Progress in Late Results Among Pediatric Cardiac Surgery Patients:
A Population-Based Six-Decade Study with 98% Follow-Up

Running title: Raissadati et al.; Late Results after Pediatric Cardiac Surgery

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Abstract

Background—Surgical treatment of congenital cardiac defects in Finland started over 60 years ago. We analyzed the survival of all the pediatric cardiac surgery patients operated on before 2010.

Methods and Results—Data was obtained retrospectively from a pediatric cardiac surgery database. Patient status was received from the Finnish population registry. Survival was determined using the Kaplan–Meier method, and the survival rate was compared to a sex- and age-matched general population. Between 1953 and 2009, 13,786 cardiac operations were performed on 10,964 pediatric patients in Finland. Follow-up coverage was 98%. The 60-year survival for the entire study was 70% versus 86% for the general population. The number and proportion of severe cardiac defects increased in the 2000s. The long-term survival of patients with severe defects improved significantly across decades. For instance the 22-year survival of patients with transposition of the great arteries operated on 1953–89 and 1990–2009 improved from 71% to 93% (hazard ratio for death, 0.29; 95% confidence interval [CI], 0.17 to 0.49; P<0.0001), respectively. The mean patient age at operation decreased from 8.9 to 2.2 years (95% CI, 6.2 to 7.1, P<0.0001). The early mortality of patients decreased from a maximum of 7% in the 1970s to 3% in the 2000s (95% CI, 0.05 to 0.08, P<0.0001).

Conclusions—Patients are diagnosed and treated at an increasingly younger age. Advanced diagnostics, surgical methods and post-operative intensive care have led to substantial improvements in both early- and late results among pediatric cardiac surgery patients.

Key words: congenital cardiac defect, cardiac surgery, cardiology, pediatric, survival, Pediatric cardiac surgery, mortality, true survival
Introduction

Although many studies have been published on pediatric cardiac surgery results, most have been limited to operations from a single center, one type of defect, a small patient population, a short follow-up time, or a limited follow-up coverage.1–6

Systematic screening of children and newborns in the Finnish healthcare system has secured exceptional accuracy in detecting and diagnosing pediatric cardiac defects. All children are treated equally regardless of socioeconomic status or place of residence. The nationwide Finnish Population Registry allows for a unique and comprehensive follow-up of practically all patients (98%).

The aim of this study was to establish the post-operative mortality and long-term survival of all pediatric patients (≤15 years of age) operated on for cardiac defects 1953–2009 in Finland. This population-based study includes 10,964 patients and 13,786 pediatric cardiac operations during 60 years of follow-up. To evaluate progress in patient care, we compared the results of operations performed across decades.

Materials and methods

Patients and data collection

We obtained patient and operational data from the custom-built ProCardio version 11 database: the Research Registry of Pediatric Cardiac Surgery (Melba Group, Helsinki, Finland) running on Filemaker Pro version 11.0v1 (Filemaker Inc, CA, USA). The database contains the records for all pediatric patients that have undergone surgery for congenital heart defects in Finland. All pediatric cardiac operations were performed at five university hospitals (Helsinki, Turku, Tampere, Oulu, and Kuopio) and one district hospital (Aurora Hospital, Helsinki) in Finland.
Since 1997 all operations in Finland have been centralized to- and performed in Helsinki university children’s hospital. We received status and date of death or emigration of patients from the Finnish Population Registry.

This study includes all operations performed between 1953 and 2009 in Finland. The only procedures excluded were patent ductus arteriosus closures in children less than one month old, due to the vast majority of their deaths being caused by prematurity. We defined children less than 15 years of age as pediatric patients.

Diagnoses were arranged hierarchically according to the severity of the defect (Table 1) as published previously. Each patient was assigned one primary diagnosis to facilitate data comparison and statistical analysis. In patients with several congenital heart defects, we chose the hierarchically higher one.

**Mortality and survival**

All patient deaths within 30 days after the operation were defined as early operative mortalities. Early mortality after the first operation for patent ductus arteriosus (PDA), atrial septal defect (ASD), coarctation of aorta (COA), ventricular septal defect (VSD), tetralogy of Fallot (TOF), transposition of the great arteries (TGA), hypoplastic left heart syndrome (HLHS), and univentricular heart (UVH) were analyzed separately. All other diagnoses were categorized as miscellaneous (misc).

Long-term follow-up started immediately after the patient’s first operation and ended at the patient’s death or October 24, 2012. For patients who emigrated, the day of emigration was marked as the last day of follow-up. Separate survival curves were created for the six most severe defects operated on 1990–2009. The follow-up for these patients excluded early mortality. Each individual patient survival was compared with that of the age-, time-, and gender-matched
population.

This research was conducted with permission from the Ministry of Social Affairs and Health and the Institutional review board and Ethical committee of Helsinki university Hospital for Children and Adolescents.

Statistics

Survival statistics and Kaplan–Meier data were obtained and analyzed using R-program and IBM SPSS Statistics version 21.0 (SPSS, Inc., Chicago, IL).

The non-parametric Mann–Whitney test was used for two-group comparisons and the one-way ANOVA with least significant difference (LSD) post-hoc test was used for comparisons between the means of multiple groups. The logrank test was used to analyze differences in the survival rates of patients operated on at different times. The Chi Square test was used to obtain \( P \) values for non-proportional hazards and to compare the number of early operative mortalities between different decades of operation. We estimated patient survival with the Kaplan Meier method. The proportional hazards assumption was tested by inspecting \( \log(-\log(\text{survival})) \) plots and by plotting Schoenberg residuals over time. Variables that failed both tests were deemed non-proportional. The univariate Cox regression model was used to estimate the hazard ratio (HR) for death in patients operated on during different decades. The univariate Cox regression model with time-dependent covariates was used to estimate the hazard ratio (HR) for death when proportionality was not met. For this model, the cutoff point was chosen manually according to where the hazards crossed, and hazard ratios reported before and after that specific point in time. \( P \) values less than 0.05 were considered significant.

Results
Patients

A total of 13,786 operations were performed on 10,964 patients during 56 years (1953–2009) of pediatric cardiac surgery in Finland. The study period ended on October 24, 2012. Follow-up was complete in 98% of patients, and ended prematurely for 177 patients (1.6%) who emigrated. Only 86 patients (0.4%) and 90 operations were excluded from analyzes due to insufficient patient and surgical data. Thus, 99.6% of all patients were included in the survival analysis.

The gender distribution was male-dominant in nearly all groups. The proportion of male patients fell between 56–67% in the more severe defect groups of TOF, TGA, HLHS, and UVH, with the TGA group having highest proportion of male patients.

Patient age at operation

Both the median and mean age at operation have decreased significantly in all diagnostic groups. Mean age at operation was 8.9 years in the 1950s gradually decreasing to 2.2 years in the 2000s (95% CI, 6.2 to 7.1, P<0.0001 for the decrease in age; Table 2, Figure 1). Similarly, the median patient age at operation has decreased from 9.0 years in the 1950s to 0.5 years in the 2000s.

The age at which patients underwent their first operation decreased substantially in all diagnostic groups, especially in the groups with severe cardiac defects (P<0.0001 for all diagnoses; Table 3). Patients with TGA, HLHS, and UVH underwent their first operation at 28, 7, and 153 days of age, respectively, 1990–2009 (Table 3).

Operations and early mortality (<30 days after the operation)

The number of operations for severe cardiac defects increased throughout the follow-up years (Table 4). Practically no HLHS patients were operated on before 1995. In the 2000s, surgical palliation of HLHS accounted for 10% of all operations performed (Table 4). The number of operations for minor defects decreased.
The average early mortality (<30 days) for all operations conducted 1953–2009 was 6% (Tables 2 and 3). Nearly all patients operated on for PDA, ASD, and COA survived their procedures, even those operated during earlier decades. For other diagnosis groups, the operative mortality decreased significantly across years (Table 3). The operative mortality of TGA operations was 13% 1953–89 and only 4% in the 2000s ($P<0.0001$; Table 3). Operative mortality of HLHS decreased from 79% 1953–89 to 7% in the 2000s ($P<0.0001$; Table 3). Similarly, the early mortality of UVH operations decreased from 20% for those conducted 1953–89 to 5% in the 2000s for all surgical stages of UVH palliation ($P<0.0001$; Table 3).

**Survival**

The vast majority of patients (8976 or 82%) were alive on the last day of the study with the follow-up time extending up to 60 years. The mean age of living patients was 30 years, ranging from 3 to 73 years. The average length of follow-up time was 25 years, ranging from 2 to 60 years (Table 5). Eleven percent (1115) of patients that survived their first operation had died. The average follow-up time for these patients was 14 years (range of 0.3 to 55 years). Patients with severe cardiac defects who survived their first operation died on average at a much younger age, e.g., 0.8 years in the HLHS group versus 30 years in the PDA or ASD group.

At the end of the 60-year follow-up period, the survival rate was 86% for the general population and 70% for patients (Figure 2A). After excluding operative mortality, the study group survival increased to 72%.

Patients with surgically corrected ASD or PDA had on average a normal lifespan (Figure 2A, Table 5). The 50-year survival for COA, VSD, and TOF patients was 70%, 77%, and 53%, respectively (Figure 2A, Table 5). Patients in the TGA and UVH group had a 45-year survival rate of 59% and 33%, respectively. There was a significant decrease in the survival curve for
TGA patients at 40 years of follow-up from 66% to 59%, which was caused by a single death event. Additionally, patients operated on for TOF had a sharp decrease in survival rate — from 58% to 53% — at 49 years, caused by only four deaths: two accidental, one related to Down’s syndrome, and one case of severe retardation. HLHS patients had the shortest follow-up time of 17 years with a 15-year survival rate of 53%.

In order to measure progress in late results, we compared the long-term survival (without early mortality) of patients operated on 1953–89 and 1990–2009 (Figures 2B and 3A-F). We found that the long-term survival of patients operated on for VSD 1990–2009 was significantly higher than those operated on 1953–89, with 95% and 87% survival at 22 years, respectively (HR for death 1990–2009, 0.36; 95% CI, 0.24 to 0.54; \( P<0.0001 \) for logrank test; Figure 3B). Moreover, patients operated on for TOF 1990–2009 had significantly improved late results compared to those operated on during 1953–89, with 90% versus 87%, respectively, at 22 years after their operation (HR for death 1990–2009, 0.52; 95% CI, 0.28 to 0.94; \( P=0.037 \) for logrank test; Figure 3C). The long-term survival rate of patients operated on for TGA 1990–2009 was also significantly higher than those operated on 1953–89 with values of 93% and 71% at 22 years, respectively (HR for death 1990–2009, 0.29; 95% CI, 0.17 to 0.49; \( P<0.0001 \) for logrank test; Figure 3D). Patients with simple TGA had a long-term survival rate of 99% at 22 years (Figure 3D). The long-term survival of patients that underwent surgery for UVH improved significantly only after the eighth post-operative year for those operated on 1990–2009 (HR for death 1990–2009, 0.17; 95% CI, 0.06 to 0.50; \( P=0.001 \) for Chi square test; Figure 3E).

HLHS was a practically inoperable cardiac defect before the latter half of the 1990s, rendering the comparison of patient survival 1953–89 and 1990–2009 to only 3 years. Nevertheless, the 3-year survival rate of patients operated on for HLHS during 1990–2009 was significantly higher
than those operated on 1953–89 at 81% versus 50%, respectively (HR for death 1990–2009, 0.16; 95% CI, 0.036 to 0.67; \( P=0.0004 \) for logrank test; Figure 3F). Eighty-nine percent (64 / 72) of all deaths in the HLHS group occurred before the second-stage operation and 93% (67 / 72) of deaths occurred within 1 year of the first operation.

**Number of operations per patient**

Of all patients alive at the closing day of the study, 15% needed reoperation surgery before the age of 15. One-third of patients with TOF or TGA needed more than one operation (Table 3). Ninety-nine percent (99%) of HLHS patients and 95% of UVH patients alive at the end of the study had more than one operation (Table 3).

**Discussion**

We previously reported long-term results of a 45-year follow-up of pediatric cardiac surgery.\(^9\)

However, the current study is unique in terms of the sheer size of the study population, the long follow-up time of 60 years, and nearly complete follow-up coverage of 98%.

The amount of pediatric cardiac operations increased substantially throughout the follow-up time. The birthrate in Finland remained relatively constant, but the improved diagnosis of cardiac defects led to this increase in operations. However, the number of operations for all defects except HLHS decreased in the 2000s. The number of children born with congenital malformations in Finland remained constant throughout the 1990s and 2000s, thus failing to explain this. On the other hand, the steady increase in the number of catheter interventions compensated for cardiac surgery, especially for less complicated defects.

**Patient age and number of operations**

Surgical skills and techniques, as well as screening for cardiac defects, have improved
dramatically throughout the years. In effect, both mean and median patient age at operation decreased steadily during the follow-up period. The average age of the patients at their first operation was highest in the ASD group and lowest in the HLHS group in the 2000s, since untreated HLHS patients are not able to live long after birth.

The rise in the number of severe defects from the 1950s to 2000s led to an increase in the average number of operations per patient. Fifteen percent (15%) of all patients who were alive at the end of the study had undergone more than one operation for their defect. Patients with HLHS and UVH required three operations to palliate their cardiac defect. In contrast, patients with simpler defects, such as PDA and ASD, required practically no subsequent procedures.

Operative results and early mortality

The risk of early mortality correlated with the severity of the defect, whereby more severe defects experienced higher early mortality. Our results show that early mortality accounted for an increasingly smaller number of deaths within all diagnostic groups, especially those with severe defects.

Enhanced surgical skills and the development of catheter techniques have contributed to the improved operative results. Moreover, the development of new inotropes (e.g. Milrinone) or therapeutic drugs for treatment of pulmonary hypertension (e.g. nitric oxide and Sildenafil), as well as technical advances in pediatric extracorporeal membrane oxygenators (ECMOs) and ventricular assist devices (VADs) have improved the postoperative care of patients. Also, advanced prenatal screening methods and diagnostic tools have led to the detection and treatment of cardiac defects at an earlier stage in life, potentially contributing to improved surgical outcomes.²¹⁰ Lastly, centralization of pediatric cardiac surgery into one center (i.e. Helsinki university central hospital) was completed towards the end of the 1990s, and has most probably
contributed to improved results during the 1990s and 2000s.

Survival

Older patients passing away due to natural, age-related factors caused the declining feature of the survival curves. The oldest patient alive at the end of the study was 73 years old. Altogether, 367 patients were over 60 years old at the end of the study period.

This study proves that the vast majority of patients with simpler defects are expected to survive up to 60 years after corrective surgery. More importantly, according to these results, the surgical treatment of PDA and ASD can be considered a curative procedure. Additionally, the majority of late deaths in these patient groups were caused by non-cardiovascular events. The late survival of COA and VSD patients has improved significantly over time. Nonetheless, after excluding early mortality, COA and VSD patients operated on 1990–2009 experienced nearly identical survival rates as the general population after 20 years of follow-up. The survival rate of patients operated on for HLHS, TGA, and UVH decreased significantly during the first year of follow-up, which was mainly caused by the high rate of early mortality. However, both early and late results of all patients operated between 1990-2009 has improved significantly when compared to those operated between 1953-1989. The explanation is most likely multifaceted, including improvements in preoperative diagnostics and care, operative treatment, postoperative care and patient follow-up.

The long-term survival of TOF patients improved significantly to 90% at 22 years for those operated on after 1990 (excluding early mortality) and compares well with those of previous studies.\textsuperscript{11–14}

In this study, TGA included both simple and complex forms of the defect. The 22-year survival rate of simple TGA patients was nearly 100% (early mortality excluded). Until 1984, the
atrial switch procedure was the primary surgical method for the correction of TGA in our institution, while, since 1990, the arterial switch operation (ASO) has been used for the correction of all TGA defects. Several studies have found the ASO method to improve late results compared with the atrial switch method.\textsuperscript{15–17} Accordingly, in our study the 22-year survival of TGA patients operated on after 1990 improved significantly, confirming the results from other studies.

Previous studies have reported a survival rate of 84–86\% at 10 years and 74–82\% at 15 years after surgical correction of UVH (including early mortality).\textsuperscript{18,19} In our study, one-third of patients were alive after 45 years of follow-up with the majority of patients dying at a young age. The late survival of patients operated between 1953–1989 and 1990–2009 was comparable until eight years after the operation, but thereafter the late survival of patients operated between 1990–2009 was significantly higher than those operated between 1953–1989.

Previous studies have reported a 39\% survival rate after stage 1 (including early mortality) and a 90\% survival rate after stage 3 HLHS operations (early mortality excluded) at 10 years.\textsuperscript{20,21} In our study, the long-term survival of HLHS patients was 81\% at 15 years after all operative stages (excluding early mortality) for those that underwent surgery after 1990. Almost all of the HLHS patients were under 1 year of age at the time of their death. The majority of deaths in the HLHS group occurred before the second operation. These results suggest that if HLHS patients survive their first operation their chance of long-term survival increases significantly. These results correspond well with previous studies where mortality after stage 1, but not stage 2 or 3, surgery remained high.\textsuperscript{22–26} However, we observed a six-fold decrease in the early operative mortality after stage I Norwood operations for HLHS from the 1990s to the 2000s. Similarly, the early mortality of patients operated on for UVH decreased significantly
from the 1990s to the 2000s, showing a potential towards improved long-term results in both the HLHS and UVH groups in the future.

**Conclusions**

In conclusion, the number of children operated on for severe cardiac defects increased steadily throughout the study period. The defects were diagnosed and treated at increasingly younger ages. Improved diagnostic methods, combined with advancements in surgical, cardiological, and intensive care skills, contributed to profoundly improve operative results and the long-term survival of patients.

**Acknowledgments:** We thank the Finnish Cancer Registry for assisting us with the statistical survival analyzes and providing us with the age-, time-, and gender-matched population data used as the comparison group in the survival study. We also extend our gratitude to the Finnish Institute for Molecular Science for their statistical advice.

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**Conflict of Interest Disclosures:** None.

**References:**


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**Table 1.** Hierarchy of Diagnoses.

Univentricular heart (UVH)
Hypoplastic left heart syndrome (HLHS)
Truncus arteriosus
Interrupted aortic arch
Transposition of the great arteries (TGA)
Atrioventricular septal defect (ASD)
Total anomalous pulmonary venous drainage
Pulmonary atresia
Tetralogy of Fallot (TOF)
Ventricular septal defect (VSD)
Coarctation of the aorta (COA)
Aortic stenosis
Pulmonary stenosis
Mitral valve disease
Partial anomalous pulmonary venous drainage
Atrial septal defect (ASD)
Patent ductus arteriosus (PDA)
Miscellaneous

**Table 2.** Mean Age at All Operations and Early Mortality (<30 days after the operation).

<table>
<thead>
<tr>
<th>Time</th>
<th>Operations</th>
<th>Early Mortality</th>
<th>Mean Age</th>
<th>Patients</th>
<th>Operations / Patient</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>no.</td>
<td>no. (%)</td>
<td>(years)*</td>
<td>no.</td>
<td></td>
</tr>
<tr>
<td>1950</td>
<td>329</td>
<td>11 (3)</td>
<td>8.9</td>
<td>323</td>
<td>1.02</td>
</tr>
<tr>
<td>1960</td>
<td>1514</td>
<td>86 (6)</td>
<td>7.1</td>
<td>1461</td>
<td>1.04</td>
</tr>
<tr>
<td>1970</td>
<td>2336</td>
<td>212 (9)</td>
<td>5.6</td>
<td>2049</td>
<td>1.14</td>
</tr>
<tr>
<td>1980</td>
<td>2973</td>
<td>197 (7)</td>
<td>3.7</td>
<td>2435</td>
<td>1.22</td>
</tr>
<tr>
<td>1990</td>
<td>3943</td>
<td>258 (7)</td>
<td>2.8</td>
<td>2842</td>
<td>1.39</td>
</tr>
<tr>
<td>2000</td>
<td>2781</td>
<td>78 (3)</td>
<td>2.2</td>
<td>1852</td>
<td>1.50</td>
</tr>
<tr>
<td>Total</td>
<td>13,876</td>
<td>842 (6)</td>
<td>5.0</td>
<td>10,694</td>
<td>1.27</td>
</tr>
</tbody>
</table>

*P*<0.0001 when comparing the mean age of patients operated on in each decade to those operated on in previous decades.
**Table 3.** Early Mortality (<30 days), Number of Operations, and Mean Age at the First Operation.

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>21 (1)</td>
<td>1 (0)</td>
<td>0 (0)</td>
<td>22 (1)</td>
<td>1.0</td>
<td>5.9</td>
<td>1.8</td>
</tr>
<tr>
<td>ASD</td>
<td>6 (1)</td>
<td>1 (0)</td>
<td>0 (0)</td>
<td>7 (1)</td>
<td>1.0</td>
<td>8.2</td>
<td>4.5</td>
</tr>
<tr>
<td>COA</td>
<td>47 (5)</td>
<td>10 (3)</td>
<td>2 (1) *</td>
<td>59 (4)</td>
<td>1.1</td>
<td>5.4</td>
<td>1.5</td>
</tr>
<tr>
<td>VSD</td>
<td>85 (9)</td>
<td>24 (3)</td>
<td>3 (1) ***</td>
<td>112 (5)</td>
<td>1.2</td>
<td>4.1</td>
<td>1.7</td>
</tr>
<tr>
<td>TOF</td>
<td>53 (9)</td>
<td>7 (3)</td>
<td>3 (1) ***</td>
<td>63 (6)</td>
<td>1.4</td>
<td>4.9</td>
<td>0.9</td>
</tr>
<tr>
<td>TGA</td>
<td>53 (13)</td>
<td>34 (14)</td>
<td>7 (4) ***</td>
<td>94 (1)</td>
<td>1.4</td>
<td>1.1</td>
<td>28d</td>
</tr>
<tr>
<td>HLHS</td>
<td>11 (79)</td>
<td>28 (42)</td>
<td>21 (7) ***</td>
<td>60 (17)</td>
<td>3.4</td>
<td>49d</td>
<td>7d</td>
</tr>
<tr>
<td>UVH</td>
<td>44 (20)</td>
<td>41 (12)</td>
<td>11 (5) ***</td>
<td>96 (12)</td>
<td>3.0</td>
<td>1.6</td>
<td>153d</td>
</tr>
<tr>
<td>Misc.</td>
<td>186 (15)</td>
<td>112 (11)</td>
<td>31 (4) ***</td>
<td>329 (10)</td>
<td>1.4</td>
<td>4.6</td>
<td>2.3</td>
</tr>
<tr>
<td>All</td>
<td>506 (7)</td>
<td>258 (7)</td>
<td>78 (3)</td>
<td>842 (6)</td>
<td>1.2</td>
<td>5.2</td>
<td>2.0</td>
</tr>
</tbody>
</table>

* P=0.017 and *** P<0.0001 when compared to early mortality 1953–1989.
† P<0.0001 in all diagnostic groups when comparing the mean age of patients operated on 1990–2009 to those operated on 1953–1989.
d = days
## Table 4. Number and Proportion of Operations Divided by Time Periods.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Total — no. (%)</th>
<th>1953–1989 — no. (%)</th>
<th>1990–1999 — no. (%)</th>
<th>2000–2009 — no. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>2356 (17)</td>
<td>1877 (26)</td>
<td>369 (9)</td>
<td>110 (4)</td>
</tr>
<tr>
<td>ASD</td>
<td>1477 (11)</td>
<td>802 (11)</td>
<td>485 (12)</td>
<td>190 (7)</td>
</tr>
<tr>
<td>COA</td>
<td>1655 (12)</td>
<td>1007 (14)</td>
<td>382 (10)</td>
<td>276 (10)</td>
</tr>
<tr>
<td>VSD</td>
<td>2124 (15)</td>
<td>991 (14)</td>
<td>724 (18)</td>
<td>409 (15)</td>
</tr>
<tr>
<td>TOF</td>
<td>1081 (8)</td>
<td>587 (8)</td>
<td>268 (7)</td>
<td>226 (8)</td>
</tr>
<tr>
<td>TGA</td>
<td>843 (6)</td>
<td>399 (6)</td>
<td>251 (6)</td>
<td>193 (7)</td>
</tr>
<tr>
<td>HLHS</td>
<td>364 (3)</td>
<td>14 (0)</td>
<td>66 (2)</td>
<td>284 (10)</td>
</tr>
<tr>
<td>UVH</td>
<td>798 (6)</td>
<td>220 (3)</td>
<td>343 (9)</td>
<td>235 (8)</td>
</tr>
<tr>
<td>Misc.</td>
<td>3168 (23)</td>
<td>1255 (18)</td>
<td>1055 (27)</td>
<td>858 (31)</td>
</tr>
<tr>
<td>All</td>
<td>13,876 (100)</td>
<td>7152 (100)</td>
<td>3943 (100)</td>
<td>2781 (100)</td>
</tr>
</tbody>
</table>

## Table 5. Average Follow-Up Time for All Patients in Each Group that Survived Their First Operation and Survival Rates Including Early Mortality for Patients in Each Diagnostic Group Operated on 1953–2009.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Average follow-up (years)</th>
<th>10-year survival (%)</th>
<th>20-year survival (%)</th>
<th>30-year survival (%)</th>
<th>40-year survival (%)</th>
<th>50-year survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>35</td>
<td>98</td>
<td>97</td>
<td>94</td>
<td>92</td>
<td>88</td>
</tr>
<tr>
<td>ASD</td>
<td>25</td>
<td>99</td>
<td>98</td>
<td>97</td>
<td>93</td>
<td>85</td>
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<tr>
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</table>
Figure Legends:

**Figure 1.** Patient Age at Operation and the Number of Operations and Birth Rate by Decade. Bars are divided according to different age groups. The continuous line represents the number of children born in the general population during each decade.

**Figure 2.** Survival of All Patients in Different Diagnostic Groups Operated on: (A) 1953–2010 and (B) 1990–2010. Follow-up started after the patient’s first operation. Early mortality is included for all diagnostic groups.

**Figure 3.** Survival of Patients in the Six Most Severe Cardiac Defect Groups Operated on 1953–1989 and 1990–2010. The numbers on the dotted lines represent the number at risk in each group at 20 years and 10 years in the HLHS group. Early mortality is excluded from all diagnostic groups. † $P=0.001$ eight years after the first operation, *$P=0.037$, **$P=0.0004$, ***$P<0.0001$ versus patients operated on 1953–1989. Data is presented in the form of Kaplan–Meier graphs and $P$-values obtained using the logrank test for proportional hazards and chi square test for non-proportional hazards (denoted by †). Hazard ratios and Confidence intervals were obtained using the Cox regression model and represent the overall hazard ratio for death for patients operated on 1990–2009.
Figure 2

A

All patients operated between 1953-2009

Survival %

Years after first operation

Legend
Pop
ALL
PDA
ASD
COA
VSD
TOF
TGA
HLHS
UVH

Follow-up time
Number at risk at 40 years
59
963
53
201
55
246
52
126
58
108
46
11
17
-
47
5

B

All patients operated between 1990-2009

Survival %

Years after first operation

Legend
Pop
ALL
PDA
ASD
COA
VSD
TOF
TGA
HLHS
UVH

Follow-up time
Number at risk at 20 years
22
712
22
152
22
140
22
68
22
116
22
40
22
29
17
-
22
15
Figure 3
Progress in Late Results Among Pediatric Cardiac Surgery Patients: A Population-Based Six-Decade Study with 98% Follow-Up
Alireza Raissadati, Heta Nieminen, Eero Jokinen and Heikki Sairanen

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