VOL. 5, NO. 11, 2019
ISSN 2405-500X/$36.00

Letters

RESEARCH CORRESPONDENCE

Sudden Cardiac Arrest in Adults With Congenital Heart Disease

Although most patients with congenital heart disease (CHD) are now reaching adulthood, the risk of sudden cardiac death remains a significant issue (1). Available data arise mostly from cohorts managed in tertiary centers. This study describes the characteristics of sudden cardiac arrest (SCA) in adults with CHD from a large population-based registry.

The Paris-Sudden Death Expertise Center study is a prospective population-based registry in the Great Paris area (population: 6.7 million), collecting all cases of out-of-hospital SCA in individuals ≥18 years of age (2). To ensure appropriate identification of all patients with CHD (including those who died in the field), medical reports were linked with the national French medico-administrative databases, with manual review and validation of diagnoses. Complexity of CHD was determined according to the last American Heart Association/American College of Cardiology classification (3). Comparisons between patients with and those without CHD were performed. To compare survival at hospital discharge, a case-control analysis (4:1) was also performed, after propensity score matching of age, sex, location of SCA, bystander cardiopulmonary resuscitation, and initial rhythm. Patients without known CHD prior to SCA were excluded from this survival analysis to avoid potential immortal-time bias (new CHD diagnosis only possible in survivors). Appropriate institutional review boards approved the investigation plan.

From May 2011 to November 2015, among the 14,049 SCA without obvious noncardiac cause, 17 cases (0.1%) occurred in patients with CHD (44.1 ± 15.6 years old; 11 males [64.7%]). The annual age and sex-standardized incidence from the general population was estimated to be 0.07 (95% confidence interval [CI]: 0.04 to 0.12) per 100,000 person-years. Five patients (29.4%) had simple CHD, 8 (47.1%) had moderate CHD, and 4 (23.5%) had complex CHD (**Table 1**).

Patients with CHD were much younger (age 44.1 ± 15.6 years vs. 70.7 ± 17.0 years; $p < 0.001$), more cases occurred during sports (17.6% vs. 1.1%; $p < 0.001$), and the initial rhythm was more often shockable (52.9% vs. 19.0%; $p = 0.002$). Survival at admission (88.2% vs. 22.3%; $p < 0.001$) and at hospital discharge (35.3% vs. 5.4%; $p < 0.001$) were higher in patients with CHD.

Except for 2 patients with SCA related to an anomalous origin of a coronary artery, all CHD were known prior to SCA. After exclusion of CHD newly diagnosed and matching, the observed difference of survival rate at hospital discharge did not remain statistically significant (26.7% vs. 16.9%; $p = 0.46$) (**Table 1**).

These data from a large population-based registry suggest the following: A) SCA in CHD patients is a relatively rare event at the community level; B) most cases occur in patients with simple or moderate CHD; and C) the survival rate is higher than in non-CHD patients but is comparable after matching.

Contrary to patients from the general population in whom SCA occurs most of the time without previously known heart disease, the CHD was recognized in all except 2 patients. Moreover, although the incidence of SCA is higher in patients with complex defects (1), this highest-risk group of patients eventually represents a minority of the total population with CHD, and the absolute number of SCA cases actually appears more important in patients with less severe phenotypes. These findings may also suggest a more careful follow-up and aggressive management in patients with complex CHD and underline the fact that appropriate long-term aftercare with cardiologists specifically trained in CHD is essential, including in simpler defects.

As recently reported, the present data also revealed a much higher survival in patients with CHD (4). Although this difference appears to be driven by younger age at presentation and other associated favorable prognostic factors, some impact of this specific setting where cardiac disease is known prior to SCA cannot be excluded (likely due to more attention and earlier SCA recognition).

We acknowledge that our population is relatively modest. Studies of SCA in CHD originate mainly from tertiary centers, with a possible overrepresentation of complex CHD. The present registry provides data from a large unselected population.

In conclusion, in this large population-based study, the incidence of SCA in patients with CHD was low in the general population. The apparent

TABLE 1 Characteristics of SCA in CHD and Non-CHD Patients

	CHD (n = 17)*	Non-CHD (n = 14,032)	p Value
Age, yrs	44.1 ± 15.6	70.7 ± 17.0	<0.001
Males	11 (64.7)	8,595 (61.3)	0.775
Public locations	5 (29.4)	2,670 (19.1)	0.349
Sport-related	3 (17.6)	150 (1.1)	<0.001
Bystander	12 (70.6)	9,812 (72.2)	0.883
Bystander-administered CPR	9 (52.9)	5,117 (38.3)	0.222
AED use	0 (0.0)	148 (1.1)	1.00
Time from collapse to basic life support, min	3.0 (0.0–7.5)	5 .0 (0.0–12.0)	0.258
Time from EMS call to EMS arrival, min	9.0 (6.3–12.5)	9.0 (7.0–12.0)	0.971
Initial shockable rhythm	9 (52.9)	2,335 (19.0)	0.002
Epinephrine use	14 (82.4)	6,031 (50.0)	0.008
Admitted alive at hospital	15 (88.2)	3,127 (22.3)	<0.001
Survival at hospital discharge	6 (35.3)	758 (5.4)	<0.001

Values are mean ± SD, n (%), or median (interquartile range). *Five patients had simple CHD (4 had isolated atrial septal defects, and 1 had isolated ventricular septal defect); 8 patients had moderate CHD (3 had an anomalous aortic origin of a coronary artery, 2 had tetralogy of Fallot, 2 had congenital aortic disease, and 1 had an Ebstein anomaly); 4 patients had complex CHD (1 had a double outlet right ventricle, and 3 patients had univentricular hearts [2 patients with total cavopulmonary derivation for: 1) pulmonary atresia with intact ventricular septum; and 2) large ventricular septal defect, 1 patient with congenitally corrected transposition of the great arteries, large ventricular septal defect, and pulmonary atresia palliated with systemic-pulmonary anastomoses].

AED = automated external defibrillator; CHD = congenital heart disease; CPR = cardiopulmonary resuscitation; EMS = emergency medical services; SCA = sudden cardiac arrest.

better survival in CHD patients mostly reflects individuals at a young age and other favorable factors at presentation. Results also underline the fact that efforts to improve risk stratification should not be restricted to complex CHD.

*Victor Waldmann, MD, MPH†
Magalie Ladouceur, MD, PhD†
Wulfran Bougouin, MD, PhD
Nicolas Combes, MD
Alice Maltret, MD
Florence Dumas, MD, PhD
Laurence Iserin, MD
Alain Cariou, MD, PhD
Eloi Marijon, MD, PhD
*Xavier Jouven, MD, PhD

*Université de Paris, INSERM, Paris Cardiovascular Research Centre, European Georges Pompidou Hospital
20-40 Rue Leblanc
75908 Paris Cedex 15
France
E-mail: victor.waldmann@gmail.com

https://doi.org/10.1016/j.jacep.2019.07.017

Please note: †Drs. Waldmann and Ladouceur contributed equally to this work. The SDEC Executive Committee is part of the ESCAPE-NET project, a €10 million funding from the European Commission (Horizon2020 program) to develop research on sudden cardiac death in Europe. Paris-SDEC 2018 Investigators Listing (Online Appendix). The Paris-SDEC activities are supported by the Institut National de la Santé et de la Recherche Médicale (INSERM), University of Paris, Assistance Publique-Hôpitaux de Paris, Fondation Coeur et Artères, Global Heart Watch, Fédération Française de Cardiologie, Société Française de Cardiologie, Fondation Recherche Medicale, as well as unrestricted grants from industrial partners (Medtronic, St Jude Medical, Boston Scientific, MicroPort and Biotronik). The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Clinical Electrophysiology* author instructions page.

REFERENCES

1. Sanatani S, Cunningham T, Khairy P, Cohen MI, Hamilton RM, Ackerman MJ. The Current State and Future Potential of Pediatric and Congenital Electrophysiology. J Am Coll Cardiol EP 2017;3:195–206.

2. Waldmann V, Karam N, Bougouin W, et al. Burden of coronary artery disease as a cause of sudden cardiac arrest in the young. J Am Coll Cardiol 2019;73:2118–20.

3. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol 2019;73:2361.

4. Vehmeijer JT, Hulleman M, Kuijpers JM, et al. Resuscitation for out-of-hospital cardiac arrest in adults with congenital heart disease. Int J Cardiol 2019;278:70–5.

APPENDIX For a list of the Paris-SDEC Investigators, please see the online version of this paper.

Epicardial Connection

The Achilles Heel of Gap Mapping After Wide Antral Pulmonary Veins Isolation

We congratulate Martins et al. (1) for their comprehensive work on mapping techniques for localization of residual gap conduction after wide antral pulmonary veins isolation (PVI). We agree with the authors that both pacing maneuvers and high-density mapping are of great value in localizing a gap along the PVI line. Nevertheless, we believe that if during sinus rhythm (SR) or atrial pacing the pulmonary vein (PV) activation sequence shows the earliest breakthrough distally inside the isolation line, instead of a gap on the line, a direct epicardial connection has to be suspected. At this point, a better understanding of the anatomic and functional substrate is needed. In a histological study on human hearts, Cabrera et al. (2) described interpulmonary myocardial connections in 83% of examined cases. Strands of atrial myocardium crossing the interpulmonary isthmus at the venoatrial junction, the so-called carina region, were found in 53% of the left and 33% of the right PVs. The authors defined the region of the interpulmonary isthmus as a common site for electrical breakthrough. Yoshida