Achievements in Congenital Heart Defect Surgery
A Prospective, 40-Year Study of 7038 Patients

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Background—This article presents an update of the results achieved by modern surgery in congenital heart defects (CHDs) over the past 40 years regarding survival and the need for reoperations, especially focusing on the results from the past 2 decades.

Methods and Results—From 1971 to 2011, all 7038 patients <16 years of age undergoing surgical treatment for CHD at Rikshospitalet (Oslo, Norway) were enrolled prospectively. CHD diagnosis, date, and type of all operations were recorded, as was all-cause mortality until December 31, 2012. CHDs were classified as simple (3751/7038=53.2%), complex (2918/7038=41.5%), or miscellaneous (369/7037=5.2%). Parallel to a marked, sequential increase in operations for complex defects, median age at first operation decreased from 1.6 years in 1971 to 1979 to 0.19 years in 2000 to 2011. In total, 1033 died before January 1, 2013. Cumulative survival until 16 years of age in complex CHD operated on in 1971 to 1989 versus 1990 to 2011 was 62.4% versus 86.9% (P<0.0001). In the comparison of patients operated on in 2000 to 2004 versus 2005 to 2011, 1-year survival was 90.7% versus 96.5% (P=0.003), and 5-year cumulative survival was 88.8% versus 95.0% (P=0.0003). In simple versus complex defects, 434 (11.6%) versus 985 (33.8%) patients needed at least 1 reoperation before 16 years of age. In complex defects, 5-year cumulative freedom of reoperation among patients operated on in 1990 to 1999 versus 2000 to 2011 was 66% versus 73% (P=0.0001).

Conclusions—Highly significant, sequential improvements in survival and reductions in reoperations after CHD surgery were seen. A future challenge is to find methods to reduce the need for reoperations and further reduce long-term mortality. (Circulation. 2015;131:337-346. DOI: 10.1161/CIRCULATIONAHA.114.012033.)

Key Words: heart defects, congenital ▪ reoperation ▪ survival ▪ thoracic surgery

Following the surgical closures of patent ductus arteriosus in 1938¹ and atrial septal defect (ASD) in 1953,² major treatment advances made surgical palliation or correction³ of nearly all congenital heart defects (CHDs) possible. Still, 232 000 deaths globally were attributed to CHD in 2010,⁴ and most victims were young. Trends in Western countries indicate, however, that this number may be substantially reduced. Nieminen et al⁵ reported major improvements in the survival of children in Finland undergoing CHD surgery up to 1989. Since then, few population-based studies have specifically targeted long-term survival after CHD surgery.⁶ ⁷

Data Collection
The study was conducted at the Departments of Thoracic Surgery, Pediatric Cardiology, and Cardiology at Oslo University Hospital, Rikshospitalet. All operations from June 21, 1971, to December 31, 2011, were registered prospectively with the use of a hand-written protocol from 1971 to 1989 (transferred to our electronic Datcor database in 1990) and directly into Datcor thereafter. All-cause mortality until December 31, 2012, was obtained from our files and from the Norwegian population registry in Statistics Norway. The study was approved by the Data Protection Officer at Oslo University Hospital. Data handling was facilitated by using each patient’s unique 11-digit social security number, which includes individual birth date. CHD diagnoses were based on International Classification of

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Cardiovascular Surgery Registry (operative from 1994) revealing that 80.1% of all CHD operations in Norway were performed at Rikshospitalet during this period. Since 2003, all CHD operations have been performed at Rikshospitalet.

From 1987 through 1998, 51 patients with hypoplastic left heart syndrome (HLHS) had their Fontan circulation completed at a surgical center in the United States (25 survivors). These operations have not been included in our database. Data on mortality in all CHDs in Norway from 1971 to 2011 were obtained through open access to official Norwegian mortality statistics at Statistics Norway.

Rapid development occurred in the availability of diagnostic tools during the long study period. In the 1970s, primary preoperative CHD diagnoses necessary for planning the operations were mainly based on clinical findings and invasive studies, but, from the early 1980s, diagnostics increasingly relied on echo-Doppler techniques and computer tomography, and, from the late 1980s, diagnostics also relied on MRI. Significant refinements in all these techniques are still taking place. However, final diagnoses (ie, applied in this article) were ultimately obtained during surgery.

Complex Defects
A CHD was defined as a defect in the structure of the heart or the great vessels that was present at birth. If >1 CHD was present, patients were classified by the most severe diagnosis according to a diagnosis hierarchy as applied previously. The following diagnostic criteria were used for complex defects (Table 1):

1. Univentricular hearts (UVH) were characterized by the presence of 1 rudimentary and 1 dominant ventricle (left or right), regardless of whether it had a double-inlet left or right ventricle or a single atrioventricular inlet.

Data on the number of operations performed at other Norwegian hospitals from 1994 to 2003 were obtained from the Norwegian Cardiovascular Surgery Registry (operative from 1994) revealing that 80.1% of all CHD operations in Norway were performed at Rikshospitalet during this period. Since 2003, all CHD operations have been performed at Rikshospitalet.

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1. Univentricular hearts (UVH) were characterized by the presence of 1 rudimentary and 1 dominant ventricle (left or right), regardless of whether it had a double-inlet left or right ventricle or a single atrioventricular inlet.
Truncus arteriosus communis was diagnosed according to Van Praagh and Van Praagh, including truncus arteriosus communis associated with an interrupted aortic arch.

Cases with interrupted or hypoplastic aortic arch (I/HAA) with or without coarctation of the aorta (COA) were grouped together. Aortic arch hypoplasia was defined according to Moulaert et al.10

Patients with simple or complex transposition of the great arteries (TGA) and biventricular anatomy were grouped together.

Atrioventricular septal defect included both the incomplete and complete forms.

Patients with totally anomalous pulmonary venous drainage were assigned to this group if this was their dominant defect.

Cases with pulmonary atresia (PA), with or without coexisting ventricular septal defect, were assigned PA as their primary diagnosis if biventricular correction was performed.

Tetralogy of Fallot (TOF) was defined according to convention. Double-outlet right ventricle included both simple and complex forms.11

According to the diagnosis hierarchy in Table 1, patients, eg, with both UVH and totally anomalous pulmonary venous drainage, were classified as UVH.

**Simple Defects**

Ventricular septal defect and COA were defined conventionally. Valvular aortic stenosis was classified as AS, valvular pulmonary stenosis as PS, and stenosis or insufficiency of the mitral valve as MV. Cases with drainage to the right atrium from ≥1, but not all, pulmonary veins, were classified as PAPVD. ASD included ASD secundum and superior and inferior sinus venous defect, but not partial atrioventricular septal defect. Patent ductus arteriosus was diagnosed in the presence of an open ductus ≥2 weeks after birth.

**Miscellaneous Defects**

The miscellaneous group (n=369) included double-outlet right ventricle (n=95), subvalvular and supravalvular aortic stenosis (n=65 and 21, respectively), Ebstein anomaly (n=11), congenitally corrected TGA (n=4), coronary artery anomalies (n=18), cor triatriatum (n=11), and vascular rings (n=48). A total of 97 patients (26.4% of all 369 patients in the miscellaneous group and 1.4% of all 7038 patients in the study) had defects lacking dominant features that justified classification into any of the previously mentioned categories.

Only patients who had their first operation before their 16th birthday were included, because this is the age threshold of the pediatric population in Norway. The term early mortality is used for all deaths that occurred <30 days after the first operation, and late mortality denotes deaths occurring later.

**Reoperations**

Dates and descriptions of all operations were recorded prospectively. The first operation was denoted the index operation. Operations both with and without cardiopulmonary bypass were included, and operations were classified according to whether they could be considered as corrections or palliations. Palliative procedures mainly included shunt operations or banding of the pulmonary artery, either as destination therapy or as steps in staged procedures aimed at final correction. Accordingly, eg, the Norwood I operation and Glenn operations were classified as a palliative procedure. For example, Fontan operations, on the other hand, were classified as corrections. The few reoperations attributable to acute surgical complications have not been included.

**Statistical Analysis**

We used Kaplan–Meier analyses to study survival after first operation (<30 days and until 16 years of age), and reoperation-free survival. In the latter case, patients who died without...
reoperation were censored at the time of death. The log-rank test (Mantel-Cox) was used to test for differences in survival between eras.

Kaplan–Meier probabilities of 5 years reoperation-free survival were computed separately for all complex defects in patients who had their first operation in 1990 to 1999 versus 2000 to 2011. Five years was chosen somewhat arbitrarily because patients operated on in 1990 to 1999 had longer follow-up than patients operated on in 2000 to 2011, and because the great majority of reoperations were performed during the first few years after the first operation in both eras (see Results).

The Mann–Whitney test was used when comparing variables not necessarily normally distributed. The binomial test was used when testing for differences in numbers of various types of operations in different eras.

Two-sided \( P \) values of \(<0.05\) were considered significant. The statistical packages Statview 5.0 and JMP 10 were used for analyses.

### Results

A total of 7038 CHD patients underwent 9380 operations during the data collection period.

One hundred eight patients (1.5%) had incomplete social security numbers and were lost to follow-up after discharge.

### Index operations

Table 2 shows the numbers and proportions of patients who had their index operation during the 4 decades of 1971 to 1979, 1980 to 1989, 1990 to 1999, and 2000 to 2011. The number of patients operated on for complex defects increased from 37 per year in 1971 to 1979 to 109 per year in 2000 to 2011. Surgery for simple defects only increased from 80 per year in 1971 to 1979 to 92 per year in 2000 to 2011.

### Long-Term Survival Trends

Substantial, gradual improvements in survival were seen over the whole study period for all complex defects (Figure 1 and Table 3). In general, most of the improvements occurred early after surgery, but also late mortality improved sequentially, particularly in UVH, TGA, and PA.

Taking into account all complex defects, early survival was 79.7% in patients who underwent surgery in 1971 to 1989 versus 94.6% in 1990 to 2011 (\(P<0.0001\)), and cumulative survival to 16 years of age was 62.4% versus 86.9% (\(P<0.0001\), respectively. Moreover, highly significant improvements were also found for simple defects like ventricular septal defect and COA.

### Table 3. Postoperative Survival in Patients Who Underwent Surgery 1971 to 1989 Versus 1990 to 2011

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<td>468</td>
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<td>(&lt;0.0001)</td>
<td>62.4</td>
<td>86.9</td>
<td>(&lt;0.0001)</td>
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<td>(&lt;0.0001)</td>
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<td>4459</td>
<td>89.5</td>
<td>96.3</td>
<td>(&lt;0.0001)</td>
<td>79.0</td>
<td>90.4</td>
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*Abbreviations are given in Table 1.
†Valvular aortic stenosis, valvular pulmonary stenosis, or mitral valve defect.
‡Atrial septal defect or partially anomalous pulmonary venous drainage.
Survival Differences in Patients Operated on in 1990 to 1999 and 2000 to 2011

Table 4 focuses on survival among patients who underwent surgery during the past 2 decades. Improvements were found particularly for the most severe defects. Early survival in complex defects among patients operated on in 1990 to 1999 versus 2000 to 2011 was 90.7% versus 97.2% (P<0.0001), and cumulative survival until 16 years of age was 80.2% versus 91.2% (P<0.0001). Significant improvements also occurred in simple defects.

Survival Differences in Patients Operated on in 2000 to 2004 and 2005 to 2011
Survival in complex defects improved significantly even among patients operated on after the year 2000 (Figure 2). Thus, early survival in patients operated on in 2000 to 2004 versus 2005 to 2011 was 93.6% versus 99.1% (P<0.0001), 1-year survival was 90.7% versus 96.5% (P<0.0001), and 5-year cumulative postoperative survival was 88.8% versus 95.0% (P<0.0003). One-year postoperative survival among patients operated on in 2005 to 2009 versus 2010 to 2011 was 95.3% versus 99.6% (P=0.003).

Among patients with UVH operated on in 2000 to 2004 (n=98) versus 2005 to 2011 (n=155), 1-year postoperative survival was 82.5% versus 94.0% (P=0.0002), and, in patients with HLHS, 1-year survival was 69.8% (n=53) versus 93.3% (n=60; P=0.0007).

Reoperations
The 2918 patients with complex defects had 4500 operations before the age of 16 years. At least 1 reoperation was performed in 985 (33.8%) patients, at least 2 reoperations were performed in 389 (13.3%) patients, and at least 3 reoperations were performed in 131 (4.5%) patients. In those with simple defects, 434 (11.6%) patients underwent at least 1 reoperation.

Among patients with complex defects who had their index operation in 1990 to 1999 versus 2000 to 2011 (767 versus 1291 patients), 308 versus 353 (40.2% versus 27.3%) had at least 1 reoperation, 156 versus 113 (20.3% versus 8.8%) had at least 2 reoperations, and 54 versus 30 (7.0% versus 2.3%) had at least 3 reoperations. Furthermore, there were 15 versus 29 catheter interventions (P=not significant). The Kaplan–Meier plots in Figure 3 show that the great majority of reoperations were performed within a few years after the index operation, and that significantly fewer patients needed reoperations in 2000 to 2011 than in 1990 to 1999, even when catheter interventions are taken into account. The estimates in Table 5 indicate trends toward increased reoperation-free survival for most complex defects, especially in patients with I/HAA and PA.

Cardiopulmonary bypass was used in 72.5% of the index operations in patients with complex defects in 1990 to 1999, and in 86.9% in 2000 to 2011 (P<0.01). Corresponding percentages for (first) reoperations were 83.9% and 92.8% (P<0.01). The proportions of index operations classified as

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<td>UVH</td>
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<td>98.0</td>
<td>0.097</td>
</tr>
<tr>
<td>VALV†</td>
<td>64</td>
<td>101</td>
<td>90.6</td>
<td>97.0</td>
<td>0.08</td>
<td>90.5</td>
<td>92.5</td>
<td>0.57</td>
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<tr>
<td>ASD§</td>
<td>239</td>
<td>187</td>
<td>99.2</td>
<td>–</td>
<td>–</td>
<td>98.6</td>
<td>97.4</td>
<td>0.31</td>
</tr>
<tr>
<td>PDA</td>
<td>215</td>
<td>280</td>
<td>95.2</td>
<td>96.7</td>
<td>0.29</td>
<td>86.3</td>
<td>93.6</td>
<td>0.0059</td>
</tr>
<tr>
<td>All simple</td>
<td>1036</td>
<td>1109</td>
<td>97.5</td>
<td>98.8</td>
<td>0.010</td>
<td>93.2</td>
<td>95.9</td>
<td>0.0083</td>
</tr>
<tr>
<td>MISC</td>
<td>84</td>
<td>172</td>
<td>91.6</td>
<td>97.7</td>
<td>0.025</td>
<td>75.9</td>
<td>90.9</td>
<td>0.0090</td>
</tr>
<tr>
<td>Total</td>
<td>1887</td>
<td>2572</td>
<td>94.4</td>
<td>97.9</td>
<td>&lt;0.0001</td>
<td>87.1</td>
<td>93.2</td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

*Abbreviations are given in Table 1.
†Maximum 13 years postoperative follow-up in patients operated on in 2000–2011.
‡Valvular aortic stenosis, valvular pulmonary stenosis, or mitral valve defect.
§Atrial septal defect or partially anomalous pulmonary venous drainage.
corrections were 70.5% in 1990 to 1999, and 78.6% in 2000 to 2011 ($P<0.01$).

**Discussion**

To our knowledge, this is the first long-term prospective study of its kind. Our data demonstrate that, over the past 40 years, there have been substantial improvements in postoperative survival within all subgroups of complex CHDs, with early mortality now approaching zero. Parallel to these improvements, there has been a decrease in the need for reoperations. This development has taken place despite the fact that increasing proportions of complex cases have been operated on, and despite a marked decrease in age at the first operation. In children with defined simple defects, survival was already so high in the late 1970s that only moderate further improvements were possible. In the following, we have therefore focused on children with complex defects.

**Number of Patients**

The birth incidence of CHD appears constant, possibly excluding the most complex defects (eg, HLHS) in some countries owing to abortions after prenatal echocardiography. The increasing number of operations for complex defects therefore implies that an increasing proportion of patients underwent surgery following improvements in preoperative diagnostics, surgical techniques, anesthesiology, and intensive care. Improved preoperative diagnostics has facilitated both the identification and characterization of patients that may be amenable for surgery. Surgical corrections that were judged impossible or too dangerous in the past, or which had not yet been developed, may now be performed with good results and acceptable risk. Notably, among recent advances are new surgical techniques such as the Norwood procedures in patients with HLHS, the arterial switch procedure in patients with TGA, staged palliation for patients with UVH, and 1-stage procedures.
palliation in patients with I/HAA and ventricular septal defect.\textsuperscript{17} Few CHDs are now beyond the reach of surgical treatment, even at a very young age, exemplified by the fact that median age at first operation in patients with complex defects was 1.6 years in 1971 to 1979 and only 0.19 years in 2000 to 2011. A sizeable proportion of children with CHD succumbed at an early age during the early phases of the study, especially those with the most severe defects. Most of these children now undergo surgery shortly after birth, thereby increasing the size and complexity of the population offered operation.

The decrease in the number of patients with certain simple CHD undergoing surgery probably follows an increase in invasive catheter treatment. For example, the number of pulmonary balloon valvuloplasty procedures increased from 89 in 1990 to 1999 to 150 in 2000 to 2011, device closures of ASD increased from 29 to 260, and balloon dilatations of COA increased from 9 to 17. An increasing availability of these techniques may have lowered the threshold for treating PS and ASD, but not COA.

Approximately 80% of all CHD surgery in Norway was performed at Rikshospitalet until 2003, and 100% was performed at Rikshospitalet from 2004 to 2011. Accordingly, the reported increase in the number of patients from 1990 to 1999 to 2000 to 2011 (Table 2) was somewhat larger than it would have been without the policy change in 2004. However, the increase in the number of complex cases was steeper than what would have been expected from a purely proportional expansion of the total patient population.

### Survival

In addition to a report from our hospital on a subcohort of 970 children operated on for CHD between 1990 and 1999,\textsuperscript{7} 3 other studies specifically targeting survival after CHD surgery have been published during the past decade, all based on retrospective data. Nieminen et al\textsuperscript{5} reported that, among patients <15 years of age who underwent surgery in 1953 to 1989 (75% in 1970–1989), 15-year survival in patients with UVH, TGA, and TOF was 45%, 65%, and 82%, respectively. In our study, 15-year survival among patients <15 years of age who were operated on for UVH in 1971 to 1989 was 36.2%; for TGA, 15-year survival was 64.7%; and for TOF, 15-year survival was 87.5%. In a study including 850 children in Denmark, Larsen et al\textsuperscript{4} reported that, among all CHD patients <15 years of age who underwent surgery in 1996 to 2002, 8-year survival was 86%. In our study, overall survival after 8 years was 90.2% among patients <15 years of age who were operated on in 1996 to 2002. Based on a population from 38 American states, Marelli et al\textsuperscript{18} found that 30-day postoperative survival in all CHD patients <18 years of age who were operated on in 2000, 2003, and 2006 was 95.8%, 95.5%, and 96.5%, respectively. However, the study did not discriminate between different CHD diagnoses, and no data on long-term follow-up were given. In our study, 30-day postoperative survival in 2000, 2003, and 2006 when taking all CHD subgroups (Table 1) together was 96.3%, 96.3%, and 97.6%.

Four additional studies on CHD survival have been published during the past decade on children with severe or critical CHD, all of whom probably underwent CHD operations,\textsuperscript{19–22} but no specific data on surgery were reported.

Based on the most recent data available in 2001, Warnes et al\textsuperscript{23} estimated that children with complex CHD born in the 1980s had a 1-year survival rate of 85%. Our data show marked further improvements in survival during the following 2 decades in all complex CHD subgroups (Figure 1). In fact, since the era studied by Warnes et al, 1-year survival in complex defects has increased almost linearly, now approaching the 100% limit (Figure 4).

Despite the early gain in survival, no increase in late mortality was seen. This appeared to be the case especially among the patients operated on from 2000 to 2011. Because nearly all (99.1%) of the children with complex defects operated on in 2005 to 2011 survived the early 30 days postoperative period, further reductions in mortality can be achieved almost exclusively from improvements in late survival. However, the potential for further improvements is narrowing—as suggested by the fact that CHD-related mortality in Norwegian children was approximately constant after 2005 (Figure 5)—in good agreement with data from the United Kingdom.\textsuperscript{24}

### Reoperations

About one-third of the patients with complex defects who were initially operated on in 1990 to 2011 needed reoperation before 16 years of age, similar to findings reported from studies in the United Kingdom\textsuperscript{25} and in the United States.\textsuperscript{26} However, parallel to the improvements in postoperative survival (Figure 2), there was also a significant increase in reoperation-free survival from 1990 to 1999 to 2000 to 2011. We are not aware of other studies reporting on sequential changes in reoperations during this period. Further studies are needed to explore the full clinical impact of these developments, eg, on quality of life.

Table 5 demonstrates increases in reoperation-free survival in most complex CHD subgroups, particularly marked in patients with I/HAA and PA, and Table 2 shows that there was a particularly marked increase in the number of patients

### Table 5. Freedom of Reoperation in Patients With Complex Defects Who Were Initially Operated on in 1990 to 1999 Versus 2000 to 2011

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
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<tbody>
<tr>
<td>Freedom of Reoperation†</td>
<td>Freedom of Reoperation†</td>
<td>P Value</td>
</tr>
<tr>
<td>UVH</td>
<td>125</td>
<td>0.16</td>
</tr>
<tr>
<td>TAC</td>
<td>23</td>
<td>0.42</td>
</tr>
<tr>
<td>I/HAA</td>
<td>66</td>
<td>0.79</td>
</tr>
<tr>
<td>TGA</td>
<td>152</td>
<td>0.87</td>
</tr>
<tr>
<td>AVSD</td>
<td>176</td>
<td>0.84</td>
</tr>
<tr>
<td>TAPVD</td>
<td>39</td>
<td>0.89</td>
</tr>
<tr>
<td>PA</td>
<td>44</td>
<td>0.32</td>
</tr>
<tr>
<td>TOF</td>
<td>142</td>
<td>0.55</td>
</tr>
<tr>
<td>All complex defects</td>
<td>767</td>
<td>0.66</td>
</tr>
</tbody>
</table>

*Abbreviations are given in Table 1.
with I/HAA, PA, and UVH being operated on during the study period. However, the reoperation-free survival figures given for patients with UVH should be interpreted with caution, because, in 1990 to 1999, not all patients with HLHS were operated on in Norway.8 Among patients who typically underwent primary correction (like TGA, atrioventricular septal defect, and totally anomalous pulmonary venous drainage), changes in reoperation-free survival were modest.

These developments coincided with declining age at the first operation and increasing proportions of corrections rather than palliations. The lower age at first operation has contributed to increasing numbers of operations in the most complex cases, because many of these patients previously succumbed at an early age. Early corrections may prevent complications caused by the hazards associated with the defects themselves. Once it has been proved that primary corrections can be performed successfully with low risk, the need for palliations and reoperations will obviously be reduced.

In summary, our data indicate that, in general, an aggressive surgical strategy aimed at primary correction instead of palliation during the very first few weeks after birth may be advocated.

Limitations
We divided CHDs into complex and simple defects based on a diagnosis complexity hierarchy adopted by Nieminen et al,5 acknowledging that the degree of severity varies widely within the complex subgroup, and even within each diagnostic category. Table 3 shows that the complex versus simple scheme fits best for patients operated on from 1971 to 1989. However, even in patients operated on from 1990 to 2011, all complex defects, with the exception of TOF, had a poorer prognosis than simple defects, which may justify the complex versus simple classification. Importantly, the assumed complexity of a certain defect depends on the assumed complexity of surgical correction, which probably changed for most heart defects during the long study period. Since risk assessment scores based on surgical complexity have been used internationally for only ≈10 years,27 such scores were not applied in the present study.

Some previous studies classified TOF as critical22 or severe,21 whereas others judged it to be moderately severe.19,23 Despite successful surgical corrections in the 1970s, we find it difficult to categorize TOF as anything but a complex defect.

The data presented show outcomes in a single, medium-sized center with low selection bias, because access to health
services in Norway is free, and because Norwegian children with few exceptions are born in hospitals. Accordingly, it is likely that the great majority of cases with complex CHD were early diagnosed and immediately referred for care. However, before 2003, some of the most severe complex cases were transferred to our institution, suggesting that postoperative mortality in children operated on before 2003 was higher than it might have been in an unselected population.

The ethical distribution in Norway changed during the 40-year study period because of considerable immigration. Among children operated on in 2000 to 2011, about one-sixth had names suggesting Asian or African origin, whereas in 1971 to 1979 only 1 such patient was operated on.

Perspectives

Essentially, our article deals with data from a continuous, 4-decade quality control and improvement project, acknowledging that controlled trials in children with CHD are difficult, if not impossible, for ethical reasons. By continuously introducing state-of-the-art improvements in diagnostics, anesthesiology, surgical techniques, and postoperative care, a level has been reached where virtually all CHDs are now within reach of surgical correction with acceptable risk. This advance makes it easier for health professionals to communicate with parents for the life of their child.

Conclusion

This prospective study covering the past 40 years of >80% of CHD surgery in Norwegian children with 98.5% complete follow-up demonstrates very marked, gradual improvements in both short- and long-term postoperative survival despite increasing proportions of complex cases becoming amenable for surgery, and despite a gradual and marked decline in age at operation. Moreover, a significant reduction in the need for reoperations is observed, probably mainly as a result of performing more primary corrections as opposed to primary palliations. Future challenges include further refinements and education in surgical techniques and implementation of risk stratification algorithms.

Acknowledgments

We express our deepest gratitude to Susan Tower Gibbs, RN at the Department of Thoracic Surgery, Rikshospitalet, for initiating the work on the database in 1971.

Disclosures

None.

References


CLINICAL PERSPECTIVE

Following major treatment advances during the past 50 years, surgical correction of nearly all congenital heart defects (CHDs) is now possible, and substantial improvements in short-term survival have been documented for most CHDs. However, few population-based studies have been published on long-term survival after CHD surgery since 1990. The present prospective study, covering the past 40 years of >80% of CHD surgery in Norwegian children <16 years of age, demonstrates sequential improvements in short- and long-term postoperative survival, even during the past 2 decades. The great majority of defined complex cases now survive childhood. These improvements have occurred parallel to declining age at operation, declining reoperation rates, and increasing numbers of complex cases becoming amenable for surgery. The data from the present study support an aggressive strategy aimed at early surgical correction in children with CHD.
Achievements in Congenital Heart Defect Surgery: A Prospective, 40-Year Study of 7038 Patients
Gunnar Erikssen, Knut Liestøl, Egil Seem, Sigurd Birkeland, Kjell Johan Saatvedt, Tom Nilsen Hoel, Gaute Døhlen, Helge Skulstad, Jan Ludvig Svennevig, Erik Thaulow and Harald Lauritz Lindberg

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