The epidemiology of persons living with Fontan in 2020 and projections for 2030: Development of an epidemiology model providing multinational estimates

Supplementary Material

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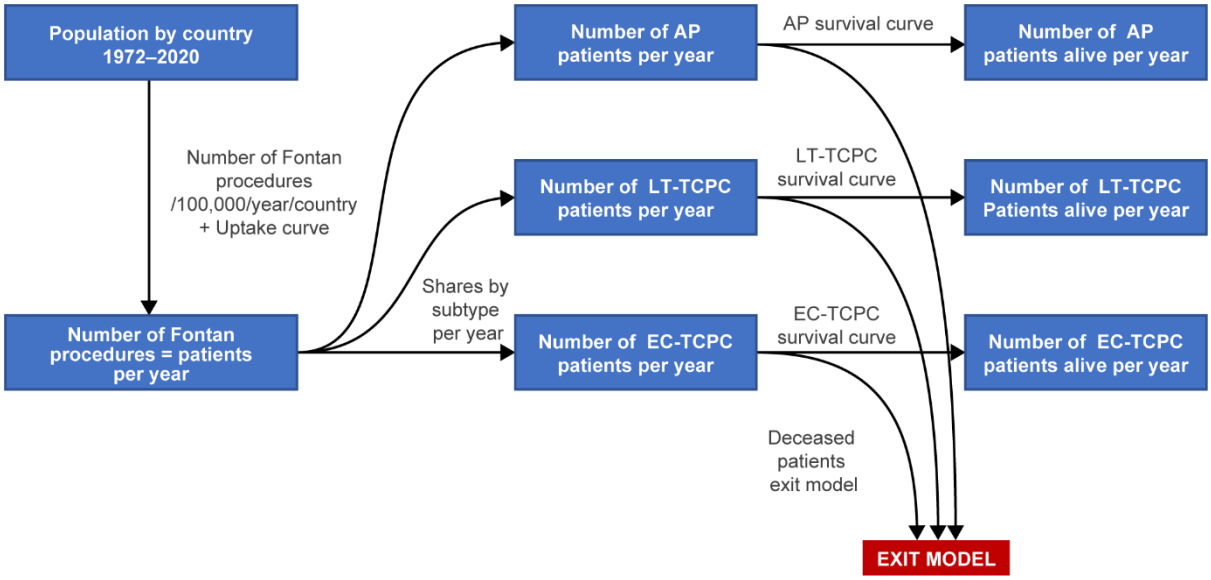
Figure S1: Overview of development of the epidemiologic model



AP, atriopulmonary connection; EC-TCPC, extracardiac total cavopulmonary connection; LT-TCPC, lateral tunnel total cavopulmonary connection.

Figure S2: Uptake curve applied to provide estimates for the number of Fontan procedures occurring prior to 2010

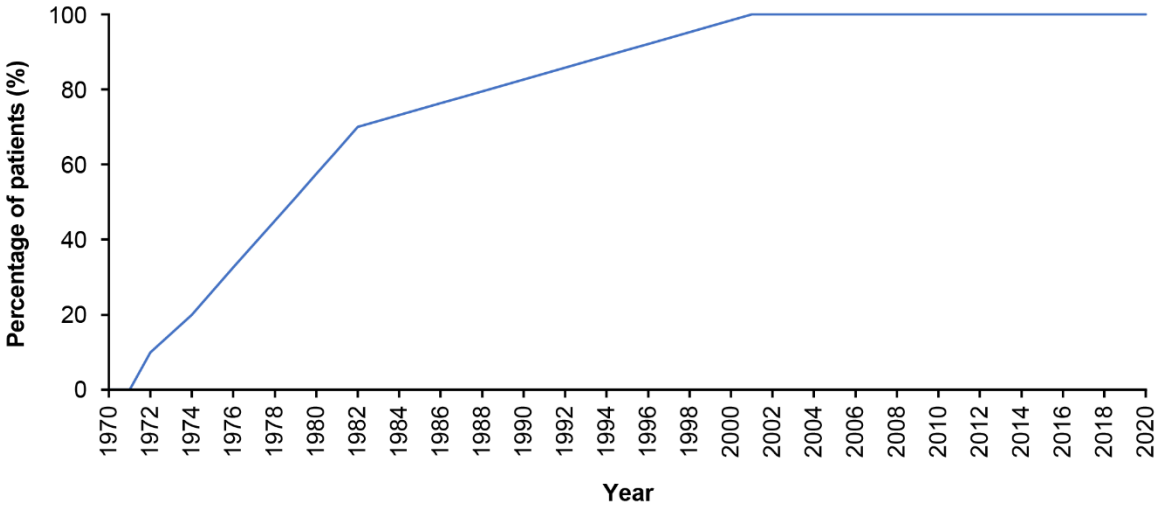
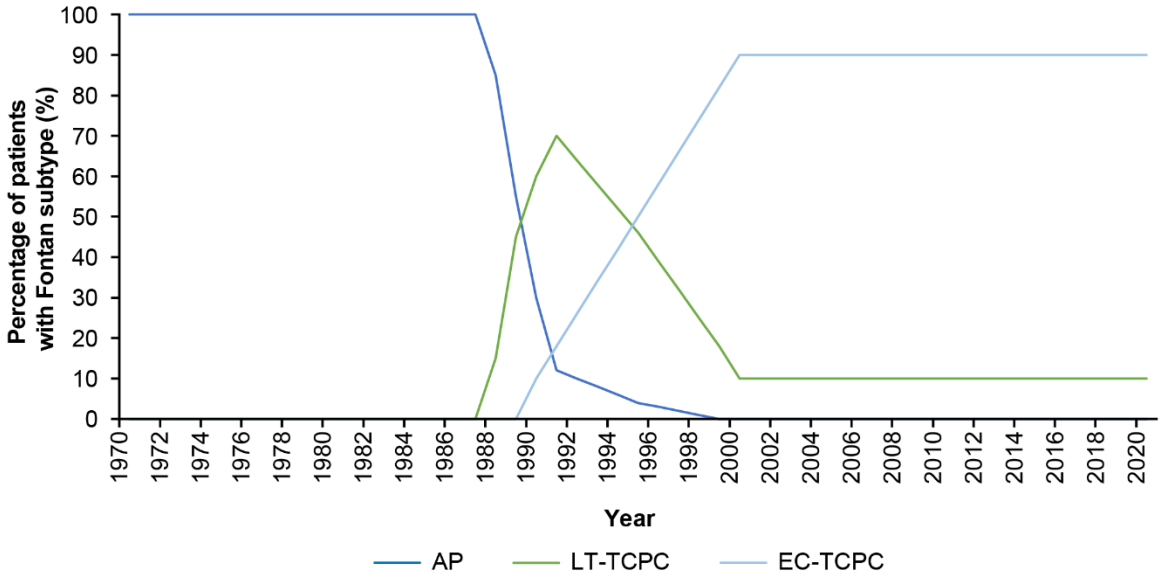
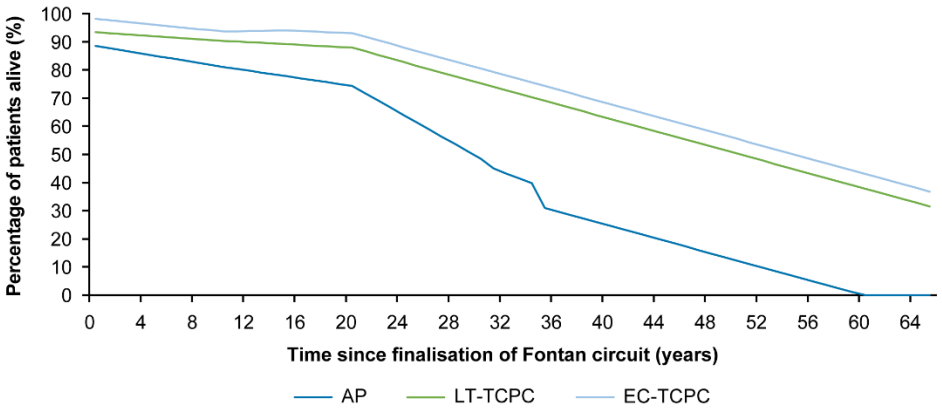


Figure S3: Uptake curve used to model the number of Fontan procedures occurring, stratified by Fontan procedural subtype, for input into the epidemiology model



AP, atriopulmonary connection; EC TCPC, extracardiac total cavopulmonary connection; LT TCPC, lateral tunnel total cavopulmonary connection.

Figure S4: Estimated survival rate (share of patients alive) in the years following completion of Fontan procedure, stratified by Fontan subtype; used as an input for generating the epidemiology model



Fontan surgical procedure	Survival rate, %						
	Time from Fontan procedure, years						
	0	10	15	20	25	30	35
AP	88.5*	81.0	77.7	74.3	61.4	48.5	31.0
LT-TCPC	93.4*	90.3	89.1	87.9	81.5	N/A	N/A
EC-TCPC	98.0*	93.6	94.0	93.0	86.2	N/A	N/A

***Calculated from 30-day mortality after the last Fontan surgery

AP, atriopulmonary connection; EC TCPC, extracardiac total cavopulmonary connection; LT TCPC, lateral tunnel total cavopulmonary connection; N/A, not available.

Figure S5: Distribution of age among persons previously palliated with a Fontan procedure between 2010–2020

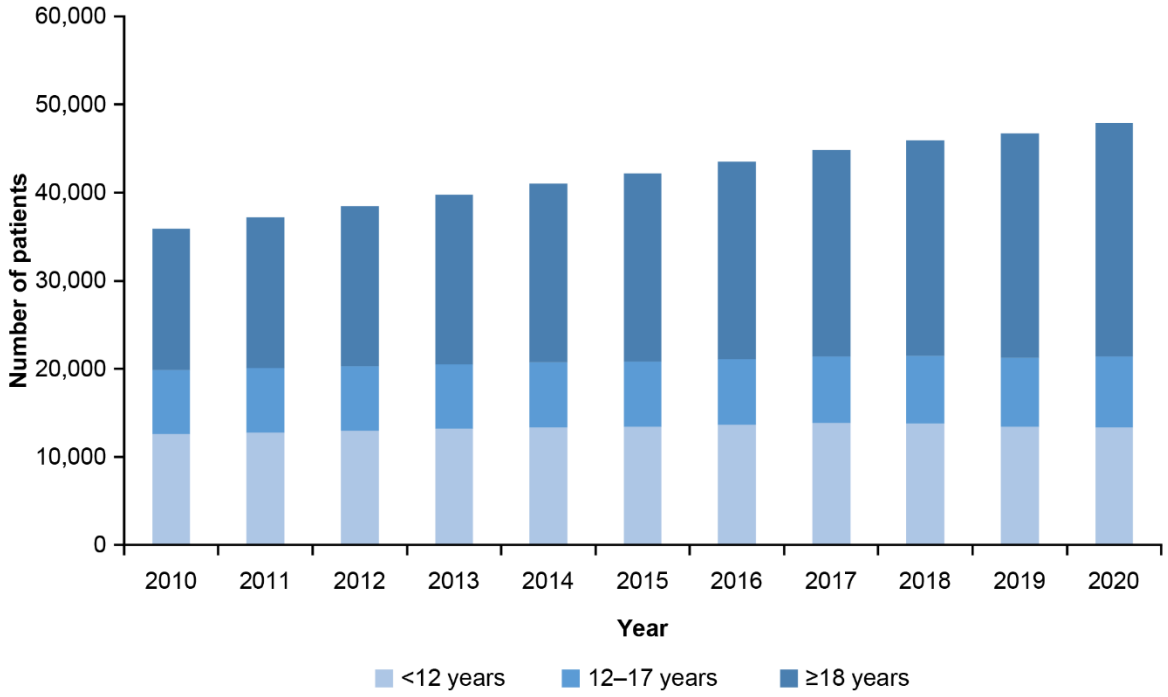


Table S1: Sources used for extraction of procedure codes and census data

Country	Database of extraction of procedure code	Database for validation and cross-checking procedure counts	Years of data available (years used in model)	Procedure codes
Australia and New Zealand	The Australia and New Zealand Fontan Registry (Kverneland *et al.* 2018)	N/A	2010–2018 (2014–2018)	N/A
France	CCAM procedure statistics	N/A	2015–2019 (2015–2019)	CCAM procedure coding (DFCA004: DZQJ002; DZQJ007; DZSA900; EQCF002; GELE001; YYYY062; YYYY189; YYYY595)
Germany	2014–2018 OPS code analysis 2014 and 2019 German heart surgery report-10%*	German Heart surgery report	2014–2019 (2015–2019)	>80% of codes were extracardiac conduit/tunnel; assumption made that any remaining AP procedures are coded as Sonstige
Italy	No sources available	-	-	-
The Netherlands	EACTS Congenital Database Zorgproducten statistics	Dx code 2535: partielle/ totale cavo/ pulmonal connection by specialist type cardio/ thoracic surgeons	2014–2018 (2014–2018)	Multiple as specialist societies can create their own procedure and diagnosis codes
Spain	CMBD dataset	N/A	2011–2015 (2011–2015)	**Up to 2015** (translation of US ICD-9-CM) [35.94] **2016** Change to ICD-10-PCS [02160JQ] **2018** Change to ICD-10-PCS, (re-coding) [06100JQ]
Sweden	NOMESCO (NCSP-S) statistics	N/A	2015–2019 (2015–2019)	NOMESCO (NCSP-S) coding allows clarification of intra/extra-cardiac approach and fenestration/shunt ±100% extracardiac TCPC [FAE40; FAE00; FAE10; FAE20; FAE30; FAE40; FAE50; FAE96]

Switzerland	CHOP Statistik	N/A	2014–2018 (2014–2018)	CHOP procedure coding based on old ICD-9-CM (same as Spain pre-2015) 1 clear code labelled as Fontan (35.94)
UK	OPCS England NCHDA	National congenital heart disease audit (NCHDA)	2016–2019 (2016–2019)	Multiple codes (K17.1–8); clear OPCS procedure coding; >85% extracardiac
US	Compile claims data	Akintoye 2019	2010–2019 (2014–2018)	Repair single ventricle; Repair modified Fontan; Bypass right atrium to right pulmonary artery with synthetic substitute, open approach

*For Germany's procedural codes, the average annual difference between GHSR and OPS code numbers was calculated.

Table S2: The number of Fontan procedures per year, obtained using Fontan procedural codes extracted from the relevant national databases

Country	Source	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019	Average number of procedures	Number of procedures (per 100,000 people per year)	Population size (2020 midyear estimate)
AUS/NZ	Australia and New Zealand Fontan registry	50	64	66	60	82	59	59	45	45	58	59	0.1935	30,391,000
CH	CHOP Statistik					24	18	18	13	17	18	18	0.2142	8,404,000
DE	Heart surgery report					215	254	186	240	200	219	219	0.2732	80,160,000
ES	CMBD dataset		55	56	73	50	34	54	53	53	49	53	0.1060	50,016,000
FR	CCAM procedure statistics						97	85	78	81	90	86	0.1270	67,848,000
IT	-											N/A	0.1060	62,403,000
NL	EACTS Congenital Database Zorgproducten statistics					29	36	26	26	20	27	27	0.1582	17,280,000
SE	NOMESCO (NCSP-S) statistics						17	24	24	25	16	21	0.2078	10,202,000
UK	OPCS/ NCHDA							179	157	171	174	170	0.2589	65,761,000
US	Compile claims data, Akintoye 2019	1,062	1,062	1,062	1,062	1,062	898	1,198	1,138	988	705	1,024	0.3078	332,639,000

AUS/NZ, Australia–New Zealand; DE, Germany; ES, Spain; FR, France; IT, Italy; NL, The Netherlands; SE, Sweden; UK, United Kingdom; US, United States.

Shading is indicative of unavailable values and subsequent estimation of data using averages of the real-world available data; to estimate the number of procedures in Italy, the data for Spain were applied.

Table S3: The numbers of patients alive in 2020 (A) and 2030 (B) stratified by age group and procedure type

A) 2020

	US	Europe	Australia/New Zealand	Total
Age group				
<12 years	7,815	4,598	449	**13,374**
12–17 years	4,726	2,721	259	**7,996**
≥18 years	14,860	9,867	755	**26,511**
Procedure type				
AP	2,270	1,657	113	**4,209**
LT-TCPC	4,740	3,085	248	**8,398**
EC-TCPC	20,390	12,443	1,102	**35,274**
Total	**27,401**	**17,186**	**1,463**	**47,881**

B) 2030

	US	Europe	Australia/New Zealand	Total
Age group				
<12 years	8,324	4,660	493	**14,009**
12–17 years	4,471	2,774	258	**7,805**
≥18 years	21,865	13,492	1,164	**37,963**
Procedure type				
AP	1,157	842	58	**2,143**
LT-TCPC	4,610	2,954	244	**8,122**
EC-TCPC	28,892	17,130	1,613	**49,512**
Total	**34,660**	**20,926**	**1,916**	**59,777**

AP, atriopulmonary connection; EC-TCPC, extracardiac total cavopulmonary connection; LT-TCPC, lateral tunnel total cavopulmonary connection; N/A, not available.