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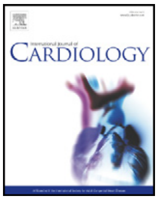

Mortality in adult congenital heart disease: Are national registries reliable for cause of death?

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ABSTRACT

Background: Statistics on cause-specific mortality are important for prognostic research. The aim of this study was to assess the utility of the national mortality registry in research on causes of death in adult patients with congenital heart disease (CHD).
Methods: The CONCOR registry of over 10,000 adults with CHD was used to verify the causes of death provided by the WHO guidelines based national mortality registry, by linkage.
Results: Of 7277 patients linked to the national mortality registry, 196 (2.4%) were recorded deceased, versus 228 deceased patients (3.1%) recorded in the CONCOR registry, during a follow-up of 25,900 patient years. Median age at death was 48.9 years. Of all deaths in the CONCOR registry, 77% had a cardiovascular origin; nearly 50% were due to progressive heart failure and arrhythmias. The national mortality registry recorded death due to progressive heart failure and arrhythmias in only 8.5%. Moreover, this registry recorded death with an 'unspecified' cause in approximately 30%, primarily containing patients who died due to progressive heart failure and arrhythmias according to their medical records.
Conclusion: WHO guidelines based national mortality registries lack the specificity and completeness needed for accurate research on causes of death in adult patients with CHD.

1. Introduction

An increasing number of patients with congenital heart disease (CHD) survive childhood due to improvements in paediatric cardiology, in cardiac surgery and due to thorough monitoring. However, increased survival is accompanied by high morbidity, mainly due to the congenital heart defect itself, and complications from interventions and residual lesions [1,2]. Furthermore, adult patients with CHD are at great risk for premature death [3]. However, little is known about the causes of death in these patients. Better insight into the mechanisms of death in adults with CHD may help improve their treatment and survival.

Mortality data can be obtained from different sources, including hospital records and national mortality statistics. The latter is a frequently used source and acquires information on deaths and causes of death from death certificates. Reliable mortality data require specific and consistent coding of causes on death certificates. Death certificates are filled out by physicians who were often not the primary caretakers. To enhance reliability, the cause of death diagnosis is centrally coded according to the 10th revision of the International Classification of Diseases (ICD), drawn up by the World Health Organisation (WHO) [4]. Regarding the cause of death diagnosis, the WHO recommends avoiding ´modes of death´ as sole cause of death on the death certificate; it is only allowed with another ´underlying´ cause of death. Examples of ´modes of death´ are cardiac arrest, cardiac failure, heart failure and ventricular failure. National mortality registries follow these coding guidelines from the WHO.

Nevertheless, the accuracy of mortality statistics obtained from death certificates has been criticized. Inaccuracies arise from various sources, such as incomplete clinical information regarding the

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circumstances of death, diagnostic difficulties, multiple problem cases, and errors in data processing [5–10].

It is unknown whether adequate clinical research on causes of death can be performed among adults with CHD using national mortality statistics, particularly since sudden death and heart failure are two important causes [1,3,11,12]. In the present study, we have used the Dutch CONCOR registry of over 10,000 adults with CHD to verify the causes of death provided by the Dutch Central Bureau of Statistics.

2. Materials and methods

2.1. CONCOR registry

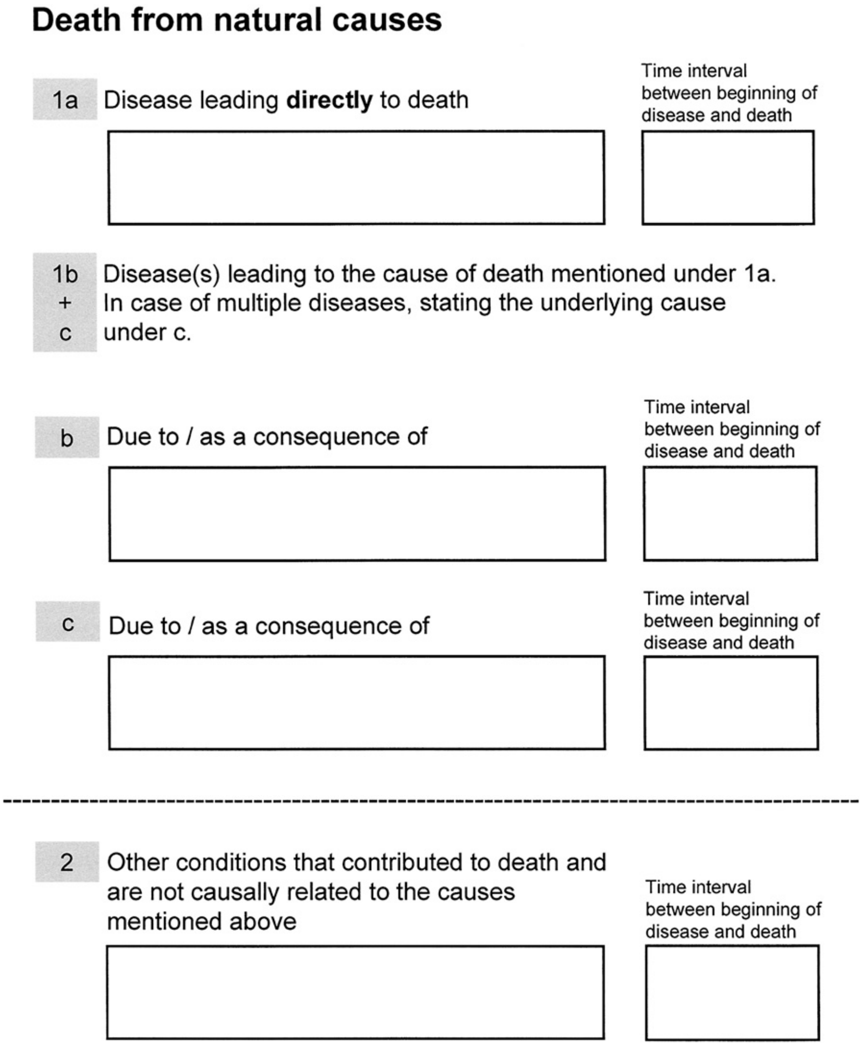
The CONCOR (CONgenital CORvitia) Dutch national registry and DNA bank of adult patients with congenital heart disease has been described in detail [13]. Briefly, CONCOR aims to facilitate research into the aetiology of CHD and on its outcome. Between November 2001 and January 2009, over 10,000 patients with CHD aged 18 years or older have been included through their treating cardiologist or via response to advertisements in local media. Clinical data such as diagnosis, clinical events and procedures — classified using the European Paediatric Cardiac Code Short List coding scheme [14] — as well as patient and family history were obtained. In patients with multiple diagnoses, a pre-specified hierarchical scheme founded on consensus-based classification of severity of diagnoses [15] was used to classify the diagnosis with the worst prognosis as main diagnosis. After entry, data on major cardiac events prior to entry and during follow-up (including death) were systematically recorded. Currently, 102 hospitals are participating, including all eight tertiary referral centres from which approximately 77% of included patients originate.

From CONCOR, date of birth, inclusion date, gender, and main congenital heart diagnosis were derived of the deceased patients recorded from January 1, 2002, to January 1, 2008. The severity of main diagnosis was categorized as mild, moderate or severe, using a consensus-based classification scheme [15]. The number of complications and interventions (surgical and percutaneous) in the medical history was obtained as well [16]. For the purpose of this study, additional data on medication use and the last known New York Heart Association (NYHA) functional class were derived from medical records, and finally the cause of death was recorded. Cause of death was determined based on chart review including available autopsy reports. Where needed, treating cardiologists and general practitioners were contacted for additional information. When there was any confusion about the cause of death, an expert panel (including 4 Grown Up Congenital Heart disease (GUCH) experts) reviewed all the available information from the medical records and assigned the cause of death after deliberation and voting. When there was a tie cause of death was stated as due to multiple causes.

2.2. National mortality registry

The second source for mortality data was the national mortality registry. By linkage of the CONCOR database (n = 7277) to the national Dutch mortality registry of the Central Bureau of Statistics (http://www.cbs.nl/), mortality data were obtained from January 1, 2002, to January 1, 2008, as recorded on the death certificate, coded by a physician according to the 10th revision of the International Classification of Diseases (ICD-10) [4]. Using a combination of zip code, gender, and date of birth, the vital status of 95% of patients was assessed; 344 patients (5%) could not be linked due to missing or erroneously registered zip codes.

The national mortality registry provides maximal 4 causes of death per subject; one primary and maximal three secondary causes, based on the death certificate (Fig. 1). In case of multiple causes of death in one subject, the most appropriate and plausible cause was chosen by our expert panel (independently from the medical records) to



Death from natural causes

1a Disease leading **directly** to death

Time interval between beginning of disease and death

1b + c Disease(s) leading to the cause of death mentioned under 1a. In case of multiple diseases, stating the underlying cause under c.

b Due to / as a consequence of

Time interval between beginning of disease and death

c Due to / as a consequence of

Time interval between beginning of disease and death

2 Other conditions that contributed to death and are not causally related to the causes mentioned above

Time interval between beginning of disease and death

Fig. 1. Part of the medical section of the death certificate.