



Mid-term outcome of Egyptian children post univentricular repair (experience of single center)



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KEYWORDS

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Abstract *Background:* More patients survive the operations for univentricular repair and the duration of follow up increases, the physicians are becoming increasingly aware of the late failure and the late complications of the surgery.

Objective: To evaluate the overall outcome of studied patients and to assess their functional status, and the occurrence of complications.

Patients and methods: This is a retrospective study; patients who underwent univentricular repair operations either Glenn or Fontan were included in the study. The following data were collected: age, sex, type of the operation, and presence of early post operative complications. The outcome was assessed by evaluating clinical status and growth of the patients during the follow-up visits. Medical and surgical complications were retrieved.

Results: Thirty-nine patients who underwent univentricular repair operations either Glenn or Fontan were included from the pediatric post operative cardiac outpatient clinic. Their age ranged from 2–18 years. 38 patients (97.4%) underwent Glenn operation followed by Fontan in 11 (27%) patients, 1 (2.6%) patient underwent Fontan without any previous operation and 4 (10.2%) patients underwent multistage palliation (prior operation to Glenn). 8 (20.5%) patients had early complications in the form of arrhythmia, bleeding, convulsions, and infection. The general functional status of patients is good; growth curves are within the normal range in 30 (76.9%), and NYHA class I in 24 (61.5%) patients. No late complications during follow-up and no mortality were documented.

Conclusion: Adequate post operative follow up is essential to improve midterm outcome of the patients.

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Abbreviations: NYHA, New York Heart Association; ACE, angiotensin converting enzyme; INR, international normalized ratio.

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Introduction

The surgical management of single ventricle physiology has been evolving for decades since the first Glenn operation¹ in which the superior vena cava is connected to the right pulmonary artery to carry the blood to the lung for oxygenation. Glenn operation is habitually part of the staging that results in a very victorious palliation total arteriopulmonary shunt by Fontan and Baude,² Glenn fell out of favor after Fontan

was introduced, subsequent to abandon; Glenn shunt was looked at again when the early versions of Fontan did not convey the expected results, the bidirectional Glenn shunt is now preferred in very small babies < 2 years,¹ and the Fontan operation has become the most common procedure performed for congenital heart disease after the age of 2 years.³ Advances in operative technique and the adequate post operative follow up have been accompanied by improvement in early survival of the patients from 75% to 83% in 1970s to over 90% in the current era.⁴ Operative mortality has decreased despite the application of the operations in patients previously considered to be high risk either due to complex forms of single ventricle or inadequate hemodynamics.³ As more patients survive the operations and the duration of follow up increases, the physicians are becoming increasingly aware of the late failure and the late complications of the operation. The aim of this study is to evaluate the overall outcome of our patients with single ventricle physiology who underwent univentricular repair operations (Glenn or Fontan) as regards their functional status, quality of life, and the occurrence of complications.

Patients and methods

Patients

We reviewed the medical records of the outpatients from the Pediatric Postoperative Cardiac Outpatient Clinic (PPCOC) in Cairo University Children's Hospital from the start of the clinic in June 2007 till December 2012, we included 39 patients with single ventricle physiology who underwent univentricular repair operations either Glenn or Fontan taking into consideration any palliative operation done before, such as shunt or pulmonary artery banding. A preoperative detailed echocardiographic evaluation was done for all patients by machine vivid 7 probe 3S, 5S, in addition to performing cardiac catheterization in patients prepared for Fontan operation to assess the existence of adequate anatomic and physiologic parameters permitting Fontan completion. These parameters included low pulmonary vascular resistance, satisfactory ventricular function, good or correctable cardiac valve function, and unobstructed or reconstructible adequate sized pulmonary arteries. The final anatomic diagnosis was based on results of two-dimensional echocardiography, cardioangiography, and operative findings. During the follow up in the clinic all patients underwent a complete evaluation in which operative, perioperative variables, and follow-up clinical data are recorded. Echocardiographic assessment is done to all patients on regular follow up periods varying from 6 months to 1 year according to the condition of the patient.

Methods

We manually reviewed the pre-completed files to collect the following data: age, sex, type of the congenital heart disease, type of the operation, age at the first operation, duration of the hospital stay post operatively, duration of follow up in the Pediatric Postoperative Cardiac Outpatient Clinic (PPCOC), presence of early complications in post operative cardiac intensive care unit, we also included the clinical status of the patients as regards the NYHA grade in the last visit, adequate growth by measuring the weight and the height on

the follow up visits, occurrence of complications during period of follow up in the clinic, the treatment prescribed to the patients either diuretics, ACE inhibitors, digoxin, pulmonary vasodilators (sildenafil), anticoagulants as marivan or antiplatelets as aspocid 5 mg/kg.

Results

The total number of patients in PPCOC was 1002 patients. 39 patients who underwent univentricular repair operations either Glenn or Fontan were included in the study and represented 3.9%. Their age ranged from 2–18 years and their age at the first operation ranged from 1–12 years. The majority 26 (66.7%) were males. Diagnosis of the studied patients is shown in [Table 1](#).

22 (56.4%) patients had single left ventricle morphology, abnormal related vessels in 16 (41%) patients, and pulmonary stenosis in 21(53.8%) patients.

38 (97.4%) patients had Glenn operation followed by Fontan in 11 (28.2%) patients, 1 (2.6%) patient had done Fontan operation solely, he was 11 years old male, with the diagnosis of double outlet right ventricle, D-transposition of great vessels, pulmonary stenosis and 4 (10.2%) patients had multistage palliation (prior operation to Glenn) in the form of pulmonary artery banding and shunt. The duration of post operative hospital stay ranged from 4–120 days.

Eight (20.5%) patients had early complications in the form of arrhythmia, bleeding, convulsions, and infection. Incidence of complications in studied patients is represented in [Table 2](#).

The follow-up period of the studied patients extended from 6 months to 6 years. The general functional status of patients is good as evidenced by: growth curves within the normal range in 30 (76.9%) patients, 8 (20.5%) patients had no heart failure, and 24 (61.5%) patients are in NYHA class I in the last visit. No late complications during follow-up and no mortality were documented. Anticoagulants in the form of oral warfarin was given to 21 (53.8%) patients followed by antiplatelets therapy in the form of aspocid 5 mg/kg while 18 (46.1%) patients were

Table 1 Diagnosis of studied patients.

Diagnosis	Number
DORV (double outlet right ventricle)	11 (28.3%)
DILV (double inlet left ventricle)	10 (25.6%)
Tricuspid atresia	9 (23.2%)
Transposition of great vessels	3 (7.7%)
Tetralogy of fallout	2 (5.1%)
Single ventricle	2 (5.1%)
Pulmonary atresia	1 (2.5%)
Complete atrioventricular canal	1 (2.5%)

Table 2 Incidence of complications in studied patients.

Type of complication	Incidence
Convulsions	3(7.7%)
Infection	2(5.1%)
Deep venous thrombosis	1(2.6%)
Intracranial hemorrhage	1(2.6%)
Nodal rhythm	1(2.6%)

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