Late Results of Pediatric Cardiac Surgery in Finland
A Population-Based Study With 96% Follow-Up

Heta P. Nieminen, MD; Eero V. Jokinen, MD; Heikki I. Sairanen, MD

Background—This population-based study characterizes the history and progress of pediatric cardiac surgery in Finland. For the first time, all defects and procedures are included in an outcome study, reflecting the true effectiveness of operative treatment.

Methods and Results—All data relating to the operations were collected retrospectively from hospital records. Current patient status was obtained from the population registry. Survival was evaluated with the Kaplan-Meier method applied to all patients and separately to subgroups of patients with the most common defects. The survival rates were compared with those of an age- and sex-matched general population. During the 37 years (1953 to 1989), 6461 patients underwent surgery; 96% of them were traced. The number of operations and the constellation of defects treated increased dramatically over time. Actuarial survival for the 45 years ended October 28, 1998 (the ending date of this study) was 78% for patients versus 93% for the general population. Survival and the number of operations per patient varied widely with the defect. The survival of patients with a surgically closed atrial septal defect was comparable to that of the general population, and such patients rarely needed a reoperation, whereas only 15% of patients with univentricular heart survived for 34 years, and almost all needed at least 2 operations.

Conclusions—The overall survival of patients with cardiac defects corrected surgically in childhood is good compared with their estimated natural course. The increasing number of surgically treatable defects and the growing number of operations per patient reflect the increasing ability to treat more difficult cases. (Circulation. 2001;104:570-575.)

Key Words: heart defects, congenital ▪ surgery ▪ follow-up studies ▪ survival ▪ population

Five hundred children are born with congenital heart disease in Finland annually.1 Heart malformations often result in high mortality and morbidity. Depending on the defect, life expectancy varies from 1 week to normal adulthood.2 In most instances, surgery is the only available treatment. Pediatric cardiac surgery began in 1938 with closure of patent ductus arteriosus. Since then, the progress has been rapid, with only a few patients remaining beyond the reach of surgical cure or palliation today.

The aim of the present study was to evaluate the late prognosis of children who underwent surgery for congenital heart defects between 1953 and 1989 in Finland. Data from all heart operations were collected retrospectively, and patients were traced to determine their subsequent course and survival.

This population-based study illustrates the history and progress of pediatric cardiac surgery in Finland. Most of the published long-term follow-up studies deal with solitary defects treated in a single institution.3–8 The present study offers unique coverage of the entire population of 5 million people during the first 37 years of pediatric cardiac surgery. The existence of a nationwide public healthcare system allowed virtually all children with congenital heart disease to be found within the limits of diagnostic methods available. All defects found were referred to care considered appropriate at any given time. A highly accurate and comprehensive national population registry enabled an essentially complete (96%) follow-up of these patients and comparison of survival rates versus the general population.

Methods
Data from all cardiac operations performed on children in Finland before 1990 were collected. Operations were performed in 5 university hospitals and 1 regional hospital. Patients were classified as pediatric if the operation took place before their 15th birthday. Data collection was limited to the end of 1989 to ensure a minimum follow-up of 8 years before the closing date of the study (October 28, 1998).

Material was collected from surgical logs, diagnosis cards, and computer files of the hospitals. Surgical logs contained daily entries of all operations performed in the hospital; diagnosis cards and computer files contained information on all patients of the hospital listed by ICD-9 (International Classification of Diseases, 9th edition) diagnosis number. If the information was incomplete, the medical records of the patients were reviewed.

Patients were given one primary diagnosis to facilitate comparison of survival between defects. Heart defects were sorted hierarchically...
patients with more than 1 defect were given the diagnosis highest on the list. The hierarchy was based on hierarchies published previously.9,10 Operative mortality included all deaths that occurred within the first 30 days after surgery. Current patient status and dates of death and emigration were obtained from the Finnish Population Registry Center. The center also provided the number of children and life tables for the general population. Complete follow-up ended either with the patient’s death or on the closing day of the study. The research was conducted with the permission of the Ministry of Social Affairs and Health.

All data were stored in a computer running a custom-made research registry program, ProCardio (Melba Group) based on FileMaker Pro 3.0 (Claris Corp). Survival was estimated with the Kaplan-Meier method for the entire patient population and separately for groups of patients with the 7 most common diagnoses: 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), or univentricular heart (UVH). Time point zero was the date of the patient’s first operation.

Late survival was calculated twice, including and excluding operative mortality. The survival and longevity of the study group was compared with time-, age-, and sex-matched general population. The comparison was made for each patient individually. For every patient and for every year of follow-up, the mortality rate of the general population was obtained. The expected probability of surviving a year was obtained for all individuals who were not censored before the beginning of the interval, regardless of their survival status. The Hakulinen method11 for combining the individual information into an estimate of the expected survival of the group was used. The effect of cardiac defect on the mortality rate was estimated by the relative survival (patient survival/control survival). All statistics were made with a survival analysis package developed at the Finnish Cancer Registry.12

Results

The first heart operation for a child in Finland was performed in 1953. A total of 7240 cardiac operations were performed on 6461 pediatric patients through the end of 1989. During these 37 years, the number of operations increased regardless of a concurrent decrease in the Finnish child population (Figure 1). Patient age at operation averaged 5.1 years (median 4.5; range 0 to 15). Mean age decreased during the study period from 8.9 to 3.4 years and the median even more from 9.0 to 1.7 (Table 2). Operative mortality varied between decades from 3.3% to 8.3%, with an overall average of 6.9% (Table 2).

Patient material varied over time, as more defects became surgically treatable. In the 1950s, most patients had surgery for PDA. During the subsequent decade, the number of operations and the variation of the defects increased dramatically as the use of open-heart surgery began (Table 3). Follow-up was complete in 96% of the patients (Table 4). Only 125 could not be traced because of illegible handwritten or incorrect markings in the old surgical logs. These patients were excluded from analysis. The follow-up ended prematurely in 129 cases because of emigration. The emigration dates were marked as the last day of follow-up for these patients. Thus, the survival analysis includes 98% of the patients.

The sex distribution of patients was nearly even: 46% males and 54% females. There were no sex differences in the incidences of VSD and UVH. A female predominance was found for PDA (71%) and ASD (60%) and a male predominance for COA (66%), TOF (60%), and TGA (67%).

Survival

A total of 433 of the 6336 traced patients died within 30 days after their first operation (Table 4). Operative mortality was lowest (1%) in the ASD group. Mortality increased as the defects became more complicated; in the UVH group, it was 25%.

### Table 1. Hierarchy of Diagnosis

<table>
<thead>
<tr>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>UVH</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
</tr>
<tr>
<td>TGA</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous drainage</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
</tr>
<tr>
<td>TOF</td>
</tr>
<tr>
<td>VSD</td>
</tr>
<tr>
<td>COA</td>
</tr>
<tr>
<td>Aortic stenosis</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
</tr>
<tr>
<td>Mitral valve disease</td>
</tr>
<tr>
<td>Partial anomalous pulmonary venous drainage</td>
</tr>
<tr>
<td>ASD, secundum</td>
</tr>
<tr>
<td>PDA</td>
</tr>
<tr>
<td>Other</td>
</tr>
</tbody>
</table>

### Table 2. Age at Operation and Operative Mortality

<table>
<thead>
<tr>
<th>Time Period</th>
<th>No. of Operations</th>
<th>Age, y</th>
<th>Mortality, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1953–1959</td>
<td>329</td>
<td>8.9</td>
<td>9.0</td>
</tr>
<tr>
<td>1960–1969</td>
<td>1505</td>
<td>7.1</td>
<td>7.3</td>
</tr>
<tr>
<td>1970–1979</td>
<td>2337</td>
<td>5.5</td>
<td>5.3</td>
</tr>
<tr>
<td>1980–1989</td>
<td>3069</td>
<td>3.4</td>
<td>1.7</td>
</tr>
<tr>
<td>1953–1989</td>
<td>7240</td>
<td>5.1</td>
<td>4.5</td>
</tr>
</tbody>
</table>
On the closing day of the study, 80% (5181) of all patients were living in Finland. Their mean age was 27.6 years (range 8.9 to 59.0 years), and their average length of follow-up was 22.3 years (range 8.8 to 45.5 years). There were 593 late deaths; these patients died at a mean age of 13.3 years (median 10.2, range 0.11 to 53.2), an average of 9.3 years (median 5.8, range 0.9 to 38.9) after their first operation.

The overall survival rate for the 45 years ended October 28, 1998, was 78% for the study group and 93% for the general population (Figure 2). The negative effect of the disease gradually disappeared over the years, and patients' risk of death became similar to control population. This was seen as planar or slightly rising curves of relative survival (Figure 3). In severe defects, the relative survival remained depressed. The sharp drops in curves were caused by a small number of living patients.

The average longevity of the patients was 10 years less than that of the general population, 58 and 68 years, respectively. After operative mortality was excluded, the survival rate increased to 84% and the average longevity to 62 years.

Surgical treatment for PDA and TOF has been available for 45 years. The actuarial survival rates of these patients in our series were 88% and 55%, respectively (Figure 2). Follow-up times and survival rates for the other common defects were as follows: COA 42 years, 79%; ASD 40 years, 95%; VSD 39 years, 79%; TGA 34 years, 49%; and UVH 34 years, 15%.

### Number of Operations per Patient

More than 1 procedure was performed on 949 patients, or 16% of those who survived their first operation. The need for multiple operations increased during the decades from 1.18 to 1.26. The number of operations needed varied with the defect. PDA and ASD patients needed rarely reoperation, 16% of patients with COA were operated on at least twice, and every third VSD, TOF, and TGA patient and 80% of patients with UVH needed several operations (Table 4). An accompanying COA increased the number of operations in VSD and TGA patients. Nine percent of patients with VSD and 6% of patients with TGA also had COA, and all those still living on the closing day of the study had required ≥2 operations. In addition, pulmonary artery banding before defect closure increased the need for operations in the VSD group. Only 11 of the 99 patients with UVH lived more than 10 years with 1 operation.

### Discussion

This population-based study traces the course of 6336 patients. With excellent cooperation between hospitals, we were able to follow most patients for the entire period of their life. The study was limited by the lack of data on patients who died before study entry or who moved out of the region.

### Table 3. Number and Proportion of Defects Divided by Decade of First Operation

<table>
<thead>
<tr>
<th>Defect</th>
<th>1950, n (%)</th>
<th>1960, n (%)</th>
<th>1970, n (%)</th>
<th>1980, n (%)</th>
<th>1953–1989, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>250 (77)</td>
<td>701 (48)</td>
<td>498 (24)</td>
<td>601 (23)</td>
<td>2050 (32)</td>
</tr>
<tr>
<td>COA</td>
<td>19 (6)</td>
<td>196 (13)</td>
<td>369 (18)</td>
<td>342 (13)</td>
<td>926 (14)</td>
</tr>
<tr>
<td>ASD</td>
<td>2 (1)</td>
<td>151 (10)</td>
<td>324 (16)</td>
<td>335 (13)</td>
<td>812 (13)</td>
</tr>
<tr>
<td>VSD</td>
<td>...</td>
<td>115 (8)</td>
<td>260 (13)</td>
<td>392 (15)</td>
<td>767 (12)</td>
</tr>
<tr>
<td>TOF</td>
<td>30 (10)</td>
<td>114 (8)</td>
<td>131 (6)</td>
<td>175 (7)</td>
<td>453 (7)</td>
</tr>
<tr>
<td>TGA</td>
<td>...</td>
<td>25 (2)</td>
<td>114 (5)</td>
<td>159 (6)</td>
<td>298 (5)</td>
</tr>
<tr>
<td>UVH</td>
<td>1 (0)</td>
<td>20 (1)</td>
<td>42 (2)</td>
<td>74 (3)</td>
<td>137 (2)</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>19 (6)</td>
<td>139 (10)</td>
<td>336 (16)</td>
<td>524 (20)</td>
<td>1018 (16)</td>
</tr>
<tr>
<td>All</td>
<td>324 (100)</td>
<td>1461 (100)</td>
<td>2074 (100)</td>
<td>2602 (100)</td>
<td>6464 (100)</td>
</tr>
</tbody>
</table>

### Table 4. Patient Follow-Up, Operative Mortality, and Need for Multiple Operations

<table>
<thead>
<tr>
<th>Defect</th>
<th>No. Traced*</th>
<th>Complete Follow-Up, †</th>
<th>Operative Mortality, ‡</th>
<th>Survived Patients, †</th>
<th>Reoperated Patients, ‡</th>
<th>Operations/ Survived Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>2050</td>
<td>1983</td>
<td>1917 (94)</td>
<td>49 (2)</td>
<td>1868</td>
<td>43 (2)</td>
</tr>
<tr>
<td>COA</td>
<td>926</td>
<td>911</td>
<td>891 (96)</td>
<td>33 (4)</td>
<td>858</td>
<td>134 (16)</td>
</tr>
<tr>
<td>ASD</td>
<td>812</td>
<td>790</td>
<td>775 (95)</td>
<td>8 (1)</td>
<td>767</td>
<td>29 (2)</td>
</tr>
<tr>
<td>VSD</td>
<td>767</td>
<td>759</td>
<td>753 (98)</td>
<td>61 (8)</td>
<td>692</td>
<td>186 (27)</td>
</tr>
<tr>
<td>TOF</td>
<td>453</td>
<td>453</td>
<td>447 (99)</td>
<td>40 (9)</td>
<td>407</td>
<td>136 (33)</td>
</tr>
<tr>
<td>TGA</td>
<td>298</td>
<td>298</td>
<td>297 (100)</td>
<td>40 (13)</td>
<td>257</td>
<td>81 (32)</td>
</tr>
<tr>
<td>UVH</td>
<td>137</td>
<td>137</td>
<td>134 (98)</td>
<td>35 (26)</td>
<td>99</td>
<td>79 (80)</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>1018</td>
<td>1006</td>
<td>993 (98)</td>
<td>167 (17)</td>
<td>826</td>
<td>261 (32)</td>
</tr>
<tr>
<td>All</td>
<td>6461</td>
<td>6336</td>
<td>6207 (96)</td>
<td>433 (7)</td>
<td>5774</td>
<td>949 (16)</td>
</tr>
</tbody>
</table>

*Patients identified from population registry.
†Patients living in Finland or who died (proportion of all patients).
‡Patients who survived first operation with complete follow-up.
§Patients who underwent ≥1 operation (proportion of those who survived).
||Number of operations for patients who survived.
able to trace all the operations performed in the whole country, and with a good national registry, we obtained survival information on 96% of these patients.

The Finnish healthcare system covers all inhabitants and reaches practically every child. The low and equal costs of health care do not restrict the availability of treatment. Therefore, the present study reveals the true population-based results of pediatric cardiac surgery.

Comparisons with the general population were possible because the population register publishes reliable life tables. Although most of the patients were still alive, calculation of longevity could be made reliably because the negative effect of the defect on mortality vanished decades after operation.

Operative Mortality

The progress of the entire chain of diagnosis and treatment is reflected in the operative mortality and in the variation of the defects treated. Early mortality increased 2.5-fold during the first decades of surgery, as more complicated defects became operable. During the last 10 years, operative mortality decreased to 6.8% as experience was gained in the treatment of patients with the most common severe defects.

Previously published results are not readily comparable to ours because this study includes all defects and procedures. Evolution of the practice of cardiology and cardiac surgery confounds comparisons even more. The population-based study conducted in the state of Oregon is closest in nature to ours, but it excludes patients with only palliation.

Operative mortality varies between defects. In our PDA, ASD, and COA groups, it varied from 1% to 4%, which compares well with previously published mortality rates of 0% to 13%. The operative mortality of an isolated VSD varies from 7% to 13%. In the present study, it was 8% for the entire VSD group.

Early mortality after corrective surgery of TOF varies widely, from 9% to 32%. Most Finnish patients underwent a corrective operation during the follow-up, but the operative mortality rate of 9% was positively affected by shunt procedures.

In most studies, patients with TGA are divided into subgroups by anatomy and surgical procedure. The 13% operative mortality in the present study includes both simple and complex TGA and covers all procedures used. In the Oregon study, the mortality rate for the corrective surgery was 22%.

Most of our patients with UVH had a palliative shunt operation. The operative mortality rate of 26% was high and comparable to the 34% rate seen in 3 months without surgery.

Late Survival

The 45-year survival rate of the patients was 15% lower than that of the general population in the present study. When operative mortality was excluded, the survival rate (84%) was only 9% less than that of the general population. The relative mortality shows that the operated defect does not significantly increase overall mortality later.

The effectiveness of surgical treatment was estimated herein by comparison of survival rates to the expected natural survival. Ideally, the results of surgery would be compared with the concurrent natural course of the defects, but as the operative treatment has proven its strength, such comparisons have become ethically indefensible.

The 15-year natural survival rate of patients with cardiac defects has been estimated to be 67%. In our material, the survival rate for 15 years after first operation was 86%. Not all children with heart defects survive to surgery, and some have a normal life expectancy without operation. However, even considering these confounding factors and the absence of reliable control data, the positive effect of surgical treatment is clear.

The overall 45-year survival rate for patients with PDA was 88% compared with 94% in the general population. The excess of deaths resulted from pulmonary hypertension, and in the 1980s, from prematurity.

Most patients (68%) with COA survive through childhood without surgery. Among our COA patients, the 15-year survival rate after operation was 92%, which is in agreement with the 90% consistently reported. An accompanying VSD reduces survival considerably.

In our hierarchy, pa-
tients with COA and significant VSD were classified in the VSD group.

If only survival is considered, the closure of an ASD is a curative procedure. After successful operation, ASD patients have a life expectancy that is similar to that of the general population, as was also seen in a study by the Mayo Clinic.2

The natural survival rate of patients with VSD is rather good; 76% of patients are estimated to live for 15 years.3 The material of the natural survival study differs significantly from ours because many defects close spontaneously.4 In the present study, the survival rate for 15 years was 83%. After exclusion of the operative mortality, the 35-year survival rate was 86%, similar to the 83% reported for isolated defects closed in the early era of cardiac surgery.3

The most dramatic improvement in survival was seen in the TOF group. The 15-year natural survival rate is 14%, whereas after successful corrective surgery, it exceeds 90%.6,17–19 In our material, which also included patients with only palliation, the actuarial survival rates for 15 years were 82% and 90% after operative mortality was excluded.

Without surgery, 38% of TGA patients reach adulthood.3 The calculated 15- and 30-year survival rates in the present study were 65% and 49%, respectively. Many patients were only palliated. In a previously published report of a portion of our data, the 15-year survival rate after Senning procedure was 90%, including operative mortality,26 which compares well with other studies in which survival ranged from 78% to 94%.21,27,28

Fewer than one third of patients with UVH survive 15 years without surgery.2 In our material, most patients received a palliative shunt in their first operation. The decline in survival is sharp: 60% for 5 years, 45% for 15 years, and 15% for 34 years. As new procedures become more common, the survival rate is improving. In selected data after the Fontan procedure, the 15-year survival rate was reported to be 74%.29

Number of Operations per Patient
The severity of the defects operated on is also seen in the number of patients who require multiple operations. Patients with PDA or ASD rarely undergo more than 1 operation. In COA patients, 2 major problems may indicate a reoperation: recoarctation and associated aortic valve disease.5,14,24 These problems caused the fairly high rate of operation per patient in the present study. Most patients with TOF were first palliated and went on to have corrective surgery later. The need for several operations for VSD and TGA patients in the present study is partly due to associated COA. Most of the UVH patients required more than 1 operation.

Conclusions
The overall survival of children operated on for heart defects is fairly good. Progress in cardiology, surgery, anesthesiology, and critical care has brought an increasing number of different defects within the scope of operative treatment. The patients, especially those with simpler defects, do not have increased risk of death years after successful operation. The growing number of operations per patient reflects the ability to treat increasingly complicated defects. However, survival is only a basic element in measuring the results of a treatment. A far bigger challenge, evaluation of the quality of life of the surviving patient, remains to be undertaken.

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References


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