Identification of patients at risk of sudden cardiac death in congenital heart disease: The PRospEctiVE study on implaNTable cardIOverter defibrillator therapy and suddeN cardiac death in Adults with Congenital Heart Disease (PREVENTION-ACHD)

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**BACKGROUND** Sudden cardiac death (SCD) is the main preventable cause of death in patients with adult congenital heart disease (ACHD). Since robust risk stratification methods are lacking, we developed a risk score model to predict SCD in patients with ACHD: the PRospEctiVE study on implaNTable cardIOverter defibrillator therapy and suddeN cardiac death in Adults with Congenital Heart Disease (PREVENTION-ACHD) risk score model.

**OBJECTIVE** The purpose of this study was to prospectively study predicted SCD risk using the PREVENTION-ACHD risk score model and actual SCD and sustained ventricular tachycardia/ventricular fibrillation (VT/VF) rates in patients with ACHD.

**METHODS** The PREVENTION-ACHD risk score model assigns 1 point each to coronary artery disease, New York Heart Association class II/III heart failure, supraventricular tachycardia, systemic ejection fraction < 40%, subpulmonary ejection fraction < 40%, QRS duration ≥ 120 ms, and QT dispersion ≥ 70 ms. SCD risk was calculated for each patient. An annual predicted risk of ≥ 3% constituted high risk. The primary outcome was SCD or VT/VF after 2 years. The secondary outcome was SCD.

**RESULTS** The study included 783 consecutive patients with ACHD (n = 239 (31%) left-sided lesions; n = 138 (18%) tetralogy of Fallot; n = 108 (14%) closed atrial septal defect; median age 36 years; interquartile range 28–47 years; n = 401 (51%) men). The PREVENTION-ACHD risk score model identified 58 high-risk patients. Eight patients (4 at high risk) experienced the primary outcome. The Kaplan-Meier estimates were 7% (95% confidence interval [CI] 0.1%–13.3%) in the high-risk group and 0.6% (95% CI 0.0%–1.1%) in the low-risk group (hazard ratio 12.5; 95% CI 3.1–50.9; P < .001). The risk score model's sensitivity was 0.5 and specificity 0.93, resulting in a C-statistic of 0.75 (95% CI 0.57–0.90). The hazard ratio for SCD was 12.4 (95% CI 1.8–88.1) (P = .01); the sensi-
tivity and specificity were 0.5 and 0.92, and the C-statistic was 0.81 (95% CI 0.67–0.95).

**CONCLUSION** The PREVENTION-ACHD risk score model provides greater accuracy in SCD or VT/VF risk stratification as compared with current guideline indications and identifies patients with ACHD who may benefit from preventive implantable cardioverter-defibrillator implantation.

**Introduction**
Congenital heart defects affect up to 1.6% of newborns. Because of surgical and medical improvements, mortality patterns have transitioned from the pediatric to the adult population. Long-term complications in patients with adult congenital heart disease (ACHD), including sudden cardiac death (SCD), are frequent. SCD constitutes 20%–40% of all deaths in the population with ACHD, and victims are on average only 36 years old. SCD is mainly caused by ventricular arrhythmias and may be prevented by implantable cardioverter-defibrillator (ICD) implantation. ICD implantation for the primary prevention of SCD is routine clinical practice in patients with ischemic or nonischemic cardiomyopathy and left ventricular dysfunction. However, because of the absence of randomized trials or accurate risk prediction algorithms and a high rate of—mainly lead-related—complications and inappropriate shocks, ICD use for primary prevention in patients with ACHD lags behind. Moreover, SCD risk differs among congenital defects, and currently, tetralogy of Fallot is the only defect listed separately in the guidelines.

Efforts to predict SCD in patients with ACHD have led to several consensus statements and guidelines with class 1, 2a, and 2b ICD indications for patients with ACHD. However, in a recent analysis, current ICD indications were moderately predictive for SCD, and many patients with ACHD still die of SCD. Risk factors for SCD in ACHD include impaired systemic ventricular function, heart failure symptoms, prolonged QRS duration, and atrial arrhythmias. Independently, these risk factors have a low predictive ability for SCD, but in combination they may provide more accurate risk stratification. An SCD risk score model, applicable to a wide spectrum of ACHD defects, may help better identify patients at risk of SCD and select patients who may benefit from primary prevention ICD implantation. We developed the PRospEctiVE study on implantable cardioverter-defibrillator therapy and sudden cardiac death in Adults with Congenital Heart Disease (PREVENTION-ACHD) risk score model, which was derived from a cohort of patients with ACHD composed of SCD cases and matched living controls. The risk score model is built to provide estimates of the annual SCD risk of patients with those congenital defects that cause the highest risk, who may benefit from ICD implantation. We hypothesize that the PREVENTION-ACHD risk score model more accurately predicts the occurrence of SCD in the general population with ACHD than do the currently accepted ICD indications.

**Methods**

**Study population**
This single-center prospective observational study was performed at the Amsterdam University Medical Centers, Amsterdam, The Netherlands, a tertiary referral center for ACHD. A detailed description of the design of this study is described elsewhere. In short, we included consecutive patients with ACHD 18 years and older presenting to the outpatient clinic from January 1, 2015, to December 31, 2015. We excluded patients with a secondary prevention ICD indication (ie, sustained ventricular tachycardia (VT), ventricular fibrillation (VF), or resuscitated cardiac arrest), those not planned for consultation within 2 years, and those with a contraindication for ICD implantation according to the guidelines.

The obligation to obtain informed consent was waived by the ethics committee at the Amsterdam University Medical Centers because of the study’s observational nature. The study was registered at ClinicalTrials.gov (identifier NCT03957824).

**Data collection**
Patients’ risk factors were obtained from medical records. Results from routine examinations, including electrocardiograms (ECGs), echocardiograms, and cardiac magnetic resonance imaging, were reviewed. The findings of these examinations were included when available within 1 year of patients’ respective outpatient appointments. Imaging reports were reviewed by imaging cardiologists, specialized in adult congenital cardiology. ECGs were measured using ImageJ (US National Institutes of Health, Bethesda, Maryland, United States).

**Risk score model**
The PREVENTION-ACHD risk score model (Figure 1) was constructed from risk factors established by a previous study in a database of 25,790 patients with ACHD. In that study, patients who died of SCD were included and matched to living controls by age, sex, diagnosis, type of surgical intervention, date of surgical repair, and treating medical center.
A careful selection process, described in detail in the design paper, was used to identify risk factors for the risk score model. The risk factors that are included in the risk score are displayed in Figure 1. The risk score model is used to calculate the individualized annual risk of SCD based on the risk factors multiplied by the baseline hazard of the patient’s congenital defect. The baseline hazard per congenital defect was derived from the CONCOR (CONgenital CORvitia) registry. All high-risk congenital defects were included in the risk score model.

For the present study, the risk of SCD was prospectively calculated for each patient using the PREVENTION-ACHD risk score model. Patients with an estimated annual SCD risk of ≥3% were considered at high risk and patients with an annual risk of <3% as low-risk controls. Patients with defects not included in the PREVENTION-ACHD risk score model have a very low incidence of SCD and were, therefore, deemed to be at low risk. Additionally, we tested the risk score model’s exposure-response relationship when the control group is partitioned into a group of (very) low-risk patients (annual SCD risk <1%) and an intermediate-risk group of patients (annual SCD risk 1% or 2%).

Outcome measurement

After ≥2 years of follow-up after the index visit, the medical record of each patient included in the study was reexamined for the predefined outcomes. The primary outcome of this study was the combined end point of SCD or VT/VF. SCD was defined as (1) proven or documented arrhythmic death, (2) arrhythmic death by exclusion (instantaneous death or circumstances compatible with SCD, without disease that would lead to death in the near future, and in the absence of a nonarrhythmic cause of death at autopsy), or (3) arrhythmic death by default (abrupt loss of consciousness and absence of pulse, without further data). VT/VF was defined as VT or VF lasting >30 seconds, aborted SCD, or appropriate ICD therapy. Appropriate ICD intervention was defined as antitachycardia pacing or ICD shock for proven VT or VF. The secondary outcome was SCD alone. These outcomes were described for patients with a high vs a low risk score and, to put the results in perspective, for patients with vs without a class 1, 2a or 2b ICD indication according to the PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. Patients with a follow-up duration of <2 years were censored at the last follow-up visit.

Statistical analysis

For normally distributed continuous variables, mean ± SD was calculated. Nonnormally distributed data were presented as median and interquartile range. Differences in normally distributed continuous variables were tested using 1-way analysis of variance; for nonnormally distributed continuous variables, the Kruskal-Wallis test was used. The Fisher exact test was used for categorical variables. Kaplan-Meier (KM)
survival estimates and plots were generated to demonstrate event-free survival from the end point of SCD or VT/VF at 2 years of follow-up; and the log-rank test was used to test the difference between high-risk and low-risk patients. Additionally, Cox proportional hazard models identified hazard ratios (HRs) between groups. The proportional hazards assumption was tested using the Schoenfeld individual test. Goodness of fit of the risk score model was tested using the Grønnesby and Borgan goodness-of-fit test. To assess the predictive value of the risk score model for SCD, the time-dependent sensitivity and specificity at 2 years of follow-up were calculated and receiver operating characteristic curves were created with area under the curve to determine the C-statistic. For all analyses, 2-tailed \( P \) values of \(< .05\) were considered statistically significant. All data were analyzed using R version 3.5.0 (R Foundation for Statistical Computing, Vienna, Austria).

### Results

The study included 783 patients (median age 36 years; interquartile range 28–47 years; \( n = 401 \) (51%) men) (Table 1). Eleven patients had an ICD at baseline. Left-sided defects, consisting of bicuspid aortic valve, aortic valve stenosis, aortic coarctation, and other aortic valve abnormalities, constituted the largest group of congenital defects (\( n = 239 \), 31%), followed by tetralogy of Fallot (\( n = 138 \), 18%) and closed atrial septal defect (\( n = 108 \), 14%).

#### Table 1 Baseline characteristics

| Characteristic                      | Low risk | High risk | \( P \) |
|-------------------------------------|----------|-----------|---------|
| N*                                 | 724      | 58        | <.001   |
| Age (y)                             | 35 (27–46) | 45 (38–55) | <.001   |
| Male sex                           | 372 (51) | 29 (50)   | .892    |
| **Main diagnosis**                  |          |           |         |
| Closed atrial septum defect         | 107 (15) | 1 (2)     |         |
| Tetralogy of Fallot                 | 126 (17) | 12 (21)   |         |
| Left-sided lesions \(^1\)           | 35 (32)  | 4 (7)     | <.001   |
| Congenitally corrected TGA          | 21 (3)   | 2 (3)     |         |
| TGA Mustard/Sennning                | 18 (2)   | 9 (16)    |         |
| Fontan circulation                  | 28 (4)   | 5 (9)     |         |
| Ebstein anomaly                     | 13 (2)   | 3 (5)     |         |
| Cyanotic non-Eisenmenger            | 0 (0)    | 10 (17)   |         |
| Eisenmenger syndrome                | 1 (0)    | 12 (21)   |         |
| **Other**                           | 175 (24) | 0 (0)     | <.001   |
| Coronary artery disease             | 12 (2)   | 4 (7)     | <.001   |
| Heart failure                       | 28 (4)   | 35 (60)   | <.001   |
| Supraventricular tachycardia        | 112 (15) | 31 (53)   | <.001   |
| Systemic ventricle                  |          |           | <.001   |
| Left (biventricular)                | 655 (90) | 31 (53)   |         |
| Right (biventricular)               | 39 (5)   | 10 (17)   |         |
| Single                              | 30 (4)   | 17 (29)   | <.001   |
| Impaired systemic ventricular function \(^1\) | 27 (4) | 18 (31) | <.001   |
| Impaired subpulmonary ventricular function \(^1\) | 47 (6) | 17 (29) | <.001   |
| Missing (patients with single ventricles) | 30 (4) | 17 (29) | <.001   |
| **QRS duration (ms)**               |          |           | <.001   |
| QRS duration ≥ 120 ms               | 115 ± 29 | 140 ± 38  |         |
| QRS duration ≥ 140 ms               | 247 (34) | 39 (67)   | <.001   |
| QRS duration ≥ 160 ms               | 149 (21) | 31 (53)   | <.001   |
| QT dispersion (ms)                  | 19 (3)   | 8 (14)    | .001    |
| QT dispersion ≥ 70 ms               | 66 ± 29  | 86 ± 27   | <.001   |
| Number of risk factors              | 1 (0–2)  | 3 (3–4)   | <.001   |
| Nonsustained ventricular tachycardia| 27 (4)   | 5 (9)     | .080    |
| Unexplained syncope                 | 7 (1)    | 0 (0)     | >.099   |

Values are presented as mean ± SD, median (interquartile range), or \( n \) (%).

*One patient with missing risk factors that resulted in an unknown risk status.

\(^1\)Left-sided lesions refer to bicuspid aortic valve, aortic valve stenosis, aortic coarctation, and other aortic valve abnormalities.

\(^2\)Other refers to patients with adult congenital heart disease defects not included in the PRospEctiVE study on implaNTable cardIOverter defibrillator therapy and sudden cardiac death in Adults with Congenital Heart Disease risk score model.

\(^x\)At least moderately impaired systolic function or ejection fraction < 40%.

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788 Heart Rhythm, Vol 18, No 5, May 2021
risk scores model. Included in the PRospEctiVE study on implantable cardIOverter de
great arteries. The risk score model had nonfatal ventricular arrhythmia.

dead during follow-up. Patients with a high risk score had an HR for SCD or VT/VF of 12.5 (95% CI 3.1–50.9; P < .001) compared with low-risk patients. The proportional hazards assumption was confirmed (P = .81), and the model showed no signs of poor goodness of fit (P = .13).

For the secondary end point of SCD alone, the KM estimate was 3.5% (95% CI 0.0%–8.1%) in high-risk patients vs 0.3% (95% CI 0.0%–0.7%) in low-risk patients (Figure 2B). The HR for SCD was 12.4 (95% CI 1.8–88.1; P = .01) for high-risk vs low-risk patients.

The PREVENTION-ACHD risk score model had a sensitivity of 0.5 and a specificity of 0.93 for SCD or VT/VF at 2 years and a sensitivity of 0.5 and specificity of 0.92 for SCD alone. The C-statistic of the PREVENTION-ACHD risk score model predicting SCD or VT/VF was 0.75 (95% CI 0.59–0.92). When predicting death from SCD alone, the C-statistic of the risk score model was 0.81 (95% CI 0.67–0.95).

Subdividing the low-risk group, 365 patients had an intermediate risk (1% or 2% annual SCD risk) and 359 patients a (very) low annual SCD risk (<1%). The KM estimates for SCD or VT/VF were 7.0%, 0.8%, and 0.3% in high-, intermediate- and (very) low risk patients, respectively, and for SCD 3.5%, 0.6%, and 0%, respectively (Figures 2C and 2D). Comparing intermediate and (very) low risk patients separately, the log-rank P values were nonsignificant (P = .33 for SCD or VT/VF and P = .16 for SCD).

### Consensus statement ICD indications

Nineteen patients had an ICD indication according to the PACES/HRS expert consensus statement. Of these, 6 already had an ICD implanted and 10 had a high SCD risk according to the PREVENTION-ACHD risk score model.

One patient had a class 1 ICD indication, 6 patients a class 2a indication, and 12 a class 2b indication. Online Supplemental Table 1 summarizes baseline characteristics of patients with and without an ICD indication. During follow-up, 2 patients with an ICD indication had VT/VF (KM estimate 10.8%; 95% CI 0.0%–24%). No patients with an ICD indication had SCD. However, 2 patients without an ICD indication had VT/VF and 4 died of SCD (total n = 6; KM estimate 0.8%; 95% CI 0.2%–1.5%) (Online Supplemental Figure 1). Patients with an ICD indication had an HR for SCD or VT/VF of 14.0 (95% CI 2.8–69.2; P = .001) compared with patients without an ICD indication.

The KM estimates for SCD were 0% and 0.5% (95% CI 0.0%–1.1%) (Online Supplemental Figure 1) in patients with and without an ICD indication, respectively. Consequently, the HR for SCD in patients with an ICD indication according to the PACES/HRS consensus statement compared with those without an ICD indication approached zero.

For SCD or VT/VF, the sensitivity of an ICD indication according to the consensus statement was 0.25 and specificity 0.98. For SCD alone, the sensitivity was 0 and specificity 0.98. The C-statistic of the consensus statement ICD indications predicting SCD or VT/VF was 0.61 (95% CI 0.46–0.76) and 0.49 (95% CI 0.48–0.49) for predicting SCD alone.

### Risk factors did not alter risk stratification status for these patients.

The PREVENTION-ACHD risk score model identified 58 high-risk patients of SCD, 2 of whom already had an ICD. Follow-up was complete in 711 of 724 low-risk patients (98%) and in all 58 high-risk patients. Loss to follow-up occurred mainly because the follow-up interval was set at >2 years by the treating physician or because patients did not return to follow-up yet. The 13 patients without follow-up were alive at follow-up, although data on ventricular arrhythmia were unavailable. Another 46 patients had a follow-up duration of <2 years. The median follow-up duration in these 46 patients was 1.5 years.

### Primary and secondary outcomes

During follow-up 10 patients died, 6 of whom after reaching the primary end point, and 6 ICDs were implanted (3 in high-risk patients).

The primary end point was reached in 8 patients (Table 2). Four patients died of SCD, and another 4 patients had aborted VT/VF. Among high-risk patients, 2 patients died of SCD, 1 patient was resuscitated for cardiac arrest, and 1 patient had sustained VT (n = 4; KM estimate 7%; 95% CI 0.1%–13.3%). The latter 2 had an ICD implanted during follow-up. Four low-risk patients experienced SCD or VT/VF (2 with SCD, appropriate ICD therapy in 1; KM estimate 0.6%; 95% CI 0.0%–1.1%) (Figure 2A). These 4 patients deceased during follow-up. Patients with a high risk score

| Characteristic | Value |
|---------------|-------|
| N             | 8     |
| Age (y)       | 44 (40–52) |
| Male sex      | 5 (62) |
| Main diagnosis |       |
| Tetralogy of Fallot | 2 (25) |
| Left-sided lesions | 3 (38) |
| Congenitally corrected TGA | 1 (12) |
| Eisenmenger syndrome | 1 (12) |
| Other*        | 1 (12) |
| ICD implanted | 1 (12) |
| High risk score |     |
| Consensus statement ICD indication | 1 (13) |
| only*         |       |
| High risk according to both models | 1 (13) |
| Outcome       |       |
| Sudden cardiac death | 4 (50) |
| Ventricular fibrillation (survived) | 2 (25) |
| Ventricular tachycardia (survived) | 2 (25) |

Values are presented as median (interquartile range) or n (%).

ICD = implantable cardioverter-defibrillator; TGA = transposition of the great arteries.

*Other refers to patients with adult congenital heart disease defects not included in the PRospEctiVE study on implaNTable cardIOverter defibrillator therapy and sudden cardiac death in Adults with Congenital Heart Disease risk score model.

1One patient with a congenital defect other than those included in the risk score model had nonfatal ventricular arrhythmia.
The PREVENTION-ACHD risk score model, designed to predict SCD in the general population with ACHD, identifies patients with ACHD at high SCD risk with an HR for SCD or VT/VF of 11.9 in high-risk patients compared with low-risk patients and a C-statistic of 0.75. Upon further dividing the low-risk control group into two subgroups (Figures 2C and 2D), the SCD and VT/VF rates were found to be lowest in the (very) low risk group compared with the intermediate risk group, although not statistically significant. Compared with currently available class 1, 2a, and 2b indications for ICD implantation, the PREVENTION-ACHD risk score model has a greater precision to predict SCD and VT/VF. The KM estimate for SCD or VT/VF was 7% vs 0.6% and for SCD alone 3.5% vs 0.3% in high- vs low-risk patients, respectively. For the consensus statement ICD indications, these estimates were 10.8% vs 0.8% for SCD and VT/VF and 0% vs 0.5% for SCD alone in patients with vs without an ICD indication, respectively. The C-statistics of the risk score model (0.75 and 0.81) exceeded that of the current ICD indications (0.61 and 0.49) when predicting SCD or VT/VF and SCD alone, respectively.

This is relevant, since the ICD indications are widely used to assess ICD eligibility. SCAD in ACHD is a major cause of death, within the population of patients with ACHD. The worldwide use of the PREVENTION-ACHD risk score model may lead to more, justifiable ICD implantations in patients with ACHD and, consequently, prevent ~50% of unnecessary SCDs from occurring. An extensively reported issue with ICD implantation in patients with ACHD is the high rate of inappropriate shocks and ICD-related complications. However, the risk of complications may be acceptable once the risk of SCD is sufficiently predictable. Additionally, a subcutaneous ICD may reduce the abundant, mainly lead-related, complications of transvenous ICD implantation in patients with ACHD.
Comparison with other studies
Several studies have described SCD in ACHD.6–8,13,14,19,20 However, this is the first study in which a risk score model for SCD in ACHD is prospectively validated in a large cohort of general patients with ACHD. With it, we deliver a fully functioning risk stratification tool that is usable in everyday practice.

Perhaps the most comparable example of a risk score to predict SCD in young patients is the Hypertrophic Cardiomyopathy (HCM) Risk-SCD score of the European Society of Cardiology (ESC): HCM Risk-SCD.21 The performance of the PREVENTION-ACHD risk score model for patients with ACHD appears to exceed HCM Risk-SCD, as the HR for SCD in high-risk patients according to our risk score model was 12.5 compared with 2.9 for HCM Risk-SCD; additionally, the C-statistic of our model is 0.75 compared with 0.67–0.70 of HCM Risk-SCD. When externally validating the HCM Risk-SCD, Vriesendorp and coworkers found a similar C-statistic of 0.69.21,22

Potential implications
This study shows that the PREVENTION-ACHD risk score model identifies patients at high risk of SCD or VT/VF, who may benefit from primary prevention ICD implantation. Moreover, it does so more accurately than the currently existing guideline ICD indications. Therefore, this suggests that the PREVENTION-ACHD risk score model may be used instead of, or complementary to, existing indications for ICD implantation. As such, a proportion of SCD victims in the population with ACHD may be prevented.

Limitations
This was a single-center study, with few events during 2 years of follow-up. More events can be expected with increased follow-up duration. However, the study was adequately powered for the primary end point at 2 years and therefore provides accurate data on the prognostic capabilities of the PREVENTION-ACHD risk score model.16 As the risk factors may be dynamic over time, each patient’s risk status should be assessed regularly. Despite the fact that the most common ACHD defects are represented in the PREVENTION-ACHD risk score model, the defects of ~20% of patients were not represented in the risk score model. As previous observations confirmed a very low SCD risk in those patients, for the present analysis those patients were considered to be at low risk; exclusion of those patients from the analysis did not alter the main conclusions, nor the performance of the risk score model.8

Risk factors, including subpulmonary and systemic function, were qualitatively assessed from echocardiograms recorded as part of standard clinical care. Magnetic resonance imaging or gated blood pool scanning could have quantified ventricular ejection fraction more precisely, but were not available in the vast majority of patients. However, the results of this study show that qualitative assessment of impaired ventricular function can be reliably used as a risk factor for SCD or VT/VF.

Follow-up data were acquired from routine follow-up appointments with the patients’ own treating cardiologist. Some low-risk patients (n = 13 [1.7%]) were, consequently, lost to follow-up, because they were not followed up within 2 years. Additionally, some patients had a follow-up of <2 years, because their next appointment was planned outside the follow-up period. Although unlikely, it cannot be excluded that events in these patients were missed.

In 67 patients (9%), ≥1 risk factors were not calculable. Because an analysis with multiple imputations may introduce bias, this was not performed. However, an analysis showed that these missing risk factors did not affect the risk status in all but 1 patient, who was excluded from the outcome analysis.

The class 1, 2a, and 2b indications in the consensus statement are not a risk score.7 Therefore, the risk score model and these ICD recommendations may not be directly comparable as such. However, these recommendations do use an abstract point-based system, with which, given enough risk factors, an ICD is recommended. Therefore, we validated the consensus statement ICD indications in the same manner as the risk score.5

Finally, the PREVENTION-ACHD risk score model is not flawless and may be further improved by additional risk stratifiers such as ECG findings, electrophysiology studies, long-term rhythm monitoring, or cardiac magnetic resonance imaging.4,23 Further development and validation of the PREVENTION-ACHD risk score model is crucial to provide physicians with an optimal tool to be able to implant ICDs in true high-risk patients.

Conclusion
The PREVENTION-ACHD risk score model provides an individualized estimation of SCD or VT/VF risk in patients with ACHD. Compared with currently available guideline ICD indications, it has greater accuracy in assessing SCD or VT/VF risk. Thus, implementation of the PREVENTION-ACHD risk score model in daily clinical practice and ICD implantation in those identified as high-risk patients may decrease the occurrence of SCD in patients with ACHD.

Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrthm.2021.01.009.

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