ORIGINAL RESEARCH

Cause-Specific Mortality in Patients During Long-Term Follow-Up After Atrial Switch for Transposition of the Great Arteries

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BACKGROUND: Little is known about the cause of death (CoD) in patients with transposition of the great arteries palliated with a Mustard or Senning procedure. The aim was to describe the CoD for patients with the Mustard and Senning procedure during short- (<10 years), mid- (10–20 years), and long-term (>20 years) follow-up after the operation.

METHODS AND RESULTS: This is a retrospective, descriptive multicenter cohort study including all Nordic patients (Denmark, Finland, Norway, and Sweden) who underwent a Mustard or Senning procedure between 1967 and 2003. Patients who died within 30 days after the index operation were excluded. Among 968 patients with Mustard/Senning palliated transposition of the great arteries, 814 patients were eligible for the study, with a mean follow-up of 33.6 years. The estimated risk of all-cause mortality reached 36.0% after 43 years of follow-up, and the risk of death was highest among male patients as compared with female patients (P=0.004). The most common CoD was sudden cardiac death (SCD), followed by heart failure/heart transplantation accounting for 29% and 27%, respectively. During short-, mid-, and long-term follow-up, there was a change in CoD with SCD accounting for 23.7%, 46.6%, and 19.0% (P=0.002) and heart failure/heart transplantation 18.6%, 22.4%, and 46.6% (P=0.0005), respectively.

CONCLUSIONS: Among patients corrected with Mustard or Senning transposition of the great arteries, the most common CoD is SCD followed by heart failure/heart transplantation. The CoD changes as the patients age, with SCD as the most common cause in adolescence and heart failure as the dominant cause in adulthood. Furthermore, the risk of all-cause mortality, SCD, and death attributable to heart failure or heart transplantation was increased in men >10 years after the Mustard/Senning operation.

Key Words: atrial switch operation ■ cause of death ■ mortality ■ Mustard procedure ■ Senning procedure ■ transposition of the great arteries

Surgical treatment of patients with transposition of the great arteries (TGA) was introduced in the late 1950s and early 1960s by Åke Senning and William Mustard.1,2 Both procedures involve redirection of the blood flow in the atria, consequently the morphological right ventricle supports the systemic part of the circulation, whereas the left ventricle supports the pulmonary circulation. Despite high perioperative mortality at the time of introduction, these procedures led to a significant improvement in survival for children with...
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TGA. However, in the early 1990s, the arterial switch operation gradually replaced the Mustard and Senning procedures as the preferred surgical technique for patients with TGA in the Nordic countries. Although early survival after the periprocedural period has been reported as good, this population is now aging. With a systemic right ventricle and tricuspid valve exposed to systemic pressure as well as the extensive atrial surgery, these patients have increased morbidity and mortality because of arrhythmia, right ventricle failure, and pulmonary hypertension as well as a risk of potential reintervention because of baffle leak or stenosis. However, short-, mid-, and long-term cause of death (CoD) in patients who have undergone Mustard or Senning corrections are not well described and were therefore the aim of this study.

**METHODS**

The authors declare that all supporting data are available within the article.

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**CLINICAL PERSPECTIVE**

**What Is New**
- Cause of death changes as patients age, with sudden cardiac death being more prominent in younger patients and heart failure/heart transplantation increasing as patients age.
- Ten years after the Mustard/Senning operation the risk of all-cause mortality, sudden cardiac death, and death attributable to heart failure/heart transplantation increases in male patients compared with female patients undergoing transposition of the great arteries.

**What Are the Clinical Implications?**
- Because death attributable to heart failure/heart transplantation seems to be increasing in aging patients with atrial correction of transposition of the great arteries, a better understanding of how to treat the failing right ventricle is needed.
- Male patients with atrial correction of transposition of the great arteries have increased mortality.

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**Nonstandard Abbreviations and Acronyms**

| Acronym | Description                  |
|---------|------------------------------|
| CoD     | cause of death               |
| HTx     | heart transplantation        |
| SCD     | sudden cardiac death         |
| TGA     | transposition of the great arteries |

**Definitions**

Date and CoD were ascertained from the medical records, national mortality databases, and death certificates. Cause-specific mortality was defined as the disease or condition directly leading to death according to World Health Organization guidance. Antecedent causes were not considered. All causes were evaluated by the same 4 coinvestigators representing each of the Nordic countries, and when the cause-specific mortality was uncertain, a consensus was reached based on medical information. Patients with insufficient data on cause-specific mortality were classified as unknown CoD.

Patients who underwent HTx were considered as an end point at the time of transplantation and combined with death attributable to heart failure (HF). Finally, patients who emigrated out of the Nordic region during the follow-up period were censored from the analysis at the time of their last contact with the Nordic health care system.

The time period for mortality was defined as short term when <10 years after the index operation; midterm follow-up was defined as 10 to 20 years after the index operation, and long-term follow-up was >20 years after the index operation.

The possible effect of surgical experience, improved surgical technique, and postoperative care was examined by dividing the observational follow-up into 2 eras according to the time of index procedure. The early era

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**Study Population**

This study is a retrospective, descriptive, multicenter study of all Nordic (Denmark, Finland, Norway, and Sweden) patients with TGA palliated with the Mustard or Senning procedure from 1967 to 2003. Patients with TGA in the Nordic countries were all operated on and followed-up at specialized congenital heart disease centers. Surgical notes as well as all medical records were reviewed case by case for the time period from 1967 until the end of 2015. The date and type of atrial procedure, reintervention, as well as heart transplantation (HTx) and last follow-up were recorded.

All citizens in the Nordic countries are assigned a unique social identification number, which makes it possible to follow their medical history over time despite change in place of residence. Nevertheless, during the early era of the atrial switch procedures, only date of birth and not the social identification number was registered in some surgical institutions in Sweden. As a consequence, some of the Mustard/Senning operated children identified from surgical notes could not be identified and were therefore excluded from the analysis. Furthermore, patients who died within 30 days after their Mustard or Senning correction (index operation) were excluded from the study (Figure 1).
was defined as surgery performed up to and including year 1982 (first half of the cohort operated on) and the late era was defined as surgery performed after 1982. Finally, TGA was considered complex in the presence of a ventricle septal defect and/or left ventricle outflow tract obstruction.

**Ethics**

This was a retrospective study based on existing patient data. The study was registered and approved by all participating institutions (Danish Datatilsynet [J.nr. 2015-41-04076/J nr 2015-231-0141], Ethical Committee of Helsinki in Finland, Regional Ethical Committee of the South-Eastern part of Norway, and Ethics Review Board [Etikprövningsnämnden] in Gothenburg, Sweden [D.nr. 174–08]). Data were anonymized before analysis and sharing across country borders.

**Statistical Analysis**

Continuous variables are presented as mean±SD and compared using the Student t test, or median (interquartile range) compared using the Wilcoxon signed rank test. Categorical variables are presented as counts and percentages and compared using χ² or Fisher exact tests.

Time 0 was set at the time of the Mustard or Senning procedure for all survival analyses. The median follow-up time was calculated using the reverse Kaplan-Meier method together with 95% CI. All-cause mortality was calculated using the Kaplan-Meier method and compared using the log-rank test. Considering other causes of death as a competing risk, the cumulative incidence of death attributable to a specific cause was estimated from Cox cause-specific hazards.

Cox regression was used to analyze the association between all-cause mortality and a subset of previously possible described variables including sex (men versus women), type of atrial correction (Mustard versus Senning), age at index procedure, era of operation (early versus late), TGA complexity (simple versus complex), and the need for a permanent pacemaker as a time-dependent variable. An interaction between sex and time ≤10 and >10 years after surgery was included because of violation of proportional rates for the variable of sex.

All statistical analyses were performed with SAS Enterprise 7.15 (SAS Institute, Cary, NC), and the null hypothesis was rejected on \( P<0.05 \).

**RESULTS**

**Identification and Follow-Up of the Nordic Mustard/Senning Cohort**

A total of 968 Nordic patients with TGA palliated with a Mustard or Senning procedure were identified, but 25 patients (2.6%) could not be identified from the surgical reports, leaving 943 identifiable patients (67.2% men). Of these, 128 patients (13.2%) died within 30 days of the index operation, leaving 814 (67.4% men) patients in the study population.

No patients were lost to follow-up, but 5 patients (3 Mustard and 2 Senning) emigrated outside the Nordic region and were censored at their last follow-up. During follow-up, a total of 191 (23.4%) patients died,
and 22 (2.7%) patients underwent HTx (Figure 1 and Table 1). None of the patients had a ventricular assist device before transplantation or as destination therapy. The mean time of follow-up was 33.6 years (95% CI, 32.9–34.1 years) after the index operation.

The Mustard and Senning procedures were performed equally in the TGA cohort (50.1% versus 49.9%), but the Mustard technique was primarily used in the early era (72.8%), and the Senning technique dominated in the late era (63.1%). As a reflection of surgical repair being undertaken at an earlier age over time, the patients undergoing the Mustard procedure were older at the time of the index procedure when compared with the Senning patients. Complex TGA was found among 29.1% of the patients, with no difference between men and women (29.6% versus 28.0%, P=0.64). During follow-up, a permanent pacemaker was implanted in 25.1% of male patients and 19.3% of female patients (P=0.07) and at a median of 15.0 years (interquartile range, 6.8–22.7 years) versus 14.7 years (interquartile range, 8.3–19.9 years) after the index operation, respectively (P=0.7). Sex and complexity of the heart lesion were equally distributed between the Mustard and Senning cohorts (Table 1).

Mortality During Follow-Up

The estimated risk of all-cause mortality in the study population reached 36.0% after 43 years of follow-up and was increased in patients palliated with the Mustard procedure as compared with the Senning procedure (P=0.012) (Figure 2).

Cause of Death

CoD was known in 78.9% of the cases. For the unknown CoD, the number was high during short-term follow-up (33.0% [32 out of 97]) but decreased during the mid- (13.8% [8 out of 58]) and long-term follow-up (8.6% [5 out of 58]). Sudden cardiac death (SCD) was numerically the most frequent known cause of mortality (n=61; 28.6%), followed by death or HTx attributable to HF (n=58; 27.2%) (Figure 3 and Table S1).

During short-term follow-up, SCD was the CoD in 23.7% of patients (23 out of 97 deaths), which increased to 46.6% (27 out of 58 deaths) during the midterm follow-up and then subsequently decreased to 19.0% (11 out of 58 deaths) during the long-term follow-up (P=0.002). Additionally, HF or HTx accounted for 18.6% of deaths (18 out of 97) during the short-term follow-up, 22.4% (13 out of 58) during the midterm follow-up, and 46.6% (27 of 58) during the long-term follow-up (P=0.0005) (Figure 3).

Implication of Type of Sex

The estimated risk of all-cause mortality was higher for men when compared with women (Figure 4), with a similar pattern for the estimated risk of SCD (Table 2, Table S1, and Figure S1). Thus, SCD occurred in 2.9% (16 out of 550 at risk) of male patients and 2.7% (7 out of 264 at risk) of female patients during the short-term follow-up (P=0.84), 5.1% (25 out of 489 at risk) of male patients and 0.9% (2 out of 227 at risk) of female patients during midterm follow-up (P=0.006), and 2.3% (10 out of 431 at risk) of male patients and 0.5% (1 out of 217 at risk) of female patients during long-term follow-up (P=0.08). The estimated risk of HTx or death attributable to HF was similar for male and female patients (P=0.1) (Table 2).

Type of Atrial Correction (Mustard Versus Senning)

During follow-up the estimated risk of SCD was similar between patients who had undergone the Mustard or

Table 1. Patient Characteristics

| Characteristic                        | Total, n=814 | Mustard, n=408 | Senning, n=406 | P value |
|--------------------------------------|-------------|---------------|---------------|---------|
| Women                                | 264 (32.4)  | 128 (31.4)    | 136 (33.5)    | 0.52    |
| Complex                              | 237 (29.1)  | 119 (29.2)    | 118 (29.1)    | 0.97    |
| Ventricular septum defect            | 202 (24.8)  | 104 (25.5)    | 98 (24.1)     | 0.66    |
| Left ventricular outflow tract       | 73 (9.0)    | 32 (7.8)      | 41 (10.1)     | 0.26    |
| Birth, y                             | 1981 (1976–1986) | 1976 (1973–1982) | 1984 (1980–1986) | <0.0001 |
| Index operation, y                   | 1982 (1977–1986) | 1977 (1975–1983) | 1984 (1981–1987) | <0.0001 |
| Age at index operation, y            | 0.9 (0.4–1.6) | 1.3 (1.0–2.4) | 0.5 (0.3–0.8) | <0.0001 |
| Postprocedure characteristic         |             |               |               |         |
| Pacemaker                            | 189 (23.2)  | 96 (23.5)     | 93 (22.9)     | 0.83    |
| ICD                                  | 25 (3.1)    | 12 (2.9)      | 13 (3.2)      | 0.83    |
| HTx                                  | 22 (2.7)    | 15 (3.7)      | 7 (1.8)       | 0.09    |
| Time from operation to HTx, y        | 22.3 (15.1–33.5) | 32.2 (16.3–34.0) | 13.4 (8.7–24.4) | 0.03    |

Data are presented as number and percentage or median (interquartile range). HTx indicates heart transplantation; and ICD, implantable cardiac defibrillator.
Senning procedure ($P=0.5$), whereas the estimated risk of death attributable to HF or HTx was increased in patients who had undergone Mustard compared with the Senning procedure (estimated 10.1% versus 5.1%, $P=0.01$) at 35 years of follow-up.

Adjusted Cox regression found no difference in the hazard rate of all-cause mortality, SCD, or death attributable to HF or HTx in patients palliated with the Mustard procedure compared with the Senning procedure (Table 3).

**TGA Complexity**

Patients with complex TGA (presence of ventricle septal defect or left ventricle outflow tract obstruction) had increased risk of all-cause mortality estimated to occur in 42.0% of patients compared with 21.8% of patients with simple TGA at 35 years of follow-up ($P<0.0001$). The risk of SCD during follow-up was 11.0% in complex TGA compared with 6.4% in patients with simple TGA ($P=0.008$). Similarly, the risk of death attributable to HF or HTx was 17.6% in patients with complex TGA compared with 4.3% in patients with simple TGA at 35 years of follow-up ($P<0.0001$).

**Risk Factors**

Adjusted Cox regression found that male sex >10 years after the index operation and complex TGA increased the hazard rate of all-cause mortality and death attributable to HF/HTx and SCD (Table 3).

**DISCUSSION**

This study of a large Mustard and Senning cohort with a long follow-up period showed that the most common CoDs are SCD and HF/HTx. There seems to be a change in CoD from midterm to long-term follow-up, with SCD initially being the most common cause, changing to HF and HTx over time. Another important finding of the study is the increased risk of all-cause mortality, SCD, and HF/HTx among men when surviving >10 years after the index operation compared with women. Risk factors associated with death attributable to all-cause mortality, HF/HTx, or SCD are male sex, >10 years after the index operation, and complex TGA.

Previous studies have reported SCD as the most common CoD in patients with TGA palliated with the Mustard or Senning procedure.\textsuperscript{5,11,15,19} These studies...
had an average follow-up time of 11 to 25 years, with findings aligned with the current study, where SCD was the most frequent cause in both the short- and the midterm follow-up groups. Based on analysis of a large younger population, it has been described that patients who survived the immediate postoperative period after the Mustard operation had a peak in mortality 8 to 15 years after surgery, with most deaths attributable to SCD. About 20% of the patients in the study had a loss of sinus rhythm, which is a common and well-described finding and may be explained by extensive atrial surgery. The loss of sinus rhythm has been described as increasing over time, but has never been associated with mortality. However, nodal rhythm shortly after the Mustard or Senning operation significantly increases the risk of developing atrial flutter, and this condition is associated with risk of SCD.

However, this alone may not explain the high risk of SCD, because the risk of loss of sinus rhythm increases over time. Nor does it explain the increased risk of death among young male patients. Two previous studies have shown that SCD happened during physical activity in about 80% of the patients. Similarly, SCD in relation to strenuous physical activity was reported in several adolescent patients in this study cohort. A possible explanation may be that in patients palliated with the Mustard or Senning procedure, atrial flutter or other supraventricular tachycardia can develop during exercise. In the case of a 1:1 or 2:1 conduction to the systemic right ventricle with abnormal systolic and/or diastolic function, this may result in low cardiac output, which may induce myocardial ischemia because of hypoperfusion of the coronary arteries, potentially leading to ventricular tachycardia and SCD. The explanation as to why death is more common in male patients remains speculative, but a possible explanation could be that they are more active and competitive when engaging in physical activity compared with female patients, especially during adolescence.

The association with HF and complex TGA has been described previously. In a study of 132 patients with TGA palliated with the Senning procedure, the systemic function of the right ventricle over time was unchanged in patients with simple TGA, but significantly decreased after 20 years in patients with complex TGA. A similar pattern has been described in a patient population with both simple and complex TGA, reporting that two-thirds of Mustard patients had right HF after 25 years but not necessarily after 14 years of follow-up. In the current study, HF or HTx was the most frequent CoD in the aging Mustard/Senning population, which could be expected with late failure of the right ventricle. In complex TGA, intraventricular surgery is performed, and in combination with presurgical cyanosis and volume overloading because of a ventricular septal defect, these factors may increase the risk of myocardial fibrosis, which is a common finding in systemic right ventricles.

Finally, the current study showed that male patients have an increased risk of all-cause mortality.
SCD, or HF/HTx when >10 years after the Mustard/Senning operation compared with female patients. Despite the fact that many of the female participants in the study most likely underwent pregnancy, this was not reflected by increased risk for HF death or HTx. Pregnancy is described as being well tolerated by these patients. However, the hemodynamic stress of pregnancy may have late adverse effects (eg, increased tricuspid regurgitation and worsening of the functional class). Therefore, in this long-term follow-up study, an increased risk of HF among women might have been expected.27–29

Limitations

Because of the nature of retrospective data collection from medical notes in several centers, this study has some limitations. As in all retrospective studies of cause-specific mortality, the validity of the CoD was dependent on the reporting physician/medical note. Furthermore, autopsy was not always performed. Despite a high percentage of known causes of death, death could not be validated in all patients. This was especially the case in the group of patients who died during childhood (aged >30 days–10 years) in the 1970s and early 1980s, where documentation in patient files compared with today were more limited. One could assume that the high number of unknown causes could partly be explained by SCD, but this remains speculative. All patients were included regardless of age at the

Table 2. Cumulative Incidence of Specific Causes of Death at 35 Years After Index Operation

| Cause of death              | Women    | Men      | P value |
|----------------------------|----------|----------|---------|
| All-cause                  | 20.3 (15.4–25.8) | 31.0 (26.7–35.3) | 0.004   |
| CD                         | 10.7 (7.4–15.4)  | 19.5 (16.2–23.5) | 0.002   |
| Heart failure/heart        | 5.9 (3.5–10.0)   | 8.9 (6.6–12.1)   | 0.1     |
| transplantation            | 4.0 (2.2–7.4)    | 9.4 (7.2–12.3)   | 0.008   |
| Sudden CD                  | 0.1 (0.1–0.2)    | 1.1 (0.6–2.5)    | 0.6     |
| Other CD                   | 2.5 (0.1–6.7)    | 2.2 (1.2–3.9)    | 0.8     |
| Operation related          | 2.0 (0.1–4.9)    | 3.6 (2.3–5.7)    | 0.2     |
| Non-CD                     | 5.2 (3.1–8.8)    | 5.4 (3.8–7.7)    | 0.8     |

The shown risk (95% CI) of specific cause of death is 35 years after the primary operation. See Figure S1. Operation-related death is further specified in Table S1. CD indicates cardiac death.

Figure 4. Risk of all-cause mortality according to sex.

Kaplan-Meier estimate of all-cause mortality for male patients compared with female patients of the full follow-up period. In the upper right is a landmark analysis of 0 to 10 and >10 years after corrective surgery.
index procedure, meaning a few outlier patients potentially had a different a priori risk of death because of higher age at the time of surgery. However, subanalysis, including 95% of patients with the lowest age at index operation, resulting in a population where all patients had been operated on before the age of 5 years, showed similar estimated risk of the specific CoDs for male patients compared with female patients as in the primary analysis (Figure S2).

**CONCLUSIONS**

Among patients with TGA corrected with a Mustard or Senning procedure, the most common CoD is SCD followed by HF or HTx. The CoDs change as the patients corrected with Mustard and Senning procedure age, with SCD as the most common cause during adolescence, whereas HF is the dominant cause in adulthood. Furthermore, this study showed that the risk of all-cause mortality, SCD, HF, or HTx was increased in men >10 years after the Mustard or Senning palliation.

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**ARTICLE INFORMATION**

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**Disclosures**

None.

**Supplemental Material**

Table S1

Figures S1–S2
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Supplemental Material
| Died due to operation                                           | Years after initial Mustard/Senning | Age at death |
|---------------------------------------------------------------|-------------------------------------|--------------|
| Information about operation not specified                     | 18                                  | 21           |
| Severe tricuspid valve regurgitation. Died due to anesthesia during tricuspid valve replacement | 10                                  | 11           |
| Information about operation not specified                     | 19                                  | 19           |
| Complications during HTx                                      | 24                                  | 29           |
| Complications from attempted transition to arterial switch    | 13                                  | 14           |
| Complications from attempted transition to arterial switch    | 34                                  | 35           |
| Information about operation not specified                     | 28                                  | 34           |
Figure S1. Estimated risks of specific causes of death.
Figure S2. Estimated risks of specific causes of death in the 95% of patients with the lowest age at index operation.