Iranian Heart Journal; 2020; 21 (4)

Original Article

Comparisons of Arterial and Venous Palliative Shunts in Adult Patients With Cyanotic Congenital Heart Diseases

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ABSTRACT

Background: Generally, 2 types of surgical operations are available for the treatment of cyanotic heart diseases: corrective and palliative. The goal of palliative surgery is to increase the pulmonary blood flow via systemic-to-pulmonary arterial shunting.

Methods: In this case-series study, we evaluated patients older than 15 years old with documented cyanotic heart diseases who underwent palliative shunting in Rajaie Cardiovascular Medical and Research Center between 2001 and 2015.

Results: Forty-seven patients at a mean age of 24.74 ± 5.67 years were enrolled in this study. Twenty-two (46.8%) patients were male. Nearly half of the study population (23 patients) had central shunting, 23 patients Glenn shunting, and 1 patient Blalock–Taussig shunting. Following surgery, there was a significant rise in the mean partial pressure of O_2 and O_2 saturation, a significant drop in the hemoglobin concentration, and a significant increase in the platelet count. Five (10.6%) patients expired during the study period. The mortality rate of Blalock–Taussig shunting and Glenn shunting was 12.5% and 8.69%, respectively.

Conclusions: In this case-series study, all the patients showed dramatic improvements in the New York Heart Association functional class, O_2 saturation, the partial pressure of O_2, the partial pressure of carbon dioxide, hemoglobin levels, and ferritin levels in the first postoperative year; however, these improvements were greater in arterial shunts. The following 10 years saw a decline in these improvements; nonetheless, the clinical status and lab data were good by comparison with the baseline.

In adults with congenital cyanotic heart diseases, palliative shunting is a good option and should be considered in patients who are inoperable due to structural problems or high risk of anesthesia or surgery. ***(Iranian Heart Journal 2020; 21(4): 49-55)***

KEYWORDS: Cyanotic heart disease, Palliative surgery, Glenn shunt, BT shunt, Central shunt

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Received: September 29, 2019 **Accepted:** April 25, 2020

Cyanotic congenital heart diseases are defined as any malformation of the heart at birth that manifests early with cyanosis. The tetralogy of Fallot, tricuspid atresia, the transposition of the great vessels, and various single-ventricle anomalies (eg, hypoplastic left-heart syndrome) are among them. Inadequate oxygenation of organs results in cyanosis (blue baby syndrome) and is due to an obstruction in the right-heart circulation or the deviation of the blood flow from the pulmonary circulation. [1-4]

Generally, 2 types of surgical operations are available for the treatment of cyanotic heart diseases: corrective and palliative.

The goal of palliative surgery is to augment the pulmonary blood flow via systemic-to-pulmonary arterial shunting or to promote the mixing of the blood through left-to-right shunting. For some patients with other cases, palliative techniques serve to "buy time" until the patient attains a more desirable age and body size for subsequent definitive repairs. [6]

The most common shunts for the palliation of patients with cyanotic heart diseases are Blalock–Taussig (BT) (arterial) shunts, central shunts, and Glenn (central/ venous) shunts.

Other famous shunts include Waterston shunts (direct side-to-side connections between the ascending aorta and the pulmonary artery) and Potts shunts (direct side-to-side connections between the descending aorta and the pulmonary artery), first introduced in 1946 and abolished in 1967 due to serious complications. [11, 12]

METHODS

Study Design

The present case-series study enrolled adult patients with documented congenital cyanotic heart diseases who underwent palliative surgery at a minimum age of 18 years for the first time or patients who had an occluded shunt or an inadequate shunt flow in Rajaie Cardiovascular Medical and Research Center between 2001 and 2015. These patients were not ideal cases for the Fontan procedure.

Data Gathering

The patients' information, including age, sex, lab data, echocardiographic data, and surgical data, was retrospectively extracted from their medical documents. A checklist was devised by the researcher to record the information for each patient.

Follow-up

The rate of in-hospital mortality was directly extracted from medical files in the hospital. The patients' cardiac status was evaluated 3 times: 1 month after shunt-creation surgery, 1 year after the operation, and more than 2 years after the operation.

Statistical Analysis

All the data were analyzed using the SPSS software, version 20. The survival analysis and the Kaplan–Meier tests were used to compare survival between the patients. For further analyses, the *t*-test, the χ^2 test, the one-way ANOVA, and correlation tests were used. A *P*-value of less than 0.05 was defined as significant.

Ethical Approval

The purpose of the study was comprehensively explained to the patients or their legal custodians, who signed a written informed consent form on the understanding that their information would be treated highly confidential. The study protocol was confirmed by the Ethics Committee of Rajaie Cardiovascular Medical and Research Center.

RESULTS

Forty-seven patients, including 22 (46.8%) male patients, at a mean age of 24.74 ± 5.67 years were enrolled in this study. Almost half of the study population (23 patients) had central shunting, 23 patients Glenn shunting, and 1 patient BT shunting.

Thirteen patients had their first shunting. In the other patients, the previous shunt was occluded in 30 cases and patent in 4 cases.

The patients were divided into 4 groups based on their preoperative left ventricular ejection fraction (LVEF): 7 (14.9%) patients had a normal LV function (LVEF > 55%), 26 (55.3%) patients had mild systolic dysfunction (45% < LVEF < 55%), 13 (27.7%) patients had moderate systolic dysfunction (30% < LVEF < 45%), and 1 (2.1%) patient had severe systolic dysfunction (LVEF < 30%).

Before shunt-creation surgery, 2 (4.3%), 4 (8.5%), 38 (80.9%), and 3 (6.4%) patients were in the New York Heart Association (NYHA) functional classes I, II, III, and IV, respectively.

Before the operation, systemic ventricular regurgitation (SVR) was mild in 17 patients, mild-to-moderate in 3, moderate in 15, moderate-to-severe in 1, and severe in 2.

The mean hemoglobin (Hb) level of the patients was 18.55 ± 3.05 before surgery, 18.08 ± 1.43 at 1 month after surgery, 18.58 ± 1.42 at 1 year after surgery, and 18.60 ± 1.38 in the longer postoperative follow-up.

The mean partial pressure of oxygen (PaO_2) increased significantly after the operation (P = 0.01). Figure 1 compares PaO_2 before surgery and in the follow-ups between the 2 shunt type groups.

O_2 saturation also increased significantly after surgery (P = 0.01). Figure 2 compares O_2 saturation before the operation and in the follow-ups between the 2 shunt type groups.

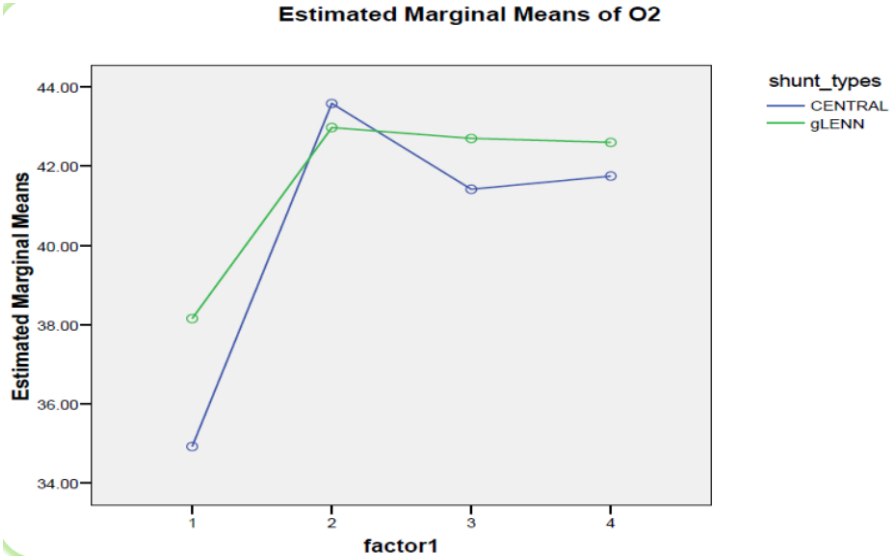

Figure 1: Comparisons are illustrated between the 2 shunt type groups in terms of changes in the partial pressure of oxygen (PaO_2) after surgery.

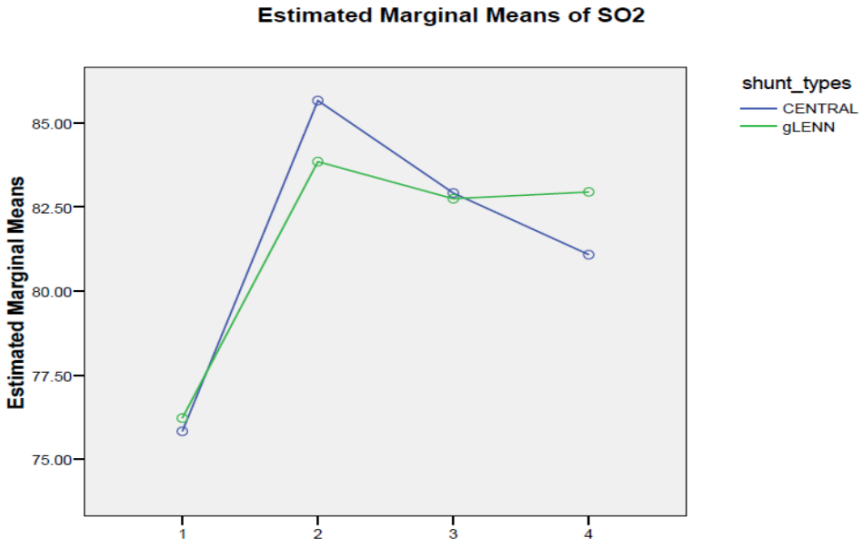

Figure 2: Comparisons are illustrated between the 2 shunt type groups in terms of changes in oxygen saturation (SO_2) after surgery.

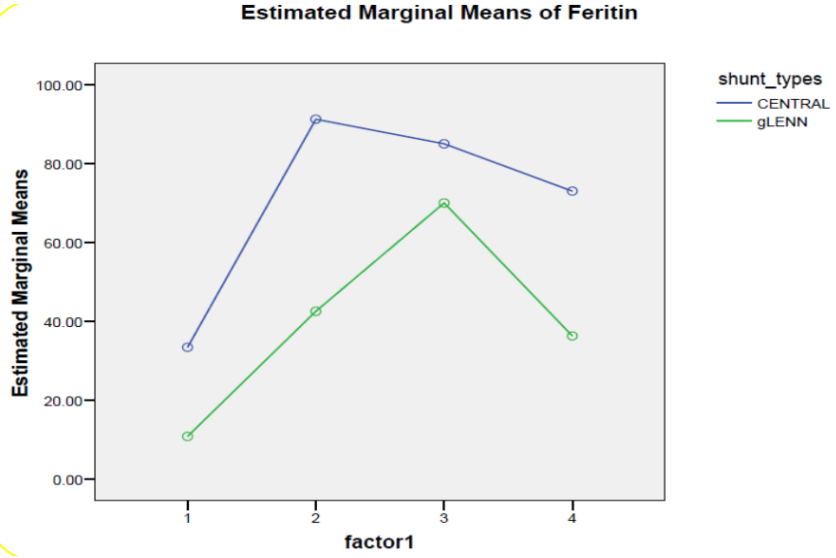

Figure 3: Comparisons are illustrated between the 2 shunt type groups in terms of changes in the ferritin level after surgery.

The mean partial pressure of carbon dioxide ($PaCO_2$) decreased significantly after surgery ($P = 0.05$).

The Hb concentration decreased significantly after the operation ($P = 0.01$), but this decline was not observed in the long-term follow-up.

The serum ferritin level rose significantly after surgery ($P = 0.02$). Figure 3 compares the ferritin levels before surgery and in the follow-ups between the 2 shunt type groups.

The serum iron level increased significantly in the follow-up tests some months after the operation ($P = 0.01$).

The very early success rate of shunt patency was 97%: 100% for Glenn shunts and 95% for central shunts. The patency rate after 1 month was 100% for Glenn shunts and 86% for central shunts. (Three patients in the central shunt group had occluded shunts after 1 month.) In the long follow-up, the patency rate was 100% for Glenn shunts as opposed to 75% for central shunts.

Five (10.6%) patients expired during the study period: 1 immediately after the operation due to refractory heart failure, 1 during the intensive unit care (ICU) stay due to shunt failure, 1 at 3 years after surgery due to valve endocarditis and sepsis, 1 at 2 years after the operation due to heart failure, and 1 while at home (sudden cardiac death, probably). Among them, 3 had central shunts and 2 had Glenn shunts.

The mortality rate of central shunts was 12.5% (3 out of 24 cases), while the mortality rate of Glenn shunts was 8.69% (2 out of 23 cases).

DISCUSSION

Our study is the first report on patients with cyanotic heart diseases who for the first time underwent palliative shunting in adulthood. In a multicenter study by Petrucci et al [13] on 1273 neonates with cyanotic congenital heart diseases undergoing BT shunting between 2002 and 2009, the mortality rate was 7.3% and the morbidity rate was 13.1%. Additionally, 33% of the deaths occurred within 24 hours after surgery, and 75% occurred within 30 days.

Another study evaluated all 2016 BT shunts created over a 62-year period since the introduction of BT shunting. According to the results, the tetralogy of Fallot was the most common indication (72%), and the total mortality rate was 14%. Further, the mean annual number of BT shunt-creation surgical operations and annual mortality exhibited a decrease over the 6 decades. [14]

Le Logan et al [15] evaluated 80 adult patients at a mean age of 36 years who had previous BT shunts and showed that BT shunting could decrease the blood flow to the right upper extremity and result in decreased hand force.

Berdjis et al [16] evaluated 200 adults at a mean age of 38 years who underwent surgery for congenital heart diseases. Fifteen patients with cyanotic heart diseases underwent 11 corrective and 4 shunt procedures. Three died due to low cardiac output or dysrhythmias (20%). Clinical status markedly improved in the survivors.

Most of these shunts are created in infancy[17]; however, adults who are not eligible cases for the Fontan operation also benefit from these surgical operations. Such adults include those who show their first symptoms in adulthood, those who are diagnosed with congenital heart diseases incidentally, or those who experience shunt failure.

It is clear that the success rate in every type of surgery depends on the technique and experience of the surgeon; thus, an evaluation of the local success rate appears to be necessary. Since no study has examined the prognosis of adult patients who undergo palliative surgeries for the first time in their adulthood, we conducted the current investigation to evaluate adult patients with congenital cyanotic heart diseases and to compare the short- and long-term results of arterial and venous shunting.

In this case series, we evaluated adolescents and adults with congenital cyanotic heart diseases who underwent palliative surgery in their adulthood. Except for 4 patients, the other cases had previously occluded palliative shunts or had their first shunt due to the late commencement of symptoms. All the patients showed dramatic improvements in the NYHA functional class, O_2 saturation, PaO_2, $PaCO_2$, Hb, and ferritin in the first year after surgery. These improvements were more remarkable in arterial shunts. In the following years, these improvements declined; nevertheless, the clinical status and lab data were acceptable in comparison with the baseline (before surgery). Our study results underscored the benefits of both central and Glenn shunts immediately after surgery, but better preservation of these improvements in the central shunt group in terms of O_2 saturation, Hb, and NYHA functional class. Our patients had nonsignificant improvements in the systemic ventricular EF and SVR in the first postoperative year, which aggregated in long-term follow-up. All the Glenn shunts, and 75% of the central shunts were patent after almost 10 years.

A significant increase in PaO_2 and O_2 saturation, together with a decreased $PaCO_2$ level, indicates improvements in the pulmonary blood flow and adequate oxygenation, resulting in decreased need for Hb to supply O_2 to the tissues and consequently decreased need for iron. Since we are not certain about the use of iron supplements by all our patients, making a judgment as regards ferritin and serum iron levels is difficult. Still, the increase in the platelet count in our study indicates improved oxygenation of the end organs.

In this study, 5 (10.6%) patients expired: 1 immediately after surgery due to refractory heart failure, 1 during ICU stay due to shunt failure, 1 at 3 years after the operation due to valve endocarditis and sepsis, 1 at 2 years after surgery due to heart failure, and 1 while at home (sudden cardiac death, probably). Among them, 3 had central shunts and 2 had Glenn shunts.

In the study by Petrucci et al, [13] the mortality rate from BT shunts was 7.3%. One-third of the deaths occurred within 24 hours after surgery, and 75% occurred within 30 days.

Another study evaluating all 2000 BT shunts created since its introduction reported a 14% mortality rate. [14]

In the study by Berdjis et al [16] on adults with congenital heart diseases who underwent corrective or palliative surgeries, 3 out of 15

(20%) patients died. Clinical status, however, was improved significantly in the survivors.

Apropos of systemic ventricular function after shunt insertion, there are 2 probabilities. Firstly, systemic shunting caused volume overload as patent ductus arteriosus on the systemic ventricle, leading to a negative effect on the systemic ventricular EF. Secondly, on the other hand, reducing cyanosis via shunt surgery had positive effects on systemic ventricular function. In our study, we found an increase in the LVEF after surgery, especially during the first year. Nonetheless, after a few years, this improvement decreased.

We also evaluated systemic atrioventricular valve regurgitation before and after shunt surgery. As a theory involving systemic ventricular function concerning the change in the grading of regurgitation, there are 2 possibilities: improvements in terms of the increased EF of the systemic ventricle and exacerbation in consequence of the volume overload of the ventricle. The changes in regurgitation grading in this study were negative. Our results showed an increase in the severity of regurgitation, which was more significant in the first year after surgery in the arterial shunt group. There was, however, no significant change in regurgitation severity after Glenn shunting.

One of the most significant factors that unfortunately we failed to consider in our study is the size of the pulmonary artery branches. In our opinion, it is a powerful marker in the patency and efficacy of the shunt. Future studies should evaluate the relationship between the size of the pulmonary artery branches and improvements in O_2 saturation and patients' symptoms.

CONCLUSIONS

In light of the results of the present study, it can be concluded that in adults with congenital cyanotic heart diseases, palliative shunting is a good option and should be considered in patients who are inoperable due to structural problems or high risk of anesthesia or surgery. These shunts are lifesaving with an acceptable quality of life even in adulthood.

Funding: This study was funded by Rajaie Cardiovascular Medical and Research Center.

Conflict of Interest: All the authors declare no competing financial interests.

Ethical Approval: All the procedures performed in the present study, which involved human participants, were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed Consent: Informed consent was obtained from all the participants or their legal custodians.

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