Role of Cardiovascular Computed Tomography Angiography in the Evaluation for Functional Single Ventricle after the Fontan Procedure

Yu Sun, Jie Hou, Junrui Xiao, Chang Liu, Miao Jing, Mingyu Zou and Benqiang Yang*

Department of Radiology, General Hospital of Shenyang Military Command, Shenyang 110001, People’s Republic of China

Corresponding author: Benqiang Yang, Department of Radiology, General Hospital of Shenyang Military Command, Shenyang 110001, People’s Republic of China, E-mail: bqyang888@sina.com

Received date: April 05, 2018; Accepted date: May 04, 2018; Published date: May 12, 2018

Abstract

Introduction: Functional single ventricle (FSV) includes a spectrum of severe congenital heart disease and is associated with high mortality, morbidity and resource usage. The Fontan procedure has been treated as the relatively ideal surgical treatment for these patients, but pathophysiological complications of the Fontan circulation are frequent and the anatomic structures of FSV patients are also complex. Even though echocardiography, CT and MRI can disclose and characterise common postoperative complications in patients with the Fontan palliation, no single imaging method is comprehensive in any given patient. Cardiovascular computed tomography angiography (CCTA), with advanced technologies and markedly decreased radiation dose, can provide better information with lower risk than other modalities. This article aims to assess CCTA modality in the evaluation for FSV with the Fontan palliation.

Materials and methods: Twenty-eight cases of FSV patients after the Fontan procedure were included in this retrospective single-center study. All of patients underwent CCTA examination on the same CT scanner. The diameters of left pulmonary artery, right pulmonary artery and descending aorta (level of diaphragm) were measured for the calculation of McGoon index to indicate the development of pulmonary artery. The qualitative evaluation of anatomic structure and postoperative complication were also completed by CCTA data. Additionally, the data’s of cardiac catheterization for these FSV patients were collected for comparison with that of CCTA.

Results: The diameters of pulmonary artery each was (1.33 ± 0.23, left), (1.38 ± 0.15, right), and the McGoon index calculated was (1.55 ± 0.13). The diameters of both left and right pulmonary arteries related well to the pressure of pulmonary arteries (left: r=−0.865 systole, r=−0.829 diastole, r=−0.867 average; right: r=−0.704 systole, r=−0.758 diastole, r=−0.743 average; P<0.001 respectively). The McGoon index also had good correlation with the resistant index of pulmonary artery (r=−0.887, P<0.001). In addition, 8 cases of Y-graft Fontan, 2 cases of atriopulmonary connection, 8 cases of lateral tunnel Fontan, and 10 cases of extracardiac Fontan procedures were detected from our study. And 2 cases of diffuse pulmonary arteriovenous malformations and 2 cases of aortopulmonary collaterals were defined as complications of the Fontan procedures.

Conclusion: With improved spatial resolution, rapid image acquisition and image post-processing technology, CCTA can supply sufficient information for both detection of anatomic structure and evaluation for FSV patients after the Fontan procedure. It has become an essential modality for congenital heart disease.

Keywords: Cardiovascular computed tomography angiography; Functional single ventricle; Congenital heart disease; Fontan procedure; Cardiac catheterization

Introduction

Functional single ventricle (FSV) is in common usage to describe a spectrum of severe congenital heart disease, with multiple anatomic structural variations but similar surgical operation strategies, including double inlet ventricle, tricuspid atresia, mitral atresia, hypoplastic ventricle syndrome, and unbalanced AV canal defect [1,2]. The Fontan procedure has been treat as a relatively ideal surgical method since its first accomplished for surgical repair of tricuspid atresia [3], and it has undergone many methodological modifications [4]. The Fontan palliation successfully directs systemic venous blood to the pulmonary artery and pulmonary venous blood to the aorta. And due to this corrective operation procedure, many FSV patients are living longer into adulthood than about three decades ago [5]. While the anatomy of FSV is complex and the postoperative pathophysiological complications are frequent, so cardiovascular imaging examination becomes a key aspect of clinical surveillance. However, interpreting imaging examination for FSV patients is a daunting task, as a result of the unusual anatomy and varied postoperative appearances that make it difficult for radiologist to maintain diagnostic proficiency.

When intervention treatment became common for congenital heart disease, cardiac catheterization was treated as the only available modality. And it was replaced by echocardiography for most indications and then most patients with congenital heart disease are managed by echocardiography alone [6]. While several limitations that hinder the application of echocardiography include degradation of acoustic windows with
surgical scarring, poor definition of vascular airway relationship, poor visualization of thoracic vasculature and so on. Cardiac magnetic resonance imaging can overcome these limitations above, but it is incompatible with most current cardiac pacemakers and defibrillators, and artifact from certain metallic coils and implants degrade image quality. Additionally, the relatively long imaging acquisition time and breath-holding required by many sequences also limit its use in certain patients [7,8].

With advanced technologies that allow for both high-quality images and markedly decreased radiation dose, cardiovascular computed tomography angiography (CCTA) has become a standard test modality for most patients. For select indications, CCTA can provide better information with lower risk than other imaging modalities. And among many advanced image post-processing technologies, iterative reconstruction can reduce radiation dose without loss of image quality [9]. As a result, CCTA examination even in the youngest patients with high heart rates can be available for diagnosis [10]. For the patients with congenition heart disease, especially tetralogy of Fallot, functional single ventricle and transposition of great arteries, CCTA may be the most useful modality to understand anatomy, surgical procedures performed and common postoperative complications [10].

The common problems in the Fontan palliation include obstruction of systemic venous and pulmonary arterial flow, semilunar valve dysfunction, aortopulmonary collateral, pulmonary arteriovenous malformation, conduit leak or thrombus in the Fontan pathway [11]. Among various modalities, no single imaging method is comprehensive in any given patient. Therefore, it is of great importance for physicians to recognize the limitations of each modality and circumvent these by application of suitable alternatives. This article is going to enable physicians to diagnose the unusual anatomy of FSV and to understand the role of CCTA in the evaluation of outcome and detection of complication after the Fontan procedure.

Materials and Methods

Study population

This was a retrospective single-centre study. Twenty-eight cases of FSV patients (20 males, 8 females; (22.3 ± 11.7) years, median=21 years, mean=22.3 years, rang from 7 to 59 years) were included in our study. All of FSV patients had undergone the Fontan procedures and the follow-up time after operation was (10.1 ± 9.4) yearsrang from 0.2 to 29 years. Our study got the approval from our institutional review ethics board.

CCTA image acquisition

The CCTA examinations were performed on the same 256-slice CT scanner (Brilliance iCT, Philips Medical Systems Inc, 595 Miner Road, Cleveland, Ohio 44143, USA). CCTA imagerings (tube voltage, 80-120 KV; tube current, 80-200 mAs; collimated detector, 128 x 0.625 mm, section thickness, 0.9 mm; reconstructed interval distance, 0.45 mm; gantry rotation time, 0.27 s; scan time, 2.5 s) were acquired with prospective ECG-gating technology (65%-75% of R-R interval). Infant underwent examination under eupnic situation and other patients were breath-hold scanning. Ionizing radiation protection was implemented by using lead collar and lead clothing. The contrast material used was iohexol (Omnipaque300; Nycomed Amersham, Oslo, Norway; 300 mg of iodine per milliliter) at a standard dose of 0.8 ml/kg and injection rate of 5 ml/s by high-pressure injector (MissouriTM, Ulrich Medical, USA), followed by 40 mL of saline. The contrast agent was given approximately half before scan and the rest of contrast is given as the acquisition timed to aortic opacification. All patients underwent CT scan with informed consent.

CCTA image analysis

All of CCTA datas were post-processed in GE AW 4.4 workstation. Two experienced radiologists (J.R.X, B.Q.Y) together analyzed imaging within 30 min per patient. Available image post-processing technologies consist of multiplanar reformation, maximum intensity projection, curved planar reformation, and volume rendering technique. The diameters of the left and right pulmonary artery and descending aorta (level of diaphragm) were measured. The McGoon index was calculated to assess development of pulmonary artery by following equation: McGoon ratio=(left+right pulmonary artery diameter)/the diameter of descending aortic (level of diaphragm). Image post-processing technologies are used to define the anatomic structure of FSV and detect post-operative complications of the Fontan circulation.

Statistical analysis

Statistical analysis was performed using SPSS software package (SPSS for Windows 11.5). The continuous data were presented as mean value ± standard deviations. The Pearson correlation analysis and Linear-regression analysis were used to assess the correlation between diameters of pulmonary arteries and pressure of pulmonary arteries. And the relationship between McGoon index and pulmonary artery resistant index was also analyzed by statistical methods above. For all of the analysis, a value of P<0.05 was considered to be statistically significant.

Results

Patient characteristics

The patient characteristics of our study are listed in Table 1.

Table 1: Baseline Characteristics of Subjects.
<table>
<thead>
<tr>
<th>Height, m</th>
<th>-</th>
<th>(1.62, 0.12)</th>
<th>1.65</th>
<th>(1.2 to 1.8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weight, kg</td>
<td>-</td>
<td>(53.1, 10.6)</td>
<td>50</td>
<td>(26 to 70)</td>
</tr>
<tr>
<td>Radiation dose, (mSv)</td>
<td>-</td>
<td>(9.52, 1.16)</td>
<td>10.4</td>
<td>(4.2 to 12.7)</td>
</tr>
</tbody>
</table>

**Fontan type**
- Atriopulmonary connection: 2
- Intracardiac lateral tunnel: 8
- Extracardiac conduit: 10
- Y-graft conduit: 8

**Follow-up time dated to operation (years)**
- (10.1, 9.4) | 6 | (0.2 to 29) |

**Postoperative complications**
- Pulmonary arteriovenous malformation: 2
- Aortopulmonary collaterals: 2
- Conduit leak: 0
- Conduit thrombus: 0

**Main malformations**
- Total anomalous pulmonary venous connection: 12
- Hepatic vein abnormal connection: 2
- Anomalous origin of coronary artery: 6

**Note:** Radiation dose is calculated by a cardiac-specific conversion factor (ie., dose-length product multiplied by 0.014 mSv/[mGy.cm]), consisting of arterial and venous phase.

**Comparison between pulmonary artery diameter and pressure**

In our study, the diameters of pulmonary artery by CCTA each was (1.33 ± 0.23, left), (1.38 ± 0.15, right). The pressure of pulmonary artery in systole and diastole through cardiac catheterization were listed as follows: (1) Left pulmonary artery: systole (26.0 ± 5.8) mmHg; Diastole (17.8 ± 5.8) mmHg. (2) Right pulmonary artery: systole (24.4 ± 4.6) mmHg; Diastole (16.9 ± 4.6) mmHg. The diameters of both left and right pulmonary arteries related well to the pressure of pulmonary arteries (left: r=-0.865 systole, r=-0.829 diastole, r=0.867 average; right: r=-0.704 systole, r=-0.758 diastole, r=0.743 average; P=0.000 respectively). The McGoon index also had good correlation with the resistant index of pulmonary artery (r=-0.887, P=0.000). In Linear-regression analysis, we found that when the diameter of pulmonary artery was narrow, the pressure of pulmonary artery would raised (Figure 1). Meanwhile, both the McGoon index and pulmonary artery resistant index can be used to indicate the development of pulmonary artery in a contrary explanation.

**Figure 1:** The results of Linear-regression analysis between diameter of pulmonary artery and pressure of pulmonary artery in systole and diastole.

**Types of the Fontan procedures, anatomies and postoperative complications**

From all of patients in our study, 8 cases of Y-graft Fontan, 2 cases of atriopulmonary connection, 8 cases of lateral tunnel Fontan, and 10 cases of extracardiac Fontan procedures were detected by CCTA data’s (Figure 2). What’s more, 2 cases of diffuse pulmonary arteriovenous malformations and 2 cases of aortopulmonary collaterals were defined as complications of the Fontan procedures (Figure 3). Neither conduit leaks nor conduit thrombus were demonstrated. As to anatomic structures, 6 cases of coronary malformation, 12 cases of total anomalous pulmonary venous connection, and 2 cases of hepatic vein abnormal connection were detected in CCTA images (Figure 4).

**Discussion**

In this retrospective single-center study, we found that CCTA played an important role in the evaluation of the Fontan procedure. The types of Fontan procedure in our study consisted of the atriopulmonary Fontan connection, the intracardiac lateral Fontan procedure, the extracardiac Fontan procedure and the Y-graft Fontan procedure. All of these Fontan conduits were clearly seen in a delayed scan that allowed for systemic venous recirculation. Furthermore, both the calculation of McGoon index to indicate the development of pulmonary artery and the detection of postoperative complication for the Fontan procedure were completed through CCTA data.
With multiple anatomic variations and high mortality, FSV has become one of the greatest clinical management challenges in congenital heart disease. The Fontan procedure was firstly reported as a surgical repair method for tricuspid atresia in the 1960s [3], and it has undergone many methodological modification [4].

Because of the surgical palliation, most FSV patients are expected to reach adulthood [5].

In our study, four types of the Fontan procedures were performed on account of the anatomic diversity and individual variation. While the opacification for the Fontan conduit was difficult, so we use a 2-phase injection protocol to complete the opacification for the inferior vena cava with arterial image acquisition and the Fontan pathway with a delay scan that allowed for systemic venous recirculation. Compared with the method above, the dual injections in both upper and lower extremities will result in swirling of contrast and unopacified hepatic venous flow into Fontan conduit that makes the diagnosis of conduit thrombus difficult [12]. And because of the later opacification of atriopulmonary Fontan pathway than other types, about additional 20 to 30 s was required for the image acquisition.

Even though successfully directing systemic venous blood to pulmonary arteries and pulmonary venous blood to the aorta, the Fontan circulation doses have many disadvantage and complications. Reasons that have yet to be fully understood, but both hepatic fibrosis [13] and protein losing enteropathy [14] related partly to high systemic venous pressure have become vexing clinical problems. Collateral systemic venous to left atrial shunts may form, which will promote progressive cyanosis [15]. And the pulmonary arteriovenous malformation is a serious complication, resulting in lower oxygenation of pulmonary venous flow [16]. Conduit leak and conduit thrombus can also be identified directly by cardiac magnetic resonance or CCTA [16] and the incidence of thrombus in Fontan patients was about 20%-30% and most thrombus were detected in the Fontan conduits or systemic veins [17]. In addition, Rathod et al. [18] reported that myocardial fibrosis was seen in about 28% of Fontan patients and the increasing extent of myocardial fibrosis...
was associated with lower ejection fraction, increased ventricular volume and mass, and a higher frequency of nonsustained ventricular tachycardia.

In our study, both the aortopulmonary collateral and the pulmonary arteriovenous malformations were detected as postoperative complications by CCTA. The aortopulmonary collaterals are vascular connections between systemic and pulmonary circulations, which will bring about numerous physiological implications consisting of ventricle volume overload and increasing incidence of pleural effusions. Furthermore, Grosse-Wortmann et al. [19] and Whitehead et al. [20] reported that aortopulmonary collaterals flow could be quantitated by cardiac magnetic resonance measurements. On CCTA imaging, we detected it through image post-processing technology without any quantitative index. As to pulmonary arteriovenous malformation, it has been treated as a complication with lack of blood reactive material supplied by hepatic vein after the bidirection Glenn procedure [21]. Wei et al. [22] had reported a FSV patient with the postoperative complication of pulmonary arteriovenous malformation in a 7-years follow-up after the bidirection Glenn procedure. However, we demonstrated two cases of diffuse pulmonary arteriovenous malformations in FSV with the Fontan procedure as one-stage operation. Therefore, it maybe the time for us to transform conception and another available explanation is in urgent need. In addition, Neither conduit leak nor conduit thrombus in the Fontan pathway were detected. The conduit leak can be identified by contrast-enhanced examinations that include MRA, CT and angiography. On MRI images, conduit leak will appear as a turbulent jet in systole and not diastole. VEC phase- contrast MRI can be used to detect relatively smaller leak which may not be clearly visualized on anatomic imaging [16]. In our study, no conduit leak was detected by CCTA and the surrounding of all the Fontan conduit were clear without the appearance of contrast agent. These results were supported by cardiac catheterisation.

Except for conduit opacification and complication of the Fontan circulation, the development of pulmonary artery is a major concern for FSV patients. In our study, diameters of pulmonary arteries were measured through CCTA to evaluate the development of pulmonary arteries for FSV patient after the Fontan procedure. Pulmonary artery blood flow encourages the development of pulmonary artery and the diameters will broaden along with its development. So, the measurement of diameter of pulmonary artery by CCTA is a competent method to assess the development of pulmonary artery and it was confirmed by cardiac catheterization data in our study.

In addition, many complex and unusual anatomic structures of FSV were detected by CCTA and it benefited from advanced image acquisition and image post-processing technologies. To a certain extent, this results highlights the clinically applicative value of CCTA modality both in pro- and post-operation. As is well known, anatomic structure and malformations play important role in surgical plan. Some physicians had reported the type of total anomalous pulmonary venous connection was a major agent for surgical complexity [23,24]. If there existed one or more hepatic vein that connected with right atrium directly, the fenestration of Fontan conduit would be not necessary [4]. From this information above, we recognize the importance of preoperative diagnosis for FSV patients. With the aid of contrast agent, radiologists can define dominant ventricle morphology type through density variation between enhanced blood and ventricle trabecular. The comprehensive diagnosis of great arteries, pulmonary vein, systemic vein and other malformations can be easily completed by CCTA with the usage of robust image post-processing technologies. While echocardiography has several limitations for comprehensive diagnosis of FSV disease. Cardiac magnetic resonance imaging modality is incompatible with most heart pacemakers and defibrillators and relatively long imaging time and requirement for breath holding for most sequences also limits its use in certain patients [25,26].

Limitations

There are many limitations in this study. Firstly, our study is a retrospective single centre study with limited samples, which will inevitably lead to bias. Secondly, the criteria for subjects was not strict and the situation of each patient was diversity. So, it is unable to conduct in-depth study. Thirdly, though cardiac ventricle function can be quantified with a retrospective ECG-gated scan with multiphase reconstruction, it was not performed in our study with lack of software for calculation. Finally, CCTA examination dose have certain ionization radiation even though the marked decrease of radiation dose with developed technology and image post-processing technology.

Conclusion

To sum up, the cardiac anatomic structures of functional single ventricle are complex and the imaging interpretation requires sufficient handle with the spectrum of anatomic abnormalities and the types of Fontan procedure. Even though the Fontan palliations can correct single ventricle, many postoperative complication linked to the Fontan circulation should arouse our concern and pathological mechanism of some complications need further exploration. With advanced technologies, CCTA can permit rapid image acquisition with excellent spatial resolution in all patients and gives aid to the diagnosis of anatomical structures and postoperative complications. It is true that CCTA has become a relatively ideal imaging modality for congenital heart disease.

References


