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Pictorial Review

Cross-sectional imaging of the Fontan circuit in adult congenital heart disease

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The Fontan circuit is the result of a palliative surgical procedure that is performed in patients with a functionally single ventricle cardiac anomaly. The success of this operation has resulted in an increasing population of adults with this anatomy and physiology. The purpose of this article is to familiarize the general radiologist with the expected anatomy and cross-sectional imaging findings, highlight special imaging considerations, and examine the common complications that are encountered in this group of patients.

Introduction

A Fontan circulation describes the surgical diversion of all systemic venous blood into the pulmonary arteries without an associated pumping ventricle. Since its conception, the Fontan procedure and its modifications have radically changed palliation of congenital cardiac abnormalities with a single functioning ventricle, in which biventricular repair is not attainable.

The uncorrected single-ventricle system results in a parallel relationship of the systemic and pulmonary circulation. In this instance, survival is only possible due to right-to-left shunting and the mixing of deoxygenated and oxygenated blood, resulting in cyanosis. This is in contrast to a normal physiological biventricular system, in which the systemic and pulmonary circulations operate in series. The major disadvantages of a single-ventricular parallel system are cyanosis with arterial desaturation and chronic volume overload of the single functioning ventricle. The latter, with time, can result in impaired systolic function and congestive cardiac failure. Furthermore, chronic cyanosis and right-to-left shunting are associated with long-term multisystem complications.

The Fontan circulation is a palliative procedure for patients born with cyanotic heart disease primarily due to a functional univentricular circulation. Underlying congenital cardiac abnormalities amenable to a Fontan procedure include tricuspid atresia, hypoplastic left-heart syndrome, and pulmonary atresia with an intact ventricular septum, amongst others.

The principal of the operation is to separate the systemic and pulmonary circulations. Thus, the extra volume load is removed from the single ventricle and cyanosis is abolished. The systemic veins are connected to the pulmonary artery without passing through a ventricle, either via the right atrium, or directly into the pulmonary arteries. The main pulmonary artery is ligated and any interatrial communication is closed (or a small fenestration left).

Functionally there is one power source, the single ventricle, for both circulations and changes in either circulation will impact on ventricular function. The Fontan state is one of chronically low cardiac output with preload starvation and a high afterload. Cardiac output is critically dependent on high systemic venous filling pressures, a low

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pulmonary vascular resistance, and the maintenance of sinus rhythm. Flow within the Fontan circuit is of a very low velocity. The key to a good cardiac output is preservation of ventricular preload and systolic function. Anatomical problems, such as an obstruction within the Fontan circuit, either at conduit level, branch pulmonary artery level, or systemic arterial obstruction (re-coarctation), will lead to diminished functional performance. Thrombus within the circulation is an often fatal complication owing to the low-flow characteristics of the systemic venous flow. The long-term effects of high hepatic venous pressures and the development of chronic liver disease are increasingly recognized.[1]

Modifications in the surgical technique have resulted in increased life expectancy and improved long-term palliation of symptoms. Consequently, current estimates suggest that in the UK there are now more adults than children living with congenital heart disease.[2,3] Radiological follow-up, predominantly with cardiac MRI, has become essential in this group of patients. The purpose of this review is to outline the history, typical anatomy, imaging considerations, and long-term complications of the Fontan procedure. The intention is to familiarize the general radiologist with the expected cross-sectional imaging findings in this group of patients, who will be more frequently encountered in the future.

History and anatomy

The Fontan operation was originally proposed in 1971 by Francois Fontan and Eugene Baudet, as an innovative palliative procedure for tricuspid atresia[4] (Fig 1). In the original procedure, Fontan and Baudet used a classical Glenn shunt, comprising a connection between the superior vena cava (SVC) and distal right pulmonary artery (RPA), with ligation of the SVC below the anastomosis and division of the RPA from the main pulmonary artery bifurcation. In addition to this, a connection between the right atrium and pulmonary artery was formed; the latter interposed with an aortic homograft (Fig 2). Since then, the procedure has undergone major technical modifications, determining the precise path deoxygenated blood takes from the inferior vena cava (IVC) to the pulmonary arteries. More in depth description of the types of congenital defects that require a Fontan procedure, types of Fontan, and management of patients with Fontan circuits can be found in an article by Paul Clift.[5]

Various techniques have been used to isolate and connect the right atrium or right atrial appendage to the pulmonary artery, collectively termed an atriopulmonary Fontan circuit. In this procedure the right atrium is anastomosed directly to the main pulmonary artery, which is divided proximally, obliterating its connection with the right ventricle[6] (Fig 3). Any associated atrial septal defect is closed. SVC and IVC blood passes into the atrium before it reaches the pulmonary arteries, bypassing the right ventricle (Fig 4). This may also be seen in association with a bidirectional Glenn shunt, a direct end-to-side connection between the SVC and RPA, which remains connected to the

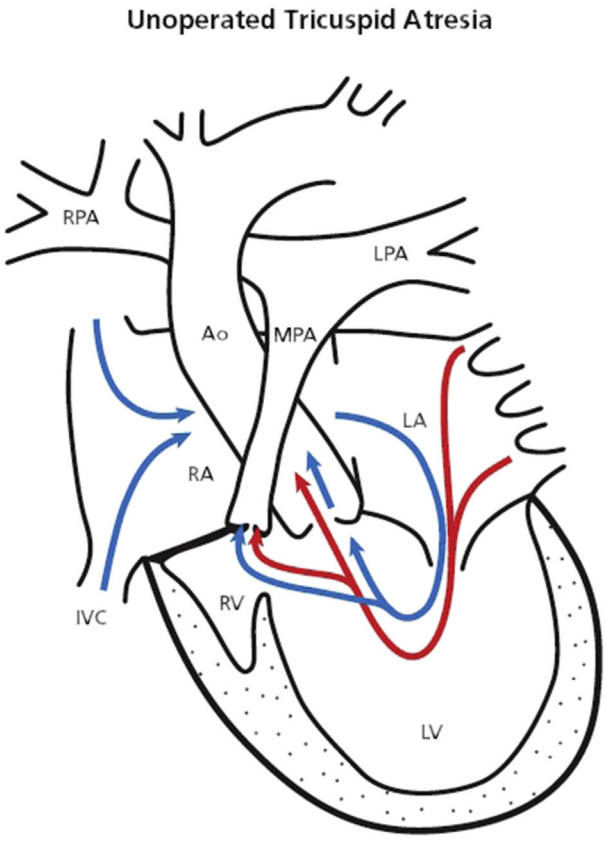

Figure 1 This diagram demonstrates the basic anatomy of tricuspid atresia. This is characterized by agenesis of the tricuspid valve and varying degrees of hypoplasia of the right ventricular inlet. This is almost always associated with an atrial septal defect and ventricular septal defect, resulting in mixed shunting and deoxygenation.

main pulmonary artery bifurcation (Figs 5 and 6). This method was the predominant technique used up to the late 1980s; however, emergent late complications led to modifications of the technique. As a result, the atriopulmonary method is no longer employed in the creation of a Fontan circuit. Nonetheless, this type of Fontan operation is still regularly encountered in adult clinical practice.

Studies performed by deLeval et al.[7] in 1988 demonstrated that reducing the turbulence of flow to the pulmonary arteries carries several benefits, leading to increased pulmonary flow and increased haemodynamic efficiency of the circuit. This led to the development of the lateral tunnel total cavopulmonary connection (TCPC; Fig 7). This comprises a bidirectional Glenn shunt and formation of a tunnel within the right atrium using the posterior atrial wall and a prosthetic patch. The tunnel baffles blood from the IVC to the transected SVC, which is anastomosed to the pulmonary artery. The proximal aspect of the main pulmonary artery is again divided from the right ventricle (Fig 8).

The newest method used in creating a TCPC is the extracardiac conduit (Fig 9), consisting of a prosthetic polytetrafluoroethylene conduit completely external to the right atrium, which connects the transected IVC and RPA (Fig 10).

The Kawashima procedure is a variant of the bidirectional Glenn operation, and is performed for intermediate-