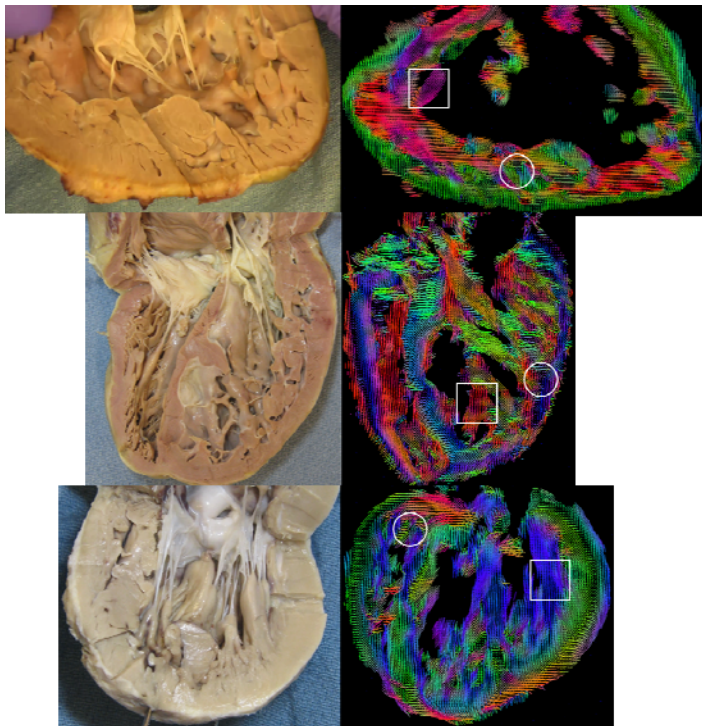


MP2-12

Myofiber organization in the failing systemic right ventricle of congenital heart diseases.

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Basis: Heart failure and arrhythmia are common complications in patients with a systemic right ventricle. Magnetic resonance imaging (MRI) studies have shown microstructural alterations in left ventricular myocardium of hearts affected by genetic and acquired cardiac diseases. In this work, we tested the hypothesis that myofiber architecture is abnormal in the systemic RV of explanted, failing hearts affected by congenital diseases. **Methods:** We used diffusion-weighted magnetic resonance imaging (DW-MRI) (3T Siemens Skyra) to examine 3 failing hearts explanted at transplantation from young patients (5-19 years) with a systemic right ventricle and a pulmonary left ventricle (1 case). DW MRI was acquired with a pulsed gradient spin echo sequence, single shot EPI readout and 1.5x1.5x1.5mm spatial resolution. Our gradient scheme contained 4 $b=0$ s/mm² images and 3 shells ($b=1000, 1500, 2000$ s/mm²) of 30 gradients each. Nine averages were acquired with both AP and PA phase encoding directions. Diffusion compartment imaging was computed to separate the free diffusive component representing free water from an anisotropic component characterizing the orientation and diffusion characteristics of myofibers. The orientation of each anisotropic compartment was displayed in glyph format and used for qualitative description of myofibers and for construction of tractograms. Blocks of ventricular myocardium were removed for comparison with diffusion imaging. **Results:** The hypertrophied systemic RV has an endocardial layer, comprising about 2/3 of the wall thickness, composed of hypertrophied trabeculae and an epicardial layer of circumferential myofibers somewhat like normal (Fig). Smaller trabeculae are organized with parallel fibers while larger, composite bundles show myofiber disarray, largely between component trabeculae. The apical whorl is disrupted and we observed myocardial whorls or vortices and abrupt fiber tract interruptions in multiple regions away from the apex. Histological exam of tissue blocks removed after imaging confirmed the diffusion imaging findings. **Conclusion:** Myofiber organization is abnormal in the failing systemic right ventricle and might be an important substrate for heart failure and arrhythmia. It is unclear if the abnormal myofiber organization is due to hemodynamic factors or intrinsic developmental problems.



HLHS (top), CTGA (middle) and DIRV (bottom). Small trabeculae with coherent fibers (boxes) and vortices (circles). Note marked disarray between trabeculae.