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Commentary

Impact of Complex Congenital Cardiac Malformations

Jesse Elliot*

Department of Physiology, University of Oxford, Oxford, United Kingdom

DESCRIPTION

The current study provides an overview of univentricular heart nomenclature and classification, epidemiology and pathological subtypes, genetic factors, physiology, clinical features, diagnostic assessment, therapy, and postoperative sequelae. Although the current report discusses issues relevant to neonates and children with univentricular hearts, the emphasis is on information of interest and relevance to adult cardiologists. The current study provides an overview of univentricular heart nomenclature and classification, epidemiology and pathological subtypes, genetic factors, physiology, clinical features, diagnostic assessment, therapy, and postoperative sequelae. Although the current report discusses issues relevant to neonates and children with univentricular hearts, the emphasis is on information of interest and relevance to adult cardiologists.

The nomenclature and classification of the univentricular heart are still debatable. The terms "single ventricle," "cor triloculare biatrium," "cor biloculare," "common ventricle," and "functionally single ventricle" have all been proposed. The debate is sparked in part by observed morphological heterogeneity and the recognition that truly solitary ventricles are extremely rare. It is more common to have a second rudimentary or hypoplastic accessory ventricle, which justifies the term "functional" single ventricle. The Atrio Ventricular (AV) connection is widely recognized as crucial in defining the univentricular heart.

A comprehensive nomenclature system should take into account atriovisceral situs, the relationship between systemic and pulmonary veins, AV valves, great arteries, and ventricular morphology. More specifically, the well-developed ventricle can be classified as left, right, or indeterminate. The left ventricle has relatively smooth walls, fine trabeculations, and no septal chordal attachments of the AV valve. In contrast, right ventricles are more coarsely trabeculated and frequently have chordal attachments of the AV valve to the septal surface. There may be a single, double, or common inlet for the AV connection. A common inlet connection is present if >75% of a common AV valve annulus empties into one ventricular chamber. Mitral or tricuspid valves have a single inlet connection and the morphological AV valve with two inlet connections.

The hypoplastic right ventricle is most commonly anterior and to the left of the left ventricle, with L-transposition of the great arteries was found to have a crude median incidence of 2.3 cases per 10,000 live births, ranging from mild left ventricular hypoplasia (amenable to biventricular repair) to aortic and mitral valve atresia with an absent left ventricle.

Even though the genetic determinants of other types of univentricular heart disease have not been thoroughly investigated, several subtypes, including DILV, single inlet, common inlet, and complex single ventricle heterotaxy syndromes, are thought to be polygenic in nature, with recurrence and transmission risks far lower than those expected from mendelian inheritance. The phenotype is assumed to be the result of additive effects of multiple genes, interactions with other genes and environmental factors, and stochastic effects in the polygenic model.

The univentricular heart's physiology is determined by key factors such as obstruction to outflow, inflow, and/or flow across the atrial septum; systemic and pulmonary venous return; pulmonary vascular resistance; and AV valve regurgitation. The neonate is dependent on right-to-left shunting through a patent ductus arteriosus to maintain systemic output when there is severe systemic outflow obstruction. Blood mixes within the atrial and ventricular chambers and is ejected primarily through the pulmonary valve to the pulmonary and systemic vascular beds *via* the pulmonary artery and patent ductus arteriosus in a ratio dependent on vascular resistances. In contrast, pulmonary blood flow is dependent on left-to-right shunting across a patent ductus arteriosus when critical pulmonary outflow obstruction exists. Again, systemic and pulmonary blood flows are interdependent and determined by resistances in the circulatory system. Although the majority of patients will be managed using a staged surgical approach in preparation for a fontan procedure, a minority will not receive fontan palliation because they have reasonably balanced systemic and pulmonary circulations or because of unfavourable hemodynamics. Adults with a univentricular heart require a thorough understanding of diverse anatomic variants, hazy cardiovascular physiology, multisystemic consequences of cyanotic heart disease, and postoperative sequelae in surgically palliated patients.

Correspondence to: Jesse Elliot, Department of Physiology, University of Oxford, Oxford, United Kingdom, E-mail: jesseE@edu.uk

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