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1 Outcomes of the arterial switch for transposition during infancy using a standardised

2 approach over 30 years

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Visual abstract 24

- Key question: What is the impact of morphological variations on early and late survival and 25
- 26 reintervention following arterial switch?
- 27 Key findings: Additional lesions and intramural coronary artery are important risk factors for
- early mortality and late reintervention 28

29 Take-home message: With a standardised approach, arterial switch can be performed with

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low mortality, moderate reintervention, and excellent long-term survival 30

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32 Abstract

33 *Objectives*: To describe the early and late outcomes of the arterial switch for transposition.

Methods: A single-centre retrospective cohort study was conducted to assess the early and late outcomes of arterial switch performed during infancy using a standardised institutional approach between 1988 and 2018, compared by morphological groups.

37 Results: 749 consecutive patients undergoing arterial switch during infancy were included, 464 (61.9%) with intact septum, 163 (21.8%) with isolated ventricular septal defect, and 122 38 (16.3%) with complex transposition with associated lesions, including 67 (8.9%) with Taussig-39 Bing anomaly. There were 34 early deaths (4.5%, 95% CI 3.1-6.1) with only 10 (2.6%) early 40 deaths since 2000. Complex morphology (OR 11.44, CI 4.76-27.43) and intramural coronary 41 artery (OR 5.17, CI 1.61-15.91) were identified as the most important risk factors for 90-day 42 mortality. Overall survival was 92.7% (95% CI 90.8-94.6) at 5 years and 91.9% (95% CI 89.9-43 94.1) at 20 years; in hospital survivors, there were 15 (2.1%) late deaths during a median 44 follow-up of 13.7 years. Cumulative incidence of surgical or catheter reintervention was 16.0% 45 (95% CI 14.5-17.5) at 5 years and 22.7% (95% CI 21.0-24.0) at 20 years; early and late 46 reinterventions were more common in the complex group, with no difference between the other 47 48 groups.

Conclusions: Using a standardised approach, the arterial switch can be performed with low
early mortality, moderate rates of reintervention, and excellent long-term survival.
Concomitant lesions were the most important risk factor for early death and were associated
with increased risk of late reintervention.

53 **Key words**: transposition, arterial switch, survival, reintervention.

55 Abbreviations

- CPB Cardiopulmonary Bypass 56
- ECLS Extracorporeal Life Support 57
- ICU **Intensive Care Unit** 58
- IVS Intact Ventricular Septum 59
- 60 ONS Office for National Statistics
- CEPTEDMANUSCR RACHS-1 Risk Adjustment for Congenital Heart Surgery 61
- TGA 62
- VSD 63
- 64

65 Introduction

66 Transposition is the most common cyanotic congenital heart defect presenting in neonates 67 and without surgical intervention, long-term survival is rare [1,2]. Since its introduction by Jatene in 1975 [3], the arterial switch has significantly improved the outcomes of children with 68 transposition and has become the procedure of choice. Advances in antenatal detection and 69 70 perioperative management [4], along with technical refinements including the Lecompte manoeuvre [5] and use of medially hinged trapdoor incisions for coronary transfer [6], have 71 led to low early mortality and excellent long-term survival [7-10]. Although technically 72 demanding, the standardised procedure is reproducible and within a mentoring framework, 73 can be taught to newly appointed surgeons without compromising outcomes [11]. 74

Morphological variations such as ventricular septal defect (VSD), obstruction in the ventricular outflow tract or aortic arch, and certain coronary artery patterns, increase the complexity of repair and have been associated with increased early mortality [12-15] but the impact on late outcomes remains uncertain. We conducted a retrospective analysis of infants with transposition treated with arterial switch at a single institution using a standardised approach over 30 years, to determine the impact of morphological variations on early and late survival and reintervention.

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83 Patients and methods

84 Ethics statement

This study was registered with Birmingham Women's and Children's Research & Development office (BWC/LA/Drury/10, 04/10/2021) and in accordance with UK National Research Ethics Service guidance, neither individual informed consent nor formal research ethics committee review was required as the study was undertaken by the direct clinical care team using information previously collected in the course of routine care.

90

91 Study population

All patients undergoing arterial switch during infancy at Birmingham Children's Hospital, UK
between January 1988 and December 2018 were included. Patients over one year of age at
arterial switch and those who underwent an alternative corrective procedure for complex
transposition were excluded (see Supplementary materials).

96

97 Operative technique

Our approach is to perform arterial switch in all patients with transposition, where it is technical feasible, within the first 2 weeks of life, unless there is an unrestricted VSD in which case we may leave longer to allow growth. In patients presenting up to 2 months, we still perform early arterial switch, with postoperative extra-corporeal life support (ECLS) if required [16]; beyond 2 months, we would consider initial pulmonary artery banding, but this is now rare in the UK.

The surgical technique for arterial switch used by all surgeons at our institution remained consistent throughout the study period [11]; for a detailed description, see Supplementary materials. In brief, the ascending aorta was transected and the coronary arteries excised with a generous cuff of aortic sinus tissue and mobilised. The resultant defects were repaired with

untreated autologous pericardium as a single patch. The main pulmonary artery was 107 transected at the same level as the aorta and coronary artery buttons relocated to medially 108 hinged trapdoor incisions using an open technique to construct the proximal neoaorta [6]. 109 Intramural coronary arteries were mobilised with generous cuffs of aortic wall, taking down the 110 111 valve commissure or laying opening the ostia as required. If unable to transfer as described above, a pericardial hood technique was used, with a bovine pericardium or homograft patch 112 to augment the receiving aortic sinus, incorporating the intramural cuff with minimal 113 mobilisation [17]. Lecompte manoeuvre was performed whenever possible [5], the neoaorta 114 reconstructed, the heart re-perfused, and reconstruction of the neopulmonary artery 115 completed while rewarming with the heart beating. Additional procedures were performed to 116 treat associated anomalies as required, a left atrial line was placed routinely, and primary 117 sternal closure was undertaken whenever feasible, or on the ICU as a delayed procedure, 118 119 usually within 24-48 hours.

120

121 Clinical variables and follow-up

Data were obtained from patient records and institutional databases, reviewed, and validated.
Morphological data were collected from detailed descriptions in the operative records and the
cohort divided into three groups:

- TGA-IVS: transposition with intact ventricular septum, including those in whom any VSD
 was deemed hemodynamically insignificant and therefore not closed,
- TGA-VSD: transposition with isolated ventricular septal defect(s),
- Complex transposition: associated anomalies such as aortic arch obstruction/interruption,
- ventricular outflow tract obstruction or Taussig-Bing.

Coronary artery pattern was described by sinus of origin, according to the Leiden convention
[18], and course around the great arteries [19]; Yacoub classification [20] was noted where
possible.

Date of arterial switch was set as baseline for follow-up. Early death and early reintervention were defined as occurring within 30 days. Potential risk factors were estimated for 90-day mortality. For late deaths, data were obtained from the UK Office for National Statistics (ONS); cause of death was obtained from hospital notes and/or post-mortem examination.

137

138 Statistical analysis

Analysis was performed using R version 3.6. Continuous variables are presented as median with interquartile range (IQR). Categorical variables are presented as frequencies and percentages. Comparisons between morphological groups were made using the Kruskal-Wallis tests for continuous variables, or Pearson's chi-squared test for categorical variables. Significance testing was two-sided with significance at p <0.05.</p>

Mortality was tracked using hospital attendance and ONS national tracing service. Patients 144 lost to follow-up were censored at the time last known to be alive. All-cause mortality was 145 estimated using the Kaplan-Meier method and comparisons made using the log-rank test. 146 Event rates for reinterventions were estimated using cumulative incidence function with death 147 as the competing risk. Patients were assessed as free from reintervention only if being 148 followed up locally, with out-of-region patients censored when last seen. To estimate risk 149 150 factors for all-cause mortality, a logistic regression model was developed using a Bayesian 151 method in R BRMS [21], with 90-day all-cause mortality as the outcome using sceptical prior distributions (see Supplementary materials). 95% compatibility intervals were derived from the 152 posterior distribution and presented. The first author and statistician had full access to all data 153 and take responsibility for its integrity and analysis. 154

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155 Results

156 Patients

During the study period, 776 patients underwent arterial switch, of whom 749 were under one year of age and included in the analysis (figure 1). Since 1988, the arterial switch has been our procedure of choice for transposition and other operations only performed when this approach was deemed unsuitable (see Supplementary materials).

161

162 Morphology

Of 749 infants undergoing arterial switch, 464 (61.9%) were diagnosed with TGA-IVS, 163 163 (21.8%) with TGA-VSD and 122 (16.3%) with complex transposition, including 67 (8.9%) with 164 Taussig-Bing. Baseline characteristics and operative data are summarised in table 1. 165 Coronary artery anatomy had the usual origin (1LOx-2R), course and branching pattern in 507 166 (67.7%) patients, originated from a single orifice in 37 (4.9%) and followed an intramural 167 course in 36 (4.8%). Non-usual coronary patterns were more frequent in the complex group 168 (62, 50.8%, p<0.001), with similar prevalence in the TGA-IVS and TGA-VSD groups (27.6% 169 and 31.9%, respectively, p=0.31). There was no difference between groups in the frequency 170 of an intramural coronary artery (p=0.23). Side-by-side great arteries were more common in 171 complex transposition (p<0.001) and typically associated with non-usual coronary patterns 172 173 (p<0.001). Detailed descriptions of the coronary patterns, including origin, course, and branching, and associated early mortality are shown in Supplementary materials. 174

175

176 Operative

Balloon atrial septostomy was performed in 532 (71.0%) patients. Thirty-one (4.1%) patients
underwent a surgical procedure prior to arterial switch, including isolated pulmonary artery

banding in 13 (1.7%), aortic arch repair with banding in 12 (1.6%), and systemic-pulmonary artery shunt +/- banding in four (0.5%), mostly performed elsewhere prior to referral in the early part of the series. Arterial switch was performed at a median age of 8 (IQR 5-16) days and beyond 28 days in 91 patients (12.1%).

Thirty-three (7.1%) patients in the TGA-IVS group had one or more small, hemodynamically insignificant VSDs identified on echo which were not closed, and often not found, during surgery. Three (2.5%) patients in the complex group had very large VSDs which were not closed at arterial switch, rather treated with concomitant neopulmonary artery banding.

187

188 Early outcomes

Nine (1.2%) infants required ECLS in the early postoperative period. Median length of stay on
ICU was three days (IQR 2-5) and length of hospital stay was nine days (IQR 7-13); both were
longer in the complex group (p<0.001).

There were 34 deaths within 30 days, with an overall early mortality of 4.5% (95% CI 3.1-6.1). 192 Early mortality was higher in the complex group (20, 16.4%, p<0.001) but similar between 193 other groups: 8 (1.7%) in TGA-IVS and 6 (3.7%) in TGA-VSD (p=0.21); there was no difference 194 between those who underwent arterial switch before or after 28 days (p=0.29). There were 12 195 intraoperative deaths but none since 1997. Early death was more common in the first four 196 years (10/136, 7.4%) than in subsequent years (24/613, 3.9%) (Supplementary Figure 2), and 197 has been low since 2000 (10/380,2.6%), with only one death (1/215,0.5%) amongst infants 198 199 with non-complex transposition and the usual coronary pattern. A further nine deaths occurred 200 during the index admission due to concomitant conditions or complications of prolonged ICU 201 stay, with an overall 90-day mortality of 5.7%.

Risk factors for 90-day mortality are shown in table 2 and figure 2. Complex morphology (OR
11.44,Cl 4.76-27.43) and intramural coronary artery (OR 5.17,Cl 1.61-15.91) were the most

important factors, whilst for isolated VSD (OR 1.94,Cl 0.73-5.06), single coronary orifice (OR
1.75,Cl 0.43-6.56) and balloon atrial septostomy (OR 0.71,Cl 0.31-1.66), the compatibility
intervals were wide so any association could not be excluded. Side-by-side great arteries were
associated with reduced risk of death compared with aorta anterior to the pulmonary artery
(OR 0.24,Cl 0.04-0.91). Overall, the model had a C-index of 0.85.

209 Early surgical reintervention was required in 32 (4.3%) patients (15 TGA-IVS, 3 TGA-VSD, 14 210 complex) and was more frequent in the complex group (p<0.001) with no difference between the other groups (p=0.59). These were most commonly coronary revision (6,0.8%, including 211 3 intramurals), pulmonary artery repair (4,0.5%), residual VSD closure (3,0.4%), epicardial 212 pacemaker implantation for heart block following VSD closure (3,0.4%) or aortic arch repair 213 (2,0.3%). Emergency takedown was performed in one (0.1%) patient for deteriorating 214 ventricular function not responding to conventional therapy. There were two (0.3%) 215 transvenous pacemakers but no other early catheter reinterventions, as per our departmental 216 policy. 217

Of the three patients with a large VSD who underwent neopulmonary artery banding, one died in the early postoperative period, one was subsequently septated, and the other was deemed unseptatable, undergoing single ventricle palliation and excluded from subsequent analysis.

221

222 Late survival

Following discharge, during a median follow-up of 13.7 years (IQR 3.8-21.3), there were 15 late deaths (2.1% of hospital survivors) at a median of 4.0 years (IQR 1.5-15.0), seven (1.5%) with TGA-IVS, five (3.1%) with TGA-VSD and three (2.5%) with complex transposition. Four patients died suddenly at home, four during or soon after reintervention, two from chronic heart failure (one whilst awaiting transplantation, one from cardiac allograft vasculopathy), one from unrelated septicaemia, and in four, the mode of death is unknown. All late deaths occurred in those with either the usual coronary pattern (1LCx-2R), circumflex from the right (1L-2CxR), or inverted circumflex/right (1RL-2Cx); none had an intramural, single orifice or interarterial
course. The incidence of sudden unexpected death in hospital survivors was 0.04% per year
of follow-up.

Of 706 hospital survivors, late outcomes were available for 621 (88.0%). Overall survival was 93.8% (95%CI 92.1-95.6) at 1 year, 92.7% (95%CI 90.8-94.6) at 5 years, 92.5% (95%CI 90.6-94.5) at 10 years, and 91.9% (95%CI 89.9-94.1) at 20 years. At latest follow-up, 446 (96.1%) with TGA-IVS, 150 (92.0%) with TGA-VSD, and 96 (78.7%) with complex transposition were alive (figure 1). Survival by group is shown in figure 3 with additional data in Supplementary materials.

239

240 Late surgical and catheter reinterventions

Following discharge, late surgical or catheter reinterventions were required in 118 (16.7%) 241 survivors (Supplementary Table 3). Eighty-two late surgical reoperations were performed in 242 66 (8.8%) patients (range 1-3), most often pulmonary artery patching (37 procedures in 34 243 patients,4.5%). Neoaortic valve/root repair/replacement has been performed in 12 (1.6%) 244 patients at a median of 13.2 (IQR 9.0-16.4) years. Late coronary reintervention was required 245 in 4 (0.5%) patients (three usual arrangement, one intramural [22]), two within the first year 246 and the others at 10 and 26 years, either button reimplantation (3) or coronary artery bypass 247 248 grafting (1). Late reoperation was more common in the complex group (25 procedures in 19 patients 15.6%, p=0.008) but similar in the TGA-IVS (39 in 32,6.9%) and TGA-VSD (18 in 249 15,9.2%) groups (p=0.39). 250

One hundred and seventy-three late catheter interventions were performed in 87 (12.3%) survivors (range 1-8). Most reinterventions were ballooning/stenting of the branch pulmonary arteries (138 procedures in 68 patients,9.1%), with ballooning of the neopulmonary valve in 12 (1.6%) and recoarctation in 6 (0.8%). Late catheter reintervention was more common in the complex group (50 procedures in 23 patients,18.9%,p=0.013) but similar in the TGA-IVS

256 (94 in 47,10.1%) and TGA-VSD (29 in 17,10.4%) groups (p=1.0).

- 257 The cumulative incidence of surgical and/or catheter reintervention censored for death was
- 258 11.4% (95%Cl 10.0-13.0) at 1 year, 16.0% (95%Cl 14.5-17.5) at 5 years, 17.8% (95%Cl 16.3-
- 19.4) at 10 years, and 22.7% (95%Cl 21.0-24.0) at 20 years. The risks of reintervention by

260 group are shown in figure 4 and Supplementary materials.

262 Discussion

263 In this study, we demonstrate that the arterial switch can be performed using a reliable 264 operative technique with consistently low early mortality, moderate rates of surgical and catheter reintervention, and excellent late survival. Our overall early mortality of 2.6% since 265 2000 compares favourably with other contemporary series [7,8,14,23]. However, patients in 266 the complex group, with associated coarctation, aortic interruption, ventricular outflow tract 267 obstruction or Taussig-Bing, and those with an intramural coronary artery were at greater risk 268 of early death. Complex transposition was also associated with increased need for 269 reintervention, but hospital survivors had similar long-term survival, 270

271

272 Risk factors for early mortality

The most important risk factors for early death were complex morphology and intramural 273 coronary artery; the latter remained associated with outcome despite a low event rate in the 274 non-complex groups. It is uncertain whether patients with a single coronary orifice had 275 increased risk of death as the compatibility interval was wide. Dealing with variations in 276 coronary anatomy is the key technical challenge of the arterial switch whilst repair of 277 associated lesions in the aortic arch or outflow tracts has greater impact on ischemic and 278 circulatory arrest times. In a meta-analysis of studies reporting patients undergoing arterial 279 switch before 2000, Pasquali et al found that intramural and single ostium coronary patterns 280 were associated with increased mortality [15]. More recently, Metton et al reported that 281 intramural patterns remain associated with early mortality in the current era [24] whilst Fricke 282 283 et al identified left ventricular outflow tract obstruction, aortic arch obstruction, and weight 284 <2.5kg as risk factors for early death, with no deaths amongst those with an intramural pattern [7]. Age at arterial switch and balloon atrial septostomy have also been identified as risk factors 285 [25] but neither were apparent in our series. Unexpectedly, we found that side-by-side great 286

arteries were associated with reduced risk of death, compared with aorta anterior to thepulmonary artery across all groups, but the reason for this is unclear.

A VSD has been identified to increase the risk of arterial switch, in both RACHS-1 and Aristotle risk-adjustment models [12,13]. Data from the European Congenital Heart Surgeons Association multi-institutional study identified VSD as the most important risk factor for early mortality [14] but may reflect the higher incidence of other associated lesions in this group, which we have classified as complex transposition. We found that an isolated VSD was less important than other factors in predicting the risk of early death.

295

296 Reintervention and late outcomes

Early surgical reintervention was required in 4.3% of patients. Late catheter and/or surgical reinterventions were performed in 15.8% of patients, comparable with previous series [8,9,26]. In this and other series, the most common reason for late reintervention was branch pulmonary artery stenosis, reflecting a proactive approach to dealing with recurrent lesions. Both early and late reinterventions were more common in those with complex transposition but unlike some other reports [8,26], an isolated VSD did not increase the risk of late reintervention.

We have previously reported on the fate of the neoaortic valve in the first decade of this cohort 303 with a 97.7% freedom from aortic valve reoperation during childhood [27]. In the current series, 304 305 neoaortic valve or root reoperation was performed in 1.6% of patients at a median of 13.2 years, which is similar to other large series: 2.7% at 13.3 years at the Mayo Clinic [9] and 4.0% 306 307 at 14.5 years in Melbourne, increasing over time [7,26]. Furthermore, early or late coronary 308 reintervention was required in ten (1.3%) patients, of whom four had an intramural and/or 309 interarterial course. No deaths beyond 30 days occurred in those with an intramural, single 310 orifice or interarterial coronary pattern. This low rate of late coronary-related complications following arterial switch using the trapdoor technique [6], or pericardial hood [17] when 311 312 required, has similarly been reported by the Melbourne group [26], in contrast to other large

series [8]. The standardised trapdoor technique therefore minimises coronary reinterventions
without an associated increase in neoaortic reoperations within the current extent of followup.

316

317 Limitations

Our findings are subject to the limitations inherent to retrospective cohort studies. Detailed 318 descriptions of morphology were obtained from contemporaneous operation notes, without 319 reference to echocardiographic or angiographic studies. We only included patients deemed 320 suitable for arterial switch and therefore patients with more complex patterns were excluded. 321 Data are limited to a single, high-volume institution with one consistent operative technique 322 used by all surgeons throughout the study period which may limit its generalisability. Our low 323 event rate for early mortality also limited the assessment of potential risk factors. In the early 324 years, many patients with complex anatomy were referred from elsewhere in the UK and 325 Europe, often following an initial palliative procedure, which may have contributed to a higher 326 throughput and complexity in this period (see Supplementary materials). 327

328

CCF.

The arterial switch has revolutionised the natural history of transposition. We demonstrate that 330 331 it can be performed with low early mortality, moderate rates of reintervention, and excellent long-term survival, using a standardised institutional technique throughout the series which 332 we have previously shown to be reproducible and suitable for mentoring to avoid a learning 333 curve [11]. Complex transposition with concomitant lesions and an intramural coronary artery 334 were independently associated with increased early mortality but with a low event rate, an 335 isolated VSD and other coronary patterns had less impact on survival. As this cohort enters 336 their fourth decade, the very late outcomes of arterial switch including the fate of the neoaortic 337 root, are not yet known. These patients require long-term follow-up to determine the ongoing 338 burden of disease and need for reintervention during adulthood. 339

340

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344

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348

Data Availability Statement: The data underlying this article cannot be shared publicly to maintain the confidentiality of the individuals involved. The anonymised data may be shared on reasonable request to the corresponding author.

352

353 **Conflicts of interest**: none.

354

355 Author contributions:

Drury: conceptualisation, data curation, formal analysis, investigation, methodology, project 356 administration, validation, writing-original draft. Mussa: conceptualisation, investigation, 357 writing-review/editing. Stickley: data curation, formal analysis, methodology, software, 358 visualisation, writing-review/editing. Stumper: resources, writing-review/editing. Crucean: 359 validation, writing-review/editing. Dhillon: resources, writing-review/editing. Seale: resources, 360 writing-review/editing. Botha: resources, writing-review/editing. Khan: resources, writing-361 review/editing. Barron: resources, supervision, writing-review/editing. Brawn: resources, 362 writing-review/editing. Jones: conceptualisation, data curation, investigation, resources, 363 364 supervision, writing-review/editing.

365 Figure legends

366

367 **Central image**. Risk factors for 90-day survival following arterial switch.

368

- **Figure 1**. Flow diagram of children undergoing surgery for transposition at our institution. REV,
- 370 Réparation à l'Etage Ventriculaire; TGA, transposition.

371

Figure 2. Forest plot of potential risk factors for 90-day mortality. The central line in each box is the mean estimate, the width of the box represents the central 50% and the whiskers corresponds to the 95% credible interval of the posterior distribution. TGA, transposition; VSD, ventricular septal defect.

376

Figure 3. Kaplan-Meier curves showing survival following arterial switch, by morphologicalgroup.

379

Figure 4. Cumulative incidence function plots for any reintervention, by morphological group:
A) transposition with intact ventricular septum (TGA-IVS), B) transposition with ventricular
septal defect (TGA-VSD), C) complex transposition.

Table 1. Patient characteristics and operative data, by morphological group.

Characteristic	Overall n=749	TGA-IVS n=464 (61.9%)	TGA-VSD n=163 (21.8%)	Complex TGA n=122 (16.3%)	p value
Age, days	8 (5-16)	7 (5-12)	12 (6.5-23)	10.5 (5-39)	<0.001
Weight, kg	3.4 (3.0-3.8)	3.4 (3.0-3.8)	3.4 (3.0-3.7)	3.4 (3.1-3.9)	0.40
Male, n (%)	531 (70.9)	327 (70.5)	120 (73.6)	84 (68.9)	0.65
Associated anomalies, n (%)					<0.001
VSD	305 (40.7)	33 (7.1) ^a	163 (100)	109 (89.3)	<0.001
LVOTO	9 (1.2)	0	0	9 (7.4)	<0.001
RVOTO	23 (3.1)	0	0	23 (18.9)	<0.001
Coarctation of the aorta	86 (11.5)	0	0	86 (70.5)	<0.001
Interrupted aortic arch	10 (1.3)	0	0	10 (8.2)	<0.001
Taussig-Bing	67 (8.9)	0	0	67 (54.9)	<0.001
Coronary artery origins, n (%) ^b					<0.001
1LCx-2R	514 (68.6)	339 (73.1)	112 (68,7)	63 (51.6)	
Usual position in sinuses (A)	507 (67.7)	336 (72.4)	111 (68.1)	60 (49.2)	
Adjacent to commissure (C)	7 (0.9)	3 (0.6)	1 (0.6)	3 (2.5)	
1L-2CxR (D)	108 (14.4)	66 (14.2)	19 (11.7)	23 (18.9)	
1RL-2Cx (E)	60 (8.0)	24 (5.2)	16 (9.8)	20 (16.4)	
1R-2LCx (E)	4 (0.5)	1 (0.2)	1 (0.6)	2 (1.6)	
Sinus 1 only, including 1RLCx	17 (2.3)	10 (2.2)	2 (1.2)	5 (4.1)	
Sinus 2 only, including 2LCxR $^\circ$	46 (6.1)	24 (5.2)	13 (8.0)	9 (7.4)	
Intramural coronary origin(s)	36 (4.8)	24 (5.2)	4 (2.5)	8 (6.6)	0.23
Single coronary orifice (B) ^b	37 (4.9)	16 (3.4)	11 (6.7)	10 (8.2)	0.048
Aorta-PA alignment, n (%)					<0.001
Aorta anterior & left of PA	30 (4.0)	23 (5.0)	3 (1.8)	4 (3.3)	
Aorta anterior to PA	459 (61.3)	311 (67.0)	99 (60.7)	49 (40.2)	
Aorta anterior & right of PA	218 (29.1)	124 (26.7)	51 (31.3)	43 (35.2)	
Side by side	42 (5.6)	6 (1.3)	10 (6.1)	26 (21.3)	
Balloon atrial septostomy, n (%)	532 (71.0)	395 (85.1)	99 (60.7)	38 (31.1)	<0.001
Additional procedure, n (%)					
VSD closure	269 (35.9)	0	163 (100)	106 (86.9)	<0.001
Aortic arch repair	84 (11.2)	0	0	84 (68.9)	<0.001
Relief of outflow tract obstruction	31 (4.1)	0	0	31 (25.4)	<0.001
Neo-PA banding, VSD not closed	3 (0.4)	0	0	3 (2.5)	N/A
LIMA-LAD coronary bypass graft	1 (0.1)	0	1 (0.6)	0	N/A
Other procedure	6 (0.8)	0	0	6 (4.9)	N/A
CPB time, minutes	129 (107-155)	115.5 (101-136)	140 (122-160.5)	164 (149-208)	<0.001
AXC time, minutes	76 (62-95)	67 (58-81)	85 (73-98.5)	110.5 (97-129)	<0.001
DHCA used, n (%) d	613 (81.8)	415 (89.4)	97 (59.5)	101 (82.8)	<0.001
DHCA time, minutes	8 (6-20)	7 (6-10)	20 (5-28)	27.5 (10-48)	<0.001
ECLS post-CPB, n (%)	9 (1.2)	5 (1.1)	0	4 (3.3)	0.058

ICU length of stay, days	3 (2-5)	3 (2-5)	4 (2-6)	4 (3-7)	<0.001
Hospital length of stay, days	9 (7-13)	9 (7-12)	9.5 (7-16)	11 (7-19.5)	0.001
30-day mortality, n (%)	34 (4.5)	8 (1.7)	6 (3.7)	20 (16.4)	<0.001
90-day mortality, n (%)	43 (5.7)	11 (2.4)	8 (4.9)	24 (19.7)	<0.001

386

AXC, aortic cross-clamp; CPB, cardiopulmonary bypass; DHCA, deep hypothermic circulatory arrest;
ECLS, extra-corporeal life support; ICU, intensive care unit; IVS, intact ventricular septum; LAD, left
anterior descending coronary artery; LIMA, left internal mammary artery; LVOTO, left ventricular outflow
tract obstruction; PA, pulmonary artery; RVOTO, right ventricular outflow tract obstruction; TGA,
transposition; VSD, ventricular septal defect.
Small, hemodynamically insignificant, not closed.

^b According to Leiden convention, with Yacoub classification in parentheses, where possible.

^c Details of origin, course, and branching patterns in Supplementary Table 2.

^d With use of single venous cannula (see Supplementary materials).

CCE

Parameter	Odds ratio	Lower 2.5% CI	Upper 97.5% CI
TGA-IVS			
TGA-VSD	1.94	0.73	5.06
Complex TGA	11.44	4.76	27.43
Usual coronary arrangement, 1LCx-2R			21
Non-usual coronary arrangement,	0.82	0.34	1.85
other than 1LCx-2R		S	
No intramural coronary artery	~		
Intramural coronary artery	5.17	1.61	15.91
More than one coronary orifice			
Single coronary orifice	1.75	0.43	6.56
Aorta anterior to PA			
Side-by-side great arteries	0.24	0.04	0.91
No balloon atrial septostomy			
Balloon atrial septostomy	0.71	0.31	1.66

Table 2. Risk factors for 90-day mortality using a multivariable Bayesian analysis.

Year of surgery and age at surgery were also included in the model (see Supplementary Figures 2 and
3). IVS, intact ventricular septum; PA, pulmonary artery; TGA, transposition; VSD, ventricular septal
defect.

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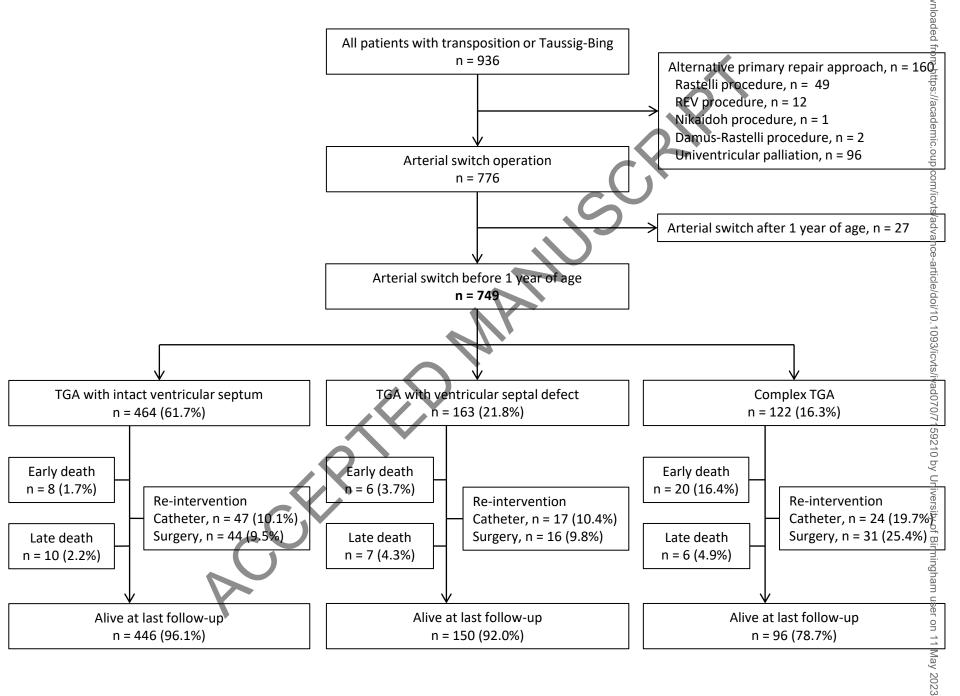
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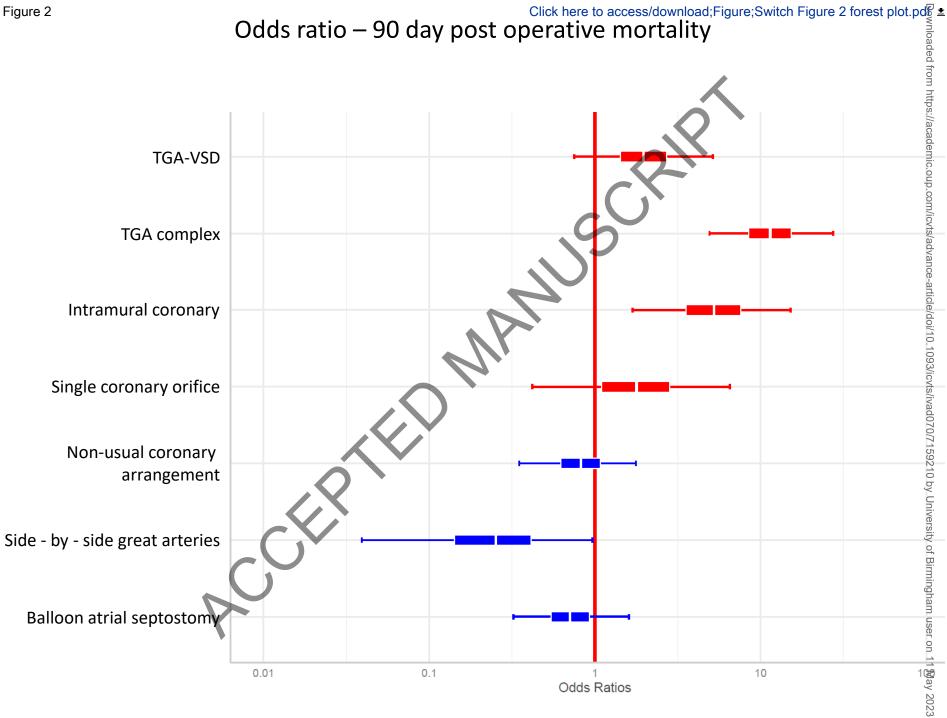
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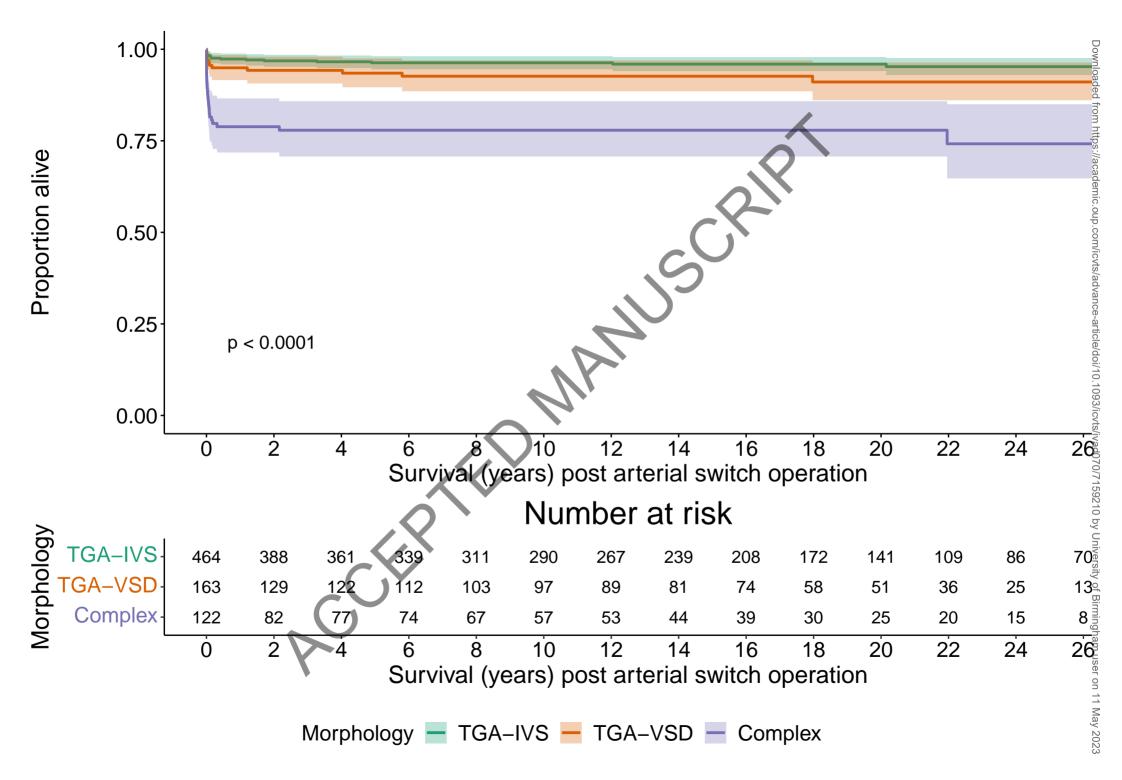


Figure 4

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