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INTRODUCTION: Isolated premature ventricular contractions (PVCs) in children are generally regarded as a benign phenomenon. Although PVCs have been shown to correlate with impaired cardiac function in adults, this correlation remains controversial in children. The influence of PVCs on systemic ventricular function is of some interest in congenital heart disease (CHD) patients, as these patients are especially prone to ventricular dysfunction. The aim of this study was to evaluate the influence of PVCs on the systemic ventricular function of patients with CHD during long-term follow-up. METHODS: The database of the Heart Center Leipzig was analyzed retrospectively. Key inclusion criteria were: CHD, age 0 – 21 years, initial systemic ventricular ejection fraction (EF) of > 0.35, follow-up of at least 30 months and at least one Holter ECG every year. Patients were classified into 2 groups in accordance with daily PVC burden (Group A > 1% PVCs/24 hours, Group B < 1% PVCs/24 hours). A subgroup A1 was defined, which presented a PVC burden of > 1% that persisted from the time of inclusion to the last follow-up. RESULTS: 97 consecutive patients were included with a median follow-up of 84 months (range 33-196). Especially patients of subgroup A1 showed a clinically significant decrease in left ventricular ejection fraction (LVEF, p= 0.03). Possible risk factors like coupling interval or the QRS duration of a PVC did not have any influence on systemic ventricular function. CONCLUSION: The current study underlines a negative influence of PVCs on the systemic ventricular function in patients with CHD during long-term follow-up.