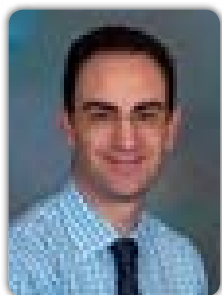


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14-3-3epsilon controls multiple developmental processes in the mouse heart

14-3-3ε plays an important role in the maturation of the compact ventricular myocardium by modulating the cardiomyocyte cell cycle via p27kip1. However, additional cardiac defects are possible given the ubiquitous expression pattern of this protein. Germ line deletion of 14-3-3ε led to malalignment of both the outflow tract (OFT) and atrioventricular (AV) cushions, with resulting tricuspid stenosis and atresia, mitral valve abnormalities, and perimembranous ventricular septal defects (VSDs). We confirmed myocardial non-compaction and detected a spongy septum with muscular VSDs and blebbing of the epicardium. These defects were associated with abnormal patterning of p27kip1 expression in the subendocardial and possibly the epicardial cell populations. In addition to abnormal pharyngeal arch artery patterning, we found deep endocardial recesses and paucity of intramyocardial coronary vasculature as a result of defective coronary plexus remodeling. The malalignment of both endocardial cushions provides a new explanation for tricuspid and mitral valve defects, while myocardial non-compaction provides the basis for the abnormal coronary vasculature patterning. These abnormalities might arise from p27kip1 dysregulation and a resulting defect in epithelial-to-mesenchymal transformation. These data suggest that 14-3-3ε, in addition to left ventricular non-compaction, might be linked to different forms of congenital heart disease.

Biography

Luca Brunelli, MD, PhD has completed his MD at the University of Genoa Medical School at the age of 25 years and his PhD at the University of Turin at the age of 39. He is an Associate Professor of Pediatrics, and Genetics, Cell Biology and Anatomy at the University of Nebraska Medical Center, and the Division Chief of Neonatology at the University of Nebraska Medical Center and Children's Hospital & Medical Center in Omaha, Nebraska. He has published more than 20 papers in reputed journals, including Circulation Research, Molecular and Cellular Biology, and Nature Methods.

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