Assessment of left ventricular myocardial fibrosis in adult patients with Ebstein's anomaly: a retrospective cohort study based on cardiac magnetic resonance and histopathological samples

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Background: The relationship between Ebstein's Anomaly (EA) and myocardial fibrosis in the left ventricle (LV) has been debated. This study aimed to determine the prevalence of replacement fibrosis in the LV using cardiac magnetic resonance (CMR) imaging and correlate it with histopathological findings. The study also investigated whether LV fibrosis is an independent risk factor for cardiovascular disease (CVD) mortality and developed a risk score for this purpose.

Methods: We performed a twelve-years (2009-2021) retrospective cohort study of adult patients with EA who underwent CMR. The CMR evaluation included a comprehensive assessment of myocardial fibrosis by late gadolinium enhancement (LGE). Four post-mortem samples were obtained from our cohort and stained using Masson trichrome to characterize LV-fibrosis. We used Cox-Regression analysis to identify and derive a prediction score that associates LV-fibrosis with CVD-mortality.

Results: We included 57 adults with EA (52% male, median age 29.52, IQR: 21.24-39.17 years). LGE prevalence by CMR was observed in 52.6% (95% CI: 39.9%-66.0%) in any chamber; LGE-LV in 29.8% (95% CI: 19.5%-42.7%). Histopathological findings revealed a mid-wall pattern with predominantly interstitial fibrosis and minimal replacement fibrosis. LV-fibrosis was associated with increased risk for CVD mortality (HR: 6.02, 95% CI: 1.22-19.91) attributable to lateral and mid-wall LV segment involvement. Our mortality score achieved an overall good prediction capacity (C-statistic: 0.93, Dxy = 0.86).

Conclusions: There is a high prevalence of replacement LV fibrosis in adults with EA, characterized by specific CMR and histological patterns. Furthermore, LV fibrosis is an independent predictor of CVD mortality, which could be integrated into a risk assessment in clinical management.

Figure 1. CMR and pathological images
Figure 1. Kaplan-Meier analysis