Patients with simple congenital heart disease (CHD), such as patent ductus arteriosus (PDA), isolated atrial septal defects (ASDs) and ventricular septal defect (VSD), either with normal pulmonary vascular resistance or mild pulmonary stenosis, are not so rare in the general population (1). In particular, these patients could present several clinical problems which are often misdiagnosed by pediatric physicians and may involve, during the life-span, different adult health professionals, such as general cardiologists or internists (1). For these reasons, large nationwide registries on CHD are of paramount importance in establishing the real role of simple CHD in the later life of these subjects (2). In a recent nationwide follow-up study performed by Videbæk et al. on a Danish cohort, a 2-fold higher risk of death was found in patients with simple CHD. In particular, the study was based on 1,241 patients with simple CHD diagnosed between the 1963 and 1973 in otherwise healthy children alive at 15 years of age (3). Patients with a combination of simple CHD and any childhood comorbidity, such as genetic syndromes, mental, kidney, gastrointestinal and pulmonary disease were excluded. At the end of follow-up, in 2012 the prevalence of alive patients with simple CHD was 1.3 per 1,000 Danish inhabitants. Authors found, both at enrolment and at the end of the follow-up, a lower prevalence of simple CHD compared to previous studies, probably because patients with comorbidities were excluded from the analysis. Simple CHDs were defined on the basis of the diagnostic criteria used in 1960s and 1970s. At that time, patients with CHD were mainly diagnosed from clinical symptoms, auscultation, chest X-ray and cardiac catheterization. In particular, the latter invasive examination was performed in the presence of a significant shunt, elevated pulmonary arterial pressure or pulmonary valve stenosis. Obviously, despite the results of Videbæk et al. (3) could not be applied in current medical practice, the question about the need of systematic long-term follow-up programs in patients with simple CHD remains an active problem. Nowadays, the increasing aging of the general population make CHD more likely to be found also in aged patients, especially the diseases at low mortality during childhood. In this perspective, new imaging techniques and endovascular treatments seem to be useful in the management of CHD in these groups of patients (4). Videbæk et al. attributed the higher mortality rate in patients with simple CHD compared to the general Danish population, to sudden death [adjusted odds ratio (aOR), 4.3; 95% confidence interval (CI), 2.9–6.5] (3). Despite simple CHDs were more frequent in women, as reported in others studies, an equal mortality risk was found between genders (3,5). Intriguingly, mortality was
increased, compared with the general population, both overall [adjusted hazard ratio (aHR), 1.9; 95% CI, 1.5–2.4] and in patients (79%) without medical follow-up (aHR, 1.7; 95% CI, 1.3–2.2). Similarly, no difference in morbidity between patients operated or unoperated in childhood (aHR, 5.5; 95% CI, 3.7–8.4 and aHR, 5.8; 95% CI, 4.6–7.5). These results are probably influenced by surgical techniques adopted at that time. Moreover, the incidence of critical cardiac comorbidity was 3.9 per 1,000 patient-years. Adult (re)operation, hospitalization for heart failure (HF) or ventricular tachyarrhythmia (VT) were the most frequent events (3). Similar results were proposed by Lin et al., which reported, after a median follow-up of 11 years, an increased risk of life-long cardiovascular major adverse cardiovascular events (MACE), including HF, stroke, acute coronary syndrome (ACS), and malignant dysrhythmia in patients with CHD (6). As already described, the risk of sudden death in patients with simple CHD is mainly caused by arrhythmias (7-9). In particular, VT seems to be the main cause of sudden death in simple CHD patients, especially in those subjects with previous history of supraventricular tachycardia (SVT), prolonged QRS duration and depressed left- and/or right-ventricular function (7,10,11). These results were in accordance with the Euro Heart Survey, which reported a similar prevalence of VT in patients with simple CHD (12). Considering both the results of Videbæk et al. (3) and others similar investigations, it is clear that one of the aims of large registry based studies on CHD is to emphasize the need for real long-term follow-up in these patients. In order to improve our knowledge about CHD, we must be able to follow our patients over 30 to 50 years, even if they have simple CHD. Obviously, a long-term follow-up is influenced by several difficulties and limitation such as economic constrains, availability of proper facilities and loss of patients. In fact, during the transition from childhood to adulthood, patients may be lost in the transition from the pediatric to adult physicians (13). However, given that current evidences indicating a high risk of cardiovascular disease (CVD) also in patients with simple CHD, is time to consider a long-term follow-up in referral specialized centers? And how long must be the follow-up period? To find answers to these questions, further studies with a long follow-up are needed. The primary aims of these future investigations should be to clarify how long will last the follow-up and which strategies will be good to protect patients from unfavourable events.

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Footnote

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