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# Arrhythmia and Mortality After the Mustard Procedure: A 30-Year Single-Center Experience

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***Objectives.*** **Our purpose was to assess the risk factors for late mortality, loss of sinus rhythm and atrial flutter after the Mustard operation.**

***Background.*** **The Mustard operation provides correction of cyanosis with low surgical risk in transposition of the great vessels. However, right ventricular failure, loss of sinus rhythm, atrial flutter and death are frequent long-term complications.**

***Methods.*** **Records of 534 children who underwent the Mustard operation at a single center since 1962 were reviewed for demographic, anatomic, electrocardiographic and physiologic predictors and outcomes.**

***Results.*** **There were 52 early deaths (9.7%). Survival analysis was undertaken for 478 early survivors with a mean follow-up interval of 11.6 ± 7.2 years. There were 77 late deaths (16.1%), with sudden death (n = 31) the most frequent cause. Survival estimates were 89% at 5 years and 76% at 20 years of age. Risk factors were an earlier date of operation, operative period arrhythmia and an associated ventricular septal defect. Risk (hazard) of late death declined in the first decade, with further peaks in the second decade. Sinus rhythm was present in 77% at 5 years and 40% at 20 years. Loss of sinus rhythm was associated with previous septectomy, postoperative bradycardia and late atrial flutter. Freedom from atrial flutter was 92% at 5 years and 73% at 20 years of age. Risk factors for atrial flutter were the occurrence of perioperative bradyarrhythmia, reoperation and loss of sinus rhythm during follow-up. Risk of atrial flutter demonstrates a late increase.**

***Conclusions.*** **Ongoing loss of sinus rhythm and late peaks in the risk of atrial flutter and death necessitate continued follow-up.**

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In 1964 Mustard et al. (1) reported a surgical method to correct the abnormal physiology associated with transposition of the great vessels. As with a similar method described by Senning et al. (2), the Mustard operation uses an intraatrial baffle to divert systemic venous return to the left ventricle and pulmonary venous return to the right ventricle. Intraatrial baffle procedures have provided excellent short-term clinical results and improved long-term survival. However, right ventricular failure, loss of sinus rhythm and development of atrial flutter are frequent long-term complications of these procedures (3–6). As a result, many centers have abandoned use of the Mustard or Senning operation in favor of the arterial switch operation described by Jatene et al. (7), which effects a more anatomic repair.

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We reviewed our case series to determine the incidence of and risk factors for late mortality, sinus node dysfunction and atrial flutter in patients after a Mustard operation.

## Methods

**Study patients.** Our study included all patients undergoing a Mustard operation at The Hospital for Sick Children between May 1963 and December 1993. Information on gender, age, body weight, underlying cardiac diagnosis, previous operation or interventional catheterization, presence of previous arrhythmias, hospital course and follow-up was obtained from hospital records.

We reviewed the results of electrocardiograms (ECGs), ambulatory ECG recordings, echocardiograms and catheterization data. All documented arrhythmias were included, except for ventricular tachycardia or fibrillation that directly preceded but was not causally related to death.

Data were obtained from The Hospital for Sick Children Cardiac Data Centre, The Toronto Congenital Cardiac Centre for Adults or from outside referring cardiologists for patients followed up elsewhere.



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**Abbreviations and Acronyms**

AV = atrioventricular
CI = confidence interval
ECG = electrocardiogram, electrocardiographic
OR = odds ratio
VSD = ventricular septal defect

**Surgical technique.** The operative technique has been previously reported (1). Operations were performed with hypothermic cardiopulmonary bypass with bicaval and aortic cannulation before 1972 and in older children and with deep hypothermia and circulatory arrest in infants <10 kg after 1972. Cold cardioplegia, either crystalloid or blood, was used beginning in 1976. Additional ventricular cooling was accomplished with either a pericardial bath or a cooling jacket (8). A pericardial patch was used to create the atrial baffle. Further modifications to the original technique were made during the series in an attempt to minimize damage to the sinoatrial node, sinoatrial conduction and the sinoatrial nodel artery (8,9).

**Definitions.** We defined the *operative period* as the duration of the hospital stay, or as 30 days from the date of operation for those discharged before 30 days. This period was used to define early survival and early or late death. *Loss of sinus rhythm* was defined as the presence of junctional or ventricular rhythm, sinus node dysfunction significant enough to require permanent pacemaker insertion, incessant atrial flutter or heart block of Mobitz II or higher grade. For survival analysis, we considered patients to have lost sinus rhythm from the date of the last ECG preceding the observation of the sinus rhythm abnormality. Patients with paroxysmal episodes of atrial flutter or other supraventricular tachycardia whose underlying rhythm was sinus based were not considered to have lost sinus rhythm. Results of all available ECGs were included in our analysis.

Although current practice would suggest the use of the term *intraatrial reentrant tachycardia*, we elected to use the term *atrial flutter* for uniformity with the diagnosis and coding used over the majority of this 30-year retrospective study and also for comparison with previous published studies of results in patients with the Mustard operation. For survival analysis of freedom from atrial flutter, patients reached the end point of this period on the date of their first documented episode of atrial flutter outside the operative period. *Heart block* was defined as the presence of high grade second-degree or third-degree heart block. *Nonsustained ventricular tachycardia* was defined as ≥3 consecutive ventricular beats but <30 s in total duration.

For our study, we used the qualitative, subjective assessments of grade of right ventricular function as normal or mildly, moderately or severely reduced by reviewing the most recent available record of two-dimensional echocardiography, cardiac radionuclide angiography or cineangiography.

Ligation or division of a patent ductus arteriosus before or at the same time as the Mustard operation was not considered an additional operation or procedure.

We considered a reoperation to be major if it involved cardiopulmonary bypass or resulted in a significant change in hemodynamic flow.

**Data analysis.** Descriptive statistics are reported as frequency, median with range or mean value with standard deviation as appropriate. Statistical Analysis Systems (10) software was used to plot time-dependent Kaplan-Meier curves for total mortality and for late death, loss of sinus rhythm and onset of atrial flutter for operative survivors only. In addition, life table analysis was used to plot estimates of instantaneous hazard of these outcomes over 2-year follow-up intervals. Independent risk factors among operative survivors for time to late death, loss of sinus rhythm and onset of atrial flutter were sought by Cox proportional hazard modeling with stepwise selection. Predictors analyzed included presence of a ventricular septal defect (VSD) or pulmonary outflow obstruction; body weight and age at operation; year of operation; preoperative balloon atrial septostomy or surgical septectomy; preoperative and operative period bradyarrhythmias, tachyarrhythmias and heart block; permanent heart block; late atrial flutter; and loss of sinus rhythm. Results are reported as odds ratios with 95% confidence intervals for significant predictors. Odds ratios >1 imply an increased risk with those <1 associated with a decreased risk of the outcome. A further analysis was performed for each outcome testing for an additional independent effect of mild, moderate and severe reduction in ventricular function relative to normal function in the subset of operative survivors for whom these data were available (341 [71%] of 478 survivors). A p value <0.05 was considered statistically significant.

## Results

**Study patients.** Descriptive characteristics of the 534 consecutive patients who underwent a Mustard operation are summarized in Table 1. Transposition of the great arteries, with either an intact ventricular septum or a hemodynamically insignificant VSD, was the most common diagnosis (two-thirds of patients). Patients operated on earlier in the series were older. Patients requiring VSD closure were older (median age 2.1 years, range 0.1 to 15.3) than those who did not (median age 1.2 years, range 0 to 18.6, $p < 0.0004$ by Kruskal-Wallis analysis of variance).

Transcatheter balloon atrial septostomy, performed preoperatively in 78% of patients, was not performed before 1967. Surgical septectomy was performed in 46% of the patients before the Mustard operation (as a primary palliative procedure in 12% and subsequent to transcatheter balloon atrial septostomy in 36%). Twenty patients (4%) had an additional procedure before the Mustard operation, 15 of whom underwent repair of aortic coarctation.

Bradyarrhythmia was noted in 12 patients (2%) and supraventricular tachycardia in 3 (0.6%) before the Mustard operation. No patient had heart block, was taking antiarrhyth-

**Table 1.** Summary of Characteristics of 534 Patients Who Underwent a Mustard Operation (including other procedures)

| | |
|---|---|
| Male | 359 (67%) |
| Median age (yr) | 1.3 |
| Range | <1 mo–18.6 yr |
| Median weight at operation (kg) | 8.8 |
| Range | 3.3–43 kg |
| Cardiac diagnosis | |
| Isolated transposition | 352 (66%) |
| Transposition+VSD | 73 (14%) |
| Transposition+PS | 50 (9%) |
| Transposition+VSD+PS | 46 (9%) |
| Other | 13 (2%) |
| Preop atrial tachyarrhythmia | 15 (3%) |
| Preop balloon atrial septostomy | 415 (78%) |
| Preop surgical atrial septectomy | 255 (48%) |
| Principal surgical procedure | |
| Mustard repair alone | 352 (66%) |
| Plus VSD closure | 113 (21%) |
| Plus pulmonary valvotomy/resection | 90 (17%) |
| Plus pulmonary arterioplasty | 43 (8%) |
| Plus LV–PA conduit | 13 (2%) |
| Plus other procedure(s) | 32 (6%) |
| Mean cardiopulmonary bypass time (min) | 79 ± 42 |
| Mean aortic cross-clamp time (min) | 40 ± 28 |
| Mean circulatory arrest time (min) | 37 ± 28 |

Data presented are mean value ± SD or number (%) of patients, unless otherwise indicated. LV–PA = left ventricular to pulmonary artery; Preop = preoperative; PS = pulmonary stenosis; VSD = ventricular septal defect.

mic medications other than digoxin or required permanent pacing before the Mustard operation.

**Operative period.** Operations were performed by four surgeons, one of whom performed 304 (57%) of the procedures. Additional major procedures performed at the time of the Mustard operation are shown in Table 1.

During their operative period, 52 patients (9.7%) died. Operative mortality was higher during the first decade of surgical experience (May 1963 to December 1972: 34 [24%] of 142) than during the second (January 1973 to December 1982: 14 [5%] of 266) and third decades (January 1983 to December 1992: 4 [3%] of 126, $p < 0.0001$).

**Early arrhythmias.** Tachyarrhythmias were noted in 61 patients (11%) during the operative period. Atrial flutter was documented in 27 patients (5%); other forms of supraventricular tachycardia, including supraventricular tachycardia not more specifically classified, were recorded for 32 patients (6%). Ventricular tachycardia occurred in two patients. Loss of sinus rhythm or sinus node dysfunction significant enough to require temporary pacing was seen in 57 patients (11%). New onset of temporary or permanent heart block was seen in 66 patients (12%). Ten patients (2%) underwent placement of permanent pacemakers during the operative period; persistent heart block was the indication in 7 of them, and sinus node disease in 3.

**Reoperation.** One or more major reoperations were required in 54 patients (10%) at a median interval after Mustard repair of 1.8 years (range 1 day to 26.2 years). Repair of an obstructed or leaking baffle was required in 25 patients (5%), semilunar valve repair or replacement in 5, atrioventricular (AV) valve repair or replacement in 4 and closure of a residual VSD or other nonbaffle systemic to pulmonic shunt in 5. Other procedures or multiple reoperations or procedures were required in 10 patients (2%). Five patients underwent heart transplantation, at 2, 14, 15, 16 and 20 years after the Mustard operation, respectively. All had associated VSDs. Rhythm before transplantation was sinus in three patients, paced (for sinus bradycardia) in one and chronic atrial flutter in one. Two of the five patients also had episodes of paroxysmal atrial flutter before transplantation.

No patient in our series underwent pulmonary artery banding and conversion to an arterial switch, as described by others (11,12).

**Follow-up period.** Follow-up data were obtained for 478 (99%) of 482 early survivors at a median interval of 11.5 years (range 0 to 30.7). Of the survivors with available follow-up data, 276 (58%) were followed up for at least 10 years, 153 (32%) for at least 15 years and 69 (14%) for at least 20 years.

**Late death and complications.** There were 77 (16%) late deaths. Unexpected sudden death occurred in 31 of these patients between 1 month and 16 years (median 3.2 years) after the operation. The cause of death was unknown in six patients.

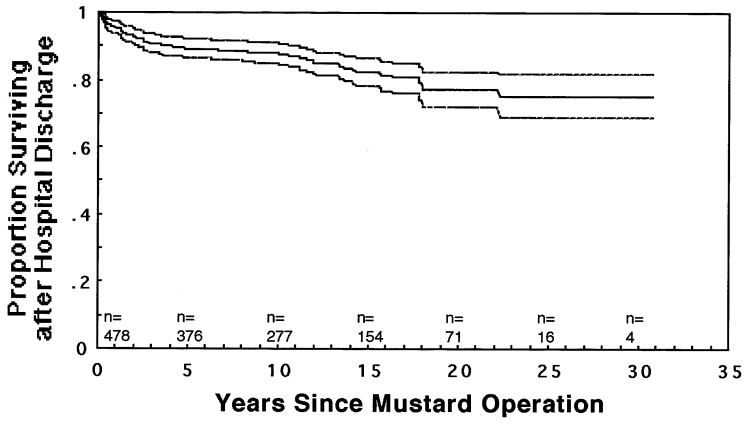


**Figure 1.** Kaplan-Meier estimates of late survival after hospital discharge after the Mustard procedure. **Top and bottom lines** indicate 95% confidence intervals.

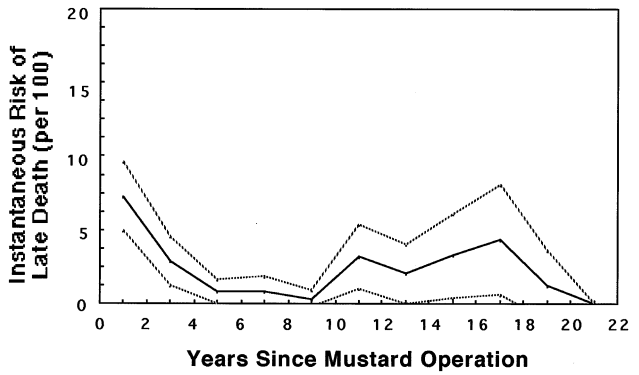


**Figure 2.** Instantaneous hazard (risk) of late death for each 2-year period after the Mustard procedure. **Top and bottom lines** indicate 95% confidence intervals.

Myocardial failure was responsible for late death in 21 patients (4%) and pulmonary artery or venous vascular disease in 10. Six patients died at the time of reoperation, and 3 died of noncardiac causes. Actuarial survival (Fig. 1) was 89% at 5 years, 88% at 10 years, 82% at 15 years, 76% at 20 years and 74% at 25 years of age. Instantaneous hazard analysis for late death (Fig. 2) is highest in the immediate postoperative period, declines to a low level from 5 to 9 years, followed by several later peaks.

Cox proportional hazard analysis indicated that the only significant independent risk factors for late death were having an associated VSD (odds ratio [OR] 2.69, 95% confidence interval [CI] 1.70 to 4.27, $p < 0.05$) and the presence of atrial tachyarrhythmia during the operative period (OR 1.99, 95% CI 1.13 to 3.50, $p < 0.02$). Later date (year) of operation (OR 0.74, 95% CI 0.59 to 0.91 [$p < 0.005$] for each consecutive 5-year increment) was protective for late death.

Independent risk factors for unexpected sudden death among operative survivors were smaller size at operation (OR 1.28, 95% CI 1.07 to 1.53 [$p < 0.008$] for each 0.1 $m^2$ decrement in body surface area); presence of atrial tachyarrhythmia during the operative period (OR 3.56, 95% CI 1.56 to 8.12, $p < 0.003$); and presence of permanent heart block (OR 5.71, 95% CI 1.32 to 24.8, $p < 0.02$).

**Sinus rhythm.** Analysis of late survival with stable sinus rhythm on serial ECGs is depicted in Figure 3. Sinus rhythm was retained in 77%, 61%, 52% and 40% of 448 operative survivors (who had measurements) at 5, 10, 15 and 20 years, respectively. Instantaneous hazard analysis (Fig. 4) demonstrates a persistent risk of loss of sinus rhythm throughout follow-up.

Independent risk factors for loss of sinus rhythm were the presence of preoperative arrhythmia (OR 7.87, 95% CI 3.84 to 16.1, $p < 0.0001$); bradycardia during the operative period (OR 1.37, 95% CI 1.01 to 1.88, $p < 0.05$); and presence of late atrial flutter (OR 3.79, 95% CI 2.73 to 5.26, $p < 0.0001$). In 62% of 54 patients who lost sinus rhythm and also experienced atrial flutter, loss of sinus rhythm occurred first.

**Atrial flutter.** Atrial flutter was documented in 67 (14%) of the 459 operative survivors for whom measurements were available. Figure 5 depicts actuarial freedom from atrial flutter in operative survivors. Survival free of atrial flutter was present in 92%, 89%, 83% and 76% of 459 operative survivors at 5, 10, 15 and 20 years, respectively. Instantaneous hazard analysis (Fig. 6) demonstrates an ongoing risk with a late peak. The bimodal pattern is similar to that previously described for late death (Fig. 2).

Independent risk factors for the occurrence of atrial flutter included the presence of bradycardia during the operative period (OR 2.67, 95% CI 1.54 to 4.60, $p < 0.0005$); permanent heart block (OR 4.17, 95% CI 1.41 to 12.3, $p < 0.01$); need for reoperation (OR 2.60, 95% CI 1.37 to 4.91, $p < 0.005$); and loss of sinus rhythm during follow-up (OR 5.69, 95% CI 3.01 to 10.80, $p < 0.0001$).

**Other tachyarrhythmias.** Supraventricular tachycardia other than atrial flutter occurred in seven patients during follow-up (AV node reentrant tachycardia in three; atrial ectopic tachycardia in one; not further defined in three). Ventricular arrhythmias included frequent premature ventricular contractions (>30/h) in 4 patients, ventricular couplets in

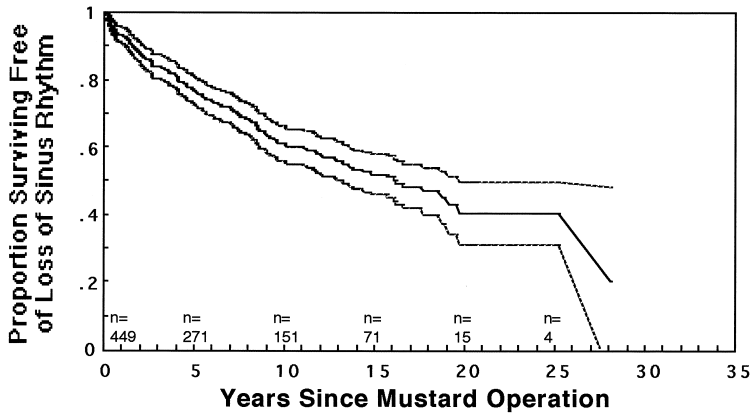


**Figure 3.** Kaplan-Meier estimates of late survival free of loss of sinus rhythm after the Mustard procedure. **Top and bottom lines** indicate 95% confidence intervals.

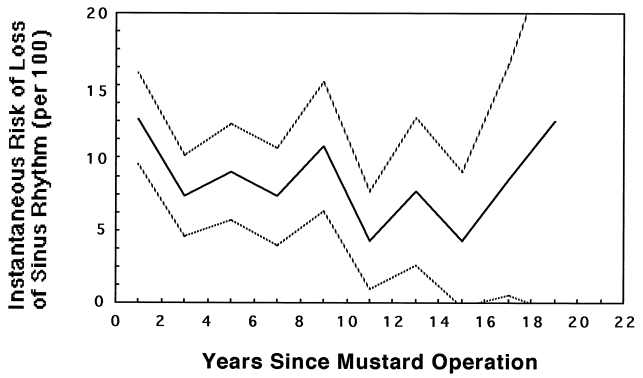


**Figure 4.** Instantaneous hazard (risk) of loss of sinus rhythm for each 2-year period after the Mustard procedure. **Top and bottom lines** indicate 95% confidence intervals.

4 and nonsustained ventricular tachycardia in 14. Sustained ventricular tachycardia (>30 s) was not seen any study patient.

**Antiarrhythmic treatment.** Nine patients were treated for ventricular arrhythmias with medication (mexiletine, sotalol or amiodarone). All but one patient diagnosed with atrial flutter was treated with digoxin. Other antiarrhythmic agents were administered to 27 patients during the postoperative period. An antitachycardia pacemaker was placed in three patients, all of whom have continued to require medication. At the time of diagnosis of atrial flutter, 22 patients were taking digoxin, whereas 37 were not. Only 1 (5%) of the 22 patients taking digoxin presented with complaints of syncope or presyncope compared with 10 (27%) of the 37 who were not taking this medication ($p = 0.04$).

**Pacemakers.** Fifty-three patients (11%) required pacemaker implantation, 10 of which were placed during the operative period before hospital discharge. Sinus node disease was the primary indication for pacing in 33 patients and heart block in 14 patients during the follow-up period. Antitachycardia pacing or the need for multiple drug treatment of atrial flutter, or both, was the indication in six patients.

**Ventricular function.** Angiographic, radionuclide or echocardiographic evaluation of right ventricular function was available in the records of 341 (71%) of 478 early survivors with follow-up. Using the most recent assessment at a median interval of 1.0 years (range 0 to 22.2) before late death or last follow-up visit, patients were designated subjectively as having normal right ventricular function or mild, moderate or severe reduction in function. Right ventricular dysfunction was severe in 23 (7%) of the 341 patients assessed.

Survival estimates, stratified for ventricular function in those patients for whom evaluations were available are shown in Figure 7. Relative to normal function, the odds ratio for time to late death from Cox proportional hazard modeling was 1.44 (95% CI 0.66 to 3.15, $p = 0.36$) for mild, 1.21 (95% CI 0.42 to 3.52, $p = 0.72$) for moderate and 8.27 (95% CI 3.22 to 21.3, $p = 0.0001$) for severe reduction in function after controlling for the effects of an associated VSD, presence of an operative tachyarrhythmia during the operative period and date of operation. Survival free of loss of sinus rhythm, stratified for ventricular function, is shown in Figure 8. Ventricular dysfunction did not significantly affect the time to loss of sinus rhythm after controlling for the significant predictors. Survival free of atrial flutter, stratified for ventricular function, is shown in Figure 9. Ventricular dysfunction was significantly related to time to onset of atrial flutter with an odds ratio relative to normal function of 2.10 (95% CI 0.98 to 4.51, $p = 0.06$) for mild, 2.95 (95% CI interval 1.23 to 7.10, $p < 0.02$) for moderate and 6.60 (95% CI 2.35 to 18.5, $p < 0.0003$) for severe reduction in function after controlling for the effects of the presence of bradycardia during the operative period, permanent heart block, need for reoperation and loss of sinus rhythm during follow-up.

## Discussion

Previous studies have suggested that both right ventricular failure (4,8,13–17) and atrial flutter (4,5) are risk factors for late death after Mustard repair of transposition of the great arteries. Our large patient cohort and considerable length of follow-up allowed for detailed analysis of independent predictors. Preoperative and operative-period information was avail-

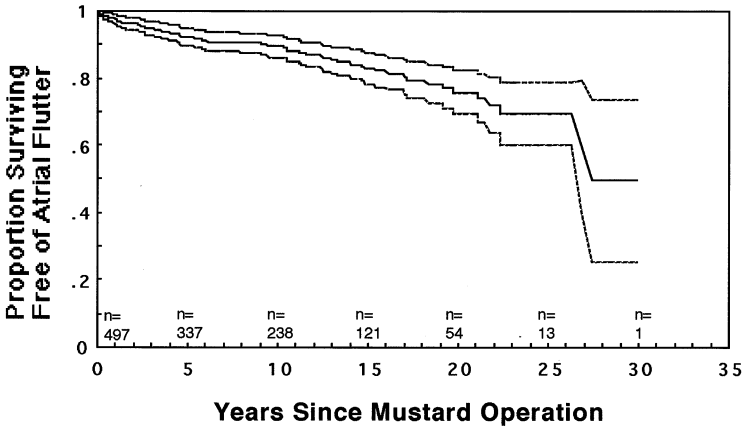


**Figure 5.** Kaplan-Meier estimates of late survival free of atrial flutter after the Mustard procedure. **Top and bottom lines** indicate 95% confidence intervals.

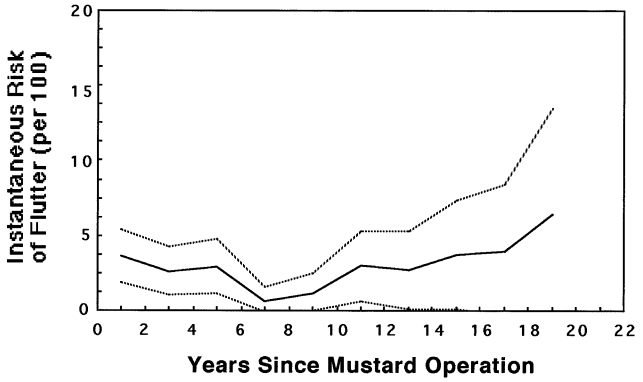


**Figure 6.** Instantaneous hazard (risk) of atrial flutter for each 2-year period after the Mustard procedure. **Top and bottom lines** indicate 95% confidence intervals.

able in all patients. Follow-up serial ECGs and other ECG information were available in a large majority of early survivors over an extended period of time.

Our study differs from that of Gewillig et al. (4), in that atrial flutter was not a significant risk factor for late death among our patients. There were two main differences in our study design: 1) We reviewed all patients up to the current era rather than restricting the study to the earlier cohort from 1965 to 1980; and 2) we did not assume patients without echocardiographic, angiographic or radionuclide assessment of the right ventricle to be normal if their physical examination and chest X-ray film did not indicate cardiac dysfunction; rather, we excluded them from that portion of the analysis. Population differences were apparent in that atrial flutter in patients in the Gewillig et al. (4) study appeared to be limited to two surgical periods, whereas in our patients' experience, it was evenly spread. However, overall frequency of late atrial flutter was similar ($p = 0.58$) between the Great Ormond Street study patients (36 [16%] of 226 survivors [4]) and our patients (67 [14%] of 478 early survivors).

In addition to atrial flutter not being a significant independent risk factor for death, another difference in our results was the demonstration of earlier date of repair as a significant risk factor. Clustering of atrial flutter in older patients who underwent operation earlier in the Gewillig et al. (4) experience may have confounded their multivariate analysis, placing undue weight on atrial flutter as a risk factor for late death. Other differences between patient groups may include different approaches to medical therapy for patients undergoing the Mustard operation in general and patients with atrial flutter specifically.

The first study to demonstrate atrial flutter as a risk factor for sudden death was that of the multicenter Pediatric Electrophysiology Group (3). That research demonstrated, by chi-square analysis, a significant association between a "dominant" tachyarrhythmia and sudden death. However, the association was only weakly significant by the Goodman-Kruskal test and did not reach significance if "nondominant" tachyarrhythmia was assessed. (We did not subclassify the occurrence of atrial flutter as dominant or not.)

**Late mortality.** Shortened life expectancy and increased risk of sudden death are well documented after atrial switch operations (9,18–20). Our findings that sudden, presumed arrhythmogenic death and myocardial failure are the most frequent cause of late death are similar to previously published results.

Earlier date of operation was an independent risk factor for late death and most likely represents improvements in surgical technique and patient selection with increased experience. Operative period atrial tachyarrythmia was found to be a risk factor for overall late death and late sudden death only, in keeping with the presumption that early arrhythmias predispose to later arrhythmias and that most sudden deaths are arrhythmogenic.

The presence of a VSD preoperatively was found to be an independent risk factor for late death. The increased risk of arrhythmia associated with ventricular surgery or presence of residual shunts are possible contributory factors. Alternatively, the later age at operation and longer period of cyanosis or volume loading in patients with VSD who underwent the operation at an older age may predispose them to late death. Pulmonary hypertension may also contribute to late death in these patients and was the direct cause of death in 10 of our patients.

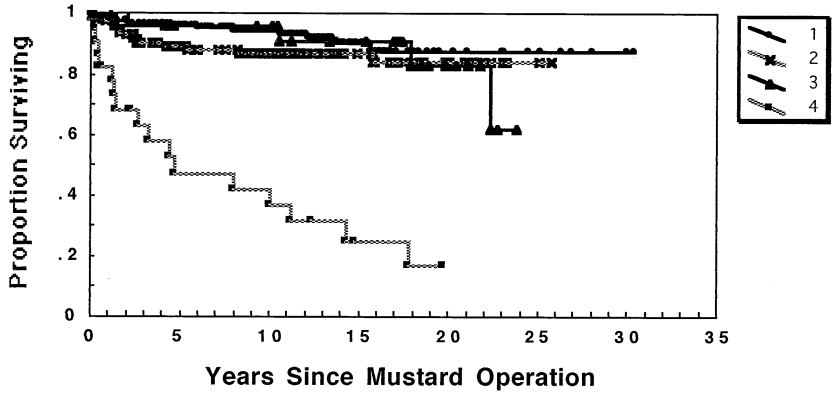


**Figure 7.** Survival after the Mustard procedure, grouped by right ventricular function: 1 = normal; 2 = mildly decreased; 3 = moderately decreased; 4 = severely decreased.

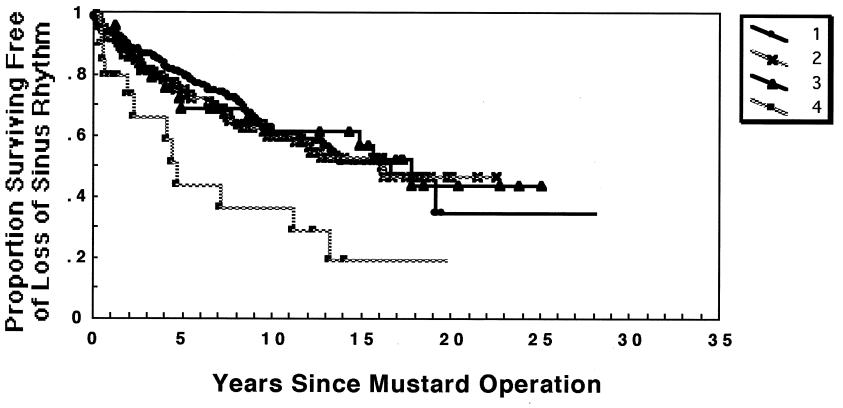


**Figure 8.** Survival with stable sinus rhythm after the Mustard procedure, by right ventricular function: 1 = normal; 2 = mildly decreased; 3 = moderately decreased; 4 = severely decreased.

**Loss of sinus rhythm and atrial flutter.** Late development of both atrial bradyarrhythmias and tachyarrhythmias are well recognized late complications of atrial baffle surgery (5,6,21). The frequency of rhythm disturbances in published reports shows a wide variation, reflecting methodologic differences. The ECGs were universally obtained at frequent intervals (usually yearly) in our cohort, whereas 24-h ambulatory monitoring was used sporadically. We chose to use only standard ECGs for our evaluation of sinus rhythm to permit more accurate comparisons between patients followed up by different physicians during different time periods.

The extensive atrial surgery involved in redirecting systemic and pulmonary venous circulations is a well documented risk factor for late atrial tachyarrhythmias and sinus node disease (5,6,21). Postulated mechanisms include disruption of sinus node arterial flow and direct damage to sinus node and atrial conduction tissue (5,6,22).

The progressive loss of sinus node function and development of atrial flutter years after operation indicate an ongoing process. The interdependence of early sinus node disease and tachyarrhythmias and the development of late sinus node disease and atrial flutter suggest that predisposing factors are similar. Similarities in the shape of the hazard curves provide further support.

**Treatment of atrial flutter.** Our findings suggest that those patients who are being treated with digoxin at time of initial presentation in atrial flutter are less likely to have symptoms of presyncope or syncope. We advocate the prophylactic use of digoxin in patients with the Mustard operation unless otherwise contraindicated.

A variety of additional medications were used to treat atrial flutter in our cohort. Because of changing practice and drug availability over the long period of this study, we were unable to assess the relative efficacies of the different treatment methods. Antitachycardia pacing was used in a small number of patients, most of whom continued to require antiarrhythmic medications.

**Right ventricular function.** Late reduction in systemic right ventricular function is a well recognized outcome after a Mustard operation; failure to improve function with exercise has also been demonstrated (15,23–25). We did not have accurate functional assessment in sufficient patients to evaluate this in our multivariable analysis. However, in those patients for whom information was available, reduced function was associated with increased mortality and atrial flutter.

A likely mechanism is that the increased end-diastolic ventricular pressure and progressive tricuspid regurgitation associated with poor ventricular function predispose the pa-

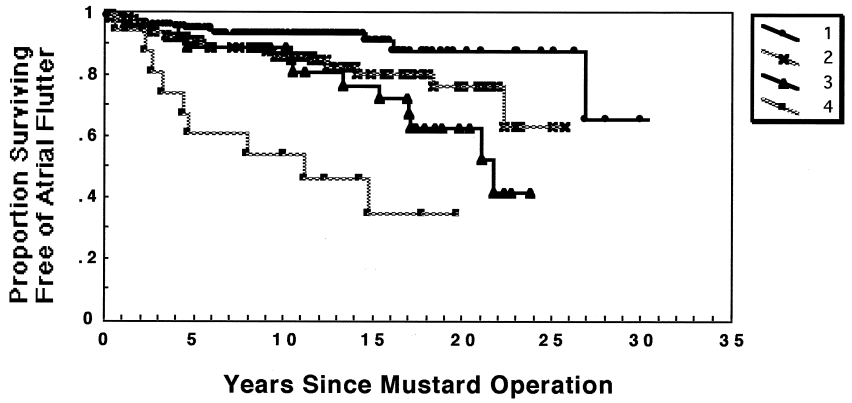


**Figure 9.** Survival free of atrial flutter after the Mustard procedure, by right ventricular function: 1 = normal; 2 = mildly decreased; 3 = moderately decreased; 4 = severely decreased.

tient to overloads in atrial volume and pressure, further contributing to the development of arrhythmias. Only prospective studies can determine whether right ventricular function can be medically improved and whether such improvement will alter outcome.

**Atrial versus arterial switch operation.** The late development of arrhythmias and cardiac failure in patients after atrial baffle procedures has resulted in a shift toward more use of the primary arterial switch procedure for the surgical treatment of transposition. Despite an initially higher surgical mortality, excellent intermediate-term results have been reported (26,27). It remains to be seen whether this continues in long-term follow-up.

**Limitations of the study.** The study was retrospective in nature and included a 30-year experience during which changes were made in diagnostic and treatment modalities. Decisions on ECG assessment and other cardiac testing were based solely on clinical grounds and were not standardized.

Analysis of right ventricular function was the most difficult variable to obtain and quantitate. We lacked an objective assessment of right ventricular function in 27% of survivors with follow-up—in many cases, because of the difficulty in obtaining noninvasive measurements before the advent of echocardiography. The lack of formal echocardiographic and angiographic criteria for assessing systemic right ventricular function, confounding variables, such as tricuspid insufficiency and VSD patches, and the subjective descriptions of different observers created further obstacles. Because of these difficulties, we did not include cardiac function as a variable in the hazard analyses.

**Conclusions.** Our findings demonstrate that ongoing and late peaks in the risk of death, loss of sinus rhythm and atrial flutter necessitate continued follow-up in patients who have undergone the Mustard operation. Patients with preoperative and perioperative rhythm disturbance are at a higher risk and may require closer observation.

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