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The functional single ventricle: how imaging guides treatment

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ABSTRACT

Functional single ventricle (FSV) encompasses a spectrum of severe congenital heart disease. Patients with FSV are living longer than decades prior resulting in more frequent imaging both for surgical planning and functional evaluation. At each stage of surgical intervention, imaging plays a critical role in detecting postoperative complications and preprocedural planning. This article describes the unique imaging findings, including complications, that are most important to the referring physician or surgeon at each surgical stage of FSV management. A description of lesions that embody the diagnosis of FSV is also included.

1. Introduction

Functional single ventricle (FSV) is a spectrum of severe congenital heart disease, with multiple anatomic variations but similar surgical treatment strategies. FSV is anatomically defined as (1) connection of both atria to the same ventricle (2:1 connection) or as (2) connection of both to atria separate ventricles, one of which is hypoplastic (1:1 connection) [1,2]. With the advent of advanced palliative and corrective surgical procedures, FSV patients are living longer into adulthood compared to two or three decades ago [3], and they are more frequently undergoing imaging to assist in clinical and surgical management. However, interpreting an imaging examination of an FSV patient is a daunting task, not only because of unusual anatomy and varied postoperative appearances but also because of the rarity of the FSV that makes it difficult for the imaging specialist to maintain diagnostic proficiency.

Surgical palliation of FSV began in 1971 with the treatment of tricuspid atresia (TA), described by Fontan and Baudet [4]. These surgical palliative techniques were then further refined in 1980 by Norwood [5,6], specifically in patients with hypoplastic left heart. Subsequently, many variations of surgical palliation have been built upon these original techniques [4] enabling neonates, who previously would have suffered significant morbidity and mortality, to have a 70% chance of survival into adulthood [3].

The goal of surgical palliation is to maximize systemic blood oxygenation by converting blood flow from a parallel to a serial circuit and to maintain the functional capacity of the systemic ventricle. This is achieved by one of the four common surgical strategies: (1) staged surgical palliation, (2) hybrid palliation that combines surgical and percutaneous interventions, (3) corrective biventricular repair, or (4) orthotopic heart transplantation.

The goal of this article is to enable the imaging specialist to successfully navigate the unusual anatomy of FSV and to understand the role of imaging in the most common operative and percutaneous procedures utilized in treatment. The article presents the anatomic lesions that are included in the diagnosis of FSV. It discusses the techniques and utility of different imaging modalities and for each surgical stage, the postoperative complications, and the information required to plan or meet criteria for the next surgery.

2. Single ventricle lesions

FSV has many anatomic presentations, which have been grouped into a number of categories [1,2,7]. Identifying ventricular morphology is the first step in assigning a patient to one of these categories.

The dominant ventricle is defined as the ventricle that is not hypoplastic and receives inflow from one atrium or both atria. The left ventricle can be distinguished by a smooth septal surface without papillary muscle or chordal attachments. Additionally in the left ventricle, the annuli of the inflow and outflow valves are in fibrous continuity. In contrast, the right ventricle is more trabeculated, usually with an identifiable moderator band. As opposed to the left ventricle, in the right ventricle, the wall has chordal attachments that connect to the septal surface, and the right ventricle contains a muscular infundibulum that separates the inflow and outflow valves. It is important to remember that neither the position of the heart in the chest nor the relationship of the great arteries or atria to the ventricle(s) can be used to distinguish the ventricles because these chambers can be inverted and the atrioventricular connections can be discordant [8]. Patients can

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also present with situs abnormalities. In some patients, the sidedness of the dominant ventricle cannot be determined due to ambiguous morphology [9].

2.1. Dominant left ventricle

2.1.1. Double inlet left ventricle (DILV)

The left ventricle receives inflow from both of the atrioventricular valves (AVVs) and communicates with the right ventricle via a ventricular septal defect (VSD), Fig. 1a and b. The right ventricle is usually hypoplastic and DILV is often associated with transposition of the great arteries, in which the aorta arises from the hypoplastic morphologic right ventricle [1].

2.1.2. Unbalanced atrioventricular canal

A common AVV is positioned primarily over one ventricle, an anomaly that falls within the broader category of atrioventricular canal/septal defects, Fig. 1c and d. The unbalanced atrioventricular canal represents approximately 10% of atrioventricular canal defects [10]. The ventricle that is partially excluded from the common AVV is often hypoplastic, which can result in single ventricle physiology [11].

2.1.3. Tricuspid Atresia (TA)

TA is characterized by plate-like atresia of the tricuspid valve, which results in a lack of direct communication between the right atrium and right ventricle. Circulation is dependent on an interatrial communication, Fig. 2a and b. Two main types of TA have been described: in the more common type, the tricuspid valve is completely absent, and in the second type, the valve is atretic [12].

2.1.4. Hypoplastic right heart syndrome (HRHS)

HRHS results from underdevelopment of the right heart structures including the right ventricle, pulmonary artery, pulmonic valve, and tricuspid valve, Fig. 2c and d. Each of these structures can be affected to a varying degree. In contrast to the DILV, the right ventricle in HRHS receives blood from the right atrium.

2.2. Dominant right ventricle

2.2.1. Hypoplastic left heart syndrome (HLHS)

HLHS is underdevelopment of the structures of the left heart including the left ventricle, aorta and aortic valve, and mitral valve, Fig. 3. As in HRHS, each structure can be affected to a varying degree. Aortic atresia is often included in the spectrum of HLHS, as it is often associated with a hypoplastic left ventricle.

2.2.2. Double inlet/outlet right ventricle

Double inlet right ventricle (DIRV) occurs when the right ventricle receives inflow from both atria via separate AVVs; there is often a VSD that allows for communication with an often hypoplastic left ventricle,

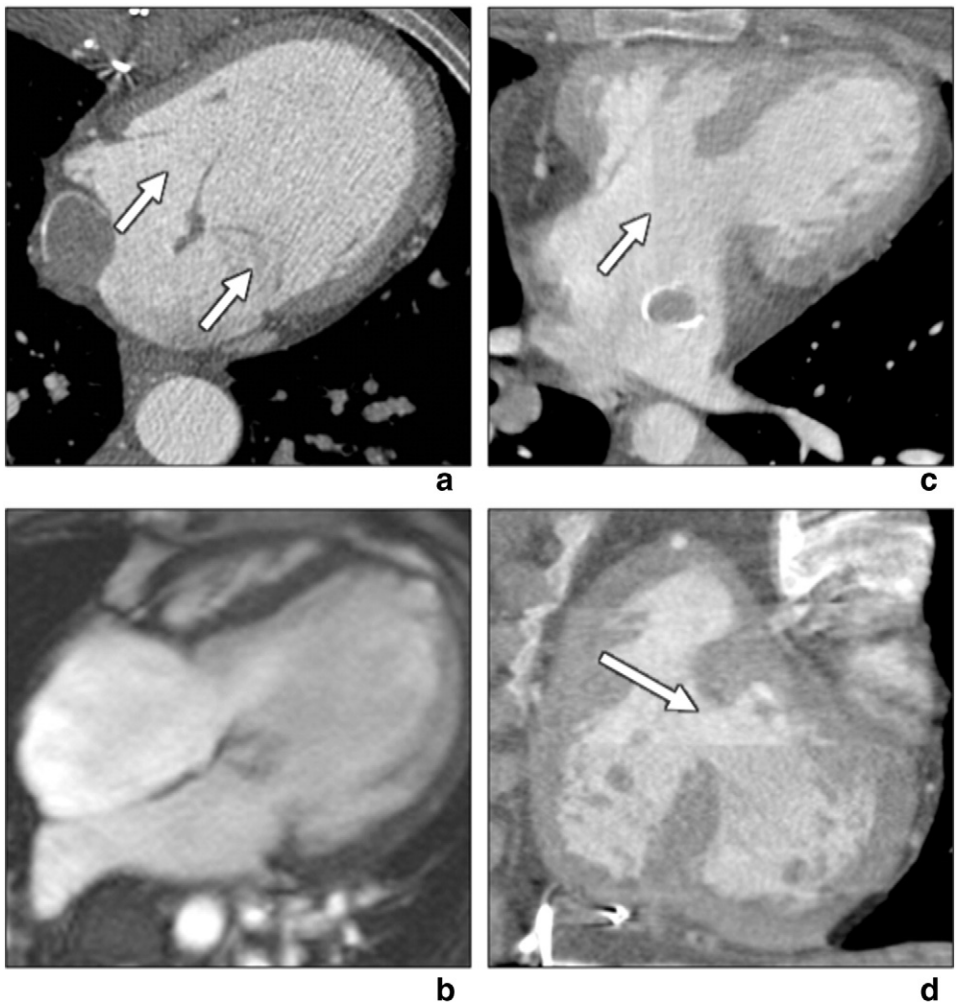

Fig. 1. Single functional ventricle lesions with a dominant left heart. DILV seen on axial CT (a) and four-chamber magnetic resonance (b) images. Both AV canal valves (arrows) are connecting to dominant left ventricle. Note the smooth border of the ventricle, which defines it as an LV. An unbalanced atrioventricular defect (arrow) seen on CT four-chamber (c) and short-axis (d). Images show a dominant left ventricle (*) and large VSD (arrow).