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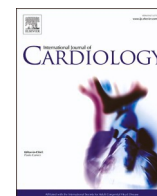
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Long-term outcome of patients with transposition of the great arteries and a systemic right ventricle: A systematic review and meta-analysis

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ABSTRACT

Background: Patients with a transposition of the great arteries (TGA) and a systemic right ventricle are at risk of heart failure (HF) development, arrhythmia and early mortality. Prognostic evaluations in clinical studies are hampered by small sample sizes and single-centred approaches. We aimed to investigate yearly rate of outcome and factors affecting it.

Methods: A systematic literature search of four electronic databases (PubMed, EMBASE, Web of Science and Scopus) was conducted from inception to June 2022. Studies reporting the association of a systemic right ventricle with mortality with a minimal follow-up of 2 years during adulthood were selected. Incidence of HF hospitalization and/or arrhythmia were captured as additional endpoints. For each outcome, a summary effect estimate was calculated.

Results: From a total of 3891 identified records, 56 studies met the selection criteria. These studies described the follow-up (on average 7.27 years) of 5358 systemic right ventricle patients. The mortality incidence was 1.3 (1–1.7) per 100 patients/year. The incidence of HF hospitalization was 2.6 (1.9–3.7) per 100 patients/year. Predictors of poor outcome were a lower left ventricular (LV) and right ventricular ejection fraction (RVEF) (standardized mean differences (SMD) of –0.43 (–0.77 to –0.09) and –0.85 (–1.35 to –0.35), respectively), higher plasma concentrations of NT-proBNP (SMD of 1.24 (0.49–1.99)), and NYHA class ≥ 2 (risk ratio of 2.17 (1.40–3.35)).

Conclusions: TGA patients with a systemic right ventricle have increased incidence of mortality and HF hospitalization. A lower LVEF and RVEF, higher levels of NT-proBNP and NYHA class ≥ 2 are associated with poor outcome.

1. Introduction

Patients with Dextro-transposition of the great arteries (D-TGA) who have had an atrial switch procedure (Mustard/Senning) or patients with a congenitally corrected transposition of the great arteries (ccTGA) who do not have had corrective surgery, have a systemic right ventricle [1]. Although D-TGA post-Mustard/Senning and ccTGA are clearly different anatomical congenital anomalies, the resemblance of all systemic right ventricle patients is that their morphological right ventricle (RV) delivers blood directly to the aorta. Patients with a systemic right ventricle form a significant part of the adult congenital heart disease (ACHD)

population (10–12%) [1]. Due to the different anatomy and physiology of the RV in comparison to the left ventricle (LV), systemic right ventricle patients are at higher risk for heart failure (HF) hospitalizations and mortality [2]. With the increasing number of adults surviving with a systemic right ventricle in the past years and the long waiting lists for heart transplantation, these patients are at increased risk for HF hospitalizations, multiple organ failure and mortality [3]. There have been many studies reporting early postoperative outcome for the paediatric population in a consistent way [4–7] but prospective studies reporting long-term outcomes in adults are heterogeneous in terms of sample size, follow-up period and description of baseline variables

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[8–10]. Therefore, we conducted this systematic review and meta-analysis of the previously published literature and formulated the following goals:

1. To investigate the yearly mortality rate in systemic right ventricle patients;
2. To compare this mortality rate with an aged-matched comparison group from previous literature;
3. To compare mortality rate of patients with ccTGA with patients with D-TGA post Mustard/Senning;
4. To investigate the yearly rate of other, secondary outcomes, such as HF hospitalizations, heart transplantation, ventricular - and supra-ventricular arrhythmias;
5. To identify factors associated with poor outcomes in systemic right ventricle patients.

2. Methods

This systematic review and meta-analysis was conducted based on the recommendations of the Cochrane Handbook for Systematic Reviews of Interventions and reported according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement [11,12]. The study protocol was registered in the international prospective register of systematic reviews of the National Institute for Health Research (PROSPERO) prior to the data extraction with registration number CRD42021282276.

2.1. Study identification

A systematic literature search of four electronic databases (PubMed, EMBASE, Web of Science and Scopus) was conducted. The search strategy consisted of medical subject heading terms and free-text words (Supplementary table 1). The initial search was performed on June 03, 2021, and was updated on June 22, 2022. Additionally, the reference lists of the included studies were screened for relevant studies. No language or time restrictions were applied.

2.2. Study selection

Inclusion criteria were formulated using the PICO format. The population included patients with a minimum age of 16 years with D-TGA repaired by either a Senning or Mustard procedure and patients with ccTGA. Because we were not interested in the short term (surgical) outcomes at infancy, we included only studies with at least 2 years of follow-up during adulthood. The intervention/exposure was a systemic right ventricle after either a Senning procedure, Mustard procedure or a diagnosis of ccTGA. The primary outcome was mortality. Also, when reported, incidence of HF hospitalization, heart transplantation and ventricular or supraventricular arrhythmias were captured as additional endpoints.

Exclusion criteria were met when study patients had an arterial switch operation in D-TGA or double switch operation for ccTGA patients, putting the left ventricle at the systemic position. Also, different articles using the same patient cohort were reviewed only if they reported on different endpoints or differed in follow-up period. Case reports, case series with fewer than 5 cases, review articles, and conference abstracts were excluded. Studies that did not report follow-up duration or studies with a follow-up duration (mean or median) of <2 years and studies that did not report on the endpoint-of-interest (i.e., mortality) were excluded.

Two reviewers (M.M.A.R. and H.Y.) independently assessed titles and abstracts for eligibility. If the title and abstract did not provide sufficient information, or in case the reviewers had doubt, independent full-text assessment was done. Any disagreement was discussed and if no consensus could be reached, a third independent reviewer (J.v.M.) made the final decision. After each selection step, inter-observer reliability

was calculated and expressed as Cohen's kappa. Studies in languages other than English were translated to English by the help of colleagues fluent in both languages.

2.3. Data collection

The data were extracted using a pre-defined form by the same reviewers. The collected data included: first author and year of publication, country in which the study was conducted, study design, number of patients, patient population (ccTGA or D-TGA or both), mean age at baseline, follow-up duration, events (including mortality, HF hospitalization, ventricular arrhythmia, supraventricular arrhythmia and transplantation), and predefined factors possibly associated with these events (e.g. NYHA class, LVEF, RVEF, NT-proBNP). If the same endpoint was assessed in multiple manuscripts of the same population, the manuscript with the longest follow-up or with more related data was included for data collection.

2.4. Quality assessment

The quality of all the included studies was independently assessed by the same two reviewers. Cohort and case control studies were assessed using the Newcastle-Ottawa Quality Assessment Form for Cohort Studies [13]. The studies were graded with stars in 3 domains of selection, comparability and outcome/exposure. Case series were graded based on a proposed tool by Murad et al. assessing criteria related to the selection, ascertainment, causality and reporting of the cases [14]. Randomised controlled trials were graded based on the Cochrane risk of bias tool [15].

2.5. Statistical analysis and data synthesis

The inter-observer agreement was calculated using IBM Statistical Package for the Social Sciences Statistics 23 (SPSS, Chicago, IL, USA) and expressed by the Cohen's Kappa.

For each outcome, the annual incidence was calculated accompanied with the 95% confidence intervals (95%CI). Statistical heterogeneity was regarded substantial if $I^2 > 50$ [12]. If ≥ 2 studies could be pooled, a summary effect estimate was calculated for each outcome as well as for the a priori defined subgroups, i.e., ccTGA and TGA with atrial switch. The meta-analysis was performed in R-meta (R v4.0.2, meta-package v4.15–1), using a random-effects model, with the DerSimonian-Laird estimator, due to the presence of clinical heterogeneity [12,16].

Meta-regression analyses based on mixed-effects models with the DerSimonian-Laird estimator was performed to regress each outcome on the subgroups and age at baseline. Here, each outcome was logit transformed to ensure a linear association between age at baseline and the outcome. The assumptions of all models were tested and verified. Meta-regression analyses were only performed if >10 studies could be included [12]. All meta-regression models yielded an estimated regression coefficient (β) with corresponding odds ratios and 95%CI. In addition, the association between right ventricular ejection fraction (RVEF), left ventricular ejection fraction (LVEF), N-terminal pro-brain natriuretic peptide (NT-proBNP) and New York Heart Association (NYHA) functional class with cardiac events was assessed. Standardized mean differences of RVEF, LVEF and NT-proBNP between patients without and with events were calculated. Furthermore, the risk ratio for cardiac events between patients with NYHA class 1 and NYHA ≥ 2 was assessed. In all analyses, $P < 0.05$ (two-tailed) was considered statistically significant.

3. Results

3.1. Identified studies

A total of 3887 records were identified through database search and

4 records through citation search (Fig. 1). After removal of 2031 duplicates, the titles and abstracts were screened and 1545 records were excluded. This yielded 311 articles for full-text review and screening. After screening the full texts, and with the addition of 4 records that were identified through citation searching, 56 articles were identified that met the inclusion criteria of the systematic review [8–10,17–69]. Three studies were removed before the quantitative analysis because (although the mean age was above 16), they also included paediatric patients in their population [46,51,70]. The Cohen’s Kappa for title/abstract and full text screening was 0.80 and 0.87, respectively.

3.2. Study characteristics and quality assessment

The characteristics of the included studies are summarized in supplementary table 2. There were 19 studies reporting patients with ccTGA, 20 studies reporting TGA with atrial switch and 17 studies included both patient populations. The studies were published from 1982 to 2022. Nine of the included studies were case series, one was a randomised controlled trial investigating the effect of valsartan on systemic right ventricle function, 35 were retrospective studies and 11 were prospective cohorts. The follow-up duration of the studies varies between 2 and 30 years.

The quality assessment of the cohort and case-control studies is included in the supplementary table 2. All case series satisfied the selection, ascertainment and reporting domains of the assessment tool used. Except for 2 case series [23,64], all others satisfied the causality domain of the assessment tool used. The only randomised controlled trial had low risk of bias in the domains of randomisation process, effect of assignment to intervention, effect of adhering to intervention, missing outcome data, and measurement of the outcome. There were some concerns for risk of bias in the selection of the reported result domain.

3.3. Incidence of mortality

Based on 53 studies including 5280 patients, the mortality incidence was 1.3 (95%CI = 1.0–1.7) per 100 patients/year (Fig. 2). The incidence of mortality was not associated with mean age at baseline (OR: 1.03 (95%CI = 0.99–1.07) (Fig. 3A) and there was no difference between the mortality incidence in patients with TGA after atrial switch (1.1 [0.7–1.7] per 100 patients/year) and ccTGA (1.9 [1.3–2.9] per 100 patients/year) when adjusted for age ($p = 0.11$) (Fig. 3B). The incidence of mortality in individual studies specifying TGA and atrial switch and ccTGA patients is shown in supplementary Fig. 1.

3.4. Incidence of heart failure hospitalization and transplantation

From the included studies, 22 manuscripts (3104 patients) reported HF hospitalization as outcome. The incidence of HF hospitalization was 2.6 (95%CI = 1.9–3.7) per 100 patients/year (Supplementary Fig. 2A). The incidence of HF hospitalization was not associated with mean age at baseline (OR: 1.06 (95%CI = 0.98–1.14) (Supplementary Fig. 2B). Patients with ccTGA had higher incidence of HF hospitalization (4.3 [2.7–6.6]) in comparison to patients with TGA and atrial switch (3.2 [2.3–4.4]) when adjusted for age ($p = 0.04$) (Supplementary Fig. 2C). The incidence of HF hospitalization in individual studies specifying TGA and atrial switch and ccTGA patients is shown in supplementary Fig. 3.

Twenty-two studies (2144 patients) reported transplantation as outcome. The incidence of transplantation was 1.0 (95%CI = 0.5–2.1) per 100 patients/year (Supplementary Fig. 4A). The incidence of transplantation was not associated with mean age at baseline (OR: 1.07 (95%CI = 0.95–1.21) (Supplementary Fig. 4B). There was no difference between the transplantation incidence in patients with TGA and atrial switch (0.3 [0.1–1.2] per 100 patients/year) and ccTGA (0.8 [0.3–2.2] per 100 patients/year) when adjusted for age ($p = 0.51$) (Supplementary

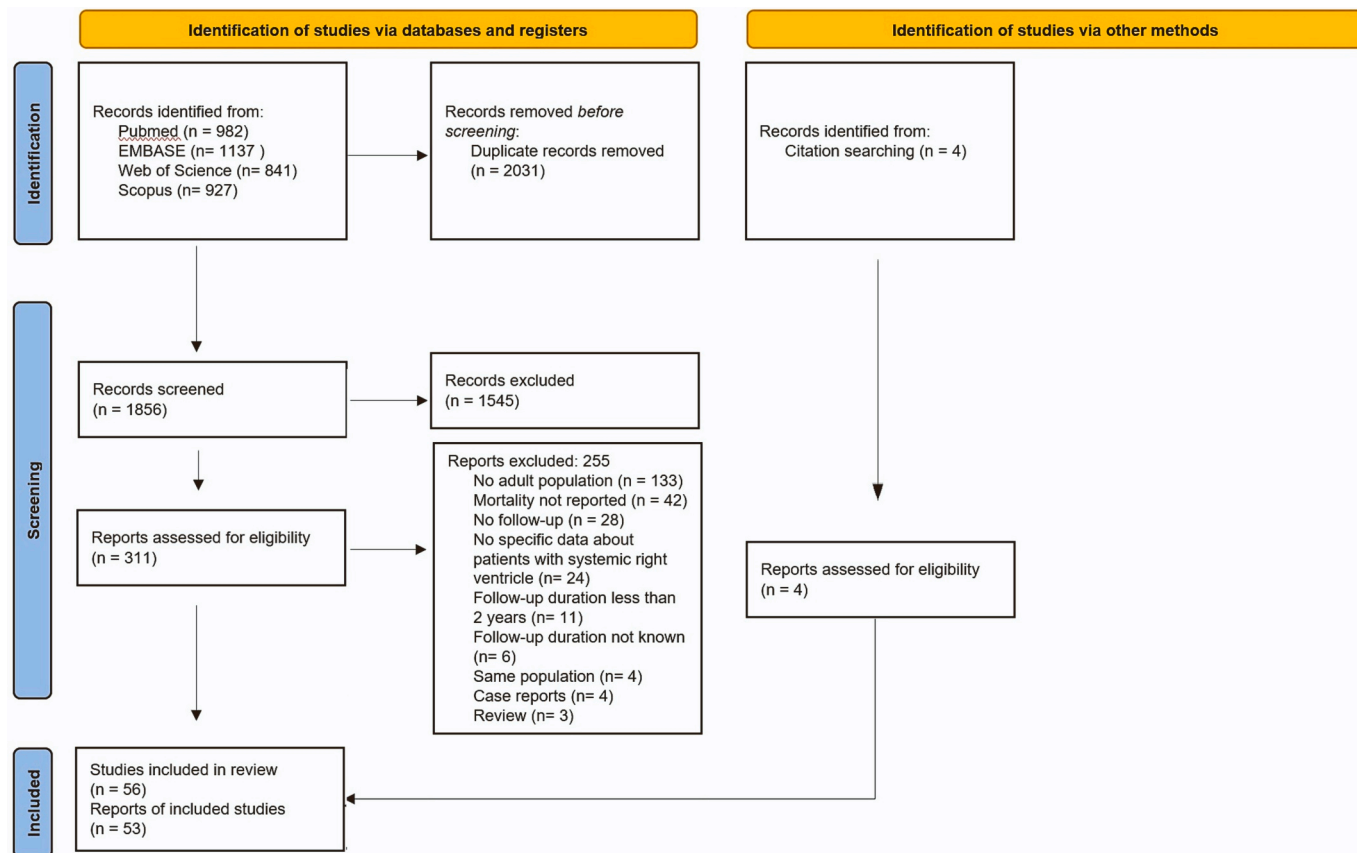


Fig. 1. PRISMA flow diagram.

Fig. 4C). The incidence of transplantation in individual studies specifying TGA and atrial switch and cTGA patients is shown in supplementary Fig. 5.

3.5. Incidence of ventricular arrhythmias

Nineteen studies (2746 patients) reported ventricular arrhythmias as outcome. The incidence of ventricular arrhythmias was 1.1 (95%CI =

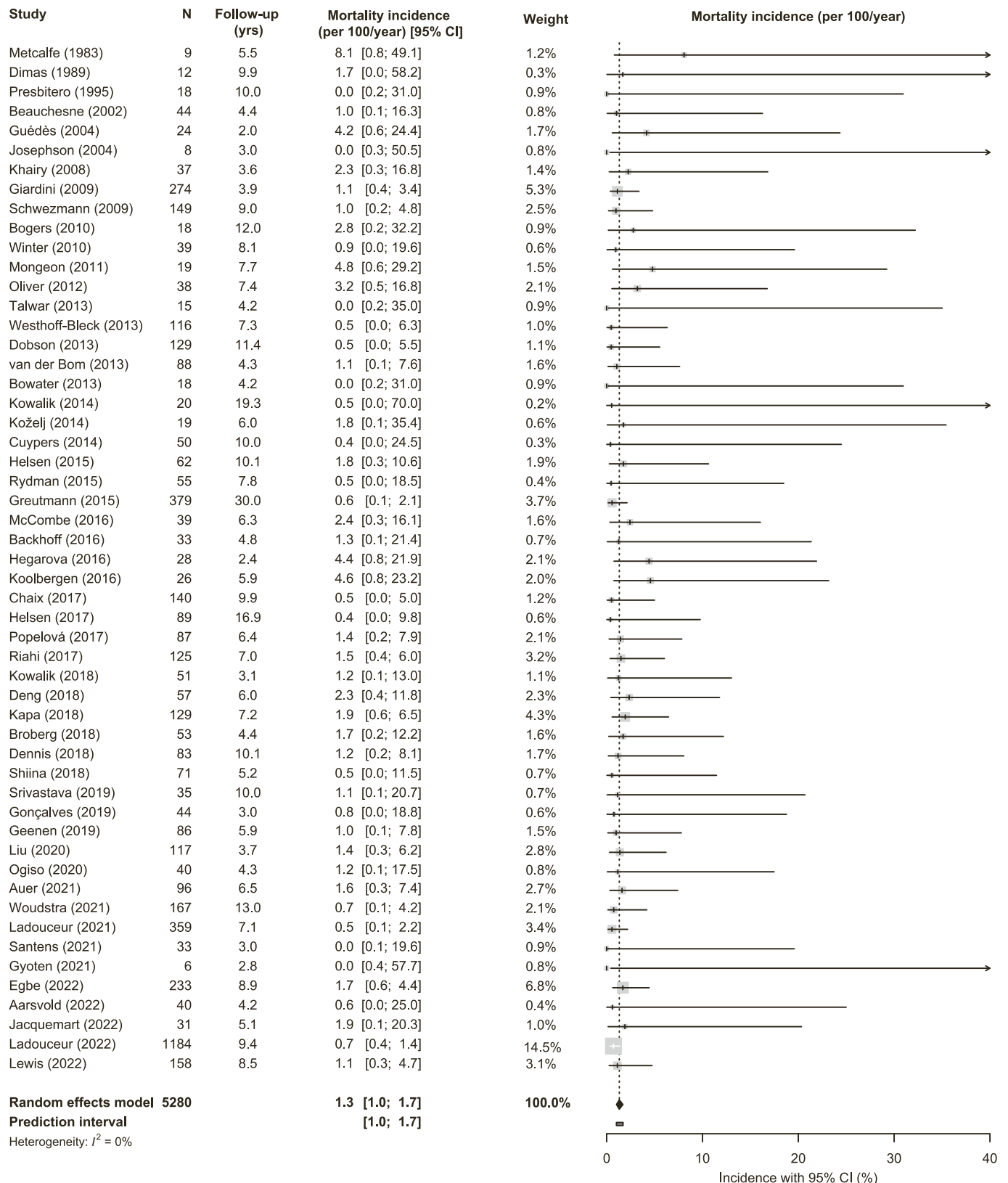


Fig. 2. Forest plot depicting studies reporting mortality. Mortality incidence (per 100 patients/year) [95% CI] is calculated for each study and shown.

0.7–1.6) per 100 patients/year (Supplementary Fig. 6A). With increasing mean age at baseline, the incidence of ventricular arrhythmias increased (OR: 1.09 (95%CI = 1.02–1.17) (Supplementary Fig. 6B). There was no difference between the ventricular arrhythmia incidence in patients with TGA and atrial switch (0.8 [0.4–1.6] per 100 patients/year) and ccTGA (2.4 [1.2–4.6] per 100 patients/year) when adjusted for age ($p = 0.20$) (Supplementary Fig. 6C). The incidence of ventricular arrhythmias in individual studies specifying TGA and atrial switch and ccTGA patients is shown in supplementary Fig. 7. Not all studies specified the type of ventricular arrhythmia. Eleven studies reported patients with sustained ventricular tachycardia or ventricular fibrillation [8,9,19,21,26,38,40,53,63,67,68]. The other studies either did not specify or were patients with non-sustained VT.

3.6. Incidence of supraventricular arrhythmias

Twenty-three studies (2649 patients) reported supraventricular arrhythmias as outcome. The incidence of supraventricular arrhythmias was 3.6 (95%CI = 2.4–5.2) per 100 patients/year (Supplementary Fig. 8A). The incidence of supraventricular arrhythmias was not associated with mean age at baseline (OR: 1.03 (95%CI = 0.97–1.10) (Supplementary Fig. 8B). There was no difference between the supraventricular arrhythmia incidence in patients with TGA and atrial switch (3.6 [2.4–5.3] per 100 patients/year) and ccTGA (4.2 [2.3–7.6] per 100 patients/year) when adjusted for age ($p = 0.44$) (Supplementary Fig. 8C). The incidence of supraventricular arrhythmias in individual studies specifying TGA and atrial switch and ccTGA patients is shown in supplementary Fig. 9. Not all studies reported if the supraventricular arrhythmias were symptomatic and required treatment. Three studies reported arrhythmias requiring cardioversion or other treatment not specified [24,61,63].

3.7. Factors associated with poor outcome

Studies comparing baseline variables in patients who had cardiac events or not were pooled if they reported baseline values of right ventricular ejection fraction (RVEF), left ventricular ejection fraction (LVEF), N-terminal pro-brain natriuretic peptide (NT-proBNP) and New York Heart Association (NYHA) functional class.

As shown in Fig. 4A, patients with poor outcome had lower RVEF with a standardized mean difference (SMD) of -0.85 (95%CI = -1.35 to -0.35) ($p < 0.001$). Patients with poor outcome also had lower LVEF (Fig. 4B) with an SMD of -0.43 (95%CI = -0.77 to -0.09) ($p = 0.013$). In Fig. 4C it is shown that patients with poor outcome have higher levels of NT-proBNP with an SMD of 1.24 (95%CI = 0.49 – 1.99) ($p = 0.001$). Patients who experienced poor outcomes were more likely to have NYHA functional class higher or equal to 2 (Fig. 4D) with a risk ratio of 2.17 (95%CI = 1.40 – 3.35) ($p < 0.001$).

4. Discussion

In the present systematic review, we aimed to investigate the incidence of mortality in patients with a systemic right ventricle. We also studied the incidence of other events such as HF hospitalizations, ventricular arrhythmias, supraventricular arrhythmias and transplantation.

With the main results of this meta-analysis, comprising studies that were published between 1982 and 2022, we were able to calculate a pooled estimate of the incidence of mortality (1.3 per 100 patients/year), the incidence of HF hospitalization (2.6 per 100 patients/year), the incidence of transplantation (1.0 per 100 patients/year), the incidence of ventricular arrhythmias (1.1 per 100 patients/year), and the incidence of supraventricular arrhythmias (3.6 per 100 patients/year). There was no relation found between the mean age of the patients at baseline and incidence of mortality. This may be related to the fact that most of the individual studies included young adults (average age at baseline 30.9 years).

Our findings highlight the increased long-term mortality, despite advances in surgical techniques and medical management, associated with a systemic right ventricle. Patients with a systemic right ventricle have a higher incidence of mortality when compared to adults aged 25–64 during the years 1968–2020 in Europe (standardized death rate ranging from 0.20 to 1.11 per 100 patients/year) [71]. In addition, the present study also confirms that patients with a systemic right ventricle have a particularly high risk of HF hospitalization, with an incidence more than twice that of mortality. This is likely due to the increased hemodynamic burden on the sub-aortic RV in these patients, which can lead to progressive ventricular dysfunction [72]. Several explanations have been suggested for the observed decline in RV function, including the longstanding pressure overload, myocardial ischemia, tricuspid regurgitation, mechanical dyssynchrony and arrhythmias [3].

Our study was able to show that patients with ccTGA have higher incidence of HF hospitalizations even after adjusting for age. This might be due to the fact that these patients are usually diagnosed later than patients with a TGA and atrial switch, which makes the systemic right ventricle more vulnerable to dysfunction and subsequent development and progression of HF [21]. The ccTGA patients also have more prevalence of tricuspid regurgitation (e.g. associated Ebstein's disease) which has a detrimental effect in worsening their HF and putting them at risk for HF hospitalizations [72]. Unfortunately, there is still a lack in proper therapeutic options to improve a failing systemic right ventricle [3]. A recent open label study has risen hope by suggesting that sacubitril/valsartan improved quality of life, functional status and imaging parameters of these patients [72]. However, large trials to confirm these findings are still lacking.

In our study, incidence of ventricular arrhythmias increases with age of patients. This might be due to the fact that older patients have had HF for a longer period and therefore have more myocardial fibrosis leading to higher risk of ventricular arrhythmias [40]. Myocardial fibrosis is not only a contributing factor for ventricular arrhythmias, but also contributes to the development of supraventricular arrhythmias. Therefore, it is important to include such imaging modalities in clinical practice and predictive models [40].

It was hypothesized that more supraventricular arrhythmias in patients with TGA and atrial switch were observed due to the atrial surgical redirection of blood flows [73]. However, no difference in the incidence of supraventricular arrhythmias between these patients and the ccTGA patients was observed. One possible reason could be that the studies reported new supraventricular events, which might be comparable in ccTGA patients, who are usually diagnosed later versus TGA patients with an atrial switch (who are already under treatment for previous supraventricular events). Another possible reason could be that supraventricular arrhythmias increase over time in patients with ccTGA [74]. In concordance with data derived from mainstream cardiology, the predictive value of RVEF, LVEF, NT-proBNP and NYHA functional class for adverse outcomes was confirmed in this meta-analysis in patients with a systemic right ventricle. RVEF was measured with echocardiography in three of the studies [34,44,49] and cardiac MRI in the other two [28,63]. In order to see if the modality used has an effect on the results, we looked at two studies [34,49] that used both modalities for measuring the ventricular function. Although there were differences in the reported ventricular function across the modalities, the difference between patients with adverse outcomes and those without adverse outcomes remained consistent. In studies reporting NT-proBNP, RVEF and LVEF [34,49], NT-proBNP was also a good predictor of outcome in one of the studies (i.e. with a high HR of 96.16 (95%CI = 11.03–838.10)). In comparison with studies which report NT-proBNP as a predictor for adverse outcome in patients with a systemic left ventricle, it seems that NT-proBNP has a higher predictive value in patients with a systemic right ventricle in this study [49]. Considering the predictive value of LVEF for outcomes in patients with a systemic right ventricle [34,63], it is important to stress that clinicians acknowledge the evaluation of the subpulmonary ventricular function in these patients.

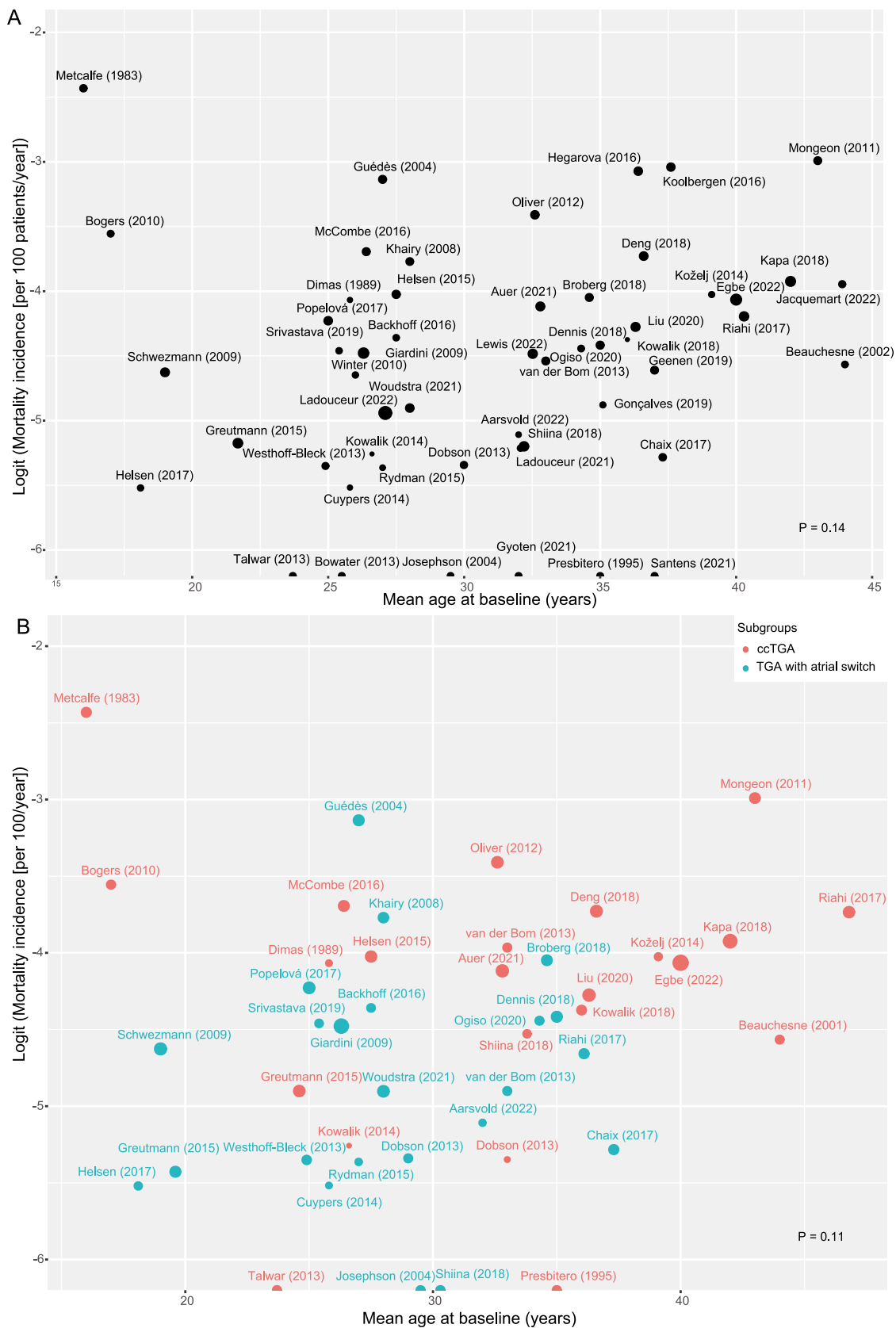


Fig. 3. Bubble plots depicting A: relationship between increasing age at baseline and incidence of mortality, B: difference between incidence of mortality in ccTGA versus TGA with atrial switch patients when adjusted for age. ccTGA congenitally corrected transposition of the great arteries, TGA transposition of the great arteries.

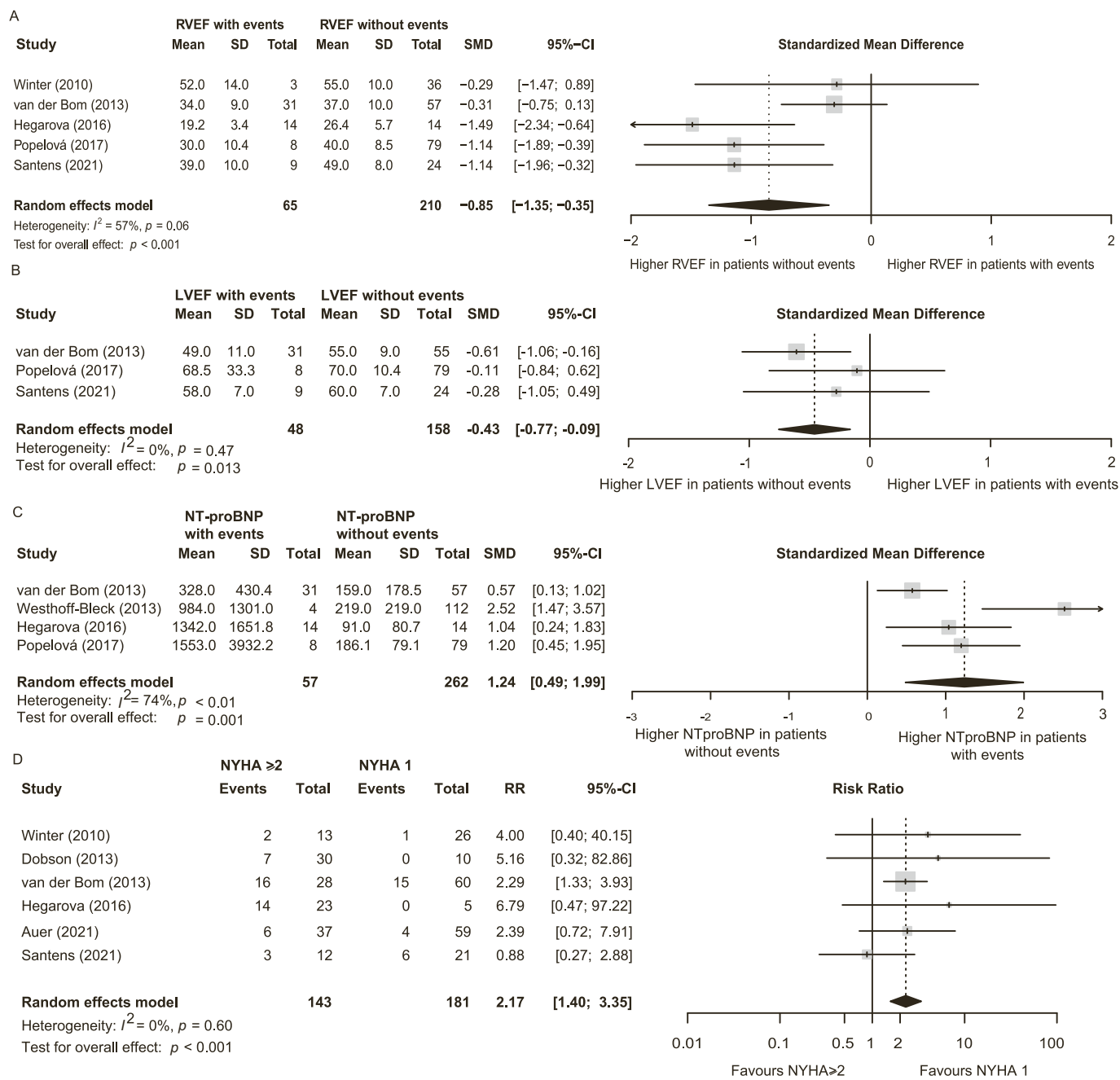


Fig. 4. Forest plots depicting A: association of lower RVEF with poor outcome, B: association of lower LVEF with poor outcome, C: association of higher NT-proBNP with poor outcome, D: association of NYHA ≥2 with poor outcome. Poor outcome included mortality, heart failure hospitalizations, ventricular and supraventricular arrhythmias.

Our study has several limitations. In this study we calculated a yearly incidence of mortality and other events based on the mean duration of follow-up of the individual studies. This assumes a linear mortality rate during follow-up. Although the data showed no statistical heterogeneity ($I^2 < 50\%$), it is preferred to include the follow-up of individual patients. This requires sharing of cohort data to perform an individual patient data meta-analysis which was not feasible at the time of writing. Future research should encourage collaborations between research groups so that sharing cohort data to perform such analysis is possible. In addition, because of the heterogeneity of the reported variables it was not possible to evaluate all the reported variables that may have predictive value for outcome. To minimize heterogeneity, we

therefore selected the variables that were uniformly reported across several studies.

In conclusion, patients with a systemic right ventricle have increased risk of mortality, heart failure hospitalizations and arrhythmia. A lower LVEF and RVEF, higher levels of NT-proBNP and NYHA class ≥ 2 are associated with poor outcome in this population.

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ijcard.2023.131159>.

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CRediT authorship contribution statement

Mohammad Mostafa Ansari Ramandi: Conceptualization, Methodology, Formal analysis, Investigation, Writing – original draft, Visualization, Writing – review & editing. **Hossein Yarmohammadi:** Methodology, Investigation, Writing – review & editing. **Barzi Gareb:** Methodology, Formal analysis, Writing – review & editing. **Adriaan A. Voors:** Conceptualization, Supervision, Writing – review & editing. **Joost P. van Melle:** Conceptualization, Methodology, Investigation, Supervision, Funding acquisition, Writing – review & editing.

Declaration of Competing Interest

None.

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