioventricular septal defect; sCHD, severe congenital heart defect; TGA, transposition of the great arteries; and TOF, tetralogy of Fallot.

is in line with what we observe across the 9 European regions, with timing of surgery for TOF at 4 to 5 months and AVSD at 5 to 6 months (Figure 1A). For critical lesions such as TGA, pulmonary atresia, and total anomalous pulmonary venous return, we also observe, as expected, rather consistent surgical timing in the neonatal period across the 9 regions. For patients with HLH or HRH, the modified Blalock–Taussig–Thomas shunt was performed at a median age of 0.4–0.9 weeks, which is in line with other reports.[18] Timing of the hemi-Fontan (or bidirectional Glenn procedure) is around 6 months (23.3 weeks in HLH), which is also well in line with other reports.[19]

We found the highest 30-day postoperative mortality rate in the neonates (6.5%), which is consistent with reports from other surgical centers.[20] It can be argued that the 30-day mortality rate is not the most optimal parameter to measure if our aim is to assess surgical outcome. Increased capacity to prolong life in the intensive care unit can decrease the 30-day mortality rate without this translating into increased survival to discharge.[21] Some authors therefore prefer the discharge mortality rate as the primary surgical outcome.[22] As such, there is a risk that our results slightly underestimate the postoperative mortality rate.

There were regional differences in postoperative mortality rates for children with sCHD, with the highest mortality rate being approximately double that of the lowest. However, it should be cautioned that when looking at data on all children with sCHDs, differences in case mix of rare conditions with high mortality such as HRH and HLH might well skew comparisons between regions. For the most prevalent diagnoses (AVSD, TGA, and TOF), we had large enough data sets to allow for a more direct comparison among regions. This showed that comparable results with respect to the 30-day postoperative mortality rate was achieved across the European regions. The only notable exception appears to be AVSD cases in Wales. One explanation for this might be that, although the surgical timing was similar, the upper limits of the 95% CI for Wales was 81 weeks, with all other regions at around 40 weeks. This shows that some patients underwent surgery in the second year of life. Later surgical repair increases the risk of pulmonary vascular hypertension and may increase the mortality rate,[23] which could be a possible explanation for the higher AVSD mortality rate in Wales.

An important limitation when looking at cardiac surgical intervention is that in most instances we have limited information about the precise repair performed. The primary challenge was differences in coding systems among European regions, making it impossible to reliably translate between systems for most procedures. Another important limitation is that we do not have data on when a diagnosis was made or the potential referral time to a tertiary surgical center. It is well established that in utero diagnosis or early postnatal diagnosis of a duct-dependent CHD can the decrease mortalityrate.[24,25] A significant number of deaths in children with a duct-dependent CHD occur in those with late or no referral to a tertiary center.[25] Most studies of mortality rates are data from tertiary units reporting their outcome in surgical series,[26] and thus to examine the true mortality rate for a specific CHD, it is important to have population-based data as in the present study.

In conclusion, there were no major differences among the 9 Western European regions in the timing and number of surgical procedures for children with CHDs. Despite an overall good prognosis for most CHDs, some lesions are still associated with substantial postoperative death and a 5-year survival of <90%. The results of this study can aid medical staff in counseling parents regarding timing of surgery and survival.

ARTICLE INFORMATION

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Affiliations

Department of Paediatrics and Adolescent Medicine, Lillebaelt Hospital, University Hospital of Southern Denmark, Kolding, Denmark (M.D., E.G.); Department of Regional Health Research, University of Southern Denmark, Odense, Denmark (M.D., E.G.); Faculty of Life & Health Sciences, Ulster University, Northern Ireland, UK (M.L., J.G.); Section of Epidemiology, Department of Public Health, University of Copenhagen, Copenhagen, Denmark (S.K.U.); Neonatal Intensive Care Unit, Paediatric Section, IMER Registry, Department of Medical Sciences, University of Ferrara, Ferrara, Italy (E.B.); Rare Diseases Research Unit, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region, Valencia, Spain (C.C., L.G.); Unit of Epidemiology of Rare Diseases and Congenital Anomalies, Institute of Clinical Physiology, National Research Council, Pisa, Italy (A.C., A.P.); Department of Knowledge Brokers, THL Finnish Institute for Health and Welfare, Helsinki, Finland (M.G., A.H.); Faculty of Medicine, Health and Life Science, Swansea University, Swansea, UK (S.J., I.S.); Population Health Research Institute, St George's, University of London, London, UK (E.L., J.T., J.K.M.); Registro IMER - IMER Registry (Emila Romagna Registry of Birth Defects), Center for Clinical and Epidemiological Research, University of Ferrara Azienda Ospedaliero-Universitaria di Ferrara, Ferrara, Italy (A.J.N.); and Malformation Monitoring Centre Saxony-Anhalt, Medical Faculty Otto-von-Guericke-University Magdeburg, Magdeburg, Germany (A.R.).

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Disclosures

None.

Supplemental Material

Tables S1–S5

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