Clinical factors affecting survival in patients with D-transposition of the great arteries after atrial switch repair: A meta-analysis

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ABSTRACT

Background: Atrial switch repair (AtrSR) was the initial operation method in patients with D-transposition of the great arteries (D-TGA) constituting the right ventricle as a systemic one. Currently, it has been replaced with arterial switch operation (ASO), but the cohort of adults after AtrSR is still large and requires strict cardiological management of late complications. For this reason, we aimed to evaluate potential long-term mortality risk factors in patients with D-TGA after AtrSR (either Mustard or Senning procedures)

Methods: We searched the MEDLINE database for suitable trials. We included 22 retrospective and prospective cohort studies of patients with D-TGA with at least 5 years mean/median follow-up time after Mustard or Senning procedures, with an endpoint of non-sudden cardiac death (n-SCD) and sudden cardiac death (SCD) after at least 30 days following surgery.

Results: A total of 2912 patients were enrolled, of whom 351 met the combined endpoint of n-SCD/SCD. The long-term mortality risk factors were New York Heart Association (NYHA) class ≥III/heart failure hospitalization (odds ratio [OR], 7.25; 95% confidence interval [CI], 2.67–19.7), tricuspid valve regurgitation (OR, 4.64; 95% CI, 1.95–11.05), Mustard procedure (OR, 2.15; 95% CI, 1.37–3.35), complex D-TGA (OR, 2.41; 95% CI, 1.31–4.43), and right ventricular dysfunction (OR, 1.94; 95% CI, 0.99–3.79). Supraventricular arrhythmia (SVT; OR, 2.07; 95% CI, 0.88–4.85) and pacemaker implantation (OR, 2.37; 95% CI, 0.48–11.69) did not affect long-term survival in this group of patients. In an additional analysis, SVT showed a statistically significant impact on SCD (OR, 2.74; 95% CI, 1.36–5.53) but not on n-SCD (OR, 1.5; 95% CI, 0.37–6.0).

Conclusions: This meta-analysis demonstrated that at least moderate tricuspid valve regurgitation, NYHA class ≥III/heart failure hospitalization, right ventricular dysfunction, complex D-TGA, and Mustard procedure are risk factors for long-term mortality in patients after AtrSR.

Key words: atrial switch repair, D-transposition of the great arteries, mustard procedure, senning procedure

INTRODUCTION

D-transposition of the great arteries (D-TGA) is a congenital heart defect characterized by atrioventricular concordance and a lack of arterio-ventricular concordance: the aorta originates from the right ventricle, while the pulmonary trunk originates from the left ventricle [1]. Currently, the surgical treatment of choice for D-TGA is arterial switch operation (ASO). The most common ASO complication, occurring in 8% of patients, is coronary artery obstruction; nevertheless, these patients are also at risk of neo-aortic root dilatation, supravalvular pulmonary stenosis, and left ventricular dysfunction, and ventricular arrhythmias [1, 2]. However, patients after atrial switch repair (AtrSR) using the Senning or Mustard methods still constitute a large group of adult

WHAT'S NEW?

Patients with D-transposition of great arteries (D-TGA) after atrial switch repair (AtrSR) are exposed to long-term sequalae requiring medical care. D-TGA is one of the conditions most prone to sudden cardiac death in congenital heart disease. However, the criteria for implantable cardioverter defibrillator implantation in primary prevention of sudden cardiac death are not clear. We evaluated potential long-term mortality risk factors in patients after AtrSR. We searched the MEDLINE database and analyzed 22 retrospective and prospective cohort studies in this meta-analysis. It showed that at least moderate tricuspid valve regurgitation, New York Heart Association (NYHA) class ≥III/heart failure hospitalization, right ventricular dysfunction, complex D-TGA, and Mustard procedure are risk factors for long-term mortality in patients after AtrSR. To our knowledge, this meta-analysis is the largest in the current literature with the highest number of risk factors for long-term mortality in the operated D-TGA population.

patients, as the 40-year survival rate described in cohorts reaches 60%–75% [3–6]. During AtrSR, the systemic and pulmonary return is redirected with an intra-atrial baffle made of a Goretex patch (Mustard) or native atrial tissue (Senning), thus the right ventricle (RV) becomes systemic. As a consequence of these anatomical alterations, patients are at risk of developing common complications like RV dysfunction or failure, progressive tricuspid valve regurgitation, bradycardia, and chronotropic incompetence, supraventricular and ventricular tachyarrhythmias [1]. According to researchers, complications of AtrSR described as risk factors for non-sudden cardiac death (n-SCD) and/or sudden cardiac death are right ventricular dysfunction (RVD), supraventricular tachyarrhythmias (SVT), tricuspid valve regurgitation (TVR), New York Heart Association (NYHA) class ≥II, and atrioventricular block [3, 7–11]. To our knowledge, this meta-analysis is the largest in the current literature with the highest number of risk factors for long-term mortality in the operated D-TGA population.

Aims

This study aimed to evaluate long-term mortality risk factors in patients with D-TGA after atrial switch operation with either the Mustard or Senning procedure.

Eligibility criteria

We included 22 retrospective and prospective observational cohort studies of patients with D-TGA with at least 5-year mean or median follow-up time after AtrSR either with the Mustard or Senning procedure, with an endpoint of SCD, SCD equivalent events (aborted cardiac arrest or appropriate ICD discharge), or n-SCD after at least 30 days following surgery. The included studies needed to describe mortality and differences between living and deceased patients in the incidence of SVT, RVD, NYHA class, the number of implanted pacemakers, number of patients with simple and complex D-TGA, or number of patients operated with Mustard or Senning procedures.

We included patients with complex D-TGA, i.e. the coexistence of an additional heart defect (ventricular septal defect, pulmonary stenosis, left ventricular outflow tract obstruction, aortic coarctation). Selected studies also reported at least one of the following conditions:

1. SVT including atrial fibrillation and regular atrial tachycardia; RVD as right ventricular ejection fraction <45% assessed by echocardiography were determined by one of the following methods:
   - Simpson apical four-chamber view;
   - Subjective assessment by an experienced cardiologist according to contraction pattern, wall thickness, ventricular dimensions, and septal movement in the presence of significant tricuspid regurgitation and a flow velocity in the ascending aorta of less than 0.7 m/sec.;
   - Partial subjective assessment, with right ventricular size and function assessed utilizing tissue Doppler imaging and tricuspid annular plane systolic excursion (TAPSE);
2. Functional NYHA class ≥III or hospitalization for heart failure (HF);
3. Atrioventricular block, sick sinus syndrome, or arrhythmia requiring pacemaker implantation; at least moderate TVR, assessed in Doppler echocardiography by an experienced cardiologist or catheterization with angiography;
4. Comparison of the number of patients operated with Mustard and Senning procedure.

Studies with no estimates of the association between risk factors and survival were excluded. We excluded studies that focused on specific subgroups of patients with D-TGA e.g., pregnant women, studies including D-TGA with other complex heart defects e.g., tetralogy of Fallot, and patients after double-switch surgery. Other exclusion criteria were studied groups <5 patients, language of the manuscript other than English, and other studies from the same research site describing the same cohort again.

Study process and search strategy

We investigated the MEDLINE database from its inception date to April 30, 2021 to find cohort studies describing long-term mortality and/or SCD risk factors in patients with D-TGA after atrial switch repair. While searching for suitable abstracts, we did not establish any language restrictions or filters. The search terms we used were (transposition of the great arteries OR TGA OR dTGA OR cTGA OR cctg OR systematic right ventricle OR atrial switch OR arterial switch

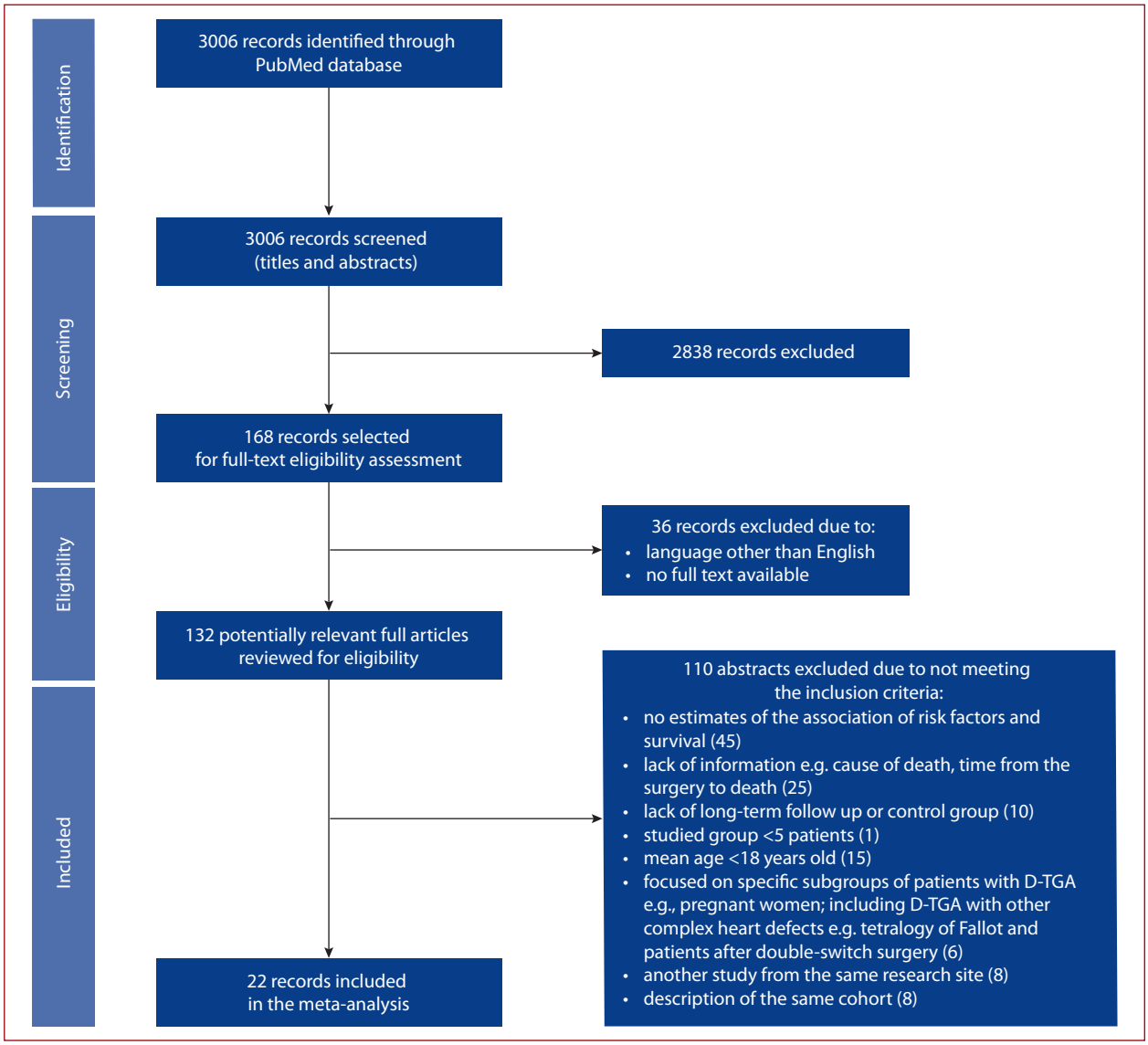

Figure 1. The study flowchart

Abbreviations: D-TGA, D-transposition of the great arteries

OR mustard procedure OR senning procedure) AND (death OR sudden death OR cardiac death OR sudden cardiac death OR outcome OR prognosis OR risk factors) AND (adults OR adolescents). One investigator (SN) analyzed the entire database searching for suitable articles and simultaneously excluded duplicates, from which 168 articles were selected for full-text eligibility assessment (Figure 1). After 36 records were excluded due to language other than English and no full-text availability, two investigators (SN and EJ) reviewed 132 potentially relevant full articles. Finally, we excluded 110 manuscripts for the reasons listed in Figure 1. Eventually, 22 records met the inclusion criteria. Additionally, we verified the references of the included articles in search of potential new studies. The studies included in the analysis were approved by an appropriate institutional review board or ethics committee and patients provided written informed consent to participate in the study. All extracted relevant information was double-checked by the investigators. Other team members (AC, PR, SG, ML, OT) were responsible for substantive oversight and resolving uncertain decisions.

Data extraction and quality assessment

The quality of the included studies was assessed by two reviewers (EJ and SN) with the use of the Quality Assessment Tool for Observational Cohort and Cross-Sectional Studies (Supplementary material, *Table S1*) [12]. We checked if the information about study patient loss, their number, and reasons for exclusion (including the safety outcome data) was provided. We extracted data describing study characteristics such as country, study design, length of follow-up, number of patients in each research group, and number of study sites. The patient population characteristics included age, percentage of females and males, mean or median follow-up time, and percentage of patients with complex D-TGA, sinus node dysfunction, baffle obstruction, and in need of reintervention or pacemaker implantation. We also

considered the numbers of n-SCD, SCD, aborted cardiac arrests, and appropriate ICDs.

Statistical analysis

The included research data were meta-analyzed using Review Manager 5.4. The random-effects model and inverse variance method were used to estimate the odds ratio (OR) of the combined endpoint of n-SCD, SCD, or SCD equivalent events. A total of 22 retrospective or prospective observational cohort studies were included in this meta-analysis [7–9, 11, 13–30]. There were seven main comparisons, which included: (1) NYHA functional class ≤II vs. NYHA ≥III/HF hospitalization [9, 11, 26]; (2) TVR <moderate vs. ≥moderate [8, 9, 11, 13, 16, 20, 21, 28, 31]; (3) Mustard vs. Senning procedure [7–9, 11, 14, 19, 20, 22, 28]; (4) complex vs. simple D-TGA [8, 9, 13, 16, 20, 21, 24, 27–29, 31]; (5) RVD vs. no RVD [8, 9, 11, 13, 15, 16, 18–23, 28]; (6) history of SVT vs. no history of SVT [7, 9, 13, 14, 16, 19, 21, 23, 27, 28, 30, 31]; (7) pacemaker vs. no pacemaker implantation [8, 9, 11, 16, 21, 23, 30]. The secondary endpoint was defined as SCD or SCD equivalent events and the random-effects model and inverse variance method were used to estimate the OR. The *P*-value <0.05 was considered as statistically significant.

The χ^2 test was used to assess the significance of heterogeneity between the results of different research and presented as the I^2 test. Significant heterogeneity was defined as I^2 >50%, and I^2 <25% was defined as non-significant heterogeneity [32].

RESULTS

Study characteristics

The included studies examined a total of 3067 patients, 73 of whom were lost to follow-up and 82 died within 30 days of the atrial switch repair. A total of 2912 patients were finally enrolled, of whom 351 met the combined endpoint of n-SCD, SCD, or SCD equivalent events. Twenty of included studies were single-center types from sites in 12 countries [7, 9, 11, 13–19, 21–24, 26–31]. The remaining 2 studies were multi-center international studies with 2 to 3 involved countries and 7 clinical sites each [8, 20]. In the described research groups, the average age ranged from 13.9 to 35 years, and men accounted for 51.3%–89.2%, with an average follow-up time from AtrSR of 9.9–30 years. The mean age at the time of AtrSR ranged from 6.9 to 46 months; 1321 patients were operated on by the Mustard method and 1340 by the Senning method; 251 cases were described as AtrSR without exact numbers on the procedure method. The proportion of patients with the D-TGA complex ranged from 0% to 49.4%, with one study also including one patient representing 1% of the study population with the Taussig-Bing anomaly [26]. Of 351 deaths, 142 (40.4%) were n-SCD, 194 (55.3%) were SCD, and 15 (4.3%) were SCD-equivalent events. The characteristics of the included studies are summarized in Supplementary material, *Table S2*.

Meta-analysis

A meta-analysis of the reported risk factors was performed (Figure 2A–B). Data on RVD was available from the highest number of publications (13 articles with a total number of 1489 patients); data on procedure type in 8 articles with 1348 patients; data on SVT in 12 articles with a total number of patients equaled 1339; complex vs. simple D-TGA was evaluated in 11 articles with 1257 patients; at least moderate TVR in 9 articles with 870 patients; implantation of pacemaker in 5 articles with 526 patients; NYHA class ≥III/HF hospitalization in 3 articles with 251 patients (Table 1).

A statistically significant relationship was found between at least moderate NYHA class ≥III HF hospitalization (*P* <0.001), TVR (*P* <0.001), type of surgical procedure (*P* <0.001), complex D-TGA (*P* = 0.005), and the combined endpoint of n-SCD, SCD, and SCD-equivalent events. We observed an association of borderline statistical significance between RVD (*P* = 0.05) and the primary endpoint. We did not observe statistically a significant (*P* = 0.09) relationship between SVT or pacemaker implantation (*P* = 0.29) and the primary endpoint (Figure 2A–B). We performed an additional analysis in which we assessed SVT separately for n-SCD and SCD/SCD equivalent events (Figure 3) [33]. The meta-analysis showed that SVT is associated with SCD (odds ratio [OR], 2.74; 95% confidence interval [CI], 1.36–5.53; *P* = 0.005). To confirm that SVT is a risk factor for SCD, we performed a separate meta-analysis for SVT as an n-SCD risk factor (Figure 3). We did not find a significant (OR, 1.5; 95% CI, 0.37–6.0; *P* = 0.57) relationship between SVT and n-SCD, which confirms that SVT is a risk factor only for SCD.

The overall heterogenity of particular analysis was considered significant for RVD (I^2 = 60%), SVT (I^2 = 53%) and pacemaker implantation (I^2 = 68%). The analysis of NYHA class ≥III/HF hospitalization (I^2 = 0%), and type of procedure (I^2 = 18%) had non-significant heterogeneity. The analysis for at least moderate TVR (I^2 = 49%) and complex D-TGA (I^2 = 48%) showed intermediate heterogeneity.

DISCUSSION

This meta-analysis demonstrated that at least moderate tricuspid valve regurgitation, NYHA class ≥III/heart failure hospitalization, right ventricular dysfunction, complex D-TGA, and the Mustard procedure are risk factors for long-term mortality in patients after AtrSR.

NYHA class

NYHA functional class appeared to be a significant risk factor for the combined endpoint of long-term mortality. Since assessment of right ventricular dysfunction by echocardiography is not standardized, we decided to evaluate the clinical status of patients with the use of

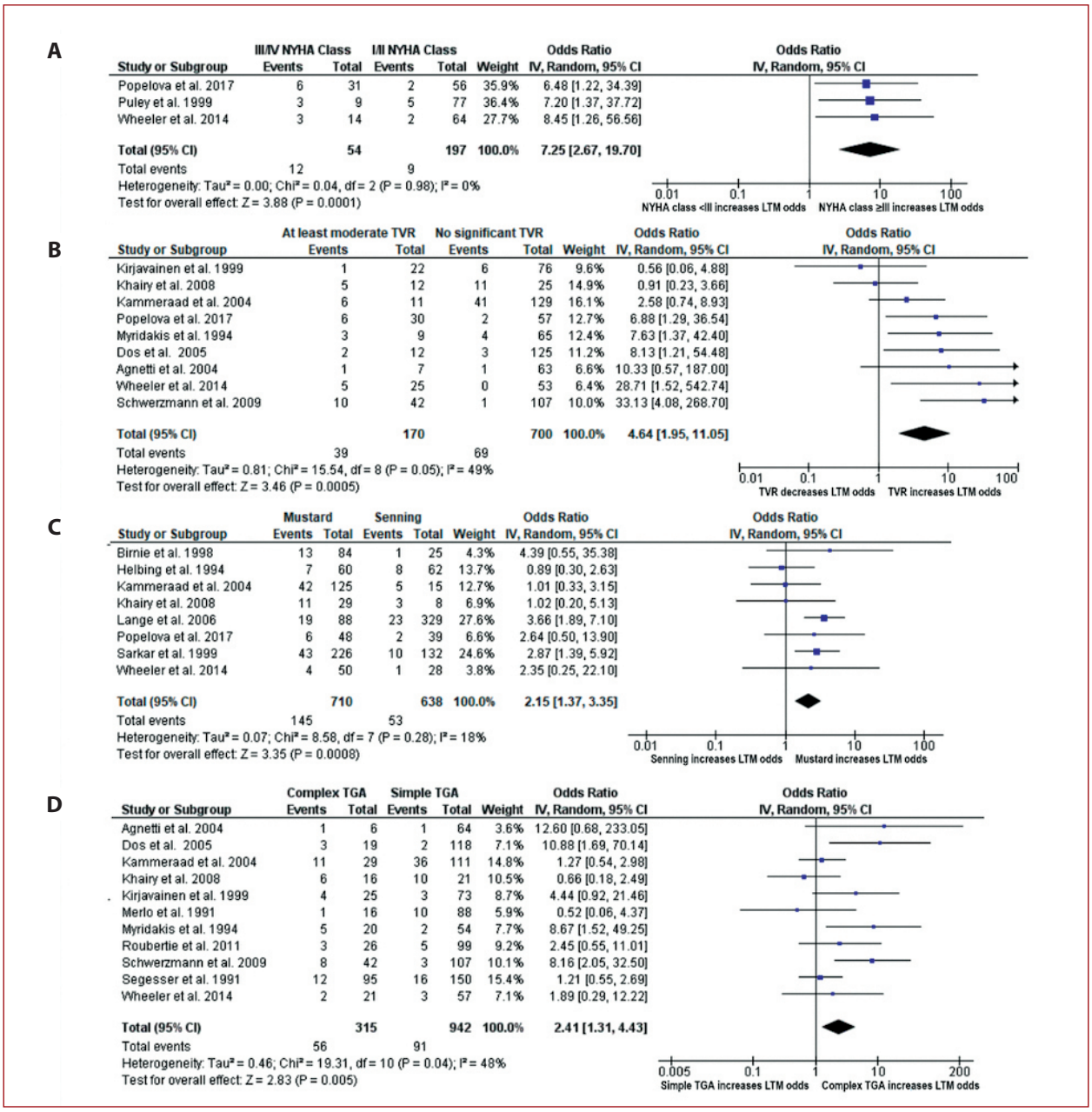

Figure 2A. Forest plots showing pooled odds ratios of NYHA >II/heart hospitalization (**A**), at least moderate TVR (**B**), Mustard procedure (**C**), Complex D-TGA (**D**). D-TGA indicates D-transposition of the great arteries

Abbreviations: D-TGA, D-transposition of great arteries; NYHA, New York Heart Association; LTM, long-term mortality;TVR, tricuspid valve regurgitation

NYHA class/HF hospitalization as a risk factor for death in patients after AtrSR. Although recent guidelines do not recommend to use the NYHA functional status in adults with congenital heart defects, this parameter has been used in numerous older publications on the survival in this group of patients [1]. Most of the patients with NYHA class I and II function are well adapted and report no clinical symptoms like dyspnea despite the objectively reduced exercise capacity in cardiopulmonary tests [11, 34]. Of the 251 patients included in the studies comparing NYHA class/HF hospitalization in living and deceased patients, as many as 54 (21.5%) patients were hospitalized for HF or were in the functional NYHA class ≥III. Previous articles, which did not show the relationship between the NYHA class and mortality, did not involve in the analysis a more objective tool, which is hospitalization for heart failure [11, 34]. In our study, clinical deterioration related to systemic ventricular dysfunction turned out to be a significant risk factor for death, which confirms that NYHA class ≥III/HF hospitalization reflects the disturbance of temporal RV compensation. Popelova et al. [11] showed a correlation between N-terminal pro-B-type natriuretic peptide (NT-proBNP) concentration and mortality, while not showing a statistically significant relationship be-

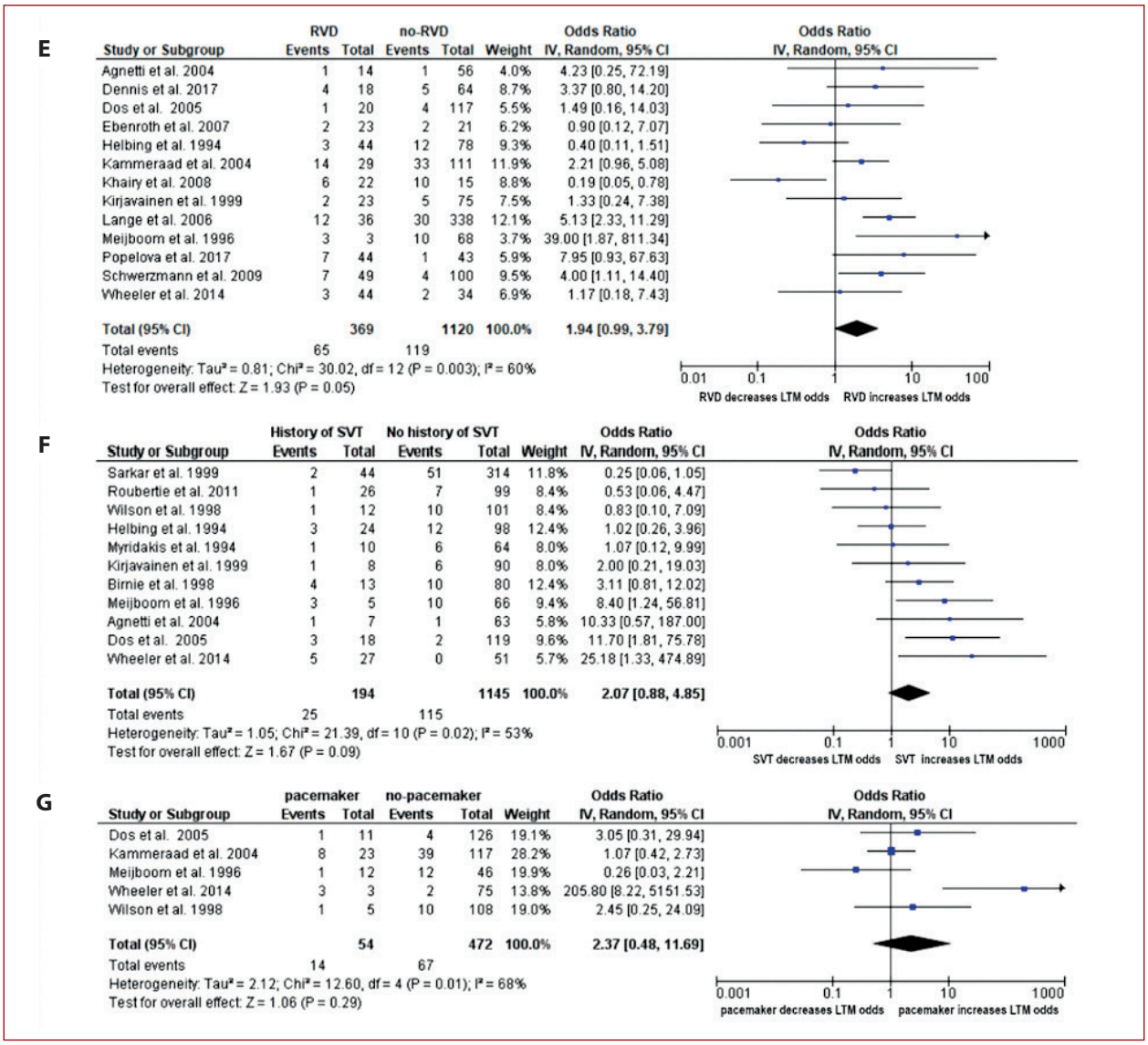

Figure 2B. Forest plots showing pooled odds ratios of RVD (**E**), history of SVT (**F**), and pacemaker implantation (**G**) for long-term mortality using a random effects meta-analysis approach

Abbreviations: RVD, indicates right ventricular dysfunction; SVT, supraventricular tachyarrhythmia

Table 1. Summary of pooled odds ratios results in the random-effect model

Comparison	Odds ratio	95% CI	*P*-value
NYHA functional class ≥III/HF hospitalization vs. NYHA Class ≤II	7.25	2.67–19.7	<0.001
At least moderate TVR vs. no TVR	4.64	1.95–11.05	<0.001
Mustard vs. Senning procedure	2.15	1.37–3.35	<0.001
Complex vs. simple D-TGA	2.41	1.31–4.43	0.005
RVD vs. no RVD	1.94	0.99–3.79	0.05
History of SVT vs. no history of SVT	2.07	0.88–4.85	0.09
Pacemaker vs. no pacemaker	2.37	0.48–11.69	0.29

Abbreviations: CI, confidence interval; HF, heart failure; other — see Figure 2

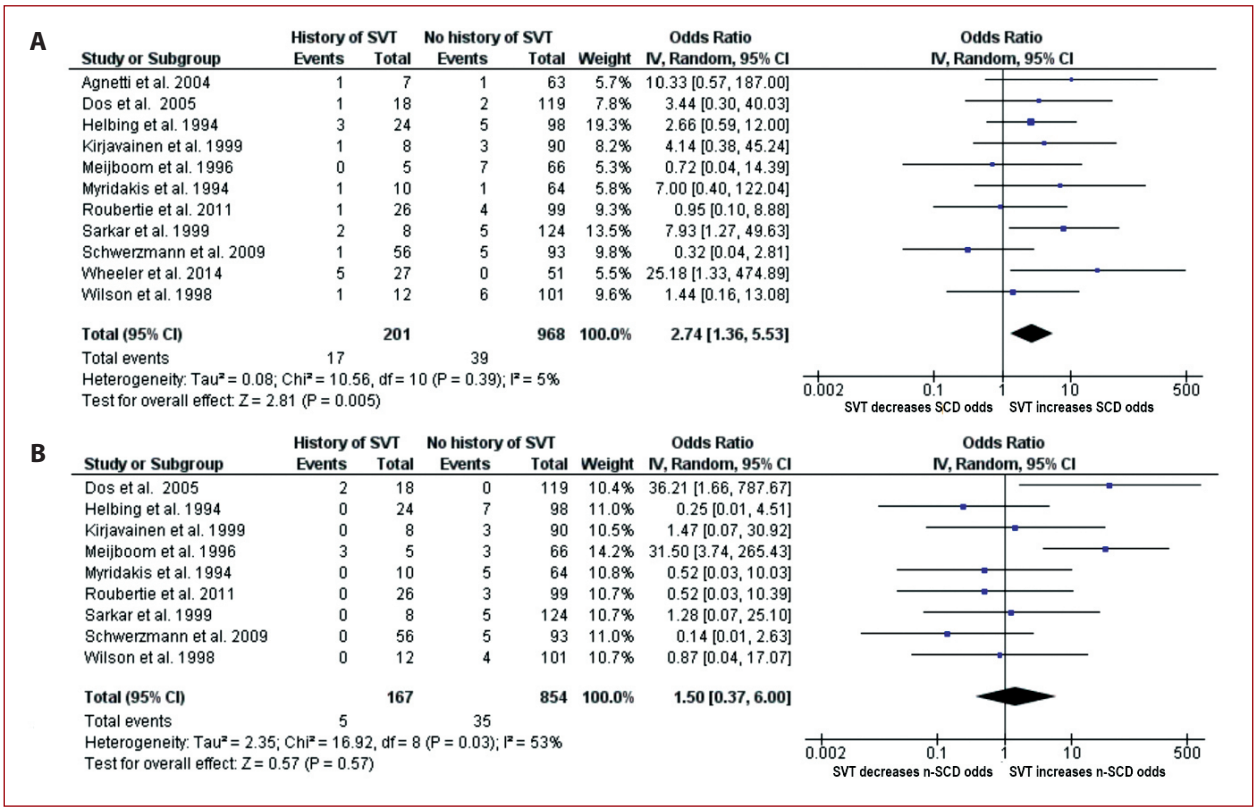

Figure 3. Forest plots showing pooled odds ratios of SVT for SCD (**A**) and n-SCD (**B**) using a random effects meta-analysis approach. n-SCD indicates non-sudden cardiac death

Abbreviations: SCD, sudden cardiac death; other — see Figure 2

tween the NYHA class and mortality. This may suggest that a combination of NT-proBNP and hospitalization for heart failure might be a more sensitive tool in assessing the clinical status and risk of death in patients after AtrSR.

Tricuspid valve regurgitation

Tricuspid valve regurgitation turned out to be the strongest risk factor for death in our review. The systematic tricuspid valve becomes progressively incompetent in patients with operated D-TGA. In most cases, TVR results from ring dilatation caused by enlargement of the failing systemic RV, but occasionally it can also be caused by surgical injury [35]. TVR is often commensurate with RVD, but it is not a constant or linear relationship as severe TVR does not develop in all patients with dysfunctional RV. The results on the use of TVR severity as an indirect parameter in assessing systematic ventricular impairment are contradictory [36, 37]. However, TV replacement/repair surgery showed stabilization of RV function and improvement of NYHA functional class [38]. In addition, TVR severity has been shown to correlate with exercise capacity of these patients assessed in cardiopulmonary tests [39]. The severity of TVR may not necessarily correspond to RVD, but it has a significant impact on the clinical status of patients with corrected D-TGA and is therefore a significant risk factor for death in this group.

Right ventricular dysfunction

To date, cohort studies have shown a statistically significant association between RVD and long-term mortality [15, 18, 21, 40]. Our meta-analysis has shown that there is a statistical tendency between RVD and long-term mortality in patients with operated D-TGA. However, a previous review article by Venkatesh et al. [33] did not confirm such results. The difference between the outcomes may be due to the discrepancy between exercise capacity, symptomatic HF, and RVD subjectively assessed by echocardiography [16, 19, 24, 41]. Additionally, systemic failure of the right ventricle due to its triangular structure and different orientation of myocardial fibers seems to have a significant diastolic component [36]. Due to this fact and the RV volume and its mechanism of adaptation to systemic load, echocardiographic assessment of RV systolic function does not always have to deteriorate. It seems that RVD evaluated by cardiac magnetic resonance will be a more reliable measure. However, the current studies, surprisingly, do not show an association between CMR parameters and patients' clinical status or exercise capacity with corrected D-TGA [42, 43].

Surgery procedure type

Mustard surgery proved to be a significant risk factor for death compared to the Senning procedure, which is in line

with the previous publications [7, 22, 33]. The use of artificial material to create baffles requires more sutures lines within the atria, poses a higher risk of baffle obstruction and thus the risk of reoperation. The articles we included were published between 1991 and 2017 reflect different surgical experiences, quality of extracorporeal circulation technology, hypothermia, use of cardioplegia, and perioperative care. All those factors could impact long-term results in this group. Right after introduction of the Mustard procedure, patients were operated on without the use of cardioplegia and cardioprotection, which also could have impacted outcomes. In the current era, the treatment of choice is arterial switch operation, but too few available data prevented us from comparing long-term mortality between AtrSR and ASO subgroups.

Complexity

The complexity of the defect turned out to be an important risk factor for the combined endpoint of long-term mortality. This can be related to the greater extent of anatomical abnormalities subsequently requiring a more extensive surgical repair, which prominently translates into worse clinical conditions. Moreover, chronic hypoxia associated with delayed AtrSR can cause myocardial ischemia and scarring. A greater extent of the surgical procedure may contribute to the excessive scar formation predisposing to iatrogenic right bundle branch block. It may also predispose to the subsequent SVT and RVD [3, 22]. The lack of a unified definition of complex TGA among the included studies may bias the outcomes of our meta-analysis. Still, previous research also described the complexity of TGA as a risk factor for death [31, 33].

Supraventricular tachyarrhythmia

We have not shown that SVT is a risk factor for the combined endpoint consisting of n-SCD and SCD with equivalent events. Since previous investigators showed such an association [33], we performed a separate analysis of SVT as a risk factor for SCD that showed a statistically significant relationship between them [33]. Additionally, the analysis of SVT as an n-SCD risk factor did not confirm such a relationship. The most common SVT occurring in patients after D-TGA correction is a cavo-tricuspid isthmus-dependent flutter, followed by a macro-reentry circuit related to surgical scars. The incidence of SVT increases with aging and affects up to a third of these patients [36, 44], with atrial fibrillation typically occurring at older age [1]. Due to the presence of stiff baffles impairing the capability to increase the preload, the high heart rate is poorly tolerated hemodynamically and can be fatal [1]. In addition, SVT may contribute to SCD with 1:1 conduction through the healthy atrial node but also because of potential ischemia of the systemic ventricle supplied by only one coronary vessel which is the right coronary artery [13, 16]. Cohort studies showed a significant statistical relationship between SVT and SCD and a poor relationship between tachyarrhythmias and overall mortality [7, 16]. Our meta-analysis confirmed these findings. However, even if SVT was found to be related only to SCD and not to the combined endpoint, it is worth emphasizing that arrhythmia may be a surrogate marker for dysfunctional systemic ventricles and indirectly may also lead to n-SCD [45].

Pacemaker

Pacemaker implantation did not show a statistically significant relationship with long-term mortality in patients after AtrSR. Due to altered heart anatomy and frequent secondary atrioventricular conduction disturbances, we assumed that this might have an impact on survival in patients with corrected D-TGA [3, 30, 46]. However, similarly to other authors, implantation of a pacemaker was not a statistically significant predictive risk factor for death [11, 30, 47]. This finding confirms that pacemaker therapy is an effective treatment for various conduction system disorders.

Study limitations

The studies included in this meta-analysis are characterized by large discrepancies in the era and quality of medical services, such as cardiac surgery, extracorporeal circulation, or postoperative care. In addition, not all studies used cardioplegia, and patients differed in comorbidities and socioeconomic conditions. This results in a very high heterogeneity of the studies included in this meta-analysis. As these were mainly retrospective studies, it should be underlined that all of them differed in study design, risk factor definitions, and endpoints. We were unable to produce a unified definition for the D-TGA complex. Additionally, we also adopted an RVEF cutoff point of <45% as a criterion for RVD, though the majority of publications defined RVD as RVEF <40%. Importantly, the rated and analyzed outcomes parameters, such as TVR, RVD, or NYHA class, were based on our subjective judgment and clinical experience. No prospective screening with echocardiography, electrocardiography, or other modality was performed, and the outcomes may be incorrectly estimated. In our opinion, the results describing the outcomes, such as right ventricular dysfunction or tricuspid valve regurgitation, may have the largest error because of our subjective assessment. Lastly, as the risk factors included in our study are derived from univariate analyses, the multicollinearity between them may occur, which should be kept in mind in clinical practice.

CONCLUSIONS

Heart failure decompensation and at least moderate tricuspid valve regurgitation have the biggest impact on long-term survival in patients after the atrial switch procedure for D-transposition of the great arteries. Right ventricular dysfunction, the complexity of the congenital heart defect, and Mustard procedure are also risk factors for mortality in

these patients. Supraventricular tachyarrhythmia is a risk factor for sudden cardiac death but does not affect all-cause mortality in this group.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska

Article information

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