

Failing Right Ventricle in Hypoplastic Left Heart Syndrome: myofiber architecture reconstruction by Diffusion-Weighted Magnetic Resonance Imaging

Campanale C. M. (1), Scherrer B. (2), Afacan O. (2), Majeed A. (3), Singh J.(2), Warfield S.(2), Sanders S. P.(2).

Ospedale Pediatrico Bambino Gesù, Roma - Italy (1); Boston Children's Hospital, Boston - USA (2); Children's Hospital of the King's Daughter's, Atlanta - USA (3)

Introduction:

Heart failure is a common cause of death in young adults with hypoplastic left heart syndrome (HLHS). Diffusion-Weighted MR Imaging (DWMRI) studies described myofiber disarray in hypertrophic and dilated cardiomyopathies. Our hypothesis is that myofiber disarray is present in the systemic right ventricle (RV) of hypoplastic left heart syndrome (HLHS).

Methods:

The explanted heart from a 14 yo with HLHS was fixed and imaged using a 3T Diffusion MR scanner. Diffusion Compartment Imaging (DCI) was computed and data displayed in glyph format for qualitative description of myofibers. Real-time Tractography was performed for 3D reconstruction. Histology sections were taken after imaging for comparison with diffusion data.

Results:

The failing systemic RV in HLHS is markedly hypertrophied, dilated and bizarrely shaped (Fig A). The epicardial layer around most of the RV is formed by a homogeneous stratum of circumferential myofibers, while the endocardial layer is remarkably disarrayed (Fig B). Some smaller muscle bundles are organized with parallel fibers (Figure C1-C3) while larger bundles, which appear to be composites, show myofiber disarray, largely between component bundles (Fig D). The apical whorl is disrupted with chaotic arrangement of fibers (Fig E). In addition, we observed myocardial whorls or vortices and abrupt fiber tract interruptions (Figure F) within the wall.

Discussion:

For the first time myofiber disarray in the RV of an HLHS has been demonstrated by DW-MRI. Myofibers and myocardial bundles arrayed at large angles result in inefficient contraction and ventricular dysfunction, as was present in this patient. These findings support our hypothesis that disordered myocardium contributes to heart failure in these patients. Further, myocardial whorls could be the substrate for arrhythmia, another frequent cause of death in young adults with congenital heart disease. Our findings support further study of myocardial architecture in congenital heart defects.

