Long-Term Outcome of Mustard/Senning Correction for Transposition of the Great Arteries in Sweden and Denmark

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Background—The atrial switch operation, the Mustard or Senning operation, for the transposition of the great arteries (TGA) was introduced in the late 1950s and was the preferred surgery for TGA until the early 1990s. The Mustard and Senning operation involves extensive surgery in the atria and leaves the right ventricle as the systemic ventricle. The Mustard and Senning cohort is now well into adulthood and we begin to see the long-term outcome.

Methods and Results—All the 6 surgical centers that performed Mustard and Senning operations in Sweden and Denmark identified all operated TGA patients. Information about death was obtained in late 2007 and early 2008 from the Danish and Swedish Centralised Civil Register by using the patients' unique national Civil Registration Numbers. Four hundred sixty-eight patients undergoing the atrial switch operation were identified. Perioperative 30-day mortality was 20%, and 60% were alive after 30 years of follow-up. Perioperative mortality was significantly increased by the presence of a ventricular septal defect, left ventricular outflow obstruction, surgery early in the Mustard and Senning era. However, only pacemaker implantation is predictive of long-term outcome (hazard ratio, 1.90; 95% confidence interval, 1.05–3.46, P=0.04), once the TGA patient has survived the perioperative period. The risk of reoperation was correlated to the presence of associated defects and where the first Mustard/Senning operation was performed.

Conclusions—The long-term survival of patients with Mustard and Senning correction for TGA appears to be primarily determined by factors in the right ventricle and tricuspid valve and not the timing of or the type of surgery in childhood. Cardiac function necessitating the implantation of a pacemaker is associated with an increase in mortality. **(*Circulation*. 2015;132:633-638. DOI: 10.1161/CIRCULATIONAHA.114.010770.)**

Key Words: heart defects, congenital ■ pacemaker, artificial ■ population ■ prognosis ■ survival ■ transposition of great vessels

Transposition of the great arteries (TGA) is a congenital heart defect where the arterial trunks are connected to the morphologically inappropriate ventricles. Unpalliated TGA has a 90% mortality within the first year of life.[1] Radical surgery in the form of the atrial switch operations was introduced by Åke Senning and William Mustard,[2,3] and these procedures were routinely performed in the 1970s. In the 1980s, the arterial switch operation (ASO) replaced the Mustard and Senning operation.[4] The initial short follow-up showed satisfactory results of ASO in comparison with the Mustard and Senning correction, but long-term results have only recently been reported.[5–8] The patient population with Mustard and Senning correction is now well into adulthood, making long-term follow-up possible. The population of patients with Mustard and Senning correction represents a clinical challenge and can provide us with important insight into the consequences of having a morphological right ventricle as a systemic ventricle. Sweden and Denmark have a population of 14.5 million. The Scandinavian social structure with general access to qualified health care and population-based validated registries creates a unique opportunity for long-term follow-up of large numbers of patients. The centralized structure of tertiary cardiology centers and congenital surgery allows us to study the whole population of Sweden and Denmark over a long period of time. The present study provides outcome data from a large, contemporary cohort of unselected, consecutive patients with TGA who underwent Mustard and Senning procedures in Sweden and Denmark from 1967 to 2003.

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Methods

We performed a retrospective analysis of the medical records of patients who had either simple or complex TGA and who had undergone either Mustard or Senning operations. Historically, Sweden and Denmark had

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6 centers performing congenital heart surgery. (The congenital heart centers were located in Copenhagen and Skejby in Denmark, and in Stockholm, Uppsala, Lund, and Gothenburg in Sweden). All operation logbooks were reviewed and, when a Mustard and Senning operation was recorded, the patient was included in the study for further review of medical records according to a prespecified protocol. Patients with TGA who had a Rastelli correction or had more complex anatomy, such as double-outlet or single-ventricle physiology, were excluded. Operation logbooks were examined by experienced cardiologists subspecialized in congenital heart disease at the respective units, who also performed the data extraction from hospital records in accordance with a prespecified data collection form. In this cohort, the first patient* was operated on in 1967 and the last Mustard operation was performed in 2003. In all 468 patients with TGA were identified and included in the study. In late 2007 and early 2008, all patients were cross-checked against the national death registries to verify mortality data and to define the date of death. The data were anonymized. Twenty-three patients from the operation diaries could not be identified and were not found in the death registry; they were presumed to have died during or immediately after the operation. The substudy in Sweden was approved by the ethics committee in Gothenburg (Dnr. 174-08, http://www.epn.se). Ethics approval for anonymized retrospective studies of registries was not required in Denmark, but the study was approved by the Danish Datatilsynet (J.nr. 2009-41-3319, http://www.datatilsynet.dk)

Statistical Analysis

The TGA patients were divided into 2 groups according to the time of surgery. The early era was defined as surgery performed before or on the median date (November 4, 1980); late era was defined as surgery performed after this date. Associations between categories of variables were measured by the χ^2 test or trend test, and Student *t* test was used for continuous variables. Binary logistic regression with backward elimination was performed to identify predictors of perioperative mortality (death within 30 days following surgery). Kaplan–Meier plots were used to illustrate survival curves, and the Cox proportional hazards model was used for initial univariate comparisons. Multivariable comparisons were performed by using a Cox proportional hazards model (fitted by backward elimination using a threshold of $P<0.1$ for elimination) after checking assumptions of proportionality. Additional Cox analyses of events (pacemaker implantation or reoperation) during the follow-up period were performed with these variables included as time-dependent covariates. Continuous variables are summarized as means and 95% confidence intervals (CI), whereas categorical data are summarized as frequencies and percentages. Statistical calculations were done with SPSS version 20.0 (SPSS Inc, Chicago, IL).

Results

This analysis included all (n=468) patients operated on for TGA in Denmark and Sweden with either the Mustard or Senning operation between 1967 and 2003. Mean age at the time of surgery was 1.9 years (95% CI, 1.7–2.2 years) and 319 (68.2%) were males. Baseline characteristics of the included patients divided by country appear in Table 1. Patients in Denmark were operated on significantly earlier than patients in Sweden, and the Mustard operation was performed significantly more frequently in Denmark than in Sweden. There were no statistically significant differences in sex or frequency of associated heart defects between the 2 countries. The majority of children (385 [82%]) with TGA were operated on before the age of 3 and very few were operated on after the age of 6 (25 [5%]). Patients operated on in the early era were significantly older (2.6. years; 95% CI, 2.2–3.0 years) than those operated on in the late era (1.3 years; 95% CI, 1.2–1.4 years), $P<0.01$. Perioperative mortality (death within 30 days following surgery) was high (93 [20%]). In multivariable logistic regression, only the era in which the patients were operated on and the presence of an associated heart defect were associated with perioperative death (see Table 2).

Table 1. Baseline Characteristics and Demographics

	Denmark, n=193	Sweden, n=275	*P* Value
Female sex, n (%)	61 (32)	88 (32)	0.93
Mustard operation, n (%)	186 (96)	124 (45)	<0.01
Age at operation, y, mean±SD	1.6±2.1	2.1±2.8	0.03
VSD, n (%)	64 (33)	70 (25)	0.07
LVOTO, n (%)	14 (7)	22 (8)	0.77
LVOTO and VSD, n (%)	5 (3)	15 (6)	0.13

LVOTO indicates left ventricular outflow tract obstruction; SD, standard deviation; and VSD, ventricular septal defect.

Median follow-up was 26.1 years (range, 0.4–60 years); 4 (<1%) patients emigrated and were lost for follow-up. During the observation period, pacemaker implantation was performed in 63 patients (15%), reoperation was performed in 27 patients (7%), and 184 (39%) of the patients died or had a heart transplant (176 [38%] died and 8 [2%] had a heart transplant).

Implantation of a pacemaker was not associated with age at operation, sex, country, era of surgery, Mustard versus Senning, or the presence of left ventricular outflow obstruction (LVOTO) and ventricular septal defect (VSD) in either uni- or multivariable analyses (data not shown).

The Kaplan–Meier curve of survival for all patients in the study shows a high perioperative mortality followed by a long period of low mortality (Figure 1).

Kaplan–Meier curves of time to reoperation divided into early versus late surgery, Mustard versus Senning, and associated heart defects are presented in Figure 2A through 2C . Even though it seems that the Mustard operation was strongly associated with reoperation after performing a multivariable Cox regression analysis, only surgery performed in Denmark was independently associated with reoperation (see Table 3). Only 5 (19%) of the 27 reoperations were performed in Sweden. Having LVOTO or VSD, or both, did not increase the risk of redo of the atrial switch.

Kaplan–Meier curves of time to death or heart transplantation after primary surgery divided into early versus late era, type of surgery, and associated heart defects are presented in Figure 3A through 3C. As presented in Table 2, only surgery in the early era (ie, surgery performed after November 4, 1980) and LVOTO and VSD were associated with increased mortality. This was true both in a univariable analysis and in a multivariable analysis including all variables (see Table 4). However, if all patients who died within 30 days of surgery (perioperative death) were excluded from the analyses, then none of the variables were associated with mortality, either in uni- or multivariable analyses (data not shown).

If pacemaker implantation was included as a time-dependent variable in a Cox regression analysis of time to death or heart transplant the results were unaltered; however, if patients who died perioperatively were excluded, then pacemaker implantation is the only variable that was associated with

*The first atrial switch operation, performed by Åke Senning in 1958,[3] was not included because the patient was not living in Denmark or Sweden. After Senning's initial landmark operation, no Mustard and Senning procedures could be found in the logbooks until 1967 (ie, 9 years later).

Table 2. Multivariable Logistic Regression Testing Factors Associated With Perioperative Death

	Univariable Analyses		Mulitivariable Analysis	
	Estimated Odds Ratio	*P* Value	Estimated Odds Ratio	*P* Value
Age at operation, y	1.01 (0.92–1.10)	0.90		
Female sex	1.09 (0.67–1.77)	0.73		
Surgery performed in Denmark	1.23 (0.80–2.04)	0.31		
Surgery performed after November 4, 1980	0.40 (0.25–0.65)	<0.01	0.39 (0.24–0.64)	<0.01
Mustard surgery	1.02 (0.62–1.67)	0.94		
Associated heart defect(s)	2.04 (1.28–3.24)	<0.01	1.99 (1.22–3.23)	<0.01

Only early surgery (early in the Mustard and Senning era) and associated defects (LVOTO and VSD) were found to influence perioperative death significantly. This analysis is from the time of surgery. LVOTO indicates left ventricular outflow tract obstruction; and VSD, ventricular septal defect.

increased mortality (hazard ratio, 1.90; 95% CI, 1.05–3.46; *P*=0.04). In a similar analysis where reoperation was included as a time-dependent variable, the Cox regression analysis showed that reoperation was not associated with increased risk of mortality and heart transplantation.

There were 6.2 million live births in Denmark and Sweden between 1967 and 2003 (Statistic Sweden [SBC] and Dansk Statistik). The incidence of TGA is 312 per million,[9] so ≈1900 children with TGA should have been born in the study period. Only 468 had a Mustard/Senning operation, and a very small number of the TGA patients had an ASO in the surgical transition period in the late1990s.

Discussion

To the authors' knowledge this is the largest study of survival after the atrial switch operation for transposition of the great arteries. The study includes all patients operated on with the Mustard or Senning operation in Denmark and Sweden from 1967 through 2003 (n=468). From the incidence of TGA in Scandinavia, one can calculate that the majority of children born with TGA must have died before a Mustard or Senning operation was even attempted. In a previous report from western Sweden early in the era, all patients born with TGA were investigated.[10] This study found that 40% children born and diagnosed with TGA died before surgery, most likely because of hypoxia or heart failure. Even after atrial septostomy, patients with TGA have a high mortality, and this is rarely discussed when comparing the Mustard/Senning operation with ASO, an operation most often performed before the child is 1 month of age. The patients in this study are similar to other populations of TGA[8,11] with a male:female ratio of 2:1. Most patients had an atrial communication made shortly after birth either with the Rashkind procedure or the Blalock-Hanlon operation. The frequency of Mustard/Senning procedures performed in this Danish and Swedish population was 35 per million, which is similar to the incidence reported by Moons and coworkers from Belgium (34 per million),[11] suggesting a similarly high preoperative mortality in children with TGA in that population. For almost 2 decades, the ASO has replaced the atrial switch operations, which means that the patient population with a Mustard or Senning repair will disappear over the next 40 years. The main reason for studying the Mustard and Senning population in the 21st century is to determine what predicts mortality in the current adult Mustard and Senning population, and it is also an opportunity to study the right ventricle in the systemic position. We found that the perioperative mortality was 20%, which is much higher than revealed in most reports.[12–15] We could find only 1 recent report of similar high mortality,[11] which is also a population-based study, and not a report from a tertiary center. We speculate that the historic reports of low perioperative mortality could be explained by patient selection in tertiary centers and publication bias, and we suggest that population-based studies better reflect the risk for a patient with a congenital heart defect. There is also a learning curve for complex surgery and postoperative care, and studies from the 1980s, late in the Mustard and Senning era, show very low perioperative mortality.[16,17] That there was a higher perioperative mortality in smaller centers like those in Denmark and Sweden than in the large international high-output surgical centers was undoubtedly also the case. It is unlikely that such differences in perioperative mortality would have been tolerated today.

The primary finding in this study is that several factors normally considered important for long-term survival (operation type [Mustard versus Senning], age at operation, institution where the operation was performed, operation early or late in the era, associated defects such as VSD and LVOTO) are not

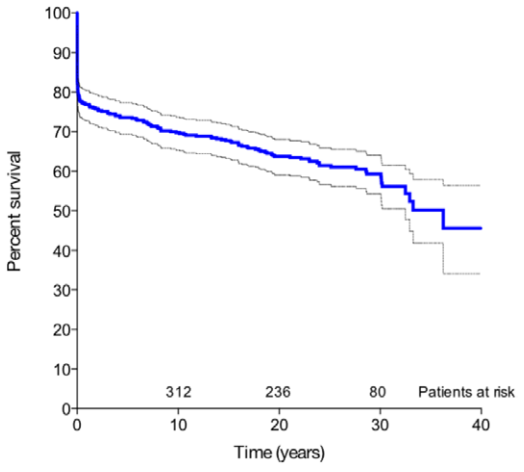

Figure 1. Heart transplantation–free survival after surgery. Kaplan–Meier plot with 95% confidence interval. Perioperative mortality was >20%. The knee on the curve is very likely the result of censoring.

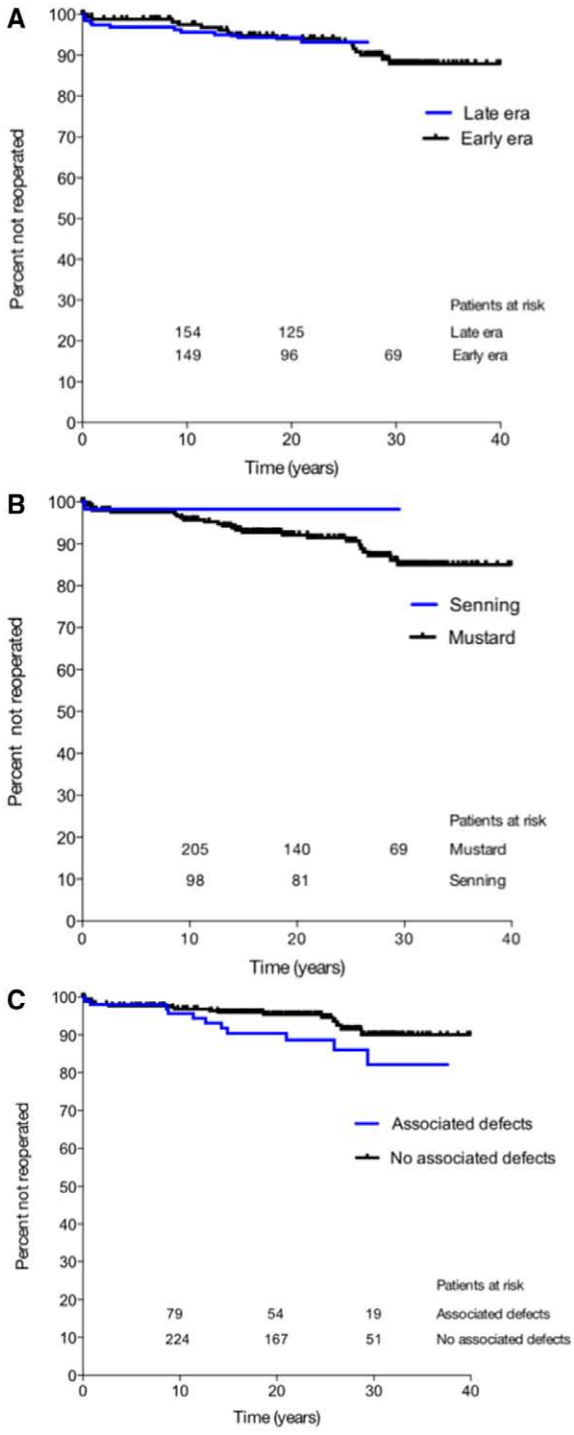

Figure 2. Reoperation-free survival. The survival curves starts at 30 days, to show the survival that is not directly dependent on the preoperative mortality. **A**, Reoperation-free survival in the early and late era. **B**, Reoperation-free survival for Mustard and Senning patients. **C**, Reoperation-free survival with and without additional lesions (VSD and LVOTO). A multivariable Cox regression analysis shows that none of these factors are significant (Table 4). LVOTO indicates left ventricular outflow tract obstruction; and VSD, ventricular septal defect.

statistically significantly associated with long-term survival. In the early part of the Mustard and Senning era, patients were operated on late. It has been suggested that longstanding preoperative hypoxia might be an important risk factor for poor long-term

Table 3. Reoperation

	Univariable Analyses		Mulitivariable Analysis	
	HR (95% CI)	*P* Value	HR (95% CI)	*P* Value
Age at operation, y	0.92 (0.75–1.12)	0.38		
Female sex	0.77 (0.49–1.22)	0.27		
Surgery performed in Denmark	7.77 (2.93–20.58)	<0.01	7.77 (2.93–20.58)	<0.01
Surgery performed after November 4, 1980	0.96 (0.42–2.23)	0.93		
Mustard surgery	5.25 (1.24–22.30)	<0.01		
Associated heart defect	1.91 (0.89–4.12)	0.10		

In a multivariable analysis only surgery performed in Denmark was associated with an increased risk of redo of the Mustard and Senning repair. CI indicates confidence interval; and HR, hazard ratio.

outcome, but we did not find any evidence for that. There are several reports describing the advantages of the Senning procedure relative to the Mustard procedure.[15,18] This study could not show any statistically significant difference in long-term survival between patients operated on with the Senning or the Mustard procedure (Figure 3). Long-term survival appears to be primarily determined by how well the right ventricle and the tricuspid valve tolerate systemic afterload and blood pressure. Patients with congenitally corrected TGA have a long-term survival similar to patients operated on with the Mustard and Senning procedures, even though many patients with congenitally corrected TGA have never undergone cardiac surgery.[19–21]

Cardiac function necessitating implantation of a pacemaker is associated with an increase in mortality (hazard ratio, 1.90; 95% CI, 1.05–3.46; P=0.04); this could be because the need for a pacemaker is a sign of other impending problems, or it could be that pacing has a deleterious effect on the systemic right ventricle.

The risk of reoperation was not associated with the type of surgery or the presence of additional defects, but was associated with who did the surgery. Denmark had a statistically significant higher number of reoperations after atrial repair than Sweden (Table 3). A less-than-perfect Mustard or Senning repair does not damage the right ventricle but will increase the risk of needing a reoperation on the atrial baffles. However, reoperation was not found to be associated with a statistically significant effect on long-term survival.

This study suggests that, when caring for survivors after Mustard and Senning repair, the clinical focus should be on the function of the right ventricle and tricuspid valve. Operation notes will be less helpful in providing information useful for predicting long-term survival. Special attention should be given to Mustard and Senning patients with pacemakers because they have increased mortality.

The population of Mustard and Senning patients will disappear over the next 40 years, but cardiologists will always have to treat patients with congenitally corrected TGA where knowledge of how the right ventricle tolerates systemic afterload

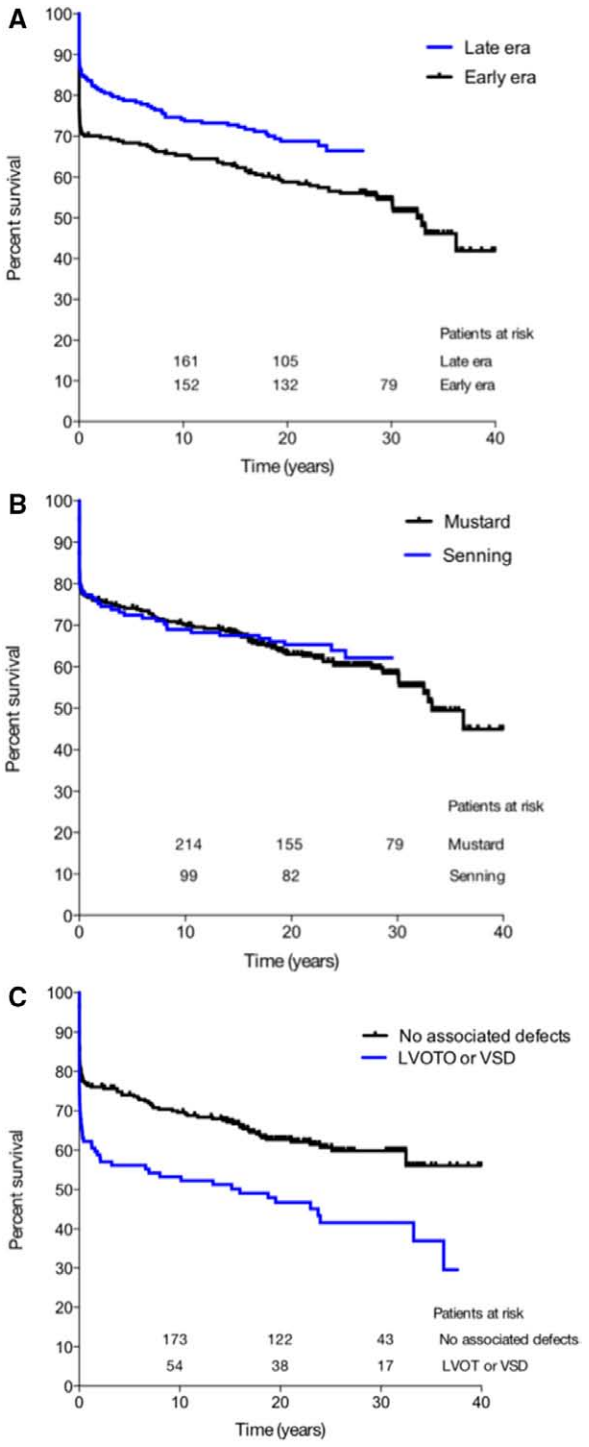

Figure 3. Kaplan–Meier curves for HTX-free survival. These survival curves start from the time of surgery and also reveal the perioperative mortality. **A**, HTX-free survival in the early and late era. **B**, HTX-free survival for Mustard and Senning patients. **C,** HTX-free survival with and without additional lesions (VSD and LVOTO). A multivariable Cox regression analysis shows that none of these factors are significant. HTX indicates heart transplantation; LVOTO, left ventricular outflow tract obstruction; and VSD, ventricular septal defect.

could be helpful. In future comparisons with ASO, one should keep in mind that one of the major advantages of the ASO procedure is, that the ASO is performed very early (2–6 weeks postpartum), because many children with TGA died before the atrial switch operation. Today, atrial correction could also be performed in the neonatal period[22] and most likely with similar pre- and perioperative mortality as the ASO. The importance of physiological correction of TGA is supported by the observation that 25 years of perfecting the Mustard and Senning surgery did not significantly change the long-term survival.

Another observation relevant to future studies of congenital heart disease is that the predictors of perioperative survival can be very different from what determines long-term survival, and survival plots without the perioperative mortality could sometimes be more clinically relevant in adult congenital heart disease.

Limitation of the Study

Twenty-three patients who underwent a Mustard/Senning procedure were not found in the death registry and they were presumed to have died early postoperatively before they were given the national 10-digit personal number in the national population registry system. If they are assumed to be dead but if they are actually still alive, the perioperative mortality is overestimated by 5% and long-term follow-up rate is reduced to 95%.

The reported number of patients with pacemaker insertion might have been underestimated, because all patients were not followed by dedicated centers and pacemaker could have been inserted at the local hospital and not found in the medical records.

Conclusion

We identified all 468 TGA patients operated on with the Mustard or Senning operation in Sweden and Denmark. The 30-day perioperative mortality was high (20%). Perioperative mortality was statistically significantly correlated with the presence of LVOTO and VSD, and whether surgery was performed before or after 1980. None of the factors important for perioperative mortality had any influence on long-term mortality. Implantation of a pacemaker was the only factor identified to have an adverse effect on long-term mortality.

Clinical Impact of this Study

What is important for long-term survival in patients palliated with the atrial switch procedures is often discussed. This study

Table 4. Univariable and Multivariable Analysis of Death and Heart Transplantation

	Univariable Analyses		Mulitivariable Analysis	
	HR (95% CI)	*P* Value	HR (95% CI)	*P* Value
Age at operation, y	0.98 (0.92–1.04)	0.44		
Female sex	1.02 (0.87–1.20)	0.76		
Surgery performed in Denmark	1.02 (0.76–1.38)	0.88		
Surgery performed after November 4, 1980	0.68 (0.50–0.92)	0.01	0.68 (0.50–0.92)	0.01
Mustard surgery	0.93 (0.67–1.28)	0.64		
Associated heart defect	1.60 (1.18–2.15)	<0.01	1.63 (1.21–2.18)	<0.01

In the multivariable analysis only surgery performed after November 4, 1980 and associated heart defect stayed in the model. This analysis is from the time of surgery. CI indicates confidence interval; and HR, hazard ratio.

finds that the type of operation, the time of operation, and even the institution of operation do not predict long-term survival and need for transplantation. Long-term survival is good in adulthood, and the knee on the Kaplan–Meier plot probably is the result of censoring. Only the need for implantation of a pacemaker is associated with poorer long-term survival. This study also suggests that in adult congenital heart disease survival curves that only look at survival in adulthood could be more relevant for clinical practice.

Disclosures

None.

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CLINICAL PERSPECTIVE

This is, to our knowledge, the largest studied cohort of patients with Mustard and Senning repair of the transposition of the great arteries. Transposition of the great arteries is a common cyanotic heart defect. The Mustard and Senning repair (atrial switch repair) of transposition of the great arteries was used in the 1960s, 1970s, 1980s, and early 1990s. Centers treating adults with congenital heart disease will have a cohort of patients who have had atrial switch repair, and the clinicians will want to identify those patients who have the highest risk of needing reoperation or who are at risk of dying. The atrial switch repair leaves the right ventricle in the systemic position and subjects the right ventricle to systemic afterload. This study finds that the type of repair (Mustard/Senning) is not statistically significantly associated with long-term survival or the risk of reoperation. The risk of reoperation is determined by who did the surgery, and the only factor we could identify that is statistically significantly associated with increased mortality is the implantation of a pacemaker.