

SUPPLEMENTAL MATERIAL

Table S1. ICD9 and corresponding ICD10 codes.

Anomaly	ICD9	ICD10
Severe CHD	74500, 74510,	Q200, Q201,
	7452-7453,	Q203-Q204,
	7456, 74600,	Q212-Q213,
	7461-7463,	Q220, Q224-
	7465-7467,	Q226,Q230,
	7471, 74720,	Q232-Q234,
	74742	Q251-Q252, Q262
Common arterial truncus	74500	Q200
Double outlet right ventricle	no code	Q201
Transposition of great vessels	74510	Q203
Single ventricle	7453	Q204
Atrioventricular septal defect	7456	Q212
Tetralogy of Fallot	7452	Q213
Tricuspid atresia and stenosis	7461	Q224
Ebstein's anomaly	7462	Q225
Pulmonary valve atresia	74600	Q220
Aortic valve atresia/stenosis	7463 (no code for atresia)	Q230
Mitral valve anomalies	7465, 7466	Q232, Q233
Hypoplastic left heart	7467	Q234
Hypoplastic right heart	No code	Q226
Coarctation of aorta	7471	Q251
Aortic atresia/interrupted aortic arch	74720	Q252
Total anomalous pulm venous return	74742	Q262

Table S2. Comparison of isolated sCHDs versus all children with associated syndromes/anomalies. Data on the all anomaly

category combines data on both isolated CHD and those with associated anomalies/genetic syndromes. This comparison is presented in

Table S1. Data is presented as medians with 95 % confidence interval in brackets. Additionally I²-values are presented for a measure of heterogeneity.

Anomaly	Surgery coding	No. Of children with anomaly	<1 year	I ²	Year 1-4	I ²	Year 0-4	I ²	Median age first operation in weeks up to 5 years of age (95%CI)	I ²	Median number of operations up to 5 years of age (95% CI)	I ²
Severe Congenital Heart Defect (sCHD)	Any surgery	5693	78% (70-85)	97,7	36% (31-42)	91,2	88% (81-92)	97,2	3.6 (2.6-4.5)	87,5	3.2 (2.6-3.9)	97,7
Atrioventricular septal defect (AVSD)	Any surgery	453	58% (48-66)	66,5	47% (36-57)	63,5	82% (70-89)	76,6	21.6 (17.7-25.4)	9,8	3.0 (2.4-3.6)	83,3
AVSD	AVSD surgery	453	29% (22-36)	47,6	21% (14-28)	50,1	51% (41-60)	62,6	38.6 (33.0-44.1)	7,4	1.0 (1.0-1.0)	0,0
Transposition of great vessels (TGA)	Any surgery	980	90% (86-94)	65,9	31% (24-38)	72,7	94% (89-96)	67,1	1.1 (0.7-1.5)	94,5	3.9 (3.2-4.6)	94,6
Tetralogy of Fallot (TOF)	Any surgery	805	90% (83-94)	77,0	41% (33-49)	73,0	97% (95-98)	18,5	24.7 (21.1-28.3)	84,1	3.0 (2.4-3.6)	93,7
Pulmonary valve atresia (PA)	Cardiac surgery	245	84% (69-92)	56,1	56% (45-66)	20,3	92% (78-98)	63,8	0.9 (0.5-1.3)	49,0	3.8 (3.3-4.2)	0,0
Total anomalous pulmonary venous return (TAPVR)	Cardiac surgery	193	87% (75-93)	47,3	13% (8-20)	0,0	93% (81-97)	58,4	2.1 (0.9-3.2)	86,3	2.0 (1.7-2.4)	98,6
Coarctation of aorta (CoA)	Any surgery	1566	81% (73-87)	88,2	33% (27-38)	63,1	89% (82-93)	84,5	2.4 (1.8-3.1)	72,0	2.4 (2.0-2.9)	85,4
CoA	Surgery for coarctation	1566	66% (54-76)	93,3	7% (5-10)	29,5	73% (61-83)	93,8	2.7 (1.9-3.6)	77,1	1.0 (1.0-1.0)	0,0

Hypoplastic left heart syndrome (HLH)	Any surgery	476	90% (81-95)	54,1	71% (48-85)	76,9	97% (88-99)	65,0	0.9 (0.7-1.0)	55,9	5.3 (3.8-6.7)	92,1
HLH	Cardiac surgery	476	88% (76-95)	70,8	68% (44-83)	78,6	97% (86-99)	78,4	0.9 (0.7-1.0)	59,8	4.4 (3.1-5.6)	92,6
HLH	Blalock shunt	476	25% (8-47)	89,1	2% (0-9)	35,6	26% (8-49)	89,5	0.9 (0.5-1.4)	47,2	1.2 (0.8-1.6)	99,9
HLH	Hemi-fontan	476	60% (41-75)	80,7	19% (4-42)	88,5	67% (46-82)	81,4	25.4 (19.8-30.9)	84,5	1.1 (0.9-1.4)	100,0
HLH	Complete fontan	94	39% (27-52)	0,0	19% (8-33)	0,0	39% (13-64)	62,9	26.5 (0.4-52.7)	92,6	1.0 (1.0-1.0)	0,0
Hypoplastic right heart (HRH)	Cardiac surgery	62	95% (83-99)	17,1	52% (24-74)	34,9	98% (88-100)	33,7	0.3 (-0.0-0.7)	0,0	5.0 (3.4-6.5)	54,9
HRH	Blalock shunt	62	48% (26-68)	39,3	2% (0-12)	0,0	51% (27-70)	39,9	0.4 (-0.0-0.9)	61,4	1.0 (1.0-1.0)	0,0
HRH	Hemi-fontan	62	58% (41-72)	0,0	24% (10-42)	5,1	81% (65-90)	0,0	26.1 (19.3-32.8)	0,0	1.0 (1.0-1.0)	0,0
HRH	Complete fontan	20	31% (0-96)	45,6	23% (5-47)	0,0	28% (6-57)	11,6	213.2 (90.4-336.0)	0,0	1.0 (1.0-1.0)	0,0
Aortic valve atresia/stenosis	Any surgery	661	63% (49-74)	83,2	38% (26-49)	73,0	69% (57-78)	71,3	3.1 (1.4-4.8)	69,7	2.0 (1.3-2.8)	85,4
Double outlet right ventricle (DORV)	Cardiac surgery	226	89% (84-93)	0,0	52% (36-67)	61,4	97% (93-99)	19,7	5.1 (2.4-7.7)	82,4	4.1 (3.4-4.9)	68,5
Common arterial truncus	Cardiac surgery	106	76% (63-84)	0,0	36% (23-48)	0,0	94% (73-99)	54,7	2.6 (1.5-3.8)	67,7	3.1 (2.3-3.8)	62,2
Single ventricle	Any surgery	222	89% (75-96)	51,3	77% (40-93)	81,9	96% (88-99)	32,0	1.8 (1.3-2.3)	0,0	5.8 (4.3-7.4)	87,5
Tricuspid atresia and stenosis	Any surgery	318	80% (67-88)	51,8	55% (40-67)	47,3	86% (75-92)	43,2	2.7 (1.0-4.5)	73,1	5.0 (4.4-5.6)	0,0
Ebstein's anomaly	Any surgery	194	35% (19-52)	64,3	29% (21-37)	0,0	45% (31-58)	37,9	13.3 (-2.8-29.5)	99,1	2.3 (1.4-3.2)	8,4
Aortic valve atresia / stenosis	Any surgery	156	87% (75-93)	32,0	59% (42-73)	27,1	96% (88-99)	32,2	1.2 (1.1-1.3)	0,0	6.0 (4.3-7.7)	71,9

Table S3. Rules for surgery definitions.

Include as surgery	Not include as surgery
Dilatations with or without stent implantation	Examinations/diagnostic procedure
Removal of foreign bodies from bronchus, lungs and oesophagus	Removal of foreign bodies in open areas (nose, ear, throat, skin)
Drainage from internal organs	Drainage with easy access by needle
Extraction of multiple teeth	Extraction of one tooth only, other dental treatment
Application of internal and external fixation to bone	Closed manipulation/application traction of bones
Harvest of skin, bone, tendon	Removal of suture, tube
	Attention, irrigation, aspiration

Table S4. All cause mortality for children with sCHD's. Data aggregated from 13 European regions.

Anomaly	1 year survival (95% CI)	5 year survival (95% CI)
Severe Congenital Heart Defect (sCHD)	88.2% (86.1 – 90.3)	87.1% (84.7 – 89.5)
Atrioventricular Septal Defect (AVSD)	89.9% (86.3 – 93.6)	87.7% (83.3 – 92.3)
Transposition of the Great Arteries (TGA)	92.5% (89.8 – 95.3)	91.7% (88.5 – 95.0)
Tetralogy of Fallot (TOF)	97.6% (96.3 – 98.9)	96.7% (95.2 - 98.2)
Pulmonary Valve Atresia (PA)	80.0% (75.4-84.9)	76.2% (70.9-81.8)
Total Anomalous Pulmonary Vein Return (TAPVR)	80.0% (75.4-84.9)	76.2% (70.9-81.8)
Coarctation of the Aorta (CoA)	94.2% (92.3 – 96.2)	93.4% (91.2 – 95.7)
Hypoplastic Left Heart Syndrome (HLHS)	54.0% (46.8 – 62.3)	51.8% (45.0 – 59.6)
Hypoplastic Right Heart Syndrome (HLHS)	72.9% (64.7-82.2)	72.7% (64.4-82.0)

Aortic atresia / interrupte aortic arch	64.5% (52.8-78.8)	63.0% (51.3-77.5)
Double Outlet Right Ventricle (DORV)	82.9% (78.8-87.3)	79.5% (74.7-84.7)
Common arterial truncus	63.9% (57.3-71.3)	61.4% (55.0-68.5)
Single ventricle	75.8% (67.2-85.5)	72.1% (63.2-82.3)
Tricuspid atresia and stenosis	80.4% (76.3-84.7)	77.9% (73.3-82.9)
Ebstein's anomaly	81.0% (73.8 - 89.0)	78.9% (70.9 - 87.8)
Aortic valve atresia/stenosis	92.2% (89.3 - 95.2)	91.3% (88.0 - 94.6)

Table S5. All-cause mortality in the first year of life for children with either Transposition of the Great Arteries (TGA), Atrioventricular Septal Defect (AVSD), Tetralogy of Fallot (TOF) and all cases of severe Congenital Heart Disease (sCHD). The CI indicates confidence intervals.

Region name	Diagnosis	1-year mortality	Lower CI 95%	Upper CI 95%
Tuscany. Italy	TGA	5.8	2.2	14.7
IMER. Italy	TGA	8.0	3.7	16.9
Finland	TGA	11.3	7.2	17.6
Wales. UK	TGA	5.1	1.9	12.9
Thames. UK	TGA	12.0	6.4	21.8
Wessex. UK	TGA	5.9	2.5	13.5
Midlands. UK	TGA	16.0	11.3	22.3
Valencia. Spain	TGA	17.0	9.5	29.2
Tuscany. Italy	AVSD	9.7	3.2	27.1
IMER. Italy	AVSD	6.3	1.6	22.8
Finland	AVSD	7.1	3.0	16.3
Wales. UK	AVSD	20.9	11.8	35.4
Thames. UK	AVSD	16.0	6.3	37.2
Wessex. UK	AVSD	12.3	4.8	29.6
Midlands. UK	AVSD	15.5	8.9	26.2
Valencia. Spain	AVSD	9.8	4.2	22.0
Tuscany. Italy	TOF	3.6	0.9	13.5
IMER. Italy	TOF	2.6	0.7	10.0
Finland	TOF	1.7	0.4	6.8
Wales. UK	TOF	1.2	0.2	8.0
Thames. UK	TOF	8.6	3.7	19.4
Wessex. UK	TOF	3.0	0.8	11.6
Midlands. UK	TOF	4.6	2.2	9.4
Valencia. Spain	TOF	3.3	0.8	12.5
Funen. Denmark	sCHD	11.3	5.8	21.3

Tuscany. Italy	sCHD	10.9	7.8	15.0
IMER. Italy	sCHD	10.0	7.4	13.5
Finland	sCHD	6.9	5.6	8.4
Wales. UK	sCHD	9.6	7.5	12.3
Thames. UK	sCHD	15.3	11.7	19.8
Wessex. UK	sCHD	13.0	10.0	16.8
Midlands. UK	sCHD	14.8	12.5	17.4
Valencia. Spain	sCHD	12.9	10.3	16.3