Clinical Surgery Journal

Case Report | Vol 4 Iss S8

Successful Fontan Operation in the Presence of a Single Lung: A Case Report

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Received: January 19, 2021; Accepted: January 22, 2020; Published Date: January 30, 2020

ABSTRACT

INTRODUCTION

The Fontan procedure involves redirecting blood flow from the lower body to the lungs. Patients get this after the Glenn procedure, usually when they are 18 to 36 months old. In this article, we report on our experience of atrioventricular valve repair and Fontan operation performed using cardiopulmonary bypass in the presence of insufficiently developed left pulmonary artery and left lung after bi-directional glen shunt.

KEYWORDS

Fontan procedure; Atrioventricular valve; Hypoplastic lung.

INTRODUCTION

In patients with single ventricle physiology, the Fontan procedure is the last-step surgery and is a palliative option. It requires a good pulmonary vascular bed and, moreover, well-preserved ventricular functions to maintain adequate systemic pressure [1]. According to the literature, providing total systemic venous return is suitable for rare patients. Relative contraindication conditions arise in the presence of a single lung [2-6]. Presence of a unilateral developed pulmonary artery or single lung requires serious planning and careful design. In addition, there has been a serious increase in valve repairs in recent years in patients with single ventricle physiology. Nevertheless, congenital mitral valve diseases in children make the treatment difficult and increase the frequency of perioperative complications.

CASE PRESENTATION

A 4-year-old male patient applied to our clinic with complaints of increased cyanosis and decreased effort capacity for the last 6 months. The bidirectional Glenn shunt operation was applied to the patient who basically had Tricuspid Valve Atresia at the age of 1. Diagnostic digital subtraction angiography (DSA) was performed by the Pediatric Cardiology unit after the patient described that her complaints had increased. In DSA, right pulmonary artery calibration was observed as appropriate for the operation (Figure 1). It was seen that the left pulmonary artery did not develop and the left lung tissue was nonfunctional (Figure 2). Echocardiography revealed moderate to severe (grade 3) atrioventricular valve regurgitation.

Citation: İbrahim Demir, Successful Fontan Operation in the Presence of a Single Lung: A Case Report. Clin Surg J 4(S8): 4-7.

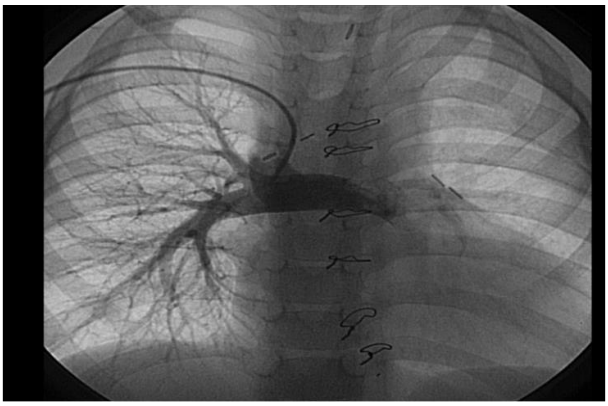

Figure 1: Well-developed and appropriate right pulmonary artery for surgery.

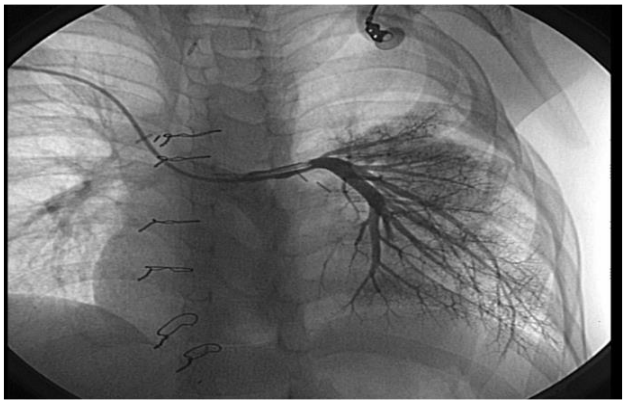

Figure 2: Hypoplastic right pulmonary artery and lung.

Operation decision was made for the patient. The atrioventricular valve was evaluated in the patient who was operated under cardiopulmonary bypass under appropriate conditions. Anterior leaflets prolapse, deepening between P2 / P3 and annular dilatation were detected. The anterior leaflet was repaired by creating 3 neocorda. P2 / P3 scallop was capped with 5/0 prolene suture. There was no regurgitation after water test (Figure 1). The main pulmonary artery pressure was found to be 17-18 mmHg in the perioperative measurements. Although antegrade flow was closed during the measurement, there was no change in pressure. Since the left pulmonary artery was hypoplastic, it was thought that a significant pressure drop could not be obtained with any procedure.

After the cross clamp was removed, the Fontan procedure was performed using an 18 mm graft. Fenestration was created between the right atrium and the graft. The operation was completed with appropriate hemodynamics.

The patient was extubated at the 5th hour without any problem. The patient was discharged on the 12th day. Oxygen saturation was monitored as 90% in the follow-up, and no regurgitation was observed in the control echocardiography.

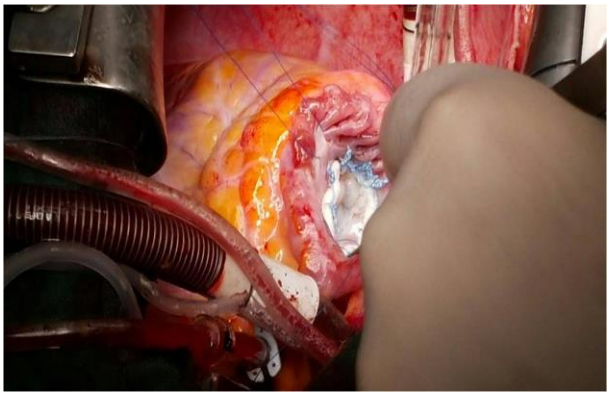

Figure 3: Atrioventricular valve after repaired.

DISCUSSION

In patients with single ventricular physiology, Fontan operation and its modifications are considered the last step of modern therapy. Passive venous flow is directed to the pulmonary circulation instead of antegrade pulmonary pulsatile flow. Therefore, proximal pulmonary artery anatomy and the state of the distal vascular structure are among the critical determinants of the Fontan procedure [3,4]. To minimize the negative effects of previous operations, damaged areas that may create resistance to passive systemic venous return should be carefully reconstructed [5,7,8].

A very rare group of patients has a functional single lung. The commonly known cause of growth retardation, stenosis or occlusion in the pulmonary artery is the long-term complication of shunt operations with short graft length [8]. If lung volume loses or decreases in some way, physiologically, the remaining lung can only expand its volume to a certain extent. However, unilateral increase in lung capacity never compensates for double lung volume

[9]. In patients with a single functional lung undergoing the Fontan operation; peak respiratory capacity, vital capacity and forced vital capacity are usually preserved in the early period and this is encouraging for surgeons. The first successful operation was published by Sade and Gillette [2] in 1989. Zachary and Jacobs (7 and 12 patients, respectively) presented their experience in a relatively large series of patients [6]. Zachary's study showed a range of oxygen saturation (87%-91%) in single-lung patients undergoing the Fontan procedure compared to functional double-lung cases [5]. Al-Khaldi successfully presented this operation in an adult patient [4]. All these results agreed that the condition of the pulmonary vascular structure is decisive for the success of the operation. In terms of Fontan operation, deterioration in the pulmonary artery structure and the presence of pulmonary vascular resistance are considered risk factors [2,6,10].

Two main factors, pulmonary artery pressure and single ventricular ejection fraction, are the determinants of Fontan operation [1]. Pulmonary vascular resistance is of high importance in the presence of nonfunctioning lung. However, operation should be considered in the presence of hypoplastic lung and pulmonary artery.

Another factor affecting morbidity and mortality in patients with single ventricle physiology is structural defects in the atrioventricular valve. Valve repair or replacement is sometimes required in this patient group. The main cause of valvular insufficiency in these patients may be the asymmetric geometry of the intraventricular muscle mass or dilatation secondary to subvalvular stenosis. Although the benefit of valve repair in patients scheduled for Fontan is still unclear, the lower risk of thromboembolism or heart block that may develop after valve replacement is one of the most important reasons that lead us to choose repair instead of replacement. For these reasons, we preferred to apply the Fontan procedure and simultaneously repair the valve because our patient did not have sufficient oxygen saturation and his exercise capacity was low.

CONCLUSION

Consequently, the state of the pulmonary circulation is crucial to the success of the Fontan procedure. Very rarely, hypoplastic pulmonary artery is considered a contraindication. When the Fontan procedure is planned in patients with single ventricle physiology, if there is amitral valve insufficiency, it is useful to plan the valve repair simultaneously.

FUNDING

None Declared.

CONFLICTS OF INTEREST

None Declared.

ETHICAL APPROVAL

Informed consent was obtained from the relative of the patient before the procedure after explaining the interventions, risks and benefits as a policy of the health system in the country.

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