Background: Repair of tetralogy of Fallot (TOF) and transposition of the great arteries (TGA), the most common congenital cardiac heart defects, often results in right ventricular (RV) volume and/or pressure overload. RV failure often ensues over time. Myocardial fibrosis may be an important correlate of RV dysfunction and adverse RV remodeling. Delayed contrast enhancement cardiac MRI (DEMRI) is known to be a sensitive technique for determining the presence, location and extent of myocardial fibrosis in vivo. The purpose of this study was to correlate the presence, location and extent of myocardial fibrosis on DEMRI with RV volume and functions, as well as QRS duration in patients with repaired TOF or TGA.

Methods: Twenty seven patients with TOF or TGA were prospectively identified and underwent cine and DEMRI. Measurements of RV morphology and function were assessed on the cine images, and the extent of hyperenhancement (HE) was assessed on delayed enhancement images. QRS duration was measured from the standard electrocardiogram.

Results: All patients, regardless of RV size and function, had evidence of some myocardial fibrosis as determined by the presence of HE. However, there was a predilection of myocardial fibrosis for specific locations. In particular, the junction of the RV freewall with the interventricular septum was commonly involved (100% at the inferior septal junction, 56% at the anterior septal junction). Six patients (22%) had direct involvement of the right ventricular free wall. The extent of HE correlated with RVEDV, RVESV, and RVEF (p<0.01). In addition, QRS duration correlated with RVEDV, RVEF, and RVEF (p<0.05, all).

Conclusion: Patients with repaired congenital cardiac heart defects such as TOF and TGA have areas of myocardial fibrosis that are located remote from sites of direct surgical intervention. The concentration of myocardial fibrosis in these unexpected locations remains to be explained. The extent of fibrosis in these patients and QRS duration correlates with adverse right ventricular morphology and function. Future investigations of the relationship between right ventricular fibrosis characteristics and outcomes in this patient population are necessary.